The Artisan aphakia intraocular lens in the paediatric eye

Sminia, M.L.

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Chapter 8

LONG-TERM FOLLOW-UP AFTER BILATERAL ARTISAN® APHAKIA INTRAOCULAR LENS IMPLANTATION IN TWO PAEDIATRIC MARFAN PATIENTS

M.L. Sminia¹, M.Th. P. Odenthal², L.J.J.M. Prick¹, J.M. Cobben³, M.P. Mourits¹, H.J. Völker- Dieben⁴

¹the Department of Ophthalmology, Academic Medical Centre, Amsterdam, the Netherlands
²the Department of Ophthalmology, Diaconessenhuis, Leiden, the Netherlands
³the Department of Paediatric Genetics, Emma Children's Hospital/ Academic Medical Centre, Amsterdam, the Netherlands
⁴the Department of Ophthalmology, Vrije Universiteit Medical Centre, Amsterdam, the Netherlands

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ABSTRACT

We present two patients with Marfan syndrome and bilateral crystalline lens dislocation who underwent bilateral lens extraction and Artisan aphakia intraocular lens (IOL) implantation in our university hospital. After a follow-up of more than 12 years a good visual outcome, no serious IOL-related complications and endothelial cell densities within the expected range for eyes without cataract surgery were found in both patients.
Zonular weakness and capsular instability are a challenge when correcting aphakia in patients with Marfan syndrome. IOL implantation can be visually rewarding in well selected cases and may improve the patient’s quality of life. We used the Artisan® (Ophtec, Groningen, the Netherlands) aphakia IOL, an iris-fixated anterior chamber lens, in two children. The clinical parameters of the four eyes reported in this paper were compared to the clinical parameters of 29 control eyes of 15 Marfan patients without a history of intraocular surgery, who visited our clinic regularly for periodical eye examinations.

**CASE 1**

In case 1 the diagnosis of Marfan syndrome was made at the age of two years, based on Ghent criteria, no *FBN1* mutation could be demonstrated. Preoperatively both eyes had a crystalline lens dislocation with a best-corrected visual acuity (BCVA) of less than 20/100 and a preoperative refractive error of about minus 20 diopters (D). The preoperative axial length was 28.5 (OD) and 28.4 millimetres (OS). At the age of nine an extracapsular lens extraction with implantation of a 7D Artisan aphakia IOL and a prophylactic encircling band were performed on the right eye. Two years later the patient underwent a prophylactic encircling band on the left eye followed one year later by extracapsular lens extraction, anterior vitrectomy and implantation of an 11D Artisan aphakia IOL. The surgeon decided to place a prophylactic encircling band to minimize the chance of retinal detachment in these axially elongated eyes, although this is not a generally accepted procedure. With a follow-up of 15.8 years (OD) and 13.3 years (OS) the manifest refraction was +2.50 +2.00 x 175 OD and +0.75 +1.00 x 160 OS, best-corrected visual acuity was 20/50 OD and 20/40 OS.

**Figure 1. Anterior segment images of case 1 and 2.**

A: An earlier model of the Artisan aphakia IOL (before 1997) in the left eye of case 1. Iris atrophy is seen at the site where the nasal claw of the IOL is attached.

B: The Artisan aphakia IOL in the right eye of case 2. Patches of radial anterior iris atrophy can be seen. These atrophic patches can be a result of multiple surgical procedures, but might also be related to the IOL. The unusual location of the peripheral iridectomy at 6 o’clock is explained by the fact that silicone oil was used as a tamponade agent at the time of pars plana vitrectomy for retinal detachment in this eye.
In case 2 the clinical diagnosis of Marfan syndrome was confirmed by a demonstration of a pathogenic $FBN1$ mutation in the third year of life. Preoperatively crystalline lens dislocation, a BCVA less than 20/200 and a refractive error of minus 10D in the right and minus 12D in the left eye were present. The preoperative axial length was 25.3 (OD) and 24.6 millimetres (OS). At the age of four the patient underwent lens extraction, anterior vitrectomy and implantation of a 21D Artisan aphakia IOL in the left eye. One year later the same procedure with implantation of a 21D Artisan aphakia IOL was performed in the right eye. At the age of five a bilateral recession of the lateral rectus muscle was performed to correct exotropia. One year later a retinal tear with a localised detachment was detected and an encircling band was placed on the right eye. Because of a recurrence of the retinal detachment, one month later, a pars plana vitrectomy, using silicone oil as a tamponade agent, was performed on this eye. The oil was removed 2.5 months later. At the age of 13 the patient received an encircling band implant for a retinal detachment in the left eye. With a follow-up of 10.3 years (OD) and 11.2 years (OS) the manifest refraction was -9.0+1.0x96 OD and -7.0+1.75x156 OS, best-corrected visual acuity was 20/50 OD and 20/25 OS.

The target postoperative refraction was emmetropia in all four eyes. After lens extraction, the remaining lens capsule was removed from the eyes using an Ocutome.

### Table 1. Clinical parameters of the 4 study eyes and 29 control eyes at the last follow-up visit

<table>
<thead>
<tr>
<th>Study eyes</th>
<th>ECD (cells/mm²)</th>
<th>CV (%)</th>
<th>Hexa (%)</th>
<th>Corneal diameter (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1 OD</td>
<td>2979</td>
<td>36</td>
<td>59</td>
<td>12</td>
</tr>
<tr>
<td>Case 1 OS</td>
<td>2741</td>
<td>43</td>
<td>52</td>
<td>12</td>
</tr>
<tr>
<td>Case 2 OD</td>
<td>2962</td>
<td>33</td>
<td>58</td>
<td>13</td>
</tr>
<tr>
<td>Case 2 OS</td>
<td>3496</td>
<td>36</td>
<td>64</td>
<td>13</td>
</tr>
<tr>
<td>Mean (SD)</td>
<td>3045 (320)</td>
<td>37 (4.5)</td>
<td>58 (5.1)</td>
<td>12.5 (0.6)</td>
</tr>
</tbody>
</table>

Control eyes n=29

<table>
<thead>
<tr>
<th>Mean</th>
<th>2905</th>
</tr>
</thead>
<tbody>
<tr>
<td>CV</td>
<td>26</td>
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<tr>
<td>Hexa</td>
<td>71</td>
</tr>
<tr>
<td>AL</td>
<td>11.7</td>
</tr>
</tbody>
</table>

| range | 2408, 3541 |
| SD    | 351 |

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| range | 2408, 3541 |
| SD    | 351 |

Table 1. Clinical parameters of the 4 study eyes and 29 control eyes at the last follow-up visit

ECD: endothelial cell density; mm: millimeter; CV: coefficient of variation of cell size; %: percentage; hexa: microscope (Topcon, Japan); mcm: micrometer; AL: axial length measured with the IOL Master (Zeiss, Germany); BSCVA: best-corrected visual acuity.

**CASE 2**

In case 2 the clinical diagnosis of Marfan syndrome was confirmed by a demonstration of a pathogenic $FBN1$ mutation in the third year of life. Preoperatively crystalline lens dislocation, a BCVA less than 20/200 and a refractive error of minus 10D in the right and minus 12D in the left eye were present. The preoperative axial length was 25.3 (OD) and 24.6 millimetres (OS). At the age of four the patient underwent lens extraction, anterior vitrectomy and implantation of a 21D Artisan aphakia IOL in the left eye. One year later the same procedure with implantation of a 21D Artisan aphakia IOL was performed in the right eye. At the age of five a bilateral recession of the lateral rectus muscle was performed to correct exotropia. One year later a retinal tear with a localised detachment was detected and an encircling band was placed on the right eye. Because of a recurrence of the retinal detachment, one month later, a pars plana vitrectomy, using silicone oil as a tamponade agent, was performed on this eye. The oil was removed 2.5 months later. At the age of 13 the patient received an encircling band implant for a retinal detachment in the left eye. With a follow-up of 10.3 years (OD) and 11.2 years (OS) the manifest refraction was -9.0+1.0x96 OD and -7.0+1.75x156 OS, best-corrected visual acuity was 20/50 OD and 20/25 OS.

The target postoperative refraction was emmetropia in all four eyes. After lens extraction, the remaining lens capsule was removed from the eyes using an Ocutome.
Postoperative topical steroid, antibiotics and mydriatics were given for a total of eight weeks. No prolonged inflammation, elevated eye pressure or IOL dislocation were encountered in any of the four eyes. At the last follow-up iridodonesis and pseudophacodonesis as well as iris transillumination defects which are characteristic for Marfan syndrome were present in all study eyes. Iris atrophy, ranging from limited to more pronounced, was observed (Figure 1).

The retina was attached in all eyes. In the 29 control eyes a partial lens dislocation was present in nine eyes, iris transillumination defects in 19 eyes and iridodonesis in nine eyes.

At the last follow-up visit corneal endothelial photographs of the four study eyes were taken. The individual endothelial cell density of the four study eyes and the 29 control eyes are presented in Figure 2. Clinical parameters of the four study eyes and all control eyes can be found in Table 1.

**DISCUSSION**

We report long-term results in four eyes of two paediatric Marfan patients, in whom we used the Artisan aphakia IOL to correct aphakia. In case 1 the surgery was performed beyond the sensitive period of visual development, which may have contributed to the
lower visual outcome in this case. Both eyes of case 2 had a retinal detachment. This is a known complication in eyes of patients with Marfan syndrome with an incidence that ranges from 5% to 25.6%, with a predilection for eyes with a lens dislocation and an increased axial length\(^1\). The eyes in both cases had a markedly long axial length, beyond the range of the axial lengths found in the control eyes.

The Artisan aphakia IOL is not approved by the United States Food and Drug Administration. For many surgeons the preferred technique in the absence of capsular support is to suture an IOL into the sulcus with scleral sutures. Late dislocation of these IOLs due to breakage of the polypropylene suture is reported in up to 24% of paediatric cases\(^3\). Long-term follow-up is very important in children to signal such late complications.

Encouraging short-term clinical results and no endothelial cell loss after Artisan aphakia IOL implantation have been reported in paediatric patient with idiopathic lens dislocation\(^4\) and with lens dislocation secondary to Marfan syndrome\(^5\). Endothelial cell loss is also found to be limited in the long-term after Artisan aphakia IOL implantation in

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**Figure 2.** The individual ECD of the study and control Marfan eyes compared to normal eyes, as a function of age.

The squares represent the 4 study eyes. The diamonds represent the ECD of the 29 Marfan control eyes. The curved line represents the mean ECD in normal eyes as a function of age, the dotted lines represent the value plus and minus two SD from the mean. The curve is created from data of the normal corneal ECD in different age groups by Möller-Pedersen et al.
More than 12 years after surgery endothelial cell densities in our cases were within the expected range for eyes without crystalline lens surgery. However, a higher coefficient of variation of cell size and a lower percentage of hexagonal cells was found, when compared to the control eyes and to normal subjects in the literature. These changes might be related to the IOL. However changes in morphology are also reported in non-operated eyes from patients with Marfan syndrome, with the most prominent changes in eyes with lens dislocation, and in both eyes of one control patient in the current study. This patient was fitted with rigid gas permeable (RGP) contact lenses to correct the refractive error, caused by dislocation of the crystalline lens out of the visual axis in both eyes. The clinical consequences of the morphological changes are unclear, but might be limited, since we found a clear cornea and a normal central corneal thickness in the four study eyes and the two eyes of the control Marfan patient with RGP contact lens wear.

We feel that the Artisan aphakia IOL may be a viable treatment option in the correction of aphakia in patients with Marfan syndrome. Larger studies on the Artisan aphakia IOL in Marfan patients are needed, to confirm our encouraging long-term clinical results and to assess the impact of the endothelial changes that we observed.
REFERENCES


