Surgical technique for congenital iris coloboma repair

Robert J. Cionni, MD, Ekaterini C. Karatza, MD, Robert H. Osher, MD, Manan Shah, MD

We describe a surgical technique for managing congenital iris coloboma. After phacoemulsification with placement of an intraocular lens in the capsular bag, coloboma repair is begun by bisecting the iris sphincter on both sides of its attachment near the chamber angle. The iris leaflets central to the sphincterectomies are approximated using a 10-0 polypropylene suture (Prolene, Ethicon, Inc.) and a modified Siepser pupilloplasty technique. The remaining peripheral iris defect is closed in a similar fashion. In patients with congenital iris coloboma, phacoemulsification with in-the-bag IOL implantation followed by this pupilloplasty technique was effective in providing functional and cosmetic repair of a congenital iris coloboma.


The introduction of the capsular tension ring (CTR) and its modifications have been helpful in managing eyes with zonular and/or lenticular coloboma. However, management of the distorted and eccentric colobomatous pupil is problematic. Leaving the pupil eccentric may result in dysphotopsia and monocular diplopia induced by the optic edge of the intraocular lens (IOL). Several surgical techniques to construct a more central and cosmetic pupil in these eyes have been developed. None results in a normal appearing or normally functioning pupil. We describe a new technique that can result in a centered, round, and normally functioning pupil.

SURGICAL TECHNIQUE

A 41-year-old white woman was referred to the Cincinnati Eye Institute with a history of poor vision caused by cataract and congenital iris coloboma bilaterally. The best corrected visual acuity was hand motion in the right eye and 20/50 in the left eye. The intraocular pressure was 14 mm Hg in both eyes. Examination revealed iris, lenticular, zonular, and retinal colobomas in both eyes (Figure 1). The coloboma in the right eye extended into the optic nerve head and macula, making visual prognosis poor. In the left eye, the coloboma extended to the temporal arcade inferiorly but did not involve the macula.

A brunescent nuclear sclerotic cataract was present in the right eye, and a +3 fetal nuclear sclerotic cataract in the left eye provoked visual complaints from the patient. The patient consented to have surgery with possible CTR implantation and pupil reconstruction in the left eye. Phacoemulsification was completed uneventfully through a superotemporal near-clear corneal incision, and a CTR (Alcon model 12) was placed through the main incision into the capsular bag. A single-piece acrylic posterior chamber IOL (Alcon model SN60WF) was injected into the capsular bag so the optic was well centered. The ophthalmic viscosurgical device (OVD) was removed from behind the IOL but left anterior to it. Acetylcholine chloride was instilled intracameraly in preparation for iris repair.

Using a Vannas scissors, 2 small incisions were made through the inferior iris sphincter, 1 nasally and 1 temporally, approximately 2.0 mm from the chamber angle (Figure 2). Two 1.0 mm peripheral corneal stab incisions were made on either side of the inferior coloboma. The pupil was reconstructed using a CIF-4 needle (Ethicon) on a 10-0 polypropylene suture (Prolene), which entered the anterior chamber through 1 of the 2 paracentesis sites. The needle was passed through the edges of the central iris leaflets, exiting through the distal peripheral corneal stab incision (Figure 3). The suture was securely tied with a modified...
Siepser sliding knot technique. Iris tissue peripheral to the reconstructed pupil was closed with several additional modified Siepser-style suture passes. The result was a centered, round pupil (Figure 4).

The OVD was removed using the automated irrigation/aspiration tip, and the incisions were hydrated and checked for stability by scleral pressure at the posterior lip of the incision. Topical povidone-iodine 5%, moxifloxacin 0.5%, and brimonidine tartrate 0.15% were instilled. The patient was examined the next day, revealing a centered and functional pupil with an uncorrected visual acuity (UCVA) of 20/20/20.

DISCUSSION

The word coloboma is Greek—koloboma, meaning mutilated or curtailed—and refers to a congenital malformation that occurs in about 1:10 000 births in which ocular structures are incompletely formed due to failure of the embryonic optic fissure to fuse. The resultant gap, notch, or hole may involve several different structures, including the eyelid, iris, lens, ciliary body, choroid, optic nerve, or retina, causing mild to severe vision loss. A complete iris coloboma involves the pigment epithelium and stroma, giving rise to the so-called “keyhole” pupil, which can be unilateral or bilateral. A partial coloboma involves the pupillary margin only, making the pupil oval. Occasionally, the coloboma affects only the iris pigment epithelium and can be seen only on transillumination. Although iris coloboma often occurs as a sporadic, isolated defect, when in the presence of microphthalmia, it is commonly associated with severe neurological or craniofacial anomalies or other multisystemic developmental defects, such as...
the cat-eye syndrome, aniridia-Wilms tumor, trisomy 13 syndrome, and CHARGE syndrome.

Iris coloboma is frequently associated with clinically significant cataracts that develop at a young age.\(^3\) Globular opacities in the anterior subcapsular region usually form inferiorly, underlying the iris coloboma with superimposed senile cataractous changes.

Few reports of cataract surgery in eyes with congenital coloboma of the iris have appeared in the literature, but most emphasize that the condition often increases the rate of surgical complications due to the compromised zonular integrity.\(^3\)–\(^10\) In 1961, Jesberg and Schepens\(^4\) reported surgery in 10 eyes—7 with retinal detachment, 2 with cataract, and 1 with glaucoma—with poor surgical outcomes (4 developed phthisis bulbi). No details of the cataract surgery were given. In 1968, Nixeaman\(^5\) was the first to document intracapsular cataract extraction (ICCE) through an iris coloboma. Vaughn and Schepens\(^6\) later reported an uneventful ICCE and IOL placement with good visual results. In 1987, Jaffe and Clayman\(^7\) reported 6 cases; 5 had phacoemulsification and in-the-bag implantation of a posterior chamber IOL with no intraoperative complications and improved postoperative vision, and 3 of the 6 eyes had a visual acuity of 20/20. Similarly, Watt\(^8\) described a similar inferior suture closure of the coloboma combined with a superior sector iridectomy to effectively move the pupil superiorly. This effectively eliminates symptoms from the inferior optic edge, while the superior optic edge is typically covered by the upper lid. Both techniques decrease the risk for dysphotopsia but leave the pupil irregular and without normal functioning.

The sphincterotomy–pupilloplasty technique we describe addresses the etiology of pupil eccentricity. By severing the iris sphincter inferiorly, the pupil’s attachment to the iris base and chamber angle is released. Following reappraisal, the pupil constricts in a normal circumferential manner and no longer moves inferiorly during constriction. In the few cases we have performed, the remaining inferior peripheral iris defect has been suture-approximated with several Siepser-style suture passes. Alternatively, the defect can be obscured by placing a sector aniridia device such as the Morcher model 96G. Although we have performed this technique in only a few patients, the results have been effective and reproducible.

**Figure 5.** Simple closure of the inferior coloboma leaves the pupil small and eccentric.

**REFERENCES**


