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Original Article

Surgical management of non-traumatic pediatric ectopia lentis A case series and review of the literature

Hugo Y. Hsu, MD [a,*]; Sean L. Edelstein, MD [b]; John T. Lind, MD [b]

Abstract

Purpose: To report a small series of pediatric patients with ectopia lentis that underwent limbal-approach lensectomy and vitrectomy and scleral-fixated intraocular lens implantation and to review the literature on the topic of surgical management of ectopia lentis.
Method: A retrospective review of 13 eyes of seven patients that underwent lensectomy, vitrectomy, and scleral-fixated intraocular lens implantation and a review of the ophthalmic literature.
Results: In our series, the average age at surgery was 70.3 ± 13.8 months and the average length of follow-up was 23.8 ± 5.9 months. The mean pre-operative visual acuity was 0.86 ± 0.17 which improved to 0.23 ± 0.09 post-operatively ($p < 0.001$). No complications were encountered in our series. A review of the literature found that amblyopia was the biggest vision-limiting factor. In general, the literature suggested that a higher percentage of eyes that were left aphakic achieved better vision than those implanted with a scleral-fixated intraocular lens. However, there may be selection bias in that more eyes receiving an intraocular lens may have pre-existing amblyopia. The complication rates for lensectomy or scleral-fixated intraocular lens implantation were low in the literature. In the latter group, suture breakage and resultant intraocular lens dislocation is a worrisome late complication.
Conclusion: Surgical intervention for ectopia lentis via vitrectomy techniques yields good result. In cases of unilateral aphakia or in settings where compliance with aphakic refractive correction is questionable and amblyopia is a constant threat, scleral-fixated intraocular lens implantation is highly encouraged. However, long-term follow-up is required due to the risk of suture breakage and resultant intraocular lens dislocation over time.

Keywords: Ectopia lentis, Marfan, Pediatric, Amblyopia, Intraocular lens

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Introduction

Ectopic lentis is defined as dislocation or displacement of the natural crystalline lens. This condition is typically divided into two cases: traumatic displacements and non-traumatic displacements. Non-traumatic ectopic lentis can be associated with systemic disease and/or hereditary causes. Familial or idiopathic ectopic lentis has no associated systemic or ocular findings. There are however many systemic diseases which can predispose pediatric patients to spontaneous dislocation of the crystalline lens.

Dislocation of the crystalline lens can be a hallmark of many diseases, some of which have life threatening manifestations. When finding a pediatric patient with non-traumatic displacement of the lens, it is important to try to identify the cause and refer the patient to proper subspecialist to address possible threatening conditions. Conditions that are associated with ectopic lentis include Marfan syndrome,

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[a] Department of Ophthalmology, Doheny Eye Institute, USC Keck School of Medicine, Los Angeles, United States
[b] Department of Ophthalmology, Saint Louis University School of Medicine, Saint Louis United States

* Corresponding author. Address: Doheny Eye Institute, 1450 San Pablo Street, Los Angeles, CA 90033, United States. Tel.: +1 323 442 6507; fax: +1 323 442 6517.
e-mail address: hhsu@doheny.org (H.Y. Hsu).

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homocystinuria, sulfite oxidase deficiency, Weil–Marchsani syndrome, Ehler–Danlos syndrome, hyperlysinemia, aniridia, and congenital glaucoma.[1]

Ectopic lentis can cause marked complications within the eye. Children can be at risk for diminished vision, marked changes in refraction with induced myopic astigmatism and monocular diplopia. If the lens becomes anteriorly displaced, pupillary block glaucoma can ensue. Posterior displacement or marked lateral displacement can cause effective aphakia, and posterior displacement can also cause potential damage to the retina.

All of these ophthalmic complications can lead to loss of vision. If this occurs during the amblyogenic age, proper visual development will not occur. Therefore, prompt recognition and treatment is of paramount importance. Medical treatment includes frequent cycloplegic refractions, glasses or contact-lens vision correction and monitoring for amblyopia. Ultimately, with marked or progressive lens displacement and subsequent poor vision and amblyopia, surgical intervention must be considered. Current strategies and issues surrounding surgical correction of ectopia lentis include how best to remove the lens and how best to visually rehabilitate the eye.

Herein we present a small case series of patients seen at our institution with ectopia lentis and report their surgical and visual outcome as well as present and review the literature on this subject.

Patients and methods

A retrospective chart review was conducted of all non-traumatic pediatric ectopic lentis treated with lensectomy and primary scleral-fixated intraocular lens (SFIOL) from January 1, 2004 to June 30, 2010. All patients were seen at the Saint Louis University Eye Institute and had surgeries either done at Saint Louis University or Cardinal Glennon Childrens' Hospital in Saint Louis, Missouri. All surgeries were performed by the same surgeon (H.Y.H.).

Approval for the study was granted by the Institutional Review Board at Saint Louis University. The patients were identified from the surgical records of the operating surgeon. Thirteen eyes of seven patients were identified fitting the above criteria. All patients referred for visually significant and/or amblyogenic ectopia lentis had undergone the same combined procedures as they were all deemed for various reasons as being unsuitable for aphakic contact-lens correction by the referring pediatric ophthalmologists.

Patient demographical information were recorded including sex, operative eye, age at time of surgery, underlying etiology, presence or absence of amblyopia, manifest refraction spherical equivalent, and pre-operative best spectacle-corrected vision (BSCVA). Intra-operative data were recorded including any intra-operative complications. Post-operative data recorded included complications, final visit manifest refraction spherical equivalent, and final BSCVA.

Surgical procedure

After induction, intra-operative axial-length measurement was taken with a contact A-scan probe (62%; 8 out of 13 cases) if the patient could not cooperate with pre-operative IOL-master biometry. Keratometry readings were taken using a hand-held auto-refractor. A superior scleral tunnel approach was used in all cases. Conjunctival peritomies were fashioned superiorly and at the 3 and 9 o'clock positions. The superior scleral tunnel wound was initiated 1.5–2.0 mm posterior to the limbus. Limbal-based partial-thickness scleral flaps were fashioned at the 3 and 9 o'clock positions with the posterior edge 3 mm behind the limbus. Two paracentesis sites were fashioned in each superior quadrant. The eyes were filled with a dispersive viscoelastic, and either bimanual or co-axial anterior-approach vitrectomy technique was employed. An initial puncture through the anterior lens capsule was done with the vitrector, and the lens substance was removed with the vitrector along with all capsule and zonular substance except in one case where the capsule was left behind. We did not find the need to specifically remove the nuclear and cortical material before removal of the capsule and zonule complex to avoid lenticular material from falling into the vitreous cavity. A generous anterior vitrectomy was performed as well. Scleral depression was typically performed in areas of intact zonules to allow for complete removal of the peripheral lens material and capsule remnant. If after a core vitrectomy vitreous was found at the wound, then repeat vitrectomy was performed to ensure that vitreous prolapse was cleared. The eye was then reformed with a viscoelastic. A 1-piece polymethyl-methacrylate (PMMA) 6-mm optic posterior-chamber intraocular lens (IOL) with haptic eyelets (Alcon® CZ70BD) was used in all cases. A double-armed 10-0 polypropylene (Prolene) suture on a CIF-4 needle (Ethicon) was threaded through the two haptic eyelets. The superior sclera tunnel wound was then enlarged to approximately 7 mm and the 10-0 Prolene needle is passed ab interno (under the scleral tunnel wound, through the pupil, under the peripheral iris) to exit approximately 1.5 mm posterior to the limbus under the partial-thickness scleral flaps. Two passes were made on either side. The lens was then guided into the eye and underneath the iris plane to rest in the sulcus plane before centration was adjusted by drawing up on the 10-0 Prolene sutures. The scleral tunnel wound was then closed with 10-0 polyglactin (Vicryl) sutures and the eye pressurized with either viscoelastics or balanced salt solution before the final intraocular lens position was secured by permanently tying the Prolene sutures. The vitrector was then used to remove all potential vitreous prolapse and viscoelastics. Miochol was injected into the eye, and the partial-thickness scleral flaps and conjunctiva were reapproximated with the 10-0 vicryl suture. Subconjunctival antibiotic and steroids were injected at the conclusion of the case.

Results

Thirteen eyes of seven patients were identified that met the inclusion criteria. 57% (4/7) were female and 54% (7/13) involved the left eye. The most common etiologies were autosomal dominant, familial ectopia lentis and Marfan syndrome, each accounting for three patients with bilateral disease. Additionally, a single patient was found to have idiopathic unilateral ectopia lentis. The average age at surgery was 70.3 ± 13.8 months (range 43–123 months) and the average length of follow-up was 23.8 ± 5.9 months (range 2–39 months). There was a high incidence of refractive or anisometropic amblyopia noted pre-operatively in all but one patient. This patient had Marfan and was the oldest

patient in the group. He was 9 years 11 months and 10 years 4 months old at the time of his surgeries. Pre-operative BSCVA was worse than or equal to 20/60 in all cases and worse than or equal to 20/200 in 46% (6/13) of cases. The average pre-operative best spectacle-corrected logMAR visual acuity was 0.86 ± 0.17 (20/144; range 20/60–20/400). Pre-operative refraction was available for 12 of the 13 eyes. Two eyes (same patient) wore high plus-power aphakic spectacle correction due to marked lens subluxation. The rest had high myopic astigmatism with an average spherical equivalent of −13.19 ± 4.61 dopters.

The vision improved in all eyes. Post-operative BSCVA was better than or equal to 20/60 in 92% (12/13) of cases and better than or equal to 20/40 in 77% (10/13) of cases. The mean post-operative spherical outcome was −0.91 ± 1.36 diopters. The worst outcome was the second eye of a child with Marfan in whom the second eye was operated on at 6-and-a-half years of age 13 months after the first eye and in whom there was only 2 months of follow-up. In this eye, the BSCVA nonetheless improved from 20/200 pre-operatively to 20/70 post-operatively. The average post-operative best spectacle-corrected log MAR visual acuity for the cohort was 0.23 ± 0.09 (20/34; range 20/20–20/70). The difference between pre- and post-operative BSCVA for the cohort was highly significant ($p < 0.001$; paired Student's *t*-test).

Intra-operative complication occurred in one case. The 10-0 Prolene suture broke along with the haptic of the PMMA lens after it had been implanted. The eye had to be reopened to explant the lens. A substitute lens of the same power was used, and the cases concluded without further issues. Post-operatively, there were no issues noted despite having to perform the lens exchange.

The most common post-operative complication was corectopia noted in 38% (5/13) of cases (due to IOL incarceration in two cases, vitreous prolapse into the anterior chamber in one case, and pupil damage from surgery in two cases). Optic capture was the next most common post-operative complication, affecting 31% (4/13) of cases. Three of these cases spontaneously resolved within 10–21 days, and their final BSCVA were 20/20, 20/25, and 20/60. In the fourth case, the final BSCVA was 20/30. In two of these cases, ocular trauma was noted to be the cause of optic capture and both self resolved. In addition, a single case each of vitreous prolapse into the anterior chamber, posterior capsular fibrosis and ocular hypertension occurred. The patient was noted to have an elevated intraocular pressure of 30 at the 1 week post-op visit, which was adequately treated using carbonic anhydrase inhibitor eye drop for several weeks. The posterior capsular fibrosis occurred in a case where the posterior capsule was not removed and the IOL was placed in the capsule with scleral suture fixation through the capsule equator. YAG laser posterior capsulotomy was performed 3 months post-operatively and the patient's vision improved from 20/50 to 20/25.

No cases of vitreous hemorrhage, choroidal detachment, retinal detachment, endophthalmitis, or Prolene suture breakage were noted in this cohort during the study period.

Discussion

When deciding the correct intervention for ectopic lentis, it is important to formulate a comprehensive plan regarding the diagnosis, surgical timing, surgical approach and plan for visual rehabilitation. As discussed previously, identifying a cause of ectopic lentis is important because this finding can help to diagnoses life threatening associated conditions. After a cause is identified, the next question a surgeon asks is whether the patient needs surgery for visual function or to prevent long-term complications. If a patient can achieve visual rehabilitation and maturation with glasses and/or contact-lens correction, many times surgery can be delayed or prevented. Despite maximal conservative management, Romano et al. reports ametropic amblyopia in 50% of patients with familial ectopic lentis. The vision in these patients ranged from 20/50 to 20/200.[2]

If optimal correction cannot be accomplished, surgical removal of the lens may be warranted.[3] If considering surgery, a thorough ophthalmic examination must be completed, as findings can help the surgeon plan for the anticipated surgery. By examining the anterior chamber depth and angle, a surgeon can better access the degree and direction of subluxation. Looking at the lens under undilated and dilated conditions can aid in assessing the degree and location of zonular dysfunction.[3] A full retinal examination must be conducted, especially in patients with Marfan syndrome, as they can be predisposed to retinal holes.[4] It is also important to consider risk of thromboembolic events in patients with ectopic lentis secondary to homocystinuria.[1] Communication is paramount with the anesthesiologist in this situation and some surgeons advocate bilateral surgery to minimize the number of general anesthetics.[1]

Once surgical removal is deemed necessary to improve a child's vision and/or to prevent irreversible amblyopia, the question is then how best to remove the subluxed lens. Traditionally, in the pre-automated vitrectomy era, removal of subluxed crystalline lens in young eyes was met with extremely high rates of complication such as vitreous loss and retinal detachments with variable and modest visual gains.[1,5–7] The advent of the close-system, automated, irrigation-vitrectomy was a game-changer for approaching these cases. Investigators favor either a limbal, anterior approach or a pars-plana approach. From either approach, there are generally good outcomes and minimal complications.[5–10] Table 1 summarizes the results of primary lensectomy via either the anterior or pars-plana approach. While the closed system allows for safe removal of the subluxed lens, the key to success and to minimize complications in all these reports is a thorough and meticulous removal of the vitreous gel and to make sure that there are no vitreous incarcerations that would potentiate retinal detachments. Theoretically, a pars-plana approach may allow for more thorough vitreous removal and spot pre-existing or iatrogenic retinal tears; however, the published literature does not support the superiority of the pars-plana approach over a limbal approach. While we utilized an anterior/limbal approach, we feel that the choice is up to the surgeon and his/her comfort level and expertise with either approach.

Once the crystalline lens is removed, optical rehabilitation of the child with aphakia and no capsular support presents the next therapeutic challenge. Options include aphakic spectacles, contact lens, angle-supported anterior chamber intraocular lens implant (ACIOL),[11,12] anterior chamber iris-enclavated IOL,[13] posterior-chamber iris-fixated IOL implant,[14,15] posterior-chamber capsule-placed IOL centered

Table 1. Primary lensectomy for ectopia lentis in pediatric patients.

	Reese and Weingeist[5]	Behki et al.[6]	Plager et al.[7]	Halpert and BenEzra[8]	Anteby et al.[9,a]	Noorani et al.[10]
No. of eyes	12	15	29	59	38	90
Age at surgery (years)	2.5–17	3.2–12.3	3–11	<3 to >5	6.4 ± 4.6	2.5–16
Length of follow-up	1–6.3 y	8–118 m	5 m–12 y	1–12 y	18 m–6 y	1–11 m
Surgical approach	PP	Limbal	Limbal	PP (54) limbal (5)	PP (27) limbal (11)	Limbal
Retinal detachment	0	0	0	1[b]	1	1
Glaucoma	0	0	0	0	0	0
Vitreous hemorrhage	0	0	0	0	1	0
Choroidal detachment	0	0	0	0	0	2
Visual outcome	20/20–20/70 (92% ⩾20/40)	20/20–20/50	20/15–6/200 (93% ⩾20/40)	80% ⩾20/40, 12% 20/40–20/70 8% <20/70	84.2% ⩾20/40	20/25-HM, 17% ⩾20/40 71% ⩾20/60

PP = pars-plana, HM = hand-motion.

[a] This report also included longer-term follow-up of a previous published cohort.[8] The details reported herein are specific to the newer cohort discussed in this publication.

[b] This case of a retinal tear in the cohort was described in a subsequent publication which provided even longer-term follow-up.[9].

with a capsular tension ring,[16] or a SFIOL implant.[11,17–24] Each of these approaches has inherent limitations and problems. Wearing aphakic spectacles is the safest but least physiologic method of optical correction, often resulting in aniseikonia and visual distortion. Aphakic glasses are not well tolerated by young children because of the glasses' weight, awkward appearance, prismatic distortion, and constriction of visual fields. Contact lens use requires ongoing care, has an inherent increased risk of corneal infection, and contact-lens intolerance can develop.[25] Both of these approaches are limited by children's and their family's compliance with using the vision-correcting devices with the constant specter of refractive amblyopia. In aphakia, good visual acuity outcomes are highly dependent on compliance with amblyopia therapy in the first several years of life. Compliance with occlusion therapy is often variable among patient populations. Epley et al. reported worse visual outcomes in their younger patient cohort due to poor compliance with optical rehabilitation and occlusion.[11] With that being said, the vast majority of the aphakic patients from the series described previously of primary lensectomy achieved good corrected visual acuities with 70–100% achieving 20/60 or better [5–10] and as high as 93% achieving 20/40 or better in one series (Table 1).[7]

IOLs obviously offer the advantage of permanent compliance, minimal aniseikonia, and an early and generally predictable refractive result. Relative short-term outcomes (9–16 months) with ACIOLs have been reported to provide good visual outcomes in young patients with no significant complications.[12] However, ACIOL implants in the pediatric population generally have had significant long-term complications, such as endothelial cell loss, corneal decompensation, iris-sphincter erosion and pupillary ectopia.[26,27] Because of these problems, posterior-chamber SFIOLs are generally preferred over ACIOLs in aphakic children with inadequate capsular support for PCIOL implantation,[11] and we agree with this general preference. Nonetheless, alternatives to conventional angle-support ACIOLs and SFIOLs do exist. The Artisan iris-enclavated IOLs have been used in a very limited basis in pediatric patients with subluxed crystalline lenses with good short-term outcomes while they have enjoyed wider usage in cases of congenital cataracts.[13] Also, posterior-chamber iris-fixated IOLs have been utilized with longer follow-up likewise with reported good visual outcomes and low complication rates in one paper [14] while another group reported good visual outcomes but with significant rates (33%) of IOL dislocation due to IOL slippage.[15] Lastly, it is possible to implant a conventional capsular tension ring or a modified scleral-fixated capsular tension ring to allow in-the-bag placement of an IOL.[16] In this report of 37 eyes, the mean post-operative BSCVA was 20/78 (range 20/32–20/200). The authors reported that if amblyopic eyes were removed from the analysis, then the mean BSCVA improved to 20/50. However, posterior capsule opacification requiring intervention occurred in 83.8% of patients (31 out of 37) during the median follow-up period of 27 months (range 1–59 months).

Scleral-fixated IOLs are the most popular means of implanting an IOL in an aphakic pediatric eye. Nonetheless, much more is known about the outcomes and complications of SFIOL in the adult population. Hyphema or vitreous hemorrhage as a complication is not surprising, considering that scleral suturing of a PCIOL requires that the needle pass through vascular tissue. In most cases, the bleeding resolves spontaneously.[28] It has been proposed that placing the sutures anteriorly (0.5–1.0 mm behind the surgical limbus) to allow the lens to sit in the true sulcus space and avoid the vascular ciliary body may reduce the risk of bleeding.[29] However there is discussion about the location of the true sulcus space relative to the surgical limbus.[30,31] The caveat to these debates is that these reports pertain to experiment in adult cadaveric eyes and surgery in adult eyes and not pediatric eyes. A survey of reports regarding SFIOL in pediatric eyes found that, when reported, the location of suture placement ranged from 0.5 to 2.0 mm posterior to the limbus.[17–19,23,24] With the exception of one report, there were relatively few incidences of intraocular bleeding, and in all cases where they are encountered, they are all self-resorbing within days. This one exception reported intraocular bleeding in 13 out of 25 eyes (52%) in which the suture was placed 1.0 mm posterior to the limbus.[24] We tend to pass the suture 1.5 mm posterior to the limbus, and in our relatively small series, we did not encounter intraocular hemorrhage. It has additionally been suggested that to prevent intraocular bleeding that the surgeon take care to maintain a pressurized globe at all times during the surgery and to minimize tissues traversed during transscleral needle passes, and we agree this recommendation.[32] Table 2 summarizes several series in the literature, including the current series, of pediatric patients that underwent either primary or secondary SFIOLs along with the complications and visual outcomes.

Retinal detachment is a potentially devastating complication after implantation of a SFIOL. In one series of adult eyes,

Table 2. Scleral-fixated intraocular lenses (SFIOL) in pediatric patients.

	Sharpe et al.[17]	Kumar et al.[18]	Zetterström et al.[19]	Epley et al.[11,b]	Jacobi et al.[21]	Bardorf et al.[22]	Buckley[23]	Asadi and Kheirkhah[24]	Current series (2012)
No. of eyes	7	11	21	18	26	43	33	25	13
Age at surgery	111.4 m (12–228 m)	6.45 ± 1.58 y	5.8 ± 2.6 y	94.7 m (21–202 m)	13.2 y (6–29 y)	1–19 y	9.7 ± 5.1 y (1–17 y)	79 ± 20.2 m (33–120 m)	70.3 ± 13.8 m (43–123 m)
Length of follow-up	26 m (3–38 m)	10.9 ± 4.3 m (4–18 m)	21.6 m (9–33 m)	10.3 m (6–26 m)	6–20 m	37 m (8–66 m)	61 ± 42 m (9–200 m)	81.1 ± 46.2 m (12–144 m)	23.8 ± 5.9 m (2–39 m)
Intraocular hemorrhage	0	0	0	1	0	6	1	13	0
Retina detachment	0	0	0	0	0	0	0	1	0
Corectopia	1	0	0	2	1	0	1	0	5
Glaucoma[a]	0	1	0	0	1	0	2	0	0
Suture exposure	1	2	0	0	2	1	0	0	0
Suture breakage IOL dislocation	0	0	0	0	0	0	4	6	0
Decentered iris-capture IOL	2	1	0	0	5	2	3	0	1
Endophthalmitis	0	0	0	0	0	0	0	1	0
Visual outcome	0.37 (20/47), 83% ⩾20/40	0.49 (20/62), 64% ⩾20/60, 55% ⩾20/40	0.53 (20/68), 47% ⩾20/60, 32% ⩾20/40	44% ⩾20/60, 39% ⩾20/40[c]	0.20 ± 0.17, (20/30)[d], 0.26 ± 0.16 (20/36)[e]	76% ⩾20/60, 73% ⩾20/40	79% ⩾20/40	0.42 ± 0.36 (20/53), 68% ⩾20/60, 64% ⩾20/40	0.23 ± 0.09 (20/34), 92% ⩾20/60, 77% ⩾20/40

m = months, y = years.
[a] Excludes transient intraocular pressure increases.
[b] SFIOL series only.
[c] "older kids" (⩾9 years old): 83% ⩾20/40.
[d] Multifocal IOL.
[e] Monofocal IOL.

retinal detachment occurred in 4.9% of 122 eyes during a follow-up period of 42 months.[33] Again the reported rate of retinal detachment after SFIOLs in children seems to be much lower. In the papers reviewed, there was one case out of 197 (Table 2). The adult eyes that suffered retinal detachments also included those that had suffered previous trauma; therefore, the cause of retinal detachment may not be due directly to the surgical implantation of a SFIOL.[34] The pediatric series reviewed also included eyes that were traumatized; additionally, patients with Marfan syndrome feature prominently in the pediatric series where there is a known increased risk of retinal detachments. It remains interesting to speculate why there seems to be a lower risk of retinal detachment in pediatric eyes.

One of the significant complications in adults who have undergone sutured IOLs is the eventual erosion and exposure of the fixation knot through the conjunctiva. Long-term transscleral suture exposure rates of 5% to 50% have been reported when sutures were covered with conjunctiva only, and exposure rates of 14.7–17.9% have been reported when sutures were covered with sclera flaps.[35–37] Again, the rate of suture erosion in pediatric cases seems to be lower. Our review of the literature revealed six cases (Table 2). In all pediatric case series, the sutures were buried under partial-thickness scleral flaps, which we advocate, which may have reduced the risk of erosion and exposure. It is also possible that exposure of suture knots was not noted in pediatric case series due to the generally short follow-up periods of published reports. Additionally, we speculate that the healthier and thicker conjunctiva and tennon's capsule of pediatric eyes may help to prevent frank suture erosion.

Besides symptomatic foreign-body sensation as a result of suture erosion and exposure, the main fear of having an exposed suture is that of endophthalmitis as infecting organisms may gain access to the eye via exposed sutures. Late endophthalmitis after SFIOL implantation is a real danger.[38] Although scleral flaps reduce the risk of suture exposure, in the long term, suture ends can erode through partial-thickness scleral flaps and conjunctiva. Indeed, there is one reported case of a late-onset endophthalmitis in a pediatric eye with SFIOL that developed 3 years after the surgery.[24] Unfortunately, there is no description of whether suture exposure was seen or part of the pathophysiology.

A disturbing complication of transscleral fixation of IOLs is the late failure of the 10-0 polypropylene sutures through degradation or trauma, which would result in lens decentration or subluxation. Histopathologic findings have shown a lack of significant fibrosis around the lens loops, suggesting that the suture remains the only support for the SFIOLs.[37] Polypropylene suture is considered to be non-absorbable; however, they do degrade slowly over time and are not "permanent".[39] One report of largely adult patients found late polypropylene suture breakage in 16 of 61 eyes (26.2%) occurring about 50 months after IOL fixation.[40] Another report of adult eyes with a shorter duration of follow-up (24 months) found a lower rate of suture breakage (2.2%) in 89 eyes after penetrating keratoplasty and SFIOL implantation.[41] In contrast, in another report with more than 10 years of follow-up of 16 adult patients with SFIOLs, no cases of suture breakage occurred.[42] Therefore, while perhaps not a certainty, the concern is that 10-0 polypropylene sutures degrade and eventual breakage will increase over time with

resultant IOL dislocation. This is of obvious concern for the pediatric patient. In pediatric series, we encountered a total of 10 cases of SFIOL dislocation due to suture breakage (Table 2).[23–24] These reports had the longest average follow-up periods of 5.1 and 6.8 years of the surveyed series. In one report, the author noted that the suture breakage occurred at 3.5, 5, 6, and 8 years.[23] Additionally, the author reported that a survey of pediatric ophthalmologists revealed an additional 10 eyes with dislocated SFIOLs from broken 10-0 polypropylene sutures and three additional eyes from another published series. In all cases, the situation was successfully dealt with surgically with good retention of visual acuities. In another report, the average time to suture breakage and IOL dislocation was 8.7 ± 1.2 years; in this report, all IOLs were explanted.[24] As a result, several recommendations have been put forth to reduce the risk of suture breakage and IOL dislocation. These include the usage of multiple sutures, thicker sutures (9-0 polypropylene), and different suture materials.[23,24,32] Further research is needed to find an ideal method and/or material for fixation of IOLs to the sclera.

Table 2 summarizes the results from the pediatric SFIOL reports in the literature. Additionally, it reports the visual outcomes reported. In general, the average best-vision ranges from approximately 20/60 to a bit shy of 20/30. Compared to reports of aphakic eyes with spectacle or contact-lens correction (Table 1), it would appear that fewer eyes with SFIOLs achieve 20/40 or better vision. However, one should keep in mind that eyes that come to SFIOLs oftentimes have failed or are intolerant of contact-lens correction or are unilateral aphakic eyes. As such there is likely a higher proportion of eyes with amblyopia in the SFIOL group, especially if the patient is young. In one report of a series of unilateral aphakic eyes, only 39% of the entire cohort achieved 20/40 or better vision after SFIOL implantation. In contrast, a subgroup analysis of patients from the cohort that became aphakic after 9 years of age, and thereby after the amblyopic age-range, revealed that 83% achieved 20/40 or better vision.[11]

In conclusion, vitrectomy techniques revolutionized the management of pediatric ectopia lentis cases due to the ability to successfully remove the subluxed crystalline lens and vitreous and avoid marked complications. The most important determinant of good visual outcome in these vulnerable eyes is amblyopia. If a child develops good vision before visually-significant lens subluxation, s/he should do well after surgery. If, however, the child is at risk of amblyopia or already has amblyopia, the visual outcome is much more guarded. Nonetheless, aggressive visual correction can still reverse the amblyopia trend. Our series with amblyopia in 6 out of 7 patients demonstrates that poor pre-operative vision can be improved if not reversed with intervention.

While the decision to implant an IOL at the time of lensectomy is a difficult one due to uncertainty about whether a child and his/her family have access to and/or will be able to comply with aphakic correction and amblyopia therapies, the low rate of complication from IOL implantation should prompt physicians treating these special patients to consider primary IOL implantation more frequently as a viable option. Even though the risks associated with IOL implantation in aphakic eyes are low and there is significant overlap of the complications from lensectomy and IOL implantation, there are, nonetheless, IOL-specific complications that are worrisome. The chief of these relates to the use of 10-0 polypropylene sutures in pediatric patient as was discussed previously. We feel that even though better methods and materials need to be developed to achieve longer-term safety for IOL implantation in the absence of capsular support, the surgical option should not be withheld from the pediatric patient facing permanent visual handicap from amblyopia. We feel that this is especially true in cases that will result in unilateral aphakia. A child with bilaterally subluxed lenses should either be bilaterally aphakic or have IOLs implanted in both eyes.

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