Management of subluxated lens in young patients

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\textbf{A B S T R A C T}

Objectives: To evaluate visual outcomes in patients treated for lens subluxation. Secondary objectives are to report best corrected visual acuity (BCVA) in LogMAR and compare the outcomes of patients managed conservatively with those treated surgically.

Methods: Retrospective comparison of BCVA in patients under the age of 50 years old with lens subluxation, managed conservatively or surgically.

Results: A total of 49 eyes of 28 patients were included. Demographic characteristics were similar in both groups. Twenty eyes were treated surgically (40.8\%) compared to 29 with medical treatment (59.2\%). Marfan syndrome (79.6\%) was diagnosed in 39 eyes. LogMAR BCVA post intervention was 0.35 ± 0.31 for medical treatment and 0.39 ± 0.32 for the surgical group, with no significant differences (p = 0.63). Improvements in LogMAR lines were 2.7 ± 4.2 and 4.11 ± 4.2 (p = 0.35), respectively. Two eyes in the surgery group developed ocular hypertension (0.04\%), none with retinal detachment.

Conclusions: The final BCVA showed no significant differences in this group of patients. BCVA depends on the visual potential of the rehabilitated eye rather than a specific type of intervention.

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\textbf{Manejo del cristalino subluxado en pacientes jóvenes}

\textbf{R E S U M E N}

Objetivos: Evaluar resultados visuales en pacientes tratados por subluxación del cristalino. Los objetivos secundarios son reportar la agudeza mejor corregida (AVMC) en LogMAR y comparar los resultados de los pacientes manejados de manera conservadora con aquellos manejados quirúrgicamente.

Métodos: Estudio retrospectivo comparativo de AVMC en pacientes menores de 50 años con subluxación de cristalino, manejados de manera conservadora con lentes o tratados quirúrgicamente.

Palabras clave:
Subluxación cristalino
Syndrome de Marfán
Aphaquia
Lentes al aire
Lentes de contacto

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Resultados: 49 ojos de 28 pacientes fueron incluidos. Las características demográficas fueron similares en ambos grupos. 20 ojos fueron tratados quirúrgicamente (40,8%) versus 29 con tratamiento médico (59,2%). 39 ojos con diagnóstico de síndrome de Marfan (79,6%). La AVMC LogMAR post intervención de 0,15 ± 0,13 para el tratamiento médico y 0,39 ± 0,32 para el grupo de manejo quirúrgico, sin encontrarse diferencias significativas (p=0,63).

Conclusiones: La AVMC final no presentó diferencias significativas en este grupo de pacientes, dependiendo del potencial visual del ojo rehabilitado más que de un tipo específico de intervención.

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Introduction

Lens subluxation can be classified as congenital, developmental or acquired. It adopts a range of presentations either as an essential or idiopathic expression or associated to other ocular pathologies and systemic hereditary diseases such as Marfan syndrome, Weil–Marchesani syndrome, homocystinuria, hyperlsinemia, Stickler syndrome, sulfite oxidase deficiency and Ehlers–Danlos syndrome, although it can also be acquired or secondary, mainly traumasms or subluxation associated to pseudo-exfoliation. Lens subluxation produces vision alterations primarily due to refractive errors and induced anisometropia and, being progressive, frequently induces refractive changes and high astigmatism. Due to the movements of the dislocated lens, the creation of intermittent phakic and aphakic visual axes can give rise to visual alterations and amblyopia if occurring at early ages, particularly if both eyes are involved. In general, the main complications of lens displacement are high ametropia, with other complications such as glaucoma or lens induced uveitis being less frequent.

Lens subluxation management is controversial. The best surgical technique and the appropriate time of intervention are not clear. In addition, there is no consensus about which patients should be treated only with conservative, nonsurgical approaches. The typical surgery indications were for subluxation compromising the visual axis and eyesight that cannot be corrected with spectacles or contact lenses. Some surgical papers reported high complication rates during and after surgery, with corresponding low visual improvement indices. However, the development of new closed surgical techniques in recent years has significantly improved visual results and long-term prognosis for said group of patients. On the other hand, the approach in our institution has varied in recent years toward more conservative managements, mainly in ophthalmic pediatricians, based on the observation that aphakic lens correction achieves good results even in small aphakic portion areas (1/5 to 1/6 of the pupil area). The primary objective of this paper is to present a group of patients treated in our institution for lens subluxation, the secondary objective being the comparison of the visual results with medical and surgical treatments.

Subjects, materials and methods

Patients

Retrospective review of records of patients treated in our institution from January 2002 up to September 2011 was done. The patients included in the review were under the age of 50 years at surgery time (conservative or surgical management) exhibiting lens subluxation. The eyes with follow-up periods under 3 months (n = 17), cataracts (n = 8), retina detachment when visiting the practice (n = 10), dislocated nucleus (vitreous or anterior chamber) (n = 5) or pseudofokia (n = 1) were excluded from the study. Similarly, patients with subluxation secondary to traumatism or pseudo-exfoliation were not considered for inclusion. The review was approved by the ethics committee of our institution.

Demographic characterization

The presurgery assessments included a full ophthalmological assessment, recording the age at the time of the intervention, the follow-up, the associated cause, the subluxation direction and amounts expressed in percentages according to the Waiss-wol classification (based on the lens position in the pupil area, grade 1: lens entirely in pupil area; grade 2: 2/3; grade 3: 1/2; grade 4: lens not in pupil area), best corrected visual acuity (BCVA) in LogMAR, phakic and/or aphakic refraction and diagnostic of amblyopia.

Intervention

The patients were recorded according to the intervention as conservative or surgical management. Conservative management is taken to be a set of nonsurgical interventions aiming at improving vision (exterior lens, contact lens, occlusion). Indications for surgery were BCVA < 20/60, progressive subluxation compromising the visual axis, imminent total dislocation or amblyopia that reverts with treatment. The different types of surgery were recorded and classified in categories: lensectomy, pseudophakia with and without capsular sac stabilization, lens fixed to the sclera or lens fixed to the iris.
Table 1 – Demographic characteristics of both groups. No significant differences prior to the intervention.

<table>
<thead>
<tr>
<th></th>
<th>Physician</th>
<th>Surgery</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>No.</td>
<td>29</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td>Sex (male)</td>
<td>21</td>
<td>10</td>
<td>0.11</td>
</tr>
<tr>
<td>Age at intervention</td>
<td>12.7 ± 12.6</td>
<td>14.5 ± 8.4</td>
<td>0.55</td>
</tr>
<tr>
<td>(years)</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Follow-up (months)</td>
<td>60 ± 58</td>
<td>53 ± 52</td>
<td>0.67</td>
</tr>
<tr>
<td>Amblyopia</td>
<td>6</td>
<td>6</td>
<td>0.45</td>
</tr>
<tr>
<td>Luxation grade (%)</td>
<td>33.7 ± 20.1</td>
<td>40.7 ± 13.8</td>
<td>0.26</td>
</tr>
<tr>
<td>Presurgery BCVA</td>
<td>0.63 ± 0.41</td>
<td>0.79 ± 0.42</td>
<td>0.22</td>
</tr>
<tr>
<td>LogMAR</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Presurgery SE in aphakia</td>
<td>10.9 ± 3</td>
<td>10.4 ± 3.4</td>
<td>0.84</td>
</tr>
<tr>
<td>Presurgery SE in phakic</td>
<td>−8.5 ± 6.9</td>
<td>−11.9 ± 10.9</td>
<td>0.39</td>
</tr>
</tbody>
</table>

Primary and secondary result

For the primary objective, the BCVA in LogMAR for the conservative managements group was compared with the surgical treatment group, whereas for the secondary objective differences in refraction and complications were targeted.

Statistical analysis

The patients on conservative management and surgical treatment were compared with the T for student test and the Chi square test. Values of p < 0.05 were considered to be statistically significant. For analyzing the data, the SPSS 16.0 for Mac (Copyright © SPSS Inc., 1989–2007) application was used.

Results

Patients

Overall, 49 eyes of 28 patients were included in the study. The demographic characteristics were similar in both groups (Table 1). Twenty eyes were surgically treated (40.8%) versus 29 with medical treatment (59.2%); 39 eyes with Marfan syndrome diagnostic (79.6%), two with Weill–Marchesani syndrome (4.1%) and 8 idiopathic subluxations (16.3%). Of the 39 eyes comprised within the subluxation records, 23 were grade 2 according to the Waiswol classification and 16 eyes were grade 3. Of the 20 operated eyes, 6 had been submitted to lensectomy without intraocular lens, 1 had been treated with intraocular lens without requiring sac stabilization methods (endocapsular rings and segments), 7 eyes with intraocular lens with sac stabilization, 1 eye treated with lens fixed to the iris and 5 eyes treated with intraocular lens with scleral fixation.

Visual acuity

LogMAR BCVA prior to the intervention of the medical treatment group was of 0.63 ± 0.41 and 0.79 ± 0.42 for the surgical group, with post-surgery LogMAR BCVA being 0.35 ± 0.31 for the medical treatment group and 0.39 ± 0.32 for the surgical management group. No significant differences were found (p = 0.63). The improvements in LogMAR lines were, respectively, 2.7 ± 4.2 and 4.11 ± 4.2 (p = 0.35). Table 2 shows the BCVA difference for aphakic correction compared to phakic correction in a group of patients in which both were obtained, with a noteworthy difference toward aphakic correction.

Spherical equivalent

The spherical equivalent (SE) was measured based on the luxation grade in the phakic and the aphakic portion. For the medical treatment group in the subgroup of phakic portion measurement, the SE prior to the intervention was of −8.5 ± 6.9 compared to post-surgery SE of −3.6 ± 15.2 (p = 0.13), while the aphakic portion measurement subgroup exhibited presurgery SE of 10.8 ± 3 compared to post-surgery SE of 9.7 ± 3 (p = 0.059). In what concerns the surgical treatment group, the phakic portion measurement subgroup exhibited presurgery SE of −11.9 ± 11 compared to post-surgery SE of 3.4 ± 7.8, yielding a significant difference (p = 0.01), whereas the aphakic portion measurement subgroup exhibited a presurgery SE of 10.4 ± 3.4 compared to post-surgery SE of 7.3 ± 7 (p = 0.29).

For the phakic portion measurement group, when comparing the post-surgery SE of the medical treatment group (−5.2 ± 15.9) with the surgery group (2 ± 5.7) no significant differences were found (p = 0.1), while for the aphakic portion measurement group when comparing the post-surgery SE of the medical treatment group (9.7 ± 3.1) with the surgical treatment group (7.3 ± 7), no significant differences were found either (p = 0.32).

Contact lenses compared with open air lenses

Of the subjects with medical treatment, 12 (26.7%) were treated with contact lenses and 17 with spectacles (37.8%), without finding significant differences for the amount of LogMAR lines gained in the open air lens group (3.7 ± 4.6) compared to the contact lens group (1.8 ± 2.1) (p = 0.17).

Complications

Two eyes of the surgery group evolved with OHT (0.04%), and none with RD.

Discussion

Lens subluxation management, surgery time and technique to be applied are subject to debate. However, early diagnosis can be useful for diagnosing diseases with systemic compromise.

Most frequent reports in the literature refer to surgical treatment of subluxated lenses. The first reports indicated a higher rate of intra- and post-surgery complications with poor visual improvement. Final VA is frequently the primary objective of the papers which are mostly retrospective or series of cases and based on a small amount of eyes (between 4 and 97 eyes). In general, surgery achieves significant improvements of the baseline BCVA, which varies in different papers from a LogMAR 2 baseline BCVA to a final BCVA between 0.7 and 0.2 LogMAR. A prospective randomized study comprising 31 eyes of 16 patients with Marfan syndrome.
which compared lens fixed to the iris with lens fixed to the sac exhibited a significant BCVA improvement in both groups, without differences between both and reaching up to 0.29 LogMAR.13 In our paper we report BCVA values similar to those obtained in the literature with the exception that one group of patients achieved those values with medical treatment only. Accordingly, final BCVA depends more on the visual potential of the rehabilitated eye than on the type of intervention.

In what concerns complications, publications provide a broader range thereof. One study with lensectomy15 reported that up to 51% exhibited immediate complications (vitreal loss, iris prolapse, corneal edema, hyphema among others), whereas a prospective lensectomy study of 95 eyes of children reported only one non-significant vitreous hemorrhage and one late case of retinal tear treated with photocoagulation.10 Other papers reported complications such as hyphema in 5%,14 corneal decompensation between 10% and 14%,5,7,13 temporary ocular hypertension between 4% and 7%,2,4,11,13 intraocular lens dislocation between 2 and 19%,4,13 and cystoid macular edema, 3%.1 As regards severe complications, 10−19% exhibited retina detachment.5,7,15 We were unable to find reports on endophthalmitis in this group of patients. In our study the frequency of complications was low, with 2 eyes exhibiting temporary ocular hypertension.

Surgical techniques are as varied as the amount of surgeons that perform them and the type of lens being implanted. Lensectomy has been applied with or without anterior vitrectomy, removal of posterior capsule with vitrector, endosacular rings, segment implant in the sac and stitching of the lens to the sclera. The development of new closed surgical techniques in recent years has been reported to provide significant improvements in visual results and long-term prognosis, even though the series report etiologies that are different to those reported in our series (such as patients with trauma and uveitis).3 However, it has not been demonstrated that one technique has special advantages over others, which explains the broad range of surgeons preferences and the particularities of each case.

Specifically in children, the results are limited mainly due to pre-existing amblyopia.1 If it cannot be treated effectively with conventional methods such as spectacles and contact lenses (e.g., high anisometropia), the surgical removal of the lens appears to be a good option. In our study we did not find differences between the surgical group vis-à-vis the medical management group. Serial controls should always be considered to correct possible refractive errors and instructions should be given for treatment with patches for amblyopia if necessary. Probably the vast majority of pediatric patients with subluxated lenses can be medically managed with spectacles or contact lenses and will probably not require surgical treatment in the mid term. An additional point for conservative management is that accommodation is maintained. Evidence indicates that patients with Marfan diagnosis and subluxed lens maintain adequate accommodation capacity.18 In any case, seeking correction following the phakic part is usually difficult due to the magnitude and irregularity of the defects. One option is to correct ametropia through the aphakic portion. Just like in operated patients we will not have accommodation but surgery will be avoided together with potential complications such as progression of refraction toward myopia, as occurs in Marfan patients. However, if the patient exhibits subluxation due to any of the above-mentioned reasons, it is foreseeable that surgery will improve visual acuity with acceptable risks according to current standards. We believe that in patients with lower risk of refractive changes, the introduction of an intraocular lens is a good option, above all in the presence of intolerance to contact lenses or very thick spectacles even though complex insertion techniques may be required (segments, rings, fixation, etc.). Even though our study involves natural limitations of retrospective analyses, it still shows that the final visual acuities are similar for both approaches and this sets the groundwork for future protocols designed in order to establish better associations.

### Conflict of interests

No conflict of interests has been declared by the authors.
REFERENCES