We present a case of decreased visual acuity due to irregular corneal astigmatism associated with intractable light sensitivity because of a fixed dilated pupil (Urrets-Zavalia syndrome) after penetrating keratoplasty. Because the corneal astigmatism was highly irregular, the corrected distance visual acuity (CDVA) was limited to 20/200 and the patient was dependent on a rigid contact lens, with very poor tolerance. The problem was addressed with a novel small-aperture supplementary intraocular implant, which reduces light entrance and minimizes the impact of corneal aberration on the optical system based on the pinhole principle. After implantation of the device, light sensitivity decreased markedly and the CDVA improved significantly, enabling the patient to discontinue contact lens use.

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High residual corneal astigmatism is a common problem after penetrating keratoplasty (PKP) and can be caused by preoperative, intraoperative, and postoperative factors.1 When astigmatism is regular, good visual acuity may be achieved with glasses. If there is an irregular component to the astigmatism, cylindrical lenses do not offer appropriate optical correction.1 Astigmatism management techniques such as suture adjustments, astigmatic keratotomy, wedge resection, and laser corneal ablation are often used, especially in postoperative regular astigmatism.1 However, these options are not appropriate in cases of irregular astigmatism. Historically, irregular corneal astigmatism has occurred in up to 20% of patients after PKP.2,3 This is an important cause of visual acuity limitation of clear grafts.2 Rigid gas-permeable (RGP) contact lenses are the ultimate treatment option available, but intolerance, bad manual dexterity, and infection risk limit their practical application. Surgical options include intrastromal corneal ring segment (ICRS) implantation, topography-guided laser ablation, and regrafting,3–5 but the results are somewhat inconsistent. Urrets-Zavalia syndrome is a serious complication associated with corneal transplantation. First described in 1963,6 it is characterized by diffuse iris atrophy and fixed mydriasis as a result of occlusion of iris vessels and consequent iris ischemia. The pinhole principle is based on the exclusion of peripheral ocular light rays of the image formation process, extending depth of focus. Since paraxial rays are less susceptible to optical aberrations, the result is a better image resolution. This principle is frequently applied in the field of ophthalmology for diagnostic reasons. When irregular corneal astigmatism is present, pinhole acuity testing usually yields a better vision.
than the best possible refraction because it limits image formation to paraxial light rays only. We present the case of a post-PKP patient who had Urrets-Zavalia syndrome and high residual irregular corneal astigmatism. This patient was treated with a device that was designed to act as an intraocular pinhole.

CASE REPORT

A 68-year-old man had had PKP in both eyes for the treatment of keratoconus 30 years earlier. Because of Urrets-Zavalia syndrome in the left eye, he ended up with a fixed dilated pupil and diffuse iris atrophy (Figure 1). There was also significant irregular residual corneal astigmatism after complete suture removal (Figure 2). The simulated keratometry (K) values were 57.2/57.9 @ 72. The patient reported that after corneal transplantation his vision did not improve, not even with glasses.

Twenty-five years after the corneal transplantation, the patient had routine cataract surgery in his left eye with in-the-bag implantation of an aspheric intraocular lens (IOL) (Acrysof SN60WF, Alcon Laboratories, Inc.). After cataract surgery, he complained of minimal improvement in visual acuity and worsening of his light sensitivity. The uncorrected distance visual acuity (UDVA) after cataract surgery was 20/800 and less than J6, and the corrected distance visual acuity (CDVA) was 20/200 with −3.50 −4.25 × 90 and J5 with +3.0 diopter (D) addition (add) (Figure 2). Because of his poor CDVA, the patient was dependent on the RGP contact lens, with very poor tolerance. The CDVA with the RGP contact lens was 20/50 and J2 with +3.0 D add. The right eye had a clear corneal graft and a normal iris, with good vision with an RGP contact lens, which the patient tolerated better than the left one.

After informed consent explaining the potential risks and benefits, it was decided to address this case with the small-aperture implant (Morcher GmbH). The device was designed to be implanted in the ciliary sulcus of pseudophakic eyes. It is made of black hydrophobic acrylic with a 1.5 mm central opening. It has a 13.5 mm overall diameter and a 6.0 mm “optic.” The 3-closed-loop haptic design is to ensure good centration and stabilization in the ciliary sulcus. The implant thickness is 350 μm and the optic has a concave-convex design to prevent contact with the primary IOL. The haptics are angulated 10 degrees to prevent iris chafing and pigment dispersion (Figures 3 and 4). They are thin, rounded, and extremely well polished. Acting as an intraocular pinhole, the implant improves image quality in highly aberrated eyes.

The implant is easily folded into a cartridge and can be injected through a 2.2 mm corneal incision under an ophthalmic viscosurgical device. It unfolds gently inside the eye and can be positioned in the ciliary sulcus with the help of a Sinskey hook (Video 1, available at: http://jcrsjournal.org).

One week after the implantation surgery (Figure 5), the intraocular pressure was 15 mm Hg and the patient noted remarkable improvement in CDVA and reduction of light sensitivity. One month after surgery, the UDVA was 20/60 and J2 and the CDVA was 20/30 with −3.00 −4.25 × 120 and J1 with +3.00 D add. Eight months after surgery there were no ocular complications.

DISCUSSION

The intraocular implant, which is based on the pinhole principle, represents an alternative to improve vision in cases of irregular corneal astigmatism and may be used in addition to other treatment options (ICRS implantation, topography-guided laser ablation). In this

Figure 1. Urrets-Zavalia syndrome: diffuse iris atrophy with paralytic mydriasis (9.0 mm pupil) after PKP.

Figure 2. Corneal tomography reveals an irregular corneal surface.
case, we addressed not only irregular corneal astigmatism but also disabling light sensitivity.

This patient came for consultation 12 months after cataract surgery because glare and light sensitivity had worsened significantly. Because he was not complaining of glare, the surgeon did not opt to address the iris defect during cataract surgery. Currently, there are many options for the correction of iris defects, which could have been performed, preferably at the time of cataract surgery.

Artificial iris implants could be an interesting option for the treatment of light sensitivity; however, the central opening of these devices is relatively large (3.5 mm or larger) to induce a pinhole effect and improve vision. Some of the implants are much more appealing in cases of light-colored irides because of an outstanding cosmetic result.

Colored soft contact lenses and corneal tattooing are other treatment options. However, they are less physiological since the pseudo-pupil is placed away from the iris plane. The poor cosmetic result may also be a relevant issue.

In some cases of large iris defects, the opacification of the anterior capsule rim may create an artificial iris effect. However, to induce a pinhole effect, the capsulorhexis would have to be very small or a severe and treacherous capsule contraction syndrome would have to occur. Furthermore, the amount of opacification of the anterior capsule is somewhat unpredictable and may not be able to minimize glare.

In this particular case, the small difference between flat K and steep K in the simulated K may lead to a false impression that this is a regular cornea. In fact, this corneal surface is very irregular, which is more evident under Placido disk keratoscopy (very distorted mires). This is the reason the patient’s visual acuity was very poor, even with glasses. We have implanted this device in cases of keratoconus, pellucid marginal degeneration, post-radial keratotomy, and post-PKP with larger amounts of astigmatism detected by simulated K, and the results were very encouraging.

To determine whether a patient is a good candidate, a simple pinhole acuity test should be done. A detailed manifest refraction should also be performed since many patients with irregular astigmatism may achieve useful vision with spectacles, avoiding surgery. Rigid contact lenses yield the best possible vision in these cases and should always be encouraged.

To avoid iris chafing and uveitis-glaucoma-hyphema syndrome, which has been reported with sulcus placement of 1-piece acrylic IOLs, the design and material of this implant were carefully selected. The problem of iris chafing is not a result of a specific type of haptic or material. It is best explained by a series of characteristics of a certain design (small overall diameter, planar design, thick haptics, rough edges of haptics and optic). We must remember that we have reports of iris chafe and pigment dispersion even with 3-piece IOLs (blue poly[methyl methacrylate] haptics and sharp-edged acrylic optic). The ideal haptic should be thin, rounded, angulated, and polished and, at the same time, strong enough to ensure centration and stabilization.
The intraocular implant has a larger overall diameter (13.5 mm) than most 1-piece IOLs for in-the-bag implantation. The 350 μm thick haptics are rounded and extremely well polished (Figures 3 and 4). They are 19% thinner than the Acrysof platform, and only 6% thicker than the Sulcoflex platform (Rayner Intraocular Lenses Ltd.), a 1-piece acrylic IOL that has proven to be safe for sulcus implantation. This first-generation prototype has already evolved into a second generation, with 250 μm thick haptics (28% thinner) that facilitate implantation. The 360-degree rounded and polished edge of the optic and the 10-degree angulation of the haptics prevent iris chafing.

The 1.5 mm pinhole central opening is very close to the nodal point of the eye, minimizing the impact of this diaphragm on the visual field. The implant is made of a special hydrophobic acrylic that blocks visible light but is totally transparent to infrared (IR) wavelengths (Figure 6). Therefore, high-definition IR images of the retina can be obtained with equipment that operates in this portion of the light spectrum, such as optical coherence tomography and scanning laser ophthalmoscopy.

Based on the initial results, we believe this technology is very promising and can be an alternative in the treatment of irregular corneal astigmatism and iris defects. This is the first case reported using this new technology. Long-term safety and efficacy of this platform must be further investigated.

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