

Pupillary capture following sutureless scleral-fixated intraocular lens in children with Marfan syndrome

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Abstract

• **AIM:** To compare the clinical outcomes between two approaches for sutureless scleral-fixated intraocular lens (SFIOL) in children with Marfan syndrome (MFS).

• **METHODS:** The study included 15 children (26 eyes) with lens subluxation due to MFS. These children underwent lensectomy, anterior vitrectomy, and sutureless SFIOL. According to the position of placement of intraocular lens (IOL) haptics, two study groups were reviewed for best corrected visual acuity (BCVA) and postoperative complications: group A, 14 eyes with haptics fixated at 2.0 mm from the limbus; group B, 12 eyes with the haptics fixated at 2.5 mm from the limbus.

• **RESULTS:** The mean axial length for all patients was 25.66 ± 2.35 mm. Postoperative BCVA in logMAR were significant improved in both groups (0.77 ± 0.32 to 0.17 ± 0.12 in group A, 0.66 ± 0.25 to 0.24 ± 0.12 in group B, both $P < 0.001$) while no significant difference between two groups ($P > 0.05$). Pupillary capture was main postoperative complication, occurring between 3d and 18mo. It occurred in 7 eyes in group A and one eye in group B ($P = 0.02$).

• **CONCLUSION:** Sutureless SFIOL is an effective treatment approach for lens subluxation in children with MFS. Pupillary capture is the main postoperative complication. Fixated IOL haptics at 2.5 mm from the limbus can reduce the occurrence of pupillary capture.

• **KEYWORDS:** Marfan syndrome; sutureless scleral-fixated intraocular lens; pupillary capture; children

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INTRODUCTION

Marfan syndrome (MFS) is a heritable disorder with an autosomal dominant mode, and affects the connective tissue. This disease is caused by the mutation from the *FBN1* gene, which codes for fibrillin-1 on chromosome 15^[1]. In clinic, the symptoms of MFS involve the eyes, skeleton, and cardiovascular system^[2]. The major manifestations in the eye are lens dislocation or subluxation, high myopia, and retinal detachment. Among these, lens subluxation is the most common complication (60% incidence) in eyes with MFS^[3]. For children with MFS, mild lens subluxation-induced refractive errors can be corrected by wearing glasses. However, severe lens subluxation-induced high refractive errors and/or anisometropia affect the development of visual function, and require surgery for correction. At present, lensectomy combined with anterior vitrectomy and sutureless scleral-fixated intraocular lens (SFIOL) has been broadly applied for the treatment of lens subluxation^[4-6]. The surgical techniques and complications have been studied. However, there were few reports on postoperative complications and treatments after the SFIOL procedure for lens subluxation in children with MFS. Therefore, the present study was designed to investigate these questions.

SUBJECTS AND METHODS

Ethics Approval This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the Ethics Committee of Southwest Hospital/Southwest Eye Hospital, Army Medical University [Third Military Medical University; Date: March 15, 2022/No.(B)KY202258]. Informed consent forms were obtained from the parents of the children before the procedure.

Patients The present retrospective study included 15 children (26 eyes) with lens subluxation caused by MFS. These children were diagnosed and treated from December 2018 to October 2021. Among them, 8 children were male and 7 children were female, and the mean age was 8.15 ± 1.41 y (range: 5-12y).

Examinations All patients received the following examinations before surgery: uncorrected visual acuity (UVA), best corrected visual acuity (BCVA), cycloplegic refraction, slit lamp microscopy, intraocular pressure (IOP), ophthalmoscopy, and measurement using the IOL Master (Model 700, Carl Zeiss,

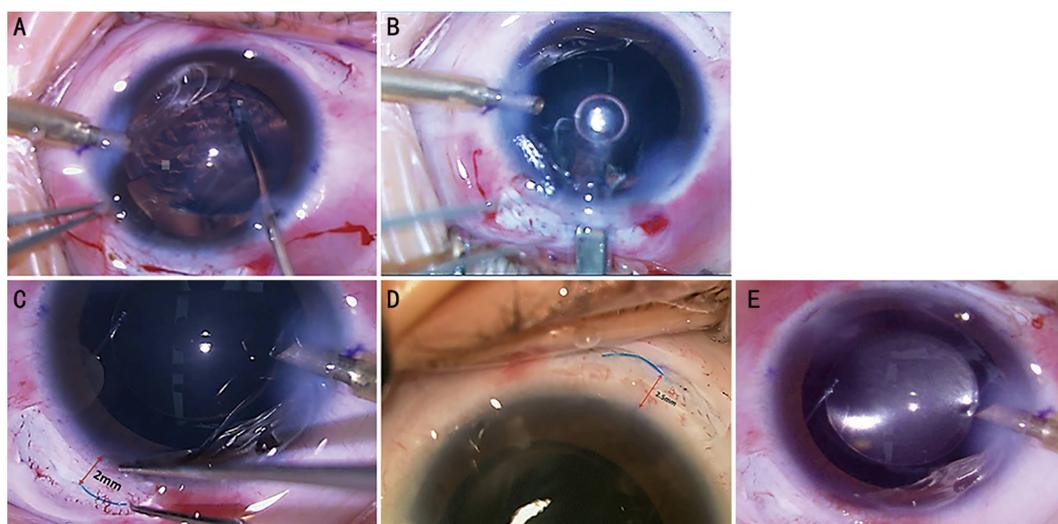


Figure 1 Surgery for lens subluxation in children with MFS A: Lensectomy combined with anterior vitrectomy; B: The IOL was implanted through the scleral tunnel; C: The IOL was fixed at 2.0 mm from the limbus (the red arrow shows the distance from the limbus to the lens haptic); D: The IOL was fixed at 2.5 mm from the limbus (the red arrow shows the distance from the limbus to the lens haptic); E: IOL position. MFS: Marfan syndrome; IOL: Intraocular lens.

Jena, Thuringia, Germany) for anterior chamber depth, cornea curvature, eye axis length, and lens thickness. The intraocular lens (IOL) diopter was calculated using the Barrett Universal II formula. The lens diopter selected for children younger than eight years old was equal to 90% of the value calculated, while 100% of the value calculated was selected for children older than eight years old.

Surgery All surgeries were conducted by the same surgeon (Qi DM), and the patients received general anesthesia (Figure 1). Before the operation, 0.5% compound tropicamide eye drops (Santen pharmaceutical Co., Ltd., Osaka, Japan) were instilled thrice in the eyes to dilate the pupils. Then, the eyelids were opened with an eyelid opener, 0.01% povidone-iodine was applied into the conjunctival sac for 90s, and this was washed out using 5 mL of phosphate-buffered saline. Next, two incisions were made along the limbus from the 11 o'clock to 2 o'clock, and from the 6 o'clock to 8 o'clock in the conjunctiva and Tenon's capsule to expose the sclera. Afterwards, an anterior chamber puncture was performed at the 7:30 o'clock (right eye) or 5:30 o'clock (left eye) to insert the irrigator. Then, include methods involved in lensectomy and anterior vitrectomy were performed through a limbus incision at the 10 o'clock, and the IOL (MA60AC, Alcon Laboratories, Huntington, WV, USA) was implanted into the eye through the cornea-scleral tunnel at 1 mm from the limbus from the 11:30 o'clock to 12 o'clock. The haptics of the IOL were placed in the 3-mm sclera tunnel at the 1 o'clock and 7 o'clock positions. Patients with IOL haptics fixed in the scleral tunnel at 2.0 mm from the limbus were assigned to group A (8 cases, 14 eyes), while patients with IOL haptics fixed in the scleral tunnel at 2.5 mm from the limbus were assigned to group B (8

cases, 12 eyes). The position of the scleral tunnel is measured using a caliper. One patient had one eye in group A and one eye in group B.

Follow-up The patients were administered with 1% prednisone acetate eye drops (6 times/day) and tobramycin dexamethasone eye ointment (before sleep) for 2wk. Then, these were changed to pranopfen eye drops (4 times/day). The follow-up periods were at two weeks, 1, 3, 6mo and then once a year after surgery. The longest follow-up duration was at 48mo after surgery. Refractive examination and glasses prescription were conducted 3mo after surgery.

Statistical Analysis SPSS 26.0 (IBM Corp., Armonk, NY, USA) was used for the data analysis. Continuous variables with a normal distribution were presented as mean±standard deviation. *t*-test was used for comparisons between two independent variables, and between preoperatively and postoperatively collected data. Categorical variables were presented as *n* (%), and were compared using Chi-square test. A *P*-value of <0.05 was considered statistically significant.

RESULTS

Age There were 14 eyes in group A, and the mean age was 8.57 ± 0.85 y (range: 8-10y). There were 12 eyes in group B, and the mean age was 7.67 ± 1.77 y (range: 5-12y). There was no statistical significance in age between the two groups ($P=0.11$; Table 1).

Eye Axis Length The mean axis length before surgery was 25.66 ± 2.35 mm, which ranged from 22.47 mm (five years old) to 29.69 mm (eight years old), for all patients. The mean eye axis length before surgery was 25.35 ± 2.62 mm, which ranged from 22.78 to 29.69 mm, for patients in group A, while the mean eye axis length before surgery was 25.11 ± 2.10 mm,

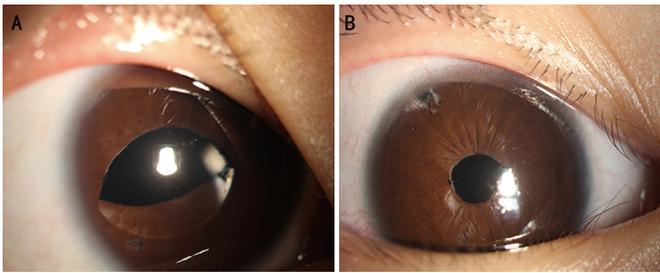


Figure 2 Position of IOL before and after laser iridotomy A: Pupillary capture after the sutureless SFIOL, with the IOL haptics placed at 2.0 mm from the limbus; B: The pupillary capture was corrected after laser peripheral iridotomy. SFIOL: Scleral-fixated intraocular lens surgery; IOL: Intraocular lens.

Table 1 Patient age and clinical measurements

Items	Group A (n=14)	Group B (n=12)	P
Mean age (y)	8.57±0.85	7.67±1.77	0.11
Mean eye axis length (mm)	25.35±2.62	25.11±2.10	0.31
Preoperative BCVA (logMAR)	0.77±0.32	0.66±0.25	0.187
Postoperative BCVA (logMAR)	0.17±0.12	0.24±0.12	0.161
Pupillary capture	7.00 (50.00%)	1.00 (8.30%)	0.02
P ^a	<0.001	<0.001	

^aSignificant difference in BCVA between the preoperative and postoperative values. BCVA: Best corrected visual acuity.

which ranged from 22.47 to 29 mm, for patients in group B. There was no statistically significant difference in eye axis length between the two groups ($P=0.31$; Table 1).

Refractive Examination and Visual Acuity The BCVA before surgery was 0.73 ± 0.28 (logMAR) for all patients. Furthermore, the spherical equivalent (SE) refraction ranged from +11.25 diopters (D) to +30.00 D in 14 cases (25 eyes), and the astigmatism ranged from 5.00 to 9.25 D in 5 cases (7 eyes). These high refractive errors and/or anisometropia were all induced by the lens subluxation. The BCVA at 3mo after surgery was 0.20 ± 0.12 (logMAR) for all patients, the SE refraction ranged from +3.75 to +1.75 D, and the astigmatism ranged from 0.75 to 1.75 D. There was a significant difference in visual acuity between the preoperative and postoperative values for all cases ($P<0.001$; Table 1). However, there was no significant difference in visual acuity between group A and group B, both before and after surgery ($P>0.05$).

Complications and Treatment The major postoperative complication was pupillary capture, which occurred in 14 eyes (7 eyes, 50%) in group A. The earliest onset was at 3d after surgery, while the latest onset was at 18mo after surgery. Self-recovery was observed in all cases with pupillary capture after pupil dilation in the supine position. However, 5 of these cases presented with recurrent pupillary capture at the subsequent follow-up, and underwent laser iridotomy (Figure 2). No recurrence was observed after the laser iridotomy. One eye with pupillary capture in group A could not be recovered after laser

iridotomy. This was finally treated successfully by re-fixing the IOL haptics to 2.5 mm from the limbus. The last follow-up was from 18mo to 4y after surgery in group A. These patients had a maintained BCVA, and no pupillary capture was observed at follow-ups. Among the 12 eyes in group B, 1 (8.3%) had pupillary capture. The onset of the pupillary capture was at 3d after surgery, and this self-recovered after pupil dilation in the supine position. There was no recurrence during the follow-up. The last follow-up was from 12mo to 3y after surgery in group B. These patients presented with a maintained BCVA, and no pupillary capture was observed during the follow-up. There was a significant difference in the incidence of pupillary capture between group A and group B ($P=0.02$; Table 1). Furthermore, one patient developed retinal detachment at 4mo after surgery in group A. This patient underwent vitrectomy with silicon oil tamponade. The eye axis length of this patient was 27.29 mm, and ora serrata dialysis was observed during the vitrectomy. The BCVA recovered to 0.3 (logMAR) after the silicon oil was taken out.

DISCUSSION

At present, the surgical treatment for ectopia lentis with MFS entails phacoemulsification and lensectomy with anterior vitrectomy. Researchers have suggested that IOL implantation should be performed at an appropriate time following the lensectomy, in order to correct the refractive error. However, challenges exist in the approach for IOL implantation. Anterior chamber IOL has a risk of corneal endothelial decompensation. Iris-claw IOL has a possibility of IOL downward displacement caused by an iris cyst and iris atrophy in the long term^[7-9]. In capsular tension ring (CTR)-assisted IOL implantation, the IOL dislocation caused by the development of a loose or broken ciliary zonule cannot be avoided. The modified CTR sutured to the sclera remains at the peripheral lens capsule, providing long-term stability to the IOL. However, this causes difficulties in treating the peripheral hole or the dialysis of the ora serrata during surgery for retinal detachment, with a high incidence in children with MFS. Furthermore, suturing in the ciliary sulcus has a risk of IOL dislocation caused by the loosening and exposure of sutures and knots in the long term^[10-13].

Due to the limitations of the above approaches, the sutureless SFIOL procedure was developed by Gabor and Pavlidis^[14]. The advantage of this surgical method is that the operation is relatively simple and there is no risk of suture loosening. The IOL haptics were fixed in the scleral tunnel at 1.5-2.0 mm from the limbus. No postoperative complication was found during the three-month follow-up period. Thus, lensectomy combined with anterior vitrectomy and sutureless SFIOL has recently become widely accepted for ectopia lentis treatment. This approach removes the entire lens and capsule, thereby preventing the dislocation of the IOL due to the damage to the

ciliary zonule. Furthermore, the IOL is located at the posterior chamber, preventing injury to the corneal endothelium. Moreover, the sutureless procedure prevents the IOL dislocation caused by the loosening or breaking of sutures or knots, and infections due to suture exposure.

At present, sutureless SFIOL fixed at 2.0 mm from the limbus remains as the most common practice^[15-18]. However, in the present study, the occurrence of postoperative pupillary capture was very high when the distance of the SFIOL fixation was at 2.0 mm from the limbus in MFS children. The reasons may be, as follows: 1) The deeper anterior chamber. Due to significant lens dislocation in children with MFS, inaccuracies often occur when measuring the depth of the anterior chamber. Therefore, we are unable to accurately measure the depth of these patient's anterior chamber. Researchers have reported that there was a positive correlation between axial length and anterior chamber depth^[19]. The mean eye axis length of these patients was 25.66 mm, which was 1.50 mm longer than that of healthy adults. Therefore, we speculate that the anterior chamber is deeper in MFS than that in normal adult. The IOL was closer to the iris when the SFIOL was fixed in the sclera using a conventional distance of 2.0 mm from the limbus. 2) At present, SFIOL is a three-piece IOL that comprises of one optic part and two tenuous haptics. After the lensectomy, the SFIOL and iris may swing due to the changes of the pupil and flow of aqueous humor between the posterior chamber and anterior chamber. Thus, when the SFIOL was fixed at 2.0 mm from the limbus in MFS children, the SFIOL was closer to the iris, it was easier to observe the pupillary capture. YAG laser peripheral iridotomy is effective for treating pupillary capture, because an additional path for aqueous humor flow is established, which reduces the pressure of the aqueous humor on the SFIOL and iris from the posterior and anterior chamber. The other complication of SFIOL was vitreous hemorrhage, postoperative hypotony, postoperative IOP rise, hyphema, tilted IOL, cystoid macular edema, retinal detachment were also noted^[20]. After surgery, one patient developed retinal detachment. No vitreous hemorrhage and hyphema occurred in all patients. The possible reason was that we used anterior chamber perfusion during the operation, the IOP was stable during the operation.

In conclusion, lens subluxation with MFS affects the visual development of children. Lensectomy and anterior vitrectomy with sutureless SFIOL is effective for improving the refractive status and visual quality. Glasses correction may give an additional improvement. For lens subluxation surgery treatment in MFS children, a high occurrence of pupillary capture was observed when the conventional approach of sutureless SFIOL was used, which fixed the IOL at 2.0 mm from the limbus. The occurrence of pupillary capture was significantly reduced

when the IOL fixation was moved to 2.5 mm from the limbus. However, the cases in the present study were limited, and the study did not have a randomized prospective design. A clinical randomized controlled study with a large sample size is warranted.

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Conflicts of Interest: Qi DM, None, Huo SJ, None; Yu T, None.

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