Traumatic Aniridia and Aphakia

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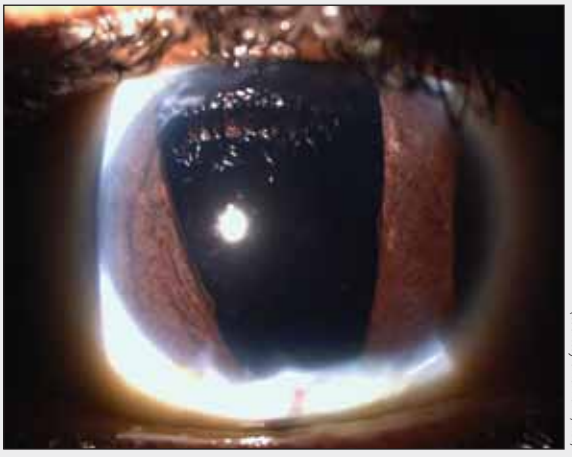
CASE PRESENTATION

A 41-year-old man presents for the surgical correction of aphakia in his left eye. He has a history of trauma to this eye at age 5, followed by "eye surgery" at age 11. The examination reveals an atonic traumatic iris defect (Figure 1). The patient's refraction is -3.00 D sphere = 20/20 OD and +10.00 -4.00 X 180 = 20/30 OS. Keratometry confirms 4.00 D of with-the-rule corneal astigmatism. His BCVA with a rigid gas permeable contact lens is 20/20 OS, but he is contact lens intolerant. What would you offer this patient for surgical rehabilitation of his aphakia and iris defect?



(Courtesy of Tal Raviv, MD)

Figure 1. An aphakic eye suffered a traumatic loss of iris tissue.

AMAR AGARWAL, MS, FRCS, FRCOPHTH, AND DHIVYA ASHOK KUMAR, MD

The case described seems challenging due to the aphakia, high corneal astigmatism, and iris defect. Without capsular support, the IOL cannot be placed in the sulcus. Because of the preexisting iris defect, the use of an iris claw lens would be difficult. The ideal option here would therefore be to implant an IOL using the glued IOL technique[1] (Figure 2) and to repair the iris defect.

We would calculate the IOL's power using the SRK II formula with a target of emmetropia. Initially, we would repair the iris defect with a modified McCannel suture[2] using 10–0 Prolene (Ethicon, Inc., Somerville, NJ). Next, we would create two partial-thickness scleral flaps of about 2.5 mm X 3 mm exactly 180° apart. Either a 23-gauge sutureless trocar infusion cannula or an anterior chamber

(Courtesy of Amar Agarwal, MS, FRCS, FRCOphth, and Dhivya Ashok Kumar, MD.)

Figure 2. In the glued IOL technique, the lens' haptics are externalized under scleral flaps created through a sclerotomy 1 mm from the limbus (A). The surgeon then tucks the haptics into a scleral pocket created by a 26-gauge needle (B). Fibrin glue seals the haptics to the sclera and seals the scleral flaps (C).

maintainer could be placed. Using a 20- or 22-gauge needle, we would make two straight sclerotomies under the existing scleral flaps about 1 mm from the limbus. A foldable three-piece lens from Abbott Medical Optics Inc. (Santa Ana, CA), Alcon Laboratories, Inc. (Fort Worth, TX), or Bausch + Lomb (Rochester, NY) would be appropriate. After implanting the IOL through the superior corneal-limbal incision, we would close the limbal wound with 10–0 monofilament nylon sutures. The haptics would be externalized and tucked at the edge of the flaps by means of a tunnel created with a 26-gauge needle. The advantage of a three-piece IOL is that its haptics will not break during externalization. Fibrin glue (Tisseel; Baxter Healthcare Corporation, Glendale, CA) would seal the haptics to the sclera and seal the flaps.

A limbal incision reduces preexisting astigmatism to a certain level.[3] A combination of surgical procedures is required in challenging cases such as this one to provide good visual and cosmetic results.

BORIS MALYUGIN, MD, PhD

This patient definitely needs to have an IOL implanted in his left eye, but the surgeon must first address several important issues, including the type of lens and its fixation as well as restoration of the iris diaphragm.

I would suture the IOL to the ciliary sulcus. Which model of IOL to use depends on the technical possibility of performing an iridoplasty in order to restore the diaphragmatic function of the pupil and repairing the iridodialysis. In Figure 1, the iris is incarcerated in the scleral-corneal scars, and a significant amount of iris tissue has been lost. Based on these findings, it will likely be impossible to form the pupil by suturing both parts of the iris. I would therefore implant an iris prosthetic device.

I also have some experience with the PMMA lenses manufactured by Morcher GmbH (Stuttgart, Germany; not available in the United States) that have a black periphery. I find they work very well but require a large incision for implantation, so they are not currently my preference.

Here in Russia, a company manufactures the foldable IOL-iris prosthetic complex in different colors (currently, 16 variations) with the results in the peer-reviewed literature.[4] I have had a good experience with this type of implant in my clinical practice and would probably choose it in this case.

An important consideration not discussed in the case presentation is the IOP. If it is close to 10 mm Hg, this finding together with the presence of the iridodialysis might indicate a ciliary body detachment. In that case, I would perform gonioscopy and/or ultrasonic biomicroscopy preoperatively, and if the ciliary body had become detached, I would address the problem during the surgical procedure. Of course, the IOP may be normal preoperatively, but such significant trauma usually dramatically affects the trabecular meshwork. That is why IOP spikes would be highly likely in the postoperative period. I would warn the patient about the possibility that he will need pressure-lowering therapy after surgery, and I would also discuss with him the option of glaucoma surgery.

Last but not least, I would try to address this patient's high astigmatism by making the clear corneal incision in the superior limbus for the IOL's implantation. An against-the-rule shift should occur after the suture's removal. The residual astigmatism should be corrected by spectacles.

DAVID C. RITTERBAND, MD

First, I would discuss realistic expectations with the patient. His eye is amenable to surgical correction, but he must be aware that more than one surgery may be required. He will likely need spectacle correction and some astigmatic treatment. He must understand that the appearance of his pupil can be improved but that it will neither function normally nor appear "normal."

I do not believe there is enough iris tissue to allow fixation of an IOL to the iris through the placement of McCannel sutures. Scleral fixation of a CZ70BD IOL (Alcon Laboratories, Inc.) would be necessary and could be performed before possible work on the iris. I would aim for a spherical equivalent of around -3.00 D. Then, a minimum of 6 weeks after surgery, I would consider performing laser refractive surgery or astigmatic keratotomy to correct the residual cylinder.

If Figure 1 shows all that remains of the iris after the instillation of a miotic, I do not believe a cosmetic appearance can be achieved by suturing that rivals the new technology available in Europe and used on a compassionate basis in the United States. Although the Ophtec model 311 aniridia lens (Ophtec BV, Groningen, the Netherlands) can be fixated sclerally and provides both an IOL and an artificial iris, it only comes in four color palettes and looks quite synthetic in eyes with residual iris. A more pleasing cosmetic appearance could be achieved with a foldable, sutureable artificial iris implant (Artificial Iris; Dr Schmidt Intraoculalinsen GmbH, SanktAugustin, Germany; http://artificial-iris.com), the color of which can be customized using photographs of the remaining iris.

GABOR B. SCHARIOTH, MD

This complex posttraumatic case requires an intensive preoperative discussion with the patient, because I envision various options for treatment.

My first choice would be a secondary implant with sutureless intrascleral fixation of the haptics.[5,6] I would place the incision superiorly to reduce the astigmatism. It would be very important to have continuous anterior chamber or pars

plana infusion during surgery to reduce the need for an ophthalmic viscosurgical device, because the viscoelastic material would be lost into the vitreous cavity. After the peritomy, I would create two sclerotomies in the ciliary sulcus, 180º from each other, and two limbus-based, parallel, intrascleral tunnels starting from these sclerotomies. Next, I would inject a three-piece IOL (AR40e; Abbott Medical Optics Inc.) onto the iris' surface. I would then grasp one haptic with an endoforceps through a sideport incision and, with a handshake technique, present it to the forceps introduced through the sclerotomy. It is important to grasp the very tip of the haptic. After externalizing this haptic, I would proceed in the same fashion with the second haptic. Then, the goal would be to introduce each haptic with a special 25-gauge forceps (DORC International BV, Zuidland, the Netherlands) into the limbus-parallel tunnel. With this technique, contact with the iris and uvea, tilting and decentration of the lens, and costs are minimal. I would target a minimally myopic postoperative refraction of about -0.50 to -1.00 D. If the patient desired an improved refractive result, I could offer refractive corneal surgery (ie, LASEK). I have had good results with this approach.

Because the case presentation does not indicate that the patient has photophobia, I would not touch the iris in a primary surgery. If needed, in a second attempt, iris sutures or an Artificial Iris could be placed.

My colleagues and I have already used the described technique for fixating a multifocal IOL. This option would have to be discussed with the patient, since he is only 41 years old. I believe that this approach would require repair of the iris defect, and I would select the three-piece Tecnis Multifocal IOL (Abbott Medical Optics Inc.) and bioptics for the treatment of residual refractive error.

SHACHAR TAUBER, MD

The conservative treatment of a rigid gas permeable contact lens has served this patient well for 30 years. It should not come as a surprise that he has developed intolerance of the lens after such a long period. Before embarking on surgical intervention, however, the treating physician should confirm that all possible medical therapies to reverse contact lens intolerance have been attempted, including the management of blepharitis and dry eye syndrome and the use of improved polymers and hybrid lenses.

Assuming that the patient is a surgical candidate, both his aphakia and iris defect will need to be corrected, preferably in one straightforward surgery. The use of an iris color-matched PMMA aniridia lens has been shown in several reports to have great success.[7,8] Recently, Dr. Agarwal described the use of this device in a sutureless sulcus-fixated technique in which the haptics are secured in a scleral tunnel.[1]

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Preoperatively, I would assess the patient's risk factors for corneal and retinal disease and for glaucoma by means of slit-lamp biomicroscopy, gonioscopy, endothelial cell counts, pachymetry, optic nerve analysis, and a careful examination of the macula and peripheral retina. I would use immersion biometry and my preferred IOL formula to order the appropriate aniridia lens from the manufacturer.

Before surgery, the patient would receive a topical nonsteroidal anti-inflammatory drug and a fourth-generation fluoroquinolone in an effort to prevent cystoid macular edema and endophthalmitis. At present, aniridia lenses are made of PMMA and thus are not foldable, so a large, shelved corneal-scleral incision is needed. I would perform a triamcinolone-assisted vitrectomy, preferably with the vitreous cutter placed through the pars plana, to ensure that the anterior chamber was free of vitreous traction. I would then place two sclerotomies under scleral flaps 180º apart and each 1.5 mm from the limbus. I would introduce the aniridia lens into the anterior chamber after filling it with viscoelastic. Each haptic would be externalized through the sclerotomy and placed through a scleral tunnel. Care must be taken with these haptics, because their angulation differs from that of most common IOLs. I would close the wound under keratometric control with 10–0 nylon sutures and prescribe an appropriate antibiotic steroid and nonsteroidal anti-inflammatory drug. Postoperative care would be similar to that for sulcus-fixated secondary IOL surgery, including careful attention to the patient's IOP and an examination of the peripheral retina and macula.

Although neither the surgery nor the surgical decisions required are complex, one issue will cause many US surgeons to consider delegating this procedure to large institutions: permission to use an aniridia lens requires an onerous application to the FDA for a human device exemption. Fortunately, the companies (Ophtec BV and Morcher GmbH) that produce the iris diaphragm lens as well as the endocapsular iris implants have a wonderful reputation for assisting US surgeons as they complete the mandated paperwork. I can only hope that, one day soon, these lenses will be available to US surgeons without this bureaucracy. ■

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