ORIGINAL RESEARCH

# Outcomes of Iris-Claw IOL Implantation in Patients with Marfan's Syndrome in Jordan

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**Objective:** The management of ocular complications of Marfan's syndrome, especially ectopia lentis, is challenging. In this study, we present the effectiveness and the safety of iris-claw intraocular lens (IOL) implantation along with lensectomy for those patients. Also, we compare the practice of implanting these IOLs either in the anterior chamber of retropupillary.

**Methods:** Retrospectively, we included all patients with Marfan's syndrome who underwent lensectomy with iris-claw IOL implantation as a result of ectopia lentis. The patients were categorized into two groups: anterior chamber iris claw IOL and retropupillary iris-claw IOL. The clinical and demographic data, the visual outcome and postoperative complications were compared. **Results:** Eighteen eyes of 10 patients were included in the study. The mean age of the patients was 19.1 years. Six patients were males. The iris-claw IOL was implanted anteriorly in 13 eyes. The visual outcome was comparable between both groups and most patients achieved improvement in the visual acuity. In addition, the postoperative complications developed similarly in both groups. However, all cases of IOL disenclavation (6 cases) developed in the anterior group. It is revealed that the age of the patient was the most significant factor affecting the occurrence of IOL disenclavation.

**Conclusion:** Iris-claw IOL (either anteriorly or retropupillary) is an effective and relatively safe method in treating ectopia lentis in patients with Marfan's syndrome. In younger patients, anterior iris-claw IOL is safer than retropupillary iris-claw IOL as the risk of disenclavation is higher in younger patients.

Keywords: Marfan's syndrome, Artisan, retropupillary, ectopia lentis

#### Introduction

Marfan's syndrome (MFS) is a genetic disorder of connective tissue associated with mutation in fibrillin-1, an important component of the elastic microfibril of ciliary zonules. It is an autosomal dominant connective tissue disorder. Early diagnosis is of crucial importance owing to the life-threatening complications of cardiovascular pathology.<sup>1–3</sup> The diagnostic criteria of MFS are included in the revised version of the Ghent criteria. According to this scoring system, MFS has been established with a score of  $\geq$ 7 points (of a maximum total of 20 points) and it is considered diagnostic.<sup>4,5</sup> In the absence of a family history of MFS, MFS is diagnosed in the presence of a ortic root dilatation combined with ectopia lentis, or a causative FBN1 mutation. In the presence of a family history, MFS is diagnosed with the demonstration of ectopia lentis, or a systemic score  $\geq$ 7 points, or aortic root dilatation.<sup>4,5</sup> Accordingly, ocular manifestations (especially ectopia lentis) are a corner stone in the diagnosis of MFS.

The main ocular features of MFS include ectopia lentis, myopia and retinal detachment.<sup>6–9</sup> Ectopia lentis is the most common one and occurring in 50–80% of patients with MFS, and it is defined as displacement or subluxation of the crystalline lens. The general feature of ectopia lentis in MFS patients is usually bilateral, symmetric and non-progressive. It may vary from a mild asymptomatic dislocation seen only with dilation of pupil to significant subluxation that places the equator of the lens in the pupillary axis. Also, the severe forms of ectopia lentis include crystalline lens dislocation

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into the anterior chamber, which may lead to pupillary block or chronic angle-closure glaucoma.<sup>3,7–10</sup> Posterior dislocation may be hazardous for the retina with a risk for retinal detachment, chronic vitritis and chorioretinitis.<sup>11</sup>

For mild cases, the functional visual acuity may be obtained with refractive aids. However, in severe ectopia lentis, unstable refractive status, glaucoma, or endothelial cell loss, surgery is recommended.<sup>12,13</sup> The optimal surgical approach is still controversial and may vary with the individual experience of the surgeon or with individual features of the patient. Lensectomy with Iris-claw intraocular lens (IOL) "Artisan<sup>®</sup>" implantation has been studied in those patients with several advantages such as good visual outcome, fewer complications, and easy placement. The iris-claw IOL may be implanted in the anterior chamber or retropupillary.<sup>8,14–20</sup>

In this study, we evaluate the practice of iris-claw IOL implantation in patients with MFS who experienced ectopia lentis in our educational institution. Also, it compares between the effectiveness and safety between anterior and retropupillary iris-claw IOL in those patients.

# **Methods**

#### Patients

Retrospectively, we evaluated the characteristics of 18 eyes of 10 patients with Marfan's syndrome who underwent irisclaw "Artisan<sup>®</sup>" IOL implantation as a result of severe ectopia lentis during the period of January 2014 to December 2021. After the approval of the Institutional Review Board at Jordan University of Science and Technology (JUST), the study was conducted at King Abdullah University Hospital (KAUH), a tertiary educational center for ophthalmic services which is affiliated with JUST. Using the paper-based and electronic documents records, demographic data (age, sex), past medical history, and the preoperative optical parameters were collected. Furthermore, the operative details, visual outcome and postoperative complications were evaluated.

The included study population was those patients with Marfan's syndrome who underwent lensectomy with a primary iris-claw IOL implantation as a result of ectopia lentis. Those patients fulfill the revised version of Ghent criteria of Marfan's syndrome. The exclusion criteria comprised patients with insufficient preoperative or postoperative data, patients with traumatic lens subluxation or ectopia lentis due to causes other than Marfan's syndrome, and patients with previous ocular surgery. The included cases of ectopia lentis were defined as crystalline lens subluxation with border affecting the pupillary axis or anterior chamber or vitreous lens subluxation. The patients were divided by location of implantation into 2 main groups: the anterior iris-claw IOL and retropupillary iris-claw IOL.

The outcome was compared between both main groups using different measures. First, the mean change in visual acuity was compared preoperatively and postoperatively during all follow-up visits. Second, postoperative complications were compared and included irregular iris shape (new postoperative irregularity or aggravated preoperative irregularity), iris tissue loss, iris-claw IOL decentration or tilt, spontaneous or traumatic disenclavation, clinical signs of endothelial cell loss (including long-term corneal edema and the development of bullous keratopathy), pigment dispersion, post-operative high intraocular pressure (IOP) which affected the vision and required the use of antiglaucoma agents or the need for glaucoma surgery and retinal detachment. All data were retrieved from visits preoperatively and at 1 week, 1 month, 3 months, 1 year, and on the last follow-up visits postoperatively.

## **Perioperative Setting**

Visual acuity was assessed by Snellen decimal projectors. Visual acuity was converted to LogMAR visual acuity. For patients with visual acuity of counting fingers, hand motion, light perception or "no light perception", they were converted according to the study of Schulze-Bonsel K et al.<sup>21</sup> IOP was measured by Goldmann tonometry, and anterior and posterior segment examination were performed through slit-lamp biomicroscopy with the required non-contact hand-held lenses. The ophthalmic examination was done by well-trained residents and confirmed by the attending consultant ophthalmologists.

The IOL power was measured either by ultrasonic biometry (Digital A/B scan 5500; Sonomed Inc., Lake Success, NY, USA (United States of America)) or by IOL Master when needed. Sanders-Retzlaff-Kraff (SRK-T) formula was used for the selection of the IOL power (other formulas were utilized such as Haigis formula in patients with high myopia and Holladay II and Hoffer Q for patients with short axial length). The optical parameters included the iris-claw

IOL power (using an A-constant of 115 for anterior iris-claw and 117 for retropupillary iris-claw IOL), keratometry readings, and axial length. Emmetropia was targeted in the eyes of patients > age 10, while hyperopia was the aim in younger patients, with values dependent on patient age (for ages 7–10: +0.5 D, ages 5–7: +1; and ages 3–5: +2). The biometry was done under general anesthesia in pediatric patients.

Six consultant surgeons performed the operations and selected to implant the IOL either anteriorly or retropupillary depending on their individual experience. The same standardized surgical technique and guidelines were applied in both groups. The lens used in this study was the Artisan<sup>®</sup> aphakia IOL (Ophtec BV, Groningen, The Netherlands) which is a polymethyl methacrylate IOL with an 8.5-mm length, 1.04-mm maximum height, and 5.4-mm optical zone width. All operations were performed under either general or local anesthesia. Two corneal side ports were performed at 3 and 9 o'clock positions. After performing the lensectomy by the vitreous cutter (either limbally through the anterior chamber in most cases or through a pars plana seclerotomies), acetylcholine 1% (Miochol<sup>®</sup> -E) was injected intracamerally through the paracentesis for miosis. A 5.5-mm corneal incision was made at 12 o'clock. For retropupillary implantation, the iris-claw IOL was inserted upside down (with its convex surface facing posteriorly), rotated by an Artisan lens forceps to a horizontal position, and centered over the pupil. The optic of the reversed iris-claw IOL was held securely using a special forceps. Next, the two haptics were gently slid behind the iris. With the other hand, a long micro-spatula was used through the side ports to tuck iris tissue into the claw. For anterior implantation, the convex surface was placed anteriorly, and the iris was enclavated at midperiphery between the claw haptics. The corneal incision was closed and secured with three simple buried interrupted 10-0 nylon sutures. In 3 cases, the procedure was combined with pars plana vitrectomy. A peripheral iridotomy (PI) was done in some cases. Postoperative therapy included antibiotic, steroid and nonsteroidal anti-inflammatory eye drops for 1 month. Selective corneal suture removal according to corneal astigmatism was performed 6 to 8 postoperative weeks. Many patients underwent prophylactic laser retinopexy especially who have retinal pathologies.

## Statistical Analysis

Extracted data were entered into a spreadsheet. Statistical analysis was performed using the IBM SPSS v.22 (Armonk, New York, USA). Data were expressed as frequency (percentage) for nominal data, mean  $\pm$  standard deviation of the mean (SD). Statistical significance between the study groups was determined using Chi-square test for categorical variables, and Student's *t*-test for continuous variables. P  $\leq$  0.05 was considered statistically significant.

# Results

## **General Characteristics**

Eighteen eyes of 10 patients with MFS who underwent lensectomy with iris-claw IOL implantation due to ectopia lentis were allocated in this study. Of the 10 patients, 6 (60%) were males. The mean age of the patients was 19.1 years. Of the 18 eyes, the left eye was involved in 9 (50%) of the cases. Iris-claw IOL was implanted in the anterior chamber in 13 (72.2%) of the cases. The mean follow-up time for the patients was 31.3 months (standard error 6.4, maximum 65 months, minimum 14 months). Table 1 summarizes the general characteristics of the included patients.

In 15 cases, the lensectomy was done through the anterior chamber along with anterior vitrectomy. In the remaining 3 cases, a pars plana lensectomy along with pars plana vitrectomy was utilized and eyes were kept flat under air. Peripheral iridotomy was created in 14 eyes (77.8%). Most patients achieved an improvement in visual acuity at the last follow-up visit. Regarding the postoperative complications, disenclavation of one or both haptics of IOL was the most significant and most commonly encountered postoperative complication. It occurred in 6 eyes (33.3%) either spontaneously or by trauma. Irregular pupil shape and iris tissue loss developed in 5 (27.8%) eyes for both. Only two eyes of the same patient developed high IOP reading, which was controlled by antiglaucoma medications. Table 2 shows the detailed outcome for every patient.

# Retropupillary versus Anterior Artisan

There was no difference between retropupillary and anterior iris-claw IOL in terms of sex, age, laterality and previous ocular diseases. Regarding the associated surgical procedure, PI was performed in the anterior chamber group (91.8% for anterior location versus 40% for retropupillary).

Patients	Laterality	Age at Operation	Sex	Iris Claw IOL Location	IOL Power	Other Ocular Diseases	Combined Procedures
I	Right eye	35	Male	Anterior	21	No	PI, PPV
	Left eye	35	Male	Anterior	20.5	No	PI, PPV
2	Right eye	6	Female	Anterior	17.5	No	PI, AV
	Left eye	6	Female	Anterior	18.5	No	PI, AV
3	Left eye	4	Male	Anterior	14.5	No	PI, AV
	Right eye	4	Male	Anterior	15	No	PI, AV
4	Left eye	23	Male	Anterior	24.5	No	PI, AV
	Right eye	24	Male	Anterior	23	No	AV
5	Left eye	39	Female	Retropupillary	6	No	PI, PPV
	Right eye	40	Female	Anterior	4	No	PI, AV
6	Right eye	17	Male	Anterior	21	No	PI, AV
	Left eye	17	Male	Anterior	22	No	PI, AV
7	Right eye	19	Female	Retropupillary	17	No	AV
	Left eye	19	Female	Retropupillary	22.5	No	AV
8	Right eye	5	Male	Anterior	24	No	PI, AV
	Left eye	5	Male	Anterior	25	No	PI, AV
9	Left eye	12	Female	Retropupillary	10	RD in the other eye	AV
10	Right eye	35	Male	Retropupillary	11	No	PI

 Table I
 The General Characteristics for Patients with Marfan's Syndrome Who Underwent Iris-Claw IOL Implantation Due to Ectopia Lentis

Abbreviations: PI, peripheral iridotomy; PPV, pars plana vitrectomy; AV, anterior vitrectomy; RD, retinal detachment.

The visual outcome was not statistically different between both groups Table 3. However, the retropupillary group achieved better visual improvement. At 1 year postoperative period, the mean change of BCVA in the retropupillary group was -0.600 LogMAR, which corresponds to an improvement in visual acuity of about 30 letters. On the other hand, the mean change of visual acuity was -0.357 LogMAR in the anterior group, which corresponds to about 18 letters of improvement.

In addition, the development of postoperative complications was comparable and not statistically significant between both groups. Irregular pupil shape was developed in 5 cases; all of them were in the anterior chamber implantation. Iris tissue loss was developed in 4 cases in the anterior group and in 1 case in the retropupillary group. High IOP was developed in 2 eyes where the iris-claw IOL was implanted retropupillary. Regarding the disenclavation of iris-claw IOL, all 6 cases developed in the anterior group rather than the retropupillary group, which carries the risk of IOL dropping into the vitreous cavity. In 4 cases (out of 6), the disenclavation was traumatic in nature. The other 2 cases were spontaneous and unnoticed trauma cannot be ruled out. The disenclavation was successfully managed by iris-claw IOL repositioning and fixation.

# Factors Affecting the Occurrence of Iris-Claw IOL Haptics Disenclavation

It was revealed that the laterality, location of iris-claw IOL, and the combined procedures did not affect the occurrence of disenclavation of the haptics. Regarding the sex, 5 cases were developed in male patients and 1 case in female but not statistically different. The only factor that was demonstrated to affect the development of the disenclavation is the age of the patients (P = 0.005). The mean age for patients with previous disenclavation is 7.8 years versus 24.7 for patients without disenclavation. It is important to notice that also patient 3 had more than one time of disenclavation in both eyes.

Patients	Laterality	Preoperative BVCA	Last Postoperative BCVA	Preoperative IOP	Postop IOP	Postoperative Complications
1	Right eye	0.3	0	17.0	14.0	No complications
	Left eye	0.3	0.2	11.0	16.0	No complications
2	Right eye	0.4	0.2	12.0	12.0	Irregular pupil shape, iris atrophy
	Left eye	1.6	0.4	12.0	12.0	Disenclavation of the haptics, pigment dispersion
3	Left eye Right eye	0.5	0.5	12.0	15.0 14.0	Disenclavation of the haptics (twice), irregular pupil shape Disenclavation of the haptics (three times)
4	Left eye	0.1	0	8.0	12.0	Iris atrophy
	Right eye	0.1	0.1	7.0	10.0	Disenclavation of the haptics, irregular pupil shape
5	Left eye	0.5	0.4	10.0	18.0	High IOP with AG use, iris atrophy
	Right eye	0.4	0.7	10.0	19.0	High IOP with AG use, iris atrophy
6	Right eye	0.7	0.2	14.0	10.0	No complications
	Left eye	0.3	0.2	15.0	10.0	No complications
7	Right eye	0.4	0.3	13	14	No complications
	Left eye	0.4	0.3	15	16	No complications
8	Right eye	0.5	0.2	12.0	15.0	Disenclavation of the haptics, irregular pupil shape
	Left eye	0.4	0.5	14.0	17.0	Disenclavation of the haptics, irregular pupil shape
9	Left eye	1.0	0.7	15.0	13	No complications
10	Right eye	1.9	0.7	10	10	No complications

<b>Table 2</b> The Visual Outcome and Postoperative Complications for	or the P	atients
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Abbreviations: BCVA, best-corrected visual acuity in LogMAR; IOP, intraocular pressure in mmHg; IOL, intraocular lens; AG, antiglaucoma.

#### Table 3 Retropupillary versus Anterior Iris-Claw IOL in Marfan's Syndrome

Variables	Posterior "Retropupillary" Artisan (%)	Anterior Artisan (%)	P-value
Age (years)	24.8 ± 5.2	16.9 ± 3.6	NS
Side of procedure (laterality)			
Right (OD)	2 (40.0)	7 (53.8)	NS
Left (OS)	3 (60.0)	6 (46.2)	
Associated ocular diseases	0 (0.0)	0 (0.0)	NS
Associated ocular procedure (during artisan implantation:			
Peripheral iridotomy	2 (40.0)	(9 .8)	0.02
Pars plana vitrectomy	14 (23.0)	19 (17.3)	NS
Anterior vitrectomy	27 (44.3)	55 (50.0)	NS
Postoperative complications			
Irregular iris shape	0 (0)	5 (38.5)	NS
Iris atrophy	I (20.0)	4 (30.8)	NS
Disenclavation of the haptics (either traumatic or spontaneous)	0 (0)	6 (46.2)	NS
Signs of corneal endothelium loss	0 (0.0)	0 (0.0)	NS
Pigment dispersion	0 (0.0)	I (7.7)	NS
Postoperative high IOP and/or use of new antiglaucoma and/or	I (20.0)	I (7.7)	NS
glaucoma surgery			
Retinal detachment	0 (0.0)	0 (0.0)	NS

(Continued)

Variables	Posterior "Retropupillary" Artisan (%)	Anterior Artisan (%)	P-value
Epiretinal membrane proliferation	0 (0.0)	0 (0.0)	NS
Macular edema	0 (0.0)	0 (0.0)	NS
Endophthalmitis/keratitis	0 (0.0)	0 (0.0)	NS
Overall visual outcome			
Change in BCVA at I week postoperative (LogMAR)	-0.425 ± 0.1	-0.114 ± 0.08	NS
Change in BCVA at 1 month postoperative (LogMAR)	-0.667 ± 0.1	-0.317 ± 0.09	NS
Change in BCVA at I year postoperative (LogMAR)	-0.600± 0.2	-0.357 ± 0.2	NS
Change in BCVA at last follow up (LogMAR)	-0.640 ± 0.1	-0.346 ± 0.1	NS

Abbreviations: SEM, standard error of the mean; OD, right eye; OS, left eye; IOL, intraocular lens; BCVA, best corrected visual acuity; IOP, intraocular pressure; NS, not significant.

# Discussion

This retrospective study compares the implantation of iris-claw IOL anteriorly versus retropupillary in patients with MFS who had ectopia lentis. It showed that the visual outcome was comparable between both groups with slight preference for the retropupillary group in the final visual acuity. In addition, both locations were safe with few side effects. However, in young patients, it was preferable to implant the IOL in the anterior chamber as the risk for disenclavation is higher. Cleary et al reported that anterior chamber iris-claw IOL is safe and effective in the correction of aphakia in children following lensectomy for ectopia lentis.<sup>17</sup> They reported their results on 3 patients with MFS. Aspiotis et al performed lensectomy with anterior chamber iris-claw IOL in 5 patients with MFS and ectopia lentis, and they reported that the BCVA improved 4 Snellen lines and endothelial cell counts remained constant during six months of follow-up.<sup>22</sup> Moreover, Sminia et al performed lensectomy with iris-claw IOL in the anterior chamber for two patients and followed them for 12 years with good visual outcomes and no serious complications.<sup>23</sup> Cevik et al reported outcomes of anterior chamber Artisan iris-claw lens implantation in children with non-traumatic ectopia lentis.<sup>16</sup> They concluded that Artisan provides good results in terms of improving uncorrected and corrected vision but involves a high incidence of postoperative complications, especially lens dislocation and retinal detachment.<sup>16</sup> In a case series by Rabie et al, the authors evaluated the outcome of lensectomy and iris-claw IOL in the anterior chamber for 12 eyes of nine patients with MFS and only one case of retinal detachment was developed, and another one case of IOL disenclavation was reported in this series during 44.5 months of follow-up.<sup>20</sup> Catala-Mora et al studied the effectiveness and safety of anterior iris-claw IOL for ectopia lentis in MFS patients, and they concluded that this technique is both safe and effective, improving vision in pediatric patients with severe ectopia lentis.<sup>24</sup> Gonnermann et al studied the posterior iris-claw IOL in patients with MFS-related ectopia lentis in 13 eves, and they reported good visual outcomes, low endothelial cell loss, and low complication rates.<sup>19</sup>

Ectopia lentis is the most common ocular sequela of MFS and varies from 50% to 80% in different studies. Ectopia lentis in MFS results from fibrillin abnormalities, which make the suspensory zonules of the crystalline lens posterior the iris. These abnormalities lead to zonular weakness and, in turn, subluxation of the crystalline lens "ectopia lentis" which is usually subluxated superior-temporally.<sup>6,12</sup> MFS results from autosomally-dominant heterozygous mutations in FBN1 gene, which in turn result in insufficiency of fibrillin-1. This leads to destruction of microfibrillar and structural architecture in the extracellular membranes.<sup>25</sup> Over 800 pathogenic mutations in FBN1 have been discovered. It was proposed that missense mutations in cysteine residues comprise a significantly higher proportion of mutations I fibrillin-1. In addition, it was found that mutations in the first 15 axons at the 5′ end are the causative in ectopia lentis.<sup>26,27</sup> This portion of the protein is thought to be integral to homodimer formation of the fibrillin-1 molecules, which eventually leads to polymers of fibrillin-1 and thus microfibrils. The mutations in FBN1 result in abnormal distribution and structure

of microfibrillar bundles in the capsule of MFS patients, particularly at the site of zonular attachment.<sup>28</sup> Subsequently, iris atrophy and iridodonesis can develop.

Many new surgical techniques were developed for ectopia lentis with advantages and good safety profile in comparison with the practice in previous decades where surgery for ectopia lentis was associated with serious intraoperative and postoperative complications that resulted in poor visual outcome.<sup>11</sup> The surgery of ectopia lentis in MFS is challenging as a result of two main factors, first, the capsular insufficiency that developed from ciliary zonular weakness. Second, the choice of IOL implant is mostly difficult.<sup>12</sup> Choices for IOL implant include iris-claw IOL either in the anterior chamber or retropupillary, anterior chamber IOL, posterior chamber scleral-fixation IOL, and scleral fixated capsular tension rings.<sup>12,13</sup> Regarding the anterior chamber IOL, these IOLs are made in a flexible open-loop pattern. They are deep in the anterior chamber and lack the stability in MFS patients, which leads to excessive movement with resultant corneal decompensation, peripheral anterior synechia, and glaucoma.<sup>12,29</sup> Scleral-fixation posterior chamber IOL is an optimal choice for implantation, which can avoid the corneal complications of the anterior chamber IOL with good visual outcome.<sup>12,30</sup> However, Asadi and Kheirkhah published a series on scleral-fixation IOL for 25 eyes of MFS children and showed a high incidence of complications including transient intraocular hemorrhage in 13 eyes, transient choroidal effusion in 2 eyes, late endophthalmitis in 1 eye, retinal detachment in 1 eye, and late IOL dislocation in 6 eyes.<sup>31</sup>

As mentioned, iris-claw IOL is an optimal and excellent option for MFS patients with an acceptable rate of complications regardless of its location. In their randomized trial, Hirashima et al studied 31 eyes of 16 patients with ectopia lentis due MFS. They categorized the patients into two groups, retropupillary group and anterior chamber group. They found that the improvement in visual acuity is similar in both groups. Although IOL disenclavation tended to occur more frequently in retropupillary group, the difference was not significant.<sup>32</sup> In our study, the improvement in visual acuity was similar in both groups. However, IOL disenclavation (as a result of iridodonesis) was seen more in the anterior group. We think that the age of the patients plays the most important role in determining the possibility of IOL disenclavation.

This study is not without limitations. First, the retrospective nature of the study with possible data inaccuracy and insufficiency is an important point. Second, the small sample size is an important point that limits the statistical analysis values. Third, variable IOL calculation methods and different surgeon handling may affect the outcome even with similar standardized protocols. Fourth, the deficiency of intraoperative images is another weakness point. Fifth, endothelial cell count is one of the important factors when comparing anterior and retropupillary iris-claw IOL. Unfortunately, the measurement tools are not available in our institution. Lastly, the rate of disenclavation is being higher in the anterior group due to the younger age of this group (selection bias).

In conclusion, MFS patients are prone for various ocular complications including ectopia lentis. Iris-claw IOL (regardless of its location) is one of the optimal choices for their ocular complications especially if can be managed by the surgeon. Retropupillary and anterior chamber iris-claw IOL are comparable in the visual outcome and post-operative complications in those patients. However, in younger patients, we would prefer to implant in iris-claw IOL anteriorly as the risk for disenclavation is higher. More randomized trials and reviews are needed to justify the results.

# **Availability Data and Materials**

The datasets generated and analyzed during the current study are available from the corresponding authors.

# Ethical Approval

This study has been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendment. This research has obtained ethical approval from Research and Ethics Committee, at Jordan University of Science and Technology and King Abdullah University Hospital, Irbid, Jordan. We confirm that the privacy of the participants was saved, and the data were anonymized and maintained with confidentiality. The need for consent was waived by our institutional review board due to the retrospective nature of the study.

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# References

- 1. Traboulsi EI, Whittum-Hudson JA, Mir S, Maumenee IH. Microfibril abnormalities of the lens capsule in patients with Marfan syndrome and ectopia lentis. *Ophthalmic Genet*. 2000;21(1):9–15. doi:10.1076/1381-6810(200003)2111-IFT009
- 2. Sarr SA, Djibrilla S, Aw F, et al. Marfan syndrome and cardiovascular complications: results of a family investigation. *BMC Cardiovasc Disord*. 2017;17(1):193. doi:10.1186/s12872-017-0629-8
- 3. McBride ART, Gargan M. Marfan syndrome. Curr Orthop. 2006;20(6):418-423. doi:10.1016/j.cuor.2006.09.007
- 4. Loeys BL, Dietz HC, Braverman AC, et al. The revised Ghent nosology for the Marfan syndrome. J Med Genet. 2010;47(7):476–485. doi:10.1136/jmg.2009.072785
- 5. De Paepe A, Devereux RB, Dietz HC, Hennekam RCM, Pyeritz RE. Revised diagnostic criteria for the Marfan syndrome. *Am J Med Genet*. 1996;62(4):417–426. doi:10.1002/(SICI)1096-8628(19960424)62:4<417::AID-AJMG15>3.0.CO;2-R
- 6. Chandra A, Ekwalla V, Child A, Charteris D. Prevalence of ectopia lentis and retinal detachment in Marfan syndrome. *Acta Ophthalmol.* 2014;92 (1):e82–e83. doi:10.1111/aos.12175
- 7. Cross HE, Jensen AD. Ocular manifestations in the Marfan syndrome and homocystinuria. Am J Ophthalmol. 1973;75(3):405-420. doi:10.1016/0002-9394(73)91149-5
- Gehle P, Goergen B, Pilger D, Ruokonen P, Robinson PN, Salchow DJ. Biometric and structural ocular manifestations of Marfan syndrome. *PLoS One*. 2017;12(9):e0183370. doi:10.1371/journal.pone.0183370
- 9. Judge DP, Dietz HC. Marfan's syndrome. Lancet. 2005;2005(366):1965–1976.
- 10. Wachtel JG. The ocular pathology of Marfan's syndrome: including a clinicopathological correlation and an explanation of ectopia lentis. *Arch Ophthalmol.* 1966;76:512–522. doi:10.1001/archopht.1966.03850010514009
- 11. Anteby I, Isaac M, Benezra D. Hereditary subluxated lenses: visual performances and long-term follow-up after surgery. *Ophthalmology*. 2003;110 (7):1344–1348. doi:10.1016/S0161-6420(03)00449-4
- 12. Esfandiari H, Ansari S, Mohammad-Rabei H, Mets MB. Management strategies of ocular abnormalities in patients with Marfan syndrome: current perspective. J Ophthalmic Vis Res. 2019;14(1):71–77. doi:10.4103/jovr.jovr\_29\_18
- 13. Simon M, Origlieri C, Dinallo A, Forbes B, Wagner R, Guo S. New management strategies for ectopia lentis. *J Pediatr Ophthalmol Strabismus*. 2015;52(5):1–13. doi:10.3928/01913913-20150714-02
- 14. Al-Dwairi R, Saleh O, Aleshawi A, et al. Anterior versus retropupillary iris-claw intraocular lens: indications, visual outcome and postoperative complications. *Opthalmol Ther.* 2022;11:32.
- 15. Català J, Cuadras D, Díaz-Cascajosa J, Castany-Aregall M, Prat J, García-Arumí J. Anterior iris-claw intraocular lens implantation for the management of nontraumatic ectopia lentis: long-term outcomes in a paediatric cohort. *Acta Ophthalmol.* 2016;95:170–174.
- 16. Çevik S. Cevik MO, Özmen A. Iris-claw intraocular lens implantation in children with ectopia lentis. Arq Bras Oftalmol. 2017;80:345.
- 17. Cleary C, Lanigan B, O'Keeffe M. Artisan iris-claw lenses for the correction of aphakia in children following lensectomy for ectopia lentis. Br J Ophthalmol. 2012;96(3):419–421. doi:10.1136/bjophthalmol-2011-300579
- Faria MY, Ferreira N, Neto E. Retropupillary iris-claw intraocular lens in ectopia lentis in Marfan syndrome. Int Med Case Rep J. 2016;9:149–153. doi:10.2147/IMCRJ.S106382
- Gonnermann J, Torun N, Klamann MKJ, Maier A-K, von Sonnleithner C, Bertelmann E. Posterior iris-claw aphakic intraocular lens implantation in subluxated lenses due to Marfan syndrome. *Eur J Ophthalmol.* 2013;24:352–357. doi:10.5301/ejo.5000366
- 20. Rabie HM, Malekifar P, Javadi MA, Roshandel D, Esfandiari H. Visual outcomes after lensectomy and iris claw artisan intraocular lens implantation in patients with Marfan syndrome. *Int Ophthalmol.* 2017;37(4):1025–1030. doi:10.1007/s10792-016-0366-5
- 21. Schulze-Bonsel K, Feltgen N, Burau H, Hansen L, Bach M. Visual acuities "Hand Motion" and "Counting Fingers" can be quantified with the Freiburg visual acuity test. *Invest Ophthalmol Vis Sci.* 2006;47(3):1236–1240. doi:10.1167/iovs.05-0981
- 22. Aspiotis M, Asproudis I, Stefaniotou M, Gorezis S, Psilas K. Artisan aphakic intraocular lens implantation in cases of subluxated crystalline lenses due to Marfan syndrome. J Refract Surg. 2006;22(1):99. doi:10.3928/1081-597X-20060101-18
- 23. Sminia ML, Odenthal MTP, Prick LJJM, Cobben JM, Mourits MP, Völker-Dieben HJ. Long-term follow-up after bilateral Artisan aphakia intraocular lens implantation in two children with Marfan syndrome. J AAPOS. 2012;16:92–94.
- 24. Català-Mora J, Cuadras D, Díaz-Cascajosa J, Castany-Aregall M, Prat-Bartomeu J, García-Arumí J. Anterior iris-claw intraocular lens implantation for the management of nontraumatic ectopia lentis: long-term outcomes in a paediatric cohort. *Acta Ophthalmol.* 2017;95(2):170–174. doi:10.1111/aos.13192
- 25. Eldadah ZA, Brenn T, Furthmayr H, Dietz HC. Expression of a mutant human fibrillin allele upon a normal human or murine genetic background recapitulates a Marfan cellular phenotype. J Clin Invest. 1995;95(2):874–880. doi:10.1172/JCI117737
- 26. Faivre L, Collod-Beroud G, Loeys BL, et al. Effect of mutation type and location on clinical outcome in 1013 probands with Marfan syndrome or related phenotypes and FBN1 mutations: an international study. *Am J Hum Genet*. 2007;81(3):454–466. doi:10.1086/520125
- 27. Trask TM, Ritty TM, Broekelmann T, Tisdale C, Mecham RP. N-terminal domains of fibrillin 1 and fibrillin 2 direct the formation of homodimers: a possible first step in microfibril assembly. *Biochem J.* 1999;340(3):693–701. doi:10.1042/bj3400693
- Mir S, Wheatley HM, Maumenee I, Whittum-Hudson J, Traboulsi E. A comparative histologic study of the fibrillin microfibrillar system in the lens capsule of normal subjects and subjects with Marfan syndrome. *Invest Ophthalmol Vis Sci.* 1998;39:84–93.
- 29. Morrison D, Sternberg P Jr., Donahue S. Anterior Chamber Intraocular Lens (ACIOL) placement after pars plana lensectomy in pediatric Marfan syndrome. *J AAPOS*. 2005;9:240–242.
- 30. Michaeli A, Assia EI. Scleral and iris fixation of posterior chamber lenses in the absence of capsular support. Curr Opin Ophthalmol. 2005;16:34.
- 31. Asadi R, Kheirkhah A. Long-term results of scleral fixation of posterior chamber intraocular lenses in children. *Ophthalmology*. 2008;115(1):67–72. e61. doi:10.1016/j.ophtha.2007.02.018
- 32. Hirashima DE, Soriano ES, Meirelles RL, Alberti GN, Nosé W. Outcomes of iris-claw anterior chamber versus iris-fixated foldable intraocular lens in subluxated lens secondary to Marfan syndrome. *Ophthalmology*. 2010;117(8):1479–1485. doi:10.1016/j.ophtha.2009.12.043

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