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Case Report

In bag IOL implantation with Cionni ring: A case report

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ABSTRACT

The supertemporal subluxation of the lens due to zonular weakness is one of the hallmarks of ocular signs with Marfan's syndrome. Cionni capsular tension ring (CTR) implantation can help to overcome this issue and correct the capsular bag decentration. We describe the visual outcome, intra and post-surgical findings of phacoemulsification with Cionni capsular tension ring and PCIOL implantation in a case of an 18-year-old-boy presented with the features of a typical Marfan's syndrome.

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1. Introduction

Marfan syndrome (MFS) is a genetic disease affecting the connective tissues. The incidence of MFS is 0.3% among the general population.¹ Fibrillin gene mutation is the main culprit of MFS, and the patient is commonly suffered from ectopia lentis (EL) or lens subluxations from the age of 3 years.²

The management of EL is one of the most crucial anterior segment surgeries. The transscleral suture fixated IOL implantation was the acceptable operating method previously. Postoperative Retinal detachment and secondary glaucoma are the reported high-risk complications.³ Moreover, per-operatively the needle needs to be passed through the vascular uveal tissue which may cause bleeding and great trauma.⁴

When performing anterior segment surgeries, implanting the Cionni-modified capsular tension rings (MCTR) has become a preferred practice instead of transscleral fixated

IOL implantation for the preservation of capsular bags of an MFS patient.⁵ As a simplified operation, MCTR implantation in a capsular bag is a simpler operating technique with minimal trauma.⁶

2. Case Presentation

An 18-year-old male, presented us with decreased vision in both his eyes (right more than left) since childhood. On ocular examination, the vision was 1/60 in his left eye and counting fingers 3 meters in the right eye, with clear cornea in both eyes. After pharmacological dilatation of the pupil for details examination, we found superior-temporal subluxations of the lens in both eyes (Figures 3 and 4). On physical examinations, the positive wrist sign (Figure 1) was present; the patient was tall (178 cm) with a thin build, had thin spider-like fingers and had a large arm span (Figure 2) to height (1.07).

A history of similar features was reported in his father and brother also. Echocardiogram revealed mild mitral and tricuspid valve regurgitation. So, the clinical diagnosis was

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Fig. 1: Positive wrist sign



Fig. 2: Large arm span in relation to height ratio



Fig. 3: Superotemporal subluxation of the lens in the right eye

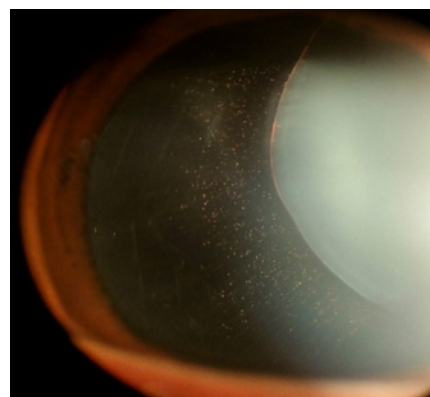


Fig. 4: Superotemporal subluxation of the lens in the left eye

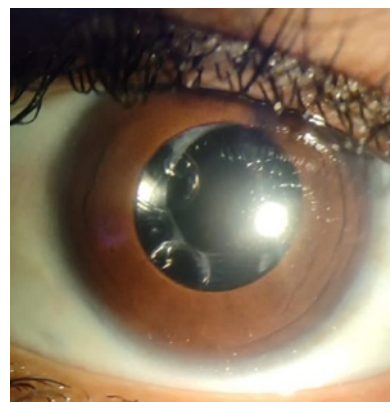


Fig. 5: Pseudophakia with PCIOL with Cionni capsular traction ring

ectopia lentis secondary to Marfan syndrome.

The patient was surgically managed with phacoemulsification with a Cionni capsular traction ring in both eyes. Postoperatively, the patient had unaided vision on 6/12 and aided vision on 6/9 (+0.75/-0.75X90°) in the right eye. On the left eye, a postoperative vision was 6/9.

3. Discussion

3.1. Ocular Manifestation of Marfan syndrome

Antoine- Bernard Marfan first narrated the Marfan Syndrome (MFS). It is a disorder of connective tissue that is caused by the mutation of fibrillin-1.⁷ The fibrillin-1 protein is a principal component of the elastic microfibril of ciliary zonules, which are responsible for ocular abnormal features like ectopia lentis, myopia, axial length, astigmatism, and flat cornea.^{8,9}

3.2. Ectopia lentis (EL) in MFS

Inherited Ectopia lentis is one of the main features of MFS, and accounts for 50%-80% of patients.¹⁰ Histologically

fibrillin is less distributed at the equator and zonular fibers in MFS patients. If visual acuity was reduced grossly and correction of the refractive status is often unstable, Extraction of the lens is required for ectopia lentis.^{11,12}

3.3. Surgical correction of EL in MFS

It is always crucial for phacoemulsification and IOL implantation for managing a patient with ectopia lentis, due to its zonular weakness and ectopic capsules which often couldn't possible for IOL implantation in the posterior chamber. Many surgeons prefer suture fixation in PCIOL procedure.

3.4. Modified capsular tension ring (MCTR)

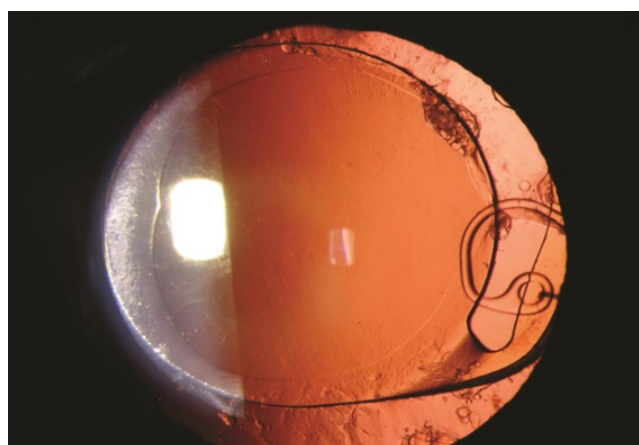


Fig. 6: Modified CTR with the addition of a fixation hook

Hara T et al. first reported the use of a Capsular Tension Ring for managing a case with mild zonular instability.¹³ Due to the decentration and imbalance of the capsular bag, CTR was reconstructed with an addition of a fixation hook (Figure 6) by Cionni and Osher.¹⁴

3.5. Safety and efficacy of modified Cionni tension ring in PCIOL surgery

It is not quite easy to perform IOL implantation in the posterior chamber of an eye with a subluxated lens. PCIOL implantation with sclera fixation or iris suture technique might cause a few complications, like retinal detachment and secondary glaucoma.¹⁵ Therefore, several surgeons prefer the introduction of a modified CTR technique for PCIOL implantation. Cionni and Osher described a modified CTR technique with a fixation hook, which did not distort the integrity of the capsular bag and obtained better zonular stability.¹⁴ The application of this modified CTR for in-the-bag IOL surgery is a relatively safe and practical choice for managing lens subluxation and zonular deficiency.^{16,17} Takimoto et al. compared the effect of CTR

with no-CTR for phacoemulsification surgery and noticed that CTR is the better choice for preventing the degree of IOL decentration and tilt, and regressed the rate of refractive prediction error.¹⁸

4. Conclusion

A modified CTR technique is the safe, and effective option for preventing reduced capsular bag shrinkage and maintaining the balance of the capsular bag of Ectopia lentis in patients with MFS.

5. Source of Funding

None.

6. Conflict of Interest


None.

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