

Iris-fixated phakic intraocular lens implantation for correction of high myopia in microspherophakia

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We report the refractive correction of high myopia in a 23-year-old patient with idiopathic microspherophakia using iris-fixated phakic intraocular lenses (pIOLs) (Verisyse/Artisan). Four years after bilateral implantation, the uncorrected distance visual acuity was 20/25 with a correction of 20/20⁻¹ in both eyes. No intraoperative or postoperative complications occurred. Iris-fixated pIOLs are not recommended for every patient with microspherophakia. However, this procedure may be an option in microspherophakic patients with appropriate anterior chamber depth and no history of lens dislocation who are likely to comply with annual eye examinations. Follow-up should include monitoring the endothelial cell count and biomicroscopy for adequate space between the pIOL, the natural crystalline lens, and the corneal endothelium. Scheimpflug photography can be a valuable tool in such cases.

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Microspherophakia is an uncommon bilateral condition in which abnormal laxity of lens zonules leads to an increased anterior-posterior diameter of the lens and development of a spherical lens. The high myopia associated with this condition, -10.00 to -15.00 diopters (D), is almost exclusively lenticular in nature.^{1,2} Because of inadequate optical correction with spectacles and patient intolerance to contact lenses, surgical correction is a valid alternative. Current available methods include clear lens extraction with posterior

chamber intraocular lens (PC IOL) placement,² pars plana lensectomy with transscleral fixation of a PC IOL,³ or an iris-sutured IOL. We report a patient with bilateral microspherophakia and high myopia who had bilateral iris-fixated phakic IOL (pIOL) implantation with a successful surgical result and satisfactory visual outcome after 4 years of follow-up.

CASE REPORT

A 23-year-old man with microspherophakia was referred to the John A. Moran Eye Center at the University of Utah for a second opinion relating to correction of high myopia. On examination, the corrected distance visual acuity (CDVA) was 20/30 in both eyes with a manifest refraction of $-13.75 + 1.75 \times 136$ in the right eye and $-14.25 + 2.25 \times 73$ in the left eye. The patient had become contact lens intolerant and was seeking a refractive surgery option as an alternative. Except for the thick crystalline lenses with a large anterior-posterior depth of 4.82 mm, slitlamp biomicroscopy was within normal limits. Anterior chamber depth (ACD) as measured by the IOLMaster (Carl Zeiss Meditec, Inc.) was 2.87 mm in the right eye and 2.80 mm in the left eye.

With pupil dilation, the equator edges and capsular zonules of each crystalline lens were clearly visible, showing an approximate diameter of 9.0 mm, and both IOLs were minimally displaced superiorly. There

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was no phacodonesis or iris transillumination. The corneal curvature was somewhat flat but within normal limits—40.0/41.7 D in the right eye and 39.4/42.6 D in the left eye. The mean axial lengths were within normal limits—24.32 mm and 24.06 mm in the right eye and left eye, respectively—suggesting that the myopia was primarily lenticular. Endothelial cell density (ECD) using the Noncon Robo specular microscope (Konan Medical) was 3077 ± 80 cells/mm² in the right eye and 2825 ± 91 cells/mm² in the left eye, and intraocular pressure (IOP) by applanation was 16 mm Hg in both eyes. In all other respects, the ophthalmoscopic findings were unremarkable.

None of the clinical features were consistent with Weill-Marchesani syndrome, Marfan syndrome, or Alport syndrome. The patient's history and extraocular physical examination were negative for findings that might lead to one of these diagnoses. The family history was also unremarkable. Bilateral idiopathic microspherophakia was diagnosed. After an extensive discussion with the patient about the diagnosis and various refractive surgery options and risks, a decision to implant iris-fixated pIOLs was made. A -14.0 D Verisyse IOL (VRSM6US) (Ophtec) was

inserted through a 6.0 mm corneoscleral incision centered on the steep axis in the left eye. Enclavations were successfully obtained at the 3 o'clock and 9 o'clock positions of the midperipheral iris with no complications. Two months later, a -13.5 D Verisyse IOL was implanted uneventfully in the same manner in the right eye. At the 24-month postoperative examination, the uncorrected distance visual acuity (UDVA) was 20/25 in both eyes with a manifest refraction of -0.25 sphere in the right eye and +0.50 +0.50 \times 39 in the left eye. The CDVA was 20/20⁻¹ in both eyes. The corneal curvature was 40.12/40.5 D in the right eye and 40.5/41.0 in the left eye.

At the 4-year examination, the UDVA remained 20/25 in both eyes with a manifest refraction of plano +0.25 \times 50 in the right eye and +0.50 +0.50 \times 50 in the left eye. The CDVA remained 20/20⁻¹ in both eyes. The IOP was 18 mm Hg in the right eye and 17 mm Hg in the left eye, and the mean ECD was 2890 ± 111 cells/mm² in the right eye and 2785 ± 78 cells/mm² in the left eye, an endothelial cell loss of 6.1% and 1.4%, respectively. The mean distance between the anterior capsule of the natural crystalline lens and the posterior surface of the IOL, measured

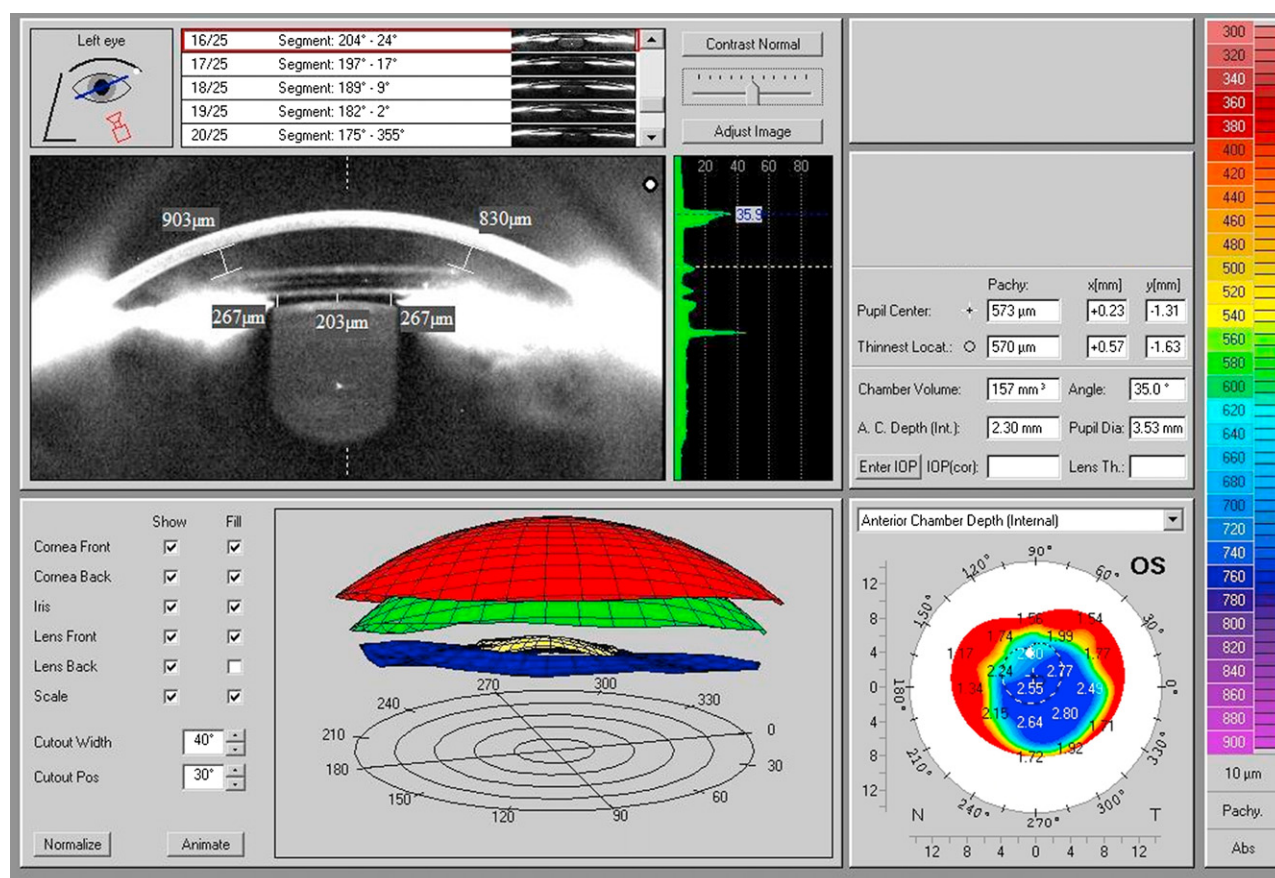


Figure 1. Scheimpflug image of the left eye showing the position of the iris-fixated pIOL.

by Scheimpflug imaging (Pentacam, Oculus, Inc.), was $265 \pm 78.9 \mu\text{m}$ (range 208 to $322 \mu\text{m}$) in the right eye and $235 \pm 57.9 \mu\text{m}$ (range 203 to $267 \mu\text{m}$) in the left eye. The ACD was 2.46 mm and 2.30 mm, respectively. The mean distance between the lens and the endothelium at the enclavation site was $0.757 \pm 0.072 \text{ mm}$ (0.71 to 0.80 mm) in the right eye and $0.867 \pm 0.062 \text{ mm}$ (0.83 to 0.90 mm) in the left eye (Figures 1 and 2).

DISCUSSION

Microspherophakia is a rare condition that is usually associated with other systemic syndromes such as Marfan, glaucoma-ectopia lentis-microspherophakia-stiffness-shortness, cri-du-chat, and Weill-Marchesani.⁴⁻⁹ Ocular complications such as glaucoma,^{7,10} ectopia lentis, high myopia, and retinal detachment are often seen in microspherophakia. Lenticular myopia is the most common cause of the high myopia because of the small spherical crystalline lens in these patients. In addition, the effective lens power may be increased by anterior subluxation, resulting in a high refractive error. The exact cause of this condition is unknown, but it is thought to be related to defective mesodermal development.¹

The high lenticular myopia in microspherophakic patients can be disabling in terms of quality of life. Unfortunately, the ideal treatment for the myopia, other than glasses and/or contact lenses, is controversial and undefined. There is one report of clear lens extraction with PC IOL implantation² and one report of pars plana lensectomy with scleral fixation IOL.³ To our knowledge, there has been no report of pIOL implantation to correct myopia secondary to idiopathic microspherophakia, although implantation of an iris-fixated pIOL has been reported in a patient with Weill-Marchesani syndrome.¹¹ The Weill-Marchesani patient was reported to have early signs of corneal decompensation in the left eye at 10 years of age, which was attributed to previous dislocation of the pIOL and the surgery required for repositioning. He maintained a visual acuity of 20/30 in the left eye and explantation was not performed, although there was corneal edema at the enclavation site. It is noteworthy that the cornea of the right eye remained clear despite an ECD of 1133 cells/ mm^2 .

The advent of Scheimpflug photography has made it possible to obtain anterior chamber data through a fast noncontact approach. The ACD and the distance

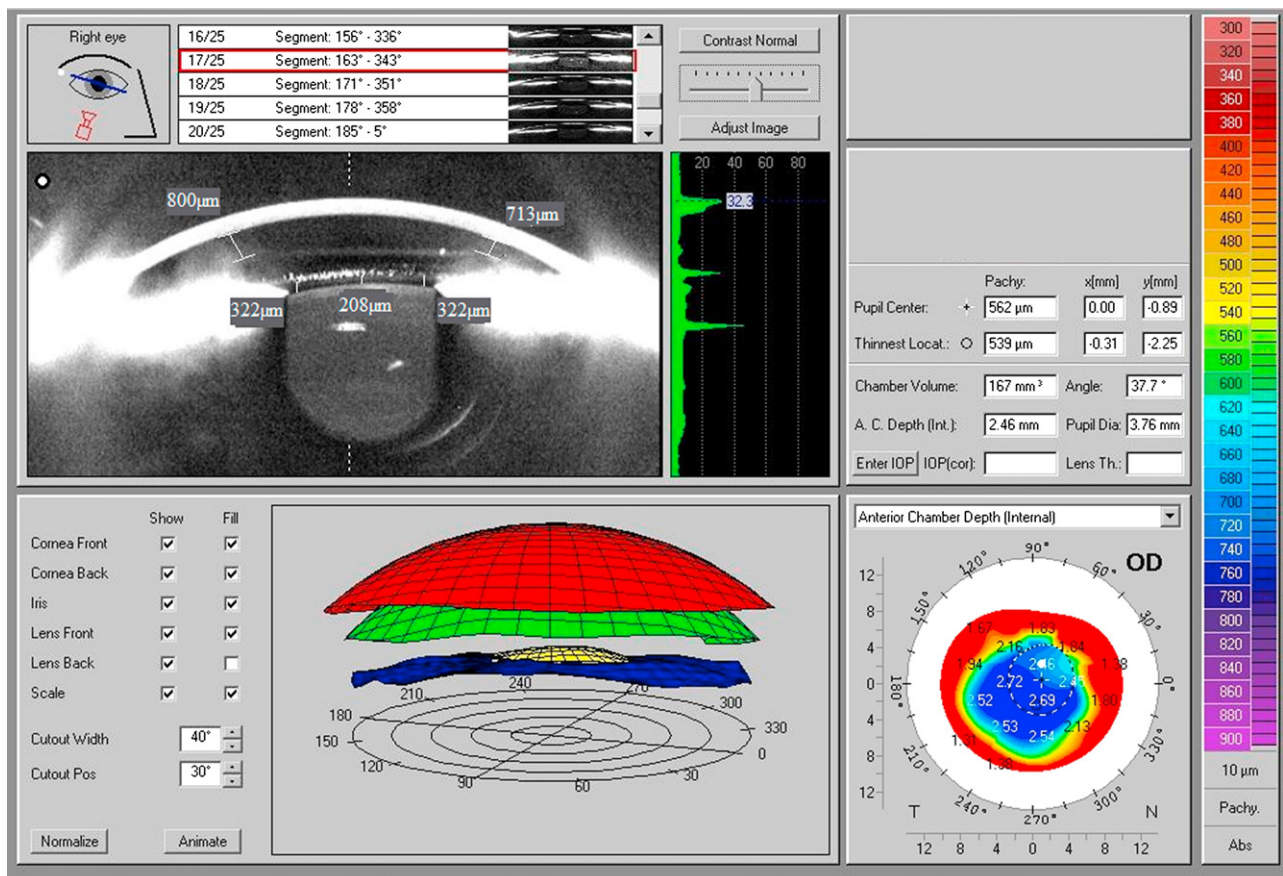


Figure 2. Scheimpflug image of the right eye with an iris-fixated pIOL.

between the pIOL and the anterior capsule of the natural lens and corneal endothelium can be collected and followed. In 2008, Kohnen et al.¹² reported the range of the distance between an iris-fixated IOL and the crystalline lens was 0.29 mm to 0.66 mm at 1 year in otherwise normal myopic patients. In our patient, the range was 0.20 mm to 0.32 mm in both eyes at 4 years. The distance between the pIOL and the corneal endothelium was 0.71 to 0.90 mm at the enclavation site in both eyes. Scheimpflug imaging showed continued preservation of the space between the iris-fixated pIOL and the anterior lens capsule and the corneal endothelium. The distance between the enclavation site and the endothelium was somewhat less in the right eye; a larger ECD decrease occurred in this eye over 4 years (6.1% right eye versus 1.4% left eye). The clinical significance of the difference is unknown as the standard deviation range of postoperative ECD was 4% (2890 ± 111 cells/mm²) in the right eye and 3% (2785 ± 78 cells/mm²) in the left eye. In addition, the postoperative ACD was larger on the right than the left, which attests to the limitation of accurate ECD measurements.¹³ At the last examination, there was no evidence of cataract formation or corneal decompensation. There was minimal IOL interaction with the surrounding tissues as shown by the visual acuity, normal IOP, quiet anterior chamber, stable IOL position; and lack of cataract formation, synechiae, iridocorneal adhesions, or iris atrophy.

Verisyse pIOL implantation may not be appropriate for many microspherophakic patients, especially those whose natural lenses have an extremely large anterior-posterior diameter or anterior dislocation that might allow contact between the iris-fixated pIOL and the crystalline lens and/or cornea. In our patient, the ACD of 2.87 mm in the right eye and 2.80 mm in the left eye met the manufacturer's recommendations of greater than 2.6 mm but fall short of the U.S. Food and Drug Administration's recommendation of 3.2 mm. There is evidence of an inverse relationship between ACD and endothelial cell loss for the iris-fixated IOL in one long-term study.¹⁴ Furthermore, the original ACD has been reported to reduce by 28% to 34% after implantation.¹⁵ In our patient, the postoperative ACD was 2.46 mm (decrease of 15%) in the right eye and 2.30 mm (decrease of 18%) in the left eye. In a young patient with minimal zonular laxity, no phacodonesis or IOL decentration, and an appropriate ACD, iris-fixated IOL implantation may be an alternative to more invasive and complicated procedures. In addition to thorough preoperative planning and discussion, patients having this procedure require follow-up at regular intervals to monitor endothelial cell count, changes in location of IOL, and potential complications such as cataract development and corneal

decompensation. The long-term results of iris-fixated pIOL implantation in this patient population are not known. Therefore, we do not recommend the procedure as a standard of treatment, but wish to present the visual outcomes in this select patient to illustrate the full range of treatment options available to microspherophakic patients.

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