# Manual Small-Incision Cataract Surgery in Pseudoexfoliation Syndrome

Pseudoexfoliation (PXF) syndrome is notorious for increasing the risk of intraoperative and postoperative complications during cataract surgery, primarily by affecting pupil size and zonular stability. In low-resource settings, where the populations with the highest cataract burden reside, cataracts are often removed using the manual small-incision cataract surgery (MSICS) technique instead of phacoemulsification. Further, MSICS may be useful in high-resource settings when the patient has dense cataracts and the surgeon wants to avoid excessive use of phacoemulsification.

## Epidemiology

Classically, PXF is associated with individuals of Scandinavian heritage, with prevalence rates up to 25%. However, among low-resource countries, there can be an unusually high prevalence of PXF with a concurrent high burden of cataract-related visual impairment. For instance, in Ethiopia, the prevalence of PXF in patients with cataracts has been reported to be as high as 39%,[1] while a recent population-based evaluation in central Ethiopia reported a prevalence of 13% in patients 40 or more years old.[2]

Although Ethiopia has one of the highest rates of PXF, there is considerable variation within Africa, as summarized in a systematic review by Olawoye et al. in 2014.[3] Prevalence studies in South Africa ranged from 5% to 16% depending on ethnic group; and clinic-based studies in Ghana, Nigeria, and the Democratic Republic of the Congo reported a prevalence of 0%, 3%, and 2%, respectively.[3]

## Pathophysiology

The defining feature of ocular PXF is deposition of extracellular fibrillar material. The origin is unclear, but the material is known to deposit systemically as well. Individuals with PXF have an increased propensity to develop cataracts, and the high PXF prevalence in Ethiopia may partly explain the high cataract burden there.

The hypothesis for the cataractogenic mechanism of PXF is that the composition of aqueous humor is altered, leading to changes in lens metabolism and subsequent cataract formation. Pupillary miosis can result from fibrosis, disorganization, or degeneration of the iris sphincter muscle. There is also an increased risk of posterior synechiae formation.[4] Further, accumulation of fibrillar material on the zonules causes zonular weakness, which increases the risk of posterior capsule rupture, zonular dialysis, vitreous loss, and retained lens fragments. If severe, this may result in lens subluxation or dislocation.

## Clinical Presentation

In the classic presentation, a concentric ring of dandruff-like material is noted on the anterior lens capsule. Other anterior segment features include corneal endothelial pigment deposition (i.e., Krukenberg spindle), hyperpigmentation of the trabecular meshwork, pupillary miosis, posterior synechiae, peripupillary transillumination defects of the iris, and white material at the pupillary margin. Signs of zonular weakness include shallow anterior chamber (AC), iridodonesis, phacodonesis, decentration of the nucleus, and vitreous prolapse in the AC.


***ENLARGED PUPIL.*** *Undilated eye after multiple iris sphincterotomies.*

Courtesy of Allison Jarstad, DO

## Surgical Management: Lens

**Expanding the pupil.** In some high-resource settings, pupil expansion may be achieved with viscodilation, intracameral dilating agents, mechanical pupil stretching, iris hooks, or various pupil expansion devices such as the Malyugin Ring (MicroSurgical Technology). However, in low-resource settings, the main technique is iris sphincterotomy.[5] This can be performed by inserting long Vannas scissors through the main incision to create

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eight evenly spaced 1-mm sphincterotomies (Fig. 1). Iridectomies have also been described for small, rigid pupils. If available, intracameral lidocaine and epinephrine can be effective as well. In addition, posterior synechiolysis may be required. Adequate pupil expansion is important when performing MSICS, as a small pupil interferes with prolapsing the nucleus into the AC, increasing zonular stress.

**Constructing the capsulotomy.** In MSICS, a large capsulotomy is preferred to deliver the nucleus.[6] Although a continuous curvilinear capsulorrhexis (CCC) may be performed, it is challenging to construct an adequately sized capsulotomy for nuclear prolapse. Various other types of capsulotomies can be performed, including a can-opener, "V," and smile- or envelope-shaped anterior capsulotomy (Fig. 2, and see this article at aao.org/eyenet for a related video). These techniques have been particularly helpful with Morgagnian, intumescent, or hypermature cataracts.[6]

In techniques with residual capsular flaps, there is added reassurance that the IOL will be inserted into the bag. When the anterior segment is filled with viscoelastic in preparation for IOL insertion, the tip of the cannula is placed inferiorly in the capsular bag, posterior to the capsular flap. As the capsular bag fills with viscoelastic, the anterior capsular flap moves anterior to the iris and folds inferiorly over the inferior iris. The folded flap thus forms a barrier to the inferior sulcus, helping to ensure that the leading haptic is not inadvertently inserted into the inferior sulcus (Fig. 3). Care must still be taken to rotate the trailing haptic into the bag.

**Envelope capsulotomy.** The envelope capsulotomy is advantageous because of the minimal zonular stress involved in its construction and the ability of the capsular opening to expand to accommodate the delivery of a bulky nucleus.[6]

To construct an envelope capsulotomy, we recommend making an initial horizontal subincisional cut in the anterior capsule with the keratome; this can be done at the same time the corneal incision is being enlarged. Two relaxing incisions are then made in the anterior capsule at either end of the horizontal anterior capsule incision. This is done by placing Vannas scissors at each end of the capsular slit with the tip of the scissors directed at a right angle to the initial cut.

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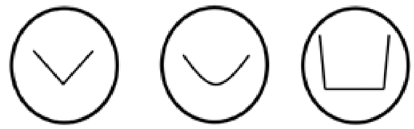

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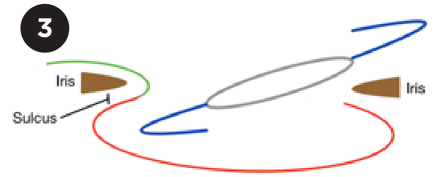


***PROCEDURES.*** *(2) Schematic diagram of "V," smile-shaped, and envelope-shaped anterior capsulotomies. (3) The leading haptic of the IOL (blue) is inserted into the capsular bag (red). Note that the anterior capsular flap (green), which is folded anteriorly over the iris, effectively blocks entry of the haptic into the sulcus, helping ensure in-the-bag placement.*

Courtesy of Stephen Lau, MBChB, and David Khorram, MD

Other advantages of the envelope capsulotomy include speed of construction; less postoperative inflammation and less corneal endothelial injury than other approaches; and it can be constructed even in the absence of a red reflex.[7,8] However, complications of the envelope capsulotomy include increased IOL tilt and decentration compared with CCC, and dyscoria caused by a residual capsular flap.[9]

**Prolapsing the nucleus.** The nucleus must be prolapsed into the AC before it is delivered through the sclerocorneal tunnel. Various prolapsing techniques have been described by Venkatesh et al.[7] Some may be accomplished mechanically with a Sinskey hook, others may be hydroprolapsed with a hydrodissection or Simcoe cannula.

In the hydroprolapsing technique, fluid is injected without decompressing the bag until one part of the equator of the nucleus is prolapsed out of the capsulotomy.[6] If done while one pole of the nucleus is pointing posteriorly and to one side, the opposite nuclear pole prolapses anteriorly. Further, partially emptying the AC of viscoelastic by pressing on the posterior lip of the incision helps promote nuclear prolapse. This maneuver minimizes zonular stress and can be performed safely, as the posterior capsule can withstand a pressure of 59 ± 10 mm Hg without rupturing. Once part of the nucleus is prolapsed, a Sinskey hook can be used to prolapse the rest of the nucleus into the AC. Alternatively, an intracapsular flip technique can be done by using the Simcoe cannula.[7]

**Delivering the nucleus.** The nucleus can then be delivered by a variety of methods such as hydroexpression, viscoexpression, vectis-assisted delivery, sandwich technique, or fishhook technique. Hydroexpression can be performed with the Simcoe cannula placed beneath the nucleus, infraducting the eye and exerting slight posterior pressure on the external lip of the incision.[6] The sandwich technique involves securing the nucleus between an irrigating vectis and iris spatula to assist in removal.[6]

The fishhook technique[10] involves using a 26- or 27-gauge needle with its tip bent in the shape of a fishhook (Fig. 4, and see this article at aao.org/eyenet for a related video). Once the superior pole of the nucleus is prolapsed into the AC, viscoelastic is injected posterior to the nucleus to separate it from the posterior capsule and underlying iris. The needle is inserted through the main incision with the fishhook tip parallel to the iris plane and slid behind the nucleus. Once under the center of the nucleus, the needle is rotated 90 degrees so that the tip faces anteriorly and impales the nucleus. The nucleus can then be delivered by pulling both needle and nucleus out of the main wound.

## Intraocular Lens Insertion

Considerations for inserting the IOL are similar to those following phacoemulsification. Because MSICS is usually used in low-resource settings, a PMMA lens is commonly used. A three-piece IOL in the bag helps provide stability in cases of zonular weakness.

The surgeon should be judicious when dialing the IOL into the capsular bag, as this maneuver exerts zonular stress. If available, a capsular tension ring (CTR) can be inserted to provide greater stability. However, use of a CTR is contraindicated if a CCC has not been made or if a posterior capsular rent occurs.

The IOL can also be placed in the sulcus if there is adequate capsular support. If a CCC was performed, the IOL may be optic-captured for improved centration. Otherwise, alternate methods of IOL implantation include AC IOLs and scleral-fixated IOLs.

## Management of a Subluxated Lens

With severe zonulopathy, the cataractous lens may subluxate. John et al. outline surgical principles for this situation.[11] These include:

- placing the incisions away from the area of zonular weakness to reduce stress on the existing zonules—but not to the detriment of the surgeon's comfort and ability to manipulate the tissue;
- making the smallest incision possible to minimize fluid egress and AC collapse; and
- using a generous amount of cohesive viscoelastic over the area of zonular dialysis to help tamponade the vitreous.

A CCC is preferred in this situation because of higher risk for a radial capsular tear when working at the lens periphery with other capsulotomy techniques. However, creating a CCC is particularly difficult in MSICS because of the larger-sized capsulotomy required, compounded by zonular weakness. Once the capsulotomy is performed, hydroexpression of the nucleus can be performed as described above.

If lens removal by means of MSICS poses difficulties, an intracapsular technique may be required to prevent the nucleus from dropping into the vitreous, potentially causing vigorous lens-induced posterior segment inflammation and retinal damage. This is particularly important in low-resource settings when vitreoretinal surgery support is not available.

If a posterior chamber IOL is chosen but is difficult to insert into the bag, the IOL may first be placed into the AC; the surgeon can then use a two-handed technique, inserting the superior haptic into the bag, followed by the inferior haptic. It is recommended that the haptics be oriented at whatever axis provides the best centration intraoperatively.[11] Although various methods of scleral fixation may be used, they may be impractical in low-resource settings.


***FISHHOOK.*** *A 26- or 27-gauge needle bent into a fishhook shape can be used for nucleus delivery.*

Courtesy of Allison Jarstad, DO

## Conclusion

Although a small pupil and zonulopathy are common in patients with PXF, these challenges can be encountered in any cataract case. Tackling these problems in a low-resource setting is particularly difficult in the absence of helpful adjuncts such as pupil expansion and capsular support devices. However, the principles described above should prove useful to surgeons who are working to cure blindness from cataract.


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