Three-Year Outcomes of Cionni-Modified Capsular Tension Ring Implantation in Children Under 8 Years Old With Ectopia Lentis

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• PURPOSE: This study evaluated visual outcomes and complications at 3 years post-implantation of a Cionni-modified capsular tension ring (MCTR) with an intraocular lens (IOL) in ectopia lentis patients ≤8 years old.

• DESIGN: Prospective clinical cohort study.

• METHODS: Included were 101 eyes from 57 patients < 8 years of age, who underwent surgery for nontraumatic ectopia lentis between November 2015 and December 2016. Exclusion criteria were planned IOL fixation in the ciliary sulcus, severe intraoperative complications, and incomplete follow-up. All eyes received in-the-bag implantation of a posterior IOL and Cionni-MCTR. Posterior capsulectomy and anterior vitrectomy were performed through the pars plana in 23 eyes of children < 5 years of age. Patients were examined at 1 day, and 1, 6, and 12 months, and at 2 and 3 years postoperatively. Outcome; measurements included best-corrected visual acuity (BCVA), intraocular pressure (IOP), IOL centration, and posterior capsule opacification (PCO).

• RESULTS: In all eyes, BCVA improved significantly after surgery, especially during the first 12 months ($P < .05$). Three years post-operatively, 44 eyes had BCVA 0.9 or better. Prophylactic Nd:YAG laser capsulotomy was performed 3 months post-surgery in 24 eyes; 34 eyes underwent this; procedure 6 months post-surgery because of PCO. A second surgery was; warranted in 4 eyes because of severe IOL decentration and combined anterior capsule contraction. No severe postoperative complications, such as retinal detachment or endophthalmitis, occurred.

• CONCLUSIONS: Implantation of in-the-bag IOL with Cionni MCTR is effective for visual rehabilitation in young children with ectopia lentis. A close follow-up of these patients is necessary to monitor IOL centration and stability. **(Am J Ophthalmol 2021;224:74–83.**

CONGENITAL CRYSTALLINE LENS SUBLUXATION IN children is mainly associated with hereditary metabolic disorders such as Marfan syndrome, homocystinuria, Weill-Marchesani syndrome, and idiopathic ectopia lentis.[1–4] Displacement of the crystalline lens commonly induces severe refractive error and anisometropia. If left untreated, children may be at a high risk of developing severe amblyopia.

However, severe zonular laxity increases the difficulty of cataract extraction and intraocular lens (IOL) implantation, especially in children, as well as the risk of severe complications such as glaucoma or retinal detachment.[5] Moreover, children's eyes have unique features, such as a high incidence of postoperative posterior capsule opacification (PCO), pronounced inflammation, probability of progressive zonular laxity, changing globe dimensions, and longer life expectancy.[6–9] Therefore, it is necessary to choose the optimal surgical method to keep the IOL stable and minimize the risk of surgery in ectopia lentis. A capsular tension ring has been used to improve the stability of implantation of the IOL in the capsule.[10–13] However, the capsular tension ring does not provide adequate support for the lens capsule in the presence of extensive zonular laxity, which is often the case in ectopic lenses.[12,13] As early as 1998, the Cionni-modified capsular tension ring (MCTR) was used in treating the severe zonular weakness for the first time by Cionni and Osher.[14]

The Cionni-MCTR provides better stability for the lens capsule without compromising the performance of the capsule. However, few case studies[15,16] have documented the use of Cionni-MCTR in young children with ectopia lentis. Thus, the study intended to prospectively analyze 3-year visual outcomes and complications in children 8 years old or younger with ectopia lentis after implantation of Cionni-MCTR combined with an IOL.

METHODS

• SUBJECTS: In this prospective study, children 8 years of age or younger with ectopia lentis (more than 90 degrees of lens subluxation) who received a sclera-fixated Cionni-

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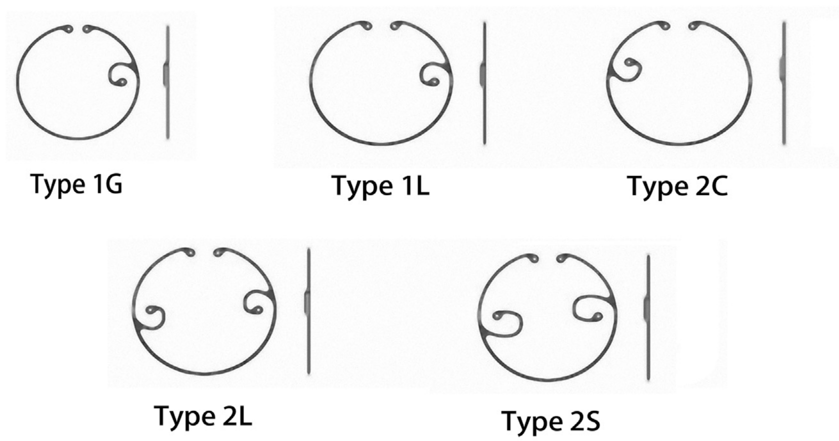

	Type 1G	Type 1L/ Type 2C	Type 2L	Type 2S
Size (open)	12.0 mm	13.0 mm	13.0 mm	13.0 mm
Size (compressed)	10.0 mm	11.0 mm	11.0 mm	11.0 mm
Zonular Damage	> 4 hours (120°)	> 4 hours (120°)	> 6 hours (180°)	> 6 hours (180°)
ACCC*	7.0 mm	7.0 mm	7.0 mm	5.0 mm
Suturable arms	One	One	Two	Two
Material	Clear PMMA	Clear PMMA	Clear PMMA	Clear PMMA

ACCC*: suggested size of anterior continuous curvilinear capsulorhexis

FIGURE 1. Characteristics of different types of Cionni-modified capsular tension ring during surgery. ACCC = anterior continuous curvilinear capsulorhexis.

MCTR in Eye and Ear, Nose, and Throat Hospital of Fudan University between November 2015 and December 2016 were included. This study was approved by the institutional review board of Eye, Ear, Nose and Throat Hospital of Fudan University. The principles of the Declaration of Helsinki were followed, and informed consent was obtained from the guardian of each patient.

Patients received in-the-bag implantation of posterior chamber IOLs with a sclera-fixated Cionni-MCTR (types 1G, 1L, 2C, 2S, or 2L; Morcher GmbH, Stuttgart, Germany) (Figure 1). Patients were not enrolled if severe intraoperative complications such as an irregular anterior capsular tear or posterior capsular rupture occurred that prevented the MCTR from being implanted. Patients were also excluded for incomplete follow-up.

Preoperative evaluation included thorough anterior and posterior segmental examinations, which mainly involved slit-lamp examination, intraocular pressure (IOP) measurement (if cooperative in the clinic), slit-lamp indirect ophthalmoscopy, and B-scan ultrasonography. The dilated slit-lamp examination assessed the severity and degree of lens subluxation. Most children who were cooperative during the examination were assessed using IOLMaster 500 (Carl Zeiss AG, Oberkochen, Germany) and Pentacam (Oculus Optikgeräte GmbH, Wetzlar, Germany) for IOL calculation. In children who were unable to cooperate with the measurement, IOL calculations were conducted preoperatively with the patient under general anesthesia, using immersion A-scan ultrasonography for axial length (AL) measurements and a hand-held keratometer for keratometry.

• SURGICAL TECHNIQUE: All surgeries were performed with the patients under general anesthesia by 1 of the 3 surgeons (Y. L., Y. J., or J. Y.). Patients were dilated preoperatively with a combination of tropicamide 1% and phenylephrine 5% eye drops. Each eye received a sclera-fixated Cionni-MCTR and an in-the-bag IOL, as previously described.[17] Posterior capsulectomy and anterior vitrectomy were performed through the pars plana in children

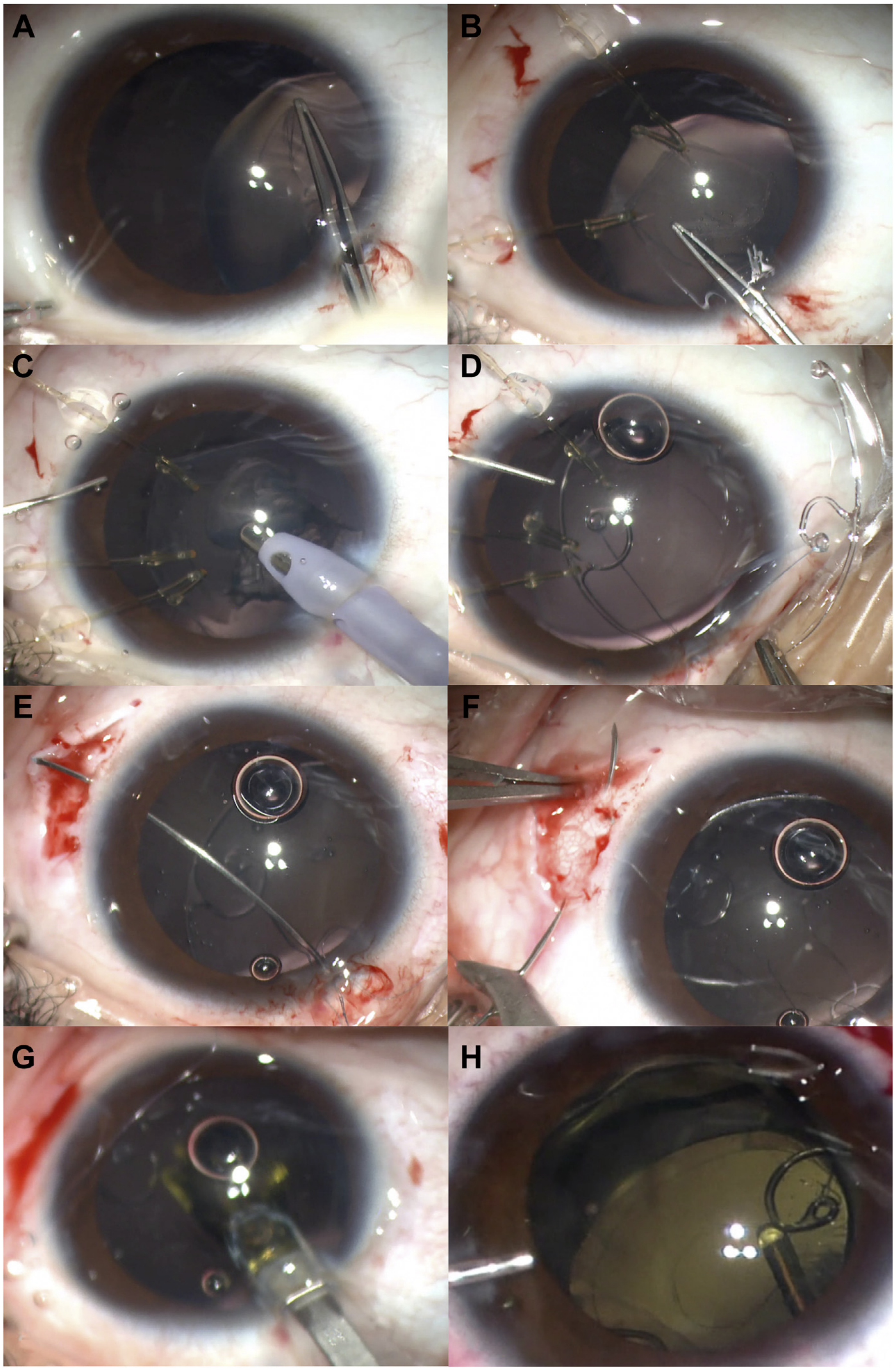

FIGURE 2. (A) Partial capsulorhexis was performed in the area of zonular dehiscence. (B) Two capsular hooks were used to fix the capsulorhexis edges and continue the capsulorhexis. (C) Lens aspiration was performed at low-flow rates. (D) Cionni- MCTR using 10-0 polypropylene looped sutures and a long, curved needle was inserted into the capsular bag. (E) The 2 needles were inserted through the main incision and passed between the iris and the anterior capsule and then outside of the scleral wall, approximately 1-1.5 mm posterior to the corneal limbus. (F) The eye underwent nonscleral flap suture fixation of MCTR implantation through the ciliary sulcus using the Z-suture knotless technique. (G) A single-piece hydrophobic acrylic IOL was implanted in the capsular bag. (H) After the main corneal incision was closed with 10-0 sutures, posterior capsulectomy and anterior vitrectomy were performed through the pars plana. IOL = intraocular lens; MCTR = modified capsular tension ring.

under 5 years of age. In children who were 5 years or older, those who were cooperative received prophylactic Nd:YAG laser capsulotomy between 1 and 3 months after surgery. If the best-corrected visual acuity (BCVA) and the severity of lens dislocation in both eyes of 1 child were similar and both fit the surgical indication, the second eye was scheduled for an operation 1 to 2 weeks after the first one, to be done by the same surgeon. If only 1 eye of the child met the inclusion criteria at the first outpatient visit, close follow-up was advised after the first surgery. Surgery for the second eye was deferred until it met the study's surgical indication criteria.

After the corneal incision, continuous curvilinear capsulorhexis (CCC) and hydrodissection were performed. Implantation of MCTR was not done if a severe anterior capsular tear occurred during CCC, and the IOL was fixed in the ciliary sulcus by transscleral sutures. Capsular bag stabilizing hooks (Mackool cataract support system; Duckworth and Kent, Hertfordshire, UK) were introduced through additional paracenteses in all cases. In the cases in which the lens dislocation was beyond 180° and severely off center, the method of 2-step capsulorhexis was used: after partial capsulorhexis was done in the area of zonular dehiscence (Figure 2, A), 1 or 2 capsular hooks were used to hang the capsulorhexis edges to centralize and stabilize the lens and continue the capsulorhexis (Figure 2, B). The size of the CCC was 3.5-4.5 mm, according to the severity of lens dislocation. Lens aspiration was performed under a low-flow rate with an infusion bottle at a height of 70-80 cm (Figure 2, C). The choice of MCTR was determined through intraoperative assessment by the surgeon based on the extent of zonular weakness and the age of the child.

A single-eyelet MCTR was implanted when the zonular loss or weakness was between 90 and 180°. In contrast, a double-eyelet MCTR was chosen when there were more than 180 degrees of zonular loss or weakness. Because only rings with 1 eyelet have a smaller size (eg, type 1G), children 5 years of age or younger with zonular loss or weakness between 90 and 180° were received MCTR type 1G implants. The technique described by Cionni and Osher[14] was used in which a single-armed 10-0 polypropylene suture with a long straight needle (AUM-5 and SC-5; Alcon, Fort Worth, Texas, USA) or a 10-0 polypropylene looped-suture with a long-curved needle (PC-9; Alcon) was threaded through the eyelets of the MCTR, and the MCTR was then inserted into the capsular bag (Figure 2, D). The needle was then inserted through the main incision, passing through the space between the iris and anterior capsule and finally out of the scleral wall, 1-1.5 mm posterior to the corneal limbus. As described by Szurman and associates,[18] an MCTR was fixed through the ciliary sulcus using the zigzag-shaped intrascleral suture (Z-suture) (Figure 2, E and F).

In those patients who received single-eyelet MCTR, the sutures were placed in the area of maximum zonular weakness. A single-piece hydrophobic acrylic IOL (ZCB00; Abbott Medical Optics, Santa Ana, California, USA; or SN60WF; Alcon) was implanted into the capsular bag (Figure 2, G), confirming the centration of the IOL. IOL power was implanted with predicted error of +4 diopters (D) in 3 years, +3 D in 4 years, +2 D in 5 years, +1 D in 6 years, and 0 D in 7-8 years. After the corneal incision was closed with 10-0 sutures, a posterior capsulectomy of 3-4 mm and anterior vitrectomy were performed through the pars plana using 25-gauge instrumentation with high cutting frequency (3,000 to 4,000 cuts per minute) and low aspiration pressure (−300 to −350 mm Hg) in children younger than 5 years old (Figure 2, H). One 25-gauge trocar cannula system was placed 2.5-3.0 mm from the limbus at 11 o'clock to introduce the 25-gauge vitreous cutter. The irrigation port was placed in the anterior segment.

Postoperative prescribed medications included topical steroids (1% prednisolone acetate starting at 4 times daily and tapered to 1 drop every week for 4 weeks), topical nonsteroidal anti-inflammatory drugs (0.1% pranoprofen, 4 times a day for 4 weeks), and antibiotic eye drops (0.5% levofloxacin, 4 times a day for 2 weeks).

• **OUTCOME MEASUREMENTS:** Patients were examined at 1 day, then 1, 6, and 12 months, and at 2 and 3 years post-operatively. The post-operative outcome measurements included BCVA, IOP, IOL centration, PCO, secondary procedure rates, and other complications.

• **STATISTICAL ANALYSIS:** Changes from mean pre-operative to post-operative BCVA were analyzed using Wilcoxon signed-rank test. A *P* value of less than .05 was considered statistically significant. Descriptive statistics and Wilcoxon signed-rank tests were used to support our findings and to compare between subgroups.

RESULTS

A TOTAL OF 131 EYES IN CHILDREN 8 YEARS OF AGE OR younger underwent surgery for ectopia lentis during the study period. Fourteen children (17 eyes) were excluded due to the planned lensectomy with or without IOL fixation in the ciliary sulcus by transscleral sutures because the crystal dislocation was too severe to implant MCTR. Seven patients (7 eyes) who received lensectomies with IOL fixation by transscleral suturing in the ciliary sulcus were excluded from the study because of severe intraoperative complications, such as an irregular anterior capsular tear or posterior capsular rupture. Six patients (6 eyes) were excluded due to incomplete follow-up. Thus, 101 eyes (57 pediatric patients) were included. The average age at surgery was 6.1 years (3-8 years). The mean value of pre-operative BCVA was 0.68 ± 0.31 logMAR, and pre-operative spherical equivalent was −9.17 ± 5.40 D (range +6.50 to −22 D). Detailed individual patient data are shown in Table 1. A total of 47 eyes (46.5%) had congenital or idiopathic ectopia lentis not associated with Marfan syndrome, whereas 48 eyes (47.5%) had Marfan-associated ectopia lentis. Six eyes (5.9%) had

TABLE 1. Preoperative and Intraoperative Characteristics	
Number of Patient Eyes	57 (101)
Females/males	29/28
Diagnosis	
Marfan syndrome	47
Congenital or idiopathic	48
Spherophakia	6
Age at surgery, y	
3~4	23
5~6	33
7~8	45
Mean ± SD axis length, mm	24.14 ± 2.44
Mean ± SD preoperative spherical equivalent, D	−9.17 ± 5.40
Mean ± SD preoperative BCVA, logMAR	0.68 ± 0.31
MCTR Type	
1g	45
1l	12
2c	17
2l	1
2s	26

BCVA = best-corrected visual acuity; MCTR = modified capsular tension ring.
These data are from subjects who completed the study.

spherophakia. The extent of subluxation ranged from 90 to 270°, except for the eyes diagnosed with spherophakia, of which the dislocation range was 360°.

All eyes had an in-the-bag posterior chamber IOL and received a Cionni-MCTR implant. Forty-four eyes (43.6%) were implanted with double-eyelet MCTR, and 57 eyes (56.4%) received a single-eyelet MCTR. Twenty-three eyes received posterior capsulectomy and anterior vitrectomy. During surgery, iris prolapse occurred in 2 eyes, hyphema in 1 eye, and vitreous prolapse in 3 eyes. Twenty-four eyes received preventive Nd:YAG laser capsulotomy between 1 and 3 months after surgery. In the other 55 eyes, Nd:YAG laser capsulotomy was performed if PCO occurred during follow-up.

The mean values of BCVA preoperatively, 1 day, 1, 6, and 12 months, and 2 and 3 years after surgery were 0.68 ± 0.31, 0.46 ± 0.22, 0.33 ± 0.17, 0.22 ± 0.14, 0.16 ± 0.13, 0.10 ± 0.09, and 0.10 ± 0.09 logMAR, respectively. Post-surgical BCVA improved significantly in all patients, especially during the first 12 months. There were no statistically significant differences between the mean BCVA values in eyes across different groups of causes ($P < .05$) (Figure 3, A). At the 3-year follow-up, the number of eyes with BCVA ≤0.5, 0.6-0.8, and ≥0.9 were 4 (4.0%), 53 (52.5%), and 44 (43.6%) logMAR, respectively (Figure 3, B). Table 2 shows the total changes in the refraction of the eyes in different age groups after IOL implantation. The differences in total changes in refraction were both significant among different age groups ($P < .05$).

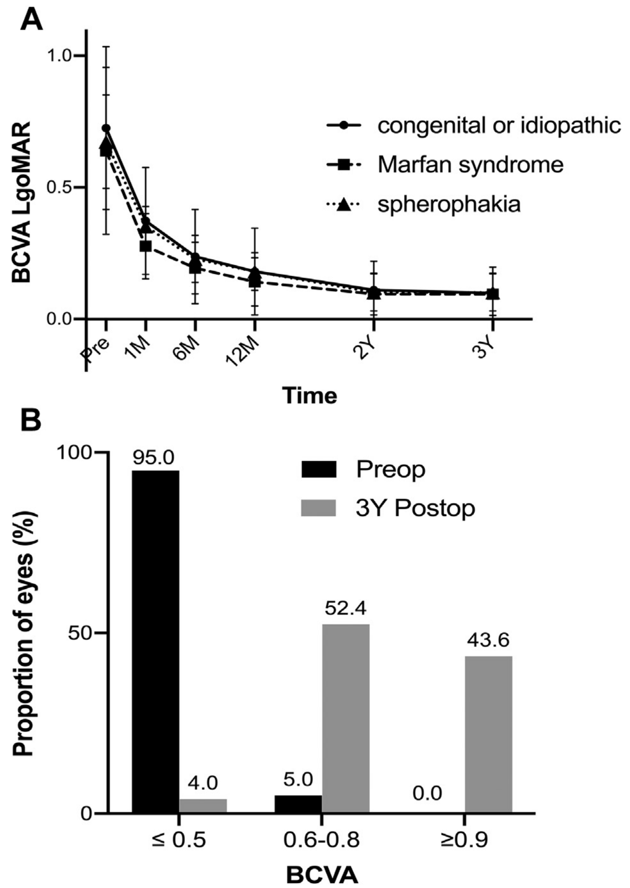

FIGURE 3. Changes in BCVA from baseline to 3 years postoperatively. (A) The temporal dynamic of BCVA in eyes at different time points indicated a significant improvement after cataract surgery during the 3-year follow-up. (B) The changes in proportion of the eyes with BCVA, ≤0.5, 0.6-0.8, and ≥09 preoperatively versus 3 years postoperatively show significant improvement at the final follow-up. BCVA = best-corrected visual acuity.

All IOLs were centered at 1 day and 1 month after surgery (Figure 4, A, B, and C).

Elevated IOP was reported 2 weeks after surgery in 3 eyes and was treated with a single topical antiglaucoma medication. Mild vitreous hemorrhage was observed 1 day postsurgery in 2 eyes and resolved in 1 week. Thirty-four eyes (33.7%) required Nd:YAG laser capsulotomy because of significant PCO between 6 months and 3 years after surgery (Figure 4, D and F). Seven eyes required Nd:YAG laser capsulotomy more than once because of the recurrence of visual axis opacification (VAO) or moderate anterior capsule contraction, among which 4 eyes had received prophylactic Nd:YAG laser capsulotomy within 3 months after the surgery. None of the children under 5 years old developed VAO during follow-up (Figure 4, E). Of those eyes that did not receive posterior capsulectomy and anterior vitrectomy or prophylactic Nd:YAG laser capsulotomy, 30 of 55 eyes (54.5%) required YAG laser

TABLE 2. Refractive Outcomes in Different Age Groups

Age at Surgery, y	n Eyes	Mean ± SD SE at 1 month Postoperative (D)	Mean ± SD SE at 3 y Postoperative (D)
3-4	23	+3.57 ± 2.13	+1.42 ± 1.29
5-6	33	+1.97 ± 1.87	+0.58 ± 0.97
7-8	45	+0.34 ± 0.96	−0.09 ± 1.01

SE = spherical equivalent.

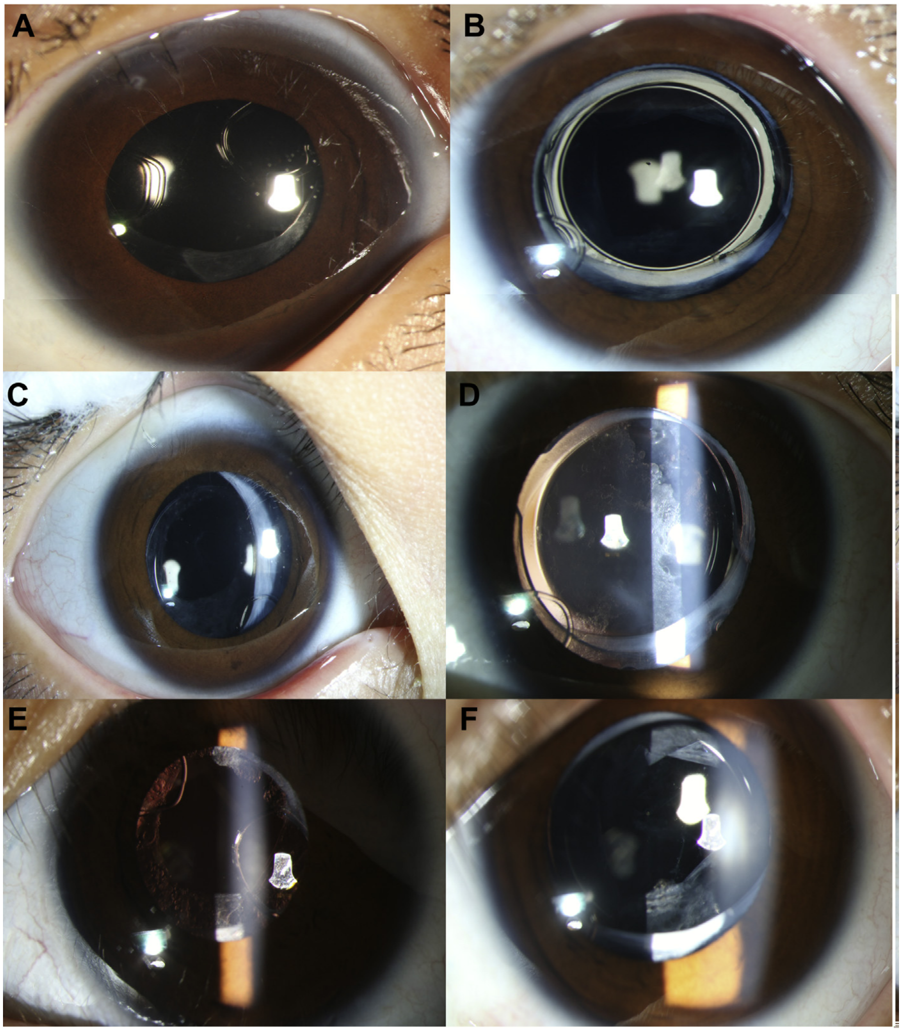

FIGURE 4. (A) Intraocular lens (IOL) and double-hook Cionni-modified capsular tension ring (MCTR) were centered at 1-month follow-up. (B) IOL and single-hook MCTR were centered at the 3-year follow-up. (C) Slight decentration of the IOL and MCTR but not vision-threatening. (D) Significant PCO was observed at 1-year follow-up in an eye without posterior capsulotomy and anterior vitrectomy during surgery. (E) Clean visual axis was observed in the eye with posterior capsulectomy and anterior vitrectomy. (F) The eye received posterior capsulotomy at 1 month after surgery due to the development of PCO but received Nd:YAG laser posterior capsulotomy again because of developing the PCO at 2-year follow-up. IOL = intraocular lens; MCTR = modified capsular tension ring; PCO = posterior capsule opacification.

Case	Diagnosis	Age (y)	AL (mm)	Pre- BCVA	3y Post- BCVA	Interval between 2 Operations (months)	Primary Surgery	Complications or Comments
1	Marfan syndrome	3	32.08	0.05	0.4	6	I/A + MCTR + IOL + PC + AV	IOL decentration + anterior capsule contraction/IOL and MCTR repositioning and vitrector-cut capsulotomy
2	Marfan syndrome	4	29.18	0.25	0.5	7	I/A + MCTR + IOL + PC + AV	IOL decentration/IOL and MCTR repositioning
3	Congenital or idiopathic	6	27.82	0.1	0.7	18	I/A + MCTR + IOL	IOL decentration/IOL exchange, sutured in the ciliary sulcus by transscleral sutures
4	Spherophakia	5	21.88	0.1	0.8	8	I/A + MCTR + IOL	IOL decentration + anterior capsule contraction/IOL and MCTR repositioning and vitrector-cut capsulotomy

TABLE 3. Summary of the Patients With Dislocated IOL

AV = anterior vitrectomy; AL = axial length; I/A= irrigation-aspiration; IOL = intraocular lens; MCTR = modified capsular tension ring; PC = posterior capsulectomy.

capsulotomy because of PCO. Iris posterior synechiae occurred in 3 eyes due to postoperative chronic inflammation, which was managed with topical steroids and mydriatic drugs.

Severe IOL decentration with or without anterior capsule contraction was observed in 4 eyes with vision impairment between 6 and 18 months post-surgery. A summary of the patients with dislocated IOL is provided in Table 3. Type of IOL, sex, axial length (AL), and suture location did not appear to be associated with an increased risk of dislocation. All 4 eyes were diagnosed with lens dislocation of more than 180° before the initial operation. Of these, 3 eyes received surgical IOL and MCTR repositioning with or without vitrector-cut anterior capsulotomy (Figure 5). The other eye underwent IOL exchange due to the serious dislocation of the IOL and a capsular tear that occurred during resuturing. The MCTR-IOL-capsular complex was replaced by a new IOL fixed in the ciliary sulcus by transscleral sutures. There were no serious complications such as endophthalmitis or retinal detachment in any pediatric patient during surgery or follow-up.

DISCUSSION

THE VISUAL OUTCOMES AND COMPLICATIONS WERE PROspectively studied in children with ectopia lentis at 8 years of age or younger, a younger age group than those in most previous studies. Furthermore, considering the high rate of PCO in young children, the surgical method of posterior capsulectomy and anterior vitrectomy after MCTR and IOL implantation were tried in children younger than 5 years old for the first time.

Ninety-nine eyes (98.0%) had BCVA of 0.5 or better at 3 years of follow-up, which was better than that of most previous reports.[16,19–21] In an earlier report from Vasavada and associates,[21] only 45.7% of the eyes achieved a visual acuity of 0.5 or better in children 2.5 to 18 years of age. Konradsen and associates[16] reported a postoperative BCVA of 0.5 or better in 25 eyes (67.6%) after Cionni-MCTR implantation in pediatric patients younger than 11 years old with ectopia lentis. Kim and associates[22] reported a BCVA of 0.5 or better in 18 eyes (94.7%) in children 18 years of age or younger using capsular tension segment and the Cionni-MCTR. We believe the most important reason behind the superior postoperative BCVA in the present study is that these patients received surgery at a very young age. Early surgical treatment potentially gives better visual outcomes in young children. It is believed that there is a declining plasticity of the central nervous system as the children age. Treatment for amblyopia is more likely to succeed if it is initiated at a younger age. Moreover, treatment of amblyopia can be initiated only after correcting refractive errors, and children older than 8 years are less responsive to treatment. As a result, under the premise of surgical safety, the earlier the surgical correction of refractive error that facilitates the initiation of amblyopia training the better the anticipated visual rehabilitation.[23,24]

Another potential reason for the good visual rehabilitation in the present study is the low rate of PCO (33.7%) due to the posterior capsulectomy and anterior vitrectomy done in the first surgery and subsequent prophylactic Nd:YAG laser capsulotomy in most of the children. In pediatric patients, VAO is almost inevitable when cataract surgery is performed without posterior capsulectomy and anterior vitrectomy.[9] Vasavada and associates[21] reported that PCO developed in 21 eyes (60%), of which 19 (54.27%) needed an Nd:YAG capsulotomy or a pars plana

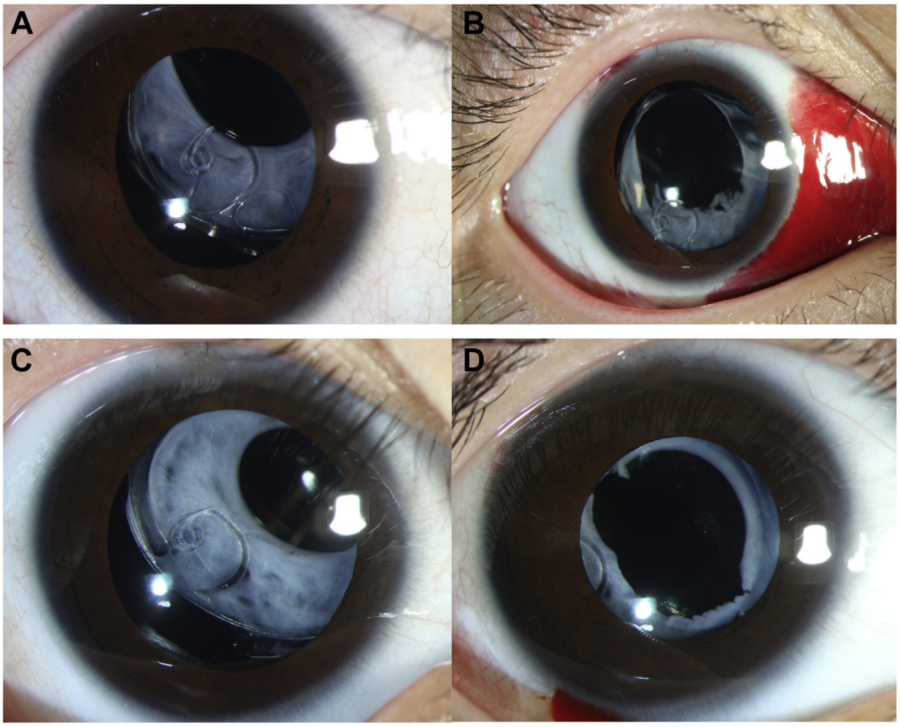

FIGURE 5. (A) One eye developed severe IOL decentration and vision impairment at 1-year follow-up. (B) One day after receiving surgical IOL/Cionni- MCTR repositioning and vitrector-cut capsulotomy. (C) One eye developed severe IOL decentration and anterior capsular contraction at 1-year follow-up. (D) One week after receiving surgical IOL and MCTR repositioning and vitrector-cut capsulotomy. IOL = intraocular lens; MCTR = modified capsular tension ring.

membranectomy to clear the visual axis in children with ectopia lentis during an average follow-up of 28 months. In another study, the PCO rate was 84% after CTR and in-the-bag IOL implantation in children with ectopia lentis.[16] In the present study, no opacification of the visual axis occurred after surgery in children who had undergone posterior capsulectomy and anterior vitrectomy during surgery. In addition, no irregular posterior capsule tears occurred during this procedure in the present study. All these outcomes confirmed the safety and effectiveness of performing the posterior capsulectomy and anterior vitrectomy through pars plana after the main corneal incision, sealed with a stable anterior chamber, with high cutting frequency (3,000 to 4,000 cuts per minute) and low aspiration pressure (−300 to −350 mm Hg). It is also safe and effective for eyes with posterior capsular left intact to receive prophylactic Nd:YAG laser posterior capsulotomy within 3 months after cataract surgery. Children may need more than 1 Nd:YAG laser posterior capsulotomy to clear the VAO. In all cases, the MCTR and IOL remained stable after the posterior capsulectomy or Nd:YAG laser posterior capsulotomy. In addition, it has been shown that the use of capsular tension rings itself may influence capsule opacification formation.[25]

In the present study, all patients were initiated on amblyopia training 1 month after surgery. Most of the children received optical correction only. Treatment with 2 hours of prescribed daily patching for 6 months was applied to 2 children who experienced incomplete improvement with eyeglasses alone, and the differences in BCVA of whom was beyond more than 2 lines between fellow eyes. Some children also received vision therapy as an adjunct, which consisted of computer programs and eye-hand coordination exercises. It is imperative to recognize postoperative complications on time and ensure amblyopia treatment and regular, close follow-up requiring cooperation from the parents. These authors plan to conduct educational sessions with the children's parents and regularly remind them of their follow-ups by phone calls to ensure that the parents bring their children to the hospital on time.

Four eyes received a second surgery due to the severely off-center IOL-MCTR capsular complex. One possible reason could be the anterior capsular contraction. The extremely small diameters of the anterior capsulorhexis increase the risk of anterior capsular contraction. In addition, in eyes with a very small capsulorhexis margin, the hook of MCTR may drag on the capsulorhexis edge and result in iris chaffing, pigment dispersion, and chronic uveitis. Achieving a continuously centered CCC of adequate size (4-4.5 mm) is crucial, not only for safe lens removal but also for successful MCTR and IOL implantation.[26] However, in eyes with an unstable lens, especially with a dislocation of more than 180°, capsulorhexis may be more difficult. In the present study, following centration of the lentis with capsular hooks, surgeons adopted the 2-step capsulorhexis to ensure the size of the capsulorhexis. In order to avoid anterior capsular contraction, prophylactic Nd:YAG laser anterior capsulotomy had been performed 1 to 3 months after surgery in eyes with very small anterior capsulorhexis.

Those 4 eyes were operated on in the early period of the study, during which a single-strain 10-0 polypropylene suture was used. It might have been due to the poor strength of the single-strand line in fixing the MCTR which caused postoperative IOL dislocation in children with severe lens dislocation. No IOL dislocation was found in eyes in the later period when the 10-0 polypropylene looped sutures were used. It is believed that the loop sutures would last longer due to less tension per strand over time with the double-stranded technique. There was no suture breakage in either suture variant. However, the follow-up period of 3 years was relatively short compared to that in the studies which observed spontaneous suture breakage and late IOL dislocation. Price and associates[27] microscopically studied the sutures of 5 dislocated IOLs and found evidence of microscopic degradation of the 10-0 polypropylene suture 7 to 14 years after implantation. Cases of suture breakage in eyes implanted with MCTR were reported with the use of both 10-0 and 9-0 polypropylene sutures.[21,28] It is believed that the 9-0 polypropylene suture has higher tensile strength[27] and was recommended for all MCTR patients, especially in young people. In a few studies, 8-0 polytetrafluoroethylene (W.L. Gore and Associates, Flagstaff, Arizona, USA) has been used. It is believed to provide better strength and stability; however, its use is off-label now. In the present study, 2 kinds of sutures were used: a single-armed 10-0 polypropylene suture and 10-0 polypropylene looped-suture. Considering its stability, the loop suture was used in this study instead of the single-armed suture when it became available. By using the loop suture, each ring of MCTR was fixed onto the sclera with 2 sutures. The authors believe that the loop 10-0 polypropylene suture can be as strong as the 9-0 polypropylene. A longer follow-up would be necessary to elucidate the probability of late IOL dislocation in 5 to 10 years when using 10-0 polypropylene looped sutures.

All children in this study were 3 years of age or older. Generally, the younger the child is, the more difficult the operation will be. Because children under 3 years old have smaller eyes and a smaller operating field, surgical procedures, such as capsulorhexis, implantation of the capsular hook, and implantation of the capsular tension ring are more difficult. In addition, the size of the eyeballs of children under 3 years of age increases rapidly, and their refractive results may be less predictable. Also, the growth of the eyeballs increases the suture tension of the fixed tension ring, increasing the probability of suture breakage.

The limitations of the current study include, first, the relatively short follow-up period and lack of a control group. Future prospective studies with a longer follow-up, such as 7 to 10 years of age, are needed to establish the long-term safety of the MCTR in pediatric eyes. Moreover, comparing the differences in visual outcomes and complication rates between the surgical technique used here with other techniques, such as sclera-fixated IOLs and iris-fixated IOLs, will require a large-scale prospective randomized clinical trial. Second, no cases of homocystinuria or other systemic diseases, which are known to be associated with ectopia lentis, were diagnosed in this series. Our hospital laboratory cannot test for it at present, and there may be a small number of cases missed because of this. We will make corrections in our future studies to reduce the probability of a missed diagnosis. Third, aspherical IOLs with negative spherical aberrations were used in this study. However, studies showed that the IOLs with greater aspherical properties incorporated in the eye were more sensitive to decentration and tilt of the IOL.[29] In eyes where the probability of IOL displacement is increased, it would be better to avoid choosing the aspherical IOLs with negative spherical aberration.

In conclusion, in-the-bag IOL implantation combined with Cionni-MCTR can effectively improve the visual rehabilitation in young children with ectopia lentis. All of the eyes showed significant improvement in BCVA in our study, and 43.6% reached a visual acuity of 0.9 or better. The visual outcomes were better in our study than those of most previous reports. One of the most important reasons was that ametropia was surgically corrected during the sensitive stage of amblyopia treatment. Posterior capsulectomy with anterior vitrectomy and preventive Nd:YAG laser capsulotomy could safely and effectively prevent PCO. The rate of IOL decentration (3.96%) was comparable to the rate of similar studies in pediatric patients. However, considering the possibility of a second operation, regular, long-term follow-up care is crucial to monitor and ensure IOL centration and stability.

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