

Clinical features of strabismus and nystagmus in bilateral congenital cataracts

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Received: 2017-09-08 Accepted: 2017-11-14

Abstract

• **AIM:** To evaluate the prevalence, clinical features, and the factors affecting onset of strabismus and nystagmus in patients with bilateral congenital cataracts.

• **METHODS:** This study evaluated 116 eyes of 58 patients who underwent lens removal for the treatment of bilateral congenital cataracts between January 1999 and January 2011. The presence and type of strabismus and nystagmus were determined before and after surgery. Type of strabismus and final visual acuity were compared in patients with and without nystagmus. Patients were divided into three groups (orthotropia/orthotropia, orthotropia/strabismus, and strabismus/strabismus) according to their preoperative and postoperative ocular alignment. Age at cataract surgery and associations of nystagmus and primary intraocular lens (IOL) implantation with strabismus were analyzed.

• **RESULTS:** Six patients (10.3%) had strabismus preoperatively and an additional 11 (19.0%) developed postoperative strabismus. Exotropia was more common than esotropia both preoperatively and postoperatively. Eighteen patients (31.0%) had postoperative nystagmus, with sensory nystagmus being the most common type. Of the 18 patients with nystagmus, 10 had strabismus, with exotropia being more common than esotropia. Postoperative visual acuity was poor in patients with nystagmus. Age at cataract surgery and rate of primary IOL implantation were significantly lower, and postoperative nystagmus was more common, in the orthotropia/strabismus group than in the other two groups.

• **CONCLUSION:** Exotropia and sensory nystagmus are common in patients with bilateral congenital cataracts. Age at cataract surgery and rate of IOL implantation are lower

and nystagmus more common in patients with postoperative onset of strabismus. Nystagmus is associated with poor visual prognosis.

• **KEYWORDS:** bilateral congenital cataracts; strabismus; nystagmus

DOI:10.18240/ijo.2018.05.16

Citation: Hwang SS, Kim WS, Lee SJ. Clinical features of strabismus and nystagmus in bilateral congenital cataracts. *Int J Ophthalmol* 2018;11(5):813-817

INTRODUCTION

Congenital cataract is one of the most important diseases that can cause visual impairment in children. It is often accompanied by other eye conditions, such as strabismus and nystagmus. The prevalence of strabismus in patients with bilateral congenital cataracts has been reported to be 25.9%-78.9%^[1-4]. However, the development of cataract surgery techniques and the early implantation of intraocular lens (IOL) have reduced the prevalence of strabismus^[5]. The types of strabismus associated with bilateral congenital cataracts can include both esotropia and exotropia, with studies differing in which is more common^[1-4,6-7]. Rates of nystagmus in these eyes have been reported to be 24.2% before cataract surgery and 20.0%-90.9% after cataract surgery, depending on the severity of the cataract^[8-9]. The prevalence rates of strabismus and nystagmus, and the types of strabismus, have been found to vary widely among studies. Moreover, few studies to date have evaluated strabismus and nystagmus in patients with bilateral congenital cataract. This study was therefore designed to evaluate the prevalence and clinical features of strabismus and nystagmus, and the factors affecting the onset of strabismus and nystagmus, in patients with bilateral congenital cataract.

SUBJECTS AND METHODS

This study included 116 eyes of 58 patients, aged 6wk to 19y, who underwent cataract surgery for bilateral congenital cataract from January 1999 to January 2011. All operations were performed by one surgeon in our department and all were followed-up for a minimum of 4y. This study was approved by an accredited institutional review board (IRB No.129792-2015-070) and complied with the Declaration of Helsinki.

Congenital cataracts were defined as those diagnosed within the first year of life or those in patients with a family history of

Strabismus and nystagmus in congenital cataracts

Table 1 Demographic data of 58 patients based on the presence of strabismus before and after cataract extraction mean±SD (range)

Variables	Orthotropia/orthotropia (n=41)	Strabismus/strabismus (n=6)	Orthotropia/strabismus (n=11)	P
Age at cataract extraction	5.9±5.0y (8wk-19y)	3.7±3.3y (7mo-12y)	0.7±1.4y (6wk-5y)	<0.001 ^a
Age at postop. strabismus			2.6±2.3y (4mo-6.6y)	
Interval ^b			1.8±1.8y (3wk-6y)	

^aANOVA test; ^bInterval between cataract extraction and onset of strabismus.

congenital cataracts. Lamellar cataract, anterior polar cataract, posterior polar cataract, posterior subcapsular cataract, nuclear cataract, total cataract, and cataract accompanied by lentiglobus were also defined as congenital cataract. Patients with other ocular diseases that could affect visual acuity or history of trauma or systemic disease were excluded.

Cataract surgery was performed under general anesthesia. A 3.2-mm main incision was made 1.5 mm behind the corneal limbus at the 12 o'clock position and a 1.0-mm side incision was made at the corneoscleral limbus at the 3 o'clock position. Following anterior continuous curvilinear capsulorhexis of 4.5-5.0 mm, hydrodissection was performed using balanced salt solution. The cataract was removed using an irrigation and aspiration technique. Except for one child aged 7mo, primary IOLs were not implanted into children less than 14 months of age; rather, primary IOLs were implanted 15mo after birth. IOL power was determined using the SRK-T value and intentionally undercorrected to be emmetropic when patients became adults. If IOL implantation was possible, posterior continuous curvilinear capsulorhexis was performed, the IOL was implanted in the bag and optic capture was performed. Alternatively, posterior continuous curvilinear capsulorhexis was performed, followed by anterior vitrectomy and IOL implantation in the bag without optic capture. Fundus examination after cataract surgery showed that all patients were normal.

The prevalence and clinical features of strabismus and nystagmus were reviewed retrospectively using patients' medical records, and the factors related to occurrence of strabismus were evaluated. Before and after surgery, the presence and type of strabismus and nystagmus were determined. Strabismus was diagnosed as a deviation ≥ 10 prism diopters at the primary position on alternate prism and cover tests. Nystagmus was diagnosed by clinical features. Manifest latent nystagmus (MLN) was defined as a decelerating velocity of the nystagmus slow phase, always beating towards the open eye. Latent nystagmus (LN) was defined as an MLN oscillation during monocular viewing but steady fixation during binocular viewing. Congenital nystagmus (CN) was defined as a slow phase that was pendular or accelerating. Sensory nystagmus (SN) was defined as nystagmus that was not MLN, LN or CN. SN was also characterized as bilateral, conjugate, pendular or jerk nystagmus.

Statistical Analysis Statistical analysis was performed using SPSS 18.0 (SPSS Inc., Chicago, IL, USA). Comparisons were performed using Pearson's Chi-square test, ANOVA, or Fisher's exact test. A *P* value ≤ 0.05 was considered statistically significant.

RESULTS

Of the 58 patients with congenital cataract, 33 were male and 25 were female. Their mean age at cataract diagnosis was 4.1±4.5y (range, 4wk-19y), their mean age at cataract removal was 4.7±4.8y (range, 6wk-19y) and the mean follow-up period was 5.5±2.1y (range, 4-12y).

Nineteen patients, of mean age 0.5±0.4y (range, 6wk-14mo), did not undergo primary IOL implantation. Of the 39 patients who underwent primary IOL implantation, 29 underwent IOL optic capture. Except for one patient who underwent surgery at age 7mo, when the primary IOL was implanted, all patients who underwent primary IOL implantation were ≥ 15 months of age. The mean age of all patients who underwent IOL implantation was 6.8±4.7y (range, 7mo-19y). Patients who did not undergo primary IOL implantation were significantly younger than those who underwent primary IOL implantation (*P*<0.001). The patient who underwent primary IOL implantation at 7 months of age was the youngest of the patients with strabismus before surgery, with early IOL implantation performed to increase eye alignment and recovery of visual function. Six patients (10.3%) with bilateral congenital cataract had strabismus preoperatively, with 11 (19.0%) developing strabismus postoperatively. Thus, at final follow-up, 17 patients (29.3%) had strabismus.

Patients were divided into three groups (orthotropia/orthotropia, orthotropia/strabismus, and strabismus/strabismus) according to their preoperative and postoperative ocular alignment. The orthotropia/strabismus group included one patient diagnosed with cataract at age 2y who did not undergo cataract surgery until age 5y; all others in this group underwent cataract surgery within 8mo (0.7y) of birth, an age significantly younger than the other groups. Mean age at strabismus occurrence after cataract surgery was 2.6y, or a mean 1.8y after cataract surgery (Table 1).

Patients who had strabismus before surgery were found to maintain the same type of strabismus after cataract surgery except for one patient. That patient, who underwent cataract extraction and primary IOL implantation at 4 years of age, had

dissociated vertical deviation before surgery and dissociated vertical deviation and exotropia after surgery. Exotropia occurred after posterior capsular opacification developed, with the patient having visual acuity of finger count preoperatively and postoperatively and SN. Of the patients with strabismus, 12 (70.6%) had exotropia and five (29.4%) had esotropia at final follow-up. Exotropia was more common than esotropia both preoperatively and postoperatively (Figure 1).

Postoperative nystagmus was observed in 18 patients (31.0%), including SN in ten, MLN in five, CN in one, and LN in two. One patient with LN before surgery developed MLN after surgery, one with MLN before surgery developed LN after surgery, and one patient without nystagmus before surgery developed LN at 4mo after surgery.

When we examined the prevalence of nystagmus according to preoperative and postoperative ocular alignment in patients with bilateral congenital cataracts, we found that 19.5% of patients in the orthotropia/orthotropia group and 50.0% of those in the strabismus/strabismus group had nystagmus. The prevalence of nystagmus in the orthotropia/strabismus group increased significantly, from 54.5% before surgery to 63.6% after surgery ($P=0.015$, Fisher's exact test).

The patient with preoperative LN who developed postoperative MLN was in the orthotropia/orthotropia group. This patient underwent surgery at 3 months of age but did not undergo primary IOL implantation. The patient with preoperative MLN who developed postoperative LN had esotropia preoperatively. This patient underwent cataract surgery and primary IOL implantation at age 7mo, making this patient the youngest in the strabismus/strabismus group to undergo cataract extraction. The patient who newly developed LN after surgery belonged to the orthotropia/strabismus group. Cataracts were extracted from this patient's right and left eyes at ages 3 and 4mo, respectively, but the patient did not undergo primary IOL implantation.

Of the 18 patients with preoperative or postoperative nystagmus, 10 had strabismus. Six had exotropia, including exotropia in three, dissociated vertical deviation and exotropia in two, and dissociated horizontal exodeviation in one. Four had esotropia, including dissociated vertical deviation and esotropia in one. Of the 40 patients without nystagmus, seven patients had strabismus, including six with exotropia and one with esotropia. Type of strabismus did not differ significantly between patients with and without nystagmus ($P=0.338$, Fisher's exact test). Preoperative and postoperative nystagmus in bilateral congenital cataracts was significantly associated with postoperative occurrence of strabismus. Of the 11 patients in the orthotropia/strabismus group, 10, including seven with exotropia and three with esotropia, did not undergo primary IOL implantation. Eight of these patients developed strabismus when they became aphakic, with the other two developing

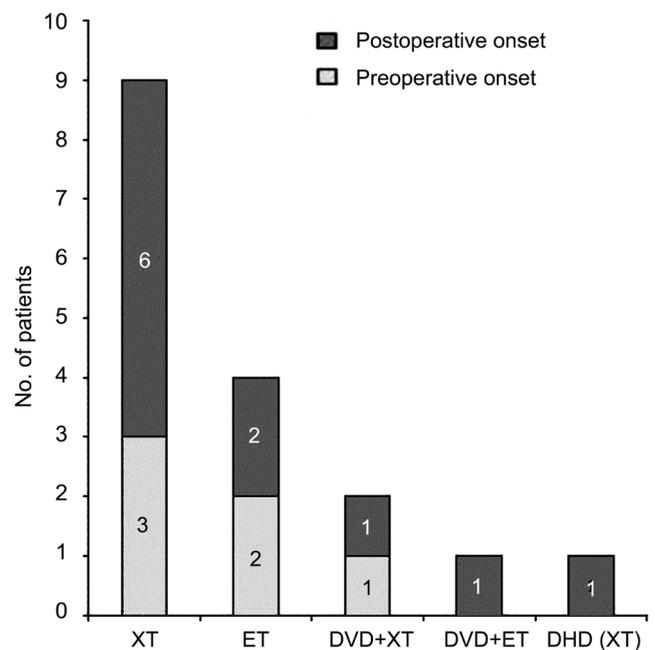


Figure 1 Type of postoperative deviation in the 17 patients with strabismus XT: Exotropia; ET: Esotropia; DVD: Dissociated vertical deviation; DHD: Dissociated horizontal deviation.

Table 2 Associations of nystagmus and primary IOL implantation with postoperative onset of strabismus

Nystagmus	Orthotropia/ orthotropia (n=41)	Orthotropia/ strabismus (n=11)	^a P
Preop. nystagmus (-)	33	5	0.050
Preop. nystagmus (+)	8	6	
Postop. nystagmus (-)	33	4	0.008
Postop. nystagmus (+)	8	7	
Primary IOL implantation (-)	8	10	<0.001
Primary IOL implantation (+)	33	1	

^aFisher's exact test.

Table 3 Associations of nystagmus with strabismus

Nystagmus	Strabismus (-) (n=41)	Strabismus (+) (n=17)	P
Preop. nystagmus (-)	33	8	0.024 ^a
Preop. nystagmus (+)	8	9	
Postop. nystagmus (-)	33	7	0.003 ^b
Postop. nystagmus (+)	8	10	

^aFisher's exact test; ^bPearson's Chi-square test.

strabismus after secondary IOL implantation. Of the patients who were orthotropic preoperatively, 18 did not undergo primary IOL implantation, with 10 of the latter (55.6%) developing strabismus after cataract surgery.

The incidence of postoperative strabismus was higher in patients without than with primary IOL implantation for bilateral congenital cataracts (Table 2). The presence of strabismus, whether preoperative or postoperative, was associated with a high probability of accompanying nystagmus (Table 3). The average postoperative visual acuity in patients with bilateral congenital cataract was 0.7 ± 0.4 (range, hand

motion-20/13). The postoperative visual acuity of 20/40 or less was found in 11 of the 40 patients without nystagmus, and 13 of the 18 patients with nystagmus. Thus, postoperative visual acuity $\leq 20/40$ was significantly more frequent in patients with than without nystagmus ($P < 0.001$, Pearson's Chi-square test).

DISCUSSION

Strabismus may be more common in patients with congenital cataracts than in the normal population because of the lack of visual stimuli due to cataracts, anisometropia or aniseikonia after cataract surgery, and/or differences in binocular visual acuity. Strabismus may be a risk factor for developmental deterioration in binocular vision and additional amblyopia and may also negatively affect the aesthetic and psychological stability of the patient^[4].

Although the prevalence of strabismus is 1.3%-4.5% in the normal population^[10-12], its prevalence in patients with bilateral congenital cataract has been reported to vary from 25.9%-78.9%^[1-4]. A study in 19 patients with bilateral congenital cataract found that 11 (57.9%) had preoperative and 15 (78.9%) had postoperative strabismus in aphakic state, with esotropia being more common than exotropia^[1]. In contrast, a study in 27 bilateral pseudophakic patients reported that seven (25.9%) had strabismus, with exotropia being more common than esotropia^[2]. Strabismus was also observed postoperatively in 102 (30.3% of 337 patients with unilateral and bilateral congenital cataract and in 72 (23.2%) of 311 patients with unilateral and bilateral acquired pediatric cataracts^[3]. Of 336 patients with bilateral cataracts, 66 (19.6%) had strabismus, with esotropia being more common in congenital and exotropia more common in acquired cataract^[3]. Another study reported that, of 65 patients with bilateral congenital cataracts without preoperative strabismus, 35 (53.8%) developed postoperative strabismus^[4]. With regard to the type of strabismus, exotropia may occur when there is low vision before the development of convergence, with esotropia occurring afterwards^[4]. Type of strabismus may also depend on refractive error in eyes with good visual acuity, with esotropia being more common in hyperopia and exotropia more common in emmetropia and myopia^[4,6-7]. The ratio of esotropia to exotropia was found to be about 1:1 in patients who developed strabismus and decreased vision between birth and age 5y, whereas exotropia was more common than esotropia in patients who developed these conditions after age 5y^[6]. In our study, strabismus was observed in six patients (10.3%) before surgery and 11 additional patients (19.0%) developed strabismus after surgery, such that the final number of patients with strabismus was 17 (29.3%). Of the six patients with strabismus before surgery, four had exotropia and two had esotropia; whereas, of the 17 patients with postoperative strabismus, 12 had exotropia and five had esotropia. Thus, in our patient population, exotropia was more common than esotropia both preoperatively and postoperatively.

A study of the relationship between age at cataract surgery and strabismus found that the incidence of strabismus was lower in patients who underwent unilateral pediatric cataract surgery before 49d after birth, because the first month of life is the most important period for the development of binocular vision and stereopsis^[5]. Although studies have reported no correlation between age at unilateral or bilateral congenital cataract surgery and strabismus, other studies have reported that younger age at surgery in patients with bilateral pediatric cataract was significantly correlated with postoperative strabismus incidence^[2,4,13-14]. Moreover, of patients who developed strabismus after cataract surgery, 70.2% did so within 1y^[4]. In the present study, patients in the orthotropia/strabismus group were significantly younger at cataract surgery (mean age 0.7y) than patients in the other two groups, with 45.5% of the former developing strabismus within 1y after cataract surgery. The more frequent occurrence of strabismus after cataract surgery despite early cataract surgery may be related to the timing of congenital cataract diagnosis. Impairments in early visual stimuli may inhibit the development of the subcortical pathway and may affect fixation stability, resulting in nystagmus. The most common initial symptoms of cataracts in children are nystagmus or random eye movement. Although these patients may be diagnosed and undergo surgery at a younger age^[15], the incidence of strabismus may be higher in patients with preexisting nystagmus. In the present study, cataract was diagnosed significantly earlier in patients with preoperative nystagmus, except for two patients who were first diagnosed with cataract in our institution at ages 18 and 19y.

Congenital cataract is accompanied by nystagmus in 20.0% to 90.9% of patients^[8-9,16]. Nystagmus is known to be more common in patients with bilateral congenital cataracts than in patients with unilateral congenital cataracts^[17]. A study of 24 patients with bilateral congenital cataract found that most had nystagmus after surgery, with these patients having a poor visual prognosis^[18]. Another study reported that nystagmus was present in 24.2% and 90.9% of patients before and after cataract surgery, respectively, but was not related to visual prognosis^[8]. Postoperative nystagmus in patients with good visual acuity was small in amplitude, intermittent, or latent, although that study, unlike the present study, included patients with other ocular anomalies or systemic disease. Another study found that only 20.0% of patients with bilateral congenital cataract had nystagmus, despite undergoing surgery after age 10mo, with nystagmus being a poor prognostic indicator^[9]. Similarly, we found that nystagmus was present in 18 patients (31.0%) after congenital cataract surgery, with postoperative visual acuity of 20/40 or less being significantly more frequent in patients with than without nystagmus.

In the present study, 10 of 18 patients (55.6%) with nystagmus also had strabismus, with this percentage significantly higher

than in patients without nystagmus. In contrast, other studies have found that the presence of nystagmus was not significantly associated with the development of strabismus^[4]. Moreover, we found that exotropia was more common than esotropia in our patients with (6:4) than without (6:1) nystagmus, with no significant difference. Other studies have found that esotropia was more frequent in patients with nystagmus, suggesting that esotropia was associated with nystagmus compensation^[1].

Of the 11 patients who developed strabismus after congenital cataract surgery, 10 did not undergo primary IOL implantation. Moreover, the incidence of strabismus was high in patients without primary IOL implantation. Studies of patients with unilateral congenital cataract found that the incidence of strabismus was lower in those who underwent primary IOL implantation than in those who underwent contact lens correction^[5,19]. Although strabismus may occur due to improper visual stimuli, studies in patients with bilateral and unilateral congenital cataract found that the incidence of strabismus was similar in patients with pseudophakia and aphakia^[13,20].

This study had several limitations, including its retrospective design. In addition, the time of cataract diagnosis was late, obscuring the exact time of cataract onset; thus, the study may have included patients with acquired cataracts. A third limitation was that most patients were first evaluated after age 1mo, which is important for visual acuity development. Fourth, the time of the first visit and the types of cataract were too diverse to analyze. For example, type of cataract and nystagmus are known to be related. One study reported that 100% of patients with total congenital cataract and 43% of those with nuclear cataract, both unilaterally and bilaterally, may have nystagmus^[21].

In conclusion, the results of this study suggest that exotropia and SN may be common in patients with bilateral congenital cataracts. In addition, strabismus after cataract surgery was associated with early cataract surgery, lack of primary IOL implantation and nystagmus. Moreover, patients with nystagmus have a poor visual prognosis, indicating that these patients require careful observation.

ACKNOWLEDGEMENTS

Presented at the 117th Meeting of the Korean Ophthalmological Society, 2017.

Conflicts of Interest: Hwang SS, None; Kim WS, None; Lee SJ, None.

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