Management of ectopia lentis in children

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Abstract:

The medical management of ectopia lentis involves refractive correction as well as co-management of any associated systemic disease. Surgical management remains a challenge, as inherent defects in the lens capsule make implantation of an intraocular lens (IOL) difficult. Multiple visual rehabilitative measures are available such as aphakic contact lenses or spectacles, capsular bag fixation with implantation of in-the-bag IOL, iris-fixated, and scleral-fixated IOL. It depends on the surgeon's expertise and discretion whether the capsular bag needs to be preserved or compromised.

Keywords:

Capsular tension ring, Cionni ring, iris fixated intraocular lens, ectopia lentis, intralenticular lens aspiration, lensectomy, scleral fixated intraocular lens, zonulopathy

Introduction

Ectopia lentis in children is difficult to manage. Management of lens subluxation in adult population is well defined and the same is being utilized in managing pediatric cases of ectopia lentis. Relatively older children can be managed according to these guidelines but for younger children, decision-making is often difficult. The management can be broadly classified into conservative and surgical.

Conservative

Optimal visual correction within the amblyogenic period is critical as amblyopia seems to be the most common cause of poor vision in cases with ectopia lentis,[1-3] which when combined with anisometropia often results in poor binocular functions.

The extent and asymmetrical nature of the zonular dehiscence determines the degree of visual impairment. For example, progressive and concentric zonular weakness in one eye allows the lens to remain centered and spherical thus giving rise to simple lenticular myopia that can be optically corrected. On the contrary, a sectoral zonular dehiscence induces significant myopic astigmatism due to lens tilt and displacement.[4]

Lenses with either very low or very high grades of subluxation respond well to optical means of visual rehabilitation. Mild subluxation allows the patient to see through the phakic portion of the visual axis with appropriate spectacle or contact lens prescription whereas higher grades of luxation permit optical correction of the aphakic portion. According to one school of thought, occlusion or patching therapy plays a vital role in the reversal of amblyopia in the presence of anisometropia and/or strabismus.[3]

On the other hand, others believe that occlusion therapy remains ineffective in amblyopia management caused by dislocation of the lens.[5] Aphakic correction tends to have better acceptance when prescribed bilaterally. Previous literature also suggests various techniques such as laser iridectomy and iris photocoagulation to enlarge the pupillary area and hence the aphakic zone.[6,7] However, these approaches tend to be more aggressive and are not currently in use due to associated photophobia.

Finally, the corrected distance and near visual acuity of the patient should determine the requirement to pursue aphakic or phakic refraction for the patient.

Literature search

A literature search was performed using PubMed Medline, the Cochrane Library Database,

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EMBASE, and Scopus (from 1970 onward), using the following terms: ectopia lentis, lens subluxation, management of ectopia lentis, visual rehabilitation in aphakia, intralenticular lens aspiration, lensectomy, SFIOL, aphakia, capsular supporting devices, intralenticular lens aspiration, lensectomy, CTR, Cionni ring, CTSs, contact lenses in aphakia, capsular bag fixation. All relevant articles were included. Priority was given to prospective studies and randomized clinical trials. However, retrospective studies and case series were included if important. Reference lists from the selected articles were further checked to obtain further relevant articles not included in the electronic database.

Surgical

When deciding the correct intervention for ectopic lentis, it is important to formulate a comprehensive outline that would include diagnosis, surgical timing, surgical approach, and visual rehabilitation.

Indications for surgery

Historically, surgery was performed to avoid sequelae such as retinal detachment, cataracts, uveitis, phacomorphic or phacolytic glaucoma, uncorrectable vision by refraction, and amblyopia.[8]

In the current scenario, indications of surgery would include the following:

1. If there is presence of significant or progressive dislocation in the younger age group
2. Lack of improvement in best corrected visual acuity with conservative management (glasses, contact lens, and/or patching)
3. For older children and adults, if poor visual acuity is attributed to subluxated lens and is not amenable to spectacle correction
4. Lens is threatening to dislocate anteriorly or posteriorly
5. Phacolytic uveitis and glaucoma
6. Visually significant cataract
7. Retinal detachment.

Timing of surgery

Although none of the studies have suggested an ideal age to undergo surgical intervention, several studies have stressed the fact of preventing dense amblyopia when lens extraction with age-appropriate visual rehabilitation is performed at an early age.[3,9-14] Romano *et al*., in their retrospective study, revealed 50% to have significant functional amblyopia in the postoperative period despite appropriate conservative measures.[14] Therefore, they recommend early surgery even before amblyopia or high axial myopia sets in,[14] while the other authors do not recommend surgery for ectopia lentis before 4–5 years of age.[15] As the visual system becomes sensitive to deprivation at 6 weeks and binocular vision development commences at 12 weeks, visual deprivation is incomplete in cases of ectopia lentis compared to that of congenital cataract owing to the older age group of presentation.[16] In the literature on pediatric cataracts, the importance of early surgical intervention is stressed.

Choice of surgical procedure

After identification of the primary pathology, the surgeon tries to find out whether surgery is required for visual rehabilitation or to prevent long-term complications.[17] If optimal correction cannot be accomplished with conservative measures, surgical removal of the lens may be warranted. 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 subluxated lens. Following removal of the crystalline lens, optical rehabilitation of the child is done in the same or staged sitting depending on the degree of zonular dehiscence as follows:

1. Superior up to 4 clock hours: Capsular tension ring (CTR) with posterior chamber intraocular lens (PCIOL) in bag; PCIOL with haptic being used to stretch the bag
2. Inferior up to 3 clock hours: CTR with PCIOL in bag
3. Between 3 and 6 clock hours: Bag fixation with modified CTR (MCTR) or single-loop Cionni ring with PCIOL in bag
4. Between 6 and 9 clock hours: Bag fixation with MCTR or double loop Cionni ring with PCIOL in bag
5. ≥9 clock hours/generalized zonular weakness: Intracapsular cataract extraction with scleral fixated intraocular lens (SFIOL)/Iris fixated intraocular lens (IOL)/anterior chamber intraocular lens (ACIOL).

With the advent of the closed automated irrigation vitrectomy system, surgeons' favor either a limbal, anterior, or pars-plana approach for zonular weakness extending more than 9 clock hours. This allows for safe removal of the subluxated lens with meticulous removal of the vitreous gel followed by implantation of a range of IOLs such as angle-supported ACIOL,[18,19] anterior or posterior chamber iris-enclavated IOL,[20-22] or a SFIOL.[23,24] Various surgical techniques for lens extraction have been described that include limbal lensectomy, intra-lenticular bimanual irrigation aspiration (I/A), pars plana lensectomy (PPL), phacoemulsification, femtosecond laser-assisted cataract surgery, and Intracapsular cataract extraction (ICCE) with or without anterior vitrectomy.

Lensectomy

For many years, the preferred surgical approach to manage the dislocated lens in these patients has been standard lensectomy with or without anterior vitrectomy. In modern days, lensectomy has a minimal role and most surgeons prefer lens aspiration in a controlled manner which avoids vitreous loss significantly.

Lens aspiration

Intralenticular bimanual I/A is preferred in cases of the anteriorly dislocated clear lens with or without microspherophakia [Figure 1]. This can be performed using bimanual intracapsular I/A under low-bottle height and a low-vacuum setting either through two small anterior capsulorhexis[25] or two small nicks created in the anterior capsule using a microvitreoretinal (MVR) blade.[26-29] Surgery can be performed under general or local anesthesia depending

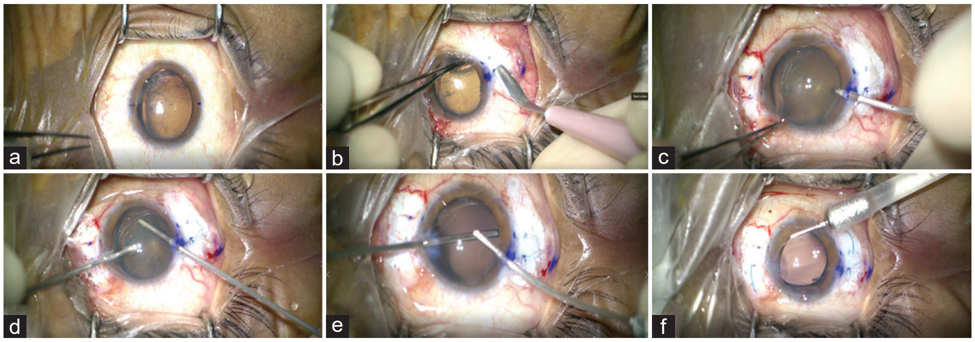

Figure 1: Intralenticular lens aspiration with intrascleral haptic fixation of posterior chamber intraocular lens (PCIOL) in a case of microspherophakia: (a) Intraoperative photograph of miscrospherophakia wherein the lens equator is visible 360°; (b) scleral pockets being created 1.5 mm posterior to the limbus using crescent blade, at 3 and 9 o'clock positions; (c) two stab incisions in the clear cornea and two small nicks in the anterior capsule are created near the lens equator with further insertion of the same microvitreoretinal blade at 10 and 2 o'clock positions; (d) bimanual intra-lenticular irrigation/aspiration is done by introducing the irrigation and aspiration probe through the two different capsular openings; (e) the vitrectomy cutter is used to perform anterior vitrectomy and remove the capsular bag; (f) fibrin glue assisted intrascleral haptic fixation of PCIOL is done under anterior chamber maintainer

on the age and cooperativeness of the patient preferably using a superior approach. Two stab incisions are made in the clear cornea using a 23G MVR blade at 10 and 2 o'clock positions. The anterior chamber is formed with sodium hyaluronate 1.4% and two small capsulorhexis of size 1.5–2.0 mm are made with Utrata forceps.[25] Alternatively, two small nicks can be created in the anterior capsule near the lens equator with further insertion of the same MVR blade.[27] A visco-dispersive substance is injected at the site of lens displacement to prevent vitreous upthrust. A gentle hydrodelineation is performed before commencing lens aspiration to reduce stress over the zonules. Bimanual intra-lenticular irrigation/aspiration is done by introducing the irrigation and aspiration probe through the two different capsular openings. The irrigation port is used to hydrate and stabilize the lens to complete the aspiration under direct visualization. This technique enables lens aspiration within the capsular bag by maintaining the anterior chamber and posterior capsular integrity thereby avoiding lens matter to drift posteriorly into the vitreous cavity.[3] The vitrectomy cutter is used to perform anterior vitrectomy and remove the capsular bag followed by stromal hydration and formation of the anterior chamber. A peripheral iridotomy is optional that can be made with the help of a vitrectomy cutter in I-A cut mode if the patient is left aphakic or implanted with anterior chamber or iris-fixated IOL. There are various advantages of this procedure over the previously described techniques such as less postoperative astigmatism, minimal chance of lens matter drops, and low risk of posterior segment complications.

Pars plana lensectomy

Combined PPL and vitrectomy have been described for removing severely subluxated or posteriorly dislocated crystalline lens. Advantages of the combined approach include a closed ocular surgical system, minimum corneal and iris trauma, good vitreous control, and an easy route to treat the retinal problems if present in the same sitting. The surgical technique consists of a standard three-port pars plana approach 3 mm posterior to the limbus in the superotemporal, superonasal, and inferotemporal quadrants. After securing the infusion cannula through the inferotemporal port, the vitreous surrounding the lens is cleared using a vitrectomy cutter. It might then be replaced by an ultrasonic phaco-fragmentation handpiece to emulsify the lens nucleus (in cases of nuclear sclerosis) followed by induction of posterior vitreous detachment and completion of pars plana vitrectomy. The peripheral retina is meticulously inspected for any pre-existing pathology by scleral indentation and treated if required followed by closure of the sclerotomy ports.

Phacoemulsification

Phacoemulsification is usually preferred in cases where the nucleus is either calcified or dense enough to preclude aspiration through a coaxial or bimanual aspiration system. A clear corneal temporal or superior incision is recommended and care must be taken such that the main incision is not lying directly over the area of zonulopathy wherever feasible. The side port entry should be made more anteriorly to prevent injury to the rhexis margin with the second instrument as the bag tends to shift anteriorly after being tented with iris hooks. Staining of the capsule with trypan blue dye is suggested as it reduces the capsule elasticity thus making penetration of the capsule easier.[30] In some scenarios, immediately after injecting dye, the red reflex can go missing due to vitreous staining. Thus, it is safer to coat a few drops of dye directly onto the anterior capsule under a viscoelastic cover. Due to the absence of zonular counter-traction and the highly elastic nature of the lens capsule, both the initiation and completion of capsulorhexis remain quite challenging and need much expertise. The capsule can be punctured with a standard bent or straight cystotome needle or the crossed-swords capsule pinch technique using two 180° opposing 30 G needle tips can also be used to pierce the capsule. In very unstable lenses, micro-forceps or a hook may be used to provide counter traction during capsulorhexis. The rhexis should be centered onto the lens capsule to ultimately achieve a round, central rhexis. Alternatively, micro-incision forceps can be used to access the capsule from multiple micro incision paracentesis around the circumference. The ideal capsulorhexis size should be such that at least a 2-mm margin between the rhexis edge and the lens equator is present to adequately support the capsular support device in the capsular bag. In cases with moderate-to-severe grades of subluxation, the capsule needs to be supported with flexible iris or capsular retractors during surgery, that are strategically placed through the limbal stab incisions.[31] After stabilization of the capsular bag with capsular supporting devices, phacoemulsification is initiated. Meticulous and repeated hydrodissection and visco-dissection form a crucial step in this technique. Mobilization of the nucleus in the bag is nearly impossible due to a lack of counter traction. Slow-motion phacoemulsification with low parameters is mandatory. If chopping can be successfully initiated, the removal of the first nuclear fragment creates more space in the bag, thereby reducing zonular stress during subsequent chopping. Devices required for long-term bag fixation and implantation of IOLs will be discussed in the further sections.

Femtosecond laser cataract surgery

This evolving technology is gradually gaining popularity and is most valued for the creation of a precise anterior capsulotomy and assisting nuclear fragmentation and softening in cases with ectopia lentis. Various studies have highlighted the reproducibility and accuracy of the capsulotomy size and centration,[32,33] thereby producing a postoperative less lens tilt and optimal visual outcome.[34,35] Despite significant subluxation, femtosecond laser remains independent of the loss of counter resistance from zonular support and enables a circular anterior capsulotomy. The laser platform enables a tremendous degree of safety and control in mobilizing the nucleus in the presence of zonulopathy. Femtosecond laser-assisted cataract surgery can allow phacoemulsification in cases with 160° of zonular dialysis with the benefits of creating minimal zonular impairment and being able to treat corneal astigmatism with relaxing incisions.[36,37] It has the potential to perform a well-centered capsulotomy for subsequent CTR or segment and IOL implantation without decentration in patients with Marfan syndrome.[38]

Intracapsular cataract extraction

ICCE is ideal for the management of more than 9 clock hours of subluxation or anteriorly dislocated lens with nuclear sclerosis. However, it requires a large incision for lens extraction that induces significant astigmatism, vitreous loss, herniation of vitreous in the anterior chamber, intraoperative hypotony, cystoid macular edema, choroid, and retinal detachment. In this technique, the entire cataractous lens with an intact capsule is delivered by rupturing the zonules by either of the following methods such as tumbling technique by giving pressure counter pressure at the opposite ends, cryoextraction (cryoprobe assisted), introducing Arruga's capsule holding forceps or the wire vectis method. This needs to be combined with meticulous anterior vitrectomy [Figure 2].[39]

Role of microscope-integrated optical coherence tomography

The standard procedures described in the previous sections can be performed with ease when the corneal clarity is excellent. However, surgery becomes challenging in the presence of corneal edema and lenticulo-corneal adhesion, and a blind effort to strip the anterior capsule from the corneal endothelium can result in Descemet membrane tear and detachment. In such case scenarios with anterior lens dislocation and lenticulo-corneal adhesion, microscope-integrated optical coherence tomography (MIOCT) has been reported to be enormously beneficial in careful peeling of the adherent anterior capsule. Following complete aspiration of the lens matter and capsular bag, MIOCT-guided identification and removal of anterior capsular tags can be ensured. Goel and Sahay *et al*., in their cases, series have highlighted the importance of MIOCT that helped in the release of lenticulo-corneal adhesions in the presence of corneal edema thus avoiding the use of a vitrectomy cutter that might have damaged the corneal endothelium further.[26,29]

Visual rehabilitation following surgery

Visual rehabilitation following lensectomy in children with

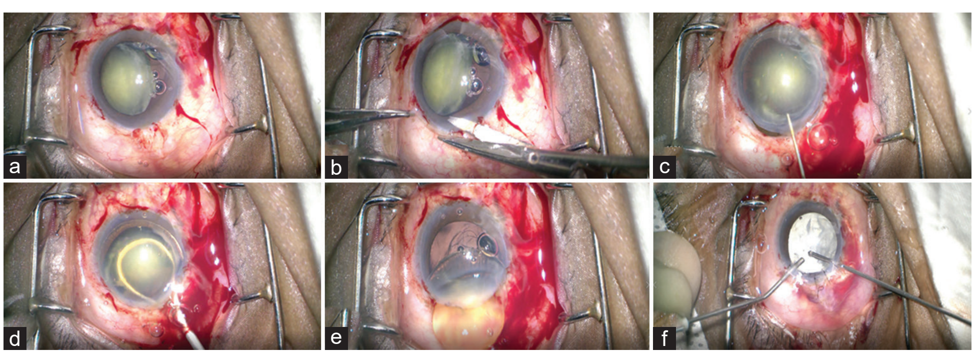

Figure 2: Intracapsular cataract extraction in case of ectopia lentis with cataract: (a) Intraoperative photograph of inferiorly subluxated cataractous lens with superior zonular dehiscence (note the broken zonules in the superior half); (b) temporal incision is being enlarged using corneoscleral scissors; (c) the cataractous lens has been maneuvered into the anterior chamber and viscodispersive agents being injected beneath the endothelium; (d) wire vectis is introduced underneath the lens; (e) lens is being delivered along with it's capsule using counterpressure with the lens hook from the nasal side; (f) After securing the main wound using 10-0 monofilament nylon sutures, triamcinolone assisted anterior vitrectomy is performed

subluxated lens is of paramount importance to prevent amblyopia and to improve their quality of life. Often infants who undergo lens extraction are left aphakic, which if not managed effectively can cause permanent visual disability. Rehabilitation must begin as soon as possible after surgery usually within 1–2 weeks.

In this section, we are going to discuss the different techniques of visual rehabilitation with lens extraction surgery or post-lensectomy aphakia management. With advances in surgical techniques and IOL designs, more options are now available for the placement of IOL in the same sitting after lens removal in patients with insufficient zonular support or subluxation. These options include an ACIOL, iris-fixated IOL (anterior chamber or retro-fixated), placement of an IOL within the capsular bag with the use of a CTR or Cionni ring or capsular tension segments (CTSs), a scleral-fixated PCIOL, and an intrascleral fixated IOL.

Spectacles

Aphakic glasses in the pediatric population are the safest approach and provides consistent visual outcome when IOL implantation is not preferable along with lensectomy. It is important to explain to the parents that spectacles will be required even if IOL is implanted in children after lensectomy. IOL implantation in children is avoided if the age is <2–3 years, the axial length is <17 mm, and WTW is <9 mm or more than 12.5 mm.[40] Pediatric eyes when left aphakic often have a reduced rate of growth or retarded axial length increase as compared to normal or pseudophakic eyes.[41]

Contact lenses

Contact lenses have an important role in the management of childhood aphakia. They are very effective in providing good-quality vision without visual field constriction and image magnification. They are particularly useful in unilateral aphakia to reduce the risk of amblyopia.

Intraocular lens placement

Recently, with improvements in surgical techniques and IOL designs, IOL placement is being performed more frequently as a primary or secondary procedure in pediatric patients with ectopia lentis requiring surgical intervention.

Anterior chamber intraocular lens

ACIOL implantation in ectopia lentis has been described in literature with good results, especially in Marfan's syndrome.[19,42,43] However, the major concerns involve pigment release, unstable IOL, and corneal decompensation. Due to the increased risk of these complications in young children with a long life expectancy, PCIOL implantation is considered a better and safer option. Closed-loop ACIOLs have a high incidence of corneal decompensation and macular edema and are not recommended.[44]

Iris fixated intraocular lens

Posterior chamber iris sutured intraocular lens

This technique has been used in pediatric eyes with a lack of zonular support and usually involves a 3-piece IOL sutured to the iris tissue with 9-0 or 10-0 polypropylene suture.

The iris enclavated IOL is the Artisan IOL developed by Worst in 1978.[45,46] It is a 3-piece rigid polymethyl methacrylate (PMMA) IOL with an anterior vault to provide clearance between the optic and IOL. It is oriented at 3 and 9 o'clock and is fixated at the mid-periphery of the iris with an enclavating needle. Surgical peripheral iridectomy is required to prevent pupillary block.

Anterior fixation

In this technique, the IOL is enclavated anterior to the iris. It is technically easier as compared to SFIOL with a shorter surgical duration. There is a lower risk of posterior segment complications. However, for the insertion of IOL, a 5–6 mm incision is required in all cases.

Posterior/retro pupillary fixation

Artisan or Verisyse iris-claw IOL can also be fixed behind the pupil to minimize the risk of endothelial cell damage. However, the technique is challenging and the outcomes in pediatric eyes with subluxation have rarely been reported.[47]

Intrascleral fixation of intraocular lens

Scleral fixation of IOLs has been the preferred technique for visual rehabilitation in children with severe subluxated lenses where preservation of capsular bag might not be possible.

Sutured scleral fixated intraocular lens

In pediatric eyes, due to the increased risk of suture breakage (24%), erosion (0–28.5%) and IOL subluxation (6.3%), retinal detachment (6.3%), vitreous hemorrhage (4.8%), large incision for IOL insertion (5–6 mm), this technique is rarely used nowadays.[45,48]

Sutureless intrascleral fixation of intraocular lens

This has been recently popularized since it eliminates the suture-related complications, reduces the risk of postoperative inflammation and glaucoma, and requires only a small 2.8–3 mm incision for foldable IOL insertion as compared to sutured SFIOL. This can be further divided into glued IOL technique described by Gabor *et al*., and popularized by Agarwal *et al*.,[49,50] and glueless sutureless SFIOL, described by Gabor *et al*., and then later modified by Yamane *et al*., and Walia *et al*.,[50-52] The outcomes of the above-described techniques are mentioned in Table 1.

SFIOL implantation is especially advantageous in eyes with low endothelial cell count, peripheral anterior synechiae, shallow anterior chamber, glaucoma, young patients, and those with absent iris tissue. This technique also has a lower risk of corneal endothelial damage, peripheral anterior synechiae, and glaucoma.[61,67] Moreover, SFIOL is placed at a more physiological position near the nodal point of the eye, thus retaining binocularity due to minimal aniseikonia. There is minimal uveal contact thus lower risk of iris pigment dispersion and glaucoma. The use of fibrin glue to allow adhesion of the scleral flap also enhances the closure of sclerotomy site thereby reducing the risk of

Table 1: Review of literature for scleral fixated intraocular lens in ectopia lentis

Author, year	Technique	Sample size and groups	Main outcome measures	Results	Complications	Conclusion
Kim, 2010[53]	Transscleral sutured IOL fixation without vitrectomy	45 eyes, 23 patients with ectopia lentis due to Marfan's syndrome Age (3–31 years) Follow-up duration up to 20 months	-	≥2 lines improvement in BCVA No vitreoretinal complications	Raised IOP (12/45 eyes) Transient pupillary capture (6/45 eyes)	This technique is safe and effective technique for eyes with ectopia lentis
Sen *et al*., 2018[54]	Sutured SFIOL with PPV (10-0 prolene)	279 eyes of 230 patients (38.7% eyes with ectopia lentis) Age (3.5–18 years) Follow-up (1–10 years)	Visual outcome and complications	BCVA improved in 97.3% eyes at 6 weeks	Serous CD (2.86%), vitreous hemorrhage (2.86%), endophthalmitis (0.72%), diplopia (0.72%), RD (5.73%), and dislocation of the SFIOL (4.7%), raised IOP (12.54%)	Sutured SFIOL is safe and effective to correct aphakia in children
Sen *et al*., 2020[55]	Sutured SFIOL with PPV (10-0 prolene)	73 eyes, 43 patients with Marfan's syndrome Age (3.5–18 years) Follow-up (6 weeks–1 year)	-	Visual acuity improved with reduction in refractive error significantly at 1 year	Raised IOP (4.1%), IOL subluxation (6.8%), RD (4.1%), CD (1.3%), pupillary capture (20.5%), vitreous hemorrhage (1.3%)	SFIOL provides good visual outcomes in eyes with ectopia lentis (Marfan's syndrome)
Kumar *et al*., 2012[56]	Glued SFIOL	41 eyes, 33 children (9/33 had ectopia lentis) Age (5–15 years) Follow-up (1–3 years)	Visual outcome, endothelial count, complications	BCVA improvement of >1 line in 53.6% Significant reduction in refractive error Mean EC loss 4.13%	Optic capture (2.4%), macular edema (4.8%), and IOL decentration (4.8%) No vitreoretinal complications, no iris damage	-
Balakrishnan *et al*., 2020[57]	Role of surgical PI in glued SFIOL and PPL	34 eyes (15 with PI and 19 without PI), 17 children with ectopia lentis Age (3.5–15 years) Mean follow-up 25.4 months	Incidence of optic capture, secondary glaucoma, IOL dislocation, or repeat surgery	Significant improvement in BCVA and refractive error	Significantly lower incidence of complications in PI group	Surgical PI in glued SFIOL surgery in children undergoing PPL reduces the optic-capture related complications
Rastogi *et al*., 2020[58]	Glued SFIOL with PPL	45 eyes, 44 children with ectopia lentis Age (6–18 years) Follow-up duration (upto 1 year)	Visual outcome and IOL tilt with UBM	Significant improvement in BCVA with reduction in astigmatism	Significant IOL tilt (>5°) in 5/45 eyes Mean ECL 3.6% RD in 2/45 eyes No IOL decentration no pseudo-phacodonesis	SFIOL is safe and effective in children Caution required in eyes with retinal degenerations and WTW>12 mm
Nb *et al*., 2018[59]	Glueless, flapless SFIOL (Gabor technique) Retrospective	40 eyes, 25 children Age (6–18 years) Follow-up (1–5 years)	Visual outcome and complications	Significant improvement in BCVA and refractive error	Transient vitreous hemorrhage and hypotony (2.5%) Hyphaema (10%) IOL subluxation (2.5%)	-
Sternfeld *et al*., 2020[60]	Glueless, flapless SFIOL (Yamane technique) Retrospective	12 eyes, 10 children (6 ectopia lentis) Mean age (10 years) Mean follow-up 8 months	Visual outcome and complications	Significant improvement in BCVA and refractive error	IOL decentration (1/12 eyes)	This technique can be adapted safely in pediatric eyes
Boral and Agarwal, 2020[61]	Glueless, flapless SFIOL (technique) Retrospective	81 eyes, 73 patients (13 patients with ectopia lentis) Age (4–78 years) Follow-up (6–51 months)	Visual outcome and complications IOL stability using ASOCT and UBM	Significant improvement in BCVA and astigmatism Stable and centered IOL with no tilt in all cases	IOL haptic dislocation (2/81 eyes) with Marfan's No other complications	-
Yen *et al*., 2009[62]	Iris sutured PCIOL Retrospective	17 eyes, 12 children (11 eyes with ectopia lentis) Age (2–15 years) Follow-up (upto 38 months)	Visual outcome and complications	Significant improvement in BCVA in all eyes	IOL dislocation (29%) with intact sutures Iris capture of IOL (1/17) RD (1/17)	Iris-fixated IOLs are reasonable alternative to sutured SFIOL in pediatric eyes

Contd...

Table 1: Contd...

Author, year	Technique	Sample size and groups	Main outcome measures	Results	Complications	Conclusion
Shah *et al*., 2016[63]	Iris sutured PCIOL Retrospective	17 eyes, 12 children (11 eyes with ectopia lentis) Age (2–15 years) Follow-up (upto 9 years)	Visual outcome and complications	Significant improvement in BCVA in 71% eyes	IOL dislocation (41%) with intact sutures (higher rate of 71% in ectopia lentis) RD (1/17)	Iris-fixated IOLs should be used with caution in pediatric eyes
Dureau *et al*., 2006[21]	Iris sutured PCIOL Retrospective	17 eyes, 9 children (6 patients with Marfan's) Age (2–10 years) Follow-up (up-to 25 months)	Visual outcome and complications	Significant improvement in BCVA in all eyes	Hyphema with pupil ectopia (1/9 patient) Aspetic endophthalmitis (1/17 eyes)	Iris sutured IOL has the advantage of small incision and the absence of transscleral sutures as compared to sutured SFIOL
Cleary *et al*., 2012[64]	Artisan anterior iris claw IOL	8 eyes, 5 children with ectopia lentis Age (3–16 years) Follow-up (4–58 months)	Visual outcome and complications	Significant improvement in BCVA in all eyes	Mean ECL 14.2% No other complications	Artisan iris-claw IOL is safe and effective in children with ectopia lentis
Català-Mora *et al*., 2012[65]	Artisan anterior iris claw IOL with pars plana cannulas	10 eyes, 5 children (4 with ectopia lentis) Age (3–16 years) Follow-up (up-to 12 months)	Visual outcome and complications	Significant improvement in BCVA in all eyes	Mean ECL 4.7% at 12 months Vitreous hemorrhage (3/10 eyes) Pigment dispersion (2/10 eyes)	Artisan iris-claw IOL is safe and effective in children with ectopia lentis
Gonnermann *et al*., 2013[66]	Posterior Verisyse iris claw IOL Reterospective cohort	16 eyes, 10 patients with Marfan's syndrome Age (9–61 years) Follow-up (6–74 months)	Visual outcome and complications	Significant improvement in BCVA in all eyes	Mean ECL 5.6% Early hypotony (15.4%) Persistent pupil ovalization (7.7%) Retinal detachment (7.7%)	Posterior iris claw IOL has good outcomes in eyes with insufficient zonular support

ASOCT: Anterior segment optical coherence tomography, BCVA: Best corrected visual acuity, CD: Choroidal detachment, EC: Endothelial cell, ECL: EC loss, IOL: Intraocular lens, IOP: Intraocular pressure, PCIOL: Posterior chamber IOL, PI: Peripheral iridotomy, PPL: Pars plana lensectomy, PPV: Pars plana vitrectomy, RD: Retinal detachment, SFIOL: Scleral fixated IOL, UBM: Ultrasound biomicroscopy, WTW: White to white

hypotony and infection. IOL tilt and pseudophakodonesis are less in sutureless glued IOL as compared to sutured SFIOL since there is only one point fixation with sutured SFIOL leading to contact of IOL with iris and iris chafing.[67] Major complications include vitreous hemorrhage, lens tilt and decentration, hypotony, secondary glaucoma, hyphema, suprachoroidal hemorrhage, choroidal effusion, cystoid macular edema, retinal detachment, and external suture erosion.

Endocapsular support devices

Endocapsular devices or CTRs are useful in eyes with compromised zonular integrity to support and centralize the capsular bag and allow for the placement of IOL in the bag. These rings are produced by Morcher Gmbh, in Stuttgart, Germany, and are made of PMMA.[68] These can be inserted into capsular bags at any point after a continuous curvilinear capsulorhexis (CCC) has been completed.

Major advantages of endocapsular devices include preservation of capsular bag, less disturbance of vitreous base, and less risk of vitreous hemorrhage and hypotony however the associated drawbacks include posterior capsular opacification, persistent iritis with prolonged steroid therapy, raised IOP, pseudophacodonesis, late IOL decentration and tilt, broken suture and need for repeat surgery, and retinal detachment.

Various commonly used devices include standard CTR, MCTR or Cionni ring, CTS. The choice of which one to use depends on the degree and the likely progression of zonular damage. In this review, we are only discussing the management of ectopia lentis in children that including mainly progressive subluxation causes.

Standard capsular tension ring

It works by redistribution of existing zonular forces and maintaining the circular diameter of the capsular bag. The standard CTR is made of PMMA material and has an oval-shaped cross-section with eyelets at both free ends. It is a compressible circular ring with two smooth-edged end terminals.

- *Prerequisites:* complete and intact CCC with an intact posterior capsule
- *Contraindications:* if CCC is not attained or posterior capsule rent is present. CTR is also not indicated in progressive subluxation like ectopia lentis.

Modified capsular tension ring

This ring is useful in extensive zonular deficiency (more than 3 clock hours or 4–9 clock hours) or progressive subluxation like ectopia lentis by allowing the surgeon to anchor the capsular bag to the sclera. It has a unique fixation hooklet designed for scleral fixation without violating the integrity of the capsular bag.

- *Prerequisites:* complete, central, and intact CCC with an intact posterior capsule
- *Contraindications:* if CCC is not attained or posterior capsule rent is present.

Capsular tension segment

This is a partial ring with 90°–120° circumference and has a radius of 5mm along with an anteriorly positioned fixation eyelet similar to M-CTR. This is useful in eyes with profound zonular insufficiency and in progressive subluxation. It provides support in the transverse plane when sutured to the scleral wall. It is often combined with CTR or MCTR to provide a circumferential support with an extra suture fixation in a particular quadrant. Cortical removal is also easier with CTS as compared to CTR.

Common limitations in the current literature investigating IOLs for correcting aphakia are small sample size, retrospective design, and lack of comparative/control data. Various approaches have been described in literature, however, till date, there is no evidence as to which of these is the best approach for pediatric ectopia lentis. It depends on the surgeon's expertise and discretion whether the capsular bag needs to be preserved or compromised.

Whatever may be the mode of postoperative visual rehabilitation, all such cases must be evaluated for coexistent amblyopia and treated accordingly. All such patients may have some underlying systemic disorders which may be life-threatening that should always be kept in mind while managing such cases.

Conclusion

The major challenge is to identify the best method of visual rehabilitation in ectopia lentis. Although the treatment with capsular bag fixation with in-the-bag IOL implantation offers the best option, it may not always be possible. Most of the visual rehabilitation methods are useful in relatively elderly children but, in those below 6 years, all these methods have their own limitations to label as gold standard treatment.

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Conflicts of interest

There are no conflicts of interest.

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