

Pseudoexfoliation Syndrome and Cataract Surgery

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Keywords

- Pseudoexfoliation • Cataract surgery • Pupil management • Cortex removal
- Anterior capsule cell removal • Late subluxation • Capsule phimosis

Key points

- Pseudoexfoliation cataracts require astute attention to detail.
- Preoperative examination is necessary to determine pupil dilatation.
- Control of intraocular pressure is important before surgery but also postoperative pressures spikes are common so same-day pressure checks are done along with more frequent monitoring for steroid and or inflammatory pressure problems.
- With careful zonular phacoemulsification techniques, used with adjunctive devices, complications during surgery are minimized and outcomes are excellent.



Video content accompanies this article at
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INTRODUCTION

Pseudoexfoliation syndrome (PXE) is an age-related disorder. Patients are characterized by production of an extracellular matrix seen commonly in the anterior segment of the eye but also present in other systemic tissues, such as the heart, lung, and liver. The association of a variant of the LOXL1 gene is present in 99.0% of patients with PXE. However, other genetic or possible environmental factors may be involved, as well as solar exposure and geographic history [1–4].

PXE is associated with glaucoma and cataract [5]. From a cataract standpoint, PXE is also associated with small pupil dilatation, zonular weakness, fragile capsules, and narrow angles. Postoperative problems can include glaucoma,

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pseudophacodonesis, and late complete capsule bag and intraocular lens (IOL) subluxation and/or dislocation secondary to the weakened zonules [6,7].

The presence of PXE in cataract surgery is associated with an overall increase in surgical complications [8,9]. There are several technique modifications as well as new devices and adjunctive measures that can reduce the incidence of intraoperative complications. Although PXE cataract surgery may be uneventful, there remains the problem of late subluxation. Late subluxation generally occurs about 8.5 years following cataract surgery (whether the original surgery was complicated or routine) [10].

CATARACT SURGERY

This article presents methods to decrease intraoperative complications, as well as technique variations that may reduce late complications [9]. Although phacoemulsification can be done with a small pupil, this can lead to a series of consequences that may ultimately induce late complications [11–14].

The small pupil leads to a small capsulorhexis size, and complete cortical removal is more difficult. The small rhexis also makes nuclear rotation more difficult (Figs. 1 and 2). All of these can lead to capsule phimosis and weakened zonules, which may lead to spontaneous subluxation, especially in patients with PXE.

If the pupil dilates poorly then the use of additional intracameral preservative-free epinephrine along with Healon 5 (Allergan, Inc, Irvine, CA) may suffice. If this does not work then the use of an iris device is indicated; either iris hooks

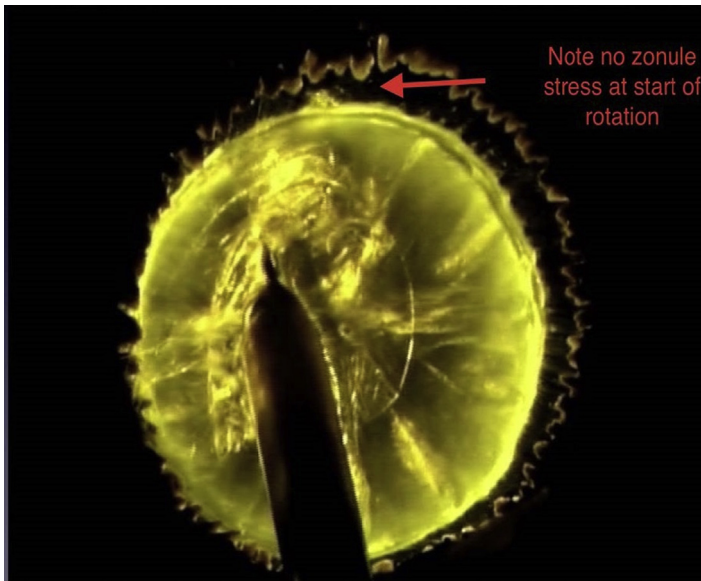


Fig. 1. At the beginning of the rotation there is no stress.



Fig. 2. As rotation continues there is significant stress on superior zonules, partially caused by small rhexis and inadequate hydrodissection.

(preferably in a diamond configuration) (Alcon, Inc, Fort Worth, TX; Microsurgical Technology [MST] Seattle WA) or a Malyugin ring (MST, Seattle, WA), which comes in 6.25-mm or 7.0-mm diameters [15,16]. I recommend a 5.5-mm to 5.75-mm diameter capsulorhexis to allow easy nuclear rotation and decrease the risk of capsule phimosis (Video 1).

Hydrodissection is critical to allowing nuclear rotation. Viscodissection may also add to reducing stress by creating and maintaining space. Multiple aliquots of BSS (Balanced salt solution) are slowly injected to achieve total cortical cleavage, using 3-mL or 5-mL syringes with a Luer-Lok (to prevent the cannula from releasing). If done correctly this separates the cortex from the capsule, allowing easy nuclear rotation [17,18].

Nuclear rotation should be as zonule friendly as possible. A bimanual technique reduces this stress by equalizing forces during the rotation (Figs. 3 and 4).

The nuclear disassembly techniques vary with the hardness of the nucleus. The most zonular friendly is the femtosecond laser, which is followed by chopping techniques (vertical or horizontal) but also includes prechop and UltraChopper. If a divide-and-conquer technique is used then grooving should be done slowly so as to not push or move the nucleus, and the nuclear rotations should be done carefully.

Divide and conquer

The nucleus is divided into 4 quadrants

In sculpting, the nucleus is grooved along 2 orthogonal axes, which creates a cruciform pattern. It is critical to understand that the maneuver is nonocclusive,

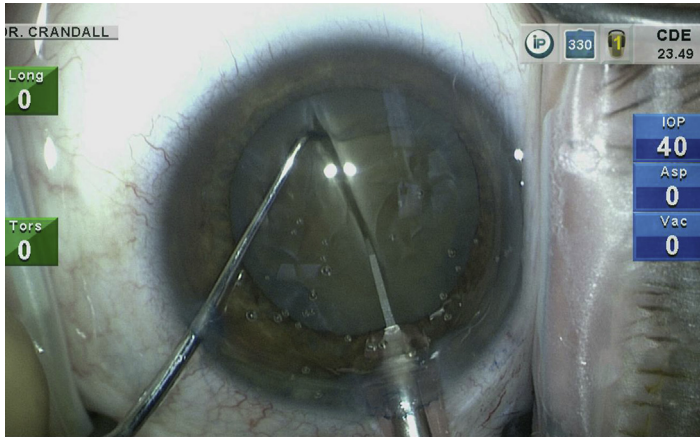


Fig. 3. Bimanual rotation: rotation started with equal rotation energy to reduce zonule stress.

which is important to reduce stress on the bag. The deeper the groove goes centrally the easier it is to divide the hemisections. Once the quadrants are free then the clinician changes to a quadrant removal mode that has increased aspiration flow rate and vacuum. Pieces are brought to a safe place in the center of the anterior chamber, just above the plane of the iris, and the power is then increased until the nucleus stays at the tip. Enough energy is used to keep the material moving into the tip. If an occlusion happens, the natural tendency is to reduce the power, but this increases occlusion, and when this is broken, surge can happen, causing a capsule tear [19].

Chop techniques allow clinicians to use less fluid during the surgery, because the time taken for a grooving maneuver is eliminated. With chop techniques it is imperative that the nucleus is impaled and held so the chopper can be used to



Fig. 4. Bimanual rotation forces are used to keep lens centered, rather than a push motion.

make the chop. It is easier to hold the nucleus with 0° or 15° tip. In a chop technique it is easier to impale the nucleus if the device is in a longitudinal and not torsional or elliptical mode. A burst mode facilitates the impaling. For a vertical chop the second instrument is placed near the tip, just in front of the phacoemulsification tip. The phacoemulsification tip is not moved, and the second instrument, moving toward the center of the nucleus, does the chopping. Once it is deep, then the chopper is moved away to separate the nucleus (Fig. 5). The nucleus is rotated and is impaled again and chopped. The clinician can repeat until 4 to 8 pieces are chopped. In a horizontal chop the nucleus is impaled, but now the chopper is placed under the anterior capsulorhexis, drawn toward the phacoemulsification tip, and then separated 90° , thereby splitting the nucleus [19] (Fig. 6, Video 2).

The UltraChopper is a surgical tool first envisioned and designed by Luis Escafe from South America. He initially used a vice to flatten a standard Kelman phacoemulsification tip to act as a cutter. The idea was presented and the patent sold to Alcon Laboratories. Although it seems initially to be dangerous it is the same size as a Kelman tip (Figs. 7 and 8). It cuts through a hard lens to allow the clinician to groove and then use a prechop instrument to divide the nucleus into quadrants or more pieces (Figs. 9 and 10, Video 3). At present, it only fits on Alcon handpieces.

Cortex removal should be as complete as possible, as well as zonular friendly. Using a Miyake-Apple technique, the authors showed that the standard radial stripping causes more zonule stress than a tangential technique (Figs. 11 and 12, Videos 4 and 5).

Another maneuver to reduce bag and zonule movement is to inject viscoelastic to stabilize the bag before removing the phacoemulsification tip, and also to inject it again before removing the irrigation and aspiration tip (Figs. 13 and 14).

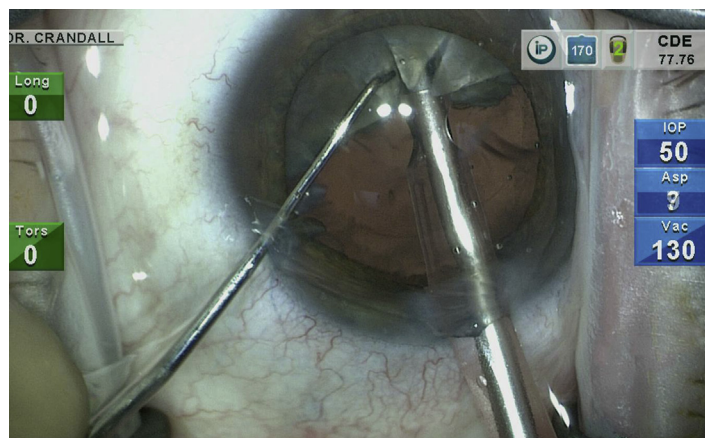


Fig. 5. Vertical chop-foot position 2. Chopper placed near tip; action is down then away from tip to split the nucleus.



Fig. 6. Horizontal chop. Nucleus impaled, foot position 2, to hold nucleus; chopper drawn toward tip then away to split the nucleus.

- Compatible with all INFINITI Handpieces
 - Optimized performance with OZil Torsional handpiece
- Allows for secure and easy division of dense nuclei
- Compatible with 0.9 mm family of Microsmooth sleeves



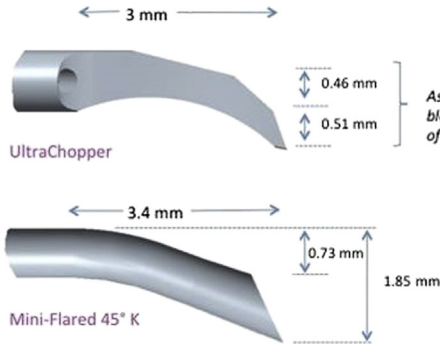



Fig. 7. UltraChopper.

Design Features:

- Distal end blade profile to enhance cutting
- Two cross-holes at distal end for fluidic control.



Assists in gauging depth of cut. 1 mm blade depth designed to reach 30%–50% of nuclear thickness.

Fig. 8. The tip on the UltraChopper is the same size as the Kelman tip.

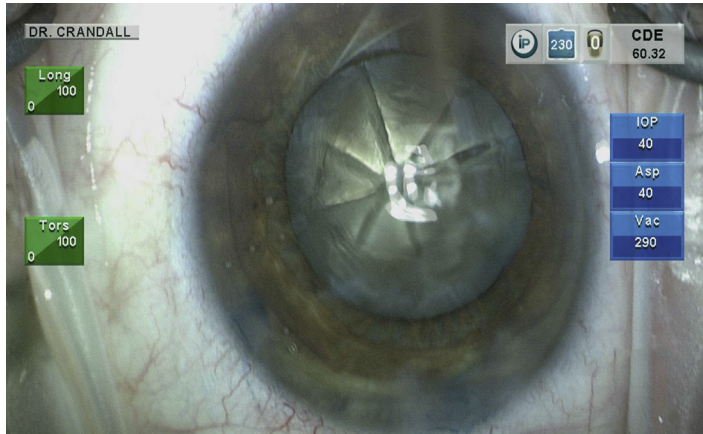


Fig. 9. Pie-shaped wedges following UltraChopper. Even though CDE is high, all energy is in the bag. Dispersive viscoelastic protects the endothelium (cell counts done preoperatively and postoperatively). CDE, cumulative delayed energy.

After cortex removal a Singer Sweep (AISCO, Epsilon) is used to remove as much of the anterior epithelial lens cells as possible (Figs. 15 and 16). Although it has not been shown to reduce posterior capsule opacification it does reduce the capsule phimosis that may reduce late bag complex subluxation [19–21].

The use of capsule tension rings can help stabilize the bag complex. The intraoperative use of capsule support devices (MST capsule hooks, Redmond, WA; MacKool capsule hooks, MSI, Phoenixville, PA; and the Capsular Tension Segment, Morcher, Germany) can provide temporary support until the nucleus and cortex removal is complete (Fig. 17). A decision then needs to

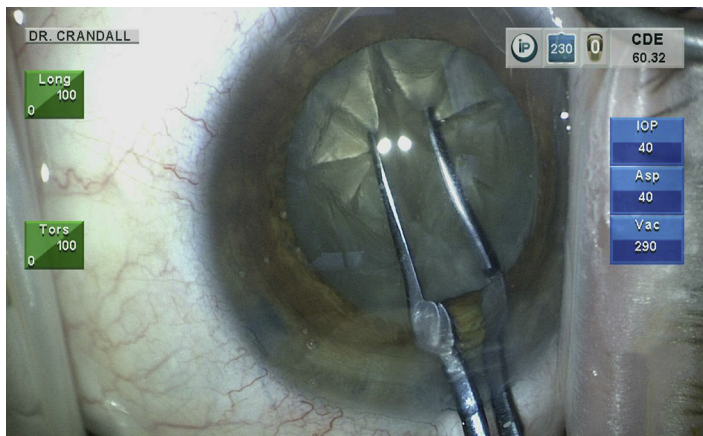


Fig. 10. An Akahoshi prechop (Aisco) or Crandall (Epsilon).

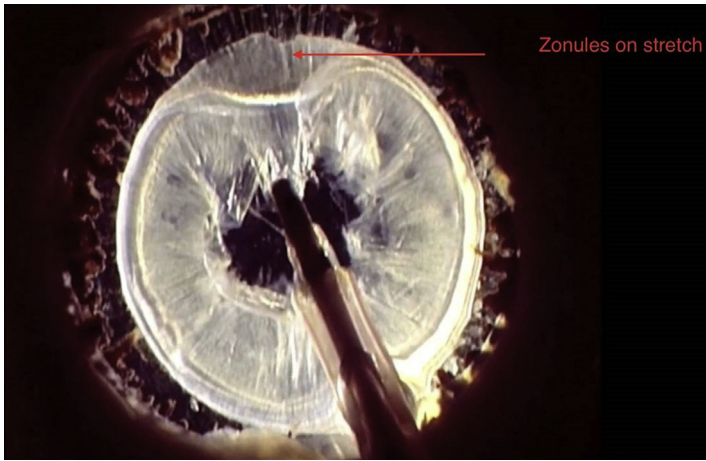


Fig. 11. Start of irrigation and aspiration (note the zonules under stress).

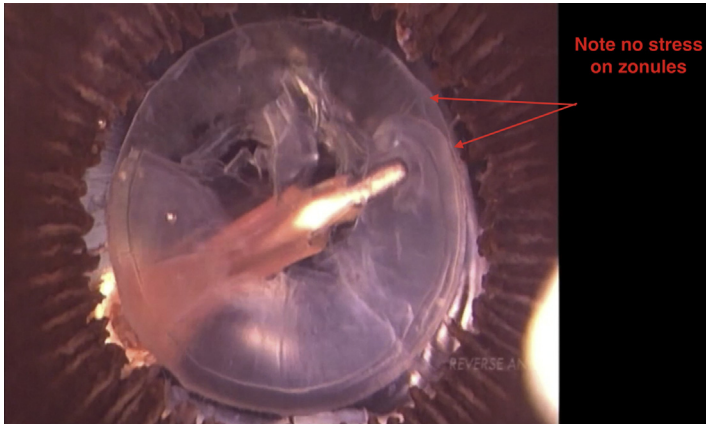


Fig. 12. Tangential stripping of cortex reduces zonule stress.

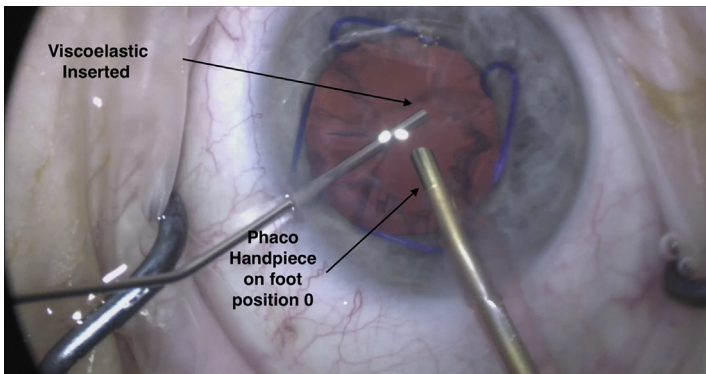


Fig. 13. Viscoelastic injection after phaco completion.

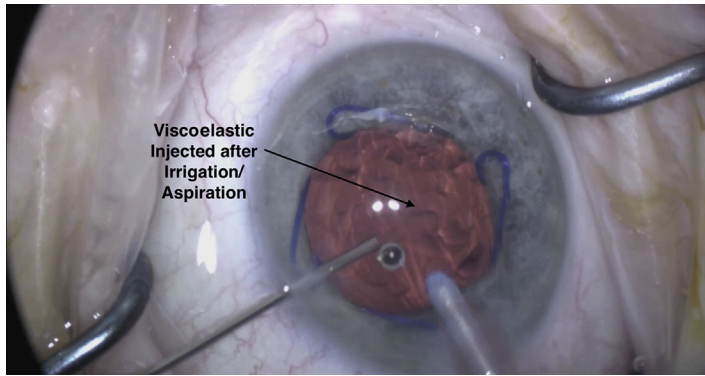


Fig. 14. Viscoelastic injection after irrigation aspiration completed.

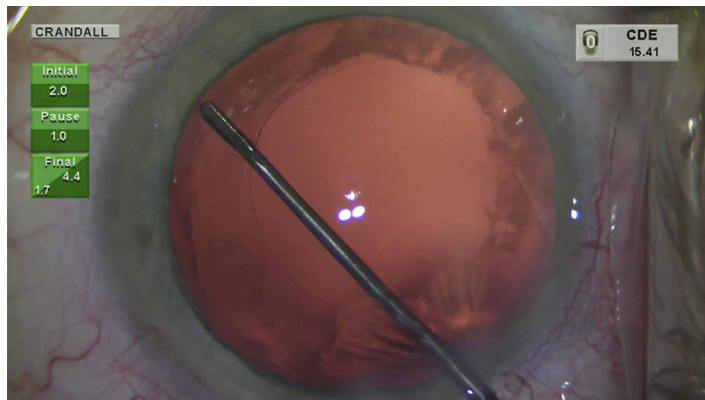


Fig. 15. Note anterior lens epithelial cells at start of sweep.

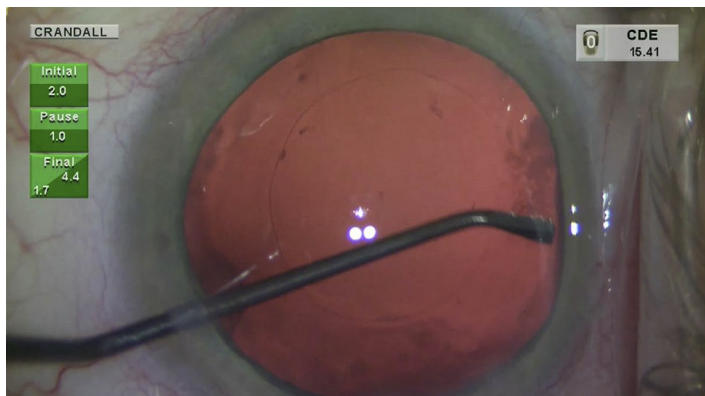


Fig. 16. Partial removal using a Singer Sweep.

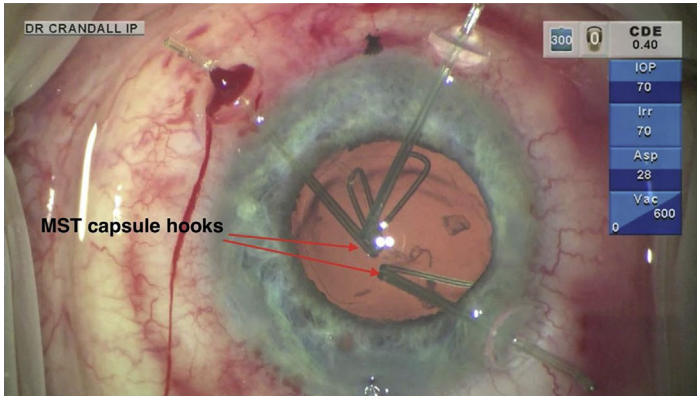


Fig. 17. MST capsule hooks supporting loose capsule.

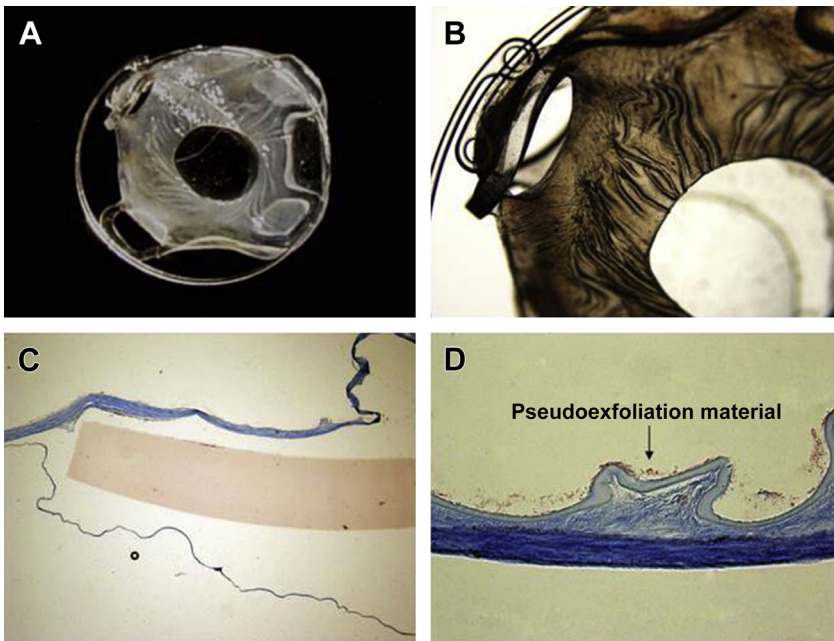


Fig. 18. Gross (A) and light microscopy (B–D) views of the specimen in case 2 showing fibrous metaplasia and contraction of the capsular bag with multiple folds, as well as anterior flexing of the closed loops. The sections cut through the specimen (C, D) show a thick fibrocellular tissue attached to the inner surface of the anterior capsule, the outline of the IOL, and a layer of amorphous substance on the outer surface of the anterior capsule (arrow) suggesting pseudoexfoliation (B: original magnification $\times 40$. C and D: Masson trichrome stain, original magnification $\times 40$ and $\times 200$, respectively). (From Zaugg B, Werner L, Neuhann T, et al. Clinicopathologic correlation of capsulorhexis phimosis with anterior flexing of single-piece hydrophilic acrylic intraocular lens haptics. *J Cataract Refract Surg* 2010;36:1608; with permission.)

be made as to the capsular tension ring (CTR) insertion versus a modified CTR (Cionni or Malyugin), iris fixated lens, or an anterior chamber IOL [22–24].

If a bag stabilizing device is in the capsule, the clinician may choose the single-piece or 3-piece IOL. However, some surgeons prefer a 3-piece IOL in order to allow for iris fixation later.

It has been shown that a CTR does not prevent all lens subluxations; however, it does make reposition by scleral fixation easier (Fig. 18). Several techniques have been developed to scleral fixate the bag [25,26]. I prefer to use a lasso maneuver [27] (Figs. 19 and 20).

The IOL selection in pseudoexfoliation syndrome

If a CTR is in place, or the surgeon is comfortable that there is no weakness of the zonules, then any IOL that the surgeon is comfortable with is fine. If there is a question, then some surgeons favor a 3-piece IOL with the optic in the bag and haptics in the sulcus, assuming that the haptics will become fibrosed in the sulcus region. There are no long-term results or randomized trials on the subject, but a 3-piece lens would be easy to iris fixate. Because the problem with PXE is progressive, the author recommends vigilance with frequent examinations (every 4–6 months); if pseudo-phacodonesis is seen, clinicians can intervene before complete subluxation occurs.

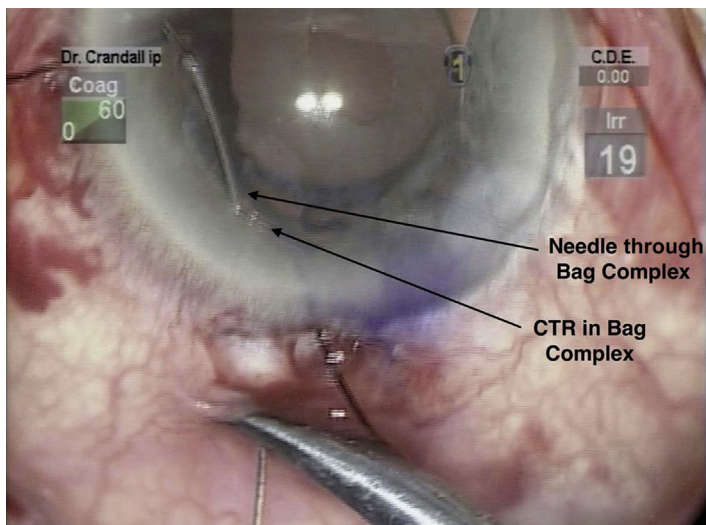


Fig. 19. A 9-0 suture on a CTS needle (Alcon) is passed under the iris 2 mm posterior to the surgical limbus, through the bag complex (in this case there is a CTR in the bag) and out through the peripheral cornea. CTS, capsular tension segment

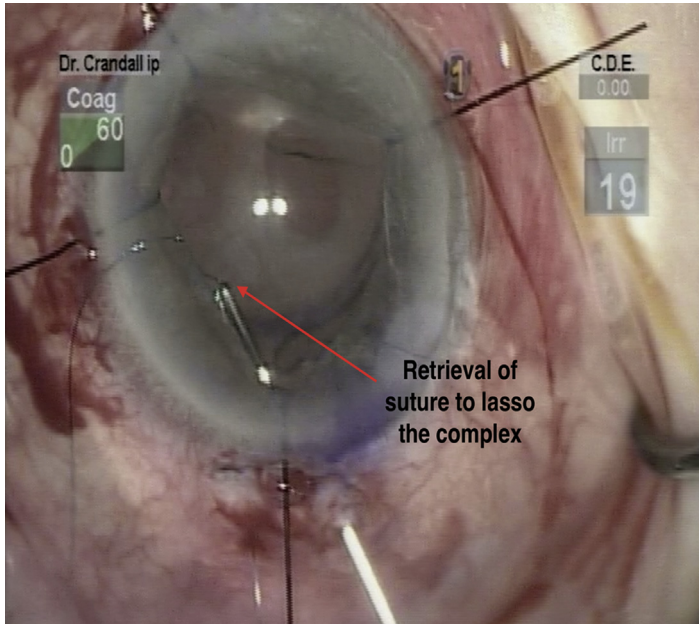


Fig. 20. A 23-gauge MVR is used to make another entrance and an MST Snare or 25-gauge ILM forceps is used to retrieve the suture to lasso the bag.

SUMMARY

Pseudoexfoliation cataracts require astute attention to detail. Preoperative examination is necessary to determine pupil dilatation. Gonioscopy (especially if the angle is narrow) looks for chamber asymmetry, which suggests zonular issues. Control of intraocular pressure is important before surgery but also pressure spikes postoperatively are common so same-day pressure checks are done along with more frequent monitoring for steroid and/or inflammatory pressure problems.

With careful zonular-friendly phacoemulsification techniques plus adjunctive devices, complications during surgery are minimized and outcomes are excellent.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.yaoo.2016.04.001>.

References

- [1] Allingham RR, Liu Y, Rhee DJ. The genetics of primary open-angle glaucoma: a review. *Exp Eye Res* 2009;88:837–44.
- [2] Stein JD, Pasquale LR, Talwar N, et al. Geographic and climatic factors associated with exfoliation syndrome. *Arch Ophthalmol* 2011;129(8):1053–60.
- [3] Pasquale LR, Jiwani AZ, Zehavi-Dorin T, et al. Solar exposure and residential geographic history in relation to exfoliation syndrome in the United States and Israel. *JAMA Ophthalmol* 2014;132(12):1439–45.

- [4] Ritch R. Exfoliation—the most common identifiable cause of open-angle glaucoma. *J Glaucoma* 1994;3:176–8.
- [5] Seland JH, Chylack LT Jr. Cataracts in the exfoliation syndrome (fibrillogluthia epitheliocapsularis). *Trans Ophthalmol Soc UK* 1982;102(pt 3):375–9.
- [6] Nauman GO, Schlotzer-Schrehardt U, Kuchle M. Pseudoexfoliation syndrome for the comprehensive ophthalmologist; intraocular and systemic manifestation. *Ophthalmology* 1998;105:951–68.
- [7] Skuta GL, Parrish RK II, Hodapp E, et al. Zonular dialysis during extracapsular cataract extraction in pseudoexfoliation syndrome. *Arch Ophthalmol* 1987;105:632–4.
- [8] Shastri L, Vasavada A. Phacoemulsification in Indian eyes with pseudoexfoliation syndrome. *J Cataract Refract Surg* 2001;27:1629–37.
- [9] Shingleton BJ, Crandall AS, Ahmed II. Pseudoexfoliation and the cataract surgeon: Preoperative, intraoperative, and postoperative issues related to intraocular pressure, cataract, and intraocular lenses. *J Cataract Refract Surg* 2009;35:1101–20.
- [10] Jehan FS, Mamalis N, Crandall AS. Spontaneous late dislocation of intraocular lens within the capsular bag in pseudoexfoliation patients. *Ophthalmology* 2001;108:1727–31.
- [11] Hayashi H, Hayashi K, Nakao F, et al. Anterior capsule contraction and intraocular lens dislocation in eyes with pseudoexfoliation and intraocular lens dislocation in eyes with pseudoexfoliation syndrome. *Br J Ophthalmol* 1998;82:1429–32.
- [12] Liu E, Cole S, Werner L, et al. Pathologic evidence of pseudoexfoliation in cases of in-the-bag intraocular lens subluxation or dislocation. *J Cataract Refract Surg* 2015;41:929–35.
- [13] Conway RM, Schlotzer-Schrehardt U, Kuchle M, et al. Pseudoexfoliation syndrome: pathological manifestations of relevance to intraocular surgery. *Clin Experiment Ophthalmol* 2004;32:199–210.
- [14] Vasavada A, Singh R. Phacoemulsification in eyes with a small pupil. *J Cataract Refract Surg* 2000;26:1210–8.
- [15] Dupps WJ Jr, Oetting TA. Diamond iris retractor configuration for small-pupil extracapsular or intracapsular cataract surgery. *J Cataract Refract Surg* 2004;30:2473–5.
- [16] Chang DF. Use of Malyugin pupil expansion device in intraocular floppy-iris syndrome: results in 30 consecutive cases. *J Cataract Refract Surg* 2008;34:835–41.
- [17] Vasavada V, Vasavada VA, Werner L, et al. Corticocapsular cleavage during phacoemulsification: viscodissection versus hydrodissection; Miyake-Apple view analysis. *J Cataract Refract Surg* 2008;34:1173–80.
- [18] Vasavada AR, Singh R, Apple DJ, et al. Effect of hydrodissection on intraoperative performance: randomized study. *J Cataract Refract Surg* 2002;28:1623–8.
- [19] Ichhpujani P, Spaeth GL, Yanoff M. Expert techniques in ophthalmic surgery. Technique for routine cataract surgery. JayPee Brothers Medical Publishers; 2015. p. 56–68.
- [20] Baile R, Sahasrabudhe M, Nadkarni S, et al. Effect of anterior capsular polishing on the rate of posterior capsule opacification: a retrospective analytical study. *Saudi J Ophthalmol* 2002;26:101.
- [21] Epstein RH, Liu ET, Werner L, et al. Capsulorhexis phimosis with anterior flexing of an accommodating IOL: case report and histopathological analysis. *J Cataract Refract Surg* 2014;40:148–52.
- [22] Bolz M, Menapace R, Findl O. Effect of anterior capsule polishing on the posterior capsule opacification—inhibiting properties of a sharp-edged, 3-piece, silicone intraocular lens: Three and 5-year results of a randomized trial. *J Cataract Refract Surg* 2006;32(9):1513–20.
- [23] Lee DH, Shin SC, Joo CK. Effect of a capsular tension ring on intraocular lens decentration and tilting after cataract surgery. *J Cataract Refract Surg* 2002;28:843–6.
- [24] Ahmed IK, Cionni RJ, Kranemann C, et al. Optimal timing of capsular tension ring implantation: Miyake-Apple video analysis. *J Cataract Refract Surg* 2005;31:1809–13.

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- [25] Cionni RJ, Osher RH. Management of profound Zonular dialysis or weakness with a new endocapsular ring designed for scleral fixation. *J Cataract Refract Surg* 1998;24: 1299–306.
 - [26] Chan CC, Crandall AS, Ahmed II. Ab externo scleral suture loop fixation for posterior chamber intraocular lens decentration: clinical results. *J Cataract Refract Surg* 2006;32: 121–8.
 - [27] Kirk TQ, Condon GP. Simplified ab externo scleral fixation for late in-the-bag intraocular lens dislocation. *J Cataract Refract Surg* 2012;38(10):1711–5.