

Visual outcome and surgical results in children with Marfan syndrome

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Abstract

Importance: To determine visual and surgical results in children with Marfan syndrome.

Background: Marfan syndrome involves ocular complications which can lead to visual disturbance and amblyopia. Data about the visual and surgical results in children with Marfan syndrome is vital for the clinical management of these patients.

Design: Retrospective data analysis.

Participants: Eighty-two eyes of 41 patients with a genetically proved diagnosis of Marfan syndrome.

Methods: Medical records of patients with Marfan syndrome were reviewed between 2007 and 2017. Ocular complications, visual acuity (VA) of patients with/without lensectomy and surgical method were evaluated.

Main Outcome Measures: VA outcomes of patients with Marfan syndrome with/without surgical repair of ectopia lentis.

Results: In 27 (66%) of the 41 patients a bilateral subluxation of the lens was visible and 14 (52%) patients received lensectomy. The mean age at initial presentation was 12.3 ± 9 years and mean follow-up was 3 years (range 1-7). VA varied from 1.2 to -0.1 logMAR at first examination. At initial presentation, mean VA was 0.1 ± 0.7 logMAR in patients with a normal lens status ($n = 14$) and 0.3 ± 0.5 logMAR in patients with subluxation of the lens ($n = 27$) ($P < .01$). VA improved from 0.2 ± 0.5 logMAR to 0.1 ± 0.5 logMAR ($P = .06$) in children with subluxation of the lens who did not need a lensectomy ($n = 12$) and from 0.5 ± 0.5 logMAR to 0.2 ± 0.5 logMAR ($P = .02$) in patients who were referred to lensectomy with/without secondary lens implantation.

Conclusions and Relevance: In Marfan syndrome subluxation of the lens was found in the majority of children. Improvement of VA resulted in children with subluxation of the lens and following lensectomy.

KEYWORDS

Marfan syndrome, ectopia lentis, lensectomy, visual and surgical result

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1 | INTRODUCTION

Marfan syndrome is an autosomal-dominant hereditary connective tissue disorder with manifestations in different parts of the body including the cardiovascular system, lung, skeleton, skin, dura mater and eye.¹ Mutations in the *FBN1* gene encoding fibrillin 1, which is a structural component of the extracellular matrix, are the most common causes of Marfan syndrome.² The prevalence varies between 1.5 and 20/100 000 in the literature, with a mean age of 19 years at first diagnosis.³⁻⁵ The disease has a high degree of clinical variability with mild to severe features and the onset of the clinical manifestations varies. It is characterized by three clinical criteria involving aortic aneurysm (and/or dissection), ectopia of the lens and systemic features together with two genetic criteria. Other manifestations include a tall and thin habitus with long fingers and toes, abnormalities of the breastbone and spine as well as cardiovascular disease.⁶

Ocular manifestations or complications vary and can include subluxation of the lens, occurring in approximately 60% of the patients, a moderate to high degree of myopia, astigmatism, strabismus and risk for retinal detachment, glaucoma, keratoconus, corneal thinning and early cataracts.^{7,8} Ectopia lentis, which mostly occurs bilaterally and superotemporally, is a key symptom of Marfan syndrome. It is often the first sign of disease and is a hallmark for the clinical diagnosis. An altered relation between the anterior zonule-lens capsule together with abnormally large capsular and zonular fibres has been observed in histopathological studies.⁷ Minimal subluxation of the lens may not cause visual disturbance. However, progressive subluxation or complete dislocation of the lens causes refractive changes and high astigmatism with visual disturbances, amblyopia and complications such as damage to the corneal endothelium, retinal detachment or glaucoma. The surgical approach for children with Marfan syndrome is lensectomy with or without implantation of an intraocular lens (IOL) to avoid visual loss and high amblyopia.

Although Marfan syndrome is a rare condition, it represents an important and challenging disease as it requires multidisciplinary clinical evaluation and management, and genetic examination. Only few data about the visual outcome and surgical results in children with Marfan syndrome and ectopia lentis are reported in the literature. Therefore, the aim of this study was to describe the ocular complications as well as the visual outcome in ectopia lentis with or without lensectomy in children with Marfan syndrome.

2 | METHODS

In this retrospective analysis, 82 eyes of 41 patients with a genetically proved diagnosis of Marfan syndrome were

evaluated regarding ocular complications and lens status. The visual outcome and surgical results were analysed for children receiving lensectomy following subluxation of the lens and for children with subluxation of the lens who did not need a lensectomy. Inclusion criteria comprised all children with a genetically proved diagnosis of Marfan syndrome who were routinely referred for clinical examination to the Department of Ophthalmology and Optometry at the Medical University of Vienna for evaluation between 2007 and 2017. The study was approved by the local ethics committee of the Medical University of Vienna. The study design adhered to the tenets of the Declaration of Helsinki.

The children were evaluated from information recorded on electronic forms regarding visual acuity (VA), refractive error, lens status, orthoptic status, general health problems (eg, heart disease, intake of medications), ocular risk factors and anomalies of the anterior and posterior part of the eye. Children with a subluxation of the lens were included for follow-up analysis regarding the course of VA, visual development as well as the surgical methods applied and results of the surgery.

2.1 | Study variables and surgical technique

The medical history, age, sex, changes in treatment or new onset of disease of the children were obtained from the data for every visit. Medical records included VA at every time point for both eyes and the refractive error or change in refraction of the patients. The full orthoptic status was recorded and a standard ophthalmological examination including evaluation of the anterior and posterior part of the eye performed.

Surgery was considered if one or more of the following criteria were observed: (a) any decrease in VA or best-corrected VA (BCVA) of less than 0.3; (b) lens opacification interfering with VA; (c) monocular diplopia; (d) progressive subluxation of the lens affecting the pupillary axis with or without elevation of intraocular pressure (IOP). These criteria were in accordance with criteria for surgery in the literature.⁹⁻¹¹ No surgical intervention was done if the subluxation of the lens was not visible without pupil dilatation in combination with a good VA. The indications for primary or secondary implantation of the IOL depended on the age of the patients and the individual patient need. Primary implantation was suggested and discussed when patients were 2 years of age or older.

All surgeries were performed under general anaesthesia. Surgical intervention was done by two surgeons and comprised lensectomy including anterior vitrectomy with/without IOL implantation. Pupils were dilated before the operation with tropicamide (Mydramide) and phenylephrine. A 2.4-mm clear cornea incision was made at the

temporal position. After instillation of viscoelastic material into the anterior chamber, paracentesis was done at the 12 o'clock position followed by a curvilinear opening of the anterior capsule (capsulorhexis). After hydrodissection with a balanced salt solution between the capsule and the cortex to allow the lens to rotate, the nucleus and cortex were aspirated bimanually within the capsular bag using the aspiration mode. Caution was taken not to damage the lens capsules until the entire lenticular material had been removed and the capsular bag emptied. Iris retractor hooks were used in cases with severe zonular weakness to stabilize the capsular bag. The capsular bag was removed in cases of complete or extensive zonular dialysis (more than 6 clock hours), significant impairment of the optic axis or if IOL implantation was hindered. Anterior vitrectomy was done in cases of questionable residual vitreous in the pupillary area. The lens capsules and the anterior vitreous were removed using the cutting mode. After the lensectomy and the anterior vitrectomy were completed, incisions were hydrated and the wounds closed with a 10-0 nylon suture. Visual rehabilitation started immediately after surgery. Contact lenses and/or glasses were prescribed for correction of the aphakia. Treatment of amblyopia was initiated immediately thereafter whenever necessary.

In the case of Artisan implantation (aphakic iris-claw lens, Ophthec BV, Groningen, the Netherlands), scleral corneal incision was made at the 12 o'clock position and the incision extended to 5.5 mm. After instillation of viscoelastic material, paracentesis was done at the nasal and temporal superior quadrant. Anterior vitrectomy was performed, and the IOL was inserted into the anterior chamber. After instillation of Miochol for constriction of the pupil the Artisan lens was enclaved retropupillary in a horizontal position. Postoperative treatment included 1% atropine once daily and 0.05% dexamethasone sodium phosphate four times daily for 4 weeks, after which the treatments were discontinued.

2.2 | Statistical analysis

Statistical analysis was performed using descriptive statistics for all variables and data frequency. The aim of the analysis was to describe the ocular complications and the VA and surgical outcomes of children with a genetically proved diagnosis of Marfan syndrome. Baseline variables were analysed for all patients with Marfan syndrome. The course of VA over time was determined for children with ectopia lentis with or without the need for surgical intervention. The change in VA over time was calculated using paired two-sided *t*-tests. The level of significance was set to 0.05. Patients were divided into age groups (0-3 years, 4-10 years and >10 years) and descriptive statistics for variables was performed. Mean VA was calculated at baseline and the final

follow-up. No *t*-test was done due to the limited patient number in the subgroup analysis. Excel and SPSS (IBM Statistics, Version 23) for windows were used for all analyses and graphs.

3 | RESULTS

We included 82 eyes of 41 patients in this retrospective analysis. The mean age was 12.3 ± 9 years and the mean follow-up of the patients with subluxation of the lens was 3 years (range 1-7 years). Table 1 shows the demographic data of all patients with Marfan syndrome at initial presentation.

3.1 | Ocular and systemic complications

Coloboma of the lens was visible in three (7%) patients, one (2%) patient had corneal thinning, two (5%) presented with nystagmus and one (2%) needed pressure lowering medication due to glaucoma. No patient presented with or developed retinal detachment, keratoconus or presented with congenital cataract.

Regarding the systemic complications of Marfan syndrome, eight of 41 children (20%) had cardiovascular disease including mitral valve insufficiency, aortic dilatation or aneurysm. Three (7%) children had skeletal deformations including scoliosis and breastbone abnormalities. One (2%) patient showed developmental delay of fine motor and language skills.

TABLE 1 Demographic data of the patients with Marfan syndrome (n = 41) at initial presentation

Age (mean \pm SD)	12.3 \pm 9
Sex, n (%)	
All (n = 41; male/female)	19/22 (46/54)
0-3 years (n = 11)	4/7 (36/64)
4-10 years (n = 13)	5/8 (38/62)
>10 years (n = 17)	10/7 (59/41)
Refractive error	
Myopia <10 dpt	27 (66)
Myopia >10 dpt	7 (17)
Other (emmetropia, hyperopia)	7 (17)
Astigmatism 0 to +2	32 (78)
Astigmatism +2 to +4	7 (17)
Astigmatism +4 to +6	2 (5)
Squint status	
Esotropia	2 (5)
Exotropia	2 (5)
Exophoria	5 (12)

Abbreviation: dpt, diopters.

TABLE 2 Lens status of the patients with Marfan syndrome

	Number of patients, n (%)
Lens status (n = 41)	
Normal lens	14 (34)
Bilateral subluxation	27 (66)
Subluxation of the lens among age groups	
0–3 years (n = 11)	10 (91)
4–10 years (n = 13)	9 (69)
>10 years (n = 17)	8 (47)
Ectopia lentis (n = 27)	
Superotemporal subluxation	16 (59)
Superonasal subluxation	6 (22)
Superior subluxation	4 (15)
Inferior subluxation	1 (4)

3.2 | Lens status and lensectomy

Table 2 presents the lens status of all patients with Marfan syndrome and patients grouped 0–3 years (n = 11), 4–10 years (n = 13) and >10 years (n = 17, range 11.5–21.8). Twenty seven (66%) of the 41 patients presented with subluxation of the lens. All of them presented with bilateral subluxation. Fourteen (52%) of the 27 patients with a mean age of 7 ± 6 years were referred for lensectomy due to significant subluxation of the lens. Same-day bilateral surgery was performed in seven (50%) of the 14 children and an IOL was implanted in six (43%) of them. Surgery was performed within 6 months after initial presentation. Thirteen (48%) of the 27 patients showed subluxation of the lens and did not need a lensectomy. Figure 1 shows a

representative image of a 15-year-old patient with ectopia lentis in Marfan syndrome.

Subgroup analysis was done divided into the age groups 0–3 years (n = 11), 4–10 years (n = 13) and >10 years (n = 17). Four (29%) of the patients who received lensectomy were in the group 0–3 years, seven (50%) in the group 4–10 years and three (21%) in the group >10 years. No patient in the group 0–3 years received primary IOL implantation. A primary IOL implantation was done in two (33%) patients in the group 4–10 years and four (67%) in the group >10 years.

No case of retinal detachment was observed following surgery. A macular oedema requiring intravitreal steroid treatment developed in one patient (7%) after surgery. Repositioning of the Artisan IOL became necessary in one patient (7%).

3.3 | Subluxation without lensectomy

Table 3 shows the characteristics of patients who did not need a lensectomy. 67% of the patients showed minor superotemporal subluxation of the lens, 25% a superonasal and 8% an inferotemporal subluxation. A total of 92% of patients showed myopia of less than 10 diopters (dpt) and were managed with spectacles for refractive correction and amblyopia prevention. No patient showed astigmatism of >4 dpt. Patching of one eye became necessary in 33% (n = 4) of the patients.

3.4 | Visual acuity

VA varied from 1.2 to –0.1 logMAR at first examination. The mean VA at initial presentation was 0.2 ± 0.5 for all

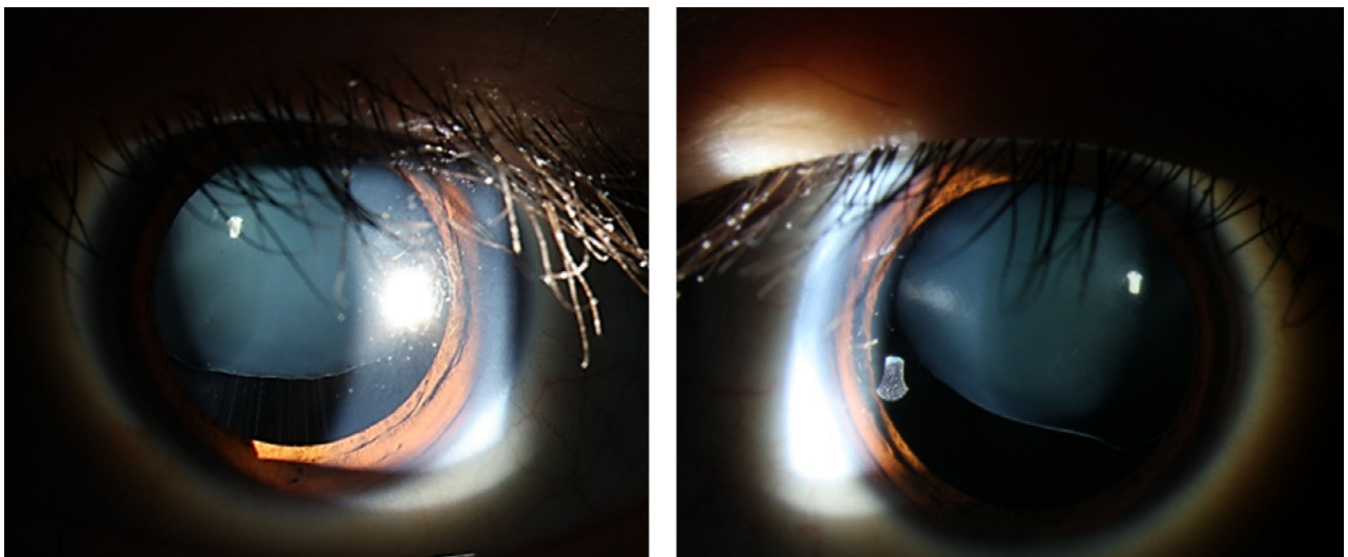


FIGURE 1 Representative image of a 15-year-old patient with ectopia lentis in Marfan syndrome. VA at last follow-up: 0.6/1.0 (right/left) Snellen equivalent; Myopia of –8.5/–7.0 dpt; positive tests for stereopsis. No lensectomy was performed due to the good visual development

TABLE 3 Characteristics of patients with subluxation of the lens who did not need a lensectomy (n = 12)

Age (mean ± SD)	12 ± 10
Refractive error, n (%)	
Myopia <10 dpt	11 (92)
Myopia >10 dpt	1 (8)
Other (emmetropia, hyperopia)	0 (0)
Astigmatism 0 to +2	8 (67)
Astigmatism +2 to +4	4 (33)
Astigmatism +4 to +6	0 (0)
Squint status	
Esotropia	0 (0)
Exotropia	1 (8)
Exophoria	3 (25)

Abbreviation: dpt, diopters.

children with Marfan syndrome (n = 41). At initial presentation, the mean VA was 0.1 ± 0.7 in patients with a normal lens status (n = 14) and 0.3 ± 0.5 in patients with subluxation of the lens (n = 24) ($P < .01$). Subgroup analysis of patients in age groups 0-3 years (n = 11), 4-10 years (n = 13) and >10 years (n = 17) showed a mean initial VA of 0.5 ± 0.6 , 0.2 ± 0.6 and 0.1 ± 0.6 logMAR. Three children who were included in the baseline evaluation were excluded from VA analysis due to insufficient data. Table 4 shows the VA of patients with subluxation of the lens at initial presentation and the final follow-up. Table 5 shows a summary of the VA of children with Marfan syndrome and subluxation of the lens.

Subgroup analysis of the patients in the age groups 0-3 years (n = 9), 4-10 years (n = 7) and >10 years (n = 8) was done for patients with a subluxation of the lens receiving lensectomy and for patients who did not need a lensectomy. The mean VA of patients receiving lensectomy was initially 0.5 ± 0.7 logMAR in the group 0-3 years, 0.7 ± 1.0 in the group 4-10 years and 0.1 ± 0.7 in the group >10 years. It was 0.3 ± 0.7 , 0.15 ± 1.0 and 0.15 ± 0.5 at

TABLE 4 Visual acuity (logMAR) of patients with subluxation of the lens at initial presentation and final follow-up

Subluxation of the lens (n = 27)	Initial presentation (right/left), mean ± SD	Final follow-up, mean ± SD	P-value
Patients receiving lensectomy ^a (n = 12)	0.5 ± 0.5	0.2 ± 0.5	.02
Patients without lensectomy (n = 12)	0.2 ± 0.5	0.1 ± 0.5	.06

^aWith/without secondary IOL implantation.

TABLE 5 Visual acuity (VA) of children with Marfan syndrome and ectopia lentis

VA of children with ectopia lentis (n = 24)	Initial	Last follow-up
VA > 0.3 logMAR (all)	50% (n = 12)	29% (n = 7)
VA ≤ 0.3 logMAR (all)	50% (n = 12)	71% (n = 17)
VA > 0.3 logMAR (lensectomy)	58% (n = 7)	42% (n = 5)
VA ≤ 0.3 logMAR (lensectomy)	42% (n = 5)	58% (n = 7)
VA > 0.3 logMAR (without lensectomy)	42% (n = 5)	17% (n = 2)
VA ≤ 0.3 logMAR (without lensectomy)	58% (n = 7)	83% (n = 10)
VA improvement		
All (n = 24)		79% (n = 19)
Eyes receiving lensectomy (n = 12)		83% (n = 10)
Eyes without lensectomy (n = 12)		75% (n = 9)
Follow-up		
1-3 years		46% (n = 11)
3-5 years		33% (n = 8)
>5 years		21% (n = 5)
Mean follow-up		3 years

the final follow-up. The mean VA of patients who did not need a lensectomy was initially 0.5 ± 0.5 , 0.2 ± 1.3 and 0.2 ± 1.3 . It was 0.15 ± 1.0 , 0.1 ± 0.8 and 0.10 ± 0.5 at the final follow-up.

Subgroup analysis of patients with primary and secondary IOL implantations showed a final VA of 0.2 ± 0.5 in the primary IOL group and 0.2 ± 0.4 in the secondary IOL group, which was not significantly different.

4 | DISCUSSION

Ectopia lentis is often the first clinical sign of Marfan syndrome and is a key symptom. The management of subluxation of the lens in patients with Marfan syndrome differs depending on preoperative findings and disease development, with the degree of lens dislocation and function playing an important role. Surgical removal of the lens becomes necessary whenever a good VA cannot be achieved and/or complications such as raised IOP develop. Our data present the outcome of patients with Marfan syndrome and subluxation of the lens with or without lensectomy and show a good functional outcome with visual improvement in the majority of the children and a low rate of postoperative complications. From 27 children with subluxation of the lens, 56% received lensectomy with or without secondary implantation of an IOL.

Some studies with/without a limited number of patients have reported the functional outcome after lensectomy with/without secondary IOL implantation in Marfan syndrome. A postoperative BCVA of $>20/30$ was reported in all children in a study in 2014 after a mean time of 5 years with aphakic correction.¹² A review including nine patients with subluxation of the lens secondary to Marfan syndrome showed significant improvement of VA (0.5 ± 0.3 logMAR at the baseline to 0.2 ± 0.2 logMAR after surgery) in lensectomy with secondary Artisan IOL implantation.¹³ VA improved from 0.5 to 0.3 logMAR in a cohort of 39 patients who were treated with 23G-vitreotomy lensectomy with and without Artisan implantation.¹⁴ The follow-up time in these studies varied between 44 months and 3 years.^{13,14} In our analysis over a mean period of 3 years, we report the outcome of patients with ectopia lentis in Marfan syndrome who received lensectomy as well as the outcome of patients who did not need a lensectomy. VA improved in 83% of patients receiving lensectomy (with/without secondary Artisan implantation) and in 75% of patients who did not need a lensectomy, with 71% reaching a final VA of <0.3 logMAR. VA in patients with subluxation of the lens who did not need a lensectomy improved from 0.2 ± 0.5 to 0.1 ± 0.5 , with 75% of eyes reaching a final VA of <0.3 logMAR.

The surgical methods for removing the lens in Marfan syndrome vary and are challenging due to weakness or instability of the zonular fibres and occurrence of loss of the capsular bag. Visual rehabilitation after surgery with contact lenses, glasses or secondary implantation of an IOL needs to be carefully managed to avoid amblyopia and achieve good VA results. The options for lens implantation include in-the-bag IOL placements, sclera-fixed posterior chamber IOLs, anterior chamber open-loop IOLs, and anterior chamber iris claw IOLs. 33% of the patients in our analysis received IOL implantations, all of which were anterior chamber iris claw IOLs. Generally, implantation of the IOL was done within one surgery in older children and younger children were left aphakic with secondary IOL implantation done later in life. The intra- and postoperative characteristics of children less than 3 years of age may differ from older children. The factors that differ include the axial length, diameter of the lens and size of the capsular bag. Use of IOLs may lead to over-sizing in the groups aged 0-3 years and the incidence of postoperative complications has been reported to be higher in younger children undergoing cataract surgery.^{15,16} We found no difference in postoperative complications in association with the younger age groups among our patients with ectopia lentis due to Marfan syndrome.

No patient in the age group 0-3 years received primary implantation of an IOL. IOLs are the standard-of-care for optical correction of aphakia in older children according to data published in 2010 by the Infant Aphakia Treatment

Study that assessed the best treatment for children with unilateral cataracts. Higher postoperative complications with IOL implantation compared with contact lens correction of aphakia have been reported for younger children. In addition, the VA outcome and the appropriate IOL power in younger children is still unknown. However, information on the management of younger children with Marfan syndrome and weak zonules is still lacking and cannot be directly compared to the management of patients with unilateral cataracts. The management of aphakia has to be decided based on individual patient need and IOL implantation considered and discussed whenever primary IOL implantation is a better option due, for example, to compliance or family circumstances.¹⁷⁻²⁰ The differences in management and complications among different age groups in patients with Marfan syndrome are of special clinical importance. However, we only did descriptive analysis because the number of patients in the subgroups of our investigation was limited.

Management of ectopia lentis in Marfan syndrome has risks and is challenging due to zonular weakness. Additive procedures can be used to ensure safe surgery with good IOL stability. Iris retractor hooks were used to stabilize the capsular bag in our series in cases with severe zonular weakness. Iris retractor hooks are easily inserted and removed making them excellent tools for decreasing the risk of complications due to an unstable capsular bag.²¹ The capsular bag was removed in the event of a complete or extensive zonular dialysis, pronounced impairment of the optic axis or if IOL implantation was hindered. Nevertheless, sclera-fixated capsular tension rings are another possibility to stabilize the capsular bag if zonular fibre weakness is severe and to give stability for IOL implantation in ectopia lentis.²² Good functional outcomes with low complication rates with both iris retractor hooks and capsular tension rings have been reported in the management of ectopia lentis in paediatric patients.^{21,23,24}

The retropupillary position of the IOL in our patient cohort was chosen for increased endothelium protection. Studies have reported 11% to 13% rates of endothelial cell loss after anterior iris-claw IOL implantation.^{8,25} Compared with retropupillary IOLs, a lower risk of endothelial cell loss was reported together with good functional outcomes.^{26,27} Endothelial cell count was not routinely done in our cohort of patients with retropupillary IOL implantation.

Various studies have evaluated the outcome of patients with subluxation of the lens undergoing lensectomy who did not receive IOL implantation. The concerns raised in these studies regarding the implantation of anterior chamber IOLs include retinal detachment, secondary glaucoma and endothelial cell loss. However, one study showed improvement in VA in children with Marfan syndrome who received anterior chamber IOL implantations, with no complications after the implantation. A study comparing the rate of retinal

detachment in patients with Marfan syndrome who were aphakic versus pseudophakic showed no difference between the groups in the occurrence of this complication.^{28,29} VA improvement with no postoperative complications was reported in patients with subluxated lenses who were followed for up to 45 months.³⁰ Only one complication, which was retinal detachment, was observed among 37 children with subluxated lenses (27 with Marfan syndrome) 2 years after surgery.⁹ This accords with our low number of complications after lensectomy and implantation of an IOL. No patient developed retinal detachment whether after lensectomy or after lens implantation. One patient developed postoperative macular oedema after Artisan implantation and re-fixation of the IOL became necessary in one patient.

Ocular manifestations or complications vary among patients with Marfan syndrome and can include subluxation of the lens, a moderate to high degree of myopia, astigmatism, strabismus and a risk for retinal detachment, glaucoma, keratoconus, corneal thinning and early cataracts. In our study, we observed a high rate of subluxation of the lens and myopia and a low rate of astigmatism and strabismus. At the time of presentation, no patient had retinal detachment, early cataract or keratoconus. One patient showed raised IOP which was controlled with local pressure lowering medication and another had corneal thinning.

The limitation of this study that needs to be mentioned includes its retrospective nature. Furthermore, the subgroup analysis comprised a limited number of patients, too small to draw general conclusions. However, with this data from a mean follow-up of 3 years we could show the various aspects of Marfan syndrome and ocular complications as well as the outcome of patients with subluxation of the lens treated with lensectomy or patients who did not need lensectomy. Good functional outcomes in both patients receiving lensectomy and patients without surgery were demonstrated.

CONFLICT OF INTEREST

None declared.

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REFERENCES

- Dietz H. Marfan syndrome. In: Adam MP, Ardinger HH, Pagon RA, et al. eds. *Gene Reviews R* [Internet]. Seattle, WA: University of Washington; 2001.
- Ramirez F, Dietz HC. Marfan syndrome: from molecular pathogenesis to clinical treatment. *Curr Opin Genet Dev.* 2007;17:252-258.
- Pepe G, Lapini I, Evangelisti L, et al. Is ectopia lentis in some cases a mild phenotypic expression of Marfan syndrome? Need for a long-term follow-up. *Mol Vis.* 2007;13:2242-2247.
- Lynas MA. Marfan's syndrome in Northern Ireland: an account of thirteen families. *Ann Hum Genet.* 1958;22:289-309.
- Fuchs J. Marfan syndrome and other systemic disorders with congenital ectopia lentis. A Danish national survey. *Acta Paediatr.* 1997;86:947-952.
- Pepe G, Giusti B, Sticchi E, et al. Marfan syndrome: current perspectives. *Appl Clin Genet.* 2016;9:55-65.
- Maumenee H. The eye in the Marfan syndrome. *Trans Am Ophthalmol Soc.* 1981;79:684-733.
- Menezo JL, Cisneros AL, Rodriguez-Salvador V. Endothelial study of iris-claw phakic lens: four year follow-up. *J Cataract Refract Surg.* 1998;24:1039-1049.
- Halpert M, BenEzra D. Surgery of the hereditary subluxated lens in children. *Ophthalmology.* 1996;103(4):681-686.
- Anteby I, Isaac M, BenEzra D. Hereditary subluxated lenses: visual performances and long-term follow-up after surgery. *Ophthalmology.* 2003;110(7):1344-1348.
- 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.
- Miraldi Utz V, Coussa RG, Traboulsi EI. Surgical management of lens subluxation in Marfan syndrome. *J AAPOS.* 2014;18:140-146.
- Rabie HM, Javadi MA, et al. Visual outcomes after lensectomy and iris claw artisan intraocular lens implantation in patients with Marfan syndrome. *Int Ophthalmol.* 2017;37:1025-1030.
- Fan F, Luo Y, Liu X, Lu Y, Zheng T. Risk factors for postoperative complications in lensectomy-vitreotomy with or without intraocular lens placement in ectopia lentis associated with Marfan syndrome. *Br J Ophthalmol.* 2014;98:1338-1342.
- Sinsky RM, Stoppel JO, Amin PA. Ocular axial length changes in a pediatric patient with aphakia and pseudophakia. *J Cataract Refract Surg.* 1993;19:787-788.
- Chrousos GA, Parks MM, O'Neill JF. Incidence of chronic glaucoma, retinal detachment and secondary membrane surgery in pediatric aphakic patients. *Ophthalmology.* 1984;91:1238-1241.
- Lambert SR, Buckley EG, Drews-Botsch C, et al. A randomized clinical trial comparing contact lens with intraocular lens correction of monocular aphakia during infancy: grating acuity and adverse events at age 1 year. Infant Aphakia Treatment Study Group. *Arch Ophthalmol.* 2010;128(7):810-818.
- BenEzra D. Cataract surgery and intraocular lens implantation in children, and intraocular lens implantation in children. *Am J Ophthalmol.* 1996;121(2):224-226.
- Lambert SR, Lynn M, Drews-Botsch C, et al. A comparison of grating visual acuity, strabismus, and reoperation outcomes among children with aphakia and pseudophakia after unilateral cataract surgery during the first six months of life. *J AAPOS.* 2001;5(2):70-75.
- Autrata R, Rehurek J, Vodickova K. Visual results after primary intraocular lens implantation or contact lens correction for aphakia in the first year of age. *Ophthalmologica.* 2005;219(2):72-79.
- Hoffman RS, Snyder ME, Devgan U, et al. Management of the subluxated crystalline lens. *J Cataract Refract Surg.* 2013;39(12):1904-1915.



22. Cionni RJ, Osher RH. Endocapsular ring approach to the subluxed cataractous lens. *J Cataract Refract Surg*. 1995;21:245-249.
23. Konradsen T, Kugelberg M, Zetterström C. Visual outcomes and complications in surgery for ectopia lentis in children. *J Cataract Refract Surg*. 2007;33(5):819-824.
24. Kim EJ, Berg JP, Weikert MP, et al. Scleral-fixated capsular tension rings and segments for ectopia lentis in children. *Am J Ophthalmol*. 2014;158(5):899-904.
25. Koss MJ, Kohnen T. Intraocular architecture of secondary implanted anterior chamber iris-claw lenses in aphakic eyes evaluated with anterior segment optical coherence tomography. *Br J Ophthalmol*. 2009;93:1301-1306.
26. Forlini M, Soliman W, Bratu A, Rossini P, Cavallini GM, Forlini C. Long-term follow-up of retropupillary iris-claw intraocular lens implantation: a retrospective analysis. *BMC Ophthalmol*. 2015;15:143.
27. Gonnermann J, Klamann MK, Maier AK, et al. Visual outcome and complications after posterior iris-claw aphakic intraocular lens implantation. *J Cataract Refract Surg*. 2012 Dec;38(12):2139-2143.
28. Anteby I, Isaac M, BenEzra D. Hereditary subluxated lenses: visual performance and long-term follow-up after surgery. *Ophthalmology*. 2013;110:1344-1348.
29. Morrison D, Sternberg P, Donahue S. Anterior chamber intraocular lens (ACIOL) placement after pars plana plana lensectomy in pediatric Marfan syndrome. *J AAPOS*. 2005;9:240-242.
30. Peyman GA, Raichard M, Goldberg MF, et al. Management of subluxated and dislocated lenses with the vitrophage. *Br J Ophthalmol*. 1979;63:771-778.

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