Pseudoexfoliation (PXF) syndrome is an age-related disease that is thought to be a systemic disorder in which hyaline material of an unknown etiology accumulates in the ocular tissue. The ocular manifestations involve all of the anterior segment, including the conjunctiva and orbital structures. There is increasing evidence of an etiologic association between PXF and cataract formation. Glaucoma occurs more commonly in eyes with PXF, the most common identifiable cause of open-angle glaucoma. Approximately 20% of open-angle glaucoma cases are associated with PXF, and approximately 30% to 50% of patients with PXF develop glaucoma.

In cataract surgery, PXF is associated with higher rates of intra- and postoperative complications. Intraoperatively, posterior capsular tear, zonular dialysis, vitreous loss, and dislocation of lens matter occur more frequently in eyes with PXF than in eyes without the condition; postoperatively, complications such as intraocular pressure (IOP) spike, corneal edema, iritis, pigment dispersion, cystoid macular edema, posterior capsular opacification (PCO), anterior capsular fibrosis, and subluxation or dislocation of the IOL-capsular bag complex are more frequent in eyes with PXF than in the general population.

The implementation of modern phaco surgery has reduced complication rates significantly; however, complications still occur as a side effect of PXF syndrome. Hence, a meticulous preoperative work-up is needed to diagnose the condition at an early stage and allow the surgeon to formulate an appropriate surgical strategy to minimize complications and optimize outcomes. My surgical strategy is highlighted in the following video on Eyetube: http://eyetube.net/?v=mefom.

**PREOPERATIVE WORK-UP**

Preoperative work-up for cataract surgery should achieve the following: (1) identify the presence of risk factors for intraoperative complications; (2) identify the presence of associated ocular comorbidities; and (3) facilitate appropriate patient counseling. Below I describe each point in detail.

**Risk factors for intraoperative complications.** Nondilating pupils and zonular instability are significant risk factors for intra- and postoperative complications. I routinely perform mydriasis evaluation to ascertain whether a small pupil strategy will be required. A viscoadaptive ophthalmic viscosurgical device (OVD) such as Healon 5 (Abbott Medical Optics Inc., Santa Ana, California) can be used to attempt viscomydriasis; however, I prefer to use iris hooks for nondilating small pupils. Good results may also be obtained using other modalities such as the Malyugin Ring (MicroSurgical Technologies, Redmond, Washington), the Beehler Pupil Dilator (Ambler Surgical, Exton, Pennsylvania), the Morcher Pupil Dilator (Morcher GmbH, Stuttgart, Germany), and the Perfect Pupil Injectable (Milvella Ltd, Sydney, Australia).

I perform thorough slit-lamp biomicroscopy to detect subtle signs of early PXF and zonular weakness. Pupillary ruff defects or pigment dispersion caution me to look more carefully for the presence of PXF in a given case. In established cases, the condition is easy to diagnose: It appears as a powdery deposit in the anterior segment including on the iris, the pupillary margin, and the surface of the crystalline lens (Figures 1 and 2). Postoperatively, these powdery deposits may also be detected on the anterior capsule and the IOL surface (Figures 3 and 4). In frank cases, the pathog-
nomonic double bull appearance on the anterior capsule is difficult to miss.

It is important to remember that the extent of PXF deposits may not correlate with the degree of zonular instability. Minimal iridodonesis is a subtle sign of early zonular instability, and an asymmetrical anterior chamber depth is an important indicator of zonular weakness. I look for asymmetry between the patient’s two eyes and also among different quadrants of the same eye. Phacodonesis and frank subluxation (after pupillary dilatation) confirm the presence of zonular weakness. Mild tilt or a slightly eccentric central nucleus picked up in the slit beam may indicate the presence of subluxation in patients with a nondilating pupil.

Associated ocular comorbidities. The presence of comorbidities such as advanced nuclear sclerosis, reduced endothelial count, and open-angle glaucoma compounds the challenges faced by the cataract surgeon. I routinely perform gonioscopy in all patients with PXF to check the status of the anterior chamber angle, as the occurrence of intraoperative complications may require implantation of an anterior chamber IOL.

I advocate early surgery for these patients, most specifically before their nuclear sclerosis has reached an advanced stage. In my experience, the majority of PXF patients present with advanced brunescent or white mature cataracts, and if this is the case I perform B-scan ultrasonography to rule out significant posterior segment pathology.

Patient counseling. I advise patients to consider early surgery to avoid operating on cataracts with harder nuclei, thus maximizing outcomes. I also explain to patients that there is an increased incidence of intraoperative complications in these challenging eyes and that the postoperative course is more intense. Patients must understand that they are required to report for more frequent postoperative visits than is routine for standard cataracts, and they are told that PCO can induce an early drop in vision, necessitating Nd:YAG capsulotomy. I advise these patients to report any visual symptoms at onset so that subluxation of the IOL-capsular complex can be picked up early and treated, thereby hopefully avoiding a major intervention.

INTRAOPERATIVE TECHNIQUES

Anesthesia. I prefer topical anesthesia for routine cataract patients, but a complex PXF case with weak zonules, subluxation, small pupil, and advanced nuclear sclerosis merits surgery under peribulbar anesthesia.

Capsulorrhexis. I always use the soft-shell OVD technique for endothelial protection in these challenging situations. I routinely stain the anterior capsule with trypan blue (0.06%) dye in eyes with PXF. Because in PXF cataracts are often advanced and provide a poor red reflex, the dye is administered to enhance anterior capsular visibility and facilitate the capsulorrhexis. The PXF material and the central zonular attachments also stand out when stained with trypan blue.

Given that the zonules may be weak, the initial puncture of the capsule is made with a 26-gauge bent needle. In
patients with significantly lax zonules, radiating wrinkles appear on the anterior capsule at the time of capsular puncture. A central rhexis with a 5.5-mm diameter is attempted. A capsulorrhexis that is too small may complicate phacoemulsification by limiting access to the nucleus, whereas a rhexis that is too large may make subsequent use of capsular support devices more difficult. However, it is safest to make the rhexis diameter larger if the nuclear sclerosis is greater. A bimanual technique may have to be employed in cases with significant subluxation.

Hydrodissection. Properly performed cortical cleaving hydrodissection is crucial. I routinely perform multiquadrant hydrodissection because it efficiently mobilizes the nucleus and greatly helps minimize zonular stress while the nucleus is rotated during phacoemulsification. I exercise extreme caution in this step when emulsifying brunescent and white cataracts associated with PXF for fear of posterior capsular blow-out. The initial rotation is achieved asatraumatically as possible by employing a bimanual technique. Viscodissection with an appropriate OVD may help in capsular-cortical cleavage and facilitate insertion of a capsular tension ring (CTR), should it be required at an early stage of surgery.

Phacoemulsification. Current phaco techniques and newer-generation phaco technology using updated power modulation and better fluidics have resulted in considerable improvement of surgical results in eyes with PXF. A correctly performed chopping maneuver minimizes trauma to the zonules and is my technique of choice for nuclear disassembly.

The benefits of a larger rhexis are appreciated at this stage, enhancing access for nucleus manipulation and transmitting less stress to the capsulozonular apparatus. I avoid cracking techniques because these maneuvers may relax the zonules in the axis perpendicular to the cracking (provided there is no imbalance of the cracking forces) but stretch the zonules in the axis of chopping.

All maneuvers should be performed slowly in the center of the pupillary zone, using a low-flow, low-vacuum technique. Anterior chamber depth should be maintained at all stages, with no sudden shallowing or deepening.

Cortex removal. Another crucial step is cortex removal, during which care should be taken to avoid stressing the zonular apparatus. Tangential stripping and aspiration of the peripheral cortex minimizes zonular stress, and manual aspiration can be helpful in difficult cases. In some challenging cases with fragile or lax zonules, I have even waited to remove the cortex after IOL implantation. Properly performed cortical cleaving hydrodissection helps by leaving behind minimal cortex for subsequent removal.

Stabilizing the capsular bag. Capsule and iris retractors, CTRs, modified CTRs, and capsular tension segments (CTSs) are helpful adjuncts that come to the rescue when one is confronted with a weak zonular apparatus in these challenging situations. When used appropriately, such devices improve outcomes and avoid complications encountered with PXF cataracts.

The zonulopathy in PXF is diffuse and progressive. Hence, appropriate modification of surgical steps and utilization of adjunctive devices are necessary when dealing with these cases. In PXF cataracts presenting with no clinical manifestation of zonular weakness, in addition to adopting a zonule-friendly cataract surgery technique, some surgeons have suggested the routine use of CTRs to reduce or delay the incidence of late postoperative complications such as capsular phimosis and dislocation. CTRs may also facilitate scleral fixation of the IOL or capsular bag complex if symptomatic subluxation develops.

I use CTRs only in the presence of clinically manifest zonular weakness. In these cases, they offer two unique advantages. First, the device helps to redistribute the pressure 360° around the capsular fornix, including areas where the zonules are dehiscent. Second, the CTR keeps the capsular bag expanded throughout the procedure, thereby ren-
The timing of CTR insertion is controversial. The device can be inserted at any stage of the cataract procedure; however, if insertion is attempted before the nucleus has been removed (especially if the nucleus is hard) the process may be more traumatic to the zonules. Therefore, I place the CTR after emptying the capsular bag.

Use of an injector is a more precise and less traumatic method for CTR insertion than a manual technique. If intraoperative stabilization of the capsular bag is required, I prefer to use dedicated capsular hooks to support the loose capsular bag. The Ahmed CTS (Morchel GmbH) may also be helpful at this stage. At the conclusion of phacoemulsification, I suture a Cionni ring (Morchel GmbH) to the sclera with 9-0 polypropylene to permanently stabilize the bag.

Cortical clean-up can be a struggle if the CTR has been placed at an earlier stage during phaco. The Henderson CTR (Morchel GmbH), with eight equally spaced indentations, has been found to be of great help in these situations.

IOL choice and lens implantation. If the capsular bag is stable—with or without a sutured capsular stabilization device—I implant a one-piece hydrophobic acrylic posterior chamber IOL in the bag. I avoid plate-haptic IOLs because management becomes difficult should postoperative subluxation occur. If the bag is unstable or absent, scleral-suturing or gluing-in of a posterior chamber IOL can be considered. A current-generation anterior chamber IOL with a quadriflex design can be yet another option if the angle is open.

POSTOPERATIVE COMPLICATIONS

IOP spike. Postoperative IOP spike is a concern in patients with PXF. I usually prescribe an appropriate topical antiglaucoma medication for a few days after surgery. A systemic carbonic anhydrase inhibitor can be used if necessary. I have found that releasing aqueous through a paracentesis is an effective strategy to decompress the eye. The IOP spike may be blunted by careful removal of the lens matter and thorough evacuation of the OVD at the conclusion of surgery.

Late complications. These can include changes to the posterior or anterior capsule and spontaneous IOL subluxation. PCO is more frequent in patients with PXF than in the general population. When it becomes clinically significant, Nd:YAG capsulotomy should be performed.

In the anterior segment, postoperative anterior capsular contraction can occur early in eyes with PXF due to an imbalance of forces caused by zonular weakness. Progressive centripetal contraction can result in progressive zonulysis. If unchecked, this may lead to capsular phimosis with or without IOL decentration. The prophylactic measures that I consider for these challenging cases include the following:

- Creating an optimally sized, round, central capsulorrhexis (with a 0.5-mm overlap on the optic edge of the IOL);
- Vacuuming and polishing the undersurface of the anterior capsule prior to lens implantation;
- Employing a zonule-friendly surgical strategy; and
- Using a CTR or modified CTR.

I consider treatment at an early stage the moment I detect the beginning of capsular contraction. I perform Nd:YAG laser relaxing anterior capsulotomy through the annular capsular band at three or four symmetrically placed locations on the capsulorrhexis margin, which greatly reduces the chance for progression to capsular phimosis, IOL tilt, and decentration.

Spontaneous IOL subluxation. This late complication associated with PXF is increasingly encountered because of the expanding pseudophakic population. This problem can arise any time between several months and 16 years postoperatively, with an average of 8.5 years after IOL implantation. Several surgical techniques are reported in the literature to manage this condition, including IOL exchange, IOL repositioning, and suturing the IOL to the sclera or iris. In a symptomatic patient, I prefer to intervene at an early stage to achieve surgical correction with minimal manipulation.

CONCLUSION

PXF can pose significant challenges for the cataract surgeon. With proper preoperative work-up, intraoperative strategy, and postoperative follow-up, the surgeon can optimize outcomes, even in these difficult eyes.

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