Pseudoexfoliation Syndrome in Cataract Surgery

BY JENNIFER CALAFATI, MD, DIAMOND Y. TAM, MD, AND IKE K. AHMED, MD
EDITED BY INGRID U. SCOTT, MD, MPH, AND SHARON FEKRAT, MD

Pseudoexfoliation syndrome (PXF) is an age-related systemic microfibrillopathy that targets ocular tissues through the gradual deposition of fibrillary residue from the lens and iris pigment epithelium, mainly on the lens capsule, ciliary body, zonules, corneal endothelium and iris.

The diagnosis of PXF is aided by characteristic findings during the ophthalmic examination. Typically, white fibrillary residue on the anterior lens capsule and pupillary margin is observed, as are pupillary transillumination defects and pigmentation of the trabecular meshwork.

PXF is a risk factor for glaucoma (most often open-angle) and has been correlated with an increased incidence of cataract formation.

Implications for Cataract Surgery

In PXF, the deposition of extracellular fibers results in predictable alterations of tissues of the anterior segment, making cataract operations potentially challenging. Surgeons must be aware of numerous intraoperative and postoperative issues in managing the patient with PXF. (See "Surgical Considerations.")

Two pathological manifestations of PXF, zonular weakness and poor pupillary dilation, have been identified as the most significant risk factors for surgical complications. Zonular weakness can be attributed to the deposition of pseudoexfoliative material on the zonular fibers and ciliary processes, resulting in a proteolytic disintegration of the zonule that can lead to spontaneous fragmentation. Poor pupillary dilation is a result of infiltration of the iris stroma with excessive extracellular matrix, causing mechanical obstruction during mydriasis.

PXF can present some unique obstacles to successful phacoemulsification. Nonetheless, with appropriate preparation and use of adjunctive devices, phacoemulsification is still the preferred method of cataract extraction in this patient population.

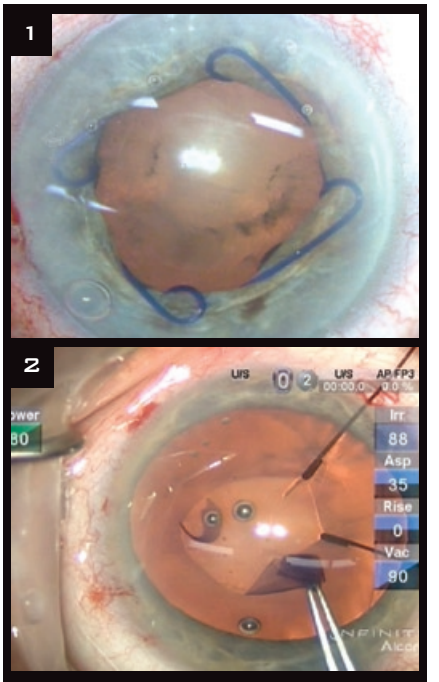
Preoperative Considerations

Before surgery, it's important to determine whether there is zonular weakness. Careful examination should be done to assess for subtle lens subluxation, zonular dialysis or iridophacodonesis. Other factors associated with zonular instability are age, shallow anterior chamber depth, cataract density, pupil size, IOP/glaucoma status, atrophy of pupillary ruff and pigment in the angle. Interestingly, the degree of PXF material visible in the eye does not seem to correlate with the degree of zonular weakness.

Anterior chamber depth is a useful predictor of intraoperative complications in eyes with PXF. An anterior chamber depth of less than 2.5 mm centrally—probably an indication of zonular instability—poses a significantly higher risk for intraoperative complications.

Zonular dialysis is typically quantified by the number of clock hours involved. If the patient has significant zonular instability, preoperative consideration of a polymethylmethacrylate capsule-supporting device (e.g., tension ring) is advisable.

Phacodonesis, a result of zonular instability, is graded on a +1 to +4 scale. When there is severe zonular instability, the surgeon may consider intracapsular extraction or pars plana lensectomy with an ACIOL or sutured



SURGERY IN EYES WITH PXF. (1) The Malyugin ring, which aids in expanding a small pupil and maintaining adequate pupil size through phacoemulsification and IOL implantation. (2) Iris hooks placed on capsulorhexis edge to aid in centration and countertraction during capsulorhexis.

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Surgical Considerations

Intraoperative	Postoperative
Dropped lens material	Acute IOP elevation
Higher risk of posterior capsular loss	Capsular contraction
Shallow anterior chamber	Corneal edema
Small pupil	Cystoid macular edema
Vitreous prolapse	Glaucoma
Zonular dialysis	IOL deposits
	IOL dislocation
	Posterior capsular opacification
	Posterior synechiae
	Prolonged anterior chamber reaction

PCIOL. Alternatively, one or more capsular tension devices can be used to safely perform endocapsular fixation of a PCIOL.

Intraoperative Considerations

A small pupil and zonular weakness must be addressed adequately during surgery to ensure surgical safety.

The small pupil. Preoperatively, NSAIDs may be given to discourage intraoperative miosis. Intraoperative measures are sometimes necessary, however. Posterior synechiae may bind down a small pupil and should be lysed with a capsulorhexis forceps prior to further maneuvers. The pupil can be enlarged using highly cohesive ophthalmic viscosurgical devices (OVD), pupil stretching devices or pupil expansion devices. Iris hooks and pupillary rings, for example, can ensure adequate dilation throughout the surgery (Fig. 1). Iris hooks also can serve to support the anterior capsulotomy if zonular weakness is significant. Care should be taken when handling the iris because injury to this tissue can occur easily.

Zonular weakness. Depending on the severity of the zonular dialysis, the surgeon may use a capsular tension ring (CTR), a Cionni modified-CTR (mCTR) or a capsular tension segment (CTS), alone or in combination.

When the degree of zonular weakness is mild, a CTR may be enough to support intraoperative maneuvers and to stabilize an IOL in the capsular bag. If obvious phacodonesis is present, it is advisable to insert a CTR early after completing the continuous curvilinear capsulorhexis to stabilize the capsular bag for phacoemulsification and cortical removal. For more advanced zonular disease, the mCTR or CTS, both of which can be sutured to the sclera for improved fixation, should be considered. Unlike the CTR, however, the CTS does not require a dialing technique during insertion, and therefore creates less stress on the zonules. Considering the long-term issues for these patients, some surgeons have advocated the routine use of a CTR in PXF cases, both as a surgical aid and for long-term IOL stability.[1,2]

In addition to the considerations above, the entire surgical process should be conducted with care.

Capsulorhexis. A sharp instrument should be used to begin the capsulorhexis, as zonular weakness results in less anterior capsular tension, which makes the initial puncture more difficult. If striae are seen during anterior capsule puncture, this may be the first clue that zonular weakness is present. In certain cases of moderate to severe zonular weakness, it may be difficult to tear the anterior capsule, and countertraction on the anterior capsule may be required. This countertraction can be achieved with an instrument such as a Kuglen hook or with iris hooks placed on the already torn edge of the anterior capsule (Fig. 2).

While capsulorhexis size is important in phacoemulsification in general, it is even more critical in an eye with zonular weakness. A capsulorhexis that is too small makes zonule-friendly phacoemulsification difficult to perform, while one that is too large may preclude the use of capsular tension devices, which require a sufficient anterior capsular edge.

Hydrodissection and viscodissection. Hydrodissection should be carried out with minimal stress on the zonule, especially if the surgeon attempts to rotate the nucleus in the capsular bag.

In the presence of zonular weakness, injection of an OVD between the lens capsule and the cortex may be useful. This lessens the likelihood of trapping cortex during CTR insertion and creates greater corticocapsular cleavage than hydrodissection alone.

Phacoemulsification. In phaco, a zonule-friendly horizontal or vertical chopping technique is recommended. It is also critical that the surgeon prevent the anterior chamber from collapsing, especially during insertion and removal of instruments from the eye. Vitreous prolapse may occur in the setting of weak or absent zonular fibers, and even intraoperative aqueous misdirection and positive posterior pressure may occur in these eyes. Keeping the chamber formed using balanced saline solution or an OVD is of the utmost importance.

Cortical removal. While nuclear and epinuclear removal often may be achieved in a zonule-friendly manner, cortical removal may be more difficult and traumatic. A tangential stripping method may be used in combination with gentle centripetal movements to allow cortical material to separate from the capsular bag. At this stage, it's important to remember that cortex may be trapped behind a capsular tension device. If this is the case, then to prevent further zonular tearing, a tangential stripping technique may be necessary. In some difficult situations, dry removal with a cannula may be required. Although these techniques may be time-consuming, they will result in the best chance of preserving zonular fibers.

IOL insertion. Although most foldable PCIOLs are reasonable options in these cases, we believe that the slowly unfolding one-piece acrylic IOL is the

best choice. This IOL design allows the surgeon to dial the IOL gently into the capsular bag and place the flexible haptics in the desired orientation with minimal capsular and zonular stress. The adjunctive use of a CTR provides excellent in-the-bag centration.

In the presence of zonular weakness, a sulcus-placed, three-piece foldable PCIOL is not recommended because of the likelihood of decentration.

Conclusion

PFX syndrome presents challenges that require careful preoperative planning and intraoperative care to ensure successful and safe surgery. The use of specialized adjunctive devices such as highly cohesive viscoelastics, pupillary expansion devices and capsular tension devices has increased the margin of safety in these potentially complex cataract surgeries. In addition, appropriate follow-up of patients after surgery is needed to assess endothelial cell function, to monitor for glaucoma and capsular phimosis and to examine for proper IOL position and centration. The risks associated with cataract surgery in the PXF patient can be minimized with the proper preoperative, intraoperative and postoperative care.

1 Bayraktar, S. et al. *J Cataract Refract Surg* 2001;27:1620–1628.
2 Hassanee, K. and I. Ahmed. *Ophthalmol Clin N Am* 2006;19(4):507–519.

Drs. Calafati and Tam are fellows, and Dr. Ahmed is assistant professor of ophthalmology and vision sciences at the University of Toronto. Dr. Ahmed is also clinical assistant professor of ophthalmology and visual sciences at the University of Utah, Salt Lake City.

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