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Comparison of the Cionni ring and Yamane techniques for intraocular lens implantation in pediatric Marfan syndrome patients with lens subluxation

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Abstract

Background To compare visual, refractive, and anatomical outcomes of lens extraction with Cionni ring and in-the-bag intraocular lens (IOL) implantation and Yamane intrascleral fixation in pediatric Marfan patients with lens subluxation.

Methods This retrospective cohort study included 21 patients (29 eyes) who underwent IOL implantation for ectopia lentis due to Marfan syndrome. Surgical techniques included Yamane sutureless scleral fixation and scleral fixation with Cionni capsular rings. Postoperative outcomes assessed were corrected visual acuity (CVA) in decimal, spherical equivalent (SE) refraction, lenticular astigmatism, IOL tilt/decentration, endothelial cell count, and complications.

Results Sixteen eyes were included in the Yamane group and 13 in the Cionni group. Both groups demonstrated a significant improvement in CVA compared to preoperative values (Yamane, $p=0.001$; Cionni $p=0.002$). A significant postoperative improvement in SE was also noted in Yamane group ($p=0.009$). The degree of IOL tilt was comparable between groups (Yamane, 0.80 ± 0.27°; Cionni, 1.33 ± 0.93°; $p=0.240$); however, IOL decentration occurred in 4 eyes in the Cionni group and 3 eyes in the Yamane group. No major intraoperative complications were reported. Among the postoperative complications, retinal pathology and glaucoma each occurred in 2 patients in the Yamane group. The median postoperative follow-up duration was 16.6 (min-max: 7–34 months) months in the Modified Yamane group and 22.6(min-max: 8–35 months) months in the Cionni group ($p=0.232$).

Conclusions In patients with lens subluxation secondary to Marfan syndrome, IOL implantation using either the Yamane technique or the Cionni ring yields favorable anatomical and functional outcomes. These methods may be considered viable options following thorough preoperative assessment, particularly when the risk of intraoperative and postoperative complications is deemed low.

Keywords Marfan syndrome, Ectopia lentis, Cionni capsular tension ring, Yamane, Lens subluxation

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Background

In pediatric patients, zonular deficiency and lens subluxation may arise from a variety of systemic conditions such as Marfan syndrome, Ehlers-Danlos syndrome, Weill-Marchesani syndrome, hyperlysinemia, and sulfite oxidase deficiency, and can also result from trauma [1, 2]. Although trauma is the most common cause of ectopia lentis overall, Marfan syndrome represents one of the most common systemic causes of non-traumatic ectopia lentis in pediatric patients, with ectopia lentis present in approximately 50–80% of cases and often serving as the initial ocular manifestation [3, 4]. Other ocular findings include high myopia, retinal detachment, glaucoma, early-onset cataract, and strabismus [5, 6]. In such cases, ensuring a clear optical axis during the critical period of visual development and managing zonular instability to prevent complications are of paramount importance.

The management of zonular weakness and lens subluxation in children is challenging due to unique pediatric tissue responses, increased postoperative inflammation, and concerns about long-term IOL stability related to their longer life expectancy [7–9]. Multiple strategies have been used to manage zonular weakness and lens subluxation, each with its own advantages and associated risks [10]. In mild cases, sulcus placement with capsular tension rings or Cionni rings allows for in-the-bag IOL implantation. In more advanced subluxation with inadequate capsular support, scleral fixation, with or without sutures is preferred. Sutured methods carry the risk of late suture erosion, while sutureless techniques such as the Yamane technique offer alternatives, but long-term data in pediatric eyes are limited and have a steeper learning curve [11, 12].

This study aims to compare the anatomical and functional outcomes, following IOL implantation using capsular bag fixation with a Cionni ring versus intrascleral fixation with a Yamane flange in pediatric patients with lens subluxation due to Marfan syndrome.

Materials and methods

This retrospective observational study included pediatric patients who underwent IOL implantation due to lens subluxation with Marfan syndrome at a tertiary ophthalmology center between 2020 and 2024. Ethical approval was obtained from the local ethics committee prior to the study, which was conducted in accordance with the tenets of the Declaration of Helsinki. A total of 29 eyes from 21 patients with Marfan syndrome were included. The study population included patients who underwent lens extraction, followed by IOL implantation using two techniques: unilateral Cionni capsular tension rings (CTR) with single-piece IOL placement in the capsular bag, and scleral fixation of a 3-piece IOL via Modified Yamane technique. All patients underwent lens extraction and IOL implantation in a single surgical session under general anesthesia. Anterior vitrectomy was performed in patients with preoperative vitreous presence. Patients who underwent pars plana lensectomy, pars plana vitrectomy at the same session or before, or lacked consistent follow-up measurements were excluded.

Preoperative data included patient age, duration of postoperative follow-up, presence of amblyopia or strabismus, axial length (AL), and any associated retinal or anterior segment pathologies. Visual acuity was recorded as uncorrected (UDVA) and corrected (CVA), using age-appropriate methods—Snellen charts for older children, and noted as "not applicable (N/A)" for children under four years of age. Eyes in which preoperative visual acuity could not be reliably measured and was recorded as 'not applicable (N/A)' were excluded from visual acuity–related analyses but were included in all other outcome analyses. Snellen measurements were then converted to logMAR. Preoperative refractive error (spherical and cylindrical components) were also recorded. Postoperative refraction was performed according to the AAO guidelines for age-based residual refraction [13]. In this case, an under-correction of approximately 20% of the calculated emmetropic IOL power was selected, given the expected myopic shift with age. IOL power calculation was performed using the SRK/T formula based on measurements obtained with the OA-2000 (Tomey Corporation, Nagoya, Japan) optical biometer.

All surgeries were performed under general anesthesia by experienced pediatric cataract surgeons. The surgical technique was selected based on capsular integrity and capsulorhexis adequacy. In eyes with an intact capsular bag and a well-centered continuous curvilinear capsulorhexis of approximately 5.0 mm, a Cionni ring with in-the-bag single-piece IOL implantation was preferred. When these conditions could not be achieved, intraocular lens fixation was performed using the Modified Yamane Technique.

Modified Yamane technique [14–17]

All surgeries were performed by the same surgeon under general anesthesia. The Yamane flanged haptic intrascleral fixation technique was performed as previously described. But the technique was slightly modified. A trocar was inserted through the sclera at the inferior nasal quadrant for pars plana infusion. The main incision was placed through the upper temporal quadrant (135°). With the help of the Mendez ring, the sclerotomy marks were made exactly 180 degrees apart (45° and 225°). Sclerotomy sites were planned 2 mm behind the limbus for patients aged 6 years and older, while for patients younger than 6 years, a 2 mm tunnel was marked 1.5 mm behind the limbus. The entire capsule was removed so that no capsular fragments were left behind, and the surgical field was prepared with an anterior vitrectomy. Through a 2.8–3.0 mm main incision, the three-piece IOL (Sensar AR40e, J&J Surgical Vision INC) was implanted into the anterior chamber. With the help of forceps, the leading haptic was inserted into a 27-gauge needle which was placed transconjunctivally through the sclera at the pre-marked area. The trailing haptic was then inserted into the second needle. First, the trailing haptic was pulled out from the needle and held 1 mm back to form a flange. Then the leading haptic was pulled out and the flange was formed by holding it 1 mm back. A disposable low-temperature cautery pen (Bovie, USA) was used to create the flange. The IOL was centralized, and the haptics were embedded in the sclera [18]. Trocar was removed and sutured with 7.0 vicryl. The main incision was sutured with 10.0 nylon suture and removed in the 1st month of the operation.

Capsular bag fixation with Cionni ring [15, 19]

A localized conjunctival peritomy was performed at corresponding to the area of zonular dialysis where the Cionni CTR was to be implanted. A 2.2 mm temporal clear corneal incision was made, and viscoelastic material was injected into the anterior chamber and the area of dialysis to tamponade the anterior vitreous. Capsulorhexis was completed with cystotome and forceps, using capsular retractors in cases of significant zonular instability. In all cases, after aspiration of the lens material via bimanual irrigation and aspiration, a single Cionni CTR segment was used for dialysis ≤ 200°. One end of a double-armed 10 – 0 polypropylene suture was tied to the Cionni CTR hook and inserted into the capsular bag. The suture needle attached to the hook was passed through the sclera 1.5 mm posterior to the limbus, at the center of the dialysis area. The fixation ring were positioned in the region of maximal zonular weakness, ensuring anterior placement relative to the capsulorhexis margin, and secured to the sclera using a Z-suturing technique. Prior to this, a conjunctival peritomy was performed in all patients in the area where the Z-suture technique would be applied. A single-piece hydrophobic acrylic monofocal IOL (SA60AT, Alcon) was implanted in the bag, and all incisions were closed appropriately.

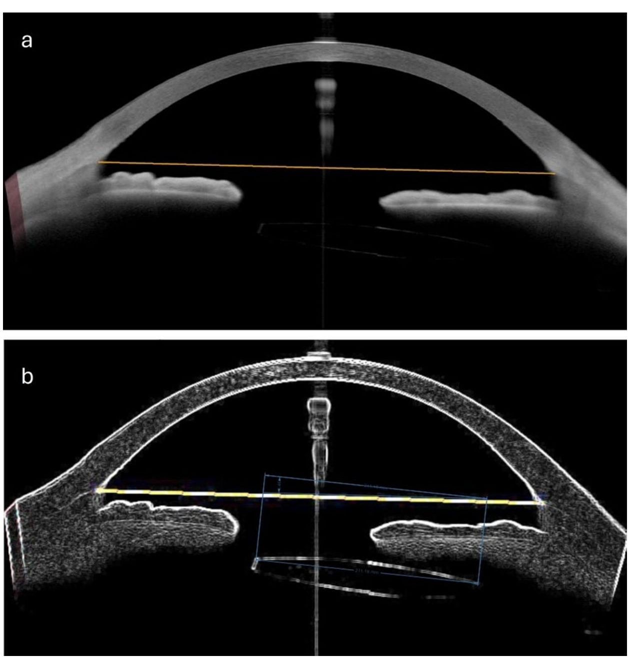

Fig. 1 **a**: Anterior segment OCT slice obtained with anterion (0-180 degrees). Orange line; spur-to-spur line (reference line) determined by the software. **b**: IOL edges were made more distinct with Image J program. Blue line on the IOL; manually drawn in CorelDRAW Graphics Suite 2024 software and determining the IOL axis. The angle between the parallel line drawn to this line with the CorelDraw program and the orange reference line was calculated with the angle calculation module

All intraoperative complications such as vitreous hemorrhage or difficulties in IOL fixation were reviewed from surgical reports. Postoperative management included topical antibiotics and corticosteroids, tapered gradually over several weeks. Follow-up visits were conducted on postoperative day 1, week 1, and month 1, and every 3 months thereafter. At the final follow-up; UDVA, CVA, manifest refraction, intraocular pressure, detailed anterior segment and dilated retinal examination findings were recorded. Lenticular astigmatism was calculated by applying power vector analysis to the difference between refractive and corneal astigmatism, with corneal data derived from the Sirius topography system (Costruzione Strumenti Oftalmici, Firenze, Italy).

Central macular thickness was evaluated using optical coherence tomography (Topcon DRI OCT Triton Swept Source), and specular microscopy data (Tomey EM-4000, Tomey Corporation, Nagoya, Japan) were collected pre- and postoperatively. Postoperative complications such as pupillary abnormalities, IOL decentration, glaucoma, retinal detachment or tears, and cystoid macular edema (CME) were documented, along with the need for any additional surgical interventions. The decentration and tilt of the IOL were assessed at 6 months using Anterion (Heidelberg Engineering Inc., Heidelberg, Germany) SS-OCT. The 0-180 degree meridian was used from these images. The spur-to-spur line determined by the software was accepted as a reference. A parallel line was drawn from one end of the IOL to the other end with CorelDRAW Graphics Suite 2024, and its angle with the spur-to-spur line was calculated (Fig. 1). IOL decentration was measured as the horizontal distance between the IOL axis and the center of the pupil. The IOL axis was defined as a straight line through the midpoint of the IOL, whereas the visual axis was determined as the line connecting the anterior corneal apex to the pupil center (Fig. 2). To ensure reliability and repeatability, all

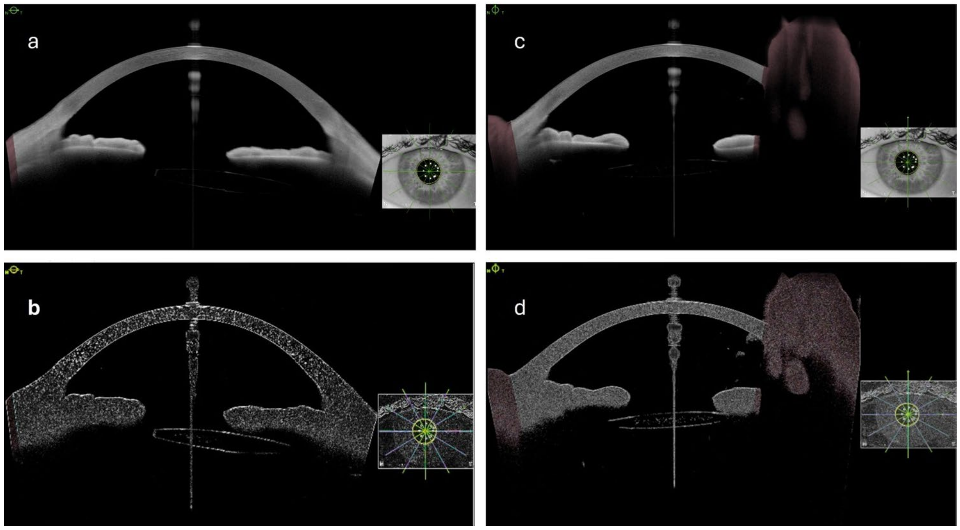

Fig. 2 **a**: Anterior segment OCT slice obtained with anterion (0-180 degrees). **b**: Optimized 0-180 degree Anterior segment OCT segment obtained with Anterion to better observe IOL decentralization. **c**: Anterior segment OCT slice obtained with Anterion (90–270 degrees). **d**: Optimized 90–270 degree Anterior segment OCT segment obtained with Anterion to better observe IOL decentralization

Table 1 Demographic characteristics of the study groups

Mean ± SD (Min-max)	Modified Yamane (n = 16)	Cionni (n = 13)	p
Age (years)	10.0 ± 5.8 (3–18)	10.6 ± 5.2 (3–18)	0.812
Sex (Male/Female)*	13/3	9/4	0.667
Laterality (Unilateral/Bilateral)*	9/7	9/4	0.48
Follow-up duration (months)	16.6 ± 12.2 (7–34)	22.6 ± 10.9 (8–35)	0.232

Differences were considered statistically significant at $p < 0.05$

* This was evaluated using the chi-square test

images were analyzed by a single clinician using a same methodology.

Statistical analysis was performed using the Statistical Package for the Social Sciences (SPSS) version 27.0. Descriptive statistics were presented as mean and standard deviation. The Wilcoxon signed-rank test was applied for within-group comparisons of paired samples, while the Mann-Whitney U test was used to compare quantitative variables between two independent groups.

Results

A total of 29 eyes from 21 pediatric patients were included in the study. Sixteen eyes (55.1%) underwent IOL implantation using the Modified Yamane technique, while 13 eyes (44.8%) received in-the-bag IOL implantation with a Cionni ring. The mean age at the time of IOL implantation was 10.0 ± 5.8 (min-max:3–18) years in the Yamane group and 10.6 ± 5.2 (min-max:3–18) years in the Cionni group, with no statistically significant difference between the two groups (p = 0.812). The median postoperative follow-up duration was 16.6 (min-max: 7–34 months) months in the Modified Yamane group and 22.6 (min-max: 8–35 months) months in the Cionni group (p = 0.232). The demographic characteristics of the two groups are summarized in Table 1.

Intraocular lens implantation resulted in a significant overall improvement in visual acuity across the cohort. Among the 15 eyes in the Modified Yamane group with measurable preoperative CVA was 0.86 ± 0.32 (min-max:0.3–1.3), compared to 0.78 ± 0.32 (min-max: 0.52–1.3) in the 12 eyes in the Cionni group. At the final follow-up, CVA improved to 0.31 ± 0.24 (min-max:0.05-1.0) in the Yamane group and 0.24 ± 0.28 (min-max:0.0–1.0) in the Cionni group, representing a statistically significant improvement in both groups (Modified Yamane: p = 0.001; Cionni: p = 0.002). At the final visit, 11 eyes (73.3%) in the Modified Yamane group and 10 (76.9%) in the Cionni group achieved a CVA of > 20/40. Eyes that failed to reach this threshold typically had amblyopia, most often due to long-standing unilateral visual deprivation. Notably, eyes with bilateral lens subluxation, generally achieved better visual outcomes than unilateral cases (Table 2).

Table 2 Visual and functional characteristics of patients diagnosed with unilateral or bilateral amblyopia

	Age	Unilateral/ Bilateral	Surgical procedure	Corneal astigmatism (D)	Lenticular astigmatism (D)	SE (D)	UDVA (logMAR)	CVA (logMAR)	IOL tilt (°)	IOL decentralization	Strabismus
1	3	U	Cionni	-2.18	-2.5	-2.75	1.3	1.0	1.29	-	
2	16	U	Yamane	-1.62	-0.75	-0.50	1.3	1.0	0.44	-	
3	18	U	Cionni	-1.22	-2.5	-0.75	1.3	1.0	0.69	+	XT
4	18	B-L	Yamane	-3.88	-0.75	-0.25	1.0	0.3	0.51	-	
5	6	B*-R	Yamane	-2.24	-1.25	-1.0	0.3	0.3	0.81	-	
6	6	B*-L	Yamane	-2.67	-3.75	0.25	0.39	0.39	1.07	-	
7	11	U	Cionni	-1.38	-3.75	-0.75	1.3	0.22	0.97	+	
8	3	U	Cionni	-4.0	-2.25	-1.5	1.3	0.39	0.79	-	ET
9	12	U	Yamane	-4.18	-0.50	-1.25	1.3	1.3	1.05	-	XT
10	6	B*-R	Yamane	-1.92	-2.25	-1.5	1.0	0.3	1.09	-	
11	6	B*-L	Yamane	-1.5	-2.25	-3.5	0.69	0.3	1.33	-	
12	5	U	Cionni	-1.22	-0.5	-0.50	1.3	1.0	0.8	+	XT
13	15	U	Cionni	-2.45	0	1.0	0.69	0.39	0.81	-	
14	18	U	Yamane	-1.27	-2.0	-3.0	1.3	0.52	3.19	-	ET

*Patients who underwent surgery on both eyes are indicated

UDVA: uncorrected distance visual acuity, CVA: corrected visual acuity, IOL: intraocular lens, SE: spherical equivalent, XT: exotropia, ET: esotropia

The mean preoperative spherical equivalent (SE) was +4.85 ± 5.06D (min:-2.0 max:14.25) in the Yamane group, and 0.73 ± 8.52D (min:-14.75 max:16.0) in the Cionni group. At final follow-up, the mean SE was −0.70 ± 1.84D (min:-3.5 max: 2.75) in the Yamane group and −0.39 ± 1.48D (min:-2.75 max:3.25), in the Cionni group (Yamane, $p = 0.009$; Cionni, $p = 0.638$). Visual and refractive outcomes for both groups are summarized in Table 3. The mean postoperative lenticular astigmatism was −1.31 ± 0.89D (min:-3.75 max: -0.25) in the Yamane group and −1.33 ± 1.08D (min:-3.75 max:0) in the Cionni group ($p = 0.846$).

IOL decentration was observed in 7 out of 29 eyes: 3 out of 16 eyes (10.3%) in the Modified Yamane group, 4 out of 13 eyes (13.7%) in the Cionni group. However the majority were mild and did not significantly affect visual outcomes, nor did they require urgent surgical intervention. Reoperation for IOL repositioning was required in only 1 of the 13 eyes (7.6%) treated with the Cionni technique due to visual acuity worse than 20/40. The mean lenticular astigmatism in patients with IOL decentration was −1.82 ± 1.27, while the mean CVA was 0.61 ± 0.03. In all patients, the mean IOL tilt angle was 0.80 ± 0.27° in the Yamane group and 1.33 ± 0.93° in the Cionni group ($p = 0.240$).

No major intraoperative complications were reported in any of the cases. Both groups showed a postoperative decrease in endothelial cell count compared to preoperative measurements, but no significant difference was observed between the two groups ($p = 0.056$). The median endothelial cell loss was −215 cells/mm² (min −495, max −41) in the Modified Yamane group and −193.5 cells/mm² (min −346, max −6) in the Cionni ring group, with no statistically significant difference between the two techniques ($p = 0.260$). Various postoperative complications occurred, particularly in eyes with more complex clinical profiles. These are summarized in Table 4. Retinal complications were observed in 2 out of 16 eyes (6.8%) in the Modified Yamane group during follow-up. In the Modified Yamane group, one eye (axial length: 25.05 mm) developed a retinal tear, while another eye (axial length: 22.22 mm) developed rhegmatogenous retinal detachment with macular involvement. The median change in central macular thickness was 30.0 μm (min −28, max 89) in the Modified Yamane group and 12 μm (min −17, max 39) in the Cionni group, and this difference was not statistically significant ($p = 0.131$). However, clinically significant cystoid macular edema developed in one patient in the Modified Yamane group.

Postoperative secondary open angle glaucoma developed in the eyes of two different patients (6.8%) who received a Modified Yamane-fixated IOL in the postoperative third month. Both glaucoma cases were successfully managed with topical medical therapy alone, and no surgical intervention was required. No cases of postoperative corneal decompensation or permanent intraocular inflammation, such as chronic uveitis, were observed. In one Modified Yamane case, early revision surgery was required due to conjunctival haptic exposure with Seidel positivity; a scleral pocket was created and the haptic was trimmed. In another Modified Yamane case, optical capture through the iris occurred as a result of optic-haptic disenclavation, necessitating a secondary Yamane procedure; in this case, a significant postoperative decrease in endothelial cell was observed due to contact between the IOL optic and the corneal endothelium (Fig. 3).

Table 3 Comparison of demographics, preoperative and postoperative values between the two groups

	Median (Min-max)	Modified Yamane (*n* = 16)	Cionni (*n* = 13)	*p*
UDVA‡	*preoperative*	1.3 (0.3,1.3)	0.91 (0.52,1.3)	0.792†
	postoperative	0.52 (0.1, 1.3)	0.39 (0.0,1.3)	
	p	0.003*§	0.022*§	
CVA‡	*preoperative*	0.69 (0.3,1.3)	0.69 (0.52,1.3)	1.0†
	postoperative	0.3 (0.05,1.0)	0.22 (0.0,1.0)	
	p	0.001*§	0.002*§	
Spherical equivalent	*preoperative*	+ 3.74 (-2.0,14.25)	-1.21 (-14.75,16)	
	postoperative	-0.62 (-3.5,2.75)	-0.50 (-2.75, 3.25)	
	p	0.009*§	0.638§	
Lenticular astigmatism		-1.12 (-3.75, -0.25)	-1.25 (-3.75,0)	0.846
IOL tilt angle		0.87 (0.44–1.07)	1.03 (0.69–3.19)	0.240
Axial length (mm)		24.45 (22.1–26.3)	23.37 (21.0-25.67)	0.699
Endothelial cell density (cell/mm^2)	*preoperative*	3369.0 (2851–4025)	2901.0 (2455–3493)	0.056†
	postoperative	3102.0 (2047–3684)	2714.0 (2487–3265)	
	p	0.003*§	0.026*§	
Central macular thickness (μm)	*preoperative*	238.5 (198–292)	282.0 (187–409)	0.809†
	postoperative	242.5 (201–272)	263.0 (166–329)	
	p	0.441	0.386	

UDVA: Uncorrected distance visual acuity, CVA: Corrected visual acuity, IOL: intraocular lens

* Differences were considered statistically significant at $p < 0.05$

† The difference (Δ) between preoperative and postoperative values was compared using the Mann–Whitney U test

§ The relationship between preoperative and postoperative values was evaluated using the Wilcoxon paired signed-rank test

‡ Data from 15 patients were analyzed in the Yamane group and from 12 patients in the Cionni group

Table 4 Postoperative complications and interventions in both groups

Post-operative Complications	Modified Yamane (*n* = 16)	Cionni (*n* = 13)
IOL dislocation/decentration	3 (10.3%)	4 (13.7%)
Amblyopia/Strabismus*		
Unilateral	3 (10.3%)	6 (20.6%)
Bilateral	5 (17.2%)	0 (0%)
Glaucoma	2 (6.8%)	0 (0%)
Cystoid macular edema	1 (3.4%)	0 (0%)
Retinal complications (tear/detachment)	2 (6.8%)	0 (0%)
Pupil malformation	6 (20.6%)	5 (17.2%)
Re-operation	3 (10.3%)	**1 (7.6%)**

***** Bilateral amblyopia was defined as a corrected visual acuity of 20/40 or worse in both eye, while unilateral amblyopia was defined as a difference of two or more lines in visual acuity between the two eyes

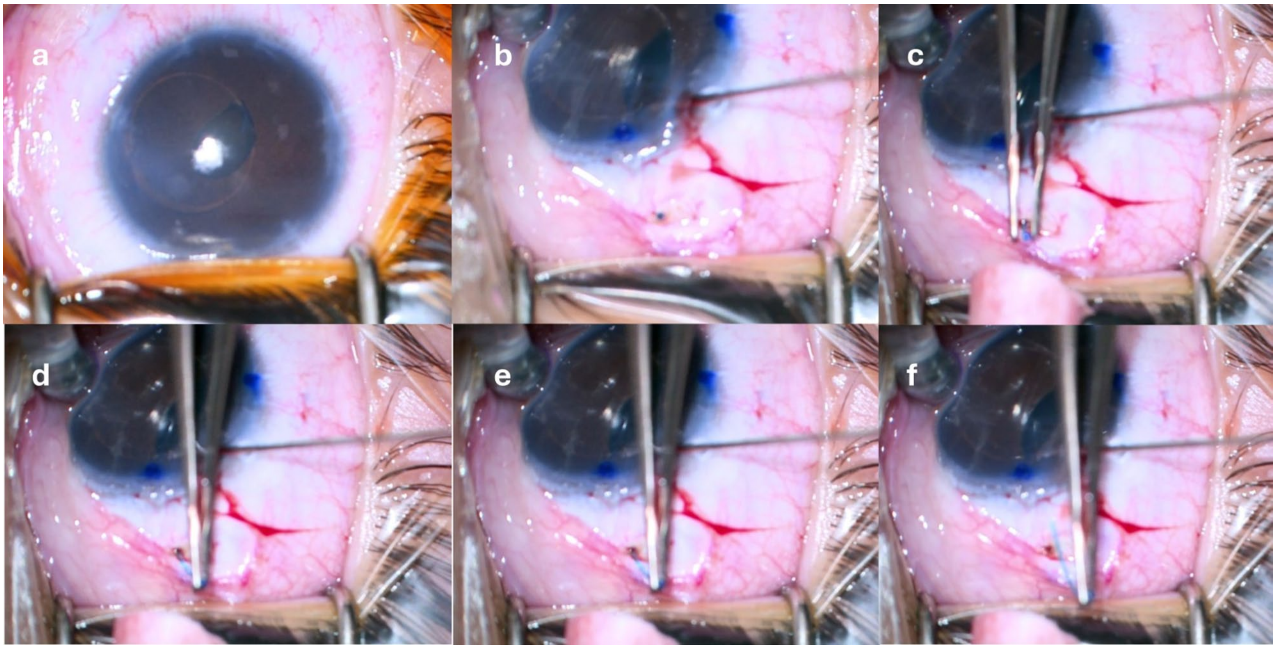

Fig. 3 **a**: Preoperative image of the capture of the optic to the iris after IOL optic-haptic dissociation. **b**: Exposure of buried haptic flange after conjunctival peritomy.**c**, **d**, **e**, **f**: Removal of the severed haptic from the scleral tunnel

Discussion

In this study, which evaluated two different IOL implantation techniques for the management of lens subluxation due to Marfan syndrome in pediatric patients, we demonstrated that successful visual rehabilitation can be achieved through individualized surgical planning. Refractive and anatomical outcomes were comparable between the two groups, with similarly low rates of perioperative and postoperative complications.

Regarding refractive outcomes, mild hyperopia or emmetropia was targeted based on the anticipated myopic shift with age and axial elongation, in accordance with the AAO [13]. However, patients showed mild myopic SE due to postoperative astigmatism. Postoperative astigmatism may result from surgical incisions, preexisting corneal astigmatism, or lenticular astigmatism related to IOL position. In our cohort, the postoperative lenticular astigmatism was 1.31-1.33D in the groups, consistent with findings from previous pediatric scleral fixation studies [15, 20]. In the Yamane group, the markedly hyperopic preoperative values, resembling those seen in aphakia, were attributed to advanced lens subluxation outside the visual axis. This finding supports the use of the Yamane technique in cases of severe zonular weakness, as described in the methods section.

Postoperative refractive values were similar between the two techniques, with slightly more hyperopic outcomes observed in the Cionni group; however, the difference was not statistically significant. Although both groups showed improvement in CVA, many patients did not achieve normal age-appropriate visual acuity due to amblyopia—particularly in those with unilateral involvement. In pediatric lens subluxation, visual development may be compromised by the severity and etiology of the subluxation, as well as coexisting visual comorbidities, especially in unilateral or syndromic cases. This finding is consistent with previous studies. For example, Shenoy et al. reported that mean CVA improved from 20/240 to 20/70 after secondary IOL implantation in a large cohort, with most children achieving better than 20/100 vision [21]. In that study, only 2 of 34 patients undergoing unilateral surgery achieved CVA better than 20/40, compared with 4 patients in our series (2 with Yamane and 2 with Cionni ring implantation). These findings underscore the importance of early optical rehabilitation and prompt amblyopia management in pediatric aphakia.

Various studies have reported differing findings regarding IOL stabilization techniques in pediatric patients [2, 21]. In our study, the mean IOL tilt angle was approximately 0.8° in the Yamane group and 1.3° in the Cionni group, with clinically significant tilt observed in one patient with Marfan syndrome who underwent the Yamane technique. Previous reports have documented tilt angles of 2.3°–3.2° with the Yamane technique [22, 23]. Sutured scleral fixation has been associated with a higher degree of tilt over time, possibly due to suture degradation [24]. In contrast, the lower tilt angles and reduced lenticular astigmatism observed with the sutureless Yamane technique may be attributed to the absence of suture-related complications. Although the Cionni ring is effective in preserving capsular bag integrity and maintaining IOL centration, decentration may still occur due to progressive zonular weakness and suture-related degradation, particularly in connective tissue disorders such as Marfan syndrome. In our series, subluxation of the IOL–capsule complex was observed in 4 patients who received a Cionni ring. Except for one Cionni case, visual acuity remained ≥ 20/40 with both methods despite significant subluxation, and no additional surgical intervention was required.

Our experience with intrascleral fixation using both techniques was generally favorable. The Yamane technique can be adapted for pediatric eyes with specific modifications, as recommended by Sternfeld et al. For instance, using 30-gauge needles instead of 27-gauge ones helps create a smaller scleral tract, better suited for the smaller anatomical dimensions of pediatric eyes [20], and may help reduce postoperative haptic exposure and prevent wound leakage. Moreover, in younger children with shorter axial lengths, care must be taken to avoid damaging the ciliary body during implantation. A notable technical challenge in these cases is maneuvering the trailing haptic into the syringe tip, especially given the narrow palpebral fissure [12]. In our practice, we modify the technique by placing the scleral entry sites slightly more anteriorly (approximately 1.5 mm behind the limbus in smaller eyes) in patients under 6 years of age and aggressively burying the haptic flanges to minimize the risk of erosion. Overall, our outcomes with the Yamane technique align with emerging literature suggesting that flanged intrascleral fixation provides stable IOL support with minimal complications in pediatric populations [9, 20].

In prior reports, early to mid-term postoperative complications such as secondary membrane formation, transient IOP elevation, and CME have been attributed to a more robust inflammatory response in children [21, 25]. However, despite this pronounced inflammatory response, the lower incidence of CME in children compared with adults has been attributed to the healthier and more youthful choroidal and vitreous structures [26, 27]. Furthermore, the lack of data on angiographically detected edema in these cases makes it difficult to accurately assess the true frequency of CME. In our study, only two cases experienced elevated IOP and one developed CME. The relatively low incidence of CME may be attributed to the use of intensive postoperative anti-inflammatory therapy and the fact that CME assessment in the modest sample size was performed only with OCT. Interestingly, the exaggerated inflammatory response in pediatric eyes may offer a benefit by promoting fibrotic integration along the scleral track of haptics, potentially enhancing long-term IOL stabilization [28].

Postoperative retinal complications unrelated to CME were observed in two case. In one of the Marfan patient, peripheral retinal tears were noted after Yamane fixation. Another one developed retinal detachment. Consistent with the literature, the risk of retinal detachment following secondary IOL surgery in non-traumatic pediatric eyes remains low [29]. However, the risk of retinal detachment in patients with Marfan syndrome is higher than in the general population, independent of surgical intervention. This risk may be reduced when the capsular bag is preserved and effective anterior vitrectomy is performed [7]. In a study by Sandhu et al. involving 41 eyes, the rate of postoperative retinal detachment was 10%, with all cases occurring in patients with Marfan syndrome [30]. In contrast, another study reported no instances of retinal detachment in patients implanted with Cionni rings [7]. In our study, no retinal tear or detachment was observed in any of the cases treated with Cionni rings. Although Marfan syndrome itself is a risk factor, the likelihood of retinal detachment may vary based on surgical technique, surgeon experience, and individual patient characteristics, and should be carefully considered in surgical planning.

In our cohort, a significant decrease in endothelial cell count was observed compared to preoperative values in both groups. However, endothelial cell counts remained within the age-appropriate normal range in all patients, and no cases of corneal decompensation were noted. One notable exception occurred in a patient with Yamane technique, where a significant postoperative decrease in endothelial cell count was observed due to endothelial-optic contact following optic-haptic separation. Most studies on pediatric endothelial cell loss have focused on anterior chamber iris-claw lenses, which are generally linked to greater cell loss [31, 32]. However, some reports found no significant difference [33]. Nevertheless, iris-claw lenses with retropupillary fixation, as well as scleral or sulcus-fixated IOLs, are considered to have a lower risk of endothelial-related complications and are thus preferable in preserving endothelial health in pediatric cases [34].

The retrospective design and relatively small sample size represent limitations of this study. However, the Marfan syndrome cases adds value, given the rarity of the condition. The heterogeneity in surgical techniques, while representative of real-world clinical practice, limits the ability to perform robust subgroup comparisons. Additionally, although follow-up extended up to nearly three years in some patients, this duration remains relatively short for evaluating long-term complications such as suture degradation, haptic erosion, or IOL dislocation. Another limitation is the lack of standardized preoperative visual acuity assessments in all patients, particularly among younger children.

Conclusions

In summary, both the Cionni ring and Yamane techniques provide effective visual rehabilitation in pediatric Marfan syndrome patients with lens subluxation, yielding comparable anatomical and functional outcomes. However, due to the dynamic nature of ocular development in children, IOL implantation presents potential short- and long-term risks. As such, long-term monitoring is essential, and surgical approaches should be tailored based on the patient's age, systemic condition, and ocular characteristics to ensure optimal outcomes.

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Author contributions

All authors read and approved the final manuscript. Fahri Onur Aydin: Conceptualization, methodology, data curation, formal analysis, investigation, resources, writing (original draft), visualization. Irem Onal: Conceptualization, methodology, data curation, formal analysis, investigation, resources, writing (original draft). Ali Ceylan: Conceptualization, data curation, resources. Yusuf Berk Akbas: Conceptualization, data curation, resources. Elif Kedek Bilmez: Investigation, data curation. Burcin Kepez Yildiz: Supervision, writing (review and editing), conceptualization. Yusuf Yildirim: Supervision, writing (review and editing).

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Data availability

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

The ethics approval for our study was obtained from the Basaksehir Cam and Sakura City Hospital Clinical Research Ethics Committee, with the approval code: KAEK/ 29.01.2025.33. For participants under 12 years of age, informed consent was obtained from their legal guardians. For participants 12 years of age and older, informed consent was obtained from both the patients themselves and their legal guardians. This study was conducted in accordance with the principles of the Declaration of Helsinki.

Consent for publication

Not applicable.

Competing interests

The authors declare no competing interests.

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