

Outcomes after lensectomy for children with Marfan syndrome

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PURPOSE	To describe the long-term incidence of retinal detachment, intra- and postoperative complications, and visual and refractive outcomes of children with Marfan syndrome following lensectomy for ectopia lentis, with or without subsequent correction of aphakia using the Artisan aphakic iris-fixated anterior chamber intraocular lens.
METHODS	The medical records of children treated at the Department of Ophthalmology, Children's University Hospital, Dublin, Ireland, from January 1, 1991, to December 31, 2011, were retrospectively reviewed. The primary outcome measure was rate of retinal detachment at final follow-up. Secondary outcomes included postoperative complications, endothelial cell count, visual acuity, refractive error, and postoperative refraction prediction error.
RESULTS	A total of 30 eyes of 15 patients underwent lensectomy. Of these, 16 eyes of 8 patients subsequently underwent Artisan implantation. Mean follow-up was 13.8 ± 5.9 years after lensectomy and 4.1 ± 2.8 years after Artisan implantation. There were no cases of retinal detachment. Best-corrected visual acuity was 0.11 ± 0.14 (logMAR) post-lensectomy and 0.12 ± 0.19 post-Artisan implantation. Endothelial cell count of the pseudophakic group was 3109 ± 458 preoperatively and 2632 ± 592 postoperatively. Mean cell loss was 15.4%. One eye required repeat surgical peripheral iridectomy for pseudophakic pupillary block. One eye required re-enclavation of a dislocated Artisan IOL haptic; the same eye required anterior vitrectomy for removal of retained lens fragment from the original lensectomy.
CONCLUSIONS	In this study cohort, there was no increased incidence of retinal detachment in children with Marfan syndrome and ectopia lentis in the setting of one or more prior intraocular procedures. A moderate rate of endothelial cell loss was observed. (J AAPOS 2016;20:247-251)

Ectopia lentis is the most common sign of ocular involvement in Marfan syndrome, affecting 50%-80% of patients, mainly between birth and age 20.¹⁻⁴ Lensectomy is performed when ectopia lentis causes unstable or progressive refractive error, with associated reduction of best-corrected visual acuity or glaucoma. Aphakia is corrected either with glasses or contact lenses, or by implantation of an anterior chamber,⁵ scleral-fixated,⁶ or iris-fixated intraocular lens (IOL).^{7,8}

Retinal detachment has been reported to occur in about 4%-24% of patients with Marfan syndrome, depending on study design, with cross-sectional studies reporting rates of

4%-10%^{4,9} and surveys reporting rates of 15%-24%.^{10,11} Previous surgery for ectopia lentis is consistently identified as a risk factor for retinal detachment in the literature. The present study aimed to assess the risk factors for postoperative retinal detachment and to report rate of endothelial cell loss in a series of children with Marfan syndrome who underwent sequential bilateral lensectomy for ectopia lentis at a single institution over a 21-year period, with or without subsequent implantation of the Artisan aphakic iris-fixated IOL (hereafter, "Artisan").

Subjects and Methods

This study was approved by the Institutional Review Board of the Children's University Hospital, Temple Street, Dublin, and complies with requirements of the US Health Insurance Portability and Accountability Act of 1996. In all cases, written informed consent for surgery was obtained from the parents/guardians.

The medical records of consecutive patients with Marfan syndrome who underwent pars plana lensectomy for nontraumatic ectopia lentis at the Children's University Hospital, Temple Street, Dublin, from January 1, 1991 through December 31, 2011, with or without subsequent implantation of the Artisan aphakic iris-fixated IOL (Ophtec BV, Groningen, The Netherlands) were

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Table 1. Follow-up of Marfan syndrome patients and interval between intraocular procedures

Follow-up	Mean \pm SD	Range
Post-lensectomy (N = 30 eyes)	13.8 \pm 5.9 yrs (3-26) ^a	3 to 26 yrs
Post-Artisan IOL implant (N = 16 eyes)	4.1 \pm 2.8 yrs ^b	9 months to 9 yrs
Interval	Median (IQR)	Range
Before contralateral lensectomy (N = 15 patients)	14 days (6-46) ^c	5 days to 1.5 yrs
Between lensectomy and Artisan IOL implant (N = 16 eyes)	8.0 yrs (4.1-10.8)	5 months to 13 yrs
Before contralateral Artisan IOL implant (N = 8 patients)	14 days (14-29) ^d	5 days to 1.6 yrs

IOL, intraocular lens; IQR, interquartile range; MFS, Marfan syndrome; SD, standard deviation.

^a22/30 >10 years.

^b12/16 \geq 2 years; 8/16 \geq 3.8 years.

^c10/15 \leq 21 days.

^d6/8 \leq 21 days.

reviewed retrospectively. All surgeries were carried out by the senior author (MOK). The diagnosis of Marfan syndrome was based on clinical criteria in the Berlin nosology¹² prior to 1997 and on clinical criteria in the Ghent nosology¹³ from 1997 onward and was made in collaboration with pediatricians.

In all patients, the indication for lensectomy was ectopia lentis causing either a reduction of best-corrected visual acuity or fluctuating refraction due to lens mobility. For patients with surgical indication for lensectomy in both eyes, the surgery was performed first in the eye with the worse visual acuity. The indications for implantation of the Artisan lens were intolerance to contact lenses or visual symptoms caused by aphakic spectacles; therefore, sequential bilateral Artisan implantation was performed. IOL power calculations were made using the Van der Heijde formula,¹⁴ according to the manufacturer's guidelines. Target refraction was emmetropia in all cases.

The following data were collected: age, sex, history of amblyopia or other visually consequential comorbidity, preoperative best-corrected visual acuity, preoperative cycloplegic refraction, anterior segment examination, dilated fundus examination, final best-corrected visual acuity, final subjective refraction. Preoperative best-corrected visual acuity was measured using a Snellen chart, bracketed HOTV letters, or line pictures as appropriate for the patient's age and ability. Postoperative best-corrected visual acuity was measured using a Snellen chart. Patients with an indication for implantation of Artisan lenses also had preoperative keratometry and anterior chamber depth measurement (IOL Master 500, Carl Zeiss Meditec, Jena, Germany) and preoperative and final endothelial cell counts (CellCheck, Model NSP 9900, Konan Medical, Irvine, CA). Contraindications to Artisan implantation included anterior chamber depth of <3.2 mm, uveitis, glaucoma, corneal endothelial cell count below 2000 cells/mm², corneal endothelial disorders, and corectopia.

Lensectomy and anterior vitrectomy was carried out through the pars plana, with an infusion cannula in the anterior chamber, as previously described.¹⁵ Subsequent Artisan implantation, in suitable patients, was carried out through a 6 mm superior corneal incision, using either an enclavation needle or enclavation forceps, as previously described.⁸ Surgery was performed under general anesthesia in all cases.

For descriptive purposes, quantitative variables were expressed as means and standard deviations, or medians and interquartile ranges, if highly skewed. Visual acuity was converted to logMAR

Table 2. Pre-lensectomy refractive error of Marfan syndrome patients^a

Mean refractive error (SE \pm SD), D	SE range, D
-3.9 \pm 9.3	-20.0 to +15.5
Myopia (eyes)	
15/20	-1.0 to -20.0
4/20	-2.5 to -6.0
5/20	-6.1 to -12.0
3/20	> -12.0

D, diopter; MFS, Marfan syndrome; SD, standard deviation; SE, spherical equivalent.

^aInformation on pre-lensectomy refractive error was ascertained from records relating to 20 of 30 study eyes.

equivalent for analysis. Numerical variables before and after surgery were compared using the *t* test for paired samples; numerical variables between groups were compared using the *t* test for unpaired samples. All statistical analyses were performed with Numbers version 2.3 (Apple Inc, Cupertino, CA). Statistical significance was defined as a *P* < 0.05 (2-tailed test).

Results

A total of 30 eyes of 15 patients (11 males) were included. The average age at lensectomy was 4.6 \pm 3.5 years (range, 1-15 years). Of these, 16 eyes of 8 patients (5 males) subsequently underwent Artisan implantation (pseudophakic group). The average age at Artisan implantation was 12.7 \pm 2.7 years (range, 9-17 years). Table 1 shows the follow-up of study patients and the interval between surgical procedures.

The mean best-corrected visual acuity before lensectomy was 0.66 \pm 0.35 (range, 0.00-1.10); after lensectomy, 0.11 \pm 0.14 (range, -0.10 to 0.48; *P* < 0.0001). The mean best-corrected visual acuity before Artisan implantation was 0.15 \pm 0.16 (range, -0.08 to 0.48); after implantation, 0.12 \pm 0.19 (range, 0.00-0.60; *P* = 0.52). There was no difference in postoperative best-corrected visual acuity between the aphakic and pseudophakic groups (*P* = 0.40). Of the 30 eyes, 28 had final best-corrected visual acuity of 0.18 or better. The remaining 2 eyes had preexisting amblyopia. Table 2 provides the preoperative refractive error of the study patients.

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