# Clinical characteristics of congenital and developmental cataract in Kazakhstan

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**Purpose:** To study and describe clinical characteristics of congenital and developmental cataract at a tertiary eye care facility. **Methods:** In this retrospective study, 942 children (1311 eyes) presenting with congenital/developmental cataract over a 10-year study period were included. Gender, age at surgery, main presenting complaint, morphologic type of cataract, laterality, family history, and associated findings were recorded. **Results:** The overall proportion of boys and girls undergoing cataract surgery was approximately equal (P = 0.110). However, in the cases of bilateral cataract, the proportion of boys was larger than girls (P = 0.028). More than half (62.3%) of the patients underwent surgery at the age of >3 years. The main presenting complaint was white pupils, accounting for 48.1% of cases. Total cataract was the most common morphologic type in all age groups. In total, 133 children out of 942 (14.1%) had a positive family history of congenital/developmental cataract. Strabismus and nystagmus were seen in 27.2% and 19.3% of the eyes, respectively. Additional ocular dysmorphology was found in 97 (10.3%) of patients. Coexisting systemic disease was found in 149 (15.8%) cases. Among syndrome-associated cataracts, Down syndrome accounted for the majority of cases. **Conclusion:** High prevalence of total cataracts as well as frequent association with strabismus and nystagmus are likely to be the consequences of delayed presentation.



Key words: Cataract, cataract surgery, child, congenital cataract, developmental cataract

Congenital and developmental cataracts are found to be among the leading causes of preventable childhood blindness worldwide.<sup>[1]</sup> The prevalence of congenital cataract varies greatly around the world.<sup>[2,3]</sup> It was found to range from 2.2 to 13.6 per 10,000 children globally.<sup>[2]</sup> The epidemiologic data is mostly focused on congenital cataract; thus, the prevalence of developmental cataract is not well understood, probably due to the difficulties in categorization and distinction between these two types of childhood cataracts.<sup>[4,5]</sup>

The etiology of congenital cataract remains largely unknown.<sup>[2]</sup> Wu *et al.*<sup>[2]</sup> in a systematic review and meta-analysis on the global prevalence and epidemiologic characteristics of congenital cataract reported that hereditary factors were responsible for 22.3% of them, while 62.2% of cases were idiopathic. Therefore, it is difficult to develop prevention strategies. As such, early diagnosis and treatment are crucial for favorable visual outcomes.<sup>[6–8]</sup> It has been reported that data on the clinical characteristics of cataract in children are useful for the proper planning of comprehensive strategies of diagnostic and treatment options.<sup>[4,9]</sup> However, there is a lack

Received: 12-Apr-2022 Accepted: 13-Sep-2022 Revision: 28-Jul-2022 Published: 30-Nov-2022 of studies on the clinical features of cataract in children from the Central Asian region.

The objective of this study was to determine the clinical characteristics of congenital and developmental cataract seen at our tertiary eye care facility over the 10-year study period.

# Methods

The institutional review board approved this retrospective chart review. All procedures conformed to the guidelines of the Declaration of Helsinki. We reviewed the medical records of all patients with congenital/developmental cataract aged 0–18 years who underwent primary cataract surgery from January 1, 2010 to December 31, 2020 at our tertiary eye care facility, which is the only tertiary eye care center providing surgery for children with congenital/developmental cataract in Kazakhstan. In clinical practice in our country, all cataracts in children are classified as congenital (code Q 12.0 in International Classification of Diseases 10), with the exception of secondary forms of lens opacity due to trauma, acquired systemic or ocular pathology. Because it is challenging to distinguish retrospectively between congenital and developmental cataracts, both of these clinical categories

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were combined in the present study and presented as congenital/developmental cataract. Cases of congenital and developmental cataract were identified using the database of the statistics department of our tertiary eye care facility. We excluded cases with missing or incomplete data, as well as cataracts with traumatic, acquired systemic (e.g., diabetes), or acquired ocular etiology (e.g., uveitis). Data on the remaining 942 patients (1311 eyes) were used for the subsequent analyses.

#### **Collection of data**

All the eligible medical charts were carefully reviewed. We recorded the following data from the medical histories of these patients: gender, age at surgery, main presenting complaint (e.g., white pupil and poor vision), laterality, morphologic type of cataract, family history, and concomitant ocular/systemic disorders and syndromes.

#### **Statistical analysis**

The data were analyzed using StatTech v. 2.4.3 (Developer-StatTech LLC, Russia). Quantitative variables were assessed for normal distribution by using the Kolmogorov–Smirnov test. Quantitative variables following non-normal distribution were described using median (Me) and interquartile range (IQR). Categorical data were described with absolute and relative frequencies. A comparison of frequencies in the analysis of multifield contingency tables was performed using Pearson's Chi-square test. One sample Chi-square test was used to measure any statistical difference between two populations on a single categorical characteristic (gender). *P* <0.05 was considered to be statistically significant.

# Results

A total of 1311 eyes of 942 patients (0–18 years old) were included in this study. According to the medical histories,

369 patients underwent surgery on both eyes within the study period (738 eyes). Furthermore, 288 patients with bilateral cataract received surgical treatment on one eye only from January 1, 2010 to December 31, 2020. Of them, 196 patients underwent the operation on the first eye and did not come for the second operation within the study period. Ninety-two patients had surgery on the first eye before January 1, 2010 and underwent operation on the second eye only within the study period.

Table 1 shows the demographic data of the patients with congenital/developmental cataract in different age groups. Males accounted for 546 (58%) of the children. The overall proportion of boys and girls undergoing cataract surgery was approximately equal (P = 0.110). However, in bilateral cases, boys presented with congenital/developmental cataracts more frequently in the following age groups: >6 months to 1 year (P = 0.001), >1 year to 3 years (P = 0.003), and >3 years to 7 years (P = 0.005). The overall proportion of boys in the bilateral group was larger than that of girls (P = 0.028). In unilateral cases, girls presented with cataracts more frequently than boys in the age group of >6 months to 1 year (P = 0.009). In the other age groups, the frequencies of both genders did not differ statistically. The overall proportion of boys and girls in the unilateral group was equal (P = 0.841).

As shown in Table 1, there were more patients with bilateral congenital/developmental cataract than unilateral (69.7% vs. 30.3%, P < 0.001). The proportion of the patients with unilateral and bilateral cataract was stable during the study period (P = 0.344) [Fig. 1].

The median age at surgery for congenital/developmental cataract was 51 months (IQR = 70 months). No patient with





unilateral cataract presented for surgery before 6 months of age [Table 1]. The analysis of the age of the patients with congenital/developmental cataract at recognition and at presentation for surgery is described in a separate report.<sup>[10]</sup> The mentioned study included 897 patients out of 942 because the information regarding the age at the time of recognition was provided in the medical histories of these 897 patients. The overall prevalence of strabismus in our study was 27.2% (356/1311 eyes), and the prevalence of nystagmus was 19.3% (253/1311 eyes). Strabismus was 2.636 times less common in bilateral cataract cases than in the unilateral cataract group (OR = 0.379; 95% CI: 0.288–0.500; P < 0.001). The presence of nystagmus was 17.920 times greater in bilateral cataracts than in unilateral cataract cases (95% CI: 7.316–43.891; P < 0.001).

Table 1: Demographic data (m=month, y=years)							
Category	Age groups						
	≤6 m	>6 m-1 y	>1 y-3 y	>3 y-7 y	>7 y	Total	
Patients n (%)							
Bilateral cataract	16 (100.0)	78 (67.3)	154 (69.1)	227 (71.6)	182 (67.4)	657 (69.7)	
Unilateral cataract	0 (0.0)	38 (32.7)	69 (30.9)	90 (28.4)	88 (32.6)	285 (30.3)	
Total	16 (1.6)	116 (12.4)	223 (23.7)	317 (33.7)	270 (28.7)	942 (100)	
Eves undergoing cataract surgery n (%)							
Bilateral cataract	18 (1.8)	112 (10.9)	246 (24.0)	369 (36.0)	281 (27.4)	1026 (78.3)	
Unilateral cataract	0 (0.0)	38 (13.3)	69 (24.2)	90 (31.6)	88 (30.9)	285 (21.7)	
Total	18 (1.4)	150 (11.4)	315 (24.0)	459 (35.0)	369 (28.1)	1311 (100)	
Gender: hovs/girls n (%)	( )	( )	· · · ·	· · · ·	( )	· · · ·	
Bilateral cataract	8 (50 0)/	52 (66 7)/	100 (64 9)/	145 (63.9)/	97 (53 3)/	402 (61 2)	
Biatoral outardot	8 (50.0)	26 (33.3)	54 (35 1)	82 (36 1)	85 (46 7)	255 (38.8)	
	0(00)/	14 (36 8)/	36 (52 2)/	48 (53 3)/	46 (52 3)/	144 (50.5)	
Unilateral cataract	0(0.0)	24 (63 2)	33 (47.8)	42 (46 7)	42 (47 7)	141 (49.5)	
	8 (50 0)/	66 (56 9)/	136 (61 0)/	193 (60.9)/	143(530)/	546 (58.0)	
Total	8 (50.0)	50 (43.1)	87 (39.0)	124 (39.1)	127 (47.0)	396 (42.0)	
Preexisting pystagmus eves $p(%)$	0 (0010)	00 (1011)	07 (0010)	()	(	000 (1210)	
Bilateral cataract	1 (22 2)	56 (50 0)	84 (34 7)	67 (18 3)	37 (13 1)	248 (24 2)	
Initiateral cataract	4 (22.2)	1 (2.6)	2 (2 7)	1 (1 1)	$\frac{37(13.1)}{1(1.2)}$	240 (24.2) 5 (1.8)	
Total	4 (22.2)	57 (38.0)	2 (2.7) 86 (23 7)	68 (14 9)	38 (10 3)	253 (10.3)	
	4 (22.2)	57 (50.0)	00 (20.7)	00 (14.3)	56 (10.5)	200 (19.0)	
Preexisting strabismus eyes n (%)	F (07 0)	04 (00 4)	70 (00 0)	70 (01 4)			
Bilateral cataract	5 (27.8)	34 (30.4)	79 (32.6)	79 (21.4)	35 (12.5)	232 (22.6)	
	0 (0.0)	16 (42.1)	32 (43.8)	46 (51.1)	30 (34.1)	124 (43.5)	
IOTAI	5 (27.8)	50 (33.3)	111 (35.2)	125 (27.2)	65 (17.6)	356 (27.2)	
Family history of congenital/developmental cataract n (%)							
Bilateral cataract	3 (18.8)	15 (19.2)	26 (16.9)	42 (18.5)	43 (23.6)	129 (19.6)	
Unilateral cataract	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	4 (4.5)	4 (1.4)	
Total	3 (18.8)	15 (12.9)	26 (11.7)	42 (13.2)	47 (17.4)	133 (14.1)	
Morphologic type of cataract (eyes) n (%)							
Total	12 (66.7)	77 (51.3)	166 (52.7)	174 (37.9)	128 (34.7)	557 (42.5)	
Polymorphic (mixed)	0 (0.0)	40 (26.7)	72 (22.9)	134 (29.2)	110 (29.8)	356 (27.2)	
Lamellar	6 (33.3)	25 (16.7)	53 (16.8)	113 (24.6)	98 (26.6)	295 (22.5)	
Posterior subcapsular	0 (0.0)	3 (2.0)	8 (2.5)	21 (4.6)	28 (7.5)	60 (4.6)	
Semi-absorbed	0 (0.0)	1 (0.7)	12 (3.8)	8 (1.7)	0 (0.0)	21 (1.6)	
Posterior polar	0 (0.0)	3 (2.0)	1 (0.3)	2 (0.4)	3 (0.8)	9 (0.7)	
Nuclear	0 (0.0)	1 (0.7)	2 (0.6)	2 (0.4)	1 (0.3)	6 (0.5)	
Anterior polar	0 (0.0)	0 (0.0)	1 (0.3)	3 (0.7)	0 (0.0)	4 (0.3)	
Anterior subcapsular	0 (0.0)	0 (0.0)	0 (0.0)	2 (0.4)	1 (0.3)	3 (0.2)	

### Table 2: The main presenting complaint of the patients according to the age groups (m=month, y=years)

The main presenting complaint <i>n</i> (%)	Age groups						
	≤6 m	>6 m-1 y	>1 y-3 y	>3 y-7 y	>7 y		
White pupil	14 (87.5)	85 (73.3)	153 (68.6)	141 (44.5)	60 (22.2)		
Poor vision	0 (0.0)	17 (14.7)	44 (19.7)	149 (47.0)	202 (74.8)	<0.001	
Strabismus	1 (6.3)	14 (12.1)	16 (7.2)	21 (6.6)	8 (3.0)		
Nystagmus	0 (0.0)	0 (0.0)	2 (0.9)	0 (0.0)	0 (0.0)		
No eye fixation	1 (6.3)	0 (0.0)	8 (3.6)	6 (1.9)	0 (0.0)		

Congenital ocular abnormalities n (%)		Total				
	≤6 m	>6 m-1 y	>1 y-3 y	>3 y-7 y	>7 y	
Persistent fetal vasculature	0 (0.0)	3 (13.0)	3 (16.7)	4 (13.8)	1 (4.0)	11 (11.3)
Microphthalmos	0 (0.0)	3 (13.0)	6 (33.3)	11 (37.9)	6 (24.0)	26 (26.8)
Microcornea	0 (0.0)	7 (30.4)	4 (22.2)	3 (10.3)	4 (16.0)	18 (18.6)
Microphthalmos + microcornea	0 (0.0)	5 (21.7)	0 (0.0)	2 (6.9)	0 (0.0)	7 (7.2)
Microphthalmos + persistent fetal vasculature	0 (0.0)	0 (0.0)	0 (0.0)	1 (3.4)	1 (4.0)	2 (2.1)
Microcornea+persistent fetal vasculature	0 (0.0)	1 (4.4)	0 (0.0)	0 (0.0)	0 (0.0)	1 (1.0)
Microphthalmos + microcornea + persistent fetal vasculature	0 (0.0)	0 (0.0)	2 (11.1)	1 (3.4)	0 (0.0)	3 (3.1)
Congenital uveal coloboma	0 (0.0)	1 (4.4)	1 (5.6)	0 (0.0)	1 (4.0)	3 (3.1)
Microcornea + congenital uveal coloboma	1 (50.0)	0 (0.0)	0 (0.0)	0 (0.0)	1 (4.0)	2 (2.1)
Microphthalmos + microcornea + congenital uveal coloboma	0 (0.0)	1 (4.4)	0 (0.0)	0 (0.0)	1 (4.0)	2 (2.1)
Aniridia	0 (0.0)	0 (0.0)	1 (5.6)	3 (10.3)	7 (28.0)	11 (11.3)
Lens coloboma	0 (0.0)	0 (0.0)	0 (0.0)	1 (3.4)	1 (4.0)	2 (2.1)
Congenital optic nerve coloboma	0 (0.0)	0 (0.0)	1 (5.6)	0 (0.0)	0 (0.0)	1 (1.0)
Microcornea + congenital optic nerve coloboma	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	1 (4.0)	1 (1.0)
Persistent pupillary membrane	0 (0.0)	0 (0.0)	0 (0.0)	1 (3.4)	0 (0.0)	1 (1.0)
Congenital glaucoma	0 (0.0)	1 (4.4)	0 (0.0)	1 (3.4)	0 (0.0)	2 (2.1)
Microphthalmos + microcornea + congenital glaucoma	0 (0.0)	1 (4.4)	0 (0.0)	0 (0.0)	0 (0.0)	1 (1.0)
Congenital ectopic pupil	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	1 (4.0)	1 (1.0)
Megalocornea	1 (50.0)	0 (0.0)	0 (0.0)	1 (3.4)	0 (0.0)	2 (2.1)
Total	2 (100.0)	23 (100.0)	18 (100.0)	29 (100.0)	25 (100.0)	97 (100.0)

#### Table 3: Congenital ocular abnormalities associated with congenital/developmental cataracts (m=month, y=years)

#### Table 4: Systemic abnormalities associated with congenital/developmental cataracts (m=month, y=years)

Systemic abnormalities n (%)	Age groups					Total
	≤6 m	>6 m-1 y	>1 y-3 y	>3 y-7 y	>7 y	
Congenital heart disease	1 (100.0)	20 (87.0)	40 (93.0)	49 (84.5)	17 (70.8)	127 (85.2)
Congenital hydrocephalus	0 (0.0)	1 (4.3)	0 (0.0)	3 (5.2)	2 (8.3)	6 (4.0)
Charcot-Marie-Tooth disease	0 (0.0)	0 (0.0)	0 (0.0)	1 (1.7)	1 (4.2)	2 (1.3)
Microcephaly	0 (0.0)	1 (4.3)	1 (2.3)	0 (0.0)	0 (0.0)	2 (1.3)
Epilepsy	0 (0.0)	0 (0.0)	2 (4.7)	3 (5.2)	4 (16.7)	9 (6.0)
Congenital hydrocephalus + Congenital heart disease	0 (0.0)	1 (4.3)	0 (0.0)	1 (1.7)	0 (0.0)	2 (1.3)
Epilepsy + Congenital heart disease Total	0 (0.0) 1 (100.0)	0 (0.0) 23 (100.0)	0 (0.0) 43 (100.0)	1 (1.7) 58 (100.0)	0 (0.0) 24 (100.0)	1 (0.7) 149 (100.0)

Table 5: Syndromes associated with congenital/developmental cataracts (m=month, y=years)

Syndromes <i>n</i> (%)	yndromes n (%) Age groups					Total
	≤6 m	>6 m-1 y	>1 y-3 y	>3 y-7 y	>7 y	
Down syndrome	1 (100.0)	8 (88.9)	17 (100.0)	11 (91.7)	8 (80.0)	45 (91.8)
Marfan's syndrome	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	2 (20.0)	2 (4.1)
Lowe's syndrome	0 (0.0)	1 (11.1)	0 (0.0)	0 (0.0)	0 (0.0)	1 (2.0)
Hallerman-Streiff syndrome	0 (0.0)	0 (0.0)	0 (0.0)	1 (8.3)	0 (0.0)	1 (2.0)
Total	1 (100.0)	9 (100.0)	17 (100.0)	12 (100.0)	10 (100.0)	49 (100.0)

In total, 133 children (14.1%) had a positive family history of congenital/developmental cataract. Positive family history was 17.163 times more common in bilateral cases compared with unilateral (95% CI: 6.279–46.914).

The most common morphologic type of congenital/ developmental cataract was total cataract, accounting for 557 (42.5%) eyes of the patients. The morphologic types of cataract in different age groups are summarized in Table 1. Total cataract prevailed in all age groups.

White pupil was the main presenting complaint in 453 (48.1%) children, while poor vision was detected first in 412 (43.7%) children. Table 2 represents the main presenting

complaint according to the different age groups. Statistically significant differences were found between the following age groups: >1 year to 3 years and >3 years to 7 years (P < 0.001); >1 year to 3 years and >7 years (P < 0.001).

Additional ocular dysmorphology was found in 97 (10.3%) patients, which was less common in bilateral cataract cases than in unilateral ones (OR = 0.504; 95% CI: 0.329–0.773; P < 0.001). Table 3 represents congenital ocular abnormalities accompanying congenital/developmental cataract cases in different age groups. The most frequently seen ocular abnormality was microphthalmos (n = 26).

Concomitant systemic disease (congenital central nervous system disease or/and anomaly of the cardiovascular system) was found in 149 (15.8%) patients. Table 4 shows concomitant systemic abnormalities according to the different age groups of the patients. Congenital heart disease (n = 127) and epilepsy (n = 9) were the most common among systemic pathology accompanying cases of congenital/developmental cataract.

Of the syndromes associated with cataracts, Down syndrome compromised the majority of cases [Table 5]. The median age at surgery was 34 months (IQR = 62 months). Thirty-nine patients (86.7%) had bilateral cataracts. Almost half of the patients (44.9%) had dense cataract.

# Discussion

The present study reports data regarding the clinical characteristics of congenital and developmental cataracts in Kazakhstan over a 10-year period. We found that the overall distribution of both genders in this study was fairly equal, which is similar to the values reported by Fakhoury *et al*.<sup>[11]</sup> and Magnusson *et al*.<sup>[12]</sup> However, in bilateral cases, boys presented for surgery more frequently than girls. In unilateral cases, the overall proportion of boys and girls was approximately equal, although the proportion of girls was statistically significantly higher in the age group of >6 months to 1 year. This finding suggests that girls with bilateral cataracts undergo surgical treatment less commonly than boys in our population.

The majority of congenital/developmental cataract cases in our study population were bilateral (69.7%), whereas unilateral accounted for 30.3%, which was consistent with the results reported in previous studies, such as 71% of bilateral cataract reported by Fakhoury et al. in France<sup>[11]</sup> and 65.8% of bilateral cataract reported by Nagamoto et al. in Japan.<sup>[13]</sup> However, data from a web-based surgical register of pediatric cataract representing Sweden and Denmark, reported by Magnusson et al.,<sup>[12]</sup> showed equal distribution of unilateral and bilateral cases, the vast majority (95,9%) of them being congenital cataract. This difference in the distribution of laterality of congenital/developmental cataract suggests that bilateral cases undergo surgery more frequently than unilateral cases, probably because bilateral cataract affects the overall child's development and leads to severe visual impairment and blindness if left untreated.

According to the main presenting complaint, over half of the children with congenital and developmental cataract (53.3% of unilateral cases and 51.3% of bilateral cases) suffered from leucocoria and 39.2% had poor vision. A Japanese study reported a lower percentage of the presence of white pupil (32.7% of unilateral cataract cases and 35.7% of bilateral cases).<sup>[14]</sup> Leucocoria was the most common complaint in children up to 3 years of age. Older patients presented for surgery with poor vision mostly. The high rate of white pupil as the main presented complaint in children younger than 3 years from birth may be explained by the fact that preverbal children cannot complain about low vision. Consequently, they present for surgery after the detection of the visible lens opacity by the parents or health professionals. According to our finding that the majority of children under 3 years present for surgery with leucocoria, we suggest that they could experience delay in the surgical treatment of congenital/developmental cataract as it is well known that visually significant congenital/developmental cataract should be removed early on to prevent irreversible deprivation amblyopia and blindness.

In our series, strabismus and nystagmus were found in 27.2% and 19.3% of the eyes with congenital/developmental cataract, respectively. The incidence of strabismus was statistically higher in unilateral cataract cases, while nystagmus was more common in the cases of bilateral cataract. Similarly, Lim *et al.*<sup>[4]</sup> also reported a higher incidence of strabismus in unilateral cataract cases. A Chinese study reported that strabismus and nystagmus were seen in 20.6% and 11.9% of patients, respectively.<sup>[9]</sup> It was established that nystagmus was seen in bilateral cataract more commonly due to the more severe vision impairment compared to unilateral cataract. Both strabismus and nystagmus are known to be signs of severe vision deprivation early in childhood, indicating unfavorable visual outcomes of cataract patients with these disorders.<sup>[9]</sup>

In this study, 133 children from 942 (14.1%) had a positive family history of congenital/developmental cataract. Similarly, Lim *et al.*<sup>[4]</sup> found the proportion of inherited cataracts to be 11.7%. A Danish epidemiologic study of 1027 cases of congenital/infantile cataract reported a higher proportion of hereditary cataract (23%),<sup>[15]</sup> which was similar to the 18.8% reported by Wirth *et al.*<sup>[16]</sup> in Australia. This difference might be explained by the different prevalence of hereditary risk factors among populations.

We found that in almost half of the cases of congenital/ developmental cataract, the morphology of lens opacity was total cataract. In a systematic review and meta-analysis presented by Wu *et al.*,<sup>[2]</sup> the most common types of congenital cataract globally were total (31.2%), nuclear (27.2%), and posterior subcapsular (26.8%). A Chinese study found that congenital cataract was total in 84.4% of pediatric patients with cataract.<sup>[17]</sup> In contrast, Holmes *et al.*<sup>[18]</sup> found that infantile cataract was total in two (13.3%) cases in a defined US population. This finding suggests the late detection of cataract in developing countries because many types of cataract slowly become total in untreated cases.<sup>[2]</sup>

Altogether, 10.3% of all patients with congenital/ developmental cataract in this study had other associated ocular abnormalities, the most common being microphthalmos. This is similar to the findings reported by SanGiovanni *et al.*<sup>[19]</sup> Magnusson *et al.*<sup>[12]</sup> reported persistent fetal vasculature to be the most common coexisting ocular abnormality in the study of operated childhood cataract in Sweden and Denmark. Around one-sixth of the children had systemic disorders (congenital central nervous system disease or anomaly of the cardiovascular system). However, Zhu *et al.*<sup>[9]</sup> reported that only 3.3% of 520 congenital/developmental cataract children had concomitant systemic disorders, of which congenital heart disease was the most frequent system abnormality. Fakhoury *et al.*<sup>[11]</sup> found that 22% of 59 congenital cataract cases were associated with systemic abnormalities.

Down syndrome accounted for the majority of cases associated with syndromes in our study. A Danish study reported that almost 50% of 60 syndrome cases were represented by trisomy 21.<sup>[15]</sup> Similarly, a Canadian study found that Down syndrome accounted for almost a third of syndrome-associated cataracts.<sup>[4]</sup> It has been reported that infants with trisomy 21 should undergo early ophthalmic examination to identify visually significant cataract.<sup>[4,20]</sup>

The strength of our study is presented by the fact that it covers almost all of the surgical cases of congenital/ developmental cataract over a 10-year period in Kazakhstan. However, it has several limitations. The limitations of our study are presented by its retrospective nature. Our report was restricted by the data provided in the medical histories of these patients. First, unfortunately, valuable information regarding possible etiology was missing in the majority of the medical histories. The information on prenatal (history of infections, especially TORCH), perinatal, and infant-based factors (delayed cry and hypoxic injury at birth) was missing, and maternal factors such as hypertension and drug use were not described. The information regarding the birth term was provided in the medical histories of only 195 patients, 163 (83.6%) of them were full birth term and 32 (16.4%) were preterm birth. Because it is not enough to be representative, we decided not to describe this finding in the manuscript. Second, there was no mention of genetic testing in the cases of inherited cataract. Third, there is a possible underestimation of some systemic disorders and syndromes associated with cataracts as there might be cases of missed diagnosis.

# Conclusion

In conclusion, we have reviewed the clinical features of congenital and developmental cataract in Kazakhstan. High prevalence of total cataracts as well as frequent association with strabismus and nystagmus are likely to be the consequences of delayed presentation for surgery in our population. It is vital to investigate the reasons and sociodemographic factors affecting the timing of presentation for surgery in cases of congenital and developmental cataract to develop strategies to overcome these barriers.

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# **Conflicts of interest**

There are no conflicts of interest.

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