

Bilateral lens coloboma in child with Marfan's syndrome treated with lens surgery

ABSTRACT

Lens coloboma can occur with ocular or systemic associations like Marfan's and Stickler's syndrome. Causes of poor vision include refractive error, anisometropia and amblyopia. Visual rehabilitation and surgery is technically challenging in these patients. Here we report a child with progressive defective vision with Marfan's syndrome and lens coloboma who underwent successful surgery with intraocular lens (IOL) placement. The bag was stabilised with capsular tension ring and iris hooks, and the IOL was further stabilised with optic capture. Postoperatively, the child regained good vision in the operated eye with very good centration of the IOL at three month follow up visit.

Keywords: Capsular tension ring, lens coloboma, Marfan's syndrome

INTRODUCTION

Lens coloboma in Marfan's syndrome is infrequently reported.^[1,2] This is due to defective or absent development of the zonules and ciliary body.^[3] Surgery is technically challenging and successful, intraocular lens (IOL) placement depends on coloboma's extent and usage of capsular supportive devices like iris hooks, capsular tension ring (CTR), Ciannis ring or segments. We present the successful visual rehabilitation following IOL placement in the left eye of a child with the above condition.

CASE REPORT

A 13-year-old boy [Figure 1] with Marfan's syndrome presented with progressively decreasing vision in his left eye of unknown duration. His best corrected visual acuity (BCVA) was 20/40 (-0.75/-3.5@180) and 20/125 (-8.0/-3.0@180) in his right eye (RE) and left eyes (LE), respectively. Slit lamp evaluation under mydriasis showed clear lens with coloboma extending from 12 to 2 o'clock (RE) and 9-1 o'clock (LE) [Figure 2] with few intact zonules. As left eye BCVA was low, we decided on surgery with IOL placement, after explaining possibility of subnormal visual outcome due to amblyopia.



Figure 1: (a) Photograph showing a 13-year-old male with tall and thin stature. He had disproportionately long arms (b) with hyper extensibility of fingers

Following lens aspiration the capsular bag was stabilised with CTR (AuroLab, Madurai, India) [Figure 3], inserting

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Submitted: 18-May-2021 Accepted: 26-May-2021 Published: 30-Aug-2022

Access this article online	
Website: www.kjophthal.com	Quick Response Code 
DOI: 10.4103/kjo.kjo_120_21	

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How to cite this article: Ganesh SC, Krishnababu, Elizabeth S, Jogitha. Bilateral lens coloboma in child with Marfan's syndrome treated with lens surgery. Kerala J Ophthalmol 2022;34:176-7.

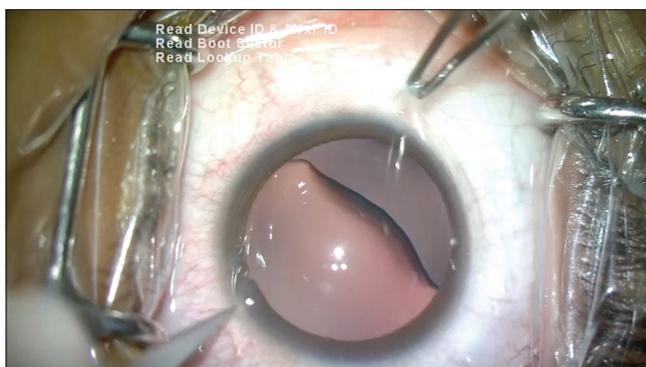


Figure 2: Picture showing lens coloboma in left eye from 9 to 1 o' clock position with intact zonules. The vision in his left eye was 20/125 (-8.0/-3.0@180)

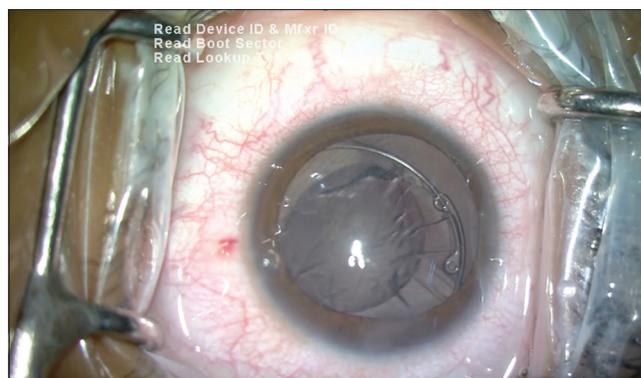


Figure 3: The capsular bag was stabilised with the capsular tension ring (Aurolab, Madurai, India), prior to aspiration of the lens cortex

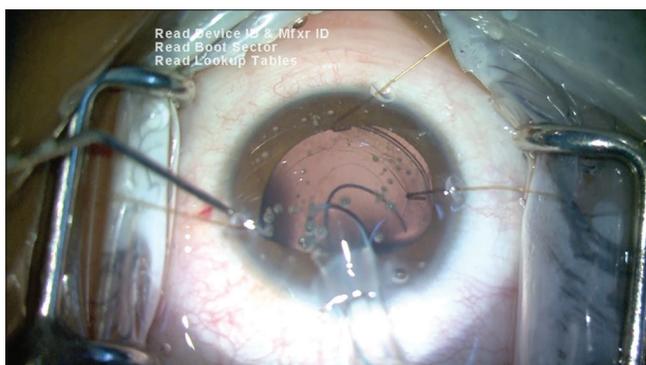


Figure 4: Three Iris hooks were used for dilatation and stabilisation of capsular bag prior to insertion of intraocular lens (Alcon Labs, Texas, USA)



Figure 5: Picture showing a well centred Intra ocular lens with capsular tension ring in the bag. Best corrected vision has improved from 20/125 to 20/63 with negligible astigmatism postoperatively (-0.75DCyl@165)

which was technically challenging due to small and irregular colobomatous bag. Three iris hooks were used to stabilise the bag prior to three piece acrylic IOL (Alcon Labs, Texas, USA) implanted in sulcus with optic capture [Figures 4 and 5].

Postoperatively, visual acuity in his LE improved to 20/63 (-0.75DCyl@165) with well centered IOL.

DISCUSSION

Visual rehabilitation is technically challenging due to capsular fornix aspiration, zonular dialysis extension, vitreous herniation and IOL decentration.^[3] Lens coloboma in Marfan's poses the difficulty of a small sized capsular bag with notching, in addition to the difficulties encountered in subluxated lens management. Also there exists a possibility of late decentration of IOL placed entirely in bag due to progressive myopia. Hence we decided on bag sulcus fixation of IOL, to aid in long term stability and centration.

The improvement in BCVA post surgery implies that the high myopia causing vision loss may have occurred beyond the amblyogenic years of his life.

IOL implantation can thus be an effective option to provide optimal visual rehabilitation in eyes with progressive vision loss in lens coloboma, in expert hands.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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