TECHNIQUE

Congenital iris coloboma repair with excision of colobomatous sphincter muscle

Gregory S.H. Ogawa, MD

Congenital iris colobomas do not usually present a major optical issue until the time of cataract surgery, when an intraocular lens is placed in the eye that is roughly half the diameter of the crystalline lens. Leaving the coloboma unrepaired or sutured closed without addressing the sphincter muscle in the coloboma often creates visual challenges for the eye postoperatively. The problem has previously been addressed, in part, with a technique that creates a scissor snip between the normal iris sphincter and the colobomatous iris sphincter, but still requires notable peripheral iris traction and root disinsertion for closure of the defect. The technique presented here removes all iris sphincter from the sides of the coloboma allowing closure of the colobomatous defect without the need to create iridodialyses. In certain cases, the use of iris diathermy can be used to create focal iris contraction to maximize sphericity and centration of the pupil.

Optic colobomas result from the failure of the optic fissure to close during embryogenesis. They can involve the iris and structures all the way posteriorly to the optic nerve. A host of associated genes and syndromes have been identified, including autosomal dominant forms that affect only the eye. The impact on vision can be minimal to total depending on the structures involved. Currently, only iris coloboma can be treated surgically.

A congenital iris coloboma develops in a manner that leaves intact and functioning sphincter muscle along its sides (see Supplemental Digital Content 1 and 2, Figures 1 and 2, available at http://links.lww.com/JRS/A220 and http://links.lww.com/JRS/A221). Congenital iris colobomas rarely need repair until the time of cataract surgery. The crystalline lens goes to, or near, the periphery of the iris coloboma, in contrast to the optic of an intraocular lens (IOL), which is generally only about half the diameter of the crystalline lens. If a surgeon performs only cataract surgery, the patient often winds up with poor quality of vision. This is due to light entering the colobomatous pupil and then going through (1) an aphakic space peripherally, (2) the peripheral lens capsule, which can become fibrotic and cause light scatter, (3) the edge of the IOL more centrally, and then (4) the optic of the IOL further centrally. These factors can even lead to patients with concomitant posterior segment colobmas involving the fovea to have challenges with an unrepaired iris coloboma after cataract surgery.

The initial iris coloboma repair procedures by many, including the author, consisted of primary closure of the coloboma at the end of cataract surgery, which generally produced small, ineffective, inferiorly located pupils, depending on the configuration of the presenting coloboma. An interesting variation of primary closure published in 1993 created a complete superior iris incision, which was then closed after primary closure of the coloboma to be able to help center the pupil.

An important advancement in the surgical treatment of congenital iris coloboma became widely known with the 2006 publication by Cionni et al. that addressed the presence of sphincter muscle in the coloboma and created a scissor snip through the sphincter at the junction of the normal-appearing iris sphincter and the sphincter in the coloboma. This technique still required some mechanical disinsertion of the peripheral part of the coloboma sphincter, but generally allowed for a better centered pupil than with primary closure alone.

In the mid-1990s, the author had the experience of creating a small, very eccentric, nonfunctioning pupil (when seen postoperatively), using the primary closure alone technique. After analyzing the situation and the role
of the coloboma sphincter, the author returned to the operating room and performed the sphincter-removing technique described below, which the author has continued to use, in dozens of cases, successfully since that time.

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This technique is specifically for iris coloboma repair in pseudophakia with a posterior chamber lens. It can be performed at the time of cataract surgery or as a secondary procedure after a previously performed cataract surgery that did not adequately address the iris coloboma.

Iris coloboma work generally needs a greater level of anesthesia than the topical anesthesia under which one commonly performs cataract surgery. The surgeon needs both reasonable ocular akinesia and patient comfort to perform the iris reconstruction. Anesthesia options include a retrobulbar block, a peribulbar block, or anesthetic injected subconjunctivally and massaged posteriorly with a muscle hook with the option of superior and inferior rectus bridle sutures for stability. General anesthesia would be reserved for select patients.

If the surgeon repairs the congenital iris coloboma at the time of cataract surgery, the cataract surgery and IOL placement should occur first. In some instances, the pupil dilates adequately with pharmacologic agents, and in other instances, the surgeon needs iris retractor hooks on the far side of the eye from the coloboma to create a large and well enough centered pupil to perform the cataract/IOL procedure. Depending on the degree of focal lens zonule absence in the coloboma region, the surgeon may need to use an intraoperative blunt-tipped capsule support hook during surgery mainly to help prevent vitreous prolapse and secondarily for capsular bag support. A capsular tension ring may be of use for the postoperative course for the same reasons. Rarely, a sutured capsular tension segment or a sutured capsular tension ring may be needed to provide adequate capsule support for IOL placement. In unusual situations, there may be frank vitreous prolapse through the zonular defect necessitating anterior vitrectomy before even starting the cataract surgery. Because the surgeon needs to pharmacologically constrict the pupil during the coloboma repair, one should try to avoid having epinephrine or phenylephrine in the cataract surgery infusion bottle as these agents usually blunt the effect of miotic agents. After completing the cataract surgery and IOL placement, one removes the ophthalmic viscosurgical device and iris hooks before proceeding to the iris repair.

The surgeon may sit superiorly or temporally to the eye during the coloboma work and may even switch positions for different parts of the procedure, all based on surgeon ergonomic comfort. Injecting a miotic agent (acetylcholine) through a paracentesis constricts the pupil—both the colobomatous part and the normal part. A self-retaining infusion cannula goes in through a conveniently placed limbal paracentesis opening to keep the eye at a normal to somewhat above normal intraocular pressure using balanced salt solution delivered via a machine capable of performing vitrectomy with a guillotine cutter. By visual inspection and manipulation with a second instrument, the surgeon determines where the colobomatous part of the pupil ends and the normal part of the pupil begins—for each side of the coloboma. Through an appropriately placed paracentesis, the surgeon uses the guillotine vitrector to remove the iris tissue containing the sphincter muscle along the sides of the coloboma. Starting from the periphery of the coloboma and working centrally toward the normal pupil allows the surgeon to see the amount of relaxation achieved and may affect just how far centrally the surgeon decides to proceed with the sphincterectomy (see Supplemental Digital Content 3, Figure 3, available at http://links.lww.com/JRS/A222); however working from normal sphincter to the periphery of the coloboma also works well. After excision of the sphincter from one side of the coloboma, the same process is performed on the other side (see Supplemental Digital Content 4, Figure 4, available at http://links.lww.com/JRS/A223). After the initial iris removal pass, one may decide to go back to certain parts to remove more tissue that appears to still have some sphincter muscle. The amount of tissue excised is often minimal in order for sphincter muscle to be removed so that only iris stroma remains; this is similar to the amount of iris one might inadvertently remove from a normal pupil to create a discontinuous pupillary sphincter band.

One sets the vitrectomy unit with low flow, low cut rate, and relatively low vacuum. For example, using a peristaltic pump machine with a 23-gauge vitrectomy probe, one might set the flow rate at 12 cc/min, the cut rate at 60 cuts per minute, and the vacuum at about 120 mm of mercury. Twenty-gauge vitrectors generally require lower vacuum, and 25-gauge vitrectors usually require higher vacuum. The bottom line is to start low with the settings and go slowly to avoid excessive removal of iris tissue. One can always turn up machine settings if not getting adequate tissue removal, but restoring iris tissue in this situation is not possible.

After satisfactory sphincterectomy of the colobomatous part of the pupil, the surgeon begins the iris suturing phase with 10-0 polypropylene on a long, curved, trans-chamber type needle. With a small-width spatula needle (eg, CTC-6L, Ethicon), one may pass the needle through the limbus at any point. The surgeon should make the initial suture pass through the iris at the end of the intact normal pupil sphincter on each side of the coloboma. One may find that a second instrument, such as an Ogawa iris reconstruction hook (Duckworth & Kent) or coaxial microforceps (multiple manufacturers), aids in supporting the iris during the needle pass to avoid stretching damage to the iris—especially as the needle goes through the first leaflet of iris tissue (see Supplemental Digital Content 5 and 6, Figures 5 and 6, available at http://links.lww.com/JRS/A224 and http://links.lww.com/JRS/A225). The second leaflet may be
supported by the inside of the limbus during this first needle pass. The surgeon then ties the arms of this suture to create the patient’s new pupil. The author preferentially uses the Ogawa in situ intraocular knot-tying technique for this procedure because of the ability to tie the knots down in the iris plane, which is particularly useful for avoiding iris damage for the most peripheral of the sutures. Excessive peripheral traction can cause dialysis, iris defects, and bleeding. Both ends of the suture are then brought outside the eye through a conveniently located paracentesis opening (see Supplemental Digital Content 7, Figure 7, available at http://links.lww.com/JRS/A226). The suture arms are tied using conventional throws outside the eye. Each throw is then brought just to the outer part of the paracentesis opening with the slack taken out of the suture inside the eye. The suture arms are then laid down on the conjunctival surface next to each other, with one arm having a little more length between the throw and where both sutures are grasped with tying forceps. That extra length creates a partial loop into which an instrument, like an IOL manipulator, is placed to pull that suture arm, and the throw, inside the eye with a pulley-like effect, while the other ends of the suture are still held with the single tying forceps outside the eye. This has 2 beneficial effects: (1) it allows the knot to be tightened with tension on the sutures to be oriented 180 degrees apart (since the IOL manipulator functions as a pulley), and (2) it keeps the knob of the IOL manipulator away from where the throws are being cinched to minimize the chance of inadvertently capturing the knob of the IOL manipulator in the throw’ (see Supplemental Digital Content 9, Figure 9, available at http://links.lww.com/JRS/A228).

When placing coaxial scissors through a paracentesis works well for cutting the tails of the sutures, leaving at least 0.5 mm of tail but not so long as to allow the tails to touch the cornea. After placing the suture at the iris sphincter, the author typically places 2 or 3 more interrupted iris stromal sutures in the defect to complete the closure. There is often a very small, insignificant, triangular defect remaining in the far periphery. (see Supplemental Digital Content 10 and 11, Figures 10 and 11, available at http://links.lww.com/JRS/A229 and http://links.lww.com/JRS/A230). When placing these additional sutures, one may need to support the iris on each side of the defect to avoid iris damage from needle traction. The author ties all the sutures in the same manner. With the colobomatous iris sphincter removed, the remaining stroma stretches well enough that one does not need to create iridodialyses of adjacent, peripheral iris tissue to close the iris defect (see Supplemental Digital Content 12, Video 1, available at http://links.lww.com/JRS/A231).

The pupil centration and sphericity may be optimized using intraocular coaxial diathermy on low settings to shrink iris tissue superiorly, nasally, or temporally. This technique, first presented by Ahmed, is useful for fine-tuning pupil geometry.

If at the end of the case the pupil seems too small, the central most suture can be removed to help enlarge the pupil. The surgeon should be aware that the appearance of the iris/pupil at the end of the case is not always the same as it is a day or 2 postoperatively. When there is a notable appearance difference postoperatively, it is usually in the direction of the pupil being smaller. If the pupil is too small, returning to the operating room to adjust the pupil size is a reasonable option.

Once the surgeon completes closure of the iris defect, they direct attention to achieving a watertight seal at all the paracenteses. The remainder of the case may be done by the surgeon’s standard protocol.

The author has used this technique for a multitude of patients, and it has reliably produced reasonably centered, round pupils, allowing the best vision of which the eye is capable with good to excellent aesthetic results. No returns to the operating room have been needed. Of note, this technique has already demonstrated transferability as the author has remotely taught 2 other U.S. surgeons this technique via videos and written and telephone communication. Both surgeons achieved good to excellent results with their first attempt.

**DISCUSSION**

Congenital iris colobomas vary considerably in exact shape, size, and location from patient to patient, so the execution of the surgical repair needs to be customized to the individual patient’s eye. The described technique provides the tools for achieving a cost-effective, useful result in most situations.

With the availability now in the United States of a custom iris prosthesis, the question arises as to whether that may be a good option for patients with congenital iris coloboma. The flexible iris prosthesis (CUSTOMFLEX ARTIFICIALIRIS, HumanOptics) is excellent at blocking light from going through the peripheral part of the coloboma. For eyes with natural iris blocking the middle of a centered IOL, this would need to be addressed with treatments like chronic pharmacologic dilating drops or excision of central natural iris. The prosthesis cost may be an impediment to its use in some situations. We do not have 25-year follow-up results yet on the available prosthesis, so that is a bit of an unknown. The time needed for device ordering, manufacturing, and delivery requires an engaged patient, so when combining that with cost and the unknown long-term results, the sort of iris repair described here will be a preferable option to a prosthesis for many patients with congenital coloboma.
WHAT WAS KNOWN

- Untreated congenital iris coloboma can negatively affect the outcome of cataract surgery due to corectopia and irregular pupil shape with associated visual artifacts.
- Primary closure of congenital iris coloboma at the time of cataract surgery is of variable visual benefit compared with untreated coloboma.
- Creating a focal separation between the pupil sphincter and the coloboma sphincter, along with disinsertion of the peripheral coloboma sphincter, likely further improves pupil centration and shape during cataract surgery but may attenuate inferior iris tissue.

WHAT THIS PAPER ADDS

- Removal of all coloboma sphincter muscle allows the juxta coloboma iris stroma to be closed without disinsertion of the peripheral sphincter in the coloboma region and likely better pupil centration and shape.
- Intracocular diathermy can be useful in optimizing sphericity and centration of the colobomatous pupil as part of repair.

REFERENCES


Disclosures: None reported.