Management of the subluxated crystalline lens

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The surgical management of ectopia lentis presents the ophthalmic surgeon with numerous challenges and options. From the clinical evaluation to the surgical approach, ectopia lentis patients require additional methodologies, techniques, and devices to ensure the best possible outcome. The continued refinement of surgical techniques and adjunctive prosthetic devices has led to incremental improvements in the ability to achieve successful in-the-bag placement and centration of intraocular lenses while reducing complications. A thorough understanding of the challenges inherent in ectopia lentis cases and the management of intraoperative complications will ensure that surgeons approaching the correction of these eyes will achieve the best possible surgical results.

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The surgical management of ectopia lentis is one of the more challenging situations an anterior segment surgeon may encounter. Ectopia lentis encompasses any displacement or malposition of the crystalline lens irrespective of the cause or association. Lens subluxation may occur from congenital or developmental conditions such as Marfan syndrome, homocystinuria, Ehlers-Danlos syndrome, hyperlysinemia, sulfite oxidase deficiency, simple primary ectopia lentis, and congenital aniridia syndrome among others.1,2 Subluxation may also be acquired due to blunt external trauma or iatrogenic zonular dehiscence induced during complicated cataract surgery. Perhaps the most common cause for adult-onset zonular weakness is that which occurs in pseudoexfoliation syndrome (PXF), in which progressive zonular degradation can result in phacodonesis and crystalline lens subluxation, in addition to late postsurgical pseudophacodonesis and intraocular lens (IOL) dislocation.3–5 Retinitis pigmentosa can result in a similar progressive zonulopathy.

Historically, when intracapsular cataract extraction was the preferred approach to both routine cataract surgery and surgical repair of the malpositioned crystalline lens, the technique for lens extraction was the same for all but the most severely dislocated lenses. However, with the advent of extracapsular cataract extraction and IOLs, techniques and approaches for ectopia lentis that are distinct and different from the typical routine cataract extraction have evolved. The ability to manage the patient with preexisting zonular weakness or lens subluxation, as well as the patient with unexpected intraoperative zonular compromise, has become an important part of the cataract surgeon’s knowledge. The goals of this paper are to review the clinical evaluation of these patients and to summarize various techniques, devices, and methodologies that are relevant to the current surgical management of ectopia lentis.

CLINICAL EVALUATION

The clinical evaluation of the ectopia lentis patient begins with an appropriate history, which should include family history, relevant trauma, and onset and types of visual symptoms. Some metabolic and genetic conditions are associated with ectopia lentis and may run in families with different types of inheritance patterns. Because certain genetic syndromes, such as Marfan syndrome, are associated with significant systemic medical problems involving other body systems, such as the cardiovascular system, a referral
to a primary care physician for a full systemic and metabolic workup is recommended.

The symptoms may include moderately blurred vision from induced lenticular astigmatism when the crystalline lens decenters to a severe decrease in vision if the crystalline lens is subluxated out of the visual axis. Because trauma is a frequent cause of ectopia lentis, patients should be questioned about a history of ocular trauma. Patient age may lead the ophthalmologist to suspect different diagnoses; pediatric patients with ectopia lentis are more likely to suffer from metabolic syndromes, whereas adult patients are more likely to present with Marfan syndrome.

**Ophthalmic Examination**

The ophthalmic examination should be comprehensive and assess the anterior and posterior segments. The degree of lens dislocation can be classified into 3 broad groups (Figure 1): (1) Minimal to mild lens subluxation in which the lens edge uncovers 0% to 25% of the dilated pupil; (2) moderate lens subluxation in which the lens edge uncovers 25% to 50% of the dilated pupil; (3) and severe lens subluxation in which the lens edge uncovers greater than 50% of the pupil.

The degree of zonular loss can be localized or extensive depending on the condition. Whereas conditions such as focal trauma or congenital defects may produce localized zonular compromise, systemic conditions such as Marfan syndrome or ocular conditions such as PXF can cause global weakness to the zonular fibers. The zonular status will determine the degree of phacodonesis that can be seen at the slitlamp during examination. The zonular fibers may be stretched or absent, and evaluation of the subluxated lens edge to determine whether it is round or flattened may indicate the status of the remaining fibers. Generally, if the lens edge is flat, the remaining zonular fibers should be fairly healthy and strong (Figure 2). If the edge of the lens remains very round in an area of absent or stretched zonular fibers, which is often a sign that the adjacent intact fibers may not have sufficient strength, the surgeon may be wise to select capsule retractors to augment structural support.

Comparing the position of the crystalline lens with the patient seated at the slitlamp and in the supine position can also help detect the degree of zonular compromise and is important in planning the surgical approach. In patients without zonular issues, there should be no change in the crystalline lens position with a change in head position. With zonular compromise, a deepening of the anterior chamber with posterior dislocation of the crystalline lens is seen when the patient is placed in the supine position. Rarely, lenses may appear approachable with the patient in an upright position but subluxate into a position better managed with a pars plana lensectomy when the patient is placed supine; thus, the importance of

![Figure 1. Mild, moderate, and severe crystalline lens subluxation.](image1)

![Figure 2. Flattened subluxated lens edge suggestive of healthy adjacent lens zonular fibers.](image2)
both an upright and a supine evaluation. If available, ultrasonic biomicroscopy (UBM) is a valuable tool in assessing the degree of zonular compromise. Since the UBM images are recorded with the patient in the supine position, information gathered will simulate the condition of the crystalline lens during the surgical procedure.

Particularly in cases of trauma, the lens capsule should be carefully examined for evidence of damage or puncture. Focal areas of lens capsule damage can pose additional challenges during planned cataract surgery.

Increased lens density can make cataract surgery more challenging, as can the presence of vitreous in the anterior chamber. When vitreous prolapses forward around the area of zonular loss, it may temporarily stabilize the crystalline lens. During cataract surgery, a partial vitrectomy will be necessary to address this prolapse.

Increased intraocular pressure may be seen in cases in which vitreous occludes the angle of the eye, in PXF, or when previous trauma that ruptured zonular fibers also caused angle trauma and recession. Similarly, prior inflammation, trauma, and vitreous prolapse can lead to corneal compromise and if that is suspected, an endothelial cell count should be performed.

Finally, the posterior segment of the eye should be carefully examined. Any evidence of retinal tears, breaks, or tufts should be sent for retinal consultation and treatment prior to performing the elective cataract surgery.

Surgical Approach
Planning

The surgical approach to phacoemulsification of the subluxated crystalline lens differs depending on the degree of zonulopathy and the underlying pathophysiologic origin. In cases in which the zonular abnormality is less than 3 contiguous clock hours, attentive surgical technique may suffice for a successful surgical outcome, although capsule retractors may provide additional support. If a posttraumatic eye has a small focal area of dialysis with an otherwise strong adjacent zonular apparatus, a capsular tension ring (CTR) may not be required. In cases of even mild zonular laxity but with a progressive pathologic state such as PXF, Weil-Marchesani, Marfan syndrome, sulfite oxidase deficiency, retinitis pigmentosa, or the like, the zonular problems can be expected to worsen over time and a CTR should be placed, if only to facilitate possible future refixation. In a young patient with such progressive diseases, a sutured CTR with scleral fixation might be prudent from the outset, even in the absence of lens displacement, although this would be up to the judgment of the individual surgeon. The relative degree of zonulopathy may not be readily apparent at the beginning of a case. Thus, surgeons who choose to tackle these cases should be prepared for greater damage than is readily apparent at the outset of the procedure.

Capsulorhexis

A complete capsulorhexis is paramount to successful phacoemulsification and bag preservation in any ectopia lentis case. The capsulorhexis in these eyes is more challenging than in a standard case because special considerations are required for the initiation and completion. First, puncturing the anterior capsule is not always easy when zonular counter traction is compromised and in young eyes in which the lens capsule is highly elastic. The use of trypan-blue dye reduces the elasticity of the capsule and makes penetration of the bag and propagation of the capsulorhexis easier, especially in young eyes. The blue staining also aids in visualization, which may be hampered in these eyes despite limited nuclear sclerosis, and in avoiding unintended capsulorhexis contact and compromise during the case.

The lack of an anteroposterior barrier in zonulopathy cases can lead to a loss of the red reflex following simple irrigation of trypan blue into the anterior chamber. For this reason, it is safer to paint a few drops of the dye directly across the anterior capsule in an ophthalmic viscosurgical device (OVD)-filled anterior chamber. The capsule can be penetrated with a standard cystotome, a microvitreoretinal blade, or a straight 25-gauge needle. If the capsule does not penetrate easily, the crossed-swords capsule pinch approach using 2 opposing 30-gauge needle tips can be used to pierce the capsule and create a starting point for the capsulorhexis (Figure 3). In the most unstable lenses, a microforceps or hook may be needed to steady the lens during the continuous tear.

The capsulorhexis should be centered on the crystalline lens, not on the pupil or the corneal apex. In the case of a misshapen lens, the shape of the capsulorhexis should follow the outer contour of the lens, whether oval, round, or kidney-bean shaped. The capsulorhexis size should take into consideration the need for at least a 2.0 mm margin between the capsulorhexis edge and the equator, since a generous anterior leaflet is necessary to keep the CTR in the bag when it is under tension. This is particularly crucial when an Ahmed segment is used since the segment’s suture creates a rotational moment arm of force anteriorly around the bag equator at its axis. Execution of the capsulorhexis is often most facile when the tear starts in a direction pulling away from the area of greatest...
zonular counter traction toward the weakened zonular fibers (Figure 4). Microincision forceps can be extremely useful in accessing the capsule from different paracentesis microincisions as this process advances around the circumference. In addition, these forceps are less likely to result in loss of OVD from the anterior chamber, which can develop with a standard capsulorhexis forceps through the relatively large main incision. This loss of OVD can result in progressive anterior chamber shallowing and incremental anterior lens movement that can complicate completion of the capsulorhexis.

Femtosecond laser–assisted cataract surgery increased in 2012 and 2013 and may offer advantages in complex cataracts such as traumatic cataracts8 and less severe cases of subluxated cataracts. The femtosecond laser does not depend on counter resistance from zonular support and is able to cut a circular anterior capsule opening despite subluxation of the lens as long as the lens is not tilted excessively after docking with the patient interface. A further benefit of femtosecond laser–assisted cataract surgery is the creation of gas bubbles in and around the nucleus during nuclear femtosecond fragmentation, leading to pneumodissection, which facilitates gentle nuclear rotation with little or no hydrodissection. Not every subluxated crystalline lens lends itself to this type of surgery; some lenses are too grossly subluxated, some may have poorly dilating pupils, and some may have a mobile crystalline lens. These represent contraindications to successful femtosecond laser–assisted phaco surgery. The ideal case is the patient with PXF or traumatic cataract in which mild phacodonesis is noted and in whom the cataract is primarily in the anatomical position.

Stabilization of the Capsular Bag

In cases of moderate zonular loss or dysfunction in the 3-to-6 clock hour range, some form of augmented capsule support will likely be needed. Flexible iris retractors placed through limbal stab incisions can be used to hook the capsulorhexis edge and support the bag.9 These work reasonably well, although there is a small chance that the hook may inadvertently tear the capsulorhexis margin. Capsule hooks, in contrast, support the bag by its equator, not the capsule margin, thereby keeping the bag distended and also reducing the likelihood of aspiration of the bag equator as the lens material is evacuated.10 In either case, when tightening the hooks, enough tension is placed to stabilize the bag, but efforts to completely recenter the bag by hook alone should not be made at this time as they may compromise the opposing healthy zonular fibers or place undue stress on the capsulorhexis during phacoemulsification. In addition, when using hooks as counter traction during capsulorhexis creation, the hooks should be placed at least 2 to 3 clock hours from the leading edge of the capsulorhexis to avoid...
tractional forces that will cause the leading edge to extend peripherally toward the bag equator (Figure 5).

A further option to stabilize the bag during phacoemulsification would be placement of an Ahmed segment. When the segment is used early in a case, OVD is placed under the anterior capsule leaflet and the cortex is pushed back, up to the equator in the quadrant of interest. The segment is then slipped into the bag fornix, leaving the fixation element anterior to the capsulorhexis. Unlike its use when the bag has been evacuated, when the segment is used before phaco, no suture is placed and the segment is stabilized by placing a flexible iris retractor through the limbus, pointing the hook’s fixation end upward, then engaging it through the Ahmed segment fixation eyelet and tightening to stabilize the bag (Figure 6). For surgeons outside the United States, the Assi Anchor and Yaguchi hook are other viable options that are smaller in profile and may be easier to insert.

When the zonular dialysis is severe, for example more than 6 clock hours, some sort of bag stabilization with hooks, retractors, or segments for phaco is nearly mandatory for safe evacuation of the capsular bag, except perhaps in very young patients in whom the bag can be evacuated by simple aspiration, sometimes even dry aspiration using a 27-gauge cannula in an OVD-filled anterior segment. Some surgeons may prefer to place a CTR early in the procedure. This approach maintains an expanded capsular bag, supports the zonular apparatus, and may facilitate phacoemulsification. However, placing the CTR (either standard or fixated) early can be much more challenging because the lens within the bag may impede the CTR progression and may strip zonular fibers. Furthermore, the CTR may trap cortical material in the equator, making subsequent removal difficult or impossible. One dictum that is followed by many surgeons is to place the CTR “as late as you can, but as soon as you must.”

**Phacoemulsification of the Loose Lens**

With a stabilized bag and pressurized OVD-filled anterior segment, the next step is evacuation of the cataractous (or misshapen) lens. Hydrodissection and viscodissection are critical tools in managing loose lens cases and should be performed in all eyes, sometimes repeatedly. In the presence of zonulopathy, with an absence of counter traction to mobilize the lens nucleus, inadequate hydrodissection or viscodissection may endanger the remaining zonular fibers and even the bag itself.

The youngest lenses can be aspirated with irrigation/aspiration (I/A) handpieces or a cannula. A 27-gauge cannula affords a tremendous degree of safety and control. If vitreous is contacted, it can be readily refluxed with minimal retinal traction. Use of automated equipment requires vigilant attentiveness. Replenishing dispersive OVD volume into the capsular fornix throughout the case maintains bag anatomy as lens material is evacuated and increases...
safety. Periodic augmentation of a dispersive OVD plug over the exposed hyaloid face will also reduce the risk for vitreous prolapse during the procedure and diminish the chances of a nuclear fragment migrating posteriorly.

Phacoemulsification within the capsular bag reduces the likelihood of errant fragments finding the posterior segment but, concomitantly, increases the odds of inadvertent bag or zonule damage by the phaco or I/A handpiece manipulations. In contrast, it is easier to emulsify the lens within the anterior chamber, but turbulence is higher and control of fragments is lower. The soft lens can usually be easily prolapsed anteriorly and because of its sticky nature is unlikely to fall into the vitreous cavity. The denser nuclei are usually hard to get out of the bag and have a greater chance of dislocating. Accordingly, sometimes the lens will dictate what it “wants” to do and the surgeon will follow suit. Lower aspiration and flow parameters, whether limited by the machine panel or by the surgeon’s judicious use of the foot pedal, will reduce turbulence, maintaining greater control over fragments of the lens between liberation from the bag and aspiration by the phaco needle.

Cortical aspiration, achieved by an automated coaxial device, bimanual aspiration, or manual dry aspiration should limit stress to the remaining zonular fibers. Stripping from the capsulorhexis margin toward the periphery minimizes strain, while centripetal maneuvers maximize tension. Tangential stripping is intermediate and is very useful in meridians distant from the dehiscence but should be used with cautious direct observation in areas adjacent to the break to prevent “unzipping” of intact zonular fibers at a dehiscence edge. Tangential stripping should always be performed toward the area of missing or weak zonular fibers to limit stress on the adjacent fibers (Figure 7). While complete cortical removal is a noble and appropriate goal, excessive efforts to remove small strands should not risk capsular or zonular damage.

### Capsular Tension Ring Selection

Deciding which CTR to use depends on the degree of and likelihood for progression of damage. When the dialysis is small, a standard CTR is usually adequate. A standard CTR can be placed by injector or manual insertion and should be inserted to direct the vector forces toward the area of greatest zonular weakness. If there is more than 4 clock hours of damage or if the lens is moderately to severely displaced, a fixation device is required. One option for fixation involves the use of a standard CTR and a ring segment in combination. This disassembled fixation ring offers the advantage of simple insertion of the CTR and fixation element with less zonular stress than would be produced with insertion of a Cionni CTR. If the capsular rim is less generous than anticipated with a larger than planned capsulorhexis, a Cionni CTR is a superior option to a ring segment. In settings in which

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**Figure 7.** Tangential aspiration of cortical material toward areas of zonular weakness.

**Figure 8.** Injection of Cionni CTR using an injector.
cost is a consideration, the total cost of a standard CTR and separate ring segment may also be higher than a Cionni CTR alone. When using a Cionni CTR, the smallest (1-G) version is most facile for all but the largest capsular bags.

**Long-Term Fixation**

Ultimately, when the crystalline lens has been removed, some mechanism of suture fixation for long-term stability is needed in any case with more than 4 clock hours of involvement. Obviously, the hooks and retractors used during the procedure cannot serve this purpose. Long-term stability can be achieved using a Cionni-style CTR (Figure 8), an Ahmed segment (Figure 9), an Assi Anchor, or in cases of severe weakness or progressive disease in younger patients, any combination of those devices. The double-fixated Cionni CTR is somewhat cumbersome to place, and with the advent of the Ahmed segment, most find it more facile to place 1 Cionni CTR and 1 segment or 2 segments and a standard CTR. Some surgeons try to preserve the capsular bag whenever possible, even with heroic measures, while others may prefer direct suture fixation of the IOL to the iris or ciliary sulcus.

Placement of Cionni CTRs in the bag should be performed in a clockwise direction. Preloading the leading eyelet with a 10-0 nylon suture makes placement around the first few clock hours much easier, adding tension to the leading suture to reduce the arc of curvature and prevent the device from hanging up on the capsular fornix (Figure 10). A microforceps grasping the nylon can also lead the element into the bag like a leash. A fixation suture through the fixation element eyelet should similarly be preloaded.

An Ahmed segment can be placed directly into the bag fornix in the desired clock hour. In any case, the fixation element should course anteriorly around the capsulorhexis margin. When a polytetrafluoroethylene (Gore-Tex) suture is used, the white suture will appear blue under a trypan blue-stained capsule, indicating that the element remains under the anterior capsule and in the bag, a definite sign of a need for repositioning that element. Once a segment has been effectively placed within the capsular bag, the fixation element is rotated to the area of greatest weakness. Some surgeons prefer ab interno placement of the sutures through the scleral wall, while others prefer to create an exterior opening in the sclera, place the blunt end of the fixation suture into the anterior chamber, and retrieve the suture with a microforceps placed ab externo through the scleral wall.

Once both sutures are externalized, a throw can be placed and the IOL placed in the capsular bag. With the globe pressurized, suture tension should be titrated to achieve maximal IOL centration. Locking throws can then be placed and the knot secured. Whether the sutures are tied under a corneoscleral pocket, tied under a scleral flap, or tied over episclera is a matter of surgeon preference, as no technique has been definitively proven superior over the
others. However, there is a general consensus that exposed subconjunctival knots should be avoided to prevent overlying conjunctival erosion and subsequent endophthalmitis.

**Suture Material**

Suture material should be permanent. Polypropylene 10-0 is unadvisable, as it will hydrolyze over time with a roughly 5- to 10-year survival time. Polypropylene 9-0 should have a longer survival time; however, to date, the interval before degradation of this suture gauge has not been reported. A polytetrafluoroethylene CV-8 suture has been used for scleral fixation off label and to date has had excellent longevity.

In cases with externally tied knots, rotation of the knot through 1 scleral opening into the eye wall is recommended and can be performed while holding the fixation eyelet of the ring/segment with a 23-gauge microforceps to avoid decentration of the capsular bag in the process. When this cannot be achieved, a patch graft is advisable.

While many options of patch material exist, costs and access can vary regionally. Split thickness cornea has the least likelihood of melt over time, does not alter cosmesis, and is reasonably easy to work with. Pericardial patch material is very easy to suture and can be more readily fashioned to different shapes than donor sclera, although its color is different than sclera and may be cosmetically unappealing in an obvious location.

**Intraocular Lens Selection**

Selection of the appropriate IOL in cases of ectopia lentis will often depend on successful completion of an intact capsulorhexis, uncomplicated lens extraction, and adequate bag fixation. If an intact, properly fixed and centered capsular bag is accomplished, and a circumferential CTR is in place (in the form of a standard CTR or Cionni-style CTR), a 1-piece or 3-piece IOL would be an appropriate IOL for insertion. Accommodating IOLs should probably be avoided because of the limited mechanical functioning that should develop from a fixed capsular bag.

A question that frequently arises surrounds the use of toric and multifocal IOLs. Although both have been used successfully in ectopia lentis cases, the inherent pathology of the zonular apparatus suggests that these IOLs should be used in only the most ideal cases. Toric IOLs require proper alignment along the steep corneal meridian and will lose their astigmatic correction with rotation away from this meridian. The inability to guarantee centration and proper alignment in addition to the possibility of late suture breakage or progressive zonular degradation requiring late lens-in-the-bag fixation (with possible IOL rotation) suggests that a better alternative for astigmatic correction in many of these patients would be limbal relaxing incisions or laser corneal refractive surgery.

Multifocal IOLs present the same challenges of adequate centration and successful determination of the effective lens position. The use of multifocal IOLs is most beneficial in young children with ectopia lentis because of their loss of phakic accommodation, yet the eyes of these children may be the most likely to have late complications due to suture degradation and breakage requiring refixation and proper centration of the capsular bag. For this reason, it is recommended that fixation sutures for these eyes be composed of 8-0 polytetrafluoroethylene (off label) rather than 9-0 polypropylene when a multifocal IOL is being considered. Regardless of the patient’s age, multifocal IOLs should be used in only the most experienced hands with appropriate informed consent and only if proper centration can be guaranteed during the surgery.

**INTRAOPERATIVE COMPLICATION MANAGEMENT**

**Vitreous Management**

Loose lens cases often have vitreous in the anterior chamber at the outset of the case and are more vulnerable to vitreous loss during the procedure. An absolute rule in these cases is to avoid vitreous traction. If vitreous is present at the beginning of the case, it must be removed from the anterior chamber before anterior segment maneuvers begin. Staining the vitreous with triamcinolone can improve visualization. Another option to aid in visualization is the use of an illuminator probe that includes irrigation. In rare cases of minimal prolapse, the vitreous can be sequestered or pushed back by the use of a dispersive OVD. In most cases of prolapse, a limited vitrectomy is required. A single-port pars plana approach has the least likelihood of damaging remaining zonular fibers or the capsule during the removal process. Once the prolapsed vitreous has been removed, placing a dispersive OVD over the exposed region should limit additional prolapse during the case. If vitreous does present again, the surgeon must stabilize the anterior chamber with OVD, halt maneuvers, and perform an additional vitrectomy.

**Capsular Bag Compromise**

Perhaps the most challenging and essential step in ectopia lentis procedures is completion of an intact anterior capsulorhexis. If the capsulorhexis starts to extend to the bag equator, attempts to redirect the
edge centrally must be made. Capsule-supporting hooks placed too close to the capsulorhexis edge may inadvertently place force on the leading edge that causes outward movement of the tear. Loosening these hooks and placing additional OVD to deepen the anterior chamber may aid in redirecting the capsulorhexis centrally. If the edge is not salvageable, microincision intraocular scissors can be used to start a new capsulorhexis by incising the capsule perpendicular to the peripheral tear and then continuing the capsulorhexis from this new starting point. The capsulorhexis can also be completed by using the scissors to create a flap at the original starting point, cutting tangentially to create a smooth edge from this new starting point. Tearing this new flap in a direction opposite the initial direction will allow the capsulorhexis to be completed to the region of the peripheral tear. In the presence of a compromised anterior capsulorhexis, a CTR should not be used. However, an Ahmed segment can be used in this scenario as long as the region of compromised anterior capsule is more than 3 clock hours away from the ultimate fixation site of the ring segment.

In the presence of posterior capsule compromise, no CTRs should be placed and attempts to place ring segments should probably be avoided except in the most ideal situations. A posterior capsule tear will usually necessitate an alternative approach for IOL insertion, outside the capsular bag.

Intraocular Lens Placement

When a compromised capsular bag is encountered, there are many alternatives for IOL placement outside the bag. Intraocular lenses should not be placed in the sulcus without some type of fixation because of the likelihood of the IOL haptics migrating through stretched zonular fibers or a frank zonular dialysis, resulting in subluxation of the posterior chamber IOL (PC IOL). Scleral fixation can be accomplished with 9-0 polypropylene or 8-0 polytetrafluoroethylene using any technique that avoids exposure of subconjunctival knots. One alternative to the use of sutures is to fixate the PC IOL by intrascleral haptic capture.19,20

Perhaps the easiest method for fixating a PC IOL in ectopia lentis eyes uses iris fixation of the haptics with 10-0 or 9-0 polypropylene. The presence of the capsule, although compromised, allows easy insertion of the IOL into the sulcus, followed by prolapse of the optic above the pupillary plane, constricting of the pupil with a miotic, and iris fixation using a Siepser slipknot technique (Figure 11).21

If the surgeon is not trained to do complex IOL fixation surgery, other options include asking the patient to wear an aphakic contact lens or implanting an anterior chamber IOL (AC IOL). An AC IOL may pose a risk to the corneal endothelium if not properly sized, resulting in long-term corneal decompensation. For this reason, it may be desirable to avoid AC IOls in patients whose life expectancy is greater than 20 years. Many of these eyes may have relatively enlarged axial lengths or anterior chambers, and an interesting alternative to the Kelman-style angle-fixed AC IOL is an iris-imbricated Worst-style IOL in which the haptics are imbricated on the iris surface and not dependent on the angle for fixation (Figure 12).

A review by the American Academy of Ophthalmology22 found no significant advantage in any of the above techniques in the absence of random controlled trials. Thus, the choice of technique and IOL is dependent on the surgeon's training and experience.
The techniques for treating ectopia lentis evolve as new adjunctive devices for facilitating the surgery are invented and become available for use and as newer innovative techniques are tried and studied. Although capsule bag fixation using prosthetic devices may be considered the current state of the art in ectopia lentis patients, other innovative approaches offer alternatives or possible improvements over current methods.

One approach used before the advent of modified CTRs involved fixation of the anterior lens capsule to the sclera, where it functioned as a support for a PC IOL and as a vitreous container. With this technique, an incision is made in the anterior lens capsule from equator to equator, oriented 90 degrees from the meridian of subluxation. One-half of the anterior capsule leaflet that is created is then folded back and sutured to the sclera using a 10-0 polypropylene suture. This sutured leaflet provides support for a sclerally positioned or fixated PC IOL, while also acting as a barrier for vitreous prolapse during aspiration of the lens.23

Another innovative method for dealing with ectopia lentis cases was described by Ventura and Endress.24 Rather than attempting to recenter the capsular bag with scleral fixation, Ventura and Endress decenter the IOL within the decentered capsular bag to achieve a centered optic. After a decentered anterior capsulorhexis is created and the lens material is removed, a CTR is placed to support the remaining zonular fibers. One IOL haptic is then amputated from the IOL and the IOL is inserted into the capsular bag, oriented so the remaining IOL haptic decenters the IOL optic toward the bag equator (Figure 13).

Another approach for dealing with the severely subluxated crystalline lens avoids removal of the lens altogether. If the patient is optically aphakic due to severe lens subluxation, placement of an AC IOL without lens extraction simplifies the procedure and avoids the potential complications due to vitreous loss or capsular bag compromise.25 A Kelman-style AC IOL or an iris-fixated Worst-style aphakic IOL can yield immediate visual recovery and markedly shortened surgical time (Figure 14). In the rare instances in which the crystalline lens may later dislocate into the vitreous cavity, the lens can be removed by a pars plana approach by a vitreoretinal surgeon, which might be the safest procedure in these severely subluxated lenses.

Although modified CTRs have become the state of the art for recentering subluxated capsular bags, insertion of the complete Cionni-style CTR can be challenging in the presence of moderate to severe subluxations or in the presence of significant zonular weakness. A recent innovation by Malyugin25 combines the ease of insertion of a traditional CTR with a fixation element that allows sclera fixation of the CTR. Wherein a Cionni-style CTR has the fixation element positioned away from the CTR endpoint and elevated above the plane of the CTR, the Malyugin modification places the fixation element on the end of the ring so it can be easily injected into the capsular bag and then manually positioned above the capsulorhexis to sclerally fixate the prosthesis (Figure 15). This device is currently undergoing development and is not available in the United States.
REFERENCES

DISCUSSION
The surgical correction for patients with ectopia lentis begins with a thorough clinical history, extensive clinical examination, and appropriate referral to medical specialists to evaluate potential metabolic and systemic abnormalities. With the advent and development of current adjunctive surgical devices, most of these cases can now be treated with excellent visual and anatomic results, much better than could be accomplished just a few decades ago. Although this paper attempts to give a rational method for approaching these cases, it in no way suggests there is only one correct method of addressing these eyes, nor was it possible to present all the potential nuances and techniques available for remedying ectopia lentis.

Each surgeon will have his or her own preferences or biases for when and how to use the various prosthetic devices, which fixation sutures and fixation methods work best in his or her hands, and which IOLs are appropriate for each clinical presentation. In addition, what is currently believed to be state of the art may in the future be considered archaic and crude. Thus, each surgeon should keep an open mind as to all alternative methods of treating ectopia lentis patients while making every attempt to be educated about the challenges and options that are present today.
1999; 99–110
24. Ventura M, Endriss D. Implantação de lente intraocular com
uma alça amputada: proposta para o tratamento cirúrgico da
subluxação do cristalino. Intraocular lens implantation with one
loop haptic amputated: a new propose to the subluxation
Available at: http://www.scielo.br/pdf/abo/v73n2/v73n2a07.pdf.
Accessed July 31, 2013
25. Hoffman RS, Fine IH, Packer M. Primary anterior chamber intra-
ocular lens for the treatment of severe crystalline lens subluxa-

OTHER CITED MATERIAL
A. Ahmed IIK, Kranemann C, Crandall AS, “Capsular Hemi-Ring:
Next Step in Effective Management of Profound Zonular Dial-
ysis,” film presented at the ASCRS Symposium on Cataract,
IOL and Refractive Surgery, San Francisco, California, USA,
April 2003
B. Chang DF, “Traumatic Cataract and Mydriasis,” Cataract &
C. Osher RH, “Weak Zonules: Identifying the Subtle Signs of Zonular
Damage,” Cataract & Refractive Surgery Today April 2005, pages
Accessed July 31, 2013
D. Rosenthal K, Personal communication, circa 2005
E. Malyugin B, “Managing Zonular Deficiency; Endocapsular De-
vices can Facilitate Cataract Removal in Patients With Weak
or Missing Zonules,” Cataract & Refractive Surgery Today