Long-Term Complications of Iris-Claw Phakic Intraocular Lens Implantation in Weill-Marchesani Syndrome

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Purpose: This study was designed to report the long-term complications of iris-claw phakic intraocular lens implantation in a patient with Weill-Marchesani syndrome.

Methods: Case report and literature review.

Results: A 26-year-old man with a history of glaucoma had bilateral phakic lens implantation for high myopia 10 years previously. Two years later, the left implant dislocated and was repositioned. Slit-lamp examination of both eyes revealed phakic implants of the iris-claw variety. There were moderate iridocorneal adhesions in the areas in which the lens haptics pinched the iris in both eyes and moderate epithelial and stromal edema over the temporal one-third of the left cornea. The crystalline lenses were clear with 3+ phacodonesis OU. Dilated fundus examinations revealed bilateral severe optic nerve cupping. Crystalline lens diameters were measured at 7.5 mm in the right eye and 8 mm in the left. Anterior chamber depths were 2.63 mm OD and 2.40 mm OS. Specular microscopy revealed central endothelial cell counts of 1133 and 587 cells/mm^2 OD and OS, respectively. Axial lengths were 23.3 mm OD and 25 mm OS. Gonioscopic examination revealed bilateral angle closure with marked peripheral anterior synechiae. Based on our findings of short stature, shortened and thickened fingers, relatively normal axial length, microspherophakia, high myopia, and glaucoma, we diagnosed the patient with Weill-Marchesani syndrome.

Conclusion: Iris claw-lens phakic lenses may be an effective surgical alternative to correct high myopia in select patients; however, it may produce long-term complications in eyes with specific features.

Key Words: iris-claw lens, phakic intraocular lens, Weill-Marchesani syndrome

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The treatment of patients with high myopia has generated controversy in recent years. Optical correction of high myopia with spectacles produces image minification, optical aberrations, limited field of vision, and a poor cosmetic appearance because of the thickness of the lenses. Contact lenses largely resolve these issues, although they entail problems of intolerance and complications derived from their use.[1] Hence, surgical procedures, such as epikeratoplasty, automated lamellar keratomileusis, excimer laser photorefractive keratectomy, laser in situ keratomileusis, phakic intraocular lens (IOL) implantation, and refractive lens exchange have been introduced to correct high myopia.[1]

The implantation of an IOL to correct high myopia was first discussed during the 1950s by Strampelli[2] and Barraquer.[3] Since then, several phakic anterior chamber IOLs (ACIOL) have been developed. Worst et al[4] performed the first implantation of the iris-claw phakic IOL.

We report a patient with Weill-Marchesani syndrome who underwent bilateral iris-claw phakic ACIOL implantation for high myopia and repositioning of the ACIOL in the left eye 2 years after the initial surgery. He developed corneal decompensation and chronic angle-closure glaucoma 10 years later.

CASE REPORT

A 26-year-old man was referred for corneal evaluation on October 17, 2003. He had a history of glaucoma since the age of 6 or 7 years and was using 2 topical antiglaucoma medications. He had undergone phakic lens implantation surgery to treat high degrees of myopia in both eyes in 1994, in Turkey. He reported that his uncorrected vision had improved dramatically after the surgery. Approximately 2 years later, the left IOL dislocated and was surgically repositioned. His visual acuity was 20/30 OU with a correction of approximately −0.75 + 4.75 at 90° in both eyes. Pupillary reactions and ocular motility were normal bilaterally. Intraocular pressures were 15 mm Hg OD and 18 mm Hg OS.

The right cornea was clear except for mild inferior punctate epitheliopathy. Anterior chamber was deep and quiet, and there was a horizontally placed iris-claw phakic ACIOL (Fig. 1). There were moderate iridocorneal adhesions in the areas in which the lens haptics pinched the iris at 3 and 9 o'clock and a peripheral iridotomy at 10 o'clock. Slit-lamp examination of the left eye revealed moderate epithelial and stromal edema over the temporal one-third of the cornea. There were posterior and anterior synechiae at 9 o'clock. There was a horizontally placed iris-claw phakic ACIOL (Fig. 2). There was a peripheral iridotomy at 2 o'clock. The crystalline lenses were clear, although there was 3 + phacodonesis OU. Dilated fundus examinations revealed severe optic nerve cupping with cup-to-disc ratios of 0.9 OD and 0.95 OS. The macula, vessels, and periphery appeared normal. Crystalline lens diameters were 7.5 mm OD and 8 mm OS. Anterior chamber depths were 2.63 mm OD and 2.40 mm OS. Central endothelial cell counts were 1133/mm^2 OD and 587/mm^2 OS. Axial lengths measured by A-scan ultrasound were 23.3 mm OD and 25 mm OS. Gonioscopy revealed bilateral closed angles with marked peripheral anterior synechiae nasally and temporally.

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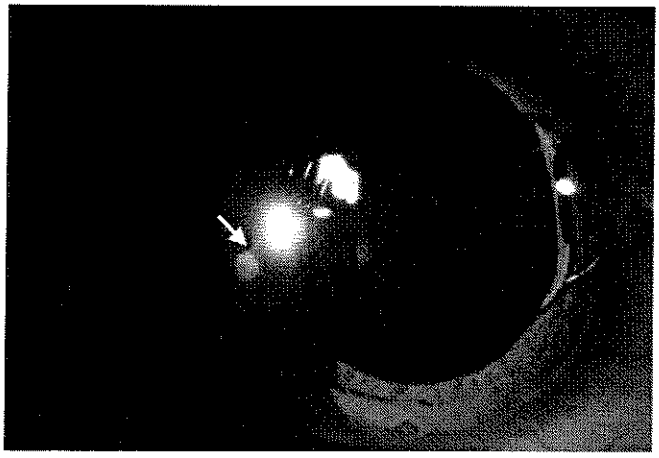

FIGURE 1. The right eye showing a clear cornea, iridocorneal adhesions at 3 and 9 o'clock and the iris-claw phakic lens in horizontal position. The arrow indicates the iridocorneal adhesion at 9 o'clock.

Based on our findings of short stature, shortened and thickened fingers (Fig. 3), relatively normal axial length, microspherophakia, high myopia, and glaucoma, we diagnosed the patient with Weill-Marchesani syndrome. Glaucoma consult was obtained for poorly controlled, advanced glaucoma. We elected to prescribe hypertonic saline, because we were concerned that explantation could cause further damage to the cornea.

DISCUSSION

Weill-Marchesani syndrome is a rare connective tissue disorder characterized by short stature, brachydactyly, joint stiffness, and characteristic eye abnormalities, including microspherophakia, ectopia lentis, severe myopia, and glaucoma.[5] Both autosomal recessive and autosomal dominant modes of inheritance have been described, and a gene for the recessive Weill-Marchesani syndrome has recently been mapped to chromosome 19p13.3-p13.2.[6]

The characteristic small, spherical shape of the lens combined with zonular laxity predisposes the eye to pupillary block and angle-closure glaucoma in Weill-Marchesani syndrome. Asaoka et al[7] reported a case and postulated a mechanism of chronic angle closure glaucoma secondary to frail zonular fibers and spherophakia in Weill-Marchesani syndrome. In our case, gonioscopic examination revealed angle closure glaucoma as well. Our patient also had 3+ phacodonesis indicative of loose zonular fibers. Although iris-claw IOLs should theoretically not disturb the angle; nasal and temporal peripheral anterior synechiae contributed to further impairment of outflow and poor control of glaucoma.

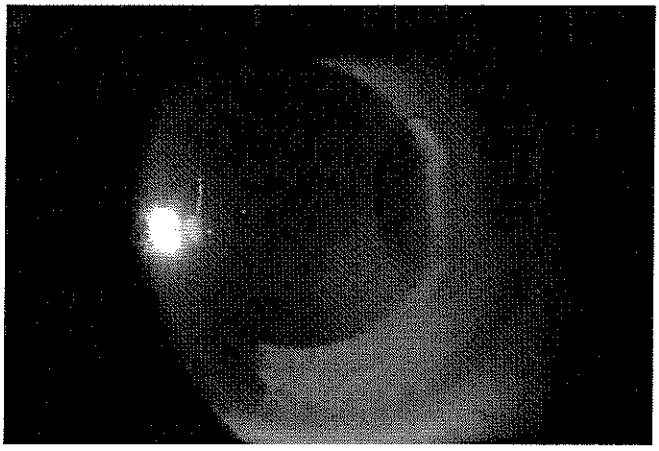

FIGURE 2. The left eye has moderate epithelial and stromal edema over the temporal one-third of the cornea.

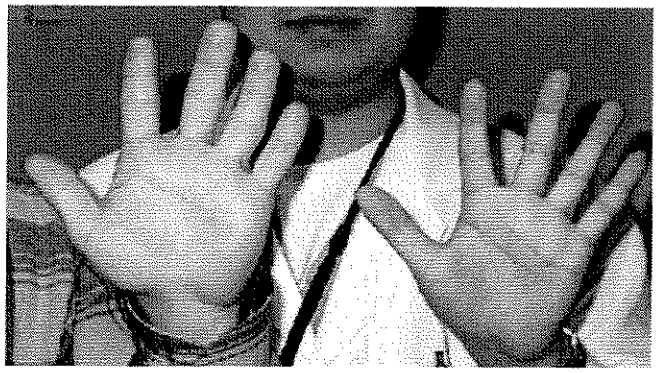

FIGURE 3. The patients' hand on the left in comparison to the hand of a normal male of almost the same age and height on the right.

Phakic ACIOL implantation is an effective surgical procedure to correct high myopia; however, there is a concern regarding long-term endothelial cell loss. Several articles have been published to assess the effect of iris-claw lenses on endothelial cells. Menezo et al[1] studied 94 eyes of 62 patients with myopia of −7.00 diopters or higher who underwent Worst-Fechner IOL implantation. Three years after surgery, 58 eyes (61%) had an uncorrected visual acuity of 20/40 or better and the mean endothelial cell loss was 17.9% at 5 years after surgery, whereas the percentage of hexagonality and the coefficient of cell variation tended toward preoperative levels. The same group reported 4-year follow-up results and suggested that there was a slight, progressive cell loss after implantation with return of morphometric changes to the preoperative levels. They concluded that endothelial loss occurred primarily during the surgical procedure.[8]

Landesz and coauthors[9] reported the long-term results of the Artisan 5-mm optic iris-claw myopia lens in 67 eyes of 38 consecutive patients with preoperative myopia ranging from −5.38 to −28.00 D. With a mean follow-up of 35 months, 67.2% of the patients had a postoperative residual refraction within ±1.00 D of emmetropia. Mean endothelial cell loss was 5.5% at 6 months, 7.21% at 12 months, 9.1% at 24 months, and 10.9% at 36 months.

Although Artisan phakic IOL has been widely used around the world over the years, it only received United States Food and Drug Administration (FDA) approval in September 2004. It is called the Verisyse lens in the United States. Pop and Payette[10] published the initial results of endothelial cell counts of 765 eyes enrolled at 25 North American sites in the FDA Ophtec clinical trial of the myopic Artisan IOL. The mean preoperative endothelial cell count

was 2631 ± 442 cells/mm^2. Percentage change from baseline at 6, 12, and 24 months was −0.09% ± 16.39%, −0.87% ± 16.35%, and −0.78% ± 17.41%, respectively. The decrease was not statistically significant. No relationships were noted between endothelial cell loss and patient age or implant power.

Ophtec does not advocate Artisan phakic lens implantation in eyes with an anterior chamber depth (ACD) below 2.6 mm. The current FDA approval indications for the Verisyse are for the ACD to be no less than 3.2 mm. The original ACDs are reported to be reduced by 28 to 34% after implantation.[11] Current ACDs in our patient are 2.63 mm OD and 2.40 mm OS. Although we were not able to identify the exact model of the iris-claw lens and preoperative ACD values are not available for our case, peripheral anterior synechiae could have further reduced the anterior chamber angle depth. The surgeon should always consider ACD before implantation of a phakic ACIOL, especially in eyes that are at increased risk for glaucoma.

To the best of our knowledge, there is no report of phakic IOL implantation to correct myopia secondary to microspherophakia in Weill-Marchesani syndrome. It is possible that the lower endothelial cell count and corneal edema in the left eye could be attributable more to the dislocated phakic IOL and the additional repositioning procedure in that eye. Nevertheless, cell counts in both eyes are significantly lower than normal for such a young age, although we do not have any data regarding preoperative endothelial cell counts.

In conclusion, iris-claw phakic IOL implantation may be an effective surgical procedure to correct high myopia in select patients; however, the surgeon should consider the risk of long-term complications in such a young patient with an unusual condition.

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