Korean Journal of Ophthalmology 22(2):87-91, 2008
DOI : 10.3341/kjo.2008.22.2.87

Clinical Outcomes of Surgical Techniques in Congenital Cataracts

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Purpose: To investigate the general clinical features of congenital cataracts and to determine their relationship to visual prognosis and surgical complications according to age at operation and surgical procedure adopted.
Method: We retrospectively evaluated 92 eyes in 61 patients with congenital cataracts who underwent cataract surgery between January 1996 and December 2006. The demographic data, surgical technique, post-operative complications, and final visual prognosis were evaluated.
Results: The average age at surgery was 3.17 years (range 1 month to 11 years), and the mean follow-up was 40.02 months (range 6 to 46 months). Of the 56 eyes that could be checked for visual acuity after cataract extraction, 29 (51.7%) had a BCVA of ≥0.5 at last visit. Unilateral congenital cataracts (p=0.025) and congenital cataracts with strabismus (p=0.019) showed significantly poorer visual outcomes. Patients with nystagmus also experienced a poor visual outcome; 6 patients (67%) had a BCVA of <0.1. Posterior cataracts had the worst visual prognosis (p=0.004). No statistically significant differences in posterior capsular opacity (p=0.901) or synechia formation (p=0.449) were observed between surgical techniques, but children younger than one year showed a higher tendency for PCO and synechia formation.
Conclusions: Anterior vitrectomy did not reduce postoperative complications. Higher rates of complications (PCO, posterior synechia) developed in children younger than one year of age.
Korean Journal of Ophthalmology 22(2):87-91, 2008

Key Words: Anterior vitrectomy, Congenital cataract, Posterior capsular opacity

Congenital cataracts are the most common cause of treatable childhood blindness and low vision, accounting for 5% to 20% of blindness in children worldwide. Foster et al[1] reported that about 200,000 children are blind as a result of cataracts. The global incidence of congenital cataracts has been reported to be 1-15/10,000 live births.[2]

Cataract surgery in children has improved dramatically over recent decades, mainly as a result of modern surgical techniques and improved intraocular lenses. However, several issues remain to be resolved, for example, when to perform surgery, how to avoid amblyopia and surgical complications like posterior capsular opacity (PCO), and how to determine the intraocular lens power in children of different ages. Several studies have concluded that amblyopia in unilateral cataracts and PCO are major complications of congenital cataract surgery.[3] PCO may markedly reduce visual acuity due to occlusion of the visual axis and result in deep amblyopia. Several techniques can avoid PCO, such as posterior continuous curvilinear capsulorhexis (PCCC), anterior vitrectomy, IOL implantation with optic capture, and the bag-in-the-lens technique. A better understanding of irreversible deprivation amblyopia and its treatment are very important.[4]

The goal of the present study was to investigate the general clinical features of congenital cataracts and to determine the relationship with visual prognosis and surgical complications according to age at operation and surgical procedure adopted.

Materials and Methods

This retrospective study included 61 children (92 eyes) with congenital cataracts who underwent surgery at the Samsung Medical Center in Seoul, Korea between January 1996 and December 2006, and were followed up for at least 6 months after surgery. Eyes with traumatic cataracts and associated retinal anomalies, systemic diseases, or neurological disorders were excluded. Thirty children underwent unilateral cataract surgery, and thirty-one underwent bilateral cataract surgery.

Study parameters included initial demographic data, age at diagnosis and operation, uncorrected and best-corrected pre- and post-operative visual acuities (if possible), intraocular pressure, type of cataract, associated ocular pathologies, other

Received: October 15, 2007 *Accepted*: April 13, 2008

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systemic disorders, and postoperative complications.

Biometry was performed using a handheld Retinomax K-plus2 autokeratometer (Nikon Inc., Japan), and axial length measurements were made using an Allergan Humphrey Ultrasonic biometer 835 (Allergan Humphrey Inc., USA). Lens power was determined using the SRK-Ⅱ, T and Hoffer Q formulae, aiming for emmetropia. However, because the indicated power was often considerably above +28 diopters, more than 1 IOL was implanted using the piggy-backing method. Three types of acrylic IOL implants were used during the study period (Table 1).

The surgery was performed in the following manner. Anterior capsulorhexis was performed using a CCC (continuous curvilinear capsulorhexis) forceps. Cataracts were aspirated via two corneal incisions, which were sutured after surgery. PCCC was always carried out as a follow-on procedure using a CCC forceps.

Patients were divided into three groups according to the subsequent surgical steps. In group 1, anterior vitrectomy and IOL implantation with optic capture was performed. The capture process was achieved by gentle pressure on one half of the IOL optic 90 degrees from the haptic-optic junction, followed by slow and gentle pressure on the other half of the optic through the posterior capsulorhexis opening. In group 2, IOL implantation with optic capture without anterior vitrectomy was performed. In group 3, no IOL was inserted and anterior vitrectomy alone was performed.

If a tear of the capsulorhexis edge was observed, optic capture was abandoned and bimanual anterior vitrectomy through PCCC was performed in all cases. Those cases were excluded from all groups (8 eyes). During follow up, spectacle correction of residual ametropia and occlusion therapy were performed as early as possible after cataract surgery. Typical occlusion therapy regimens for infantile onset were 6 to 8 hours per day and for later onset, 2 to 6 hours per day in unilateral cases, with adjustments according to the degree of amblyopia in bilateral cases.

Statistical data were obtained using SPSS 9.0 software. Qualitative data (complication rates between the two age groups and between the three different groups associated with surgical procedures) were compared using the chi-square test. Because group distributions were not Gaussian, quantitative data (visual acuities between unilateral and bilateral cataracts and between patients with and without strabismus) were compared using the Mann-Whitney U test, and ranks of visual acuity associated with the type of cataract were obtained using LSD: multiple comparisons with ranks. P-values of <0.05 were considered significant.

Table 1. IOL implants used during the study period

	"A" Constant	Number of patients
Alcon Acrysof MA30BA	118.9	59
Alcon Acrysof SA60BB	118.4	1
HOYA VA60BB	118.7	11

Table 2. Demographic data

Total (persons/eyes)	61/92
Sex (M:F)	26 (43%) : 35(57%)
Laterality (uni-:bi-)	30 (49%) : 31(51%)
Strabismus	17 (28%)
Nystagmus	9 (15%)
Mean age at diagnosis (years)	2.82 (±3.41)
Mean age at operation (years)	3.17 (±3.53)
Mean follow-up (months)	40.02 (±29.99)

* M=male; F=female.

Results

Demographic data and clinical features

Sixty-one patients (26 males, 35 females) were included in this study. Sixty-two eyes from 31 patients underwent surgery for bilateral congenital cataracts and thirty eyes from 30 patients underwent surgery for unilateral cataracts. The mean age at the time of surgery was 3.17 years (range 1 month to 11 years), and the mean follow-up period was 40.02 months (range 7 to 97 months). Of the 61 patients, 17 patients (28%) had strabismus and 9 patients (15%) had nystagmus prior to cataract removal. Postoperatively, the corrected and uncorrected visual acuity could be checked in 56 eyes (60.9%) with a Snellen acuity chart.

Unilateral congenital cataracts (logMAR 0.71 vs 0.25, *p*=0.025) and congenital cataracts with strabismus (logMAR 0.93 vs 0.31, *p*=0.019) showed significantly poorer visual

Table 3. Clinical features and visual outcomes (Best Corrected Snellen visual acuity)

Clinical feature (n)		n (%)		
		HM-<0.1	0.1-0.4	>0.4
uni- or bilateral	bilateral (32)	0 (0)	10 (31)	22 (69)
	unilateral (24)	5 (21)	11 (46)	8 (33)
accompanying strabismus	monocular (13)	3 (23)	8 (62)	2 (15)
	alternate (4)	0 (0)	2 (50)	2 (50)
nystagmus (9)		6 (67)	2 (23)	1 (10)

* HM=hand movement.

outcomes than bilateral cataracts and cataracts without strabismus, respectively (Mann-Whitney U test). Patients with nystagmus experienced poor visual outcomes as well; 6 patients (67%) had a BCVA of <0.1 (Table 2, 3).

Types of cataracts

There were four types of congenital cataracts (lamellar, nuclear, posterior and anterior polar). Nuclear cataracts had a significantly poorer visual outcome than lamellar cataracts (logMAR 0.74 vs 0.32, p=0.016). Posterior cataracts, which included both posterior polar and posterior subcapsular opacities in this study, had a significantly poorer outcome than nuclear cataracts (logMAR 0.41 vs 0.32, p=0.004) (LSD: multiple comparisons with ranks). Lamellar cataracts had the best visual prognosis, while posterior cataracts had the worst. Posterior cataracts consisted of 21 cases (91.3%) of unilateral cataracts and 2 cases (8.7%) of bilateral cataracts (Table 4).

Complications

No statistically significant differences in posterior capsular opacity (p=0.901) or synechia formation (p=0.449) were observed between the three different groups associated with surgical procedures, as mentioned above (Chi-square test). One eye (6.2%) in group 2 and 2 eyes (10%) in group 3 developed secondary glaucoma (Table 5).

We also compared the complication rates between the patients who underwent cataract surgery before and after 1 year of age. The younger patients showed a significantly higher rate of synechia formation than the older (p=0.002). However, there was no statistically significant difference in terms of PCO formation (p=0.503), although the younger group showed a higher tendency for PCO formation (Chi-square test) (Fig. 1).

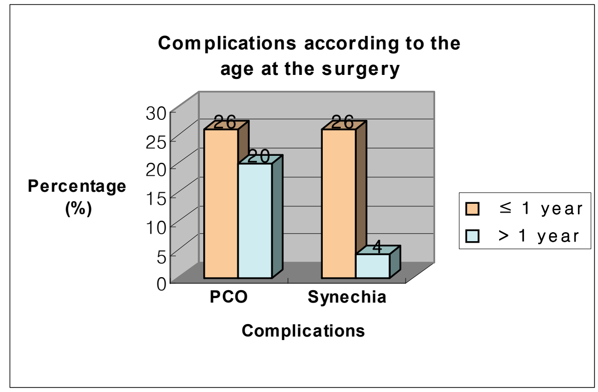

Fig. 1. Comparison of complication rates between the two age groups, before and after 1 year of age at operation (n=92 eyes). A significant difference in synechia formation was observed between these two groups (p=0.002), though there was no significant difference in PCO formation (p=0.503) (Chi-square test).

Table 4. Cataract types and visual outcomes (Best Corrected Snellen visual acuity)

type of cataract (N)	n (%)			Total (n)
	HM-<0.1	0.1-0.4	>0.4	
lamellar (6)	0	0	6 (100)	6
nuclear (62)	0	12 (43)	16 (57)	28
posterior (23)	5 (24)	9 (43)	7 (33)	21
anterior polar (1)	0	0	1 (100)	1
Total (92)				56

* HM=hand movement; N=total number of eyes in study population; n=total number of eyes that could be checked for best corrected visual acuity (BCVA)

Visual outcomes according to age at operation and laterality

Of the 56 eyes that could be checked for visual acuity, 25 eyes (45.4%) had BCVA of ≥0.5 after cataract extraction at last visit, although the statistical significance of visual acuities according to age at operation and laterality could not be evaluated due to unequal sample size in all of the age groups (Table 6).

Discussion

The visual prognosis of children with congenital cataracts has improved dramatically. Today, most pediatric ophthalmologists agree that earlier detection, prompt treatment and management of amblyopia, advances in microsurgical techniques and instrumentation, and IOL developments have contributed greatly to the management of congenital cataracts.[5,6]

Table 5. Postoperative complications according to surgical techniques

Procedure (n)	n (%)						
	PCO	Synechia	Eccentric pupil	Decenteration	Pigmentation	RD	Glaucoma
group 1 (48)	7 (15)	5 (10)	0 (0)	1 (2)	1 (2)	1 (2)	0 (0)
group 2 (16)	3 (19)	2 (13)	1 (7)	0 (0)	1 (7)	0 (0)	1 (7)
group 3 (20)	3 (15)	5 (25)	2 (10)	1 (5)	0 (0)	0 (0)	2 (10)
Total (84)							

* PCO=posterior capsular opacity; RD=retinal detachment.
* group 1=anterior vitrectomy and IOL implantation with optic capture; group 2=IOL implantation with optic capture alone (without anterior vitrectomy); group 3=anterior vitrectomy alone without IOL implantation.

Table 6. Visual outcomes (Snellen) according to age at operation and laterality

V/A/Age	n (%)								Total (n)
	0 mon~3 mon		3 mon~18 mon		18 mon~3 yrs		3 yrs~		
	bi-	uni-	bi-	uni-	bi-	uni-	bi-	uni-	
HM<0.1	0 (0)	0 (0)	1 (25)	3 (33)	0 (0)	2 (33)	0 (0)	2 (25)	8
0.1-0.4	4 (67)	6 (86)	1 (25)	4 (44)	2 (67)	3 (50)	1 (8)	2 (25)	23
>0.4	2 (33)	1 (14)	2 (50)	2 (23)	1 (63)	1 (17)	12 (92)	4 (60)	25

* V/A=visual acuity; HM=hand movement; mon=months; yr=years; uni-=unilateral; bi-=bilateral.

In this study, we investigated the general clinical features and surgical outcomes of congenital cataracts. It was found that in congenital cataracts, the presence of strabismus, nystagmus and posterior cataracts are associated with poor visual outcomes, which confirms previously published data.[7]

In contrast to adult cataract surgery, pediatric cataract surgery is often accompanied by postoperative complications, such as posterior capsular opacity (PCO), posterior synechia, secondary glaucoma, fibrinoid reaction, and pupil decenteration.[7] Of these, opacification of the visual axis is the most common and most severe vision-threatening complication in children, particularly in the youngest. Even when posterior capsulorhexis has been performed, ingrowth of lens epithelial cells (LEC) on the vitreous surface or on the back of the IOL optic can be found months after surgery. Moreover, the incidence of PCO after cataract surgery is nearly 100% in infants. Therefore, Nd:YAG laser posterior capsulotomy or a surgical procedure should be adopted for PCO removal.[8,9] As mentioned, several methods have been proposed to minimize PCO, e.g., anterior vitrectomy, IOL optic capture, and 'bag-in-the-lens' techniques. Fenton and O'Keefe[10] reported that 15.6% of children with PCO who underwent PCCC and no vitrectomy required Nd:YAG capsulotomy. Anterior vitrectomy is performed to remove the scaffold of LEC migration and regeneration. It has also been postulated that even in the absence of LEC migration, vitreous opacification may be the result of a primary response to the contact between the anterior vitreous face and the IOL optic, which is another reason why routine anterior vitrectomy should be performed.[11] However, some complications such as increased IOP, macular edema, IOL decentration, and vitreous traction may occur after anterior vitrectomy.[7] To minimize these complications, several studies have reported that PCCC with optic capture prevents PCO of the visual axis even in the absence of an anterior vitrectomy.[12,13] Gimbel and DeBroff[14] first found that IOL implantation with optic capture prevented the development of posterior capsular opacity without anterior vitrectomy. When optic capture is performed, the leaflet of the anterior and posterior capsulorhexis are apposed and sealed in front of the IOL optic, except at the haptic-optic junction. Theoretically, LECs will proliferate between the anterior and posterior capsules, preventing migration over the anterior hyaloid face. In contrast, Koch and Kohnen[15] reported that 4 of 5 patients who underwent optic capture without anterior vitrectomy developed secondary cataracts at 2.5 years postoperatively. Other studies have concluded that anterior vitrectomy is beneficial when performed routinely with posterior capsulorhexis with optic capture, especially in children younger than 5 years.[16]

We did not find any significant differences between surgical procedures in terms of postoperative complications (e.g., PCO, synechia), which we attribute to different age distributions and short follow-ups. The mean ages (years) at operation in groups 1, 2, and 3 were 3.18 (±2.82), 3.78 (±2.56), and 1.02 (±2.36), and the corresponding mean follow-ups (years) were 3.45 (±1.80), 3.38 (±1.98), and 1.95 (±2.22). Thus, the mean age was lower and the mean follow-up period was shorter in group 3 than in the other two groups, which could indicate that the complication rate in group 3 is significantly underestimated.

Previous reports in the literature have recommended early surgery, i.e., at less than 8 weeks in bilateral cataracts, which is at odds with our finding of no relationship between the age at operation and the surgical outcome.[5,6,17] We assumed that patients with a relatively lower cataract density would have presented and been operated on later than patients with a higher cataract density, and would have achieved better visual outcomes. In the present study, we did not evaluate cataract density, which is a notable study limitation.

The optimal timing of cataract surgery in children with congenital cataracts remains an open question. Improvements in our understanding of critical periods of visual development have led to the acceptance of surgical interventions for dense cataracts within the first 3 months of life, possibly as early as 6 weeks for unilateral disease.[18] In contrast, it has been proposed that children with bilateral congenital cataracts should be treated during a period, i.e., extending to 8 weeks of life.[19]

Vishwannath et al.[20] cautioned that although surgery for visually significant cataracts must be performed within this critical period to prevent amblyopia, complications rates are higher in earlier surgeries. Chak et al.[17] also reported a significant association between lower age at surgery and serious postoperative complications. In this study, complications (PCO, synechia) tended to increase in younger children.

Secondary glaucoma, which is usually of the open-angle type, is one of the most serious complications that occurs after pediatric cataract surgery. Vishwanath et al.[20] reported that 50% of children who had undergone bilateral lensectomies during the first month of life developed glaucoma in one or both eyes at the 5-year follow-up as compared with only 15% of children who underwent surgery at a later time. Lundvall and Kugelberg[6] found that 80% of children with bilateral congenital cataracts who developed glaucoma underwent cataract surgery during the first 4 weeks of life. Accordingly, they proposed that cataract surgery should be deferred until after the first 4 weeks of life. In the present study, 3 eyes (3.2%), all of which underwent surgery within the first 6 months after birth, developed secondary glaucoma, which is comparable with the results of the reports previously mentioned. Further evaluation and long-term follow-up is recommended in this area.

Another limitation of this study is neglect of the degree of compliance with occlusion. Chak et al.[17] concluded that compliance with occlusion was the factor most strongly associated with visual outcome in bilateral and unilateral disease, which represents a critical limitation of the present study.

In conclusion, the present study revealed that visual outcome was found to depend on laterality, cataract type, and associated ocular pathologies (strabismus, nystagmus). We found that anterior vitrectomy had no additional effect in reducing postoperative complications. However, this could have been due to the different age distributions and unequal sample sizes in our study groups. Nevertheless, we detected higher rates of complications (PCO, posterior synechia) in children younger than one year. Thus, we recommend that further larger-scale, longer-term prospective studies, which take into account the degree of cataract density and compliance with amblyopia treatment, be conducted to avoid selection bias and to achieve higher statistical power. Finally, we advise that practitioners follow not only advances in surgical techniques, materials, and instrumentation, but also improvements in the management of amblyopia, particularly in terms of early detection and occlusion therapy to improve visual prognosis in cases of congenital cataracts.

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