

Clinical Profile of Pediatric Cataracts in a Tertiary Eye Care Center in Mongolia

Shamsiya Murat^{1,2}, Erdenebileg Nasantogtokh², Gombojav Davaa³, Ganbaatar Erdenetuya⁴, Davaatseren Uranchimeg¹

¹Department of Ophthalmology, School of Medicine, Mongolian National University of Medical Sciences, Ulaanbaatar, Mongolia; ²Department of Ophthalmology, National Center for Maternal and Child Health of Mongolia, Ulaanbaatar, Mongolia; ³Department of Epidemiology and Biostatistics, School of Public Health, Mongolian National University of Medical Sciences, Ulaanbaatar, Mongolia; ⁴Department of Endocrinology, School of Medicine, Mongolian National University of Medical Sciences, Ulaanbaatar, Mongolia

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Corresponding Author

Shamsiya Murat, MD
Department of Ophthalmology,
School of Medicine, Mongolian
National University of Medical
Sciences, Ulaanbaatar 14210,
Mongolia
Fax: +976 9909-9348,
E-mail: muratshamsiya@gmail.com

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Objective: To describe the clinical characteristics of pediatric cataracts in children undergoing surgery in our tertiary care center in Mongolia. **Methods:** Patients aged ≤ 18 years who underwent pediatric cataract surgery over a three-year period at our pediatric hospital in Mongolia were included. The data collected included: demographic information, family history, laterality, morphology, etiology, and coexistent ocular and systemic disease, age at diagnosis. **Results:** Over the 3-year study period, 118 children underwent surgery for pediatric cataracts. Sixty participants (53.4 %) had congenital and 92 (78 %) had bilateral cataracts. The most common etiology of pediatric cataracts was idiopathic (34 cases, 28.8 %). Lamellar cataracts were the most commonly observed pediatric cataract (23.8 %). Microphthalmos/microcornea was the most frequent ocular malformation seen in pediatric cataracts. **Conclusions:** The most common etiology of congenital cataracts was idiopathic, whereas developmental cataracts were most commonly hereditary. In almost all patients, the main morphology of congenital cataracts was nuclear, and the main morphology of developmental cataracts was lamellar. In congenital and developmental cataracts, the main associated ocular disorder was microphthalmos/microcornea. The most common associated systemic disorder in developmental cataracts was Type 1 diabetes and in congenital cataracts was Down syndrome.

Keywords: Etiology, Congenital, Cataract, Morphology, Developmental

Introduction

Worldwide, the prevalence of pediatric cataracts is 0.32 - 22.9 per 10 000, and congenital cataracts is 0.63 - 9.74 per 10,000, depending on the country [1]. Over 90 % of pediatric cataracts are congenital or developmental [1]. The prevalence of blindness

(best-corrected visual acuity less than 3 / 60) from cataracts in children in developing countries is probably 1 to 4 / 10,000 and approximately 0.1 to 0.4 / 10,000 children in industrialized countries. This difference reflects the better prognosis for vision obtained when children are diagnosed early and managed by pediatric ophthalmologists. Globally, an estimated 200,000

children are blind from bilateral cataracts. The cause of pediatric cataracts is often unknown [2]. According to Bulgan's study [3], 34 % of severe visual impairments of children in both eyes are caused by lens disorders in Mongolia.

A recent study demonstrated that identifying the clinical profile and analyzing the relationships among the different variables are clinically important for earlier diagnosis and early treatment of pediatric cataracts to prevent amblyopia in children [4].

The most common morphologies of pediatric cataracts include nuclear, lamellar and total [4 - 6]. Morphology varies according to eye laterality and cause. An inherited cataract is usually lamellar and bilateral. The visual outcome depends on the morphology of the cataract. For example, if the cataract is total, the visual prognosis is poor [7]. In the developed world, the common etiology of congenital/ infantile cataracts was idiopathic and then hereditary, and most cases were bilateral [8 - 10]. In the study in Toronto, with systemic disease, more than half of cataracts were steroid-induced, and the next most common cause was juvenile idiopathic arthritis. According to TORCH infection associated with cataracts, rubella still accounted for most cases, with the next most common being cytomegalovirus infection. The most frequently seen ocular abnormalities in unilateral cataracts were persistent fetal vasculature, retinochoroidal coloboma [11], and in bilateral was microphthalmos [9]. The most common systemic disorders were congenital heart disease and cleft lip and palate [11]. In a recent study from Japan, total and lamellar cataracts and nystagmus were significantly greater among bilateral cases [4]. In contrast, posterior subcapsular/polar cataracts and strabismus were more frequent in unilateral cases.

Severe childhood visual impairments are caused by lens disorders in Mongolia, but information on the prevalence, etiology, morphology, and comorbidity of pediatric cataracts is lacking [3]. In this study, we defined the clinical profiles of a series of pediatric cataracts and captured the age of onset and laterality.

Materials and Methods

Study design

The National Center for Maternal and Child Health is Mongolia's only pediatric cataract surgery center. Therefore, our center

sample is likely to be nationally representative. We assumed the sample size was a proportion in an infinite population. We chose a confidence interval of 95 %, a margin of error of 5 %, and a predictive value of 0.5 for this calculation. We performed an analytical cross-sectional and comparative analysis of cataracts morphology, etiology, comorbidity and demographic variables with age at onset and laterality of 118 patients. Records of pediatric patients who had undergone surgery at the National Center for Maternal and Child Health for congenital or developmental cataracts from January 1, 2018, to December 31, 2020, were evaluated.

Data collection

We collected the following information from the patients' medical records: demographic variables, visual acuity, family history, cataract morphology, cataract etiology, coexistent ocular anomalies, associated systemic disease or syndromes, and age at onset, initial hospital visit and surgery. Visual acuity was evaluated using the Snellen chart and Lea symbol. In patients who could not perform visual acuity testing, we assessed fixation and following pattern. We diagnosed familial cataracts using family member screening. Children who did not cooperate were examined under general anesthesia with an operating microscope. The morphology of each cataract was determined using a slit lamp and operating microscope. Complete ophthalmologic examination included the presence of nystagmus, strabismus, anterior segment findings, measurement of horizontal corneal diameter, intraocular pressure measurement, keratometry, and fundus evaluation with indirect ophthalmoscope after pupil dilation, axial length measurement and ultrasound B scan done in patients with a total cataract. We checked toxoplasmosis, other, rubella, cytomegalovirus, and herpes infections titers, and plasma calcium and phosphorus, and fasting blood sugar levels. When necessary, a pediatrician was consulted.

Study case definition

Congenital and developmental cataracts are categorized separately in the standard disease classification system. Cataracts were classified as congenital (ICD - Q12) if recognized at or within 1 year of birth or if accompanied by nystagmus, nuclear, polar, lenticonus, or persistent fetal vasculature cataract. They were classified as developmental (ICD H26.0) if zonular and onset was after 1 year of age [7, 12].

Classification of cataract

Cataracts were classified according to the most likely etiology: (1) idiopathic, (2) inherited, (3) ocular disease-associated, (4) syndrome or systemic disease-associated, (5) metabolic, (6) trauma- or laser treatment-associated, (7) drug-induced, (8) Rubella- or TORCH infections-related and (9) prematurity-associated [13].

Cataract morphology included the following types: nuclear, cortical, lamellar, sutural, anterior and posterior subcapsular, lenticonus, persistent fetal vasculature, total, anterior and posterior polar, and combined [13, 14]. After the etiological subdivision, the patients were further divided into clinically defined groups of (1) isolated cataracts, (2) no systemic disorder associated with additional ocular dysmorphology, and (3) cataracts with systemic anomalies regardless of additional ocular dysmorphology [8, 14]. History details, including presenting complaints, were noted from the records. The cases were distributed according to unilateralism or bilateralism. If family history was positive, the cataract was considered to be hereditary.

Inclusion criteria

Children aged 0 - 18 years old.

Exclusion criteria

Children with post-traumatic cataracts, lens dislocation, or previous surgery in the eye were excluded from the study.

Statistical analysis

Continuous variables (age) were presented by their central tendency and variability. Continuous variables included age at onset, first hospital visit and surgery. All continuous variables were normally distributed. The student t-test was used to determine the mean age difference between congenital and developmental cataracts at their age of diagnosis, initial hospital visit and surgery. Descriptive analysis of category variables was calculated in frequency and percentages. Category variable association was found by applying Pearson's chi-square and Fisher's exact test. The chi-square test was only used when the number of participants per cell was sufficient. We used a chi-square test to compare the cataract type (development and congenital) by location, gender, and laterality eye. The confidence interval for the analysis was calculated as 95 % and

was considered statistically significant if the p-value was < 0.05 . Some factors affecting congenital cataracts were assessed by binary logistic regression analysis. We used SPSS version 26.0 to perform the analyses.

Ethical statements

The study was conducted in accordance with the Declaration of Helsinki, and the Research Ethical Committee of the Research Ethical Committee of the Mongolian National University of Medical Sciences approved the study protocol (No.2019/3-02).

Results

In total, 118 individuals under 18 years of age were seen for pediatric cataracts from January 2018 to December 2021 at the Mongolian National Center for Maternal and Child Health. Sixty-five individuals (55 %) were male; 55 (47 %) were from rural areas.

Ninety-two (78 %) had bilateral cataracts, and 26 (22 %) had unilateral. Sixty-three (53 %) cases were congenital and 55 (47 %) were developmental cataract. Among congenital cataract cases, 73 % were developmental cataracts and 84 % were bilateral ($p = 0.036$). Compared to developmental cataracts, congenital cataracts presented much earlier (7.6 ± 1.3 months vs. 59 ± 15.6 months, $p < 0.000$), and consequently had their first visit to the hospital earlier (17.1 ± 8.7 vs. 78 ± 15.4 months, $p < 0.000$), and had surgery earlier (33.4 ± 8.9 months vs. 102 ± 21.8 months, $p < 0.000$) (Table 1).

The most common etiology of pediatric cataracts in our study was idiopathic (41 cases, 35 %), followed by hereditary (31 cases, 26 %). The etiologies of congenital and developmental cataracts were statistically significantly different (Table 2).

The etiology was usually hereditary, followed by idiopathic and associated systemic disorders in bilateral cases. In unilateral cases, the most common etiology was idiopathic, followed by those with an associated ocular disorder. Regarding drug-induced cataracts, one child with aplastic anemia had cataracts due to prolonged use of corticosteroids. Another developed a cataract due to corticosteroids used for unilateral eyelid and facial hemangiomas.

Congenital and developmental cataract morphologies were different in the study population (Table 3). In unilateral cases, the most common morphology was persistent fetal vasculature

Table 1. Demographic characteristics.

Variables	Congenital (n = 63)	Developmental (n = 55)	Total (n = 118)	p-value
	Mean ± SD	Mean ± SD	Mean ± SD	
Age at recognition	7.58 ± 1.3	59.6 ± 15.6	21.6 ± 9.6	0.000 [†]
Age at presentation	17.07 ± 8.65	78.8 ± 15.4	46.3 ± 11.3	0.000 [†]
Age at surgery	33.4 ± 8.9	102.2 ± 21.8	65.2 ± 12.4	0.000 [†]
	N (%)	N (%)	N (%)	
Gender				
Male	36 (57.1)	29 (52.7)	65 (55.1)	0.842 [†]
Female	27 (42.9)	26 (47.3)	53 (44.9)	
Location				
Urban	31 (49.2)	31 (56.4)	62 (52.5)	0.031 [†]
Rural	32 (50.8)	24 (43.6)	56 (47.5)	
Laterality				
Unilateral	17 (27.0)	9 (16.4)	26 (22.0)	0.037 [†]
Bilateral	46 (73.0)	46 (83.6)	92 (78.0)	
Total	63 (53.4)	55 (46.6)	118 (100.0)	-

[†]Student's T-test, [†]Pearson's chi-square test

Table 2. Etiology of pediatric cataracts by age at onset.

Etiology	Congenital (n = 63)	Developmental (n = 55)	p-value
	N (%)	N (%)	
Idiopathic [†]			
No	38 (60.3)	39 (70.9)	0.000
Yes	25 (39.6)	16 (29.1)	
Hereditary [†]			
No	55 (87.3)	32 (58.2)	0.000
Yes	8 (12.7)	23 (41.8)	
Associated ocular disorder [†]			
No	51 (80.9)	53 (96.4)	0.000
Yes	12 (19.1)	2 (3.6)	
Associated systemic disorder [†]			
No	50 (79.4)	49 (89.1)	0.000
Yes	13 (20.6)	6 (10.9)	
Metabolic			
No	63 (100.0)	49 (89.1)	
Yes	-	6 (10.9)	
Drug-induced			
No	63 (100.0)	53 (96.4)	
Yes	-	2 (3.6)	
Premature [†]			
No	58 (92.1)	55 (100.0)	
Yes	5 (7.9)	-	

[†]Pearson's chi-square test, [†]Fisher's exact test

Table 3. Morphology of cataracts and onset time.

Morphology of cataract	Congenital (n = 63)	Developmental (n = 55)
	N (%)	N (%)
Total	15 (23.8)	9 (16.4)
Nuclear	14 (22.2)	-
Lamellar	7 (11.1)	22 (40.0)
Membranous	6 (9.5)	-
Mixed	2 (3.2)	8 (14.5)
Polar	-	4 (7.3)
Cortical	3 (4.8)	3 (5.5)
Persistent fetal vasculature	7 (11.1)	-
Lenticulus	5 (7.9)	-
Sutural	2 (3.2)	-
Different bilateral	2 (3.2)	9 (16.4)

Table 4. Patients' comorbidity, age of onset of cataract and laterality.

Co-morbidities	Congenital n = 63 N (%)	Developmental n = 55 N (%)	p – value	Unilateral n = 26 N (%)	Bilateral n = 92 N (%)	p-value [†]
Ocular disorders						
Microphthalmos						
Yes	8 (12.7)	2 (3.6)	0.000	-	10 (10.9)	
No	55 (87.3)	53 (96.4)		26 (100.0)	82 (89.1)	
Persistent fetal vasculature						
Yes	7 (11.1)	-		7 (26.9)	-	
No	56 (88.9)	55 (100.0)		19	92 (100.0)	
Leber'amaurosis and keratopathy						
Yes	-	2 (3.6)		-	2 (2.2)	
No	63 (100.0)	53 (96.4)		26 (100.0)	90 (97.8)	
Uveitis and secondary glaucoma						
Yes	1 (1.6)	2 (3.6)	0.000	1 (3.8)	2 (2.2)	0.000
No	62 (98.4)	53 (96.4)		25 (96.2)	90 (97.8)	
Congenital glaucoma						
Yes	1 (1.6)	-		-	1 (1.1)	
No	62 (98.4)	55 (100.0)		26 (100.0)	91 (98.9)	
Atrophy optic nerve						
Yes	1 (1.6)	-		-	1 (1.1)	
No	62 (98.4)	55 (100.0)		26 (100.0)	91 (98.9)	
Ptosis						
Yes	1 (1.6)	-		1(3.8)	-	
No	62 (98.4)	55 (100.0)		25 (96.2)	92 (100.0)	
Systematic disease						
Neurological						
Yes	5 (7.9)	1 (1.8)	0.000	-	6 (6.5)	
No	58 (92.1)	54 (98.2)		26 (100.0)	86 (93.5)	

Continued

Down syndrome					
Yes	8 (12.7)	3 (5.5)	0.000	-	11 (11.9)
No	55 (87.3)	52 (94.5)		26 (100.0)	81 (88.1)
Metabolic					
Yes	-	6 (10.9)		-	6 (6.5)
No	63 (100.0)	49 (89.1)		26 (100.0)	86 (93.5)
Aplastic anemia					
Yes	-	1 (1.8)		-	1 (1.1)
No	63 (100.0)	54 (98.2)		26 (100.0)	91 (98.9)
Juvenile rheumatoid arthritis					
Yes	-	2 (3.6)		-	2 (2.2)
No	63 (100.0)	53 (96.4)		26 (100.0)	90 (97.8)
Other disease					
Yes	1 (1.6)	3 (5.1)	0.000	-	4 (4.5)
No	62 (98.4)	52 (94.5)		26 (100.0)	88 (95.6)

† - Fisher's exact test

Table 5. Multiple logistic regression analysis to determine the risk factors for bilateral cataracts.

Variables	OR	95% CI		p-value
		Lower	Upper	
Cataract type				
Congenital	Reference			
Developmental	1.89	0.76	4.67	0.169
Location				
Rural	Reference			
Urban	1.21	0.12	1.32	0.381
Age				
< 1 year	Reference			
1 – 6 years	1.24			0.127
> 6 years	1.12	0.97	1.36	
Gender				
Female	Reference			
Male	1.4	0.23	1.46	0.241

Reference group, OR - Odds ratio, CI – Confidence interval

(27 %, $p = 0.041$), lamellar for bilateral cases (30 %, $p = 0.002$) and total (24 %, $p = 0.044$). No cases of bilateral persistent fetal vasculature and lenticonus were recorded.

Microphthalmos/microcornea (8 %, $n = 10$) and persistent fetal vasculature (6 %, $n = 7$) were the most common comorbidities in pediatric patients with cataracts. In addition, 9 % ($n = 11$) of all cases were diagnosed with Down syndrome. Lebers' amaurosis was associated with keratoglobus and

keratoconus.

Congenital cataract was more commonly associated with neurological disorders, including microcephaly, Walker Dandy syndrome, and epilepsy. The most common metabolic disease was type 1 diabetes. Heart defects did not occur alone and, in all cases, were associated with Down syndrome (Table 4). None of the risk factors for bilateral cataracts in bilateral cataracts were significant (Table 5). Age and male gender tended to increase

Table 6. Multiple logistic regression to determine the risk factors in developmental cataracts.

Variables	OR	95% CI		p-value
		Lower	Upper	
Location				
Rural	Reference			
Urban	1.33	0.65	2.76	0.438
Age				
< 1 year	Reference			
1 – 6 years	1.24			
> 6 years	1.12	0.97	1.36	0.127
Gender				
Female	Reference			0.241
Male	1.4	0.23	1.46	0.241

Reference group, OR - Odds ratio, CI – Confidence interval

the risk of a developmental cataract (Table 6).

Discussion

Located in the capital city of Mongolia, the National Center for Maternal and Child Health provides tertiary health care for adults and children in 21 provinces. Our study's greatest strength is that we have described and compared the etiology, co-morbidities, and diagnosed age of pediatric developmental and congenital cataracts in the central Asian population.

We classified cataracts as congenital if the patient had a nuclear, polar, persistent fetal vasculature or lenticonus cataract that occurred before 1 year of age based on parents' reports and if the child had nystagmus. We classified a cataract as developmental if the patient had lamellar morphology that developed after 1 year of age [7, 12, 15, 16]. Scott et al. noted that many of these cataracts are congenital; in some cases, it can be difficult to establish the age of onset with certainty [8, 17]. In some studies, up to 3 months of age [18, 19] was considered congenital, while in other studies, one year of age was the cut-off [15, 16]. Researchers note that time of onset has a strong influence on final visual acuity [18]. Laterality is very important in pediatric cataracts because it causes unilateral low vision. In our study, unilateral cataracts occurred in 22 % of patients, and bilateral cataracts occurred in 78 %, similar to reports from other countries [4, 9, 20 - 22]. Regarding gender, 55 % were male in our study, similar to other countries [7, 9, 21, 23].

In our study, 27 % of bilateral cataracts and 47 % of unilateral cataracts were idiopathic. In a study from Japan [4] and one

from Denmark [8], 36 – 50 % of congenital cataracts and 68 – 87 % of unilateral cataracts were idiopathic, respectively. These percentages were higher than in our study. Hereditary cataracts occurred in 26 % of patients in our study. In comparison, 33 % had bilateral cataracts, and 4 % had unilateral, similar to reports from Denmark [8] and Britain [9], but slightly higher than the 19 % reported in a study in Australia [10].

In our study, bilateral congenital cataracts were more likely associated with systemic disorders (16 %). This was similar to rates of 16 % and 17 %, respectively, reported in Australia and China and lower than the rate of 21% reported in Britain [10, 24, 25]. Down syndrome was the most common chromosomal disorder associated with cataract [4, 8] syndrome in our study (9 %), similar to reports of 11 % in Japan [4], 5 % in Australia [10], 1 % in China [23], and 9 % in a British [9] study. Other disorders such as deafness and muteness occurred in 2 %, similar to a study from China [23]. Type 1 diabetes occurred in 5 % of patients in our study. According to the study by Wirth [10], diabetic cataracts occurred in only 0.8 %, less than in our study.

In our study, 13 % of individuals with hereditary cataracts also had microcornea/microphthalmos, similar to other studies [8, 14]. However, hereditary cataracts are more likely to occur in isolation [8]. When viewed in conjunction with eye disorders, 42 % of unilateral cataracts in our study were associated with another ocular disorder. This was similar to reports from other countries. For example, in a study from Britain, 52 % were associated with an ocular disorder [25]. In a survey from Rahi, 51 % had an associated ocular disorder [9]. In a study by Haargaard, 66 % had an ocular disorder [8]. These studies

indicate that unilateral cataracts were more often associated with ocular disorders than bilateral cataracts. The predominant ocular pathology in our study was persistent fetal vasculature, which accounted for 27 %, respectively, similar to that of a Japanese study. In fact, many studies [8 - 10, 18, 26] have reported persistent fetal vasculature as the most common ocular disorder associated with unilateral cataracts. In our study, 17 % of bilateral cataracts were associated with ocular disorders, compared with 21 % in a Chinese study [26] and 29 % in Halbasic's study [27]. In our study, 29 % of congenital cataracts were associated with an ocular disorder, similar to 23 % reported by Wu [24] but higher than 14 % reported by Rahi [9] and 3 % reported by Dohvoma [28]. We found microcornea/microphthalmos was the most common cause of congenital (13 %) and inherited cataracts (13 %) compared with 11 % [20] and 26 % of cases in other countries [23] and 2 % in China [26].

According to Nadeem [29] and Viola [28], nuclear cataracts accounted for 4 % and 11 % of bilateral cataracts, respectively, whereas we found a higher proportion of nuclear cataracts (23 %) in bilateral cases. We found a lower proportion of lamellar cataracts (11 %) compared with Jain [14] and Angra's [5] (33 % and 52 %, respectively). In the Dohvoma study, total cataracts were present in 54 % of bilateral cataract cases, but we found a low proportion of total cataracts (24 %) [28]. In a study by Epring, only 2 % of subjects had different morphology in each eye compared with our study, in which we found a higher rate. However, the morphology of the two eyes did not differ in congenital cataracts [22]. When we noted different morphologies in bilateral cataracts, usually one eye was lamellar, and the other eye was total, suggesting the lamellar cataract was progressing.

One form of developmental cataract is traumatic etiology. According to a study in Mongolia, traumatic cataract was more common in boys, while no significant gender difference was identified in other types of cataract [29, 30]. According to a study in Niger, children with cataracts were recognized at 13.5 months and diagnosed at 48 months [31]. We found the cataract was first recognized relatively late at a mean age of 21.8 months, but the diagnosis was made at 46 months. In a study in India, children with congenital cataracts underwent surgery at a mean age of 48.2 months, 15 months later than our study, and children with developmental cataract surgery underwent surgery at a mean age of 99.7 months, 2 months earlier than in our series [10]. According to a Chinese study, children with congenital cataracts

underwent surgery at an average age of 27.6 months, 6 months earlier than our study [26].

There are a few limitations to our study. The study was retrospective, and we did not perform genetic tests for idiopathic and hereditary causes. In the future, we plan to do more genetic and endocrinological analyses to determine the etiology of pediatric cataracts. We are also planning to study the results of surgical treatment in relation to the etiology and morphology.

Conclusions

In this case series, the most common etiology of congenital cataracts was idiopathic, while the etiology of developmental cataracts was hereditary. In terms of morphology, the morphology of congenital cataracts was nuclear and was commonly associated with Down syndrome. Regarding developmental cataracts, the morphology was lamellar and was associated with Type 1 diabetes. Moreover, both for congenital and developmental cataracts, the most common associated ocular disorder was microphthalmos/microcornea. Our research continues, and we have taken the first step toward the early detection of congenital cataracts by conducting the red reflex test in our hospital and the maternity wards of 6 local hospitals.

Conflict of interest

The authors declared no conflicts of interest.

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