New Management Strategies for Ectopia Lentis

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ABSTRACT
Ectopia lentis refers to displacement of the crystalline lens in the setting of various systemic and metabolic disorders. A literature review was conducted to investigate the management of non-traumatic ectopia lentis in the pediatric population, particularly focusing on surgical intervention. Both limbal and pars plana approaches for lensectomy are well established in the literature. Surgical options for intraocular lens implantation in pediatric eyes with ectopia lentis include anterior chamber intraocular lenses and iris-fixated or scleral-fixated posterior chamber intraocular lenses. Recently, the use of capsular tension rings has also been described with promising results. Visual rehabilitation and treatment of amblyopia are essential for patients within the amblyogenic age group following surgical intervention. [J Pediatr Ophthalmol Strabismus. 2015;52(5):269-281.]

INTRODUCTION
Ectopia lentis is a rare condition associated with displacement of the crystalline lens of the eye. The lens is defined as subluxed when it is displaced but still contained within the pupillary space, whereas a luxated lens is completely detached from the ciliary body and can be loose within either the anterior chamber or the vitreous cavity.1,2 Displacement of the lens may induce refractive changes such as astigmatism, myopia, or aphakic hyperopia. Patients may also experience diplopia, particularly if the equator of the lens crosses the visual axis.1

The underlying pathophysiology of ectopia lentis involves the disruption or dysfunction of the zonular fibers.1 Nearly 50% of all cases of lens dislocation are caused by trauma, although ectopia lentis may occur in predisposed eyes even in the absence of trauma because this condition frequently occurs in the setting of systemic disease or metabolic disorder.3 Isolated ectopia lentis, also called simple ectopia lentis, is an autosomal dominant disorder. This condition may present at birth or later in life, and the lens generally displaces upward and temporally. Another heritable condition is ectopia lentis et pupillae, in which both the lens and pupil are displaced, usually in opposite directions. This disorder is autosomal recessive and generally bilateral.1 Microspherophakia, an additional heritable disorder often associated with Weill-Marchesani syndrome, is often bilateral and characterized by a small and spherical lens. Patients with microspherophakia typically have abnormalities in the zonular apparatus, accounting for the high incidence of lens dislocation in these eyes.4

Although there are several systemic disorders associated with ectopia lentis, Marfan syndrome, Weill-Marchesani syndrome, and homocystinuria account for more than 75% of reported non-traumatic lens displacements. Marfan syndrome is
an autosomal dominant disorder characterized by an abnormality in the gene encoding the fibrillin-1 protein, accounting for the typical phenotype of long, thin limbs, tall stature, joint laxity, and cardiac abnormalities. Up to 80% of patients with Marfan syndrome develop ectopia lentis, and 50% of these cases occur before the age of 5 years. Lens displacement is usually bilateral and symmetric, and frequently occurs in the superotemporal direction. In contrast, Weill–Marchesani syndrome, an autosomal recessive disorder characterized by short stature and short digits, is associated with inferior dislocation of the lens. Likewise, up to 90% of patients with homocystinuria experience inferonasal lens displacement. Lens dislocation in these eyes occurs from progressive zonular degeneration. These patients are usually tall and thin, and approximately 50% have developmental delay. Thromboembolic events are common in these patients and may cause death at an early age; thus, all patients with non-traumatic lens dislocation without an obvious cause should undergo biochemical screening for homocystinuria.

The management of ectopia lentis, particularly in children, is challenging. The goal of this review is to discuss the management of non-traumatic ectopia lentis in the pediatric population, focusing particularly on the variety of surgical options that exist for these patients.

**VISUAL SEQUELAE AND CONSERVATIVE MANAGEMENT**

The most common cause of uncorrectable poor vision in non-traumatic ectopia lentis is amblyopia. Amblyopia, as well as anisometropia, can lead to poor binocular fixation. Progressive lenticular myopia and astigmatism are often the first signs of lens dislocation; both may occur when the zonules are disrupted and the lens curvature increases, or when a displaced lens is tilted. Myopia in patients with Marfan syndrome can also be due to long axial length.

The lens may assume a range of positions, including minimal dislocation, pupil bisection, dislocation into the anterior chamber, or posterior displacement into the vitreous. The edge of the lens can be detected in the pupillary space using retinoscopy. The degree of decreased vision depends on the position of the lens. If the lens is displaced posteriorly, despite being central, impairment may be minimal. Some researchers believe that the position of the ectopic lens relative to the pupil correlates more closely with the degree of amblyopia than do other factors such as etiology, age, and visual acuity. Romano et al. suggested that it was possible to predict which children with ectopia lentis would develop significant amblyopia. If the edge of the lens was 1.3 mm from the center of the pupil (range: 0.3 to 2.3 mm), there was high probability that the patient’s vision would not be correctable with conservative measures.

Optimal visual correction within the amblyogenic period is critical. Patching may be a part of this therapy. However, others hold that occlusion therapy cannot address the poor visual development caused by dislocation of the lens. Phakic refractive surgery typically demonstrates a high degree of myopia and astigmatism. In cases where lens tilting or movement creates an aphakic space for refraction, aphaic refraction may be advocated. Some patients could use an aphakic portion of vision for distance and a phakic portion for near vision, and some advocate that phakic and aphakic refraction could be used for monovision, although this would interrupt binocular vision. There is some controversy using phakic and aphakic positions as monovision because it would eliminate fusion in these children.

Aphakic correction may achieve better vision, depending on lens position and pupillary space. Manipulation of the iris has been proposed to expand the pupillary space by pharmacologic mydriasis, and historically by laser iridectomy and iris photocoagulation, although these more aggressive approaches are not current and are controversial. Nd:YAG laser zonulysis has also been performed to create an aphakic space for refraction. Photophobia may result from these measures, but visual acuity may still be optimized and not all patients require pupillary dilation. However, others believe that the pupillary space is still too small after pharmacologic dilatation and that such an approach to aphakic refraction is ineffective. Ultimately, the decision to pursue aphakic or phakic refraction must be guided by the patient’s best-corrected visual acuity (BCVA) and as a response to amblyopia therapy.

**INDICATIONS FOR SURGERY: HISTORICAL AND CURRENT REVIEW**

Historically, surgery was performed to avoid sequelae of ectopia lentis, including retinal detachment, cataracts, uveitis, a lens in the anterior chamber or posterior cavity, imminent complete displacement,
phacomorphic or phacolytic glaucoma, uncorrectable vision by refraction, and amblyopia.\textsuperscript{5,26,27} Historical approaches, including removal of the lens with capsule forceps, open sky vitrectomy with cryoextraction, discission and aspiration, and intracapsular or extracapsular extraction, were used in the 1960s, 1970s, and early 1980s, but are no longer reported.\textsuperscript{5}

Before more advanced techniques of microsurgery, visual outcomes from surgical lensectomy were not reliably improved, and complication rates, especially retinal detachments, were high.\textsuperscript{8,9,26,28,29} Furthermore, anesthesia risk for patients with homocystinuria was great.\textsuperscript{30} Therefore, conservative, non-surgical management and avoidance of surgery were advocated.\textsuperscript{5,31} Instead of surgery, pupillary dilation and massage were regularly advocated to move an anteriorly displaced lens back behind the iris.\textsuperscript{5,30,32,33} Nevertheless, outcomes with conservative management could also be poor.\textsuperscript{12}

More modern closed microsurgical techniques with the vitrector revolutionized the surgical approach for ectopia lentis in the 1980s and 1990s.\textsuperscript{34} Peyman et al.\textsuperscript{35} first reported success using combined lensectomy and vitrectomy with vitrophage in ectopia lentis in 1979, with visual improvement in all 11 eyes regardless of anterior or posterior surgical approach. Girard\textsuperscript{36} reported similar results in 1981. The closed system technique relieves traction on the retina and vitreous; it also avoids damage to the cornea and trabecular meshwork.\textsuperscript{12}

Improvement in anesthesia techniques has made surgery for patients with homocystinuria safer.\textsuperscript{11} In a 1998 study of patients with homocystinuria and ectopia lentis, only 1 of 82 procedures under general anesthesia was associated with a complication.\textsuperscript{37} The single complication was a transient cerebrovascular accident that occurred in a patient with no special precautions taken before or during surgery, and the patient recovered with no long-term sequelae.\textsuperscript{37}

Even with the improved safety profile of surgical intervention in more recent years, indication for surgery is not lens subluxation alone. Most commonly, surgery is performed for a lack of improvement in BCVA with conservative management. The current BCVA threshold for surgery is reported at 20/70\textsuperscript{12,16,34} or 20/60.\textsuperscript{6,10,13,38-40}

Additional indications for surgery as historically considered still apply\textsuperscript{12,17,40} and also include glare, monocular diplopia, and contact lens intolerance.\textsuperscript{12,16,39,40} Visual rehabilitation may be more effective in a child with bilateral than unilateral aphakia, similar to children requiring visual rehabilitation after cataract surgery.\textsuperscript{17} A small retrospective review of children with ectopia lentis, myopia, and angle-closure glaucoma showed that lensectomy reverses angle-closure glaucoma when performed early and mitigates the angle-closure glaucoma when performed later.\textsuperscript{41} In another study, patients with homocystinuria with a high prevalence of mental delay and inability to test visual acuity were operated on for lens dislocation, pupillary block, and optical aberrations. In all 37 eyes with anterior chamber dislocation, conservative measures were first tried but failed, and surgery became necessary.\textsuperscript{37}

Much of the available literature stresses that surgical intervention with appropriate visual rehabilitation can prevent a dense amblyopia if performed early enough, although no controlled trials or retrospective studies have specifically identified an ideal age for intervention.\textsuperscript{6,11-13,16,42,43} Romano et al.\textsuperscript{43} retrospectively reviewed patients with genetic ectopia lentis and found that 50% had significant ametropic functional amblyopia (visual acuity: 20/50 to 20/200), despite appropriate conservative management. The authors recommended early surgery before amblyopia or high axial myopia develops, especially in patients with significant lens displacement within the center of the pupil that may increase the risk of developing amblyopia.\textsuperscript{43} Others believe that earlier surgery for ectopia lentis before ages 4 or 5 years remains controversial.\textsuperscript{44}

In a study of a single family with familial simple ectopia lentis, the surgical outcomes in children between the ages of 14 months and 11 years were found to be superior to the outcomes in their relatives who underwent surgery at a later adult age, including outcomes for associated glaucoma; the authors still concluded that lensectomy should not be performed for subluxation alone in a patient at any age.\textsuperscript{25} These authors believed that adverse sequelae from the subluxation must be present to justify surgery.

In the literature on pediatric cataracts, the importance of early surgical intervention is stressed, as the visual system becomes sensitive to deprivation at 6 weeks and binocular vision that initially develops at 3 months of age.\textsuperscript{45} However, these patients with cataracts often experience more complete visual deprivation than those with ectopia lentis, and children with ectopia lentis are usually older than those with congenital cataracts, even if still within the amblyogenic period.
SURGICAL APPROACH

Surgical approach remains a matter of much debate, with no randomized controlled studies in the literature specifically comparing limbal to pars plana approaches in patients with ectopia lentis. Each approach has anatomically strategic advantages and avoids some risks and adverse sequelae of the other approach. Surgeon comfort level with either approach may be an important factor.

Limbal

A limbal approach is superior for lenses dislocated into the anterior chamber. This approach also requires less instrumentation, avoids using a posterior infusion line, and allows for aspiration with an intact posterior capsule, therefore with reduced risk for trauma to the vitreous base and retina.\(^6\)\(^,\)\(^10\)\(^,\)\(^13\)\(^,\)\(^46\)\(^,\)\(^47\) Avoidance of a sclerotomy also diminishes the risk for vitreous hemorrhage or retinal detachment related to this procedure.\(^13\) The pars plana does not reach adult proportions before 7 years of age, and many authors believe that a posterior approach may be particularly risky for this immature anatomy.\(^15\)\(^,\)\(^46\)\(^,\)\(^47\) The limbal approach may be easier to perform for surgeons more experienced with anterior segment than posterior segment surgery,\(^13\) and some advocate that pars plana lensectomies (PPLs) should be performed only by vitreoretinal surgeons.\(^10\)\(^,\)\(^46\)

Numerous studies support the limbal approach for patients with non-traumatic ectopia lentis.

In 1990, Bekhi et al.\(^6\) published a review of 15 eyes with non-traumatic ectopia lentis operated on with limbal lensectomy and anterior vitrectomy between 1977 and 1989. Average patient age at surgery was 6.15 years. Preoperative visual acuity after amblyopia treatment ranged from 20/60 to 20/200. Improvement in postoperative visual acuity was documented in all eyes, ranging from 20/20 to 20/50 during a follow-up period ranging from 8 to 118 months (median: 33 months). The only major complication was secondary membrane formation that was treated with Nd:YAG capsulotomy.\(^6\)

Plager et al.\(^11\) performed a similar retrospective study in 1992 of 29 eyes in children with ectopia lentis operated on with the limbal approach and anterior vitrectomy. All eyes had visual improvement; 27 of 29 eyes had postoperative visual acuity of 20/40 or better, and the poorer visual outcomes were attributed to amblyopia. No significant complications occurred. Two patients with homocystinuria had bilateral simultaneous surgery under one anesthetic, with no systemic or ocular complications.\(^11\)

Subsequent studies confirmed these findings, with all but a few patients obtaining improved visual acuity, most to better than 20/40, and those with poorer visual outcomes could generally be attributed to amblyopia. Complications from these studies included: transient anterior chamber hyphema, vitreous hemorrhage necessitating a trabeculectomy in a patient with a preoperative pupillary membrane and glaucoma; intractable uveitis requiring a secondary vitrectomy in an eye with preoperative dislocation of the lens into the anterior chamber; and eyes with vitreous strands to the wound, requiring secondary vitrectomy or Nd:YAG laser treatment in some patients.\(^13\)\(^,\)\(^47\)

A larger retrospective review by Kim et al.\(^16\) of 78 eyes of children with ectopia lentis, also treated with limbal lensectomy and anterior vitrectomy, reported that surgical trauma to the zonules was avoided by performing hydrodelineation and maintaining the capsular bag until the end of surgery. The average patient age at surgery was 5.7 years and the average length of follow-up was 7.1 years. Visual acuity improved in all eyes except two, which was attributed to preoperative amblyopia. One patient with Marfan syndrome developed a retinal detachment 9 years after surgery.\(^16\)

PPL

A pars plana approach gains access to any lens or fragments in the posterior chamber, and it also minimizes the chance of displacing the lens more posteriorly during lensectomy.\(^10\)\(^,\)\(^34\)\(^,\)\(^39\) The pars plana approach also avoids possible complications of the limbal approach, most commonly vitreous loss, vitreous adherence to the wound or incarceration, and need for further surgery due to these complications.\(^10\)\(^,\)\(^14\)\(^,\)\(^15\)\(^,\)\(^39\)\(^,\)\(^46\)\(^,\)\(^48\) A pars plana approach allows for complete vitreous removal, which can reduce traction on the retina, and any necessary treatment to the retina can be administered at the time of lensectomy.\(^14\)\(^,\)\(^46\) Additional sequelae that PPL helps to avoid include anterior synechiae, anterior wound dehiscence, pupillary block glaucoma, bullous keratopathy, epithelial or stromal ingrowth, surgically induced astigmatism, and iris trauma.\(^34\)

Despite the theoretical risk of retinal detachment associated with intraocular surgeries in high myopic eyes and some types of ectopia lentis, many studies with PPL showed no retinal detachments.\(^10\)\(^,\)\(^15\)\(^,\)\(^39\)\(^,\)\(^40\)
One group of researchers reviewed 18 eyes after lensectomy for ectopia lentis and noted no retinal detachment. The authors suggested that the low incidence of postoperative detachment may be due to relative stabilization of axial length after lensectomy. The use of closed intraocular microsurgical techniques and improved surgical equipment have allowed greater intraoperative control with fewer complications, including retinal detachment. 

In a study by Hubbard et al., 46 40 eyes with Marfan syndrome and ectopia lentis that underwent PPL with complete vitrectomy and intraoperative panretinal photocoagulation (PRP) reported improved vision in all patients, but concluded prophylactic laser treatment was unnecessary. The postoperative rate of retinal detachment was 6%, which is higher than in other recent studies of ectopia lentis in children, but this rate is in part attributed to the high predisposition of eyes with Marfan syndrome to develop a retinal detachment. 

In 1987, Reese and Weingeist reviewed 12 eyes treated with PPL and anterior vitrectomy. Etiologies of ectopia lentis included idiopathic, Marfan syndrome, familial, and congenital. The average patient age was 7.8 years, and the average length of follow-up was 5.1 years. All but one patient improved to a BCVA of 20/40 or better, and the one poor outcome was attributed to the high predisposition of eyes with Marfan syndrome to develop a retinal detachment. 

Additional studies of non-traumatic ectopia lentis in children treated by PPL with anterior vitrectomy had similar results. Complications included: posterior vitreous detachment, transient vitreous hemorrhage, peripheral anterior synechiae, contact lens-related complications, and a single retinal detachment 2 years after surgery in a child with Marfan syndrome. 

Some reviews reported cases with both limbal and pars plana approach. Hakim et al. published a retrospective review of 44 eyes in children with ectopia lentis. The surgeons performed both PPL and limbal lensectomies, according to individual surgeon preference. Of note, all PPLs in their study were performed by a vitreoretinal surgeon. It is not clear whether the location of the subluxed lens influenced the selected approach, but the authors did mention that in case of a hard nucleus, a PPL approach was advantageous should the lens fall to the posterior chamber intraoperatively. All patients had anterior vitrectomies. All eyes had improvement in vision, and 75% had a postoperative visual acuity of 20/40 or better; poorer outcomes were attributed to amblyopia. Six of 26 limbal lensectomy eyes required removal of vitreous strands to the entry sites with a second surgery, and 1 of 18 PPL eyes had secondary removal of residual lens. Two of 4 eyes that had 360° of retinal cryotherapy during PPL developed tears, but none developed detachments. No retinal detachments or glaucoma complications were reported. 

The authors concluded that limbal lensectomy and PPL were both successful techniques. The limbal approach requires less instrumentation and reduces possible trauma to the vitreous base and retina. This is particularly relevant in children, because the pars plana does not reach adult proportions. PPLs were all undertaken by vitreoretinal surgeons in their study.

Anteb y et al. reviewed cases where 59 eyes of children with ectopia lentis were operated on between 1980 and 1994 and an additional 38 eyes were operated on between 1994 and 2001. The surgeons performed either limbal lensectomy or PPL with combined anterior vitrectomy in all children. For the later cohort, PPL was used in 71% of eyes for “marked phacodonesis” or “nearly total subluxation,” and the limbal approach was used in 29% of eyes for “less marked subluxation.” Patient age at the time of surgery was not statistically different between the two surgical groups, and postoperative visual acuities were only marginally better in the later cohort (84.2% BCVA 20/40 or better vs 80%). Complications included one eye in each study period with a retinal tear or detachment. The authors concluded that surgery with either the limbal or PPL approach using an ocutome vitrectomy probe was successful over long-term follow-up. The study also showed little change in approach or results between the 1980 to 1994 and 1994 to 2001 cohorts.

**Anterior Vitrectomy**

Before the advent of automated vitrectomy, the most common complication of lensectomy for ectopia lentis was vitreous loss. Automated vitrectomy with “in-the-bag” aspiration allows for total removal of the anterior vitreous and diminishes vitreous loss, secondary membrane formation, pupillary block, and retinal detachment. Anterior vitrectomy became common practice for lensectomy in children with ectopia lentis to prevent not only vitreous loss and displacement, but also posterior opacification postoperatively. Posterior capsular opacity (secondary membrane) is a common complication and
major risk factor of amblyopia after lensectomy in children. A primary vitrectomy may be needed during lensectomy for both ectopia lentis and cataract surgery in young children.

A retrospective review of children with ectopia lentis who had lensectomies with intraocular lens (IOL) implantation in posterior capsules left intact on first surgery for suspicion of zonular weakness was performed. Thirty-one of 37 (84%) eyes developed visual axis opacification, and 26 had a secondary surgery; 21 of these had discission of the posterior capsule with anterior vitrectomy and 5 had Nd:YAG capsulotomy. Five eyes had more than one surgery for the posterior visual axis opacity. 42

Similar to children after cataract surgery, pediatric patients with ectopia lentis who have lensectomies are prone to develop visually significant posterior capsular opacity, leading to amblyopia without prompt treatment. Forbes and Guo 45 stated that the child's vitreous is dense and can act “as scaffold for the growth of cells even in the absence of a posterior capsule” when reporting on posterior capsular/vitreous face opacification in pediatric cataract surgery. For these patients with pediatric cataract, they advocate either primary posterior capsulotomy with anterior vitrectomy or primary posterior capsulotomy with optic capture via the limbal or pars plana approach. 45 Nd:YAG capsulotomy is an option for posterior capsule opacification, but it may require multiple procedures and high energy. Although their discussion did not address patients with ectopia lentis, it is important to consider better managing anterior vitreous face to achieve best visual outcomes in patients with ectopia lentis. 45, 49

Similarly, Kugelberg et al. 50 randomized patients with pediatric cataract who all had a posterior capsulorhexis to vitrectomy or no vitrectomy. Patients younger than 5.2 years were more likely to require secondary procedures for posterior capsular opacification without primary vitrectomy. In another study, similar results directed recommendations for posterior capsulotomy and anterior vitrectomy in children younger than 5 years. 51 However, these studies evaluated children with cataracts, not ectopia lentis, and the main outcome was posterior capsular/vitreous face opacity, not BCVA.

**VISUAL REHABILITATION**

In cases of postoperative aphakia, visual rehabilitation must begin as soon as possible, usually within 1 to 2 weeks after surgery; traditionally, this rehabilitation has been with refraction, glasses or contact lens fitting, and any necessary occlusion therapy. 10, 38 Anteby et al. 38 gave children younger than 3 years a prescription for monovision, whereas those older than 3 years had multifocal glasses or contacts for distance and reading glasses. Some believe children with bilateral aphakia can tolerate aphakic spectacles well. 5 Others argue that aphakic glasses are heavy, awkward, constrict visual fields, and offer prismatic distortion. 52, 53 Drawbacks of contact lens use include cost, possible infection, displacement of lenses, and noncompliance with use and care. 14

Recently, IOL placement has been performed more frequently as a primary or secondary procedure in pediatric patients with ectopia lentis requiring surgical intervention. Although IOL implantation is the subject of much debate, it is considered appropriate management for children older than 2 years with pediatric cataracts. 45 Compared to contact lenses, IOLs have reduced the chances of inducing corneal epithelial erosion or infection, and are not dependent on patient and caretaker cooperation. 16 The option of IOL placement depends on the amount and degree of capsular support that exists. Options include anterior chamber IOL (AC-IOL), iris-fixated IOL, and sutured or sutureless transscleral-fixated IOL; each has its own associated benefits and risks. 54 More recently, some surgeons have begun using capsular tension rings (CTRs) and capsular tension segments (CTSs) in their surgical management of patients with ectopia lentis. Each of these options is discussed below.

**AC-IOL**

The original design of the AC-IOL was a closed-loop, rigid lens; the newer and more frequently used design is that of an open-loop, flexible AC-IOL. These lenses have been associated with iritis, pigment dispersion, glaucoma, hyphema, corectopia, iris-sphincter erosion, corneal endothelial decompensation, and cystoid macular edema. 55, 58 Many of these side effects occurred with either open-loop or iris-fixated AC-IOLs. 58 Although the newer open-loop design is less associated with the above risks, 37 such complications in the pediatric population are more likely to occur over time given the obvious longer life expectancy of children. For this reason, AC-IOLs are not typically the first choice in pediatric patients with ectopia lentis who require
lensectomy; however, they may be considered in the absence of adequate capsular support.

Closed-loop, semiflexible anterior chamber implants are not commonly used due to high incidence of corneal edema and persistent cystoid macular edema. Smith et al.\textsuperscript{59} evaluated 53 eyes that underwent implantation of closed-loop AC-IOL (Surgidev Style 10 Leiske IOL; Surgidev Corporation, Goleta, CA). Surgery for IOL removal or exchange was needed in 34 (64%) of 53 eyes and corneal transplantation was needed in 26 (49%) of 53 eyes. Given the intractable inflammatory sequelae and complications associated with closed-loop AC-IOLs, the authors strongly urge discontinuation of these lenses.\textsuperscript{59}

Some surgeons are cautious about using AC-IOLs, specifically in patients with Marfan syndrome who require lensectomy.\textsuperscript{60} Because these patients tend to have large eyes with deep anterior chambers and recessed angles, the standard AC-IOL may be too small for implantation, and thus IOL dislocation, corneal decompensation, iritis, or glaucoma may develop. Another concern in patients with Marfan syndrome is their increased risk of retinal breaks and detachment, and the presence of an AC-IOL may make subsequent examination of the retinal periphery difficult.\textsuperscript{61} This is especially concerning in children with Marfan syndrome who will require routine retinal evaluation for several decades.

In 2005, a retrospective study evaluated AC-IOL placement in pediatric patients with ectopia lentis from Marfan syndrome.\textsuperscript{62} Eight eyes with BCVA of 20/50 to 20/400 were implanted with a single-piece, polymethylmethacrylate AC-IOL (model MTA4UO; Alcon Laboratories, Inc.). Average postoperative BCVA at the most recent follow-up visit (mean: 12.7 months [range: 9 to 16 months]) was 20/32 (range: 20/50 to 20/20). Complications included pigment deposition on the AC-IOL (3 eyes), iritis (1 eye), iatrogenic sector iridotomy (1 eye), and fine keratic precipitates (1 eye).\textsuperscript{62}

Another retrospective study evaluated 4 eyes in children with idiopathic lens subluxation who underwent lensectomy via sclerocorneal tunnel incision and iris fixation of an AC-IOL (Artisan model 305, Ophthec USA, Inc., Boca Raton, FL) as a single procedure.\textsuperscript{63} Preoperative BCVA was worse than 6/12 in all eyes. Patients were observed for a minimum of 8 months, and all eyes gained at least two Snellen lines of BCVA. No postoperative complications were reported. Interestingly, whereas no patient underwent a preoperative corneal endothelial cell count, two of the three patients who had unilateral surgery exhibited no endothelial cell loss after 8 months compared to their non-operative eyes. This is in contrast to data showing 40% to 44% endothelial cell loss in children following AC-IOL placement.\textsuperscript{64}

Much of the data on pediatric AC-IOLs comes from studies on pediatric cataract extraction without ectopia lentis. In one report, 10 children underwent AC-IOL placement as a secondary procedure and were observed for 12 to 78 months (mean: 49.2 months).\textsuperscript{65} Among patients who had reached visual maturity prior to becoming aphakic, postoperative BCVA was at least 20/40 in all eyes following AC-IOL placement. However, among amblyopic eyes BCVA in 40% was 20/400 and no eyes achieved BCVA better than 20/40. The most common complication related to AC-IOL placement was pigment deposition on the IOL surface, followed by corectopia, and haptic migration with subsequent glaucoma. A concern of AC-IOL insertion is the greater postoperative recovery time and astigmatism compared to the foldable posterior chamber IOL (PC-IOL).\textsuperscript{66}

Iris-fixated PC-IOL

In the absence of adequate capsular support, a second option for IOL placement in children with ectopia lentis is an iris-fixated PC-IOL.

Some studies have looked at iris-fixated PC-IOL following PPL in children with ectopia lentis. A study from 2008 evaluated 22 pediatric eyes with ectopia lentis (average age: 5.5 years) in which PPL-vitrectomy was performed; 12 eyes then underwent iris-fixated PC-IOL implantation, whereas the remaining eyes were left aphakic.\textsuperscript{67} After an average follow-up of 19.8 months, mean BCVA in cases of iris-fixated PC-IOL improved from 20/117 to 20/35, and more than 80% achieved BCVA of 20/40 or better. IOL dislocation occurred in 4 eyes after an average of 5 months due to rotation of the IOL haptic out of the suture loop. Following IOL resuturing, all 4 eyes achieved a BCVA of at least 20/30.

Other retrospective data have shown similar results for children with ectopia lentis who undergo PPL-vitrectomy with primary or secondary placement of an iris-fixated PC-IOL.\textsuperscript{68} In all cases, a three-piece foldable PC-IOL was sutured to the iris using a modified Siepser sliding knot.\textsuperscript{69} IOL disloca-
tion occurred in several eyes after 6 months (range: 1.8 to 9.5 months), and did so independently of patient age, IOL model, or meridian of suture placement. None of the IOL dislocations were the result of suture compromise or erosion. Outcome following IOL repositioning seemed to vary. One patient with Marfan syndrome developed iris capture following IOL repositioning, but was successfully managed and remained stable by postoperative month 17. Another patient with Marfan syndrome developed retinal detachment 8 months after IOL repositioning, and the visual acuity declined from 20/50 to hand motions.

A modified technique of iris suture fixation of a foldable PC-IOL has been described in children with ectopia lentis for placement within the sulcus space in the region of intact zonule, using the remaining part of the sulcus to guide the proper positioning of the IOL haptic and iris sutures. This study looked at 17 eyes of 9 pediatric patients with ectopia lentis, the majority of whom had superior lens subluxation associated with Marfan syndrome. Lensectomy was performed in all eyes via an anterior approach through a small (3 × 2 mm) capsulorhexis, followed by ablation of the inferior zonule, capsular bag, and anterior vitreous with a vitrectome. A superior crescent of zonule and capsular bag were left intact to provide sulcus support to the superior IOL haptic. The monobloc hydrophobic acrylic IOL (SA60; Alcon Laboratories, Inc.) was used for this study. Both the superior and inferior haptics were sutured to the posterior iris. Mean BCVA improved from 20/153 (range: 20/400 to 20/50) to 20/32 (range: 20/50 to 20/20) over an average follow-up period of 16.3 months (range: 12 to 25 months). No eyes experienced IOL dislocation throughout this period, but one eye developed hyphema, synechiae, and corectopia requiring pupilplasty after 2 months, and another eye developed aseptic endophthalmitis requiring vitrectomy.

Literature on adult patients with iris-fixated PC-IOLs illustrates iris pigment epithelial loss as a potential intraoperative complication; however, subsequent development of pigment dispersion glaucoma has not been reported. Postoperative complications include the formation of peripheral anterior synechiae overlying the IOL haptics, suture breakage, and suture erosion into an iris vessel leading to hyphema. Some authors support using a 9-0 instead of 10-0 polypropylene suture to decrease the risk of suture breakage.

**Scleral-fixated PC-IOL**

In addition to the iris-fixated PC-IOL, another viable option for pediatric eyes in the absence of adequate capsular support is the transscleral-fixated PC-IOL. This technique, originally described in adults more than 30 years ago, has been used (albeit cautiously) in pediatric patients since the 1990s. In 2010, Kim evaluated 22 eyes of 11 pediatric patients with ectopia lentis secondary to Marfan syndrome who underwent lensectomy and concomitant transscleral-fixated PC-IOL placement. Average age at the time of surgery was 9.8 years (range: 4 to 17 years), and mean follow-up was 10.4 months (range: 2 to 20 months). Average BCVA improved from 1.0 logMAR (range: 2.0 to 0.5 logMAR) to 0.18 logMAR (range: 0.5 to -0.1 logMAR). Postoperative complications included pupillary capture of IOL (2 eyes) and elevated intraocular pressure (IOP) (5 eyes). No eyes developed retinal detachment in the follow-up period, which the authors believed was due to visco-dissection of the posterior capsule from the anterior vitreous face prior to PC-IOL insertion.

An earlier review included children with inadequate capsular support secondary to Marfan syndrome (7 eyes), essential lens dislocation (7 eyes), spherophakia (5 eyes), and perforation with lens injury (2 eyes). Sixteen of these 21 eyes underwent lensectomy via limbal incision and concomitant transscleral-fixated PC-IOL placement (model CZ70BD; Alcon Laboratories, Inc., or model 722Y; Pharmacia & Upjohn, Kalamazzo, MI). Mean age at the time of surgery was 5.8 years (range: 1 to 11 years), and average follow-up was 21.6 months (range: 9 to 33 months). BCVA improved in 15 eyes and remained unchanged in 4 eyes; vision in the remaining 2 eyes could not be measured preoperatively or postoperatively given concurrent developmental delay. Postoperative complications included mild fibrinoid anterior chamber reaction (2 eyes), posterior synechiae formation and cells on the IOL surface (4 eyes), and IOL subluxation into the anterior chamber that was managed successfully with pilocarpine (2 eyes). Because both cases of IOL subluxation occurred with the CZ70BD lens (Alcon Laboratories, Inc.), it is possible that this was due to the relatively small optic size (6.5 or 7.0 mm) compared to the pediatric pupil, which is often 7.0 mm in darkness. No eyes in this review developed cystoid macular edema, hemorrhage, or retinal detachment.
A case report from 2002 described a 5-year-old boy with ectopia lentis and subsequent bilateral superonasal subluxation who underwent bilateral transscleral-fixed IOL placement. An IOL (Acrysof; Alcon Laboratories, Inc.) was inserted via a polypropylene suture bridging the posterior chamber by way of two sclerotomies placed 180° apart. After the sutures were adjusted to center the IOL appropriately, the sutures were then secured beneath each scleral flap. Although complicated by an IOP spike on the first postoperative day, both eyes achieved BCVA of 20/40 within 2 weeks (preoperative visual acuity was 20/40 in the right eye and 20/60 in the left eye).74

Some surgeons advocate the importance of anterior vitrectomy when performing lensectomy with transscleral-fixed PC-IOL placement in children with ectopia lentis. One retrospective review evaluated 17 pediatric eyes with ectopia lentis, including Marfan syndrome (9 eyes), ectopia lentis et pupillae (3 eyes), simple ectopia lentis (2 eyes), sporadic spherophakia (1 eye), and homocystinuria (2 eyes).40 All eyes underwent PPL and anterior vitrectomy; 16 eyes were left aphakic, whereas one eye that had been aphakic for 6 years underwent secondary placement of a transscleral-fixed PC-IOL for contact lens intolerance. BCVA in this patient, who had undergone amblyopia treatment following lensectomy, improved from 20/1600 to 20/100 after placement of a +26.5-diopter PC-IOL; her refraction changed from -8.00 +1.25 × 90 to -1.00 +3.00 × 80. The only postoperative complication in this patient was transient IOP elevation to 28 mm Hg in both her operated and unoperated eyes at postoperative year 5, which resolved spontaneously without optic nerve changes.40

Another retrospective review evaluated 6 pediatric eyes with ectopia lentis secondary to Marfan syndrome who underwent lensectomy and anterior vitrectomy with concomitant transscleral-fixed PC-IOL implantation.75 Average age at the time of surgery was 66.5 months (range: 60 to 73 months), with a mean follow-up of 71.5 months (range: 12 to 84 months). BCVA improved from 20/400 to 20/320, 20/63, and 20/32, respectively, in 3 eyes, and from 20/50 to 20/40 in one eye. Of the remaining 2 eyes, one remained stable at central, steady, and maintained in a preverbal child, and one worsened from 20/200 to no light perception due to endophthalmitis. Late IOL decentration occurred in one eye (postoperative month not indicated) due to breakage of the 10-0 polypropylene suture.75

A literature review by Johnston and Charteris found that although combined PPL and anterior vitrectomy was safer in patients with ectopia lentis who have a greater than baseline risk of retinal detachment, performing a complete pars plana vitrectomy in these eyes may reduce this risk. The authors pointed out the challenge in placing sutures precisely within the ciliary sulcus, because in eyes without zonular tension, including those with ectopia lentis, the ciliary processes tend to contract, thereby narrowing the ciliary sulcus and making PC-IOL placement challenging. Furthermore, to reduce the risk of hemorrhage induced by damage to the major arterial circle of the iris, needle passes need to be made through the avascular portion of the ciliary body when suturing the PC-IOL into place. The authors also recommended using a single-piece polymethylmethacrylate IOL with a large optic (range: 6.5 to 7.0 mm), haptic-to-haptic diameter of 12.0 to 12.5 mm, posteriorly angled haptics, and preferably haptics with eyelets to allow easier fixation. Although their review was not specific to either pediatric patients or those with a history of ectopia lentis, the authors point out that for patients with Marfan syndrome who have undergone vitrectomy, the above average incidence of hypoplastic iris sphincter muscles may put them at risk for IOL tilt and pupillary capture of the IOL optic.57

Legitimate concern exists regarding postoperative complications following pediatric transscleral-fixed PC-IOL placement, including pupillary abnormalities, glaucoma (including pigmentary glaucoma), IOL tilt or decentration, pupillary IOL capture, uveitis, hemorrhage, cystoid macular edema, retinal detachment, suture erosion or breakage, and endophthalmitis.65,67,76-78 Intraoperatively, problems such as hypotony, hyphema, or suprachoroidal hemorrhage can occur given the degree of manipulation required to insert and adequately secure a transscleral-fixed PC-IOL.55 Furthermore, suture erosion through the conjunctiva may lead to endophthalmitis; for this reason, it is common practice to bury suture knots within the sclera or to conceal them beneath a scleral flap.55,74

A large concern with pediatric eyes is IOL subluxation, because lifelong IOL stabilization is obviously an important surgical goal in children. Interestingly, the sole support for the IOL postoperatively appears to be the suture itself rather than fibrosis. Gonioscopy performed at postoperative month
36 in one study, found that contrary to previous hypotheses, no fibrous proliferation had formed around the IOL haptics. Some authors recommend using 9-0 instead of 10-0 polypropylene sutures for both transscleral- and iris-fixated PC-IOLs, particularly in the pediatric population who require IOL stabilization for many decades. Data on children who undergo primary or secondary transscleral-fixated PC-IOL implantation for congenital or traumatic cataracts have demonstrated surgical stability over a broad range of follow-up periods (range: 4 to 40 months). Postoperative BCVA in non-amblyopic eyes improved or remained stable in most cases. The most common postoperative complications appear to be persistent inflammation, increased IOP, and IOL decentration. Less frequent complications include vitreous hemorrhage, cystoid macular edema, and suture erosion through the conjunctiva. In some cases, suture erosion has been managed by shrinking the suture ends with an argon laser. To decrease the risk of suture erosion and endophthalmitis, one may bury suture knots within the sclera, cut suture ends flush with the knot, and cauterize suture ends if they begin to erode through the conjunctiva, or use a scleral flap or donor scleral patch intraoperatively. Some authors advocate haptic fixation via sutureless techniques, including scleral flaps or tunnels or intrascleral gluing.

In addition to the aforementioned risks, another concern with transscleral-fixated PC-IOLs is that inserting them, particularly placing the haptics in the proper location, is essentially a blind procedure. When using an ab interno technique, the needle tip is obscured by the iris (blind approach), and for this reason many surgeons advocate using an ab externo technique to minimize the intraocular manipulation. The latter approach also offers the benefit of passing the suture through the sclera while the eye is closed and firm, which is of particular benefit in vitrectomized eyes. Endoscopic visualization has been employed to overcome the difficulty of visualization during ab interno sulcus IOL placement.

Another obstacle to placement of a sulcus-fixated IOL is the resultant astigmatism from the relatively large corneoscleral wound that is required for insertion of a single-piece polymethylmethacrylate IOL. Foldable IOLs pose risk given the lack of sufficient location on the haptic to secure a suture, predisposing to possible lens rotation and movement. Certain IOLs may prove more fitting for this use; not only can a suture be more firmly secured to a lens with a terminal knob on its haptic, but newer flexible lenses permit small eyes to grow without placing tension on the fixation suture. Furthermore, these lenses may provide more stability because greater flexibility allows for a longer area of contact between the haptic and ciliary sulcus. Conversely, greater flexibility, particularly at the haptic–optic junction, compounded with a smaller optic size (5.5 mm versus 7.0 mm), may predispose these lenses to subluxation or visual disturbances in the setting of large pupils.

### CTR

The CTR is a promising option for patients with ectopia lentis who require lensectomy. Inserted into the capsular bag, this device provides both intraoperative and postoperative stabilization of the capsular bag–IOL complex. The standard CTR is an open, compressible ring of polymethylmethacrylate with a single eyelet at each end. By providing centrifugal force at the capsular equator that is distributed equally around the zonular apparatus, the CTR stabilizes weak zonules by recruiting stronger ones. It also decreases the occurrence of postoperative capsular traction and posterior capsular opacification formation. The ideal CTR is slightly larger than the capsular bag itself, so that once in place the ends overlap, thus providing complete circumferential support. Another advantage to the CTR is that it promotes symmetric postoperative capsular contraction, unlike the less controlled changes observed in the absence of a CTR. The literature shows promising results for both adult and pediatric patients with ectopia lentis who have undergone lensectomy and in-the-bag IOL implantation with the aid of a CTR.

Currently, there are no standard guidelines for choosing a CTR size. Lens capsule diameter determines which CTR size is appropriate, and although related to axial length, it correlates better with the white-to-white measurement. Thus, when choosing among the three sizes of currently approved CTRs, the white-to-white should first be measured with a caliper or the IOLMaster (Carl Zeiss Meditec, Jena, Germany). In addition to lens capsule diameter, the surgeon should also subjectively assess the degree and type of zonular laxity. An eye with a small white-to-white measurement and not much zonular laxity could do well with a small CTR. On the
other hand, an eye with a large white-to-white measurement and marked laxity might do better with a larger diameter CTR. The surgeon may insert the CTR into the capsular bag before or after nuclear extraction, being cautious not to entrap cortical material beneath the CTR if placed prior to lensectomy. The CTR should not be used in the presence of an anterior or posterior capsular tear, including noncontinuous capsulorhexis, because ensuing forces may extend small tears that are already present. Also in cases involving a previously decentered and fibrosed capsule, suturing a standard CTR through the capsular bag may induce astigmatism or capsular tear. It is important to note that the CTR is indicated in cases of mild, generalized zonular weakness or localized zonular dialysis of less than 3 or 4 hours. It is not adequate for recentering a severely subluxed lens or preventing progressive zonular loss. For either of these conditions, one may consider a modified CTR or CTS, both of which may be sutured to the scleral wall. Thus, it is important for the surgeon to consider whether zonular integrity is expected to worsen over time. For instance, in comparison to ectopia lentis associated with metabolic disorders with generalized zonular dysfunction/ laxity, lens subluxation due to trauma appears to respond well to the use of a CTR, because the remaining zonules are strong and healthy.

Because capsular tears may result from suturing the CTR through the capsular bag, the modified CTR was designed to address this complication. The modified CTR has one or two fixation eyelets that protrude 0.25 mm anteriorly from the ring; once the modified CTR is in place, these eyelets sit anterior to the anterior capsule and provide an anchor for sutures while allowing for uncompromised integrity of the capsule itself. Typically, the eyelets are sutured in the direction of zonular weakness. An adequately sized capsulorhexis (5.5 mm) decreases the risk of pigment dispersion or inflammation induced by mechanical trauma of the eyelets against the iris. Some authors believe that the modified CTR gives the surgeon greater control of the capsule during insertion of the IOL. Another benefit to this ring is a square-edge design that decreases the incidence of posterior capsular opacification formation.

A retrospective study of children and adults with Marfan syndrome (56 eyes), idiopathic ectopia lentis (30 eyes), and Weill–Marchesani syndrome (4 eyes) looked at lensectomy with modified CTR and PC-IOL implantation. A modified CTR was placed either before phacoemulsification and sutured with 10-0 polypropylene suture or after phacoemulsification and sutured with 9-0 polypropylene suture. BCVA improved in 79 eyes, remained stable in 9 eyes, and worsened in one eye. Postoperative complications included symptomatic posterior capsular opacification requiring Nd:YAG capsulotomy (18 eyes), iritis treated with prolonged topical steroid therapy (3 eyes), increased IOP treated topically (2 eyes), pseudophacodonesis (1 eye), and focal retinal detachment after 1 month that was successfully managed with photocoagulation (1 eye). Late IOL decentration occurred in 6 eyes, 5 of which underwent resuturing, and suture breakage occurred in 9 eyes after an average of 1 year, 5 of which underwent resuturing. All cases of decentration or suture breakage occurred in the setting of 10-0 polypropylene suture.

Another study from 2007 evaluated 37 eyes in 22 children with ectopia lentis who underwent lensectomy with insertion of a standard CTR (2 eyes) or modified CTR (33 eyes). Each ring was fixed to the sclera with a 10-0 polypropylene suture passing either through the eyelet and capsular bag in cases with CTR or through the anterior eyelet in cases with modified CTR. All eyes underwent in-the-bag placement of a single-piece (SN60AT/ SN30AL; Alcon Laboratories, Inc.) or multi-piece (MA60BM/MA60AC/MA30BA; Alcon Laboratories, Inc.) acrylic IOL. Average BCVA improved from 0.26 (range: 0.015 to 0.65) to 0.59 (range: 0.2 to 1.0), and among non-amblyopic eyes postoperative BCVA ranged from 0.4 to 1.0. Postoperative complications included visual axis opacification (31 eyes), IOL dislocation requiring secondary suturing (2 eyes), ocular surface discomfort related to sutures requiring trimming under anesthesia (2 eyes), and anterior synechiae formation (1 eye). No patients in this study developed glaucoma, retinal detachment, or endophthalmitis during the follow-up period.

In addition to the modified CTR, another implant that can be sutured to the scleral wall without passing the needle through the capsule is the CTS. This ring segment is designed for placement within the capsular bag over an area of zonular weakness and, unlike the standard CTR, can be used in cases of profound zonular insufficiency. Its design is 120° with a radius of 5 mm and anteriorly positioned eyelets. Because it can be implanted without a dialing
technique, the CTS can be inserted with less force compared to the CTR and may even be safe to use in the setting of discontinuous capsulorhexis, anterior capsular tear, or posterior capsule rent.\textsuperscript{81,82} Interestingly, multiple CTSs may be placed within the same capsular bag, or may be used concomitantly with a CTR. Furthermore, the CTS offers the option of removal from the eye following lensectomy if so desired.\textsuperscript{82} Surgical success has been demonstrated in adults with Marfan syndrome who undergo phacoemulsification followed by in-the-bag IOL insertion with the aid of one or two CTSs. After one year, all 10 eyes in this study maintained IOL centration, with 8 eyes achieving BCVA of at least 6/12; however, there remains limited data on CTS use in children with ectopia lentis.\textsuperscript{60}

\textbf{REFERENCES}


