A 41-year-old man with congenital glaucoma has had several repeated filtering procedures in both eyes. After a perforating injury that occurred when the patient was 10 years old, the right eye became phthisic and was removed. In the left eye, intraocular pressure (IOP) was successfully regulated. During the past years, the remaining left eye developed nuclear cataract; however, surgery was delayed because of risk considerations. Within months, the visual impairment became disabling and the patient was referred for cataract surgery.

The patient presented with a buphthalmic left eye (Figure 1). A basal coloboma with a scarred filtering bleb was visible superiorly, and a prominent avascular and cystic filtering bleb was visible in the superior-temporal quadrant (Figure 1, top right). The IOP was 8 mm Hg. The cornea had Descemet tears (Haab striae) and diffuse stromal edema (Figure 1, top left), which significantly obscured detailed visualization of the anterior segment, even under oblique slitlamp illumination. Medical dilation of the small pupil was moderate. The eye had a dense brunescent nuclear cataract (Figure 1, bottom). A small translucent mass dusted with pigment granules, which appeared to represent localized vitreous prolapse, was noted in the pupillary plane, suggesting possible zonular deficiency. However, no lentodonesis was detected, even with gentle knocking on the retrolimbal area. No red light was reflected from the retina. Ultrasonography showed an attached retina, a large excavation of the papilla, and a highly reflective retrolental membrane of unknown nature that was in contact with the opacified lens. Axial length was 31.0 mm. Keratometry of the swollen cornea was approximately 33.00 diopters (D). Visual acuity was 1/30 with a refraction of approximately –11.00 D. In the months before referral for surgery, the patient had been able to perform computer work.

With the patient was positioned under the operating microscope, visualization was judged to be inadequate to allow controlled anterior segment surgery due to the light scatter from the stromal edema of the cornea.

Considering the unclear but obviously complicated anatomy and the difficulties in visualization, what preoperative and intraoperative approach would you use for this patient’s only eye?

This patient has problems usually encountered in old age in an only eye. Echography shows an attached retina, which provides the reason to proceed with surgery despite the unclear anatomy. The patient is young, so general anesthesia should be used and would be preferred.

The first goal is to improve visibility. Descemet-stripping endothelial keratoplasty (DSAEK) should be attempted, keeping in mind that any surgery under current conditions will cause further corneal decompensation. Surgery can be performed in the usual way (by removing the recipient endothelium) or if endothelial removal is impossible because of poor visualization or anatomic distortion, by adding the donor lamella. The cornea should improve in 2 to 3 months, after which I would reevaluate the eye for cataract surgery, which is feasible in eyes that have had DSAEK.

The subsequent cataract surgery must take into account the hardness of the cataract, the presence of
the translucent mass, and the poor endothelial function as negative factors and the absence of lentodonesis as the only positive factor. Two 1.0 mm incisions are enough to attempt pupil dilation with 1:100,000 epinephrine and to test the nature of the small translucent mass, which would eventually be removed by careful anterior vitrectomy. Use of a highly adhesive ophthalmic viscosurgical device (OVD) is mandatory.

If the lens is not too hard, coaxial microincision phacoemulsification is probably the best option. The small instruments have advantages in eyes with a narrow pupil, and anterior chamber stability is much better with phaco machines that do not require high irrigation. A small capsulorhexis should be created to save the peripheral capsule for IOL support. If the lens is too hard for phacoemulsification, extracapsular surgery with an inferotemporal approach and surgical dilation of the pupil should be planned. Lens expression should be avoided in favor of OVD-aided lens extraction. This type of surgery allows better visualization of the anterior segment anatomy, although it can lead to late bleb failure.

During or after lens removal, the posterior capsule must be evaluated to determine the nature and optical consequences of the membrane seen on echography. If no vascularization is present, the posterior capsule and the membrane can be transfixed with a sclerotomy and vitrectomy used to create a round hole approximately 5.0 mm in diameter.

The biometric calculations anticipate an aphakic refraction of approximately +8.00 D and probably more hyperopic refraction after DSAEK. Therefore, all attempts should be made to implant an intraocular lens (IOL). With good capsule support, even under the best conditions, in-the-bag IOL implantation would likely be impossible; therefore, I would select a hydrophobic acrylic IOL designed for sulcus implantation (ie, 13.0 mm long). If capsule support is not suitable for sulcus implantation, I would select a 3-piece posterior chamber IOL (PC IOL).

If extracapsular surgery were performed, I would suture the iris to avoid postoperative diplopia and carefully close the corneoscleral wound to avoid aqueous leakage that could impair bleb function.

The patient should be told of the possibility of early or late corneal decompensation with the need for repeat DSAEK. He is only 41 years old, and it is likely these will not be the last surgical procedures in his only eye.

Roberto Bellucci, MD
Verona, Italy

In this complicated case, a preoperative endothelial cell count would help determine whether cataract extraction and IOL implantation should be combined with penetrating keratoplasty (PKP). In any case, a topical nonsteroidal antiinflammatory agent should

Stephen Kohnen, MD
Aachen, Germany
be administered 3 days preoperatively to decrease the chance for cystoid macular edema.

If only cataract surgery is performed, I would prefer phacoemulsification. Preoperatively, I would use topical glycercine as a dehydrating agent to clear the cornea and then administer topical anesthesia. The phaco incision should be temporal and corneal to avoid the cystic bleb site. First, I would inject an OVD through a paracentesis to increase IOP, facilitating creation of the main incision. I would use a dispersive OVD to protect the corneal endothelium followed by a cohesive OVD to form the anterior chamber, dilate the pupil, and facilitate capsulorhexis. To widen the pupil further, I would perform multiple sphincterotomies using microscissors. The anterior lens capsule would be stained with trypan blue under an OVD cushion. I would use Vannas scissors to trim the prolapsed vitreous knuckle flush with the iris to avoid traction on the vitreous. Next, I would create a capsulorhexis using a fine forceps, guiding the capsulorhexis within the limits of the pupil. I would not use hydrodissection because the fluid wave would not be visible, which would increase the risk for capsular block syndrome.

For nucleus removal, I would use low energy and fluidics, exerting minimum stress on the zonules and working in the capsular bag. Although the patient is 41 years old, the lens color and density indicate a very hard cataract. Phaco machine settings would be vacuum 100 mm Hg for sculpting and 300 mm Hg for chopping; aspiration rate 30 cc/min for all stages; bottle height 65 to 75 cm. I would use a flared, 45-degree angled Kelman tip, and 100% continuous torsional ultrasound with a minimum of 60% power. To emulsify the nucleus, I would use a 2-nuclei concept—a combined debulking and drilling technique. First, the superficial one third of the nucleus is shaved off to debulk the nucleus. A wide groove is made in the remaining part of the nucleus down to the deepest fibers to crack the nucleus; 5 to 6 holes are drilled in the remaining nuclear shell to weaken it. Moving to stage 2, the phaco tip is buried between the drilled holes and, with a Neuhan nucleus dividing hook, the nucleus is split into smaller pieces that can be removed with less energy and without stress on the zonules. I would use intermittent phaco bursts to avoid corneal wound burn. A dispersive OVD could be repeatedly injected during emulsification for added corneal protection.

After cortical aspiration, the bag would be inflated with a cohesive OVD. A foldable IOL would then be implanted in the capsular bag after the integrity of the remaining zonules was checked and a capsular tension ring (CTR) inserted, if necessary. The corneal wound would be closed with a single 10-0 nylon suture to prevent postoperative hypotony and wound leak.

Yehia M.S. Mostafa
Cairo, Egypt

- The best option is microincision cataract surgery with DSAEK. The technique would comprise topical or sub-Tenon anesthesia, epithelial removal to improve anterior segment visualization, cataract emulsification through a 1.7 mm scleral microincision placed temporally 1.0 mm behind the limbus (far from previous filtering surgeries), anterior vitrectomy to prevent further vitreous traction and postoperative filtering bleb occlusion, a 5.0 mm capsulorrhexis, CTR implantation to prevent vitreous prolapse, phacoemulsification, in-the-bag implantation of a 3-piece C-loop hydrophobic IOL for stability, Descemet membrane stripping, and endothelial–stromal graft introduction through an enlarged cataract incision.

The approach has several surgical risks. Scleral perforation might occur with sub-Tenon anesthesia. If retrobulbar anesthesia were required in this buphthalmic eye, it could thin the ocular tissue (especially the sclera) and increase IOP, endangering the glaucomatous optic nerve. Thus, I would use general anesthesia to avoid posterior pressure and eye movement.

In addition, poor visualization of anterior segment alterations and a possible zonular defect increase the risk for dropped nucleus. Thus, I would consider open-sky cataract surgery followed by PKP, which would provide excellent intraoperative visualization and prevent anterior–posterior pressure on the crystalline lens and thus the risk for posterior lens luxation into the vitreous cavity. Lens luxation would jeopardize the visual prognosis because it would require complicated posterior maneuvers in the vitreous cavity.

The efficiency of previous filtrating surgeries would be threatened by the scleral incision and conjunctival peritomy required for DSAEK and by prolonged postoperative steroid therapy. The main threat is keratoplasty (especially PKP) because of possible synechia formation; however, the risk is probably low here because of the deep anterior chamber, large corneal diameter, and wide iridocorneal angulation.

The vitreous in the anterior chamber, which probably reflects a zonular defect, is another risk factor. The absence of crystalline lens movement does not guarantee stability. The vitreous is plugging the zonular defect, and lentodonesis may present when the vitreous cushion is removed by vitrectomy. This is the main argument for an open-sky technique.
The highly reflective retrolental membrane might indicate a posterior polar cataract, which risks posterior capsule opening and dropped nucleus and would require treatment (eg, posterior capsulorhexis and vitrectomy) before IOL implantation. Again, good surgical visualization would be required for these maneuvers.

Considering these factors, I would perform combined PKP and open-sky cataract extraction, adapting the graft to the corneal diameter and using iris stretching or iris retractors, if needed. The technique would comprise corneal trephination under a cohesive OVD, anterior vitrectomy with low vacuum, a 5.0 mm open-sky capsulorhexis assisted with blue dye, hydrodissection to ease nucleus delivery out of the bag without exerting posterior pressure, CTR insertion with the capsular bag filled with cohesive OVD, and removal of the remaining cortex with a double-flow cannula. I would examine the posterior capsule, in particular the dense posterior opacity. If the opacity were a posterior polar cataract remnant, I would carefully remove it. I would perform a posterior capsulorhexis under dispersive OVD followed by dry vitrectomy for adequate in-the-bag implantation of a 3-piece hydrophobic IOL. Finally, I would suture the donor cornea under cohesive OVD.

Dominique Pietrini, MD
Paris, France

■ I would recommend the following 2-step procedure: Remove the cataract without implanting an IOL; for better visualization, remove the corneal epithelium and carefully dilate the pupil with iris retractors or a distension ring; remove the cataract by phacoemulsification using a temporal approach to avoid the filtering bleb; stain the capsule with trypan blue and create a capsulorhexis using a capsulorhexis forceps for better control; and fill the anterior chamber with a dispersive OVD. Because of the reduced visualization through hazy cornea, the capsulorhexis should have a large diameter to prevent injuring the anterior capsule rim during phacoemulsification. (If phacoemulsification were not feasible [nucleus too hard, visualization inadequate, procedure too dangerous with high risk for posterior capsule rupture], extracapsular cataract extraction could be performed.) The next step would be to lower the infusion bottle to limit flow and pressure in the anterior chamber to the minimum required for phacoemulsification to avoid overinflating the filtering bleb, risking rupture. Intracapsular cataract extraction should be avoided in patients with 1 remaining eye because of the high risk for posterior segment complications (eg, intraoperative choroidal hemorrhage, postoperative retinal detachment). If phacoemulsification were successful, all remaining cortex material must be carefully removed using low IOP and a bimanual technique for best access to the capsular bag fornix. Because of the large globe, the postoperative refractive error should be minimized by removing only the opacified crystalline lens and not implanting an IOL because of the higher risk for postoperative problems (impaired fundus visualization) and difficult IOL positioning due to poor visibility.

If the cornea is permanently opacified postoperatively, a second surgery should be planned. I would recommend DSAEK because only Descemet membrane and endothelium are exchanged, leaving the corneal stroma and Bowman membrane untouched. Because of the buphthalmic cornea, only the central cornea can be treated. A large transplant (>9.0 mm) should be used. The technique would comprise removing the central Descemet membrane; filling the anterior chamber with air; preparing the corneal graft with the artificial anterior chamber of an automated lamellar therapeutic keratoplasty system, cutting and then rejecting a 350 μm flap; using a trephine to punch out the deep cornea with some stroma, Descemet membrane, and endothelium; coating the endothelium with a dispersive OVD; introducing the IOL into the anterior chamber through a 3.5 mm clear corneal incision using a 10-0 nylon traction suture; centering the transplant with the anterior chamber filled with air; and closing the wounds with nylon sutures to maintain stable pressure. The air would remain for 15 minutes, after which the anterior chamber would be filled with a balanced salt solution with little air remaining. The pupil would be dilated; the patient would remain supine for 1 hour. I would not implant an IOL as a third step because it would be in the patient’s only eye. Because of the large buphthalmic eye, the postoperative refractive error would be modest. Spectacles would be my first choice to correct the aphakia.

Later, if visual acuity significantly worsens, neodymium:YAG laser capsulotomy could be performed, although it may increase the risk for posterior segment complications.

Christian Skorpik, MD
Vienna, Austria

■ A case with congenital glaucoma, corneal decompensation, vitreous prolapse, narrow pupil, and dense brunescent cataract is a challenge for every anterior segment surgeon. The fastest rehabilitation would be
a combined operation including perforating keratoplasty, anterior vitrectomy, cataract removal, and implantation of a PC IOL, if the zonules are strong enough. The disadvantage of this procedure is rather long-lasting intraoperative hypotony with the risk for choroidal hemorrhage or severe vitreous loss if the zonular apparatus is weak.

Considering that the patient is monocular, the safest course of action would be a 2-step procedure. The first step would be keratoplasty and removal of prolapsed vitreous with a microstripper. Then, after 3 to 6 months, when the cornea allows good visualization, phacoemulsification could be performed with the help of iris hooks to dilate the pupil. The technique would include posterior capsulorhexis, removal of the retrolental membrane with the vitreous stripper, and implantation of a PC IOL. If the zonules were weak, I would use a CTR and, if necessary, suture it transsclerally with 10-0 polypropylene. A good option would be a Cionni CTR, which was designed for transscleral fixation. I would avoid implanting an anterior chamber IOL (AC IOL) because in highly myopic eyes, there is an increased risk for retinal complications. With an AC IOL in the eye, subsequent gas or silicone-oil tamponade would be impossible.

Andreas Forrer, MD
Aarau, Switzerland

This case represents an interesting treatment dilemma. Two approaches can be considered in this relatively young patient. One is simultaneous cataract extraction, PKP, and IOL implantation (triple procedure). If possible, a continuous capsulorhexis should be created and the IOL placed in the capsular bag. Phacoemulsification must be used if the nucleus appears to be too large to pass through the capsulorhexis. Accurate prediction of IOL power is difficult in such a case because of the imprecision of keratometry, AL, and anterior chamber depth measurements after PKP. I would perform this complex procedure using general anesthesia. The advantages of this option are that it would require only a single surgical treatment and patient rehabilitation may be faster. However, the case states that in the “months before referral for surgery, the patient had been able to perform computer work,” which suggests that PKP may not be needed and that cataract surgery alone may be sufficient to significantly improve the visual function.

The second option would be phacoemulsification with IOL implantation alone. However, visualization under the operating microscope appears to be inadequate to allow controlled anterior segment surgery. Even so, and with careful evaluation of the anterior segment photograph, I believe that phacoemulsification with foldable IOL implantation would be possible. Use of trypan blue and transcorneal illumination with a fiber-optic light probe should provide adequate visualization during surgery. A PKP could be considered later if the visual outcomes of the cataract surgery were inadequate.

With both options, removal of the retrolental membrane by pars plana vitrectomy should be postponed until better visualization is achieved. Neither surgical treatment can be regarded as safe in this case. If I had to choose, I would begin the treatment with option 2; that is, cataract surgery with IOL implantation alone.

Wojciech Omulecki, MD, PhD
Lodz, Poland