International Journal Dental and Medical Sciences Research
Volume 4, Issue 5, Sep-Oct 2022 pp 468-470 www.ijdmsrjournal.com ISSN: 2582-6018

Management of Ectopia Lentis in a case of Marfan syndrome

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Submitted: 10-10-2022 Accepted: 22-10-2022

ABSTRACT:
This was a case of 14 years old male presented to Department of ophthalmology Acharya Vinod Bhave Rural Hospital, Sawangi, India on 6th of April with Diminution of vision for 5 years. Patient was examined and had high Arched Palate ,long Axial skeleton and Bilateral Ectopia Lentis keeping in these patient was Diagnosed as a case of Marfans syndrome. Patient was given refraction and prescribed glasses for Improvement of vision and was referred to department of cardiology for further investigation and Management as it is important to have a multi-disciplinary approach in a case of Marfans syndrome to avoid any dangerous or life threatening complications before going for surgery for Ectopia Lentis.
Key words: Marfans syndrome, Ectopia Lentis

I. CASE PRESENTATION:

A 14 year old Male Patient presented to ophthalmology outpatient department with complaints of diminution of vision in both eyes for 5 years. Late presentation was due to financial reasons. Patient had no cardiac history of breathlessness and fatiguability. Developmental hiatory and behaviour was normal for Age. The child was first born in a family of two and had no similar cases in his family.

On Physical examination Patient was thin built, tall, with long Extremities. The patient also had other features high Arched Palate and Long fingers (arachnodactyly)

Cardiovascular examination of the patient was done which was within normal limits, ECG was normal, Echocardiography showed good LVEF and was normal for this Patient. Chest was clear and no abnormality was detected in Abdominal and CNS examination. Patient was given fitness for surgery for ectopia lentis.

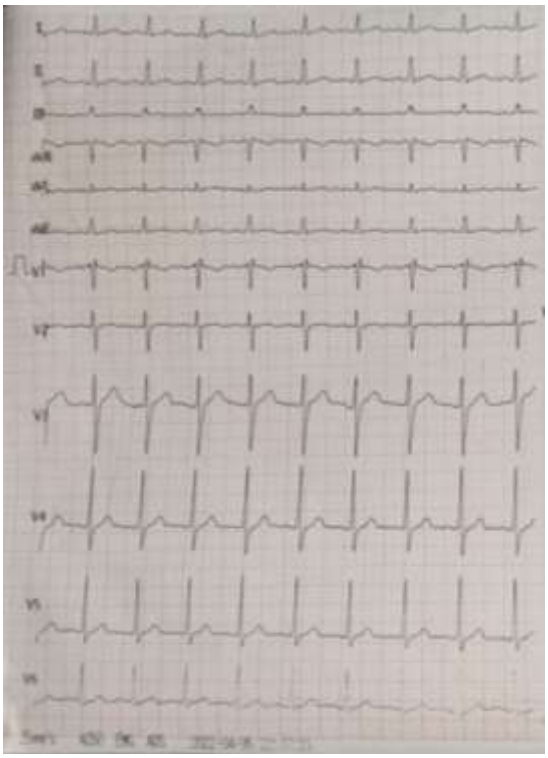

Fig1-ECG of the Patient

Ocular Examination was done and the best corrected visual acuity in both eyes was 6/60. On slit lamp examination both eyes had temporal dislocation of lens and Zonules could be seen. Anterior segment in both eyes were quiet, Pupils were reactive to light. Fundus examination on dilatation showed pink optic disc with 0.3 CDR and flat retina. Intraocular Pressure could not be assessed but appeared normal on Digital Tonometry. Patient was advised to undergo Lens Extraction with IOL implantation. A-scan was done after Keratometry readings to calculate the IOL power for the Patient.

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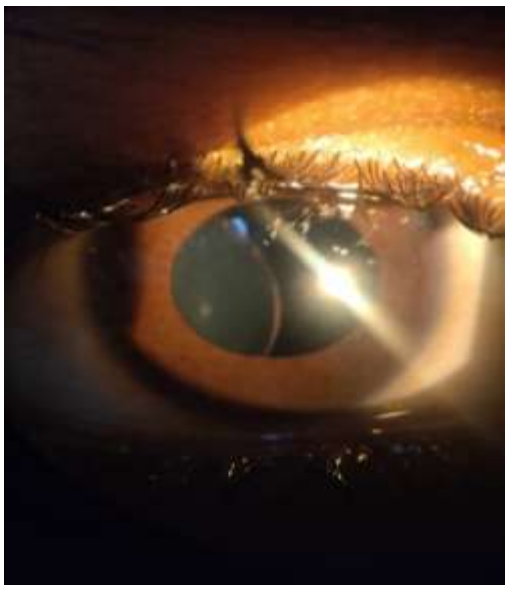
Fig 2(a)-Temporal subluxation of right eye lens

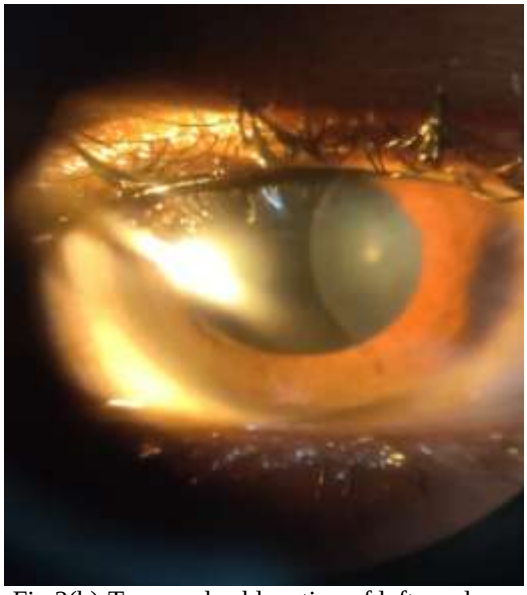
Fig 2(b)-Temporal subluxation of left eye lens

Patient was taken for Surgery the following day under Peri bulbar block. Posterior capsule could not be separated from the lens and therefore ICCE was done followed by Anterior vitrectomy in right eye. SFIOL was implanted following lenticular extraction. Eye was Padded and opened the next day. On post-op day 1 Corneal edema was present, IOL was central in position, Patient was giving vision of 6/36 with pinhole. Post-op day 3 Corneal edema had subsided and the Pin hole vision was now of 6/18 and Patient was discharged.

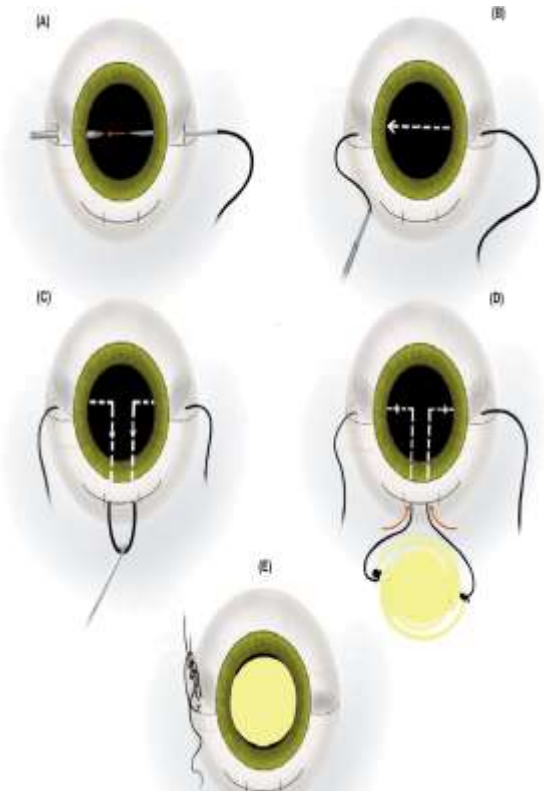
Fig-3 Steps of SFIOL Procedure.

Patient had come for Follow-up after 15 days when Anterior chamber was quiet, unaided vision was 6/18 and improved to 6/9 with pinhole. Patient was advised refraction after another 15 days.On follow up after 1 month anterior chamber was quiet and best corrected visual acuity was of 6/9 ,after which patient was advised to undergo lens extraction with IOL implantation in left eye.

II. DISCUSSION:

Marfans syndrome is a genetic disorder of connective tissue origin mostly affecting skeletal, cardiovascular and ocular systems [1]. It can also affect other tissues like skin, adipose , lung and muscle [2,3]. Occurrence of Marfans syndrome is of 8-10 per 1 lakh people annualy [4]. Cause of marfans was identified to be fibrillin-1 (FBN-1) gene mutation on chromosome number 15. Fibrillin is a glycoprotein which provides elasticity and structural support to connective tissue of eye [5]. Marfan syndrome does not have unique symptoms or signs, but mainly diagnosed with the dislocated lens (ectopia lentis), long fingers (arachnodactyly) and elongated limbs (dolichostenomelia) and aortic root dilatation seen in some cases . various other clinical features are associated with this syndrome, mainly involving skin, skeleton and the joints.

Marfan syndrome can lead to poor vision due to subluxation of lens, retinal detachment, and

myopia [6]. Almost half of the cases of this syndrome are diagnosed by ophthalmologists. Early Intervention is required to avoid Amblyopia although patient presented late which might have led to amblyopia in this patient.

Refractive error in marfan due to subluxation can be managed with refractive glasses but in most of the cases surgery is indicated. Lens extraction with IOL implantation is donewhere IOLcould be of various methods in a patient with no zonular or posterior capsule support, these include Iris claw lens, ACIOL, Scleral fixating IOL (SFIOL). Like in this case of marfans SFIOL was implanted into the eye.

SFIOL with sutures have showen to have good results in the outcome of cases with ectopia lentis in Marfans syndrome, but also there has been some amount of Decentralisation of lens due to the sutures or absorption of the sutures in later years, though there are these difficulties it still is a better option than Iris claw or ACIOL which is why it was preffered in this case of Marfans syndrome.

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