Uniocular Axial Congenital Cataract

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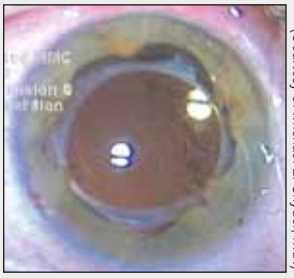
CASE PRESENTATION

A now 3-year-old female was born with a uniocular axial congenital cataract in the central fetal nucleus. Sphincterotomies were performed to create an optical iridectomy by enlarging the pupil in an effort to allow the child to see around the cataract. Despite her compliance with patching therapy, amblyopic treatment failed to progress. At 2.5 years of age, the patient underwent a cataract extraction, primary posterior capsulorhexis, and the placement of a PCIOL with a targeted refraction of +4.00 D. Amblyopic therapy continued without success. In fact, the parents note that, in any outdoor setting as well as indoors in rooms with abundant ambient light, the young girl either squints or closes the affected eye, thus possibly adding a component of occlusion to the amblyopia.



(Courtesy of Michael E. Snyder, MD.)

Figure 1. An examination with the patient under anesthesia shows the resting state of the iris and the IOL in position. The white letters on the screen are an artifact of the video overlay.

An examination of the patient's unaffected fellow eye is unremarkable and demonstrates 20/20 vision. The operated eye sees 20/200 (Allen card pictures) with best correction in a darkened room. The child closes the eye when the room's fluorescent lights are on. The pupil measures 8 mm in its resting state. Numerous sphincterotomies are visible circumferentially, and a single-piece PCIOL is well positioned in the capsular bag (Figure 1). A posterior capsulorhexis is present with a thin layer of lens epithelial cells across the hyaloid face. Fundus findings are unremarkable.

What would you recommend for this patient?

M. BOWES HAMILL, MD

The patient's pupil is too large, and enough light is getting around the lens' edge to cause photophobia, which interferes with her vision and consequently contributes to the failure of amblyopic treatment. Although optical sphincterotomies can be very helpful for localized lenticular opacities, they have unfortunately resulted in a large pupil in a pseudophakic eye. When creating optical iridectomies, I have found it helpful to make two radial incisions into the iris that start from a common point on the iris sphincter. The orientation and angle of the incisions are determined by the location of the opacity and the required degree of pupillary opening. The resultant keyhole-shaped pupil allows the patient to see around the opacity. An advantage to this approach is that, in the event that the opacity is removed at a later date, it is fairly easy to repair the incisions by joining the apex of the incised tissue to the adjacent pupillary sphincter's edges. Frequently, only a single stitch is required.

In this case, one could consider closing each sphincterotomy individually. Given the appearance of the iris in Figure 1, however, this approach would be technically difficult. I would prefer to lyse any posterior synechiae so as to free up the iris. Subsequently, a 360º cerclage suture could be passed in the method of Ogawa to remanufacture a circular pupil. I would close any visually significant gaping of the iris radial to the cerclage suture with individual sutures. Because of the poor healing qualities of the iris, I would employ a permanent suture such as 10–0 Prolene (Ethicon Inc., Somerville, NJ). The cerclage suture also has the benefit of allowing the surgeon to manufacture a pupillary aperture of whatever size desired. For this as well as most cases, a midposition pupil is probably ideal for giving reasonably good cosmesis, allowing a sufficient opening to permit a fundus examination in the future, and blocking enough light to prevent photophobia in normal lighting conditions. Another advantage of the cerclage approach over closing each sphincterotomy with an individual suture is that, should further surgical intervention be

required, the surgeon could open the entire pupillary aperture by removing the single cerclage suture, thus allowing the pupil to dilate to its preoperative state.

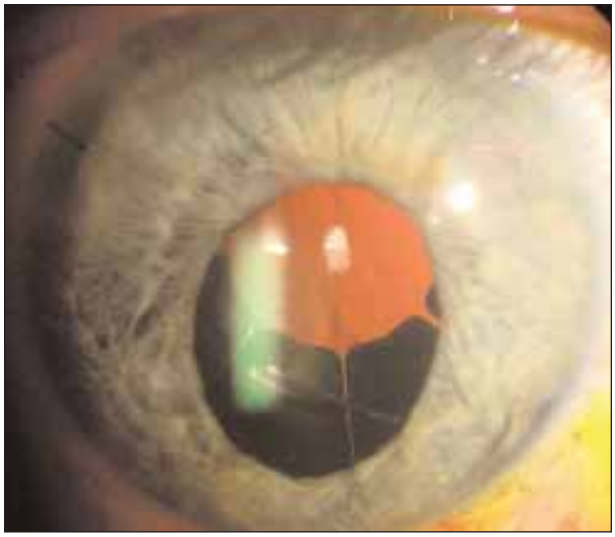
SAMUEL MASKET, MD

Prior to the development of current, effective surgical methods and appropriate lens implants for managing pediatric cataracts, surgeons performed optical iridectomy in an attempt to improve vision and allow the treatment of amblyopia. We now recognize, however, that enlarged or deformed pupils are associated with undesirable glare disability.[1] Given the general success of present-day cataract surgery for children, I believe optical iridectomy to be an obsolete procedure and would not have considered it for this child. Moreover, the surgeon could have considered pharmacologic pupillary dilation as a reversible alternative.

Although this patient seems to be significantly photophobic, children with amblyopia or strabismus often close the involved eye in light, because suppression is hindered under bright lights; this phenomenon may persist throughout life despite treatment. Nevertheless, we should assume that at least part of this child's response to light is due to her enlarged pupil and should address this problem surgically.

Another matter of concern is the opacified hyaloid face. Although the posterior capsule was opened during surgery, it appears that an anterior vitrectomy was not performed. Lens epithelial cells may migrate freely across the hyaloid face and use it as a scaffold, which will preclude a clear optical path and contribute to amblyopia. Generally, performing an anterior vitrectomy along with the posterior capsulorhexis prevents this phenomenon. Some surgeons also prolapse the optic of the IOL behind the posterior capsulotomy, as originally proposed by Gimbel,[2] in order to create another barrier to the migration of lens epithelial cells across the visual axis. The clouded anterior hyaloid in this case must also be addressed at surgery for the enlarged pupil.

When planning surgery for complex cases such as this one, the surgeon should have a series of plans in case the preferred course is not possible. My desire would be to reopen the anterior capsulotomy and implant two of the Rasch-Rosenthal-Masket (50E) or aniridic model 50F rings (Morcher GmbH, Stuttgart, Germany) to create a 3.5-mm pupil at the iris plane (Figure 2). Although this approach might be difficult or impossible with an open posterior capsule, in this case, the capsule should be stable after a true posterior capsulorhexis. In the United States, one must obtain an exemption for compassionate use from the FDA in order to implant the Morcher devices. Afterward, a sutured-pupil cerclage would achieve improved aesthetics. If it were not possible to reopen the anterior capsulotomy, I would perform only the cerclage for the enlarged pupil. Given the age of this child, I would hesitate to implant any aniridic devices in the ciliary sulcus. Finally, the opacified hyaloid could be addressed with a single-port pars plana automated vitrector. The IOP could be maintained by infusion through one of the corneal paracenteses that was established for the iris suturing. In a child of this age, the globe should be entered approximately 2.5 mm posterior to the limbus. A limited anterior vitrectomy would clear the visual axis and likely prevent recurrent opacification.



(Courtesy of Samuel Masket, MD.)

Figure 2. Dr. Masket used Rasch-Rosenthal-Masket (50E) aniridic endocapsular devices to create an artificial pupil for a case with a surgically repaired 6-clock-hour iridodialysis.

FRANCIS W. PRICE, JR, MD

No matter what is done, the child will likely remain amblyopic, but her sensitivity to light might be improved. The biggest problem appears to be the large, fixed pupil. The placement of an artificial iris implant could be expensive and require enrollment in a study or a special exemption. The most cost-effective treatment would be to suture the iris to reduce the pupil to a 2- or 3-mm diameter if the iris is not too friable or stiff/scarred. I would use a purse string of 9–0 Prolene on a 4-mm, straight tapered needle. The process can be slow and tedious, but it creates a round pupil. Any of a variety of intraocular forceps might be used to grasp the needle and pass it with multiple bites through the edge of the pupil before eventually tying the suture intraocularly.

FREDERICO F. MARQUES, MD, AND DANIELA M. V. MARQUES, MD

We would not recommend repairing the iris first. An inflammatory response due to the patient's age increases her

chance of developing glaucoma and an inflammatory papillary membrane. Instead, we would first try to optimize the amblyopic eye by using the total refraction (distance and near) on a bifocal, darkened (as in sunglasses) spectacle lens for the patient's affected eye and complete occlusion of the fellow eye. We would re-evaluate her photophobia and BCVA in 3 weeks. If treatment resulted in progress and was acceptable to her parents, we would continue the therapy and reevaluate both of the patient's eyes every 3 weeks. We would take care not to induce amblyopia in her unaffected eye by alternating occlusion (6 days with the unaffected eye occluded followed by 1 day with the affected eye occluded). Future iris repair would be associated with a lesser chance of a strong inflammatory reaction and a better visual outcome.

If therapy with spectacles failed, we would inform the patient's parents that the only remaining option would be to repair the iris, although an improvement in BCVA would not be guaranteed. When reconstructing the pupil, it would be important that the final size not be too small to allow a retinal examination, because this patient is at increased risk for retinal detachment. The pupil must not be so large, however, as to cause photophobia. We think a diameter of 3 to 4 mm would be adequate.

We would try a cerclage technique involving multiple catches of the iris margin over 360º. We would pass 9–0 polypropylene sequentially via four paracenteses to create a pupil of approximately 3.5 mm in diameter. Postoperatively, we would carefully manage ocular inflammation with topical and, if necessary, oral steroids. Occlusive therapy would proceed as described earlier.

KENNETH J. ROSENTHAL, MD

Because the rare contraindication to surgery in infants concerns anesthesia-related risks, the problem now presented could have been avoided had the patient undergone definitive cataract surgery initially. In any case, when cataract surgery was eventually performed, it should have incorporated iris repair.

The patient now experiences a reduced quality of vision because of:

(1) ghosting due to the simultaneous projection of both a focused image through the IOL and an unfocused image from light entering peripherally to the IOL onto the retina

(2) increased higher-order aberrations caused by light entering unconstrained through the dilated pupil and incorporating the optics of the peripheral cornea

(3) amblyopia because of voluntary lid closure caused by photophobia.

A trial with a masking contact lens might delay surgery, but, in my experience, patients generally have difficulty wearing such contact lenses. Furthermore, because the condition in this case is unilateral, maintaining a small pupil in mesopic conditions may deepen the amblyopia due to her limited vision through an inappropriately small pupil.

Before surgically repairing an iris, I will normally prepare an iris repair "kit" with a variety of iris prosthetic implants, all of which would require preapproval from the FDA for compassionate use. In this case, I would have available both a "modular," in-the-bag iris prosthesis such as the Rasch-Rosenthal-Masket (50E) Morcher device or the Hermeking Iris Prosthetic System (Ophtec BV, Groningen, The Netherlands), as well as a device intended for placement in the sulcus.

First, I would perform an iris cerclage incorporating both the more centrally located, scalloped pupillary edge (corresponding to the papillary ruff) as well as the curved portion of the iris lying in between these points. I would tie the knot with a temporary single throw. Although the pupil might not reassume its original aperture, it might be within the edge of the IOL. It would be important not to make the suture overly tight, which would create too small a maximal pupil size. An alternative approach would be only to incorporate the more central iris points, which correspond to the original papillary ruff, in a cerclage suture. This technique could be supplemented by multiple interrupted sutures to close the remaining radially oriented defects. In patients as young as this one, I frequently use (off label) a durable suture material such as Gore-Tex (W. L. Gore & Associates, Inc., Newark, DE), because this material does not show signs of biodegradation, is very flexible, and has great tensile strength.

If the cerclage left some gaps through which light might penetrate, or if the iris has lost enough elasticity that the iris sectors cannot be completely approximated, I would untie the cerclage suture and attempt to reopen the capsular bag with a 30-gauge needle or a Chang hydrodissection cannula to lift the edge of the anterior capsulorhexis followed by viscodissection of the IOL from the capsular bag. I would then insert a modular iris prosthetic device. If the bag were too fragile or fibrosed, or if I noticed zonular instability, I would instead suture in a model 311 Ophtec Iris Prosthesis (Sundmacher design; Ophtec BV) without an optic in the sulcus anterior to the existing IOL. Again, I would use Gore-Tex sutures off label. This approach would require a 10-mm incision, which lengthens healing time and can result in surgically induced astigmatism. I would then retie the cerclage suture(s).

All of these solutions would produce a 4-mm pupillary aperture, adequate for comfort in average lighting conditions, good visual acuity, minimal higher-order aberra-

tions, and reduced ghost images.

One might wish to measure the anterior segment structures prior to surgery. Specifically, one could measure the white-to-white and sulcus-to-sulcus diameters with imaging technology such as anterior segment optical coherence tomography or Scheimpflug photography. One could then order a custom sized iris prosthesis if these structures are smaller than in an adult.

Vigorous amblyopia therapy should recommence as soon as adequate healing has taken place.

JOHN C. HART, JR, MD

The timing of cataract surgery in patients with unilateral congenital cataract is paramount. In infants, not only does the cataract blur the image, it also disrupts the development of the visual pathways in the central nervous system.[3] The critical age for the surgical treatment of dense, unilateral cataracts is from birth to 6 weeks. Thereafter, the chance of improving visual acuity beyond 20/80 decreases.[4] A trial of patching may be indicated, if the level of visual loss seems disproportionate to the density and size of the cataract. The pharmacologic dilation of the pupil can occasionally be helpful as an adjunctive treatment.[5] In this case, cataract surgery was delayed, thereby limiting the potential for the patient's visual rehabilitation from amblyopia. The iridotomy performed instead leaves the patient with debilitating glare, which may exacerbate her amblyopia.

The patient squints or closes the affected eye in any lighted setting. Potential factors contributing to this problem include strabismus, glare from light entering the eye outside the IOL, and increased spherical aberration from light entering the eye unrestricted by the pupil.

Initially, the patient should be evaluated for strabismus. Then, placing an opaque soft contact lens with a small pupillary opening would eliminate glare from the iridotomy as well as correct residual refractive error and decrease spherical aberration. If the patient will not tolerate a contact lens, then the surgical repair of the iris could be considered to make the pupil more physiologic in size and thereby decrease glare and spherical aberration. Because simple sphincterotomies were used to enlarge the pupil, presumably, no iris tissue was removed.

I would begin by creating four 1-mm paracenteses at the limbus 3 clock hours apart. Ectropion uvea, caused by membranes on the surface of the iris, is apparent along the two superior sphincterotomies. A small amount of Triesence (Alcon Laboratories, Inc., Fort Worth, TX) instilled in the anterior chamber would help in the identification of these membranes and decrease postoperative inflammation. I would inject a dispersive ophthalmic viscosurgical device into the anterior chamber to provide stability and create working space. Gently stretching the iris with Ahmed microgaspers (Microsurgical Technologies, Redmond, WA) would release adhesions within the iris. After stripping the membranes identified on the iris, I would perform a sphincterotomy repair using a 10–0 Prolene suture on a CTC 6-L needle (Ethicon Inc.). First, I would place a simple interrupted suture near the pupillary border to oppose the cut sides of a sphincterotomy. An additional interrupted suture might be required to close the sphincterotomy completely. The six larger sphincterotomies would need closure. To perform this suturing technique, I would pass the needle through a paracentesis, grasp the iris with microgaspers, and pass the needle through the iris. Then, I would grasp the adjacent iris with the microgaspers to drive the needle through. I would dock the needle tip in a 27-gauge cannula to guide the needle out of the anterior chamber, through a paracentesis. I would then tie the suture with a Siepser sliding knot.

After the surgical repair of the iris, amblyopia therapy should resume. I would explain to the patient's parents that the prognosis for further visual recovery was extremely guarded.

KEVIN M. MILLER, MD

The primary outcome of any intervention should be a reduction in the patient's sensitivity to light and glare. Any gain in visual acuity should be considered secondary.

I would have done several things differently before the patient reached this point. I think the adverse consequences of a large optical iridectomy in a young child far outweigh any of the procedure's small benefits. Cataract surgery should have been performed first, if the cataract was visually significant at the time of the iridectomy. Moreover, I would have targeted the eye for emmetropia and placed the lens in the ciliary sulcus. It is highly likely that the patient would have been myopic by the time she became a teenager or young adult. I would have planned to exchange the implanted IOL for an emmetropic sulcus-fixated lens when her eye's growth stabilized. This approach would have made myopic amblyopic therapy much easier than it will be now.

The pupil of the affected eye is markedly enlarged, and its margin is irregular from the posterior synechiolysis. There appear to be multiple small posterior synechiae, although a slit-lamp examination would be necessary to confirm this observation. The posterior capsulorhexis seems small, and the peripheral posterior capsule appears to be mildly, if not significantly, opacified. Lastly, Elschnig pearls are present on the anterior vitreous face. It is the combination of the large pupil, the opacified peripheral posterior capsule, and the lens epithelial cells proliferating across the anterior hyaloid that is contributing to her exquisite sensitivity to glare.

I would recommend a pars plana vitrectomy to remove the lens epithelium from the anterior hyaloid. At the

same time, the posterior capsulorhexis should be enlarged to just inside the border of the IOL's posterior edge. I would then perform a purse string pupilloplasty to bring the pupil down inside the anterior capsulorhexis. It is questionable whether a 10–0 Prolene suture will last the rest of the patient's life and not cheese wire through the pupil, but simply removing most of the anterior hyaloid opacification and posterior capsular opacification will significantly reduce her sensitivity to glare. If the purse string pupilloplasty does not last, it can be repeated.

One could consider implanting a foldable artificial iris (HumanOptics AG, Erlangen, Germany), but I do not know whether the FDA would grant the necessary exemption for compassionate use for a 3-year-old. I would also worry about the long-term sequelae of a new product. I would not implant devices from Morcher GmbH or Ophtec BV intended for the capsular bag in this patient. First, sulcus-fixated devices do not work very well in my experience. Second, the size of incision that would be necessary for a sulcus-based implant would not be warranted in a young child.

After the vitrectomy and purse string pupilloplasty, I would allow the patient time to heal and then fit her with a contact lens to correct her hyperopia, possibly targeted for mild myopia. I would resume the patching therapy. At age 3, it is likely that she will experience some gain in vision once she is able to use the eye under standard lighting conditions.

The long-term visual prognosis for this patient is relatively poor. Deprivation amblyopia secondary to congenital cataract is very difficult to treat. Nevertheless, it would be worth a concerted effort until the patient enters elementary school. The reduction in her sensitivity to light and glare that surgery could provide should be of major benefit to her. ■

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