Master Techniques in Cataract and Refractive Surgery

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INTRODUCTION

Pseudoexfoliation (PEX) syndrome is an ocular mystery. Many investigators over the years have attempted to clarify and identify its peculiar etiology, appearance and propensity for ocular pathology.\textsuperscript{1,2} Many associations have been established, complications treated, and manifestations documented. It has been found sporadically in many populations worldwide, yet its exact makeup and point of origin in the eye remain obscure.\textsuperscript{3} PEX syndrome has the potential to create difficulties and catastrophic complications in cataract surgery. The study of PEX syndrome is important.

In this chapter we will describe in detail our current technique, designed to prevent and reduce the incidence of complications in cataract surgery for PEX patients. We will also offer a review of current information on this intriguing disease.

PEX syndrome is a systemic degenerative disorder that is characterized in the eye by deposits of an irregular meshwork of fibrillar eosinophilic material. This material may be found on the structures of the anterior and posterior chambers. This condition may be associated with cataract and glaucoma. The precise composition of the PEX material has not yet been identified. Studies suggest an important role of proteoglycans in the pathogenic pathway in PEX syndrome.\textsuperscript{4-11} PEX syndrome has been suggested to include a blood-aqueous barrier impairment associated with higher protein content in the aqueous humor. It has also been suggested that PEX syndrome may be associated with elevated serum amyloid levels,\textsuperscript{12} but this has not been substantiated.\textsuperscript{13}

In PEX syndrome, conjunctival biopsy may reveal the presence of PEX material, even though the conjunctiva does not display clinical manifestations.\textsuperscript{14} PEX material has been documented on the ciliary processes and zonule,\textsuperscript{15} but PEX syndrome is more commonly associated with deposition of material on the anterior lens surface, which is more readily visualized with dilated pupils.\textsuperscript{16} Deposition is more marked in the mid-periphery of the lens; with a translucent central zone surrounded by an intermediate clear zone (iris movement denudes the capsular surface of PEX material) (Figure 14-1). Deposition material is also frequently seen at the pupillary margin and is often associated with iris transillumination defects (“moth-eaten” appearance).\textsuperscript{17} Posterior synechiae are often associated with PEX syndrome. Broad posterior synechiae and miosis may prevent adequate viewing of the anterior lens capsule, making the clinical diagnosis of PEX syndrome difficult. In eyes with broad, circular posterior synechiae, the possibility of PEX syndrome should be considered.\textsuperscript{18} Pigment and PEX material (flakes) are sometimes found on the corneal endothelium. A reduction
in the number of endothelial cells may be present.\(^{14,19,20}\)

One of the major difficulties encountered with PEX is glaucoma. PEX glaucoma is likely secondary to the accumulation of PEX material in the trabecular meshwork.\(^{21-23}\) The meshwork is frequently pigmented in a patchy fashion in contrast to the dense, homogeneous deposition seen in pigmentary dispersion syndrome.\(^{16}\) Pigment dispersion syndrome should be included in the differential diagnosis for PEX syndrome as it is also associated with the deposition of material within the ocular tissues. Pigment dispersion syndrome and pigmentary glaucoma tend to occur in younger, myopic patients, with pressure spikes seen after exercise.\(^{24}\) In contrast, PEX syndrome tends to occur in the sixth decade and beyond. In pigment dispersion syndrome, Krukenberg’s spindle, a triangular-shaped adherence of pigment to the corneal endothelium is present due to the current flows of the aqueous humor. In PEX syndrome, white fibrillar material in no apparent pattern may be noted on the endothelium. In pigment dispersion syndrome, radial, midperipheral iris transillumination defects can be seen with retroillumination. In PEX syndrome, iris atrophy can also be seen, but it usually has a peripupillary distribution (Figure 14-2). In pigment dispersion syndrome, a peripheral iridotomy has been shown to change the dynamics and architecture of the lens-iris diaphragm, often affecting a cure.\(^{25}\)

PEX glaucoma is likely secondary to the accumulation of PEX material which blocks aqueous outflow in the trabeculum.\(^{26,27}\) IOP tends to be higher than in eyes with primary open-angle glaucoma (POAG).\(^{28,29}\) PEX glaucoma is associated with greater visual field loss and worse optic nerve cupping.\(^{30,31}\)

PEX glaucoma tends to be less responsive to medical therapy than POAG\(^{31-35}\) and surgical treatment is more commonly necessary.\(^{31,36}\) In PEX glaucoma, argon laser trabeculoplasty is initially effective in lowering IOP,\(^{37,39}\) but there is a significant loss of effect over long-term follow-up.\(^{37}\)

Filtering surgery for PEX glaucoma has similar results to POAG.\(^{40}\) In true exfoliation of the lens, which is secondary to trauma, chronic exposure to heat, or inflammation, elevated pressures are not typically seen.

The association of PEX syndrome with phacodonesis and spontaneous subluxation of the lens is due to zonular breaks at the insertion of the zonular fibers into the ciliary body epithelium and not at the insertion of the zonule into the lens capsule.\(^{41,42}\) Some studies imply a genetic role at the cellular level in the pathogenesis of PEX syndrome.\(^{43-46}\)

Different reports have given a wide range in the prevalence of PEX according to age and sex distributions, which may be due to genetic factors, differences in diagnostic technique, or differences in the populations studied.\(^{47-53}\) The prevalence of PEX syndrome has been studied in Scandinavians and other Europeans,\(^ {48,54}\) Japanese,\(^ {15}\) Australian aborigines,\(^ {49}\) Australian non-aboriginal adults,\(^ {55}\) Navajo Indians,\(^ {56}\) natives of India,\(^ {57}\) and Pakistan,\(^ {58}\) Bantu tribe of South Africa,\(^ {52}\) African-Americans,\(^ {59}\) and others.

PEX syndrome tends to be bilateral but is usually clinically asymmetric rather than unilateral. Upon clinical examination, many patients with PEX syndrome reveal only unilateral ocular involvement. This has been investigated by transmission electron microscopy and immunohistochemistry. When 1 eye demonstrates clinical evidence of PEX syndrome, alterations can be found in the anterior segment tissue of the fellow eye. Because PEX syndrome is associated with glaucoma and is an important risk factor for complications during cataract surgery, the potential involvement of both eyes in the PEX process is important.\(^ {60,61}\)

In PEX syndrome, involvement of the lens, zonule, ciliary body, iris, trabecular meshwork, and corneal endothelium may result in open-angle glaucoma, angle-closure glaucoma, phacodonesis, lens dislocation, and/or poor pupillary dilation. When performing cataract surgery in the presence of PEX syndrome, special consideration must be given to the
increased risk of complications such as lens subluxation, zonular dialyses or breaks, posterior capsular rupture, vitreous loss, subluxation of the IOL, hemorrhage, formation of posterior synechiae, and corneal endothelial decompensation.

Even though PEX syndrome has traditionally been associated with increased risk, modern cataract surgery with appropriate surgical technique and preventative measures makes it possible to achieve good results, avoiding the increased complication rate attributed to PEX syndromes.

**INDICATIONS AND SPECIAL CONSIDERATIONS FOR CATARACT SURGERY IN PEX SYNDROME**

Cataract surgery should be performed when reduced visual function impairs the quality of life sufficiently to warrant the risk of surgery. The increased risk of complications associated with PEX syndrome must be balanced against the experience and expertise of the operating surgeon. In the best of hands, when all resources are brought to bear, it is possible to reduce the risk of complications considerably. Most of the potential complications associated with PEX syndrome can be either prevented or readily managed during surgery, even when there is a complete dislocation of the lens. The surgeon should fully discuss with the patient the indications, risks, and benefits for the proposed cataract surgery. The decision to perform or delay the surgery and whether there is a benefit in referring the patient to a more experienced surgeon should be considered.

A comprehensive ophthalmic examination should include a complete history of current and past medical pathology including specific questions about systemic illnesses (ie, diabetes, systemic hypertension, ischemic heart disease, chronic pulmonary disease, renal disease, obesity, mental status) and all medications taken. A careful personal and familial ocular history is also important (cataract surgery on the fellow eye, glaucoma, trauma, inflammatory episodes, amblyopia, infections, previous ocular surgery complications, and any topical medications).

Various aspects of visual function should be considered. Testing may include visual acuity at distance and near, visual field testing, color vision, contrast sensitivity, light adaptation, and depth perception. Cataracts may coexist with other causes of decreased visual function. In the presence of significantly decreased visual acuity and a dense cataract, the ophthalmologist may evaluate entoptic phenomena, use the Potential Acuity Meter, perform laser interferometry, A - and B-scan ultrasonography, and/or visual electrophysiology (electroretinography-ERG and visually evoked potentials [VEP]) to determine the possibility of visual rehabilitation.

Brisk pupillary reflexes suggest good retinal function. The extent of pupillary dilation with mydriatics should be evaluated. If the pupils will not dilate widely, posterior adhesions may be present or the dilator muscle is weak. It should be anticipated that appropriate measures might be necessary to enlarge the pupil during surgery to allow adequate access to the lens. These may include stronger mydriatics, topical non-steroidal anti-inflammatory drugs (NSAIDs), epinephrine in the irrigating solution, lysis of posterior adhesions, pupillary expansion devices, iridectomy, stretching the pupil, or sphincterotomy.

Preoperative examination of the eyelids and lacrimal apparatus should identify cases of blepharitis, entropion, lagophthalmos, keratoconjunctivitis sicca, and dacryocystitis. This will enable appropriate measures to be taken to reduce the risk of infection, keratitis, or wound-related complications. Corneal endothelial dystrophy, as seen frequently with PEX syndrome, might lead to clinically significant corneal decompensation. Iridodonesis and phaco-donesis may herald a dislocated or subluxated lens and the surgeon should be prepared for the possibility of vitreous loss during surgery. Fundus examination with pupillary dilation should be performed to detect peripheral retina pathology.

If the patient has medically controlled glaucoma, it may be anticipated that intraocular pressure will remain under control with the same or less medications after surgery. If glaucoma control is poor, a combined trabeculectomy and cataract procedure should be considered. The combined procedure has greater associated risk than phacoemulsification with IOL implantation alone.

Prior to scheduling surgery, all patients should be fully informed of risks and benefits, the alternatives and elective nature of their procedure. Options for optical correction, including the different types of implants and their desired postoperative refractive status—both eyes focused at distance, monovision, and multifocal IOLs should be considered. A final visual outcome of emmetropia to mild myopia is usually ideal, but the refractive error and overall status of the fellow eye should be considered carefully since anisometropia may not be well tolerated. Appropriate informed consent is obtained.

**SURGICAL TECHNIQUE**

**Anesthesia: The Rand-Stein Analgesia Protocol**

Surgical technique and good results partially depend on good anesthesiology. The Rand-Stein Analgesia Protocol (RSAP) is an intravenous technique for providing profound ocular and body analgesia virtually without sedation. Anxiety and patient cooperation are managed separately with intravenous sedative medication. The control of pain, anxiety and patient cooperation are even more important in
the presumed fragile ocular environment of PEX syndrome. We have consistently used the RSAP on all of our cataract surgery patients.95 We have not had an intraoperative conversion to local anesthesia in more than 15,000 cases. This technique can be expected to allow the PEX syndrome patient to undergo a controlled, painless and anxiety free, cataract procedure using a suture-less corneal incision with virtually no probability of having to rely on or convert to local anesthesia.

Reviewing the literature, we find that general anesthesia is seldom used for cataract surgery. However, local (retrobulbar or peribulbar) anesthesia with intravenous sedation is still in common usage. Retrobulbar and peribulbar anesthesia techniques for cataract surgery are associated with potentially disfiguring, blinding, and life-threatening complications.96-99 The very nature of a blind injection into the periocular tissues carries with it the potential for catastrophic retrobulbar hemorrhage, which can cause permanent blindness.94 Many local anesthesia blocks fail to provide adequate ocular analgesia.95

Careful anesthesiology monitoring is indispensable to prevent and control complications in elderly patients as they often have serious associated systemic disease, such as coronary artery disease, hypertension, diabetes, and/or chronic lung disease.96,97

With phacoemulsification, the necessity for complete ocular akinesia has been eliminated. Topical anesthesia techniques significantly reduced the risk of surgically induced diplopia, amaurosis, ptosis, lid ecchymosis, and pain associated with injection anesthesia.98 Topical anesthesia, however, is inadequate for providing profound internal analgesia for the eye and offers no remedy for the management of the uncooperative patient.99,100 Approximately 10% of topical anesthesia patients require intraoperative conversion to local anesthesia with the eye already surgically opened.101 Topical anesthetics and intracameral anesthetic agents have the potential to cause endothelial cell injury,102 and they can damage the ocular surface in older patients with dry eye and blepharitis.103 The RSAP eliminates the risks of topical and local anesthesia. The RSAP offers the benefits of reduced morbidity while providing control of the patient’s ability to cooperate.

The RSAP uses low-dose intravenous Alfentanil HCl (Taylor Pharmaceuticals, Decatur, Ill) for its intense, rapid-onset analgesia without sedation.104,105 Low-dose Methohexital Sodium (Eli Lilly, Indianapolis, Ill) provides a rapid-onset, ultra-short-acting sedative effect that precisely controls the patient’s state of alertness. Preoperative Midazolam HCl (Abbott Laboratories, North Chicago, Ill) can be used optionally for preoperative anxiety.

Alfentanil HCl is reversible with Naloxone HCl (Abbott). Midazolam HCl is reversible with Flumazenil (Romazicon, Roche, Nutley, NJ). Methohexital sodium requires no reversal agent because of its ultra-short duration of action.

Droperidol (American Regent Laboratories, Shirley, NJ) can be used for its antiemetic function when nausea is present and an additional sedative effect is desired. We use Metoclopramide (Baxter Healthcare Corporation, Irvine, Calif) for nausea when no additional sedation is needed (Table 14-1).

THE CATARACT PROCEDURE IN PEX SYNDROME

Cataract surgery in PEX syndrome has the potential to become complicated and extensive due to inherent structural weakness. By utilizing a precision microsurgical approach, these complications can be significantly reduced, yielding consistently better postoperative results. Cataract surgery in PEX syndrome will frequently encounter small pupils, shallow anterior chambers, posterior adhesions, weak zonular support, partial subluxation, or complete dislocation of the crystalline lens. Final placement of the implant may be adversely affected by inadvertent stress exerted upon the zonular structures during surgery, resulting in subluxated or dislocated lens implants. This may become apparent during the intraoperative, postoperative, or even in the long-term postoperative period. The principles that will be described here may be useful for performing better surgery for all surgical patients, but become more critical in the unstable ocular environment of PEX syndrome.

Preoperative Considerations

In patients with PEX syndrome, intraoperative pupillary size can be expected to be significantly smaller compared to normal patients undergoing cataract surgery. Postoperatively, IOP and aqueous cell response is similar in both groups, but a significantly higher flare response has been observed in PEX syndrome patients.106 Topical Ketorolac Tromethamine 0.5% (Acural, Allergan, Irvine, Calif) is an effective inhibitor of miosis during extracapsular cataract extraction and IOL implantation. It provides a stable mydriatic effect throughout surgery.107

Prep and Drape

After instilling tetracaine hydrochloride drops (Alcon, Fort Worth, Tex), sterile prep and drape (Cataract Pack # 6974-03, Alcon) are performed. While the surgeon is scrubbing, analgesia and sedation are initiated in accordance with the RSAP guidelines.

Speculum and Eye Wash

A speculum (Barraquer Adult Speculum, Bausch & Lomb Surgical, St. Louis, Mo) is placed. The eye is washed with BSS (Sterile Irrigating Solution, Alcon). 5% Iodine-Povidine (Aplicare, Branford, Conn) is placed in the con-
junctival cul-de-sac for 30 seconds. Antibiotic drops, such as Gentamycin. (American Pharmaceutical Partners. Los Angeles, Calif) and/or Cefalozin (Apothecon, Bristol-Myers Squibb, Princeton, NJ) are placed in the cul-de-sac, prior to the first incision, the counter-incision.

**Surgical Incisions**

For the past 15,000 cataract surgeries with foldable lens implantation, we have utilized CCIs. Obviously, some PEX syndrome patients required sclero-corneal incisions, such as those in whom combined cataract surgery and trabeculectomy or larger, PMMA implants (retinal pathology) were indicated. We believe that CCIs are the procedure of choice for PEX syndrome patients. CCIs generate less inflammation, irritation, pain, and redness, because the conjunctiva is not traumatized. The conjunctiva is conserved for possibly needed glaucoma surgery in the future. CCIs generate minimal astigmatism in the axis where they are made (flattening this axis approximately 0.50 to 0.75 D in the authors’ experience). The incision can be made in the steepest axis and the counter-incision approximately 90 degrees away (Figure 14-3). We will describe the procedure for a right-handed surgeon in a slight with-the-rule astigmatism eye.

The procedure is designed to provide the gentlest tissue-handling possible of the ocular structures, preserving the integrity of structures that might be significantly weakened. Careful attention to the principles of precision microsurgery are strictly adhered to, including frequent refocusing of the microscope and a 3-D proprioceptive technique, which are continuously employed to significantly reduce stress on the cornea and zonules.

The procedure starts with a counter-incision at the 2- to 3-o’clock position through the posterior limbus, 1 mm in size. By cutting the tip off of an eye spear (Cellulose Sponge Spear, Hurricane Medical, Brandenton, Fla), approximately halfway down, the spear can be used as a blunt instrument. This avoids using a forceps that can cause a conjunctival hemorrhage. The sponge is placed on the limbus at 180

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**Rand-Stein Analgesia Protocol Summary**

**Preoperatively (In The Preoperative Area)**

- **Midazolam HCl**, 1 mg IV in preoperative area after vitals signs confirmed stable. If needed, additional 1 mg IV, 10 to 15 minutes after first dose.
- **Reversal agent**: Fluromazenil, 2 cc (0.2 mg) IV

- **Methohexitol sodium**, 1 cc (10 mg) IV, PRN, 10 to 15 min before transfer to the operating room. Used only in cases with severe preoperative anxiety.
- **Reversal agent**: None needed, short acting, less than 3 to 5 min, if inadvertent overdosage occurs, use simple. If SaO₂ falls below 90%, suspend administration and remind patient to breathe in and out deeply. If needed, Ambu ventilation until spontaneous respiration returns, usually 3 to 5 min.

- If prior history of nausea or vomiting, (previous anesthesia), pre-treat with:
  - **Droperidol**, 1 to 2 mg (75 mcg/Kg) IV, on arrival (for sedative/antiemetic effect), or
  - **Metoclopramide**, 10 mg, IV, on arrival (for a pure antiemetic effect, without sedation)

**In the Operating Room**

- **Tetracaine HCl**, 1 drop previous to washing, prepping and draping the eye.

- **Alfentanil HCl**, (500 mcg/cc) 4 to 6 doses of 125 mcg (1/4 cc), every 30 to 45 seconds. For inadequate analgesia or to prolong the analgesia effect: additional Alfentanil HCl, 125 mcg (1/4 cc) IV, every 30 to 45 seconds, until relief of pain. If SaO₂ falls below 90%, suspend administration and remind patient to breathe in and out deeply, as needed.
- **Reversal agent**: Naloxone HCl, 0.2 to 0.4 mg (1/2 to 1 cc)

For intraoperative anxiety or persistent anxiety, squeezing, poor cooperation:

- **Methohexitol sodium**, 1 cc (10 mg) IV, every 2 minutes until relief of anxiety, and reassertion of control.

**Postoperatively**

- If nausea or vomiting during or after surgery
  - **Metoclopramide**, 10 mg IV

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**Table 14-1**
degrees from where the counter-incision is to be made, minimizing conjunctival trauma while providing sufficient grip to prevent the eye from moving (Figure 14-4). The tip of a 1-mm (30-degree angle) Crystal Keratome (HUCO Vision SA, St. Blaise, Switzerland) is placed at the posterior margin of the limbus and is advanced at an angle that will penetrate into the anterior chamber, making a corneal tunnel of approximately 2 mm in length and 1 mm in width. The anterior chamber is then filled with Viscoat (Alcon Ft. Worth, Tex) without overfilling. The viscoelastic solution protects the corneal endothelium and deepens the anterior chamber (Figure 14-5).

A Bechert Rotator (Bausch & Lomb Surgical, St. Louis, Mo) is then inserted into the counter-incision and braced against the edge of the incision to prevent the eye from moving. Placing the tip of a 3.2-mm (60-degree angle) Crystal Keratome at the posterior margin of the limbus, approximately 90 degrees from the counter-incision, pressure is applied pushing at an angle that will allow penetration into the anterior chamber after producing a corneal tunnel of approximately 2 to 3 mm in length and 3.2 mm in width (Figure 14-6). Initially, the incision is engaged with a slight downward direction. Then the 3.2-mm Crystal Keratome is quickly re-directed so it becomes parallel to the plane of the cornea and enters the anterior chamber more or less horizontally. This consistently creates corneal incisions with self-sealing valves. Additional Viscoat may be injected into the anterior chamber, to protect the cornea during the capsulorrhexis.

Posterior Adhesions and the Small Pupil:

If there are posterior adhesions of the iris, or if the pupil does not dilate well for any reason, this must be addressed before doing the capsulorrhexis. Two ideal instruments are the Bechert Rotator and the Kuglen Hook (Bausch & Lomb Surgical St. Louis, Mo). These instruments can be inserted through the incisions and by “pulling” in opposite directions, they can effectively stretch the pupillary margins.
enlarging the pupillary aperture (Figure 14-7). Alternatively a pupil-stretching device can be used. We no longer use these devices because they are cumbersome and time consuming. If the pupil cannot be enlarged sufficiently using a bimanual stretching techniques, we prefer to make a series of appropriate sized radial sphincterotomies. Although not as cosmetically attractive, these sphincterotomies produce a much safer pupil access environment and assure much better postoperative retina visualization.

Capsulorrhexis

The advantages of "in-the-bag" PCIOls make the CCC the preferred method of capsulotomy. Especially important in PEX syndrome, the force is applied tangential to the zonule when creating a continuous circular tear. This reduces direct traction on the zonule and the risk of zonular dehiscence. The smooth edge capsulotomy with the absence of irregular anterior capsular tags or flaps reduces the risk of inadvertently pulling on the capsule, causing disinsertion. If a capsular tag becomes engaged in the automated tip during emulsification or during irrigation/aspiration of cortical material, this can cause zonule and/or capsular dialysis and lead to vitreous loss. When a posterior capsular tear occurs (with or without vitreous loss), an intact anterior capsular ring can still provide excellent support for a PCIOl with the optic placed anterior to the capsulotomy and the haptics placed in the ciliary sulcus.

Utilizing different instruments, 2 basic physical principles can be applied during CCC—shearing (cutting) and ripping (tearing). When cutting (shearing) the anterior capsule, the vector forces created by the instrument that generates traction (from A to B) on the capsular flap (CF) is parallel to the vector in which the cut is made (from A to B) (Figure 14-8). When tearing (ripping) the anterior capsule, the vector of the force created by the instrument that generates traction (from A to B) on the capsular flap (CF) is not parallel to the vector in which the cut is made (from A to B) (Figure 14-9).

These two concepts have been explained in a 2-D plane (X = 9 to 3 and Y = 6 to 12). In order for an appropriate capsulorrhexis to occur, all traction in the third dimension (anterior-posterior or Z) should be eliminated or carefully controlled. The anterior chamber should be adequately filled with viscoelastic solution to avoid displacement of the lens (too much viscoelastic will displace the lens posteriorly and not enough anteriorly). When the lens capsule is displaced anteriorly or posteriorly, the zonule will exert traction on the capsule. These forces should be neutralized to avoid an equatorial extension of the capsulorrhexis. The anterior capsule can also be pushed posteriorly against the anterior cortical masses; this generates vectorial forces that will alter the radius of curvature of the capsulorrhexis. Understanding of these principles can be useful to reduce the radius of curvature (in order to bring the capsulorrhexis towards the center and away from the periphery). If too much pressure is applied, the cystotome might tear the anterior capsule or even rupture the posterior capsule.
Our capsulorrhexis is created utilizing only a bent 22-gauge needle (Becton Dickinson and Company, Franklin Lakes, NJ). Although we do not find it necessary, the CCC can also be done with Utratta Forceps (Bausch & Lomb Surgical, St. Louis, Mo). To bend the 22-gauge needle, the needle is grasped in the non-dominant hand with a Castroviejo Needle Holder—Heavy (Bausch & Lomb Surgical). The needle’s tip is grabbed with another needle holder in the dominant hand and bent away from the bevel until approximately an 80- to 90-degree angle is created (Figure 14-10).

Our technique for capsulorrhexis:\textsuperscript{109} is as follows: The bent needle is placed in the lower left quadrant (approximately at 4:30) (Figure 14-11). Using a relatively quick sweeping motion (the needle is twisted or rotated while traction is exerted from 4:30 toward the 8- or 9-o’clock position), a triangular, anterior capsular tear is made from the 4:30 position until approximately the 6-o’clock position (Figures 14-12 and 14-13). The initial motion is to pull the capsulotomy tangentially when close to the site of tearing, but as the tear becomes more peripheral and away from where the bent 22-gauge needle is grabbing the anterior capsule, the more radial a force needs to be exerted (toward the center of the anterior capsule).

The bent 22-gauge needle is then placed at the 6-o’clock area, near the tear, and gentle pressure is applied on the anterior capsule while the needle creates traction towards the 10- or 11-o’clock position. As previously described, traction initially is tangential to the capsulotomy, but traction shifts to a radial force (toward the center) as the tear extends peripherally. This brings the tear to the 9-o’clock position (Figure 14-14). Once again, the bent 22-gauge needle is repositioned near the tear at the 9-o’clock area, and pressure is applied on the anterior capsule while the needle creates traction towards the 12-o’clock position. As the tear becomes more peripheral and away from where the bent 22-gauge needle is grabbing the anterior capsule, the more radial force is exerted (toward the center of the anterior capsule) (Figures
14-15 and 14-16). The bent 22-gauge needle is then placed near the tear at the 12-o’clock position, and traction is generated towards 4-o’clock position (Figure 14-17). As previously described, this motion is continued by a more radial traction vector (toward the central capsule), and continued until the CCC is completed.

It is very important to observe the striae (stretch marks) formed on the anterior capsule because they will predict where the tear is going. This is even more important when the pupil dilates poorly, because capsulorrhexis can be created under the iris without direct visualization. This procedure requires expertise and shouldn’t be attempted before mastering the above technique. To minimize the possibility of the capsulotomy flaring out, downward pressure is exerted onto the nucleus to keep the capsulotomy centripetal (turns inward, toward the center). When the capsulotomy size needs enlargement (a larger radius of curvature or to flare out), the needle is placed more superficially and ahead of the already torn portion of the capsule so that it extends outward (centrifugal). In this manner, the capsulotomy can be kept reliably on course.

An important point is to apply only enough pressure to keep the needle from slipping off the capsule edge as it tears. If too much force is applied, the epinucleus becomes scuffed, the surgeon will not be able to identify the cut capsule edges and will loose control of the capsulotomy.

**Hydrodissection**

The main purpose of the hydrodissection maneuver is to float the nucleus out of the bag to reduce the stress that can be transferred to the zonular elements. This is essential in reducing complications in cataract surgery for patients with PEX syndrome. Hydrodissection uses BSS under pressure to separate the capsule from the cortex and cortex from the...
nucleus. By separating the different layers of the lens, the nucleus can be floated out of the bag and freely rotated during emulsification without stressing the fragile zonular system.

A 25-gauge hydrodissection cannula (Bausch & Lomb Surgical, St. Louis, Mo) is placed approximately 90 degrees with a tangent line to the edge of the CCC. The tip of the cannula is introduced just underneath the anterior capsule approximately 2 mm in the direction of the equator, as far as direct visualization permits. It should be placed at approximately 180 degrees from the counter-incision. BSS is then continuously injected under pressure with the tip held firmly in contact with the underside of the anterior capsule flap, forcing the fluid to flow all the way around the lens. We want a free egress of fluid from the eye during this maneuver. It helps to wash out the viscoelastic situated between the area of the hydrodissection and the 3.2-mm incision, in order to prevent initial overfilling of the eye. The hydrodissection cleavage can take place between the capsule and cortex (Figure 14-18), or between the cortex and nucleus (Figure 14-19), accomplished by burying the cannula deeper within the lens substance. Either way, the nucleus is separated from the capsule and any force exerted upon the nucleus will no longer be transmitted directly to the zonule structure. Several hydro-dissecting BSS injections might be needed in order to completely loosen the nucleus until it floats out of the bag, at least 180 degrees. When changing the position of the hydrodissection cannula, the surgeon should avoid Descemet’s membrane and endothelial trauma.

**Phacoemulsification (Nucleus Equatorial Reduction Technique)**

We prefer machine settings that are relatively aggressive for our phacoemulsification machine (Diplomax, American Medical Optics, North Andover, Mass). The ultrasound is set to 100% power, but it is flexibly controlled in a linear and pulsed ultrasound mode. The maximum aspiration rate is set to 34, also in linear mode. The maximum vacuum limit is at the 150 level. These high levels provide maximum power and force, but remain entirely adjustable for appropriate intraoperative modulation. The bottle height is approximately 33 inches above the level of the patient’s eye.

Viscoat is placed between the prolapsed nuclear equator and the corneal endothelium. Maintaining a protective film of thick viscoelastic material is necessary to protect the endothelium. Phacoemulsification starts by simply sculpting out the central nucleus to remove the central bulk of the lens. A vital consideration is that the surgeon must continuously maintain a constant depth of the anterior chamber with stable inflation of the posterior capsule. This prevents chamber collapse with its potential damage to the capsule, zonules, or cornea through inadvertent stress or contact with the surgical instruments. This is accomplished by making sure that the aspiration port of the phacoemulsification tip is always occluded with lens material whenever the aspiration mode (position 2 or 3 on the foot switch) is engaged.

During all manipulations, it is important to apply 3-D thinking to the surgical process. This can considerably reduce or eliminate much of the tissue distortion that can occur. It is common for less-experienced surgeons to regard their instruments as being fixed in a horizontal plane. If the phaco tip is directed downward, there is no need to commit the rest of the instrument to descend in the same plane. This can cause needless tissue distortion, trauma, and unnecessary stress. The instruments should be conceptualized to rotate around the incision as if it were a fulcrum, central to all movements in all 3 dimensions.

In the emulsification process, the phacoemulsification tip is initially held in an almost vertical position as it enters the eye and engages the nucleus. It then slides forward assuming a more horizontal direction as it advances toward the 6-o’clock position of the nucleus. This can be better under-
stood if we consider the direction of movement of a spoon as it enters a bowl of cereal or when scooping ice cream from a large bowl.

We perform several passes in order to debulk the center of the lens nucleus (Figure 14-20). Occasional air bubbles and lens debris may be trapped in the protective layer of Viscoat. This material can be useful in confirming that the endothelium is well insulated and protected. It is not necessary to perfectly visualize all of the structures and lens material at all times, as long as we can be confident that the posterior capsule remains expanded with no potential for anterior chamber collapse and the cornea is adequately shielded. We can not overemphasize the key element of continually maintaining an occluded aspiration port on the ultrasonic probe while aspiration is engaged (position 2 and 3 on the foot pedal). Otherwise, fluid will be suctioned out of the eye faster than it can be infused and the posterior capsule will come forward, increasing the risk of capsule rupture and vitreous loss. Additional Viscoat can be periodically instilled to maintain the protective coating of viscoelastic material.

After the initial sculpting, the lens is floated up by hydrodissection. When hydrodissection is infused at about the 9-o’clock position, the equatorial lens will tend to dislocate and tilt upward at approximately the 3-o’clock position. The anterior equator of the left side of the nucleus will usually float up over the level of the capsulotomy and over the pupillary plane (Figure 14-21). In this position, the nucleus can readily be maneuvered with a Bechert Rotator placed through the counter-incision. Additional hydrodissection may be necessary to refloat the nuclear tilt from time to time.

Once the lens has been tilted into the pupillary plane, it can be assumed that it has been sufficiently floated out of the bag and the zonules will no longer bear all of the pressure of the phacoemulsification process. The lens removal strategy is to sequentially reduce the equatorial diameter. As the equator-to-equator lens diameter is reduced, the ability of the lens to exert force on the intraocular structures is diminished. To avoid endothelial trauma, ultrasound fragmentation should always occur at or below the level of the pupil (iris). The nucleus is maintained at approximately the pupillary plane with a Bechert Rotator holding up the lens through the counter-incision as ultrasonic fragmentation is performed. The equatorial reduction technique gradually wears away the equator, as the lens is gradually rotated 360 degrees. The phacoemulsification tip slides sideways or is directed peripherally through the equatorial nucleus from the center outward, amputating a portion of the equator. Counter-traction is applied by pushing the nucleus with the Bechert Rotator. The force applied may be used to produce a nuclear cracking to reduce the nucleus into smaller, more manageable pieces (Figures 14-22 and 14-23).

As the phacoemulsification tip advances peripherally, the Bechert Rotator controls the nucleus, "feeding" the phacoemulsification tip, while holding the nucleus at the pupillary plane. As the equator is reduced, the lens diameter shrinks, lessening the forces against the capsule (posterior and equatorial). The nucleus is slowly rotated in a bimanual fashion utilizing the Bechert Rotator and the phaco tip, until the entire equatorial nucleus has been removed. A mixture of lens material, air bubbles, and viscoelastic between the phaco tip and the cornea may partially impede visualization but this buffer zone protects the endothelium. A combination of feeding the lens material, bimanual nuclear cracking and short horizontal movements of the phaco probe can facilitate the nuclear removal process, all the time keeping force from impacting the lens zonule. The last nuclear fragments should be emulsified by holding them against the phaco tip, while reducing the ultrasound energy applied with the foot pedal. The use of low ultrasound energy reduces the tendency for the phaco probe to repel lens fragments and can actually be
more efficient in removing the smaller particles. Nuclear fragments can be held against the phacoemulsification tip, to emulsify them or to crack them into smaller pieces. This can prevent fragments from being directed against the endothelium by the turbulence (Figure 14-24).

Sometimes, lens material will remain hidden under the iris or in the peripheral anterior chamber. We use a Bechert Rotator to move the iris, stretching the pupil to the angle while the I/A tip is still irrigating in the anterior chamber to search out and reveal any nuclear or cortical fragments that may remain.

Management of the Cortex: Irrigation and Aspiration

It is quite common in PEX cataracts to have significant amounts of thick cortex. Much of the thicker cortical material can be removed with the phacoemulsification tip using I/A only, without ultrasound or with very short, low power bursts to increase the flow of lens material into the phaco probe (Figure 14-25). The remaining cortical material is removed with a 0.3-mm I/A tip, maximum aspiration rate at 36 (linear mode), maximum vacuum limit at 100 and a bottle height of approximately 33 inches above the level of the patient’s eye.

When pulling on cortex in a radial fashion, significant zonular stress can be created. A significant zonule stress reduction can be effected by capitalizing on our ability to expand the space between the anterior and posterior capsule at the equator of the lens. This is where the equatorial cortex is anchored. To remove the cortex without stressing the zonules, we initially engage the cortex by inserting the I/A tip into the cortex at the equator of the capsule with the aspiration port pointing upward. As the aspiration begins and the tip becomes occluded, we then push the phaco tip downward, toward the optic nerve. This pushes the posterior capsule down and away from the anterior capsule, widening the space between the anterior and posterior capsules. The equatorial cortex will virtually deliver itself without the need to pull radially, sparing significant zonular stress (Figure 14-26).

Management of the Posterior Capsule:

Vacuuming of the posterior capsule can be performed with a 0.3-mm I/A tip, maximum aspiration rate at 14 (linear mode), maximum vacuum limit at 40 and a bottle height of approximately 33 inches above the level of the patient’s eye, if zonular integrity is sufficient. If capsular striae enter the aspiration port and remain fixed and do not readily move as the capsule is rasped clean, the aspiration and movement should be stopped and the probe removed under irrigation only. It may be necessary to back-flush the aspiration line to free the capsule from the aspiration port.
If there is danger of capsular rupture or zonular dehiscence, it is better to leave the capsule somewhat clouded because this can be handled with a YAG laser later on. Intentionally leaving some capsule debris on the equatorial or central aspect of the capsule can provide a measure of support in very fragile capsules. The integrity of the capsule and zonular structures is the primary concern for proper implant fixation.

Capsule expansion rings can be effective in bridging the areas of weak zonules. We find them generally unnecessary, but they will become widely available in the future. Further experience may result in a greater role of these devices in PEX cataract surgery.

**IOL Implantation**

When considering which PCIOL to use, the implant material is important. We prefer silicone lenses. These lenses are relatively weightless and are inherently less stressful on the posterior capsule on insertion. Silicone lenses have been associated with a significantly lower degree of posterior capsular opacification. One study reported that the mean percentage area of posterior capsular opacification for hydrogel lenses was 63%; for PMMA, 46%; and for silicone, 17%. Less posterior capsule opacification was associated with less Nd:YAG laser capsulotomies.

When inserting a flexible silicone lens implant, care should be exerted to avoid the stress of inappropriate rotation of the implant as it is placed. As the implant unfolds, the lead haptic or the unfolding optic can cause traction on the capsule structures. In PEX cataract, it is especially important to prevent insertional haptic incarceration in the flaccid posterior capsule, which can literally rip the capsule and zonules off of their insertion points.

The technique is to insert the implant with only horizontal insertion force, negating all vertical forces. To accomplish this, we rotate the implant cartridge selectively in order to deliver the implant without any vertical insertion force. The implant is advanced far enough for the lead haptic to come to lie horizontally over the iris at the 2- to 3-o’clock position (down and to the left in the surgeon’s view) (Figure 14-27). This is accomplished by turning the inserter upside-down, rotated clockwise approximately 180 degrees. As the implant is further unfolded, it will leave the optic upside down unless an immediate 180 degrees counter-clockwise twist of the inserter is made to deliver the lens and the following haptic right side up, in the horizontal plane. If the iris is not used to hold the lead haptic during this maneuver, it is possible for the lead haptic to turn vertical and incarcerate itself in the fragile PEX capsule. The iris can securely hold the lead haptic in place and prevent it from twisting until the optic is correctly delivered horizontally. The lead haptic will advance to where it delivers itself into the capsule bag in most cases, or at least it will come to lie over the inferior iris, where it can be directly inserted in the bag (Figure 14-28). The implant is then manipulated until visual verification confirms that the lead haptic is in the capsular bag inferiorly.

To complete the insertion and place the following haptic in the bag, we use a bimanual insertion technique. Through the counterincision, the nondominant hand utilizes a Kuglen hook to grab the anterior capsule and lift it upward toward the incision site. A Bechert Rotator is then utilized to rotate the implant only slightly, while directing the implant downward, into the posterior capsule. There is little rotation, only enough to allow the haptic to slide over the bag. The essential force is downward toward the optic nerve. In this manner, the following haptic will deliver itself into the bag with little or no rotational stress on the zonules (Figure 14-29). A common mistake is to rely only on horizontal movements, pushing the implant far to the side. This can place severe stress upon the zonules and capsule structures. It is not necessary to actually visualize the implant going into the bag.
If the Kuglen hook holds the anterior capsular bag while the implant is being eased downwards with slight rotation, the haptic will almost invariably end up in the right place.

**Cleanup and Final Inspection**

The viscoelastic is removed from the anterior chamber with the I/A tip (same settings as aspirating cortex). If the capsular structures are exceptionally fragile, some Viscoat® may be left in the anterior chamber. If so, the patient can be given oral acetazolamide (Diamox, Lederle Pharmaceutical div. American Cyanamid Company, Pearl River, NY) 500 mg, which can be repeated 10 to 12 hours later.

Before terminating the surgery, a 25-gauge air needle with BSS irrigates the anterior chamber above the iris plane to wash out any loose viscoelastic material and to provide a small turbulence in the anterior chamber which can liberate any possible hidden fragments of nuclear or cortical material.

The placement of a small bolus of Viscoat under the inner aspect of the 3.2-mm incision assures a better initial seal and prevents leakage during the early postoperative period (Figure 14-30). Using the 30-gauge cannula, the anterior chamber is refilled with BSS to the proper tension. The epithelial site of the 3.2-mm incision is pressured with a cellulose spear and an adequate seal is confirmed (Figure 14-31). Before removing the lid speculum, a corneal shield (Surgilens, Bausch & Lomb Surgical, St. Louis, Mo) soaked in Gentamycin and/or Cefazolin solution can be placed over the cornea (Figure 14-32).

**Recovery**

Using the RSAP, patients are expected to be alert at the end of the surgery and are transferred to the recovery room by wheelchair. They are monitored for 30 minutes. A light snack is offered and instructions are given. The patient is discharged by the anesthesiologist and allowed to return home.

**Postoperative Management**

Postoperative management is very liberal. The patient can go home without a patch on the eye. A shield is worn for only 1 night. By the next day, patients are allowed to golf, play tennis, or return to work. Only restrictions against contamination such as avoiding rubbing or swimming are imposed.

If too much viscoelastic remains in the anterior chamber, intraocular pressure can become elevated. We can often anticipate this and pretreat with Acetazolamide. For the few patients in whom the pressure goes up unexpectedly, causing pain or discomfort, the patient can be brought back to the office. At the slit lamp, a small amount of fluid is allowed to escape by exerting a slight downward pressure on the scleral portion of the counterincision with a sterile disposable needle. This allows a safe and gradual reduction of pressure without rapid depressurization, and without violating the sterility of the incision.
Follow-up visits are scheduled on the next day after the procedure, and at the 1- and 3-week periods. Special attention is given to the possibility of elevated intraocular pressure and/or inflammation.

**COMPLICATIONS**

Complications associated with PEX syndrome can be medically or surgically treated. PEX glaucoma has already been discussed. We will review some complications of cataract surgery in patients with PEX syndrome.

Patients with PEX syndrome and cataracts tend to have poorly dilating pupils, synechiae and tenuously supported capsular bags. The combination of these findings significantly increases the intraoperative and postoperative risk in these patients.

In PEX syndrome, a shallow anterior chamber may be associated with zonular instability and the cataract surgeon should be aware of a higher risk of intraoperative complications. In eyes with pseudoexfoliation, an anterior chamber depth of less than 2.5 mm was associated with a risk of 13.4% for intraoperative complications compared to an overall incidence of intraoperative complications of 6.9% and an incidence of 2.8% for an anterior chamber depth of 2.5 mm or more.111

The odds ratio for intraoperative complications such as capsular tears, zonular break, and vitreous loss was estimated to be 5.1 for patients with PEX compared to normal patients. PEX was associated with a statistically significant increase in intraoperative complications during cataract surgery (p<0.0001).112

PCIOLs are susceptible to dislocation secondary to insufficient capsular or zonular support, or following trauma, in PEX syndrome (Figure 14-33). A dislocated PCIOL may be repositioned or removed and replaced with an appropriate IOL. Repositioning the dislocated PCIOL into the ciliary sulcus is generally considered the best option. There are many techniques to reposition and obtain adequate stability of the dislocated PCIOL including scleral fixation of the dislocated IOL. Endoscopy, though not always available, allows viewing of the retropropillary area and verification of precise haptic placement. Sometimes repositioning of a PCIOL cannot easily be accomplished. Removing it and replacing it with an ACIOL is an option that is frequently less traumatic and involves less risk, but elevated IOP may be more pervasive113 (Figure 14-34). Care must be taken during all steps of the phacoemulsiﬁcation procedure not to use excessive force which could easily rupture the zonule, cause a "dropped nucleus," or plainly lead to vitreous loss which is associated with a higher risk of endophthalmitis. In our practice, when vitreous is lost, broad spectrum oral antibiotics are prescribed to obtain adequate vitreous antimicrobial concentrations, such as ofloxacin (Floxin. Ortho-McNeil Pharmaceutical, Raritan, NJ) 400 mg twice daily by mouth for 1 week.
Even when the capsule is compromised, an IOL can be placed in-the-bag. Despite perfect placement, long-term difficulties can be encountered. Patients with PEX syndrome may have a higher risk for dislocation of endocapsular PCIOLs. A study reported a mean time from IOL implantation to dislocation of 85 months after surgery. They were treated with IOL exchange.114 In a patient with PEX, 12 years after cataract surgery, liberated lens cortical material after spontaneous dislocation of a PCIOL was associated with lens particle glaucoma in patients.115 Despite the association of PEX syndrome with subluxated in-the-bag IOLs after cataract extraction, risks can be reduced by not using a foldable IOL, using IOLs with larger optics, and early Nd:YAG anterior capsulotomy.116

Due to focal zonular lysis in PEX syndrome, the capsular bag may "shrink" in areas, making it difficult to place an implant in-the-bag. Implanting a CTR before phacoemulsification of the nucleus has been suggested as an appropriate method to reduce the risk of zonular separation. In some studies, it increased the rate of endocapsular IOL fixation, and improved postoperative UCVA.117 In patients with PEX syndrome that undergo phacoemulsification with continuous curvilinear capsulorrhexis, cataract extraction, and IOL implantation (even with an endocapsular ring), anterior capsule fibrosis with complete occlusion of the capsule opening (causing significant visual loss) can occur. After a Nd:YAG laser anterior capsulotomy, visual acuity can be restored. Endocapsular ring implantation does not prevent anterior capsule contraction syndrome but can prevent IOL decentration.118

Pressure spikes can easily occur, sometimes as a result of pupillary dilation liberating fibrillar material; therefore, IOP control is extremely important. Pressure should be monitored carefully in the immediate postoperative period. Over the long term, patients tend to do well. Six and 12 months after phacoemulsification with an IOL implant, patients with PEX syndrome have a greater postoperative IOP reduction than patients with POAG and cataract control groups.119 IOP decreased after phacoemulsification cataract surgery in the presence of PEX similarly as in normal eyes.120 If a more severe glaucoma is present, a combined clear cornea phacoemulsification, IOL implant, and trabecular aspiration in patients with PEX glaucoma may be a safe and effective way to control IOP with fewer postoperative medications than clear cornea phacoemulsification with IOL implant alone. A statistically significant decrease in postoperative IOP has been found.121 Considering IOP elevation in PEX glaucoma is due to obstruction of the intertrabecular spaces by exfoliation material, Jacob et al122 recommended bimanual trabecular aspiration with a 400 µm-in-diameter intraocular aspiration probe. Trabecular debris and pigment is cleared with a suction force of 100 to 200 mmHg under light tissue-instrument contact. Irrigation of the anterior chamber is performed via a separate irrigation cannula. There is a slight regression in effect over time, attributed to liberation of exfoliative debris.

Endothelial cell loss has been found in PEX syndrome patients. This coupled with the poor lens support, and possibly elevated IOP may lead to an increased risk of corneal decompensation. Concomitant Fuch’s dystrophy, also more commonly found in the elderly population (as is PEX syndrome) may compound the loss of endothelial cells and lead to pseudophakic bullous keratopathy.

Phacoemulsification cataract surgery is considered safe for most eyes with PEX, even though significantly more complications, such as capsular/zonular tear or vitreous loss may occur intraoperatively. Also, there may be an increased inflammatory response postoperatively, associated with increased flare in the aqueous humor.123 This finding may suggest a reason for adding an NSAID to the postoperative regimen of steroids to reduce the potentially increased incidence of CME.
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