Lenticular Abnormalities in Children

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ABSTRACT

Purpose: To study the lenticular problems in children presenting at an apex institute.

Methods: Retrospective analysis of records (< 14 years) of new lens clinic cases was done.

Results: Of 1,047 children, 687 were males. Mean age at presentation was 6.35 ± 4.13 years. Developmental cataract was seen in 45.6% and posttraumatic cataract in 29.7% of patients. Other abnormalities were cataract with retinal detachment, persistent hyperplastic primary vitreous, subluxated lens, micro/spherophakia, cataract secondary to uveitis, intraocular lens complications, cataract with choroidal coloboma, and visual axis opacification.

Conclusion: Developmental and posttraumatic cataracts were the most common abnormalities. Delayed presentation is of concern.

INTRODUCTION

An estimated 1.4 million children younger than 15 years are blind worldwide.1 Eighty percent of blindness is avoidable (ie, readily treatable and/or preventable). Approximately three-quarters of blind children in the world are in developing countries in Africa and Asia.2

Lens abnormalities are an important cause of childhood blindness. Studies done in schools for the blind in South India and Chile showed lens abnormalities contributed to 7.4% and 9.2%, respectively.3 In a study4 done in a blind school in Karnataka, the major causes for vision loss were congenital anomalies (microphthalmos and anophthalmos; 35.7%), corneal condition (14.9%), cataract or aphakia (11.4%), and retinal disorder (19.9%).

The human lens starts developing from the fourth week of intrauterine life.5 It reacts to any insult by losing its clarity and developing opacity. This manifests as congenital cataract at birth and later as developmental cataract. However, such a presentation is not the only form of cataract in childhood. Due to its physiological and metabolic complexities, posttraumatic, complicated, and other forms such as metabolic cataract are also seen.

The lens clinic is our specialized clinic to tackle the problem of pediatric lenticular abnormalities. We collected data from children who came to the lens clinic between January 2008 and December 2009. We aimed to establish the types of cases, the etiology for which they were referred to us, and the complexities of presentation of those cases.
PATIENTS AND METHODS

Retrospective collection of data from the lens clinic records was done from January 2008 to December 2009 and all new cases were considered. All children 14 years or younger were selected. Detailed notes were made, including age at presentation, gender, ocular and systemic history, history of trauma, and any ocular surgery. The morphology of cataract was noted. A separate record was made for cases of choroidal coloboma, subluxation, micro/spherophakia, and findings suggestive of any retinal pathology or persistent hyperplastic primary vitreous (PHPV). Cases of cataract with media opacities and problems related to intraocular lenses (IOLs) were also recorded. Surgical cases with clear media, refractive error, and amblyopia were not considered.

Patients were divided into ten broad categories depending on etiology and associated features: (1) developmental cataract, (2) posttraumatic cataract, (3) cataract with retinal detachment, (4) PHPV, (5) subluxated or dislocated lens, (6) cataract secondary to uveitis, (7) micro/spherophakia, (8) complications of cataract surgery, (9) cataract with choroidal coloboma, and (10) visual axis opacification.

The developmental cataract group was further divided into subgroups on the basis of cataract morphology (nuclear, zonular, polar, and mixed). The posttraumatic cataract group was divided into closed globe and open globe injuries. The subluxated lens group was divided into nontraumatic and posttraumatic, which included both cataractous and clear lens subluxation. The nontraumatic group was further subdivided into Marfan syndrome, homocystinuria, and other (undiagnosed cause or idiopathic ectopia lentis). Cases that had already undergone surgery elsewhere, were not satisfied by the surgical outcome, and wanted a second opinion were grouped under complications of cataract surgery (including optic capture, decentration IOL, dislocated IOL, and broken haptics). The visual axis opacification group mainly had cases with posterior capsule opacification, vitreous face opacification, and pre/retro lenticular membranes.

Data were compiled and analyzed by SPSS software (SPSS, Inc., Chicago, IL).

RESULTS

A total of 1,047 children were registered. Of these, 687 children (65.7%) were male. The mean age at presentation was 6.35 ± 4.13 years (range: 0.05 to 14 years). The youngest patient was a 20-day-old male infant with bilateral congenital cataract. Detailed analysis of groups is described in the table.

Developmental cataract was the most common diagnosis (45.6%) in our study (Fig. 1). We included both congenital and developmental cases in this group. The congenital cases were usually present at birth and had a nuclear element alone or with zonular involvement, whereas developmental cataract was usually not seen at birth. Of those with developmental cataract, zonular cataract accounted for 38%, followed by nuclear cataract (22%), mixed morphology (23%), and others that consisted of total cataract, membranous cataract, polar, and subcapsular cataract (17%). The mean

<table>
<thead>
<tr>
<th>Code</th>
<th>Frequency</th>
<th>Male (%)</th>
<th>Bilateral (%)</th>
<th>Age (Y)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1:   Developmental cataract</td>
<td>477 (45.6%)</td>
<td>63.7</td>
<td>44</td>
<td>4.53 ± 3.92</td>
</tr>
<tr>
<td>2:   Posttraumatic cataract</td>
<td>311 (29.7%)</td>
<td>69.1</td>
<td>4</td>
<td>7.73 ± 3.53</td>
</tr>
<tr>
<td>3:   Cataract with retinal detachment</td>
<td>18 (1.7%)</td>
<td>61.1</td>
<td>33.3</td>
<td>8.95 ± 4.16</td>
</tr>
<tr>
<td>4:   Persistent hyperplastic primary vitreous</td>
<td>13 (1.2%)</td>
<td>46.2</td>
<td>20</td>
<td>4.61 ± 3.53</td>
</tr>
<tr>
<td>5:   Subluxated/dislocated lens</td>
<td>62 (5.9%)</td>
<td>69.4</td>
<td>54</td>
<td>9.36 ± 3.56</td>
</tr>
<tr>
<td>6:   Cataract with uveitis</td>
<td>37 (3.5%)</td>
<td>67.6</td>
<td>38</td>
<td>7.79 ± 3.31</td>
</tr>
<tr>
<td>7:   Micro/spherophakia</td>
<td>16 (1.5%)</td>
<td>56.3</td>
<td>94</td>
<td>6.56 ± 4.52</td>
</tr>
<tr>
<td>8:   Complications of cataract surgery</td>
<td>31 (3.0%)</td>
<td>74.2</td>
<td>29</td>
<td>8.07 ± 3.73</td>
</tr>
<tr>
<td>9:   Choroidal coloboma</td>
<td>12 (1.1%)</td>
<td>33.3</td>
<td>60</td>
<td>8.5 ± 4.16</td>
</tr>
<tr>
<td>10: Visual axis opacification</td>
<td>70 (6.7%)</td>
<td>67.1</td>
<td>61</td>
<td>7.58 ± 3.51</td>
</tr>
</tbody>
</table>

*Values given as mean ± standard deviation.
The mean age for developmental cataract was 4.53 ± 3.92 years, 63.7% were male, and 44% had bilateral cataract. Abnormal ocular movements such as nystagmus/nystagmoid movements were observed in approximately 15% of developmental cases. This was common with cataract at earlier onset, especially those younger than 6 months and cases with total cataract. Esotropia was more frequent than exotropia and approximately 30% of cases with developmental cataract had squint.

Posttraumatic cataract was the second largest group (29.7%; Fig. 2). Trauma included both open and closed globe injury, with most being open globe injury (70%). Cases of repaired corneal/corneoscleral perforations were also included in this group. The mean age was 7.73 ± 3.53 years, 69.1% were male, and only 4% of these cases were bilateral, which included firecracker injuries and road traffic accidents.

A total of 18 (1.7%) patients had cataract with retinal detachment that was diagnosed on B-scan ultrasonography. Twelve of these had history of trauma and the rest were bilateral and referred from a retina clinic for cataract management. The mean age of presentation of this group was 8.95 ± 4.16 years, 61% were male, and approximately 33% were bilateral.

Thirteen (1.2%) patients had cataract in association with PHPV, confirmed clinically and with ultrasonography. We had cases without classic microphthalmos and even bilateral cases. The mean age at presentation for this group was 4.61 ± 3.53 years. Of these, 46.2% were male and approximately 20% were bilateral.

Subluxated or dislocated lens was present in 62 (5.9%) patients (Figs. 3A and 3C). Of these, 32 patients had nontraumatic subluxation and 30 patients had history of trauma. In the nontraumatic subgroup, 8 patients were diagnosed as having Marfan syndrome and 3 had homocystinuria. The remaining 21 cases were not systemically evaluated and were grouped as undiagnosed. The mean age of cases with a subluxated or dislocated lens was 9.36 ± 3.56 years. Of these, 69.4% were male and 54% were bilateral.

Secondary cataract due to uveitis was present in 37 (3.5%) patients. These included cases with signs of uveitis, such as synechia, keratic precipitates, vitreous or retinal findings, and previously documented episodes of uveitis. The mean age in this group was 7.79 ± 3.31 years. Of these, 67.6% were male and 38% were bilateral. One had hyperoleon with complicated cataract (Fig. 3B).

Sixteen (1.5%) patients were diagnosed as hav-
ing micro/spherophakia. These cases had the complete lens visible on dilatation and also presented with anterior luxation of lens. One of these cases had systemic features suggestive of Weil–Marchesani syndrome (Fig. 3C). The mean age of these cases was 6.56 ± 4.52 years. Of these, 56.3% were male and 94% were bilateral.

Thirty-one (3.0%) patients presented with surgical complications (Fig. 4). Two of these cases had a broken haptic, 20 were cases of partial optic capture, and the remaining 9 cases were total optic capture. The mean age was 8.07 ± 3.73 years. Of these, 74.2% were male and 29% were bilateral.

Twelve cases (1.1%) presented with cataract associated with choroidal coloboma (Fig. 3D). Their mean age was 8.5 ± 4.16 years, 33% were male, and 60% were bilateral.

The visual axis opacification group comprised 70 patients with a mean age of 7.58 ± 3.51 years (Figs. 4B and 4C). Of these, 50 patients had posterior capsular opacification where posterior capsulorrhexis was not done by the operating surgeon. Fifteen were cases of prelenticular membrane and 5 had retrolenticular membrane that caused visual axis opacification. Males formed 67.1% of this group and 61% were bilateral.

**DISCUSSION**

The mean age of children presenting in our pediatric lens clinic was 6 years. We analyzed the mean age in different groups. The mean age for developmental cataract in our series was 4.53 ± 3.92 years. We combined all congenital and developmental cataract cases in one diagnosis. Hence, the mean age is older than reported in other studies.

In a study done in South India, 65 children with developmental cataract were included and their mean age at presentation was 53 months. The mean age depicts the delay in presentation of these cases. Mwende et al. found a mean delay in presentation of congenital cataract ranging from 9 to 18 months compared with a 24-month delay in presentation in children with developmental cataract. They found literacy of the mother and distance from the hospital to be the most important reasons for this delay. A similar study from Taiwan found the delay to be between 1 and 157 months.

The age pattern explains that many children are devoid of any medical examination for the early years of their life. This figure points to the fact that before surgery itself, visual potential in many cases is limited. Fixation develops in the early months of life and most children presented to us well after this age. Compared with all other categories, the mean age was higher in the posttraumatic group (7.7 years) and in the subluxated or dislocated lens group (9.36 years).

When analyzing the age pattern of various groups, we found it correlated with the presenting complaint. Common symptoms for developmental cases were leukocoria, abnormal eye movements, or disinterest in surroundings. Smaller eye or leukocoria was the main complaint in patients with PHPV; however, we did have cases of PHPV without classic presentation of microphthalmos. In other subtypes, especially with subluxated or dislocated lens, micro/spherophakia, and visual axis opacification, the mean age was older and most of these patients were brought in with complaints of poor vision noticed either by teachers or while playing with other children. The gap of approximately 4 years in presentation of developmental cataract and visual axis opacification or cataract surgery complications, such as optic capture or broken haptic, is not reflective of the pathology of condition. This gap is the true reflection of poor follow-up for such cases, which is in fact due to poor understanding of the condition by parents and possible improper counseling by the treating physician.

Males outnumbered females (ratio almost 2:1) in our study, except in the PHPV and choroidal coloboma groups, where females were more prevalent. Both of these are congenital anomalies and the lim-
The most common pathology in our study was developmental cataract (45.6%). The morphology varied widely from a white dot in the anterior lamellae to total opacities. Zonular cataract accounted for the majority, followed by nuclear, anterior and posterior capsular, anterior and posterior polar, total, and membranous cataract. We had 30 rubella cataracts, with the most common morphology being total cataract and others being membranous and nuclear cataract. Rubella cataract was commonly associated with atrophied iris and nondilating pupil.

Morphology of cataract has also varied in other studies. Lim et al. found posterior subcapsular cataract to be most prevalent; however, this was due to steroid use in their study. A Danish study that included congenital/infantile forms found nuclear/zonular cataract to be common.

Traumatic cataract formed the second largest group in our study and the nature of injury was penetration in more than half of the cases. The pattern of injuries has varied in different studies. In a study done in Germany, blunt trauma was found in 56.1% and open globe injury in 43.9%. Singh et al. reported penetrating trauma in 54.1% and blunt trauma in 39.3% of the cases.

Eighteen children in our study had cataract with retinal detachment. This group consisted of all patients with cataract and retinal detachment. Approximately one-third of these had bilateral retinal detachment, probably secondary to retinopathy of prematurity. History of trauma was found in most of the unilateral cases. These patients were either referred from a retina clinic for cataract surgery or they were diagnosed primarily on ultrasonography. Cataract in retinal detachment points to chronicity of the disease and is a poor prognosis for functional and surgical success of the condition.

In our clinic, most of the PHPV cataracts presented as posterior plaque with vascularization of the opacity. Mean age at presentation in this group was well below the average age in other groups because these cases are congenital and usually present with leukokoria in early life. In their work on PHPV, Hunt et al. found the mean age of presentation to be 44 days. Fifty-five eyes of 50 patients were included in the study, with 31 (62%) males and 19 (38%) females. Five (10%) of the patients were diagnosed as having bilateral PHPV. Pollard found 2 of 83 (2.4%) cases to be bilateral, whereas Haddad et al. found 7 of 62 (11%) cases to be bilateral. Twenty percent of our cases were bilateral.

We had a total of 22 cases of a subluxated or dislocated lens in our study. Nine eyes had subluxation due to trauma and 13 had a nontraumatic cause. Das et al. found posttraumatic subluxation in 7 cases and nontraumatic in 11 cases. In his study on hereditary causes of subluxation, Dehghan found the mean age of presentation to be 13.8 ± 9.1 years. He found 79.5% of cases had Marfan syndrome, 4.1% had homocystinuria, and 8.2% were simple ectopia lentis.

In our series, 16 cases of micro/spherophakia were identified on clinical examination. Most of them had complaints of poor vision. One patient presented with acute angle closure.

The prevalence of congenital coloboma is estimated to be 4.89 per 100,000 newborns, in which ocular structures are incompletely formed due to failure of the embryonic fissure to close. Coloboma is transmitted as autosomal dominant with variable penetrance and bilateral in approximately 60% of cases. In our series, 60% of cases were bilateral.

The visual axis opacification group was important in our study, having 70 cases. Although surgeons mostly agree on the necessity of posterior capsulectomy in children younger than 6 years, many younger children in our study (n = 50) underwent surgery elsewhere and no posterior capsulectomy was performed. Other causes of visual axis opacification included incomplete vitrectomy and postoperative inflammation. This highlights the necessity of proper posterior capsulectomy and anterior vitrectomy in pediatric cases.

We had few cases of broken IOL and total or partial optic capture, with partial optic capture being more common. These were cases that underwent surgery elsewhere and the patients were not satisfied with the visual outcome.

Our series is likely the largest reported series from northern India that discusses the lenticular problems in children in detail. However, because the center is tertiary, the data of more than 1,000 cases may not be the true representation of actual prevalence. This is an inherent pitfall in retrospective studies from an apex institute.

This study points to the fact that given proper and timely care, many lenticular problems can be addressed and rehabilitated. Delay in presentation...
to an ophthalmologist is common in many parts of northern India for cases of developmental/congenital cataract. Trauma is a significant cause for cataract in the pediatric age group. With the increasing number of surgeries for pediatric cataract, more problems related to cataract surgery (eg, optic capture) and cases of visual axis opacification are expected.

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