Implantation of an implantable collamer lens and iris cerclage for the treatment of Urrets-Zavalia syndrome

Abstract
A 27-year-old woman with bilateral high myopia had anterior chamber iris-claw phakic intraocular lens implantation in her left eye under local anesthesia and with no intraoperative complications. The preoperative slit-lamp examination was normal, and she had no relevant past medical history. On the second postoperative day, patient developed fixed, dilated mydriasis, and one month later, an IOL subluxation was noted on examination. She had no history of intraocular hypertension and no mydriatic drops were used. She was ultimately diagnosed with Urrets-Zavalia syndrome. We removed the IOL and observed the patient for six months after fitting her with a colored contact lens. During this period of time, she complained of glare and photophobia and developed contact lens intolerance. To bring more comfort to the patient, we implanted a posterior chamber Implantable Collamer Lens (ICL) and performed iris cerclage in the left eye with good aesthetic and refractive outcome. Posterior chamber ICL implantation with iris cerclage proved to be a simple and effective approach in the management of this situation.

Keywords: Implantable collamer lens; Iris cerclage; phakic intraocular lens; Urrets-Zavalia syndrome.

Introduction
Paralytic mydriasis and iris atrophy, first noticed following penetrating keratoplasty in eyes of patients with keratoconus, was named Urrets-Zavalia syndrome.\(^1\) Later reports have also indicated the presence of this syndrome following intracameral gas injection for treatment of acute corneal hydrops\(^2\), deep
lamellar keratoplasty, argon laser peripheral iridoplasty for glaucoma, trabeculectomy and implantation of phakic intraocular lenses (IOLs) for myopia. It is also frequently related to high postoperative intraocular pressure (IOP). Herein, we describe a case of Urrets-Zavalia syndrome after an uneventful implantation of an iris-claw phakic IOL and the postoperative follow-up.

**Case report**

A 27-year-old woman with bilateral high myopia had anterior chamber iris-claw phakic IOL implantation in her left eye (LE) with no intraoperative complications. She had no relevant past medical history. The preoperative slit-lamp examination was normal; in fact, both pupils were light-reactive, isochoric, and 5.75 mm in diameter under scotopic illumination. Preoperative best-corrected visual acuity (BCVA) was 20/20 bilaterally with a refraction of -11.00 -1.25 x 105 in the right eye (RE) and -11.25 -1.50 x 150 in the left eye (LE). The IOP was 14 mmHg in the RE and 13 mmHg in the LE. The anterior chamber depth was 3.35 mm in the RE and 3.41 mm in the LE. The limbus-to-limbus measurements were 11.02 mm in the RE and 11.25 mm in the LE, and the preoperative corneal topographic findings as measured by the rotating Scheimpflug camera (Pentacam®, OCULUS Optikgerate GmbH, Wetzlar, Germany) were normal in both eyes. Central endothelial cell count (CECC) was 2790 cells/mm² in the RE and 2812 cells/mm² in the LE. The surgical procedure in the LE was performed under topical anesthesia with oxybuprocaine 0.4% and intracameral lidocaine 2%. After a clear corneal superior incision was made, an intracameral injection of acetylcholine 1% was administered, and the IOL was implanted on the iris with the help of sodium hyaluronate 1% (Healon®). After a peripheral iridectomy was performed, the ophthalmic viscosurgical device was removed from the anterior chamber via irrigation and aspiration with a balanced salt solution. No mydriatic drugs were given before, during, or after surgery.

On the second postoperative day, the patient developed fixed, dilated mydriasis in her LE. No reaction to light or 0.125% or 1% pilocarpine, and the IOP was 15 mm Hg in both the RE and the LE (Figures 1 and 2). One month later, she developed IOL subluxation in the LE. We removed the IOL and observed her for six months after fitting her with a colored contact lens. During this period, she developed contact lens intolerance, which led to symptoms such as incapacitating glare, photophobia, and decreased vision. At this point, specular microscopy revealed a CECC of 2695 cells/mm² in the LE. We decided to implant a posterior chamber Visian implantable collamer lens (ICL) (STAAR Surgical, Inc. Nidau, Switzerland) to correct the refractive error and to protect the lens, and simultaneously performed an iris cerclage. The cerclage was performed using a curved needle and 10-0 Prolene sutures with a running suture technique (Figures 3 and 4).

After five weeks of follow-up, she no longer suffered from glare or photophobia, and the visual acuity in the LE on examination was 20/25; moreover, the IOP was 14 mm Hg in the LE, and the CECC was 2771 cells/mm² in the RE and 2480 cells/mm² in the LE. Ultimately, we are planning to implant an ICL in the RE as well.
Discussion

Urrets-Zavalia syndrome has been reported most frequently following penetrating keratoplasty for keratoconus. However, it can accompany virtually all anterior segment surgeries. For example, Yuzbasioğlu et al reported one case of fixed paralytic mydriasis following anterior phakic angle-supported IOL implantation. In that case, the primary causative factor was probably iris ischemia secondary to IOP elevation up to 60 mmHg on the first postoperative day. Later, Park et al. reported a case of Urrets-Zavalia syndrome following anterior segment iris-claw phakic IOL implantation without high IOP. The pathogenesis of this syndrome remains uncertain. To date, iris ischemia secondary to high IOP, low sclera rigidity, intrinsic iris abnormalities, and intraoperative damage of the iris vessels or parasympathetic nerves have been identified as the main risk factors. However, other unidentified mechanisms must justify the occurrence of this syndrome after successful surgery with no postoperative complications or intraocular hypertension.

Our patient did not experience pre or postoperative IOP elevation, and no mydriatic drugs were used on her before, during, or after surgery. Adie’s tonic pupil and third nerve palsy associated mydriasis were excluded as potential causes, because no response was observed to the 0.125% and 1% pilocarpine instillation, respectively. The iris-claw IOL subluxation was probably due to pronounced iris atrophy. Posterior chamber ICL implantation offers a good refractive solution, and the iris cerclage using a running suture technique resulted in a good aesthetic and functional result. This technique minimizes any stretching and distortion of the iris and creates a pupil that is precisely sized and rather round. Additionally, the implantation of an ICL prevents any damage to the lens. Also, it is noteworthy that the patient’s CECC was at a sufficient level to withstand another intraocular surgery. The minimum recommended CECC for phakic IOL implantation is 2500 cells/mm² in patients over 21 years of age. The results from various studies on ICLs reveal an initial loss of endothelial cells; i.e., probably related to the surgical procedure, followed by stabilization over time. A large clinical trial showed that the cumulative corneal endothelial cell loss was between 8.4% and 8.9% over the first 3 years and between 8.4% and 9.5% over the first 4 years depending on the method of calculation. In this particular case, the initial cell loss was predictably higher due to increased anterior chamber manipulation, but the long-term variation in the endothelial cell density of this patient tends to approximate that obtained in these clinical trials.

Given the patient’s intolerance to colored contact lenses and the poor results obtained in the first experiments with phakic iris implants, the only alternative solution to manage the dysphotopic complaints would be to perform a corneal tattoo. Good functional and cosmetic outcomes have been reported with keratopigmentation using new micronized mineral pigments. Alió et al. recently reported the successful management of a case of Urrets-Zavalia syndrome using a double intrastromal tunnel femtosecond-assisted keratopigmentation technique.

In our opinion, posterior chamber ICL implantation with iris cerclage is another effective and safe approach to manage this syndrome, provided the patient has normal endothelial cell counts and morphology. Moreover, it enables the surgeon to manage the refractive error and the iris atrophy in the same procedure while sparing the healthy corneal tissue.

REFERENCES