Intraocular Lens Implantation in Marfan’s Syndrome

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ABSTRACT

**Purpose:** To describe the results of the removal of ectopic lenses from patients with Marfan’s syndrome using the scleral fixation method.

**Methods:** Intraocular lenses (IOLs) were implanted in six eyes affected by lens dislocation (ectopia lentis) using either the scleral fixation method (five eyes) or a silicone IOL in the capsular bag (one eye). Mean patient age ranged from 8-11 years and follow-up ranged from 7-20 months.

**Results:** Functional success was obtained in all eyes. Postoperative visual acuity was 20/20 to 20/40. One patient showed a dislocation of the IOL in the anterior chamber. In three eyes, an opacification of the posterior capsule was treated using an Nd:YAG laser.

**Conclusion:** Intraocular lense implantation using the scleral fixation technique is the first choice in patients with Marfan’s syndrome because it reduces the complications of IOL decentration.


INTRODUCTION

Marfan’s syndrome, also known as congenital hypoplastic dystrophia mesodermalis, is a rare disorder of the connective tissue that affects the skeletal system, the cardiovascular system, the eyes, and sometimes the central nervous system. The syndrome is hereditary through an autosomal dominant trait and affects approximately 1 in 10,000 people. The characteristics of this disease include long and slender skeletal extremities, arachnodactyly, chest abnormalities (pectus excavation or pectus carinatum), and most significantly, cardiovascular anomalies including dilatation of the aortic root with aortic regurgitation, mitral valve prolapse, and sometimes dissecting aortic aneurysm. As far as the eyes are concerned, the syndrome is identified by ectopia lentis (present in 60%-80% of cases) with straining of the zonular fibers and spherochakia with associated myopia (axial or lenticular), and more rarely, keratoconus, heterochromia iridis, or retinal detachment.

MATERIALS AND METHODS

Intraocular lenses (IOLs) were implanted in six eyes affected by lens dislocation (ectopia lentis) in
TABLE

PRE- AND POSTOPERATIVE VISUAL ACUITY

<table>
<thead>
<tr>
<th>Sex/Age at Surgery</th>
<th>Eye</th>
<th>Surgical Procedure</th>
<th>Visual Acuity</th>
</tr>
</thead>
<tbody>
<tr>
<td>F/11 y</td>
<td>OS</td>
<td>Lensectomy-vitrectomy; sclerally fixed IOL (+25)</td>
<td>20/70 (22-3 [180°])</td>
</tr>
<tr>
<td></td>
<td>OD</td>
<td>Extracapsular cataract extraction; silicone IOL in the bag (+25)</td>
<td>20/40 (21-3.25 [180°])</td>
</tr>
<tr>
<td>M/8 y</td>
<td>OD</td>
<td>Intracapsular extraction-vitrectomy; sclerally fixed IOL (+16)</td>
<td>20/200 (216)</td>
</tr>
<tr>
<td></td>
<td>OS</td>
<td>Extracapsular cataract extraction; sclerally fixed IOL (+18)</td>
<td>20/200 (211)</td>
</tr>
<tr>
<td>M/8 y</td>
<td>OS</td>
<td>Lensectomy-vitrectomy; sclerally fixed IOL (+25)</td>
<td>20/200 (22-1 [180°])</td>
</tr>
<tr>
<td></td>
<td>OD</td>
<td>Extracapsular cataract extraction; sclerally fixed IOL (+25)</td>
<td>20/70 (22-2 [180°])</td>
</tr>
</tbody>
</table>

children with Marfan’s syndrome. Patients underwent cataract removal and IOL implantation using either the scleral fixation method (five eyes) or a silicone IOL in the capsular bag (one eye).

Lens power was calculated with the Binkhorst equation after keratometry and biometry, with the help of A-diagnostic ultrasonography. Polymethylmethacrylate IOLs with a 7-mm biconvex body and a total length of 12.5 mm were implanted. Although larger lenses are commonly used in other countries, we have encountered no disadvantages using smaller lenses. All surgeries were performed under general anesthesia with orotracheal intubation.

Surgical Technique

The sclera was exposed from 3 o’clock to 9 o’clock with a 180° peritomy. Incisions were made at 3 o’clock and 9 o’clock to form two triangular scleral flaps with sides of approximately 3 mm based on the limbus. Paracentesis was performed at 5 o’clock.

The anterior chamber was opened with a sclerocorneal incision, and viscoelastic substance was inserted. Next, anterior capsulotomy or capsulorrhesis was performed. The lens substance was aspirated manually or mechanically, and an anterior vitrectomy was performed.

A Prolene monofilament was inserted through the sulcus at the level of the scleral flaps at 1.5 mm of the limbus. The monofilament was extracted using an IOL hook through the sclerocorneal aperture and attached “ab externo” to the eyes of the loops. The IOL was inserted into the posterior chamber with simultaneous tension of the threads to ensure correct location of the IOL. The monofilament was sutured to the sclera, and the scleral flaps were sutured, covering the knot to protect it. The viscoelastic substance was aspirated from the anterior chamber, and the sclerocorneal opening was sutured with separate stitches.

Postoperatively, antibiotics and corticosteroids were instilled four times daily as well as timolol 0.5% twice daily for 20-30 days. In Italy, the use of a topical beta blocker to prevent increased intraocular pressure is common. In addition, betamethasone and amoxicillin (or cefotaxime) were administered for the first 4 days postoperatively.

RESULTS

Patients underwent clinical examination 15, 30, 60, and 90 days postoperatively, and visual acuity was evaluated from a minimum of 7 months to a maximum of 20 months (Table).

In one case, an early postsurgical complication arose consisting of dislocation of the IOL in the anterior chamber. This IOL subsequently was repositioned. In the case with the longest follow-up, an opacification of the posterior capsule developed and was treated using a Nd:YAG laser. To date, there has been no evidence of hemorrhage, suture or scleral erosion, endophthalmitis, retinal detachment, uveitis, or glaucoma, which are complications frequently mentioned in the literature.
These results confirm that the implantation of a posterior chamber, sclerally fixed IOL is an effective technique in the case of ectopia lentis in young patients. Continued follow-up will give indications as to the long-term safety of the technique.

**DISCUSSION**

In the past, the management of patients with ectopia lentis has been controversial, principally because of the numerous complications that arose after surgery. In 1972, Jensen and Cross described the results of 47 cases of removal of ectopic lenses from patients with Marfan's syndrome using various techniques and carried out by a number of surgeons. They reported immediate complications (prolapse of the iris, corneal edema, recurrent blood in the anterior chamber, and vitreous loss) in 51% of cases and detachment of the retina in 19% of cases. Detachment of the retina also has been reported in cases in which ectopia lentis has not been treated surgically, since this disorder leads to fragility of the retina. Hence, techniques that involve a risk of retinal trauma should be avoided. The use of phacoemulsification with vitrectomy has shown to be the best method for extraction of an ectopic lens.

Implantation of an intraocular lens is the most commonly used technique for the recovery of sight in children suffering from aphakia. This approach avoids the use of contact lenses, which are problematic in young patients for a number of reasons including the frequent occurrence of infection, the ease with which the lenses may be misplaced, and the necessity for a high degree of collaboration from the parents. The use of an IOL also eliminates the onset of anisometropia and aniseikonia, which occurs particularly when the cataract is monolateral, thus achieving effective prevention of amblyopia and strabismus.

In young patients with Marfan's syndrome, the lack of adequate capsular support causes problems with IOL implantation in the posterior chamber and suggests the suitability of IOL implantation in the anterior chamber or in the posterior chamber with scleral fixation. Secondary IOL implantation in the anterior chamber commonly causes complications such as increased intraocular pressure, endothelial damage with subsequent keratitis bullous, uveitis, and pupillary block.

Several techniques are now available for scleral-fixed IOL implantation. One of these uses IOLs with positioning holes on the haptics and a double-armed Prolene suture with two passes through the sclera. The suture ends are tied together and a knot is rotated to bury it, eliminating the need for scleral flaps. This technique should eliminate IOL decentration and erosion by Prolene suture. Intracocular lens implantation with scleral fixation promises to be a reliable and virtually risk-free technique, as suggested by our trials, and has resulted in good recovery of visual acuity in our young patients.

**REFERENCES**