We describe a technique for image-guided femtosecond laser–assisted cataract surgery in a case of Peters anomaly type 2. The femtosecond laser technology enabled reliable construction of a complete anterior capsulotomy despite central corneal opacification and a tented anterior capsule. Use of an image-guided femtosecond laser for cataract extraction in Peters anomaly type 2 is a safe method for removing these complex cataracts while preserving capsular and corneal integrity.

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Peters anomaly is a rare congenital ocular disorder characterized by central corneal opacification with absent endothelium, Descemet’s membrane (DM), and posterior stroma. This ocular malformation is believed to be due to abnormal migration of neural crest cells toward the posterior cornea during embryogenesis. Type 1 Peters anomaly is characterized by corneal leukoma with iridocorneal adhesions. Type 2 is characterized by corneal leukoma with cataract and corneolenticular adhesions.

In patients with Peters anomaly type 2, visually significant cataracts can be present at birth or develop early in life. Poor visualization through the cornea, tenting of the anterior capsule, and traction from corneolenticular adhesions on adjacent DM can make manual capsulorhexis construction and cataract surgery challenging. As a result, cataract extraction without concurrent penetrating keratoplasty (PKP) has rarely been reported in Peters anomaly patients. However, the prognosis for PKP in these patients is extremely poor. In a review of the literature by Bhandari et al., PKP in Peters anomaly type 2 failed in 85.8% of reported cases. Given these poor outcomes, cataract surgery alone is an important option when vision loss is out of proportion to the degree of corneal opacification in Peters anomaly patients.

With femtosecond laser technology and intraoperative image guidance, options for overcoming major challenges in otherwise difficult cataract cases are now available. We present the use of image-guided femtosecond laser technology for performing cataract surgery alone in Peters anomaly type 2.

SURGICAL TECHNIQUE

Preoperative assessment of pupil dilation (4.0 mm or greater) and presence of iridocorneal or iridolenticular adhesions is important in determining whether a patient with Peters anomaly is a candidate for femtosecond laser–assisted cataract extraction. Iridocorneal or iridolenticular adhesions that encroach on the central 4.0 mm of the visual axis may interrupt the laser capsulotomy and may necessitate decentration of the capsulotomy or exclusion of the patient. Preoperative anterior segment optical coherence tomography (AS-OCT) can identify areas of potential traction on adjacent DM from corneolenticular adhesions and determine whether the anterior chamber depth (ACD) is adequate for the laser to cut the anterior capsule without damaging the overlying cornea (Figure 1).

In almost all cases, general anesthesia will be required given the young age of most of these patients. Coordination with the anesthesia team is important to ensure the patient can be safely kept under general anesthesia with appropriate monitoring during administration of the laser, transportation from the laser room to the operating room, and
performance of the manual portion of the surgery. To our knowledge, no hazard risk is associated with having the laser activated while the patient is under general anesthesia.

After administration of general anesthesia, the surgical eye is docked to the femtosecond laser. A lid speculum is used to open the lids. Forceps are used to center the globe and minimize ocular tilt while the patient interface is docked. During docking, air bubbles between the cornea and the contact interface should be minimized since they may obstruct laser delivery (Figure 2). The smaller, steeper corneas seen in young children may require slight manipulation of the eye with forceps or slight movement of the head while the eye is docked under suction to burp out retained bubbles. Shadowing in the path of the laser on the intraoperative AS-OCT overlay may be due to bubbles and may warrant redocking (Figure 3).

Adjustments should be made to the capsulotomy size and centration based on the surgeon’s view of the eye provided by the laser platform (Figure 2). Care should be taken to avoid any iridolenticular adhesions. The diameter of the capsulotomy should be large enough to ensure that the central corneal opacity does not obscure the path of the laser. Widening the diameter of the capsulotomy or decentering the capsulotomy may be necessary to create adequate ACD between the corneal endothelium and the distorted anterior lens capsule (Figure 3).

Using the AS-OCT imaging data provided by the femtosecond laser platform, the height of the anterior capsulotomy should be adjusted to encompass the anterior capsule surface within the anterior and posterior limits of the laser cut for the entire 360-degree circumference of the capsulotomy (Figure 3). Care should be taken to ensure that the capsulotomy laser cut does not hit the cornea. Since the distorted anterior capsule may have multiple peaks and troughs along the circumference of the intended capsulotomy, aligning the anterior and posterior limits of the laser at a single peak or trough, respectively, (the method typically used to compensate for simple lens tilt) may result in an incomplete capsulotomy. Intraoperative AS-OCT providing a scrolled scan of the circumference of the capsulotomy is crucial for visually confirming that the range of the laser cut includes the anterior capsule for the entire 360-degree circumference of the capsulotomy (Video 1, available at: http://jcrsjournal.org). After capsulotomy adjustments have been made, the laser can be delivered per routine.

In the operating room, two paracentesis incisions are made to facilitate cataract removal. Entering the anterior chamber with the paracentesis blade should be done carefully since the anterior capsule may be tented if a complete laser capsulotomy was not achieved. Trypan blue followed by injection of an ophthalmic viscosurgical device (OVD) to clear the dye can then be used to stain the margins of the capsulotomy. A complete capsulotomy should be visible with a notable gap between central anterior capsule tissue that is still adherent to the cornea and peripheral anterior capsule that has relaxed and flattened (Figure 4). Gentle manipulation of the adherent anterior capsule tissue with sweeping motions of the OVD cannula can identify the separation and confirm a
complete capsulotomy. With flattening of the anterior capsule, there is dilation of the capsulorhexis diameter. For this reason, it may be acceptable and even desirable to make a slightly undersized capsulotomy during the laser portion of the procedure.

After it is confirmed that a complete capsulotomy was performed, anterior vitrectomy should be performed to trim and debulk the adherent central anterior capsule and optimize the surgical view. In cases in which preoperative AS-OCT shows traction on DM, a high cut rate should be used to prevent further detachment of DM. The anterior vitrector and bimanual irrigation/aspiration can then be used to remove the lens nucleus and residual cortical material. An intraocular lens (IOL) can be placed into the capsular bag and a posterior capsulotomy created if indicated (Video 2, available at: http://jcrsjournal.org).

Case Report

A 6-year-old African American boy with bilateral Peters anomaly type 2 and speech delay was evaluated for cataract surgery in his left eye. The child had a long history of multiple surgeries in his right eye, including multiple failed grafts and keratoprostheses, which ultimately left him with only light perception visual acuity in that eye. His parents had noticed worsening vision in his better-seeing left eye over the past 14 months.

On examination, the uncorrected distance visual acuity in the left eye was counting fingers, down from a baseline of 20/100. Slitlamp and AS-OCT examinations of the left eye were notable for a central cornea scar approximately 4.0 mm in diameter with clear peripheral cornea. An underlying corneolenticular adhesion and a cloudy cataract were also noted (Figure 1). Peripherally, the anterior chamber was deep, with 1 clock hour of iridolenticular adhesion noted inferonasally. Posterior examination was limited but unremarkable.

The decrease in vision was attributed to cataract progression. Given the clear peripheral cornea and prior failure with corneal transplantation in the right eye, the decision was made to proceed with cataract surgery without corneal transplantation. Cataract extraction with IOL implantation was performed alone using the described femtosecond laser–assisted cataract surgery technique on a LenSx femtosecond platform (Alcon Surgical, Inc.).

At the 1-month follow-up, examination under anesthesia was notable for a central corneal scar with clear peripheral cornea and a well-centered posterior chamber IOL (Figure 5, A). At the final examination, 7 months after surgery, the corrected distance visual acuity was 20/200 at distance and 20/60 at near (with the child maneuvering his head and the near card to see around the residual central corneal opacity at near). Remnants of the adherent anterior capsule were noted on AS-OCT imaging (Figure 5, B).

DISCUSSION
Cataract surgery in Peters anomaly type 2 can be challenging due to the presence of corneal scarring, which obscures the view; and corneolenticular adhesions, which distort and place tension on the anterior capsule. Manual continuous curvilinear capsulotomy techniques that rely on a flat anterior capsule surface can be difficult to control when the capsule is tented up. A manual can-opener capsulotomy may be an alternative but carries the risk for radial capsule tears during hydrodissection, hydrolinelineation, and lens removal. Manual manipulation of the anterior capsule in the presence of corneolenticular adhesions may also lead to traction and detachment of adjacent DM. Because of these challenges, cataract surgery is rarely performed without concurrent penetrating keratoplasty in cases of Peters anomaly type 2.

Our experience suggests the femtosecond laser has the potential to perform a well-centered complete capsulotomy in cases of Peters anomaly type 2 in which the peripheral cornea remains clear. Care must be taken when adjusting the height and depth of the femtosecond laser cut to ensure a complete capsulotomy and avoid damaging the endothelium. Anterior segment OCT can accurately determine the appropriate position and tilt of the anterior capsulotomy despite the distorted anatomy. In cases in which the anterior chamber is shallow, an increase in the diameter of the capsulotomy may be necessary to move the incisions more peripherally, where the chamber is deeper. In cases with significant tenting of the anterior capsule, the final diameter of the anterior capsulotomy is likely to be larger than expected after the capsulotomy detaches from the cornea and lies flat on the lens surface. With minor intraoperative adjustments to the femtosecond laser based on the associated intraoperative AS-OCT, femtosecond laser–assisted cataract surgery appears to be safe and effective in cases of Peters anomaly type 2.

WHAT WAS KNOWN

• Femtosecond laser–assisted cataract surgery is an effective technique for performing routine cataract surgery in patients with clear corneas.
• Cataract surgery alone is rarely performed in patients with Peters anomaly type 2 despite the fact that graft failure following penetrating keratoplasty (PKP) surgery with cataract extraction is very high in these patients.

WHAT THIS PAPER ADDS

• Femtosecond laser–assisted cataract surgery is a useful technique for performing laser capsulotomy and cataract removal without PKP in patients with Peters anomaly type 2.
• Intraoperative ocular coherence tomography can provide accurate and reliable guidance for placement of the femtosecond laser–assisted anterior capsulotomy despite distorted anatomy in patients with Peters anomaly type 2.
REFERENCES


Figure 1. Preoperative AS-OCT (A and B) and slitlamp photograph (C) of the left eye of the patient with Peters anomaly type 2. A broad corneolenticular adhesion (A and B) was noted in parallel slices, with evidence of potential traction on adjacent Descemet’s membrane (A, arrow) noted on an inferior slice.

Figure 2. Intraoperative view of the left eye after docking on a femtosecond laser platform. A: Initial docking resulted in a large retained air bubble (white arrow) due to the small, steep cornea. B: After burping out the air by manipulating the eye under suction, only a small air bubble remains (yellow arrow). The capsulotomy is adjusted to avoid the central corneal scar, the inferonasal iridolenticular adhesion (asterisk), and residual air bubbles under the contact patient interface.
**Figure 3.** Intraoperative AS-OCT of the left eye showing (A) a scrolled cross-sectional display along the intended circumference of the capsulotomy and (B) a single axial cross-section along the axis of greatest lens tilt. **A:** The height and position of the capsulotomy is adjusted to ensure that the anterior (*white arrow*) and posterior limits (*gray arrow*) of the laser cut contain the anterior capsule for the full 360-degree circumference of the capsulotomy. Due to multiple peaks (*yellow arrowheads*) and troughs (*blue arrowheads*) in the distorted anterior lens capsule, alignment of the anterior and posterior limits of the laser cut on a single peak or trough can result in an incomplete capsulotomy. **B:** On axial cross-section, care should be taken to ensure the intended path of the laser (demarcated by the *purple lines*) does not hit the cornea. Since the anterior chamber in Peters anomaly type 2 is deeper peripherally, smaller capsulotomies (*yellow lines*) are more likely to hit the cornea. Air bubbles (*asterisk*) between the cornea and the patient interface can cause shadowing.

**Figure 4.** Intraoperative photograph of the left eye taken after trypan blue staining of the anterior capsular. Spontaneous separation and flattening of the peripheral capsule tissue from the central capsule tissue (which remains adherent to the cornea) after femtosecond laser capsulotomy results in a visible gap (*arrows*) and dilation of the initial capsulotomy diameter.

**Figure 5.** Postoperative photograph (A) and AS-OCT (B) showing a central corneal scar with adherent remnants of the anterior capsule.

**Video 1.** Key intraoperative laser parameter adjustments for successful completion of a femtosecond laser capsulotomy in Peters anomaly type 2.

**Video 2.** Surgical technique for cataract removal after femtosecond laser capsulotomy in Peters anomaly type 2.