Original article

Congenital cataracts: complications and functional results according to different surgical techniques

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ABSTRACT

Purpose: To evaluate visual results, refractive error and complications in a series of congenital cataracts treated with different surgical techniques.

Methods: Retrospective review of 51 eyes with congenital cataracts which were surgically treated between 1989 and 2005. Eleven were unilateral cataracts and 40 were bilateral cataracts. Thirty-three eyes were treated with a primary intraocular lens (IOL) and 18 were aphakic eyes.

Results: The best outcomes were observed in bilateral cataracts and in those that received early primary IOL. Three eyes achieved 0.8-1, two eyes achieved 0.6-0.7. All of them were eyes with primary IOL. Seventeen percent of unilateral cataracts and 23.8% of bilateral cataracts achieved 0.4 or better. The most serious complication was visual axis opacification (VAO), 27 of 51 eyes developed VAO. Fifty percent of eyes with primary IOL and 63% of aphakic eyes developed VAO. The VAO developed sooner in the IOL group (6.5 months on average). There was a high myopic shift in 3 eyes. There was a significantly greater myopic shift in the unilateral cases.

Conclusions: The best acuities were achieved in the eyes which had been treated with early primary IOL, but they had a higher rate of complications (VAO), thus requiring reoperation. Final refraction in the unilateral group was significantly more myopic than in the bilateral group.

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Introduction

Congenital cataracts continues to be an important problem in what concerns management. It still constitutes one of the most important causes of blindness both in developing and developed countries. The prevalence of blindness due to congenital cataracts ranges between 5% and 20% according to the series1.

The main cause of loss of vision related to congenital cataracts is amblyopia.

The therapeutic approach to congenital cataracts has evolved rapidly in recent years. An early diagnostic and the introduction of new intraocular lenses with a broader range of power as well as improved adaptation to small sacs contribute to a better visual prognosis for these patients. Similarly, a more effective post-surgery amblyopia therapy and a correct and comprehensive visual rehabilitation treatment also contribute to improve said visual prognosis.

At present, many research groups are utilizing intraocular lenses (IOL) in increasingly younger children. For infants under two years of age there is still controversy regarding the use of IOL for treating congenital cataracts2,3.

Subjects, material and methods

An epidemiological, descriptive and longitudinal study which included all the children with isolated surgical congenital cataracts intervened in the Doce de Octubre Hospital of Madrid between 1989 and 2005. The eyes with other associated malformations as well as acquired pediatric cataracts were excluded. A total of 51 eyes were included in the study. The following parameters were analyzed: final visual acuity (VA) based on the surgery technique, early and late complications and final refractive error.

VA was measured utilizing the Pigassou test and in younger or non-cooperative children the preferential gaze (Teller). Objective tests were utilized for calculating the refractive error. In this case, skiascopy was utilized, inducing pharmacological midriasis with cyclopentholate 1% in children over one year of age and 0.5% in front below the year.

VA and refraction were measured at diagnostic time and prior to surgery. Then, the VA and refraction were monitored the day after the operation, after one week and one month and subsequently every three months or whenever the VA indicated a refractive change.

A slight myopic defect was defined up to 3 negative dioptres, a medium defect between 3 and 6 negative dioptres and a severe defect over 6 dioptres. A slight hypermetropic defect was up to 3 positive dioptres, a medium defect between 3 and 6 positive dioptres and a severe defect over six dioptres.

As regards the surgical treatment, different surgical techniques have been implemented depending on the historic stage of the cataracts. In the older cataracts IOL was not used because we didn’t have at the time lenses adapted to pediatric eyes or experience with IOL in children. In some more recent
bilateral cataracts, IOL was not utilized in the same surgical intervention, opting instead for optical correction after surgery and a subsequent secondary IOL implant. Similarly, the oldest IOL were rigid PMMA lenses whereas the new ones were straight-edged flexible acrylic IOL, better suited to pediatric sacs.

For selecting the power of the IOL keratometry was utilized together with ultrasound to measure the antero-posterior axis. The IOL power was calculated utilizing latest generation formula suited for short eyes (SRK T, Hoffer Q and Holladay I). After obtaining the theoretical power of the IOL, we corrected for the age of the child, with hypocorrection between 10% and 30%, seeking a refractive defect of approximately 5-6 positive dioptres in infants under one year, 4 positive dioptres in children between one and two and approximately 2-3 positive dioptres in children between 2 and 4. In this way we can control the residual myopic error derived from the growth of the eyes, particularly in children under two years of age.

In all cases of our hospital, post-surgery amblyopia was treated.

Early complications were considered to be those which occurred in the first month after surgery, while late complications were those occurring over one month after surgery.

In the group of 51 eyes with congenital cataracts intervened in our hospital, 22% were unilateral while 78% were bilateral. Both were treated as independent groups due to the different visual prognosis. Similarly, the surgery groups with and without IOL were treated independently. We also differentiated between cataracts intervened with rigid IOL and those intervened with flexible IOL.

In the group of unilateral cataracts, 73% were eyes intervened with IOL while 27% were intervened without IOL. In the bilateral cataract group, 63% were eyes intervened with IOL whereas 37% were aphakic eyes.

### Results

The mean age of surgery was of 23.5 ± 39.4 months (including initially nonsurgical cataracts which subsequently evolved to surgical).

In the bilateral cataract group, the mean time between surgery of one eye and the contralateral eye was of 3.4 ± 9.79% of the bilateral cataract group received surgery in both eyes at the same time. Different surgical techniques were utilized on the basis of the historic stage (Table 1).

Thirty-three eyes received primary IOL while 18 eyes remained in postsurgical aphakia.

### Complications

Visual axis opacification (VAO) was the most important complication, which was developed by 27 eyes. In addition, this complication arose more frequently and earlier in eyes with IOL (Table 2).

Other complications were much less frequent or important. The most frequent early complication was an inflammatory reaction in the anterior chamber, which occurred in 10 eyes. Early glaucoma arose in two eyes. The most frequent form of glaucoma in our series was late glaucoma, which appeared in 31% of aphakic eyes.

### Final visual acuity

In the unilateral cataract group, the best VA was 0.8-1, which occurred in one eye which was pseudophakic. In the bilateral cataract group we found VAs better than 0.8 in only two eyes which were also pseudophakic (Table 3).

As regards the eyes in which IOL was utilized, better VA results were obtained utilizing acrylic and flexible IOL than when using rigid and less adaptable IOLs (Table 4).

### Refractive defect

In the unilateral pseudophakic cataract group there was only one case of slight residual myopic defect and only one eye with a high residual myopic defect.

In the bilateral pseudophakic cataract group there were 5 eyes with slight myopic defect, two eyes with medium myopic defect and two eyes with high myopic defect.

### Discussion

#### Surgical procedure

In recent years, surgical procedures have improved and adapted to pediatric eyes together with technical developments.
However, the use of IOL in children under two years of age remains controversial\textsuperscript{3,5}. The currently used technique (37%) consists in continuous circular capsulorrhexis, lens aspiration, manual posterior capsulotomy and anterior vitrectomy. The IOL is inserted in the capsular sac. In the first cataracts of the series, PMMA rigid IOL were utilized, which later evolved in material and morphology to the current acrylic lenses, involving a reduction of posterior capsular opacification (PCO).

In our study we utilized lenses with powers between +20.50 and +26.50 dioptres. The power is downwardly corrected between 20\% in older children and 30\% in the younger ones. At present, it is perfectly accepted to use IOL in pediatric patients, even those under two. The IOL ensures a complete and constant refraction in the growing eye, reducing the risk of amblyopia due to deprivation. An increasing number of surgeons are adopting this practice\textsuperscript{2,4,6,7}.

Complications

The most important complication of congenital cataract surgery is the visual axis opacification (VAO) due to its amblyogenic capacity. Any delay in its diagnosis could involve the development of irreversible amblyopia. The intensity and precocity of opacification his greater in younger children. The rate and intensity thereof was reduced with the systematic application of posterior capsulotomy and anterior vitrectomy as a part of the surgical procedure. Even so, this complication still occurs frequently in infants under one year of age. A factor that influences the appearance of VAO is the use of IOL. In our study, 50\% of pseudophakics developed VAO. In the aphakic group, 65\% of eyes developed VAO. This factor is independent of the bilateral characteristic of the cataracts.

Accordingly, we can say that in our study VAO is more frequent in the aphakic group (p=0.6).

The mean age of patients that are affected by VAO in our study was of 8.5±10.9 months after surgery. We observed that VAO occurred earlier when IOL is utilized then when the patient is left aphakic. Even though posterior capsulotomy and anterior vitrectomy is performed, the risk of VAO continues to exist because the IOL acts as a scaffolding for the proliferation of fibroblastic cells or, in the absence thereof, the anterior and posterior capsules serve as scaffolding for said cells\textsuperscript{8}.

In the group of pseudophakics affected by, up to 58\% of required surgery for their treatment whereas in the group of aphakic patients 41\% of eyes required surgery for treating VAO. Accordingly, it was observed that the rate of reoperations due to VAO was higher when IOL was utilized.

In what concerns other complications, the most frequent in both groups of the study was the inflammatory reaction in the anterior chamber. It was a very frequent but hardly an aggressive complication with small consequences because in all cases it remitted with topical corticoid treatment. In our study, early glaucoma arose in two eyes (15.4\%). Both eyes were pseudophakic.

Late glaucoma was more frequent in the aphakic group, occurring in 57\% of aphakic eyes while in the pseudophakic group it occurred in 42\% of eyes (p=0.2).

### Table 3 – Final visual acuity in the congenital surgical cataracts group per localization and use of IOL in the surgical technique

<table>
<thead>
<tr>
<th>Final visual acuity</th>
<th>No cooperation</th>
<th>&lt; 0.1</th>
<th>0.1</th>
<th>0.2-0.3</th>
<th>0.4-0.5</th>
<th>0.6-0.7</th>
<th>0.8-1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral (n=11)</td>
<td>OL yes (n=8)</td>
<td>18.18</td>
<td>27.27</td>
<td>9.09</td>
<td>0</td>
<td>9.09</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>OL no (n=3)</td>
<td>0</td>
<td>18.18</td>
<td>0</td>
<td>9.09</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Bilateral (n=40)</td>
<td>OL yes (n=25)</td>
<td>27.5</td>
<td>5</td>
<td>10</td>
<td>10</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td>OL no (n=15)</td>
<td>5</td>
<td>5</td>
<td>2.5</td>
<td>10</td>
<td>15</td>
<td>0</td>
</tr>
</tbody>
</table>

### Table 4 – Final visual acuity result in the surgical congenital cataract group per implanted IOL type

<table>
<thead>
<tr>
<th>Final visual acuity</th>
<th>No cooperation</th>
<th>&lt; 0.1</th>
<th>0.1</th>
<th>0.2-0.3</th>
<th>0.4-0.5</th>
<th>0.6-0.7</th>
<th>0.8-1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rigid IOL (n=4)</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Flexible IOL (n=20)</td>
<td>12</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Total (n=24)</td>
<td>13</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

Statistical significance p=0.03.
Visual acuity

The VA analysis was studied independently according to laterality (unilateral-bilateral) and use of IOL. In addition, within the IOL group we differentiated one rigid IOL subgroup and another flexible IOL group.

One of the most important results of this study is that all the eyes with a final VA of 0.8-1 were pseudophakic (p=0.008) and all the eyes with final VA of 0.6-0.7 were also pseudophakic.

The best VA achieved in the aphakic group was 0.4-0.5 (33%). In 27.8% of aphakic eyes, a final VA of 0.2-0.3 was achieved.

In addition, we observed better VAs in the bilateral cataract group compared to the unilateral cataract group, regardless of the surgical technique applied. Only 16.7% of unilateral cataract achieved a final VA of 0.4 or better while 23.8% of bilateral cataracts achieved a final VA of 0.4 or better.

Accordingly, the best functional results were achieved in children with bilateral congenital cataracts who were intervened in their first year of life utilizing IOL.

Refractive defect

Two children with unilateral cataracts and nine with bilateral cataracts developed a myopic defect and all were obviously pseudophakic eyes.

There are still unresolved questions about the impact of IOL in the growth of a pediatric eye, particularly in infants under one year. Vasavada et al found severe myopic defects in children intervened in their first two years of life. The myopic defect was more acute in unilateral vis-à-vis bilateral pseudophakic eyes.

On the other hand, it was observed that visual deprivation produces by itself an elongation on the axial axis. Vasavada et al described that the VAO did not influence the growth rate of the axial axis.

The current situation is that there is no consensus about the variables that modify the growth of the axial axis in pseudophakic pediatric eye, particularly in infants under one year of age. It is known that some factors such as the age at the time of the surgery, aphakia, pseudophakia, laterality and visual deprivation can influence said growth.

In what concerns the election of the IOL power to be utilized in a pediatric eye, different aspects have been taken into account. Everybody agrees that if we implant an IOL having the power corresponding to the emmetropia existing at that moment, an important myopia will develop in a few months after surgery as a consequence of the growth of the pediatric eye. Therefore, it seems logical to correct downwards the power of the lens calculated for the emmetropia in order to avoid this myopic defect in younger children, particularly in infants under one year of age.

Some studies suggest a post surgery refraction of +6 dioptres in infants one year old. A defect of +2 dioptres in children aged 1-3 and a defect of +2 dioptres children aged 2-4.

It is important to take into account that in infants under one year of age with eyes having antero-posterior axes under 200, large and important residual refractive errors can appear when calculating the IOL. This is because the calculation formulae are not designed for small eyes, and not even the formulae for short axes (Holladay, Hoffer Q) achieve greater precision in calculating IOL powers. It would be necessary to establish new calculation formulae for pediatric eyes.

In our study we have achieved the best functional results when surgery was carried out at an early stage in patients in which continuous circular capsulorrhexis, posterior capsulotomy, anterior vitrectomy was performed and we have utilized flexible lenses implanted in the capsular sac. As this is an infrequent but difficult pathology to manage, we do not have action protocols ensuring the best functional results. They are different approaches to this pathology according to the different study groups.

The aim of this descriptive study is to make a self-assessment of our results according to the different techniques and, together with the experiences of other groups, approach this complicated pathology in the best possible manner in order to obtain the best functional and refractive results for these patients.

REFERENCES