Long Term Follow-up of Retropupillary Fixation of Iris-clipped Intraocular Lens for Children with Inadequate Capsular Support

Alvina Pauline D. Santiago, MD[1,2,3] and Noel S. Cariño, MD[1]

[1]*Department of Ophthalmology and Visual Sciences, College of Medicine and Philippine General Hospital, University of the Philippines Manila*
[2]*The Medical City, Pasig City*
[3]*St. Luke's Medical Center, Quezon City*

ABSTRACT

Children with weak or absent capsular support requiring lens removal and intraocular lens implantation present as one of the more challenging cases in pediatric ophthalmology practice. The authors present a case series with postoperative follow-up of at least five years after retropupillary fixation of iris-clipped lenses. All cases had improved visual acuity with only minor focal iris atrophy as complication in one case. Retropupillary fixation of iris-clipped IOL should be included in an ophthalmologist's armamentarium when operating on cases with weak or absent capsular support.

Keywords: iris-clipped intraocular lens, lens subluxation, retropupillary fixation

INTRODUCTION

Children with weak or absent capsular support requiring lens removal, either because of trauma, subluxation, or genetic abnormalities affecting the zonules present a predicament to the ophthalmologist as to the optimal technique of intraocular lens (IOL) implantation. In previous years, transscleral fixation has been used but carries an increased risk of intraocular hemorrhage, choroidal effusion, increased intraocular pressure, retinal detachment, suture breakage, IOL dislocation and late endophthalmitis.[1,2] Glued intra-scleral fixation carries the risk of IOL decentration and intraocular hemorrhage.[1] Iris-sutured IOLs have been associated with high prevalence of IOL dislocation.[1] Anterior fixation of iris-claw IOLs (e.g., Artisan®)[3] carries the risk of de-enclavation, endothelial cell loss, pupillary block, and glaucoma.[1] Capsular tension rings with IOL implantation[4,5] have also been suggested but reports of severe IOL decentration, anterior capsule contraction,[6] and the risk of continuing stretch or rupture of lens fibers leading to subluxation of the IOL-capsule complex preclude its universal application.

In recent years, the same Artisan® lens previously placed in the anterior chamber has been placed behind the iris to correct aphakia and is secured by iris enclavation.[7,8] This allowed the IOL to be situated in a more anatomic position in the posterior chamber and reduced the risk of endothelial cell loss.[1] It was considered a fast and relatively atraumatic technique, with less glare and reduced higher order aberrations compared to an IOL implanted in the anterior chamber.[9,10] Despite the iris enclavated in the lens, dilation of the pupils

Corresponding author: Alvina Pauline D. Santiago, MD
Department of Ophthalmology and Visual Sciences
Sentro Oftalmologico Jose Rizal
Philippine General Hospital
University of the Philippines Manila
Taft Avenue, Ermita, Manilla 1000, Philippines
Email: adsantiago1@up.edu.ph

may still be performed without hampering the view of the posterior pole, permitting evaluation and treatment delivery if necessary.

We report our series of three cases without adequate capsular support in whom retropupillary fixation of an iris-clipped IOL (Artisan®) were done. All three cases had follow-up of at least five years. All three cases are mutual patients of both authors.

The study describes the indication for lens removal, age at intervention, ophthalmologic co-morbidities, and the postoperative course with follow-up of at least five years, including details on final refraction and best corrected acuity. Complications, if present are also reported.

The technique of surgery was discussed with the responsible parent and patient prior to signing of informed consent for the procedure.

Technique of Surgery

The retropupillary fixation of an iris-clipped lens follows the technique described by Mohr.[9-10] Biometry is performed with calculations using a modified A-constant of 116.7. Depending on the patient's age at the time of the procedure, postoperative target refraction was tailored according to the patient's needs, biased towards hyperopia (as a myopic shift remains expected due not only to growth but also to the systemic pathology). Desired dilation is only about 5 mm, achieved after instillation of only 1 drop tropicamide-phenylephrine just before commencement of the procedure.

The main corneal slot incision is done according to the surgeon's preference, as both superior, temporal, or a supero-temporal location can be utilized. Creating a scleral tunnel positioned 1-2 mm behind the corneal entry is likewise an option. Preplaced stab incisions at 3 and 9 o'clock positions are placed for superior corneal slot (this becomes 6 and 12 o'clock when the corneal opening is done temporally (Figure 1). These stab incisions represent the area where an instrument for enclavation of the iris will be introduced.

A lensectomy is performed using the surgeon's preferred technique (extracapsular extraction, mechanical irrigation and aspiration, mechanical lensectomy). At least a good anterior vitrectomy is performed, utilizing diluted triamcinolone (10 mg/ml, 0.1 cc with 0.9 cc of balanced salt solution) to stain the vitreous if necessary. Total vitrectomy is combined with the lensectomy when there is significant lens dislocation or subluxation. Laser retinopexy is performed either on the slit-lamp or via endolaser if there are co-existing posterior pole pathology.

The corneal incision is enlarged to about 5-6 mm to permit entry of a single-piece iris-clipped IOL. The Artisan® lens convexity is reversed, with the convex lens oriented towards the retina, to permit posterior vaulting of the lens. With an IOL lens holder, the lens is introduced into the anterior chamber and placed sitting on top of the iris. The IOL is then rotated with the enclavation sites positioned at the surgeon's 3 and 9 o'clock. The IOL is regrasped with a lens

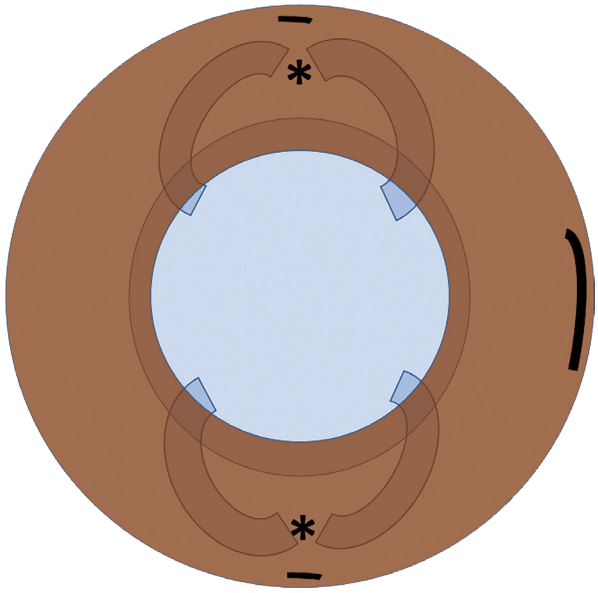

Figure 1. ***Schematic Diagram for Posterior Iris Enclavation.*** The intraocular lens (IOL) vaulting is reversed to avoid iris chafing. With a temporal corneal incision *(long black slightly curvilinear line near limbus)* where the IOL is initially introduced, the area of the enclavation can be done at the 6 and 12 o'clock position with a second instrument (e.g., cyclodialysis spatula) through the side ports created *(small black lines)*.

holder using the surgeon's nondominant hand (the left for right-handed surgeons), the horizontal haptic then introduced just behind the iris. The IOL is held slightly anteriorly, tenting the iris, permitting the outline of the enclavation site to be visualized. A second instrument is introduced at 3 o'clock and the enclavation of the iris is performed with a downward tap on the area of the clip. The surgeon then regrasps the IOL using the opposite hand, and the enclavation through the 9 o'clock slot is performed in the same manner.

Some surgeons proceed with just one hold on the IOL. Both haptics are introduced just behind the iris, and the IOL held slightly anteriorly to outline the areas of enclavation. With a second instrument at the preplaced 3 or 9 o'clock, an instrument is introduced and a downward tap at the area of enclavation is performed to capture the iris. Pushing this instrument further permits enclavation at 9 o'clock without the need to change hands.

The anterior chamber is cleaned of viscoelastic, and the slot sutured using nylon 10-0 interrupted sutures, which are subsequently buried. Intracameral preservative-free antibiotic such as moxifloxacin or levofloxacin is usually placed but left to the surgeon's discretion. The conjunctiva is then draped over the sutures at the end of the procedure.

Postoperative regimen consists of topical steroids (e.g., prednisolone 1%), topical antibiotics (e.g., moxifloxacin), and a weak cycloplegic agent (e.g., tropicamide). Final refraction is performed around 6-8 weeks from the procedure.

CASES

All three cases were diagnosed as Marfan syndrome by a pediatric geneticist. The IOL implanted was an Artisan® iris-fixated lens, with reversal of vaulting done during implantation, using an A-constant of 116.7 for IOL calculations. The optic size was 5.4 mm, with overall IOL length of 8.5 mm. Postoperative regimen for all cases was moxifloxacin, prednisolone 1%, and atropine 1%.

Case 1

At 9 years of age, on initial consultation, a patient diagnosed with Marfan syndrome showed right eye preference with a refraction through the phakic portion of -7.75 -3.25 x 180 with a vision of 20/40-. The left eye's refraction was -8.00 -5.00 x 180 but the patient only counted fingers at 2 feet despite correction. Both lenses were subluxated superiorly, with the right eye showing stretched lens fibers from 4 to 10 o'clock, while the left eye showed similar stretching of the lens fibers from 2 to 8 o' clock. The right lens still occupied about half of the dilated pupil, while the left occupied a third. The left lens also showed beginning cortical cataracts as well as a straightening of the inferior lens edge from 4 to 5 o'clock consistent with a lens coloboma. Dilated funduscopy did not reveal any posterior pole pathology. The patient was initially managed conservatively with best correction and patching of the right eye for amblyopia as definitive cardiac intervention was not performed until the patient was 15 years of age. Cardiac status stability was required before an elective eye surgery could be performed.

At about age 15, the lens subluxation progressed in the left eye, making it difficult to refract through the phakic portion. The right eye showed a refraction of -9.50 -3.50 x 180 through the lens, with a +10.50 through the aphakic portion. Her vision through either portion was 20/50. The aphakic refraction of the left eye was +7.50 with which she also saw 20/50. The patient, however, preferred to wear the high myopic correction consistent with the right eye refraction. For various medical reasons, only conservative management was done as her cardiac procedure was prioritized. Eventually, it became increasingly difficult to perform objective refraction, relying mostly on subjective refraction on follow-up, which maintained her vision in the right eye at 20/50. The left eye deteriorated to only finger counting vision.

Parental permission for the definitive management for the lens subluxation was given only when the patient turned 20 years old (Figure 2).

Because of the patient's age, emmetropia was targeted as the postoperative refraction. The left eye surgery was performed first, with peripheral laser retinopexy done prior to undergoing lensectomy, vitrectomy, and retropupillary fixation of an Artisan® IOL. The right eye had the procedure performed about four months later.

At her last evaluation at age 25, the right eye revealed a refraction of -3.25 -0.50 x 180, with an improved 20/30-

Figure 2. ***20-year-old patient with Marfan syndrome.*** Notice the wingspan that is longer than her height.

vision. The left eye refraction was -0.75 -0.25 x 180 with 20/100 vision. Reading adds were given for near work. There was a large angle exodeviation for which elective strabismus surgery was advised. The retropupillary-fixated IOL remained enclavated at 6 and 12 o'clock in either eye, with iris dimpling corresponding to the area of enclavation seen on slit-lamp biomicroscopy. Dilation showed essentially normal posterior poles with attached retina in both eyes.

Case 2

This patient first consulted at age 4 years for blurring of vision. At that time, she had medial displacement of both lenses with absence of lens attachments in the temporal quadrants of both eyes. Her vision through the phakic portion with an initial refraction of -4.50 OU was 20/100 in either eye. Although Marfan syndrome was suspected, the full features meeting the diagnosis were not met until the child was 13 years old.

For the next six years, she was managed conservatively. Expectedly, her myopia with astigmatism increased as the lens subluxation progressed. When refraction through the phakic lens could no longer be performed, aphakic correction was tried but the patient was unhappy with this correction.

At age 11, she subsequently underwent argon laser cerclage OU for peripheral retinal pathology prior to a lensectomy and anterior vitrectomy with iris-clipped lenses implanted behind the iris (Figure 3). Two months post-operatively, with a refraction of +2.00 -0.50 x 180 OD and +2.00 -0.50 x 180 OS, patient saw 20/50 with either eye, improved to 20/40 when tested binocularly. With addition lenses of +3.00, the patient read 1.25M (slightly larger

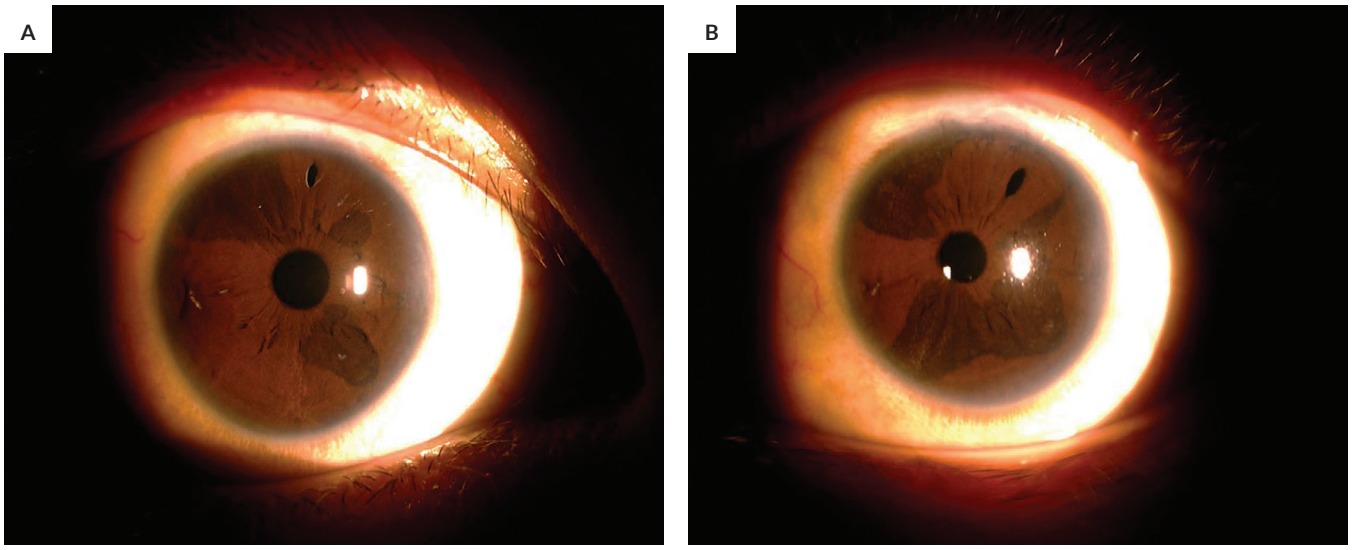

Figure 3. ***Right and left eyes of Case 2.*** Iridotomy is situated superiorly in both eyes, with iris imbricated near the 3 and 9 o'clock positions.

Table 1. Postoperative Endothelial Cell Count (ECC) of Patient 2

Timing postop (years)	Right Eye		Left Eye	
	ECC (cells/mm²)	CV*	ECC (cells/mm²)	CV*
2	3215	29	3333	30
3	2724	40	3378	30
4	3058	36	3278	20

**CV: Coefficient of variation*

than newsprint). Endothelial cell count and pachymetry preoperatively were attempted but was not completed due to the patient's young age precluding full cooperation for the procedure.

Serial postoperative endothelial cell counts (ECC) and the corresponding coefficient of variation (CV) are summarized in Table 1. Normal value (NV) for ECC is ≥2000 cells/mm² with coefficient of variation ≤30.

These values show that ECC remained adequate even after four years from surgery. No further cell counts were done.

She was regularly seen over the next nine years, with the last refraction at age 21 recorded as -1.00 -1.00 x 180 OD, and -1.00 -0.50 x 180 OS. These allowed her to see 20/20 in the right, and 20/25 in the left. She saw Jaeger 1+ with addition lenses of +3.00. The posterior chamber IOL remained in place, with iris enclavation noted at 3 and 9 o'clock positions. The pupils remained dilatable for posterior pole evaluation where the retina remained attached. Laser cerclage marks were seen in the periphery.

Case 3

A 4-year-old boy with a history of outward drifting of either eye noted two years prior consulted only when the child was noted to come within several inches of the television when watching. His initial examination revealed a 30 PD of exodeviation with no eye preference. Cycloplegic refraction of -4.50 -1.00 x 180 was obtained in the right eye, but vision was only 20/200 using picture charts, whereas the left eye saw 20/200 with -3.50 -1.50 x 180. There was note of bilateral lens subluxation medially and nasally. Eighty percent of the lens was still visible through the dilated pupil. Stretched zonules were seen through the 1.5 mm peripheral zone of the pupil beyond the edge of the lens. Dilated indirect ophthalmoscopy showed essentially normal findings. His father also had exodeviation but no family history of lens subluxation was elicited.

Over the next three years, he was managed conservatively with phakic refraction, achieving 20/50 best vision in either eye obtained with a refraction of -8.25 -1.50 x 180 and -6.75 -1.75 x 180 for the right and left eye, respectively. The patient's deviation was reduced to 8 PD of exotropia on both the simultaneous prism cover test and alternate prism cover test.

Expectedly, myopia progressed with vision also deteriorating to 20/100 in either eye even when a higher spectacle correction through the phakic portion of -10.50 -1.50 x 180, -8.50 -1.75 x 180 were given. Eventually, refraction through the phakic portion became difficult, as the lens subluxation slowly progressed where only ¼ of his lens could be seen through a dilated pupil. Aphakic refraction was not tolerated, iris-clipped IOL implantation was suggested.

At age 7, he underwent peripheral laser retinopexy, vitrectomy, and retropupillary iris-clipped IOL implantation in both eyes. One month postoperatively, vision in the right with refraction of -0.25 x 90 was 20/30- while the left eye refraction was +1.00 with a vision of 20/30-. His refraction five years postoperatively, at his last visit, showed the patient

requiring -1.00 -0.50 x 090 OD while OS required -1.25 -1.00 x 090 with which he saw 20/30 in either eye. Reading addition lenses required to see at least Jaeger 1 was only +2.00. Lenses remained clipped at 3 and 9 o'clock for both eyes and stayed stable. Pupils remained easy to dilate, and the posterior pole can be evaluated following cycloplegia with tropicamide and phenylephrine combination drops.

A summary of the features of the three cases presented are outlined in Table 2.

DISCUSSION

The degree of lens subluxation in Marfan syndrome is varied and can occur in different directions. The lens can displace superiorly, medially, and even inferiorly as these cases showed. Initial management remains to be conservative, with best correction provided either through the phakic or aphakic portion as the patient can tolerate. However, there are instances when the shift to aphakic correction is not tolerated as these three cases showed. Also, as the lens gets

Table 2. Summary of Patients who Underwent Retropupillary Fixation of Iris-clipped Lenses

	Case 1	Case 2	Case 3
Age at Presentation	9	4	4
Initial Phakic Refraction	OD: -7.75 -3.25 x 180 OS: -8.00 -5.00 x 180	OD: -4.50 OS: -4.50	OD: -4.50 -0.50 x 180 OS: -3.50 -1.50 x 180
Initial best vision (with correction)	OD: 20/40- OS: Counting fingers, 2 feet	OD: 20/100 OS: 20/100	OD: 20/200 OS: 20/200
Presurgical refraction	Phakic: OD: -10.50 -3.50 x 180 OS: unable Aphakic OD: +8.50 OS: +5.50	Phakic: OU: unable Aphakic: not tolerated, not recorded	Phakic: OD: -10.50 -1.50 x 180 OS: -8.50 -1.75 x 180 Aphakic: OD: +10.50 OS: +7.50
Vision prior to surgery	OD: cc 20/50 OS: Counting fingers *(preferred myopic Rx)*		OD: cc 20/100 OS: cc 20/100
Estimated degree of subluxation	OD: 50% OS: 80%* **With cortical cataracts*	OD: 60% OS: 70%	OD: 75% OS: 75%
Direction of subluxation	Superonasal OU	Medial OU	Inferior OU
Age at surgery	20	11	7
Laser Retinopexy	Yes	Yes	Yes, with vitrectomy
Target refraction	Emmetropia	+1.00	+1.00
Postoperative refraction	OD: -1.75 -0.75 x 90 OS: +1.50 -1.25 x 180 Adds: +3.00	OD: +2.00 -0.50 x180 OS: +2.00 -0.50 x 180 Adds: +3.00	OD: -0.25 x 90 OS: +1.00 Adds: +3.00
Postoperative 2 months VA (with correction)	OD: 20/40+ OS: 20/100	OD: 20/50 OS: 20.50 OU: 20/40 Near: 1.25M	OD: 20/30- OS: 20/30-
Age at last follow-up	25	20	12
Total follow-up	16 years	16 years	7 years
Total post-surgical follow-up	5 years	9 years	5 years
Latest refraction	OD: -3.25 -0.50 x 180 OS: -0.75 -0.25 x 180 Adds: +3.00	OD: -1.00 -1.00 x 180 OS: -1.00 -0.50 x 180 Adds: +3.00	OD: -1.00 -0.50 x 090 OS: -1.25 -1.50 x 090 Adds: +2.00
Best vision with correction	OD: 20/30- OS: 20/100-	OD: 20/20 OS: 20/25 Near: cc J1+	OD: 20/30 OS: 20/30 Near: cc J1+
Postoperative problems			
Focal Iris Atrophy	(-)	(-)	Yes
Glaucoma	(-)	(-)	(-)
Postop dilation	(-)	(-)	(-)
Loss of enclavation	(-)	(-)	(-)
Inadequate view of retina	(-)	(-)	(-)

displaced, the edge of the lens can become troublesome, prompting the need to consider earlier surgical removal when this obstructs the visual axis.

All the cases showed better vision after the subluxated lenses were replaced with a retropupillary placed iris-clipped lens. The surgery probably removed aberrations and distortions that patients used to experience when viewing through a part of the lens other than the center.

Targeted postoperative refraction for children with connective tissue disorder such as Marfan syndrome should probably be maintained at least at a slight hyperopia, anticipating further myopic shift, even after cataract extraction and even after age at surgery of 7 to 20 years. Dilation for posterior pole evaluation despite enclavation was achievable as evidenced by these cases.

Careful patient selection necessitates that those with iris disease or atrophy should be carefully evaluated for the potential for IOL enclavation that may loosen because of iris pathology. Although ideal, endothelial cell count should be obtained preoperatively, and followed serially postoperatively.[11] This, however, was often limited by the lack of cooperation in younger patients, as well as unavailability of the machine at institutions where patients consulted. Standardizing preoperative and postoperative protocols for centers or surgeons who will perform the procedure is recommended.

The lens removal may be approached anteriorly, through a scleral or corneal incision, or posteriorly through a pars plana approach, but is probably largely dependent on surgeon preference. Laser retinopexy was also best delivered via endolaser procedure through the help of a retina colleague but slit-lamp delivery of laser treatments can also suffice, especially for the older, more cooperative patient. Laser retinopexy is indicated for eyes with significant peripheral lattice degeneration and/or holes.

We have observed that superior scleral incisions with appropriate suture removal left very small residual refraction, of with-the-rule astigmatism (Cases 1 and 2). Temporal incisions, such as the one done in Case 3, produced an against-the-rule astigmatism. We also tended to remove sutures later than what we would have done for an eye without connective tissue pathology.

With careful patient selection and attention to peripheral retinal pathology requiring retinopexy, patients did well, even after 5 to 9 years as evidenced by this series, and 13.8 + 5.9 years in a large series by Manning et al.[11] All patients in our series had the benefit of good peripheral examination, one required that this be done under general anesthesia preoperatively with the endolaser procedure for peripheral cerclage delivered in the operating room. Postoperative dilation was not hampered by the iris enclavation and still permitted a good visualization of the retina periphery.

Some of the complications to monitor include loss of enclavation (threatening a dropped IOL in the posterior pole and its consequences), pseudophakic pupillary block (which can be prevented by a surgical peripheral iridotomy or iridectomy), and maybe even glaucoma. Although not seen in our series and Manning's,[11] retinal detachment associated with high myopia should be monitored closely.

CONCLUSION

Retropupillary fixation of an iris-clipped intraocular lens is a viable procedure even for children without capsular support. It is more physiologic when positioned behind the iris. With careful patient selection and attention to peripheral retinal pathology requiring retinopexy, patients do well, even for the long term. These patients should be closely monitored for complications due to either the surgery or the pathology of Marfan syndrome.

Statement of Authorship

Both authors contributed in the identification of cases for surgery, conceptualization of work, recording of clinical course and findings, performing and documenting of surgeries, drafting and revising of manuscript, and final approval of the version to be published.

Author Disclosure

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