

Surgical technique for management of isolated lenticular coloboma with high corneal astigmatism

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We describe a surgical technique for the correction of isolated congenital lenticular coloboma associated with high corneal astigmatism. Transscleral fixation of the capsular bag with a single eyelet Cionni capsular tension ring was followed by in-the-bag implantation of a toric intraocular lens (IOL). This led to complete correction of the lenticular defect and perfect alignment of the toric lens. In this case, the child attained an unaided distance visual acuity of 20/30 following amblyopia therapy and a well-aligned toric IOL at 12 months of follow-up. This technique can be used in cases with concomitant lenticular coloboma and significant corneal astigmatism.

Key words: Astigmatism, capsular tension ring, cionni ring, coloboma, toric intraocular lens

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Lens coloboma is a congenital anomaly of the capsular bag due to abnormalities of the zonules and ciliary body. It is not a true coloboma because there is no actual loss of lens substance, but a secondary manifestation of congenital absence of zonules in a quadrant of the eye.^[1,2] Severe anisometropia and amblyopia can be commonly found in unilateral cases because of the disfigurement of the lens, although lens coloboma may be bilateral or unilateral.^[3] Irregular astigmatism due to the abnormal lens margin can itself cause significant visual compromise, but when associated with significant corneal astigmatism too, it can be a management challenge. We describe the surgical technique for the management of such cases with correction of the colobomatous defect along with toric intraocular lens (IOL) implantation.

Surgical Technique

A 9-year-old child with unilateral congenital lenticular coloboma and corneal astigmatism of more than 2.0 diopter was planned for surgical correction with toric IOL implantation under general anesthesia. In the preoperative area, the 0° and 180° axis of the right eye were marked under topical anesthesia using a bubble toric marker (Nuijts-Lane Preoperative Toric Reference Marker, ASICO, IL, USA) with the child in sitting position. After induction of general anesthesia, the right eye was prepared and draped using aseptic technique.

In the case described below, the lenticular coloboma extended from 6 to 11 o'clock position. A fornix-based

conjunctival flap was raised at 8:30 position and the bleeding points were cauterized. A triangular partial thickness scleral flap was raised under the conjunctival flap in the quadrant of the coloboma using a crescent knife. The main port incision was made at its predetermined location of 110° guided by the Callisto Eye (Carl Zeiss Meditec, Germany). After forming the anterior chamber with viscoelastic, an oval continuous curvilinear capsulorhexis was completed using a utratras forceps [Fig. 1a]. To stabilize the capsular bag, three capsular hooks were applied to the rhexis margin in the quadrant of the coloboma followed by a gentle multiquadrant cortical cleaving hydrodissection [Fig. 1b]. The cortical material was aspirated using bimanual irrigation and aspiration (I/A). A 10-0 polypropylene suture was passed under the scleral flap 1.5 mm posterior to the surgical limbus to enter into the anterior chamber. Care was taken to not inadvertently rupture the capsular bag with the suture needle, and viscoelastic was used to tamponade the anterior capsule before passing the needle. The needle was guided out by docking it in a 26G needle placed through a previously made limbal paracentesis at 2:30 position [Fig. 1c]. This end of the suture was tied securely to the eyelet of the fixation hook of a Cionni capsular tension ring (CTR) which was then dialed into the capsular bag through the paracentesis at 2:30 position using a Sinskey hook.

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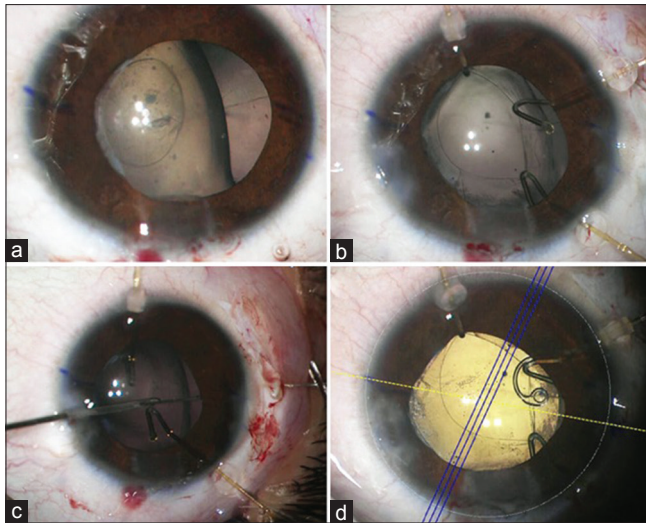


Figure 1: Surgical technique. (a) Surgeons view showing the lens coloboma with a completed continuous curvilinear capsulorhexis. (b) Capsular hooks supporting the capsular bag after aspiration of the cortical material. (c) Needle on a 10–0 polypropylene suture passed under the scleral flap being guided out by docking in a 26-gauge needle. (d) Toric intraocular lens implanted in the capsular bag and perfectly aligned with the virtual guidance marks of Callisto eye after viscoelastic removal

A toric IOL (AcrySof IQ model SN6AT5, Alcon, Fort Worth, Texas, USA) was implanted into the capsular bag and dialed a few degrees short of the target axis. Viscoelastic was removed with bimanual I/A, and the IOL rotated into its predetermined axis using virtual guidance marks of the Callisto Eye [Fig. 1d]. The capsular hooks were then safely removed to obtain a circular rhexis from the initial intentional oval design due to correction of the coloboma by the Cionni ring. The Cionni ring was secured to the sclera with the 10–0 polypropylene suture under the fashioned partial thickness scleral flap followed by standard steps of closure of the scleral and conjunctival flaps. The limbal paracentesis was hydrated with balanced salt solution, and the alignment of the toric IOL rechecked at the end of the procedure [Video 1].

Case Report

A 9-year-old boy was brought by his parents with complaints of decrease of vision in the right eye since birth. The patient gave no history of any previous ocular trauma or surgery. Initial examination revealed a retinoscopy of $-4.5\text{DS}/-2.25\text{DC}$ @ 110° with a best-corrected visual acuity of counting fingers at 3 M in the right eye and unaided visual acuity of 20/20 in the left eye. Intraocular pressures were 16 mmHg OU by Goldmann applanation tonometry. Slit lamp examination under mydriasis demonstrated a clear lens with a straight temporal margin indicative of a coloboma extending from 6 to 11 o'clock position. There was also the presence of posterior synchia nasally from 2 to 4 o'clock with a quiet anterior chamber [Fig. 2a]. Fundoscopy showed a normal fundus with no coloboma of the retina, choroid or the optic nerve. The optical biometry with the IOL Master (Carl Zeiss Meditec, Germany) showed an axial length of 23.85 mm in the right eye with keratometry readings of 40.86 D/43.27 D \times 82°. The SRK-T formula predicted an IOL power of 22.5

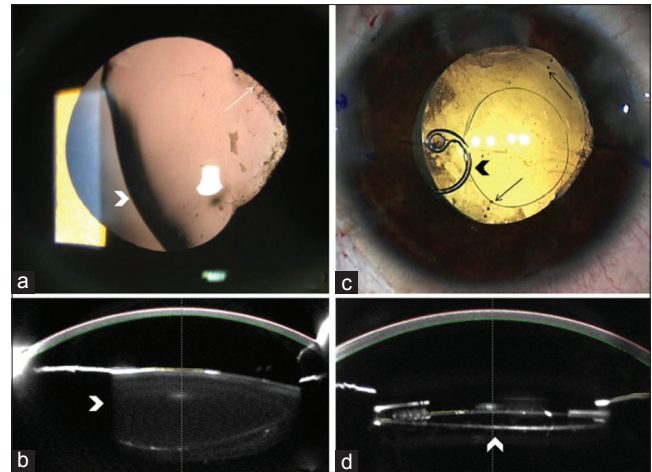


Figure 2: Pre- and post-operative comparison. (a) 9-year-old male child with right eye lenticular coloboma extending from 6 to 11 o'clock and posterior synechiae from 2 to 4 o'clock. (b) Preoperative Scheimpflug imaging depicting the abrupt ending of the lenticular margin. (c) Same patient after phacoaspiration with transscleral fixation of capsular bag with Cionni ring and in-the-bag implantation of toric intraocular lens. (d) Postoperative Scheimpflug imaging demonstrating a well-centered intraocular lens

diopter to obtain emmetropia. Scheimpflug images obtained with the Pentacam HR (Oculus, Germany) demonstrated the abrupt ending of the lenticular margin [Fig. 2b]. Given the markedly reduced vision due to the lenticular coloboma along with the high corneal astigmatism of 2.41 D, the patient was planned for surgical coloboma correction with toric IOL implantation under general anesthesia as per the technique described above.

Postoperative course

The child attained an uncorrected distance visual acuity of 20/60 at 1 week postoperative visit with no residual refractive error for distance on retinoscopy and subjective refraction. The IOL was well centered in the capsular bag [Fig. 2c]. Amblyopia therapy was initiated for the stimulus deprivation amblyopia, and at the last follow-up 1 year later, the child had improved to 20/30 in the right eye with good compliance to patching. The IOL maintained its alignment and centration throughout the follow-up period. Postoperative scheimpflug imaging revealed the reformation of the contour of the capsular bag and the well centred IOL [Fig. 2d].

Discussion

We describe a case of lenticular coloboma with significant corneal astigmatism managed successfully with modified CTR and in the bag toric IOL implantation. CTR has for long been used to facilitate cataract surgery in patients with profound zonular weakness that may be inherent in conditions such as Weil Marchesani syndrome, Marfan's syndrome as well as secondary causes such as trauma.^[4] CTR can be very useful in the surgical management of lenticular coloboma because there is a zonular loss to be compensated for and there is no actual loss of lens tissue. Mizuno *et al.*^[5] first described the use of CTR for the surgical management of lenticular coloboma where they successfully used the device in one eye of a patient with bilateral coloboma of the lens.

However, they did not recommend a routine use of a CTR in lenticular coloboma.

Toric IOLs have been shown to be safe and effective in children over the age of 8 years.^[6] When planning the use of a toric IOL in these patients, the decision regarding CTR use can be a tough one. An implanted CTR can rotate in the capsular bag after surgery leading to the loss of capsular bag symmetry with subsequent decentration and rotation of the toric IOL. It has been estimated that approximately 1° of off-axis IOL rotation results in a loss of up to 3.3% of IOL cylinder power. Complete loss of cylinder power occurs when the IOL is misaligned by more than 30°.^[7] Cionni and Osher^[8] designed a modification of the conventional CTR, also known as the Cionni ring. This modified CTR has 1 or 2 fixation hooks with eyelets that permit suture fixation of the CTR to the sclera. Implantation of a Cionni ring into the capsular bag to correct the coloboma along with transscleral suture fixation of its eyelet can ensure adequate centration and also prevent subsequent rotation of the CTR postoperatively. Gurler *et al.*^[3] first described the use of Cionni ring in patients with congenital lenticular coloboma. They advocated the use of a conventional CTR in cases of lens coloboma of <120° and a Cionni CTR in cases of lens coloboma of more than 120°. However, a toric IOL was not used in any of the eyes in their series. We recommend the use of a Cionni ring in all cases of lens coloboma to be taken up for toric IOL implantation to ensure adequate centration and stability which are required to achieve optimum results with these IOL.

One potential long-term complication of our technique can be the rupture of the scleral suture leading to decentration of the CTR-IOL-capsular bag complex. Cionni *et al.*^[4] in their series of 90 eyes noted a 10% rate of suture breakage (10–0 polypropylene) and recommended the use of 9–0 polypropylene for Cionni ring fixation. However, the risk of suture breakage continues to exist even with the use 9-0 polypropylene. Gimbel and Amritanand^[9] have described a technique for tripod fixation of the Cionni ring-IOL-capsular bag complex in the event of this unfortunate complication. Posterior capsular opacification requiring capsulotomy can be another cause of rotation of the toric IOL. Both these complications warrant a watchful and long-term follow-up in such patients.

Conclusion

We describe a technique for surgical management of congenital lenticular coloboma with high corneal astigmatism using a single procedure resulting in complete refractive correction. With adequate amblyopia therapy following this procedure,

gratifying visual outcomes can be achieved in such young patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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