Outcomes of Iris-Claw Anterior Chamber versus Iris-Fixated Foldable Intraocular Lens in Subluxated Lens Secondary to Marfan Syndrome

Denise E. Hirashima, MD, Eduardo S. Soriano, MD, Rodrigo L. Meirelles, MD, Gustave N. Alberti, MD, Walton Nosé, MD

Purpose: To compare the outcome of phacoemulsification using 2 different iris-fixation techniques for intraocular lens (IOL) replacement, a foldable posterior chamber IOL (PCIOL; AcrySof MA60AC, Alcon Laboratories Inc, Fort Worth, TX) and an iris-claw anterior chamber IOL (ACIOL; Artisan, Ophtec BV), for treatment of subluxated lenses in patients with Marfan syndrome (MFS).

Design: Randomized, controlled trial.

Participants: A total of 31 eyes of 16 patients with subluxated lenses associated with MFS and a preoperative corrected visual acuity (CVA) ≤20/40 based on the Early Treatment Diabetic Retinopathy Study chart.

Methods: Patients were enrolled and the eye with worse visual acuity was randomly assigned to 1 of 2 study arms: phacoemulsification and iris-fixated PCIOL or phacoemulsification and iris-claw ACIOL; the second eye of the same patient received the other IOL type. Preoperative and postoperative ophthalmologic examination, optical coherence tomography, and endothelial cell counts were performed.

Main Outcome Measures: We recorded CVA results at 3, 6, and 12 months, complications, endothelial cell loss, and central retinal thickness.

Results: In the iris-fixated PCIOL group, CVA was significantly improved at 3 ($P = 0.011$; n = 16), 6 ($P = 0.006$; n = 16), and 12 months ($P = 0.002$; n = 16). In the iris-claw ACIOL group, CVA was significantly improved at 3 ($P = 0.001$; n=15), 6 ($P = 0.001$; n = 15), and 12 months ($P = 0.009$; n = 12). The CVA results did not differ significantly between groups. Dislocation of the IOL occurred in 3 of 16 (18.75%) eyes in the PCIOL group. Retinal detachment occurred in 3 eyes (2 in the PCIOL group and 1 in the ACIOL group) and was successfully repaired. Postoperative foveal tomograms in both groups revealed a decrease in the mean foveal thickness (MFT; ≤172 μm) in 54.16% of the patients.

Conclusions: The iris-sutured PCIOL and iris-claw ACIOL produced comparable improvements in CVA at 3, 6, and 12 months postoperatively. Although IOL dislocation tended to occur more frequently in the iris-fixated PCIOL group, the difference was not significant. At 6 months postoperatively, all study patients tended to have a thinner MFT. None of the patients in either group developed cystoid macular edema.

Financial Disclosure(s): The authors have no proprietary or commercial interest in any of the materials discussed in this article. *Ophthalmology 2010;117:1479–1485 © 2010 by the American Academy of Ophthalmology.*

Marfan syndrome (MFS) was first described in 1896 by Antoine-Bernard Marfan.[1] An inherited disorder of the connective tissue, MFS is caused by a mutation of the extracellular matrix protein fibrillin 1 (15q21.1).[2–5] The incidence of classic MFS is approximately 2 to 3 per 10 000 individuals.[6] It is a multisystem disorder with manifestations typically involving the skeletal, ocular, and cardiovascular systems.[7] Diagnosis and management benefit from a multidisciplinary assessment, generally including a geneticist, ophthalmologist, and cardiologist.[6,8]

Ocular involvement in MFS is subdivided into major and minor criteria. Ectopia lentis of any degree is a major criterion. At least 2 of the minor criteria must be present for consideration as ocular involvement: an abnormally flat cornea, increased axial length, and hypoplasia of the ciliary muscle or iris, causing decreased mydriasis. Mean keratometry values <42 diopters (D) are highly correlated with a diagnosis of MFS.[9] Other secondary manifestations are early severe myopia, retinal detachment, early cataracts, glaucoma, and amblyopia.[6,8] Ectopia lentis occurs in 50% to 80% of patients with MFS. Management initially involves optical refraction but may require lens removal when visual acuity becomes uncorrectable or the refractive status is unstable.[10–12]

Ectopia lentis is likely caused by a structural deficiency of FBN1; histologic studies reveal reduced fibrillin at the equator and fewer zonular fibers in MFS patients.[6,13,14] Operative correction poses an interesting challenge. Zonular weakness and ectopic capsules complicate phacoemulsification and lens implantation procedures. Since the publica-

ISSN 0161-6420/10/$–see front matter
doi:10.1016/j.ophtha.2009.12.043

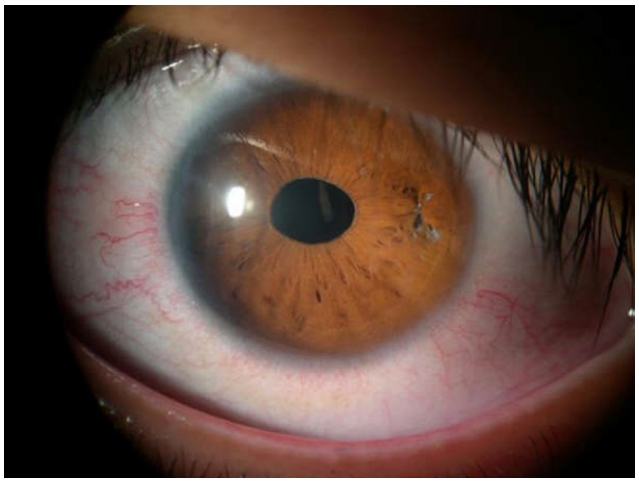

Figure 1. Iris-fixated posterior chamber foldable intraocular lens.

tion of a large series of cases on surgery for ectopia lentis by Jarrett in 1967,[15] several operative approaches have been used. No large-scale studies to date, however, have evaluated the efficacy and complication rates of the various surgical options available.

In the present study, we have evaluated the outcome and complication rate of 2 different surgical approaches used for the correction of lenticular subluxation in MFS: phacoemulsification with an iris-fixated posterior chamber intraocular lens (PCIOL) or an iris-claw anterior chamber IOL (ACIOL) implantation.

Materials and Methods

The present study was an interventional, randomized, controlled trial performed at the Federal University of Sao Paulo. University Ethics Committee approval and informed consent were obtained. The study was performed according to the tenets of the Declaration of Helsinki.

Participants

Inclusion criteria were patients with a diagnosis of MFS, referred after genetic and cardiologic evaluation in the University Marfan Syndrome Group Study with ectopia lentis and a corrected visual acuity (CVA) ≤20/40 based on the Early Treatment Diabetic Retinopathy Study (ETDRS) logarithmic chart (Lighthouse Inc., New York, NY).

Patients were enrolled and the eye with worse visual acuity was randomly assigned to 1 of 2 study arms: phacoemulsification and PCIOL (AcrySof MA60AC, Alcon Laboratories, Inc, Fort Worth, TX) group or phacoemulsification and ACIOL (Artisan, Ophtec BV, Groningen, The Netherlands). The second eye of the same patient received the other IOL type. Patients <15 years of age were excluded.

Intervention

The patients underwent a full ophthalmologic examination, including, CVA, keratometry, pachymetry, slit-lamp evaluation, intraocular pressure measurement, posterior segment evaluation, endothelial cell count (Topcon SP-2000P, Topcon Corporation, Tokyo, Japan), and retinal central thickness using macular optical coherence tomography (OCT; Stratus OCT, Carl Zeiss Ophthalmic Systems, Inc., Humphrey Division, Dublin, CA).

Initially, the foveal thickness was measured with OCT2, but the equipment was changed to an OCT3 (Stratus, Carl Zeiss Meditec, Dublin, CA) in the middle of the study. The data from the OCT2 were excluded and only data from the OCT3 (Stratus) were used in the analysis. The standard fast macular thickness scan protocol was selected to obtain 6 consecutive macular scans centered on the fovea, equally spaced 30° apart. The mean foveal thickness (MFT) was defined as the average thickness of the central sector with a 1000-μm diameter, which was measured both preoperatively and postoperatively. Data with a signal strength of no less than 4 and/or a center point standard deviation of ≥20 μm were excluded.

When measurements using an optical biometer (IOL Master, Carl Zeiss Meditec, Jena, Germany) were not possible due to lens opacities or a subluxated lens, biometry was performed using an immersion technique with an ultrasonic biometer (OcuScan, Alcon).

Eyes selected to receive the iris-claw ACIOL underwent peripheral iridotomy using a neodymium–yttrium aluminum garnet (YAG) laser (YAG 3000LE, Alcon) before surgery.

Operative Technique

Two surgeons performed the procedures (WN, DH). After intravenous sedation and topical 1% tetracaine or per ocular ropivacaine injection, a corneal incision was made temporally in the right eye and nasally in the left eye with a disposable 2.75-mm keratome, and a 1.0-mm peripheral paracentesis was created 180° away from the main corneal incision. In patients with topical anesthesia, preservative-free 1.0% lidocaine hydrochloride was injected into the anterior chamber. The chamber was then filled with dispersive viscoelastic (Viscoat Laboratories, Inc, Alcon Laboratories, Fort Worth, TX) to protect the endothelial cells and tamponade the vitreous behind the elongated zonular fibers. Sodium hyaluronate (1.0% Provisc Laboratories, Inc, Alcon Laboratories) was then used to deepen the anterior chamber. Capsulorrhexis was performed using Utrata forceps, followed by hydrodissection. Aspiration or phacoemulsification was performed slowly. Carbachol and 1.0% sodium hyaluronate were then placed in the anterior chamber.

The iris-fixated PCIOL was folded to create a "moustache" haptic configuration in a Buratto inserter. The lead haptic was

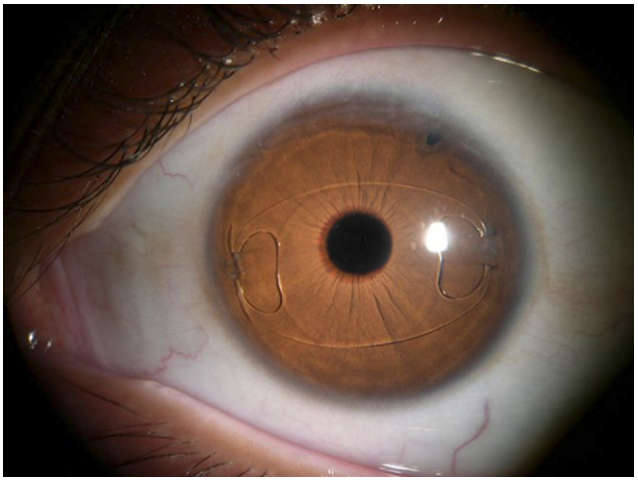

Figure 2. Iris-claw anterior chamber intraocular lens.

compressed gently against the edge of the optic as the IOL entered the incision. The IOL was slowly unfolded with both haptics projecting through the pupil, while the optic was held just above the iris plane supported by a spatula until completely captured by the pupil and stabilized.[16] The haptics were placed at the 3 and 9 o'clock meridians.

To fixate each haptic to the peripheral iris, a 10.0 polypropylene suture on a long curved needle (PC-7; Alcon Laboratories, Inc) was passed through the cornea, through the peripheral iris, behind the haptic, and out through the iris and peripheral cornea, perpendicular to the haptic position. Both suture ends were retrieved through the main corneal and paracentesis incisions using a Hirschman iris hook. The knot was tied with a double throw, and slid down onto the haptic, followed by a third throw to secure the knot. The maneuver was repeated to lay down a second, modified McCannel iris suture on top of the first.[17] After performing the same procedure with the second haptic, the optic was prolapsed into the posterior chamber, and the viscoelastic was removed (Fig 1).

For implantation of the iris-claw ACIOL, a 5.2-mm corneal incision at 12 o'clock and 2 paracenteses at 2 and 10 o'clock were created to introduce the IOL and enclavation needles, respectively. Carbachol was injected to constrict the pupil. The anterior chamber was filled with high viscosity viscoelastic to create space and protect the endothelium. The iris-claw ACIOL was introduced with the haptics at 3 and 9 o'clock centered on the pupil. While securely holding the lens body with forceps, an enclavation needle was used to fixate the IOL at the iris midperiphery (Fig 2).

An anterior vitrectomy was minimally performed in both techniques when the vitreous prolapsed the anterior chamber and after IOL fixation to remove the capsular bag and viscoelastic.

The primary postoperative outcome was CVA at 3, 6, and 12 months. The secondary postoperative outcomes were endothelial cell loss, macular thickness with OCT, and complications.

Statistical Methods

For descriptive purposes, qualitative variables are expressed as percentages and quantitative data are expressed as mean and standard deviation. The Student *t* test was used to compare baseline data. A Wilcoxon test was used to compare postoperative CVA with baseline values within each group, and a Mann–Whitney *U* was used to compare CVA between IOL groups. Analysis of variance was used to compare endothelial cell count within each group and between groups. The Fisher test was used to compare the percentage of eyes with CVA that had improved by 3 lines, remained stable (within 3 lines), or decreased by 3 lines on an ETDRS chart at 3, 6, and 12 months between groups. The Fisher test was also used to compare groups at each time point (preoperatively, and 3, 6, and 12 months postoperatively) with regard to the percentage of eyes in the range of ≥20/40, ≤20/50, and ≤20/200, and to compare MFT. The Kaplan-Meier curve was used to compare complications between groups. $P<0.05$ was considered significant.

Sample Size

We included 31 eyes with a significance level of 5% and 13% power for a variable percentage of eyes with CVA improvement of >3 lines, stable (within 3 lines), or a CVA decrease of >3 lines on an ETDRS chart.

Results

Between May 2001 and June 2006, 31 eyes of 16 patients (8 males, 8 females) with MFS were enrolled in the study. The average age at the time of surgery was 25 years (range, 15–40). Mean keratometry of the 31 eyes was 40.18±1.89 D. The lowest mean keratometer reading was 37.00 D; 70.96% of patients had mean keratometer readings ≤42 D (22/31). Mean central corneal thickness was 521±61 μm and mean axial length was 25.47±2.14 mm. The longest eye measurement was 32.16 mm. One patient exhibited a microcornea. Phacoemulsification was performed in all but 1 patient owing to extension of the subluxation. In this case, the lens was removed intact through a large limbal incision using a loop hook without complications.

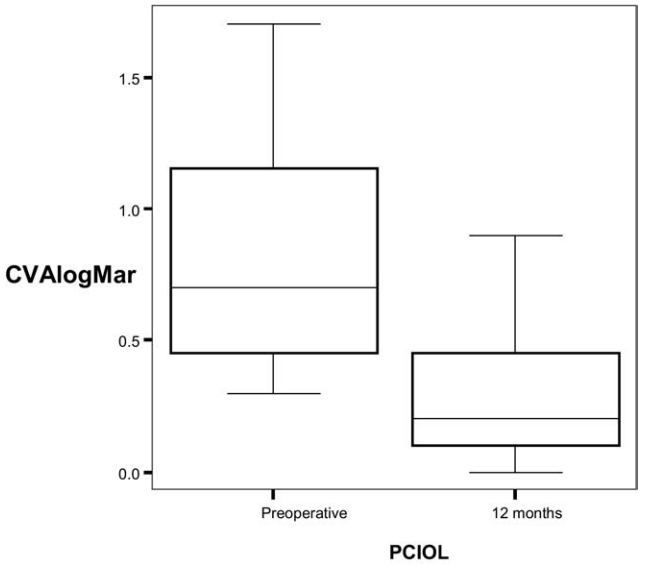

Figure 3. Box plot of iris-fixated posterior chamber intraocular lens (PCIOL) corrected visual acuity (CVA; logMar) preoperative and 12 months postoperatively. logMar = logarithm of the minimal angle of resolution.

The PCIOL group comprised 16 eyes of 16 patients with a mean age of 26 years (range, 16–22). Compared with the baseline values, visual acuity was improved at 3, 6, and 12 months postoperatively. The preoperative CVA mean of 0.83±0.46 (range, 0.30–1.7) improved to 0.39±0.38 (range, 0.10–0.30; $P = 0.001$; n = 16) at 3 months, 0.33±0.40 (range, 0.00–0.20; $P = 0.006$; n = 16) at 6 months, and 0.29±0.26 (range, 0.00–0.90; $P = 0.002$; n = 16) at 12 months (Fig 3).

The ACIOL group included 15 eyes of 15 patients with a mean age of 25 years (range, 16–40). Compared with the baseline values, visual acuity was improved at 3, 6, and 12 months postoperatively. The preoperative CVA mean of 0.87±0.51 (range, 0.3–1.7; n = 15) improved to 0.41±0.42 (range, 0.1–1.7; $P = 0.001$; n = 15) at 3 months, 0.39±0.43 (range, −0.1 to 1.7; $P = 0.001$; n = 15) at 6 months, and 0.48±0.61 (range, −0.1 to 1.7; $P = 0.009$; n = 12) at 12 months (Fig 4).

To evaluate the effects of the 2 procedures based on the ETDRS chart, we compared the percentages of eyes with 3 lines improvement, eyes with no change in visual acuity (within 3 lines), and eyes with >3 lines visual acuity decrease at 3, 6, and 12 months. There was no difference between groups at any time (Table 1). Further, the percentages of eyes with CVA ≥20/40, ≤20/50, and ≤20/200 on the ETDRS chart preoperatively and at 3, 6, and 12 months postoperatively did not differ between groups (Table 2).

Among all patients in the study, the proportion of legally blind eyes (≤20/200) changed from 35% (11 eyes) before surgery to 6.45% (2 eyes) 6 months after surgery. Of the 2 eyes with a CVA

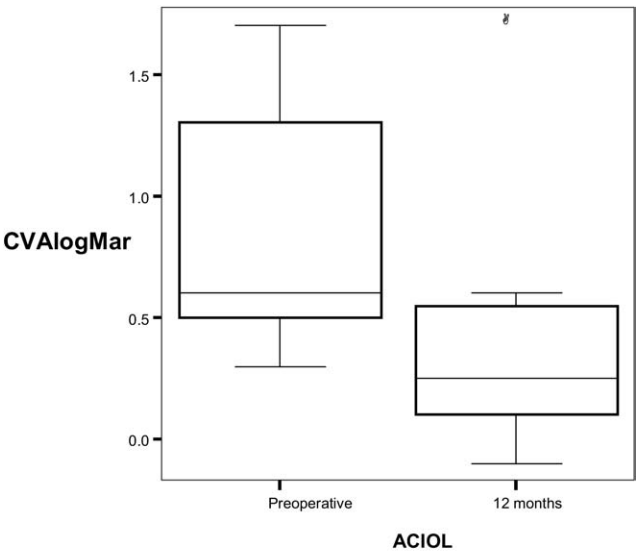

Figure 4. Box plot of iris-claw anterior chamber intraocular lens (ACIOL) corrected visual acuity (CVA; logMar) preoperative and 12 months postoperatively. logMar = logarithm of the minimal angle of resolution.

of ≤20/200, one was severely amblyopic and the other had a retinal detachment that was successfully repaired.

Complications in the PCIOL group included transient ocular hypertension (n = 1), iris atrophy (n = 5), retinal detachment that was successfully repaired (n = 2), IOL capture (n = 1), and IOL dislocation (n = 3). In 2 cases, the dislocated IOL haptics were sutured using the same technique, and good visual acuity and position were ultimately achieved. At 6 months postoperatively, 1 patient achieved a CVA of 20/40 and the other patient a CVA of 20/30. Another patient who experienced blunt trauma to the face, which caused the lens to dislocate to the posterior vitreous, underwent posterior vitrectomy and IOL scleral fixation by another service; the CVA after surgery was 20/60.

Table 1. Visual Acuity Changes According to Early Treatment Diabetic Retinopathy Study Lines at Each Follow-up Visit in Relation to the Preoperative Values

Visual Acuity Changes	Iris-Fixated Posterior Chamber Intraocular Lens, Total (%)	Iris-Claw Anterior Chamber Intraocular Lens, Total (%)	*P* Among the Groups
3 Months			
Improvement >3 lines	11 (68.75%)	11 (73.33%)	
Stable within 3 lines	4 (25.0%)	4 (26.67%)	0.616
Decrease >3 lines	1 (6.25%)	0 (0.0%)	
Total (n)	16	15	
6 Months			
Improvement >3 lines	12 (75.0%)	11 (73.33%)	
Stable within 3 lines	3 (18.75%)	4 (26.67%)	0.561
Decrease >3 lines	1 (6.25%)	0 (0.0%)	
Total (n)	16	15	
12 Months			
Improvement >3 lines	12 (75.0%)	8 (66.67%)	
Stable within 3 lines	3 (18.75%)	4 (33.33%)	0.497
Decrease >3 lines	1 (6.25%)	0 (0.0%)	
Total (n)	16	12	

Table 2. Comparison between Iris-Fixated Posterior Chamber Intraocular Lens and Iris-Claw Anterior Chamber Intraocular Lens of the Percentages of Eyes with a Corrected Visual Acuity of ≥20/40, ≤20/50, and ≤20/200 Preoperatively and at 3, 6, and 12 Months Postoperatively

Range	Iris-Fixated Posterior Chamber Intraocular Lens, Total (%)	Iris-Claw Anterior Chamber Intraocular Lens, Total (%)	*P* Value
Preoperative			
≥20/40	1 (6.25%)	2 (13.33%)	
≤20/50	10 (62.50%)	7 (46.67%)	0.631
≤20/200	5 (31.25%)	6 (40.0%)	
Total (n)	16	15	
3 Months			
≥20/40	9 (56.25%)	10 (66.67%)	
≤20/50	6 (37.50%)	4 (26.67%)	0.810
≤20/200	1 (6.25%)	1 (6.66%)	
Total (n)	16	15	
6 Months			
≥20/40	11 (68.75%)	10 (66.67%)	
≤20/50	4 (25.0%)	4 (26.67%)	0.992
≤20/200	1 (6.25%)	1 (6.66%)	
Total (n)	16	15	
1 Year			
≥20/40	10 (62.50%)	7 (58.33%)	
≤20/50	6 (37.50%)	3 (25.0%)	0.221
≤20/200	0 (0.0%)	2 (16.67%)	
Total (n)	16	12	

Complications in the ACIOL group included transient ocular hypertension (n = 1), iris atrophy at the enclavation site (n = 5), retinal detachment that was successfully repaired (n = 1), pupillary block (n = 1), and IOL pigment deposition (n = 1).

The mean endothelial cell density in both groups was significantly lower after surgery at 6 ($P = 0.003$) and 12 ($P<0.001$) months. Endothelial cell loss did not differ between groups. The mean cell density in the PCIOL group was 3109.3±496.5 (n = 14) preoperatively and 2797.7±279.5 (n = 14) and 2808±202.9 (n = 5) at 6 and 12 months postoperatively, respectively (Fig 3). Two patients with retinal detachment were excluded. The PCIOL group had 10% endothelial cell density loss 6 months after surgery (Fig 5).

The mean cell density in the ACIOL group was 3178.2±419.4 (n = 14) preoperatively, and 2933.6±254.1 (n = 14) and 2734.1±290.7 (n = 7) at 6 and 12 months postoperatively, respectively. The results of the endothelial cell count of the 1 patient with retinal detachment after IOL implantation were also excluded. The ACIOL group endothelial cell density loss after 6 months was 8% (Fig 3).

Postoperative MFT was 170±21 (n = 12) in the PCIOL group and 173±25 (n = 12) in the ACIOL group. There was no difference between groups. Subluxated lenses and cataracts affect OCT image quality. Because preoperative measurements were obtained in only 7% of the eyes in the PCIOL group and in 20% eyes in the ACIOL group, these measurements were not compared. Three eyes were diagnosed with retinal detachment after surgery on postoperative days 6, 7, and 23; tomograms of these eyes were excluded. Postoperative tomograms indicated that 54.16% (13/24) eyes had an MFT <172 μm. Clinical cystoid macular edema was not diagnosed in either group for up to 6 months postoperatively.

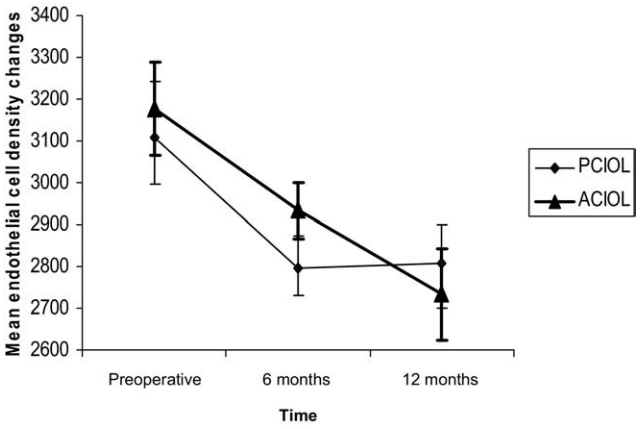

Figure 5. Mean endothelial cell density changes in the iris-fixated posterior chamber intraocular lens (PCIOL) and iris-claw anterior chamber intraocular lens (ACIOL) group.

Discussion

Since Jarrett's publication of a large series of cases (n = 166) on surgery for lens subluxation 1967,[15] various operative techniques and results have been reported for subluxated lenses and aphakia correction in patients with MFS. Nevertheless, the management of ectopia lentis remains challenging and certain controversies remain unresolved.[12,15]

Surgical options include a pars plana or limbal approach lensectomy,[12,18–21] intracapsular cataract extraction,[22,23] lens irrigation and aspiration,[24–27] and phacoemulsification with a Cionni-modified capsular tension ring.[28] The surgeon must then choose between aphakia, which was the most common selection in the early 1990s,[19,20,24,25] and IOL implantation, for which there are also many options: anterior chamber open loop IOL,[12,21] anterior chamber iris claw,[26] posterior chamber sclera-fixated,[22,23] or iris-fixated.[27] A recent metaanalysis of the safety and efficacy of open-loop anterior chamber, scleral-sutured posterior chamber, and iris-sutured posterior chamber IOLs for correction of aphakia in eyes without adequate capsular support revealed no differences between them.[29]

In the present study, we managed 31 subjects using an anterior chamber lens approach and IOL implantation with either an iris-fixated foldable PCIOL or an iris-claw ACIOL in patients with MFS. The CVA at 3, 6, and 12 months postoperatively improved in both groups to a favorable visual outcome, similar to previous reports. Kazemi et al[12] described 36 consecutive cases retrospectively reviewed in 4 categories of lens subluxation, including 14 patients with MFS treated by pars plana lensectomy-vitrectomy with an anterior chamber open loop flexible IOL. The average visual acuity in all 4 groups improved from a preoperative score of 20/163 to 20/36.[12] Morrison et al[21] reviewed 8 eyes of pediatric patients with MFS that underwent pars plana vitrectomy, lensectomy, and primary ACIOL placement in which best CVA improved from a preoperative score of 20/80 to 20/32.

The number of eyes with legal blindness owing to retinal detachment and amblyopia was reduced in the present study from 35% (n = 11) preoperatively to 6.45% (n = 2) postoperatively. In 1980, Maumenee[10] reported a visual acuity of ≤20/200 after treatment in 9% of patients with MFS. The most frequent causes of limited vision in that study were retinal detachment without lens surgery, followed by amblyopia, retinal detachment after lens surgery, and glaucoma.[10]

In our PCIOL (n = 16) group, CVA improved by ≥3 lines on the ETDRS chart in 75% of the patients at 6 months postoperatively. In the ACIOL (n = 15) group, CVA improved ≥3 lines in 73.33% of the patients at 6 months postoperatively (Table 1). Improved CVA was also reported in other studies: Halpert and BenEzra[20] described 59 eyes with subluxated lenses (27 eyes in patients with MFS) that underwent pars plana lensectomy combined with anterior vitrectomy. After surgery, 88% of the patients achieved an improvement of ≥2 lines on the Snellen chart.

Although a quality-of-life questionnaire was not administered as part of this study, most of our patients reported better quality of life after surgery. Seven patients returned to school, 1 patient was able to return to work, and 5 patients >18 years old were able to walk independently after surgery.

Complications in both groups were similar, including transient ocular hypertension, iris atrophy at the enclavation site, retinal detachment that was successfully repaired, and IOL pigment deposition. In the ACIOL group, a pupillary block occurred in 1 patient despite previous laser iridotomy, which was successfully treated with surgical iridectomy. In the PCIOL, there was both IOL capture and IOL dislocation.

The PCIOL group had a higher rate of IOL dislocation (3 eyes) compared with the ACIOL group (0 eyes), although this differences was not significant. In the iris-fixated PCIOL group, IOL dislocation generally seemed to occur later; the dislocations occurred 14, 24, and 30 months postoperatively. One occurred after a blunt trauma to the face, and the 2 others occurred after haptic slippage through a 3 o'clock suture, temporally in the left eye. It is likely that with IOL rotation the haptic slipped through the suture and dislocated the lens, although it remained fixated. We believe that fixation of the IOL haptics at 12 and 6 hours will avoid this decentration. Kaiura et al[30] described haptic slippage in 3 of 4 cases of iris-sutured IOLs.

Iris-claw ACIOL implantation after phacoemulsification for a subluxated lens has been successfully performed since its first description by Gabor.[31] Although there is a slight learning curve, the advantages include a closed globe with controlled intraocular pressure during the procedure, secure wound closure, negligible astigmatism, and minimal operative trauma.[31] One possible complication is the potential for increased endothelial cell loss.

A multicenter study of the Artisan phakic IOL suggested that corneal endothelial cell loss is stabilized to a physiologically normal level after 3 years.[32] Because our follow-up was too short to evaluate endothelial cell loss, we cannot infer much about endothelial damage from our findings. The cell loss we measured at 6 months postoperatively, however, was close to that expected secondary to operative trauma (PCIOL, 10%; ACIOL, 8%); to date, none of our patients have presented with corneal edema.

The patient in the ACIOL group with retinal detachment had the same complication in the fellow eye, which underwent the iris-fixated PCIOL procedure. Individuals with MFS are more predisposed to retinal detachment. The most significant risk factors include increased axial length, positive family history, prior ocular surgery, and dislocated lens.[6,10]

In the PCIOL group, complications included 2 eyes with retinal detachment, which was successfully repaired—one was the patient described and the other occurred in the eye with the longest axial length in the study (32.16 mm). Complications of iris-fixated PCIOL reported in the literature are glaucoma escalation, IOL dislocation, graft failure, and cystoid macular edema.[33]

It was difficult to obtain OCT before surgery in a majority of the patients in the present study owing to subluxated lenses, opacities, and fixation loss. Therefore, we could not compare preoperative and postoperative results. Image quality in OCT is reduced by cataract. Macular thickness measurements are slightly increased after uneventful cataract surgery.[34,35]

Chan et al[36] reported an MFT of 212±20 μm in 37 healthy subjects as measured by OCT3, and proposed that a foveal thickness >252 μm indicates macular thickening and a foveal thickness <172 μm indicates macular thinning. Based on these proposed measurements, the MFT of the PCIOL (n = 12) and ACIOL (n = 12) groups in our study at 6 months postoperatively indicated no foveal thickening. Between the 2 groups, 13 (54.16%) of the postoperative tomograms showed a value < 172 μm. We did not observe any evidence of clinical cystoid macular edema. The OCT findings suggested that patients with MFS generally have a thinner MFT compared with normal eyes.

One limitation of the present study was the small sample size, which restricts our ability to definitively interpret these data. Nevertheless, the findings of the present study indicate that both iris-claw and iris-fixated IOL implantation procedures produce similar results in terms of visual acuity improvement at 3, 6, and 12 months in patients with ectopia lentis secondary to MFS. The complication rates, most notably IOL dislocation, however, were more common in the PCIOL group. Endothelial cell loss was comparable between the 2 groups. Another limitation of the present study, however, is that we do not have any long-term data regarding the safety of these 2 procedures.

Further, large-scale studies are needed to help elucidate the safety and efficacy of different surgical techniques for ectopia lentis correction in patients with MFS.

References

1. Marfan AB. Un cãs déformation congénitale des quatre membres, plus prononcée aux extrémités, caractérisée par l'allongement dês os avec un certain degré d'aminicissement. Bull Mem Soc Méd Hop Paris 1896;13:220–6.
2. Sakai LY, Keene DR, Engvall E. Fibrillin, a new 350-kD glycoprotein, is a component of extracellular microfibrils. J Cell Biol 1986;103:2499–509.
3. Hollister DW, Godfrey M, Sakai LY, Pyeritz RE. Immunohistologic abnormalities of microfibrillar-fiber system in the Marfan syndrome. N Engl J Med 1990;323:152–9.
4. Dietz HG, Cutting GR, Pyeritz RE, et al. Marfan syndrome caused by a recurrent de novo missense mutation in the fibrillin gene. Nature 1991;352:337–9.
5. Kainulainen K, Pulkkinen L, Savolainen A, et al. Location on chromosome 15 of the gene defect causing Marfan syndrome. N Engl J Med 1990;323:935–9.
6. Judge DP, Dietz HC. Marfan's syndrome. Lancet 2005;366:1965–76.
7. Pyeritz RE, McKusick VA. The Marfan syndrome: diagnosis and management. N Engl J Med 1979;300:772–7.
8. De Paepe A, Devereux RB, Dietz HC, et al. Revised diagnostic criteria for the Marfan syndrome. Am J Med Genet 1996;62:417–26.
9. Heur M, Costin B, Crowe S, et al. The value of keratometry and central corneal thickness measurements in the clinical diagnosis of Marfan syndrome. Am J Ophthalmol 2008;145:997–1001.
10. Maumenee IH. The eye in the Marfan syndrome. Trans Am Ophthalmol Soc 1981;79:684–733.
11. Nelson LB, Maumenee IH. Ectopia lentis. Surv Ophthalmol 1982;27:143–60.
12. Kazemi S, Wirostko WJ, Sinha S, et al. Combined pars plana lensectomy-vitrectomy with open-loop flexible anterior chamber intraocular lens (AC IOL) implantation for subluxated lenses. Trans Am Ophthalmol Soc 2000;98:247–51.
13. Mir S, Wheatley HM, Hussel IE, et al. A comparative histologic study of the fibrillin microfibrillar system in the capsule of normal subjects and subjects with Marfan syndrome. Invest Ophthalmol Vis Sci 1998;39:84–93.
14. Traboulsi EI, Whittum-Hudson JA, Mir SH, Maumenee IH. Microfibril abnormalities of the lens capsule in patients with Marfan syndrome and ectopia lentis. Ophthalmic Genet 2000;21:9–15.
15. Jarrett WH II. Dislocation of the lens: a study of 166 hospitalized cases. Arch Ophthalmol 1967;78:289–96.
16. Condon GP. Simplified small-incision peripheral iris fixation of an AcrySof intraocular lens in the absence of capsule support. J Cataract Refract Surg 2003;29:1663–7.
17. McCannel MA. A retrieval suture idea for anterior uveal problems. Ophthalmic Surg 1976;7:98–103.
18. Peyman GA, Raichand M, Goldberg MF, Ritacca D. Management of subluxated and dislocated lenses with vitrophage. Br J Ophthalmol 1979;63:771–8.
19. Hakin KN, Jacobs M, Rosen P, et al. Management of the subluxed crystalline lens. Ophthalmology 1992;99:542–5.
20. Halpert M, BenEzra D. Surgery of hereditary subluxated lens in children. Ophthalmology 1996;103:681–6.
21. Morrison D, Sternberg P Jr, Donahue S. Anterior chamber intraocular lens (ACIOL) placement after pars plana lensectomy in pediatric Marfan syndrome. J AAPOS 2005;9:240–2.
22. Omulecki W, Nawrocki J, Palenga-Pydyn D, Sempinska-Szewczyk J. Pars plana vitrectomy, lensectomy, or extraction in transscleral intraocular lens fixation for the management of dislocated lenses in a family with Marfan's syndrome. Ophthalmic Surg Lasers 1998;29:375–9.
23. Siganos DS, Siganos CS, Popescu CN, Margaritis VN. Clear lens extraction and intraocular lens implantation in Marfan's syndrome. J Cataract Refract Surg 2000;26:781–4.
24. Maumenee AE, Ryan SJ. Aspiration technique in the management of the dislocated lens. Am J Ophthalmol 1969;68:808–11.
25. Seetner AA, Crawford JS. Surgical correction of lens dislocation in children. Am J Ophthalmol 1981;91:106–10.

26. Lifshitz T, Levy J, Klemperer I. Artisan aphakic intraocular lens in children with subluxated crystalline lenses. J Cataract Refract Surg 2004;30:1977–81.
27. Dureau P, de Laage de Meux P, Edelson C, Caputo G. Iris fixation of foldable intraocular lenses for ectopia lentis in children. J Cataract Refract Surg 2006;32:1109–14.
28. Konradsen T, Kugelberg M, Zetterström C. Visual outcomes and complications in surgery for ectopia lentis in children. J Cataract Refract Surg 2007;33:819–24.
29. Wagoner MD, Cox TA, Ariyasu RG, et al; Ophthalmic Technology Assessment Committee Anterior Segment Panel. Intraocular lens implantation in the absence of capsular support: a report by the American Academy of Ophthalmology. Ophthalmology 2003;110:840–59.
30. Kaiura TL, Seedor JA, Koplin RS, et al. Complications arising from iris-fixated posterior chamber intraocular lenses. J Cataract Refract Surg 2005;31:2420–2.
31. Gabor R. Artisan IOL after phacoemulsification in subluxated lenses [letter]. J Cataract Refract Surg 2002;28:2064.
32. Budo C, Hessloehl JC, Izak M, et al. Multicenter study of the Artisan phakic intraocular lens. J Cataract Refract Surg 2000;26:1163–71.
33. Schein OD, Kenyon KR, Steinert RF, et al. A randomized trial of intraocular lens fixation techniques with penetrating keratoplasty. Ophthalmology 1993;100:1437–43.
34. von Jagow B, Ohrloff C, Kohnen T. Macular thickness after uneventful cataract surgery determined by optical coherence tomography. Graefes Arch Clin Exp Ophthalmol 2007;245:1765–71.
35. van Velthoven ME, van der Liden MH, de Smet MD, et al. Influence of cataract on optical coherence tomography image quality and retinal thickness. Br J Ophthalmol 2006;90;1259–62.
36. Chan A, Duker JS, Ko TH, et al. Normal macular thickness measurements in healthy eyes using Stratus optical coherence tomography. Arch Ophthalmol 2006;124:193–8.

Footnotes and Financial Disclosures

Originally received: April 4, 2009.
Final revision: December 24, 2009.
Accepted: December 29, 2009.
Available online: May 13, 2010. Manuscript no. 2009-466.

Federal University of Sao Paulo (UNIFESP), Sao Paulo, Brazil.

Financial Disclosure(s):
The authors have no proprietary or commercial interest in any of the materials discussed in this article.

Correspondence: Denise E. Hirashima, MD, R. Mussumés, 378 CEP02130-070 São Paulo-SP Brazil. E-mail: dehirashima@me.com.