

Review

Pattern of Refractive Correction and Timing of Stage II IOL Implantation after Congenital Cataract Extraction

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Abstract

Congenital cataract occurs during infancy when the axial length and corneal and visual function are in the sensitive stages of rapid development. Inappropriate surgical intervention not only fails to restore visual function, but also causes irreversible serious influences upon eyeball development in children diagnosed with congenital cataract. At present, the uncertainty of selection of intraocular lens (IOL) degrees during the eyeball development period is averted by using a main treatment of congenital cataract that includes two-stage surgery: stage I cataract extraction and stage II IOL implantation. However, the accurate selection of a refractive correction method and the timing of IOL implantation during stage II surgery for aphakic eyes remains controversial following stage I cataract extraction. This review retrospectively summarizes the current progress and existing problems indicated by related recent studies focusing on refractive correction pattern and IOL implantation timing. (*Eye Science* 2014; 29:237–242)

Keywords: congenital cataract; stage II IOL implantation; surgical timing

Introduction

Congenital cataract is a partial or complete clouding of the lens of the eye that is observed with one year after birth. The pathogenesis of congenital

cataract is complex; it occurs both unilaterally and bilaterally, with approximately 1/3 of cases correlated with hereditary factors, 1/3 associated with environmental factors, and the other 1/3 resulting from unknown causes. The severity of congenital cataract is varying and complex. Surgical timing should be determined based upon each individual situation rather than as early as possible. Cases presenting with dense congenital cataract during infancy, especially at 6 weeks to 3 months after birth (the critical period for visual acuity development¹) should be treated with stage I simplex cataract extraction as early as possible after diagnosis. Postoperatively, a suitable refractive correction method should be adopted to prevent the incidence of low vision and blindness.

Gradual improvement of surgical techniques and instruments has led to wide performance of stage I simplex cataract extraction in babies and infants with congenital cataract². However, the changes in refraction caused by rapid development of the eyeball create a series of difficulties to refractive correction treatment for aphakic eyes. The questions then become how to select the optimal refractive correction method after stage I simplex cataract extraction and how to choose the perfect timing of stage II intraocular lens (IOL) implantation. We will address these questions in this review.

Ocular characteristics of healthy and congenital cataract children

Healthy children are characterized by having rapid development of the refraction system and gradual maturation of visual function.

The average axial length of newborns is 16.8 mm and it increases at the greatest rate before the age of two years (an average increase of 4 mm per year). For a child aged 7 years, the axial length is approx-

DOI: 10.3969/j.issn.1000-4432.2014.04.010

Funding: The Pearl River Science and Technology New Star (Grant No. 2014J2200060) Project of Guangzhou City, the Guangdong Provincial Natural Science Foundation for Distinguished Young Scholars (Grant No. 14050000151) and the Cultivation Projects for Young Teaching Staff of Sun Yat-sen University (12ykpy61) from the Fundamental Research Funds for the Central Universities.

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imately 23 mm and this increases to 23.6 mm by adulthood.

The corneal diameter of newborns is 9-10 mm and it increases at the greatest rate before the age of 1 year, so that children aged 3-4 years have a similar corneal diameter to adults (11-12 mm).

Both corneal curvature and axial length of infants change during aging. The overall diopter of newborns ranges from +2.00 to +3.00 D as presbyopia, which is gradually altered to emmetropic eyes by age 6-7. This inclination of the diopter towards myopia is known as the myopia shift^[3].

The visual acuity of newborns is approximately 0.01. The fixation reflex is formed at 2-3 months after birth. A 3-month-old infant has an established bilateral visual function. An infant possesses stereoscopic vision at approximately 6 months after birth. The period of 6 weeks to 3 months after birth is the critical timing for visual acuity development¹.

However, children with congenital cataract present a pattern of eye growth that differs significantly from that of healthy infants^{4,5}. During early development of the eyeball (the critical period of visual acuity development) in children with congenital cataract, form deprivation alters the normal development of eyeball and leads to form deprivation myopia. In addition, animal and clinical research has confirmed a positive correlation between the growth of axial length and form deprivation^{6,7}. The development of axial length in children suffering from congenital cataract is influenced by multiple factors following cataract extraction.

Chinese scholars found that the axial length was significantly increased postoperatively in children without IOL implantation, especially before the age of two years. Age at operation, preoperative axial length, and postoperative best corrected visual acuity were also significantly correlated with the growth rate of axial length⁸. Younger children presented with higher corneal curvature and shorter axial length. No statistical significance was observed between male and female counterparts ($P>0.05$). The axial length was significantly longer in unilaterally affected children than in bilaterally affected counterparts. In addition, the corneal curvature was equally significantly higher in the unilaterally affected eye than in the fel-

low eye and the axial length was significantly longer than in the fellow eye (all $P<0.01$)⁸. Phakic eyes had a tendency toward myopia. The subjects undergoing IOL implantation at a younger age had a more significant inclination toward myopia⁹. Consequently, the timing of IOL implantation has a strong effect upon the development of axial length in children with congenital cataract.

Refractive correction method after congenital cataract extraction

Affected children present with special changes in refractive development, which creates issues in the selection of a proper method for refractive correction in these children in clinical settings. At present, children with congenital cataract are treated with refractive correction methods for aphakic eyes that mainly include common frame glasses, corneal contact lenses, press-on optics, and IOL. The advantages and disadvantages of these four approaches of refractive correction are illustrated in Table 1.

Making the best choice among these four approaches of refractive correction remains an issue in clinical settings. In China, affected children aged < two years with bilateral aphakic eyes mainly receive frame glasses and press-on optics for refractive correction. Those aged < two years with unilateral aphakic eyes receive corneal contact lenses for refractive correction. IOL implantation is recommended for the children aged > two years with unilaterally/bilaterally affected eyes^{10,13}. The Infant Aphakia Treatment Study Group from the U.S. demonstrated that children with unilateral congenital cataract did not significantly differ in terms of postoperative visual acuity after cataract extraction before the age of 7 months when corneal contact lens and IOL implantation groups were compared. Instead, the children in the IOL group presented with more postoperative complications and higher re-operation rates when compared with their counterparts in the corneal contact lens group. Consequently, this study group recommended that IOL implantation should not be performed before cataract extraction for these children; these patients should wear corneal contact lenses to correct refractive error¹².

Most scholars from the U.S. also have suggested that IOL implantation following cataract extraction

Table 1 Advantages and disadvantages of common methods of refractive correction

	Advantages	Disadvantages
Common frame glasses	Economical, easy adjustment and replacement	In only applies to bilaterally affected children, thick and heavy lenses, poor compliance, likely to cause image distortion and visual field constriction, poor efficacy of refractive correction.
Corneal contact lenses ^{11,12}	Light-weight, easy adjustment and replacement, good imaging quality, and high efficacy of refractive correction	Inconvenient to wear and take off and difficult to restore and process
Press-on optics ¹³	Light-weight, aesthetic and comfortable, good compliance, easy adjustment and replacement, convenient to restore and process, low incidence of image distortion	Easy aging and contamination, slightly poor light penetration, slow improvement in visual acuity
IOL implantation ¹⁰	Conforms to eye physiological characteristics and optical effects, high efficacy of visual acuity correction	Difficult replacement, high price, multiple postoperative complications, probably requires repeat surgeries

should be conducted at an age \geq two years for children with congenital cataract^{14,15}. In developing countries, environmental, cultural, and economic discrepancies have led to consideration of IOL implantation as a unique approach for visual acuity correction early after cataract extraction. IOL implantation could improve the visual acuity of the affected children¹⁶⁻¹⁸.

Timing of stage II IOL implantation

The timing of stage II IOL implantation is important to avoid postoperative complications and to enhance the visual function of the affected children as much as possible. Chinese scholars have suggested that children with bilateral congenital cataract but without eye or systemic diseases should undergo IOL implantation at the age of 2-3 years¹⁹⁻²¹. Children aged < 2 years are characterized with rapid development of the eyeball, unstable development of corneal curvature and axial length, and immature development of the ciliary muscle and lens capsule. Therefore, accurate calculation of the diopter of IOL is difficult. Children aged < 2 years are also likely to experience a greater number of postoperative complications after IOL implantation¹⁹⁻²¹. Hence, most scholars have recommended that children aged < 2 years and with aphakic eyes wear corneal contact lenses or frame glasses to correct refractive error.

As mentioned above, both corneal contact lenses and frame glasses have disadvantages, such as poor compliance, little adaptability, and low effectiveness

of visual correction, etc. The delay in IOL implantation probably misses the optimal period of visual development and negatively affects the visual function. Recently, the accuracy of selection of IOL refraction degree has been improved, along with the introduction of new formulas for diopter and myopia shift estimation. For children aged 2-3 years, the clinical efficacy of stage II IOL implantation has been significantly enhanced and the re-operation rate for refractive correction has been greatly reduced. However, irreversible inhibitory amblyopia could be induced in children's aphakic eyes after unilateral cataract extraction. Most researchers therefore recommend performing stage I IOL implantation for children with unilateral congenital cataract.

Stage I IOL implantation yields multiple postoperative complications, but most complications can be alleviated by laser treatment. This operation strategy could prevent the need for repeated stage II IOL implantation, reduce economic burden, create conditions for recovery of binocular visual function, and improve the survival and quality of life of the affected children^{19,21}. Consequently, ophthalmologists in China now recommend that cataract extraction and stage I IOL implantation should be performed simultaneously at an appropriate age for children with unilateral congenital cataract, which agrees with some international viewpoints. In 1993, 12.9% of physicians from the U.S. chose IOL implantation after congenital cataract extraction for children aged < 2 years²² and

that percentage had increased up to 81.9% by 2001²³. However, clinical experience and observation during the past decade have demonstrated that undesirable postoperative visual effects are observed and increasingly more postoperative complications occur in certain children. Hence, the surgical timing of IOL implantation is now a controversial issue.

In 2010, the U.S. Infant Aphakia Treatment Study (IATS) conducted a multi-center random clinical trial consisting of 114 children from 12 centers undergoing surgery within 7 months after birth. The authors found that the grating visual acuity did not significantly differ between children receiving IOL implantation at age of 1 and their counterparts wearing corneal contact lenses²⁴. The 5-year follow up also revealed that the average visual acuity in children aged 4-5 did not significantly differ after cataract extraction regardless of whether they underwent IOL implantation. In contrast, the incidence of postoperative complications and the re-operation rates were significantly higher compared in the IOL implantation group than in the corneal contact lens group. In addition, IOL implantation caused significantly more economic burden and higher risk than did the use of corneal contact lenses.

However, researchers from the Europe and America have recently found that early IOL implantation could lead to better binocular vision and stereoscopic vision, etc. Some scholars have even suggested that early IOL implantation probably contributes to the growth of the eyeball in pediatric patients²⁵. Magli et al²⁶. conducted a long-term clinical trial and confirmed that the visual acuity and postoperative complications did not significantly differ between children with congenital cataract undergoing stage I and II IOL implantation after cataract extraction combined with posterior capsulorhexis and anterior vitrectomy. Carrigan et al²⁷. demonstrated that the cost was significantly higher for cataract surgery combined with stage I IOL implantation than for corneal contact lenses. Therefore, most researchers from Europe and America recommend performing stage II IOL implantation in children aged ≥ 2 years regardless of unilateral or bilateral eye diseases¹².

Scholars from South Korea demonstrated that the age of two years is the optimal time for IOL im-

plantation. Otherwise, the prognosis of visual acuity is likely to be reduced. In a retrospective study conducted by Seoul National University between 1993 and 2004, 37 children with bilateral congenital cataract underwent early cataract extraction combined with refractive error correction by glasses and stage II IOL implantation. Postoperative visual acuity was then assessed. The researchers conducted an 81.4 month follow up and concluded that children aged < 8 weeks had UCVA of 20/50 after undergoing cataract extraction combined with refractive error correction by wearing glasses, followed by stage II IOL implantation at approximately age two. Among these patients, 44% of eyes obtained UCVA $\geq 20/40$ ²⁸. The UCVA was also significantly better in this group than in the cataract extraction and stage I IOL implantation group and the stage II IOL implantation group (aged > 2 years)²⁹.

Previous studies have indicated that long-term visual acuity can be accurately estimated in children aged 7 years³⁰. Therefore, Korean scholars suggested that IOL implantation should be performed at approximately age 2 to correct early visual acuity, which is of significance for improving the prognosis of visual acuity. Consideration of multiple factors, such as the rapid and unpredictable development of infant eyes, the difficulty in selection of IOL degree, and final postoperative visual function, has led to the proposal that the age of two years is the optimal time for IOL implantation.

Conclusion

Congenital cataract is a primary cause of low vision and blindness in children, which not only worsens social and family burdens, but also creates various challenges for clinical treatment. At present, further investigation is still needed regarding the selection of appropriate postoperative refractive correction and the proper timing of IOL implantation. Many Chinese scholars have proposed that stage II IOL implantation should be performed at age 2-3 years in children with bilateral congenital cataract. For those diagnosed with unilateral congenital cataract, stage I IOL implantation should be conducted immediately following cataract extraction regardless of age. However, ophthalmologists abroad have recommended

performing stage II IOL implantation at approximately 2 years of age, regardless of unilateral or bilateral congenital cataract. Pediatric ophthalmologists should pay attention to each step of treatment, aiming to obtain the highest clinical efficacy.

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