Original Article

Ten-year Etiologic Review of Chinese Children Hospitalized for Pediatric Cataracts

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Abstract

Purpose: Our aim was to obtain a better understanding of the etiologies and characteristics of pediatric cataracts treated at a single facility in China.

Methods: Medical records accrued over a 10-year period (from August, 2003 to July, 2013) at Shengjing Hospital of China Medical University were reviewed retrospectively, identifying all patients treated for various subtypes of pediatric cataract. A database with 367 subjects under 14 years of age (598, including second-round surgeries) was generated.

Results: Of this cohort (n=367; males: 232, 63.2%; females: 135, 36.8%), 200 patients (54.5%) had bilateral cataracts, and 258 (70.3%) were under 3 years of age. In all age groups and in all subtypes of pediatric cataract, males were most commonly affected. Congenital cataract was the most prevalent subtype, accounting for 296 patients (80.7%). Most congenital cataracts were associated with other ocular or systemic abnormalities; and in 48 patients (16.22%), they were hereditary. Traumatic cataract was the most common subtype (85.92%) of acquired cataract. The few instances of cataracts due to steroids (n=3) or to metabolic disorders (n=2) occurred in males and involved both eyes.

Conclusion: The majority of pediatric cataracts in this patient population were congenital in nature. A significant lag in ophthalmologic evaluation of Chinese infants was evident and should be addressed by educating both children and parents on risk factors for cataract development. Regular assessments are especially important in children subjected to long-term systemic steroid treatments. (***Eye Science 2014; 29:138-142***)

Keywords: pediatric cataract; etiology; hospitalization

DOI: 10.3969/j.issn.1000-4432.2014.03.003

Funding: National Natural Science Foundation of China (No. 30973276)

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Introduction

Pediatric cataract is a broad term that refers to any form of cataract developing between birth and 14 years of age. Congenital cataract is the most common subtype, with a prevalence in developing countries of 5–15 per 10,000 children[1,2]. Pediatric cataracts are the leading cause of blindness in children worldwide[3]; for example, a survey conducted in Cambodia showed that visual impairment and blindness is traceable to the lens in 27.4% of children[4].

To date, surgery is still the sole means of treating pediatric cataracts[5-8]. Advanced surgical techniques and timely intervention allow early extraction of cataracts, assuring proper optical correction and treatment/prevention of otherwise irreversible deprivation amblyopia[9]. The epidemiology of previous hospital-based sampling may be representative of a broader population.

Through our efforts, the etiologies and characteristics of pediatric cataracts were reviewed in a large, diverse cohort of sufferers hospitalized in Shengjing Hospital over the past decade. The details provided are intended to provide insight into children's eye health in China.

Materials and methods

Review materials

Complete medical records of 367 children hospitalized at Shengjing Hospital of China Medical University between August, 2003 and July, 2013 for pediatric cataracts were reviewed retrospectively. All patients were under 14 years of age and presented with various subtypes of cataract as the sole criterion for inclusion.

Patient stratification

Subjects were grouped according to etiology as follows: 1) Congenital cataracts: both idiopathic and genetic in nature, or associated with other systemic and ocular abnormalities, and 2) Acquired cataracts: induced by trauma, stemming from complications, and due to steroidal or metabolic influence.

Statistical analysis

A descriptive statistical analysis was performed using Microsoft Excel. All original medical records were collected(hard copy from August, 2003 to December, 2008; digital format from January, 2009 to July, 2013) to create a database, including age at clinical presentation, gender, home address, cataract laterality and morphology, family history, primary and secondary diagnosis, and any other coexisting ocular or systemic abnormalities. Visual acuity was not addressed in this analysis, considering a majority of the children were too young for testing. All data were categorized, computed, checked, and then analyzed.

Results

Overall, 17,272 admissions were recorded by the Eye Department of Shengjing Hospital between August, 2003 and July, 2013, including 4162 children (24.1%). The 367 patients hospitalized (598 exposures) for pediatric cataracts (males: 232, 63.2%; females: 135, 36.8%; bilateral cataracts: 200, 54.5%) represented 14.4% of all pediatric admissions. Only 16 patients (4.4%) were from regions of China other than the Northeast.

Age

Cataract prevalence declined sharply as a function of age (Figure 1), with 258 patients (70.3%) <3 years old. Cataract subtypes also varied by age. The vast majority (93.8%) of cataracts seen in children < 1 year old were congenital. Mean age of traumatic cataracts was 78.5 months.

Gender

Males in all age groups and in all subtypes of cataract were more commonly affected (male to female ratios by subtype: congenital, 3:2; acquired, 3:1)(Figure 2). All cataracts due to steroids (n=3) and metabolic disorders (n=2) developed in males.

Cataract laterality

Most congenital cataracts were bilateral (65.9%), rather than unilateral (34.1%; right, 17.9%; left, 16.2%); whereas all acquired cataracts related to trauma were unilateral (right, 55.7%; left, 44.3%), and all cataracts due to steroids and metabolic disorders were bilateral (Figure 3).

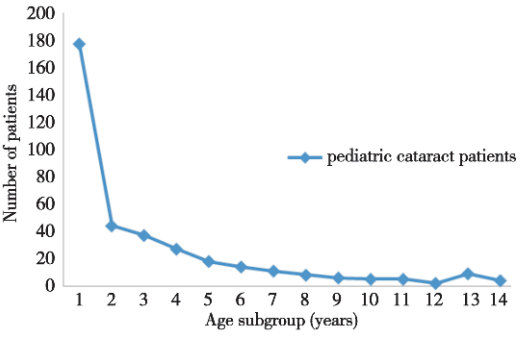

Figure 1 Distribution of pediatric cataracts by age

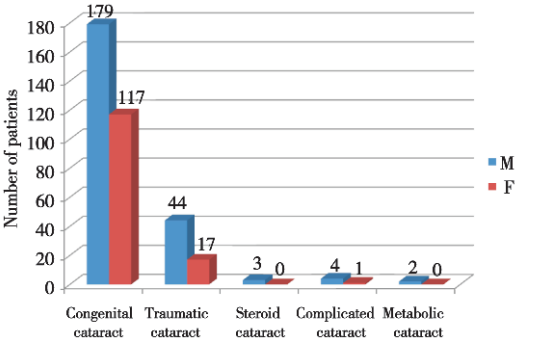

Figure 2 Distribution of pediatric cataracts by subtype and gender

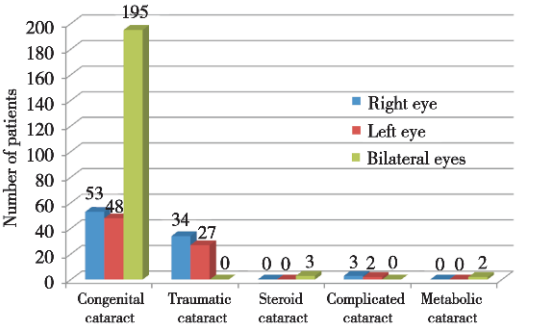

Figure 3 Distribution of pediatric cataracts by subtype and laterality

Congenital cataracts

Congenital cataracts (296/367, 80.7%) were the most common overall and were generally seen in

males (179/296, 60.5%) rather than females (117/296, 39.5%). Forty-eight (16.2%) congenital cataracts were genetic anomalies with autosomal dominance, 10 (3.4%) were associated with older parents, 5 (1.7%) occurred in twins, 1(0.3%)involved a set of triplets, 11(3.7%) were due to intrauterine infections (rubella), and 8 (2.7%) developed in premature infants.

The most frequently associated ocular anomaly was nystagmus (Table 1), which was found in 249 patients (84.1%) with congenital cataracts. Strabismus was present in 212 patients(71.6%). Other documented ocular abnormalities were as follows: 1) pseudo microphthalmus (76/296, 25.7%); 2) persistent hyperplastic primary vitreous (PHPV; 6/296, 2.0%) 3) persistent pupillary membrane (PPM; 4/296,1.4%); 4) congenital glaucoma (2/296, 0.7%); 5) structural abnormalities (2/296, 0.7%); and 6) Morning glory syndrome (1/296, 0.3%).

Table 1 Other ocular abnormalities associated with congenital cataracts

Documented ocular abnormalities	Total	Percentage
Nystagmus	249	84.1%
Strabismus	212	71.6%
Pseudo microphthalmus	76	25.7%
PHPV	6	2.0%
PPM	4	1.4%
Congenital glaucoma	2	0.7%
Structural abnormalities	2	0.7%
Morning glory syndrome	1	0.3%

PHPV, persistent hyperplastic primary vitreous; PPM, persistent pupillary membrane

Congenital heart disease (septal defects, patent ductus arteriosus, valvular stenosis, etc.) accompanied 31.4% (93/296) of congenital cataracts. Other systemic abnormalities documented with congenital cataracts included cerebral palsy(n=5), Down's syndrome (n=4), deafness (n=3), polydactyly (n=1), spina bifida (n=1), and situs inversus viscerum (n=1) (Table 2).

Acquired cataracts

Traumatic cataracts constituted 85.9% of all acquired cataracts. Five patients developed cataracts as complications secondary to anti-glaucoma surgery (n=2), iritis (n=2), and vitreous hemorrhage (from retinal detachment in aplastic anemia; n=1). Three patients also suffered cataracts from long-term steroid exposure for kidney conditions (n=2) and during high-dose treatment of Crohn's disease (n=1). In two patients, cataracts resulted from metabolic disorders (galactosemia and Wilson's disease).

Table 2 Systemic abnormalities associated with congenital cataracts

Documented systemic abnormalities	Total	Percentage
Congenital heart disease	93	31.4%
Cerebral palsy	5	1.7%
Down's syndrome	4	1.4%
Deafness	3	1.0%
Polydactyly	1	0.3%
Spina bifida	1	0.3%
Situs inversus viscerum	1	0.3%

Discussion

In this investigation, congenital cataracts (296/367, 80.7%) clearly predominated, most of which were bilateral (65.9%) and found in males (M/F ratio, 3:2). Similar results were reported by Haargaard et al. in Denmark[10], where 1027 children were afflicted with largely bilateral (64%) and male-predominant (males, 529; females, 498) congenital/infantile cataracts.

We discovered that only 16.22% congenital cataracts in our patient population were familial in nature, which was considerably lower than the range (1:4–1:3 ratio) cited by Yi et al[11]. Although Perucho-Martínez et al[12]. of Madrid also reported a 17% rate of inheritable congenital cataracts, this figure was considered low, given that primary mutations could present as isolated occurrences. Another study by Lim et al[13]. of Toronto indicated a lower rate (11.7%), while maintaining that some idiopathic cataracts likely were inheritable but could only be recognized over time or through improved access to molecular genetic testing. We concur with the latter interpretation, acknowledging that our inability to detect such primary mutations may have skewed our rate of inheritable cataracts to a lower value.

In patients with congenital cataracts, an interesting phenomenon emerged in the form of a definitive link with congenital heart disease (31.4%; Table 2). It is thus fair to speculate that the lens of the eye may share structural proteins with the cardiac framework

and heart valves. Certainly, this assumption is tentative and in need of validation. Other systemic abnormalities observed in this context were deafness, cerebral palsy, Down's syndrome, polydactyly, spina bifida, and situs inversus viscerum, all at low incidences (0.3-1.7%, Table 2).

We also identified nystagmus (84.1%) and strabismus (71.6%; Table 1) as the top two ocular complications of congenital cataracts. Following cataract extraction, nystagmus may be relieved to some extent through prompt optical correction and treatment of amblyopia, but in the majority our patients, nystagmus persisted to some extent. In addition, 76 eyes (25.7%) with congenital cataracts were smaller than normal. Average corneal diameter varied from 6–9 mm, and axial length varied from 16–20 mm, suggesting pseudo-microphthalmus. In this regard, second-round intraocular lens implantation may be impacted.

Screening for lens opacity is routinely practiced in developed countries, enabling ophthalmologists to identify and treat cataracts without delay. Half of the children in a UK study were diagnosed by the age of 10 weeks, and only 30% were diagnosed after the first year of life[14]. On the other hand, only one in five children in our cohort was diagnosed by the age of 3 months, and 43.9% were diagnosed after the first year of life. According to You et al[9]., the mean delay in diagnosis of congenital cataracts for Shandong province of China was 35.7 months; and no cataract surgeries were done within the first 3 months of life. These findings underscore a significant lag in ophthalmic evaluation of infants in China that deserves attention.

Traumatic cataracts (85.92%), all of which were unilateral, constituted the most common subtype of acquired cataracts in our analysis. Eye injuries are the chief cause of non-congenital unilateral blindness in children[15], with males typically affected more often than females[16]. Our male to female ratio of 5:2 was weighted accordingly but was lower than the 4:1 ratio reported by Johar et al. for Western India. Traumatic cataracts also tend to occur in older children (mean age, 78.5 months)[17]. Because these cataracts are largely preventable, children and parents should be forewarned that blunt or perforating trauma, ocular foreign bodies, chemical burns, and radiant energy may encourage the development of cataracts[18].

The few cataracts we encountered due to steroid use (n=3) and metabolic disorders (n=2) were bilateral and involved males, in a manner seen with nephrotic syndrome[19]. This is perhaps more of an issue in the Kingdom of Saudi Arabia, where 4.6% of pediatric cataracts are attributable to long-term systemic steroid use[20]. Nevertheless, it is especially important that children subjected to this type of treatment undergo regular ophthalmic evaluations.

Acknowledgment

The authors thank the patients and their family members for their cooperation and all the people who helped us complete the research successfully. This work was supported by National Natural Science Foundation of China (No. 30973276)

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