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Journal of
Clinical & Medical Surgery

Case Report

Retro Pupillary Posterior Iris Fixation of the Iris-Claw Lens Implantation for the Management of *Ectopia Lentis* in Cohen Syndrome

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Article Info

Received: Jun 10, 2022
Accepted: Jul 11, 2022
Published: Jul 18, 2022
Archived: www.jclinmedsurgery.com

Abstract...

Cohen Syndrome (CS) is a rare genetic disorder. It is characterized by craniofacial malformations, hypotonia, obesity, non-progressive intellectual disability, retinal dystrophy. We present a case of CS in a 29-year-old female with subluxated Posterior Polar Cataracts (PPCs) in both eyes. Our objective is to achieve the best strategical surgery with the retropupillary fixation of iris-claw lens.

Keywords: Cohen syndrome; Posterior polar cataract; Iris-claw lens; Iris fixation of posterior chamber intraocular lens; Retropupillary fixation lens.

Introduction

Cohen Syndrome (CS) is an autosomal recessive genetic disorder caused by an altered gene located on chromosome 8. CS is characterized by intellectual disability, neutropenia and truncal obesity, hypotonia, microcephaly, abnormalities face, hands, and feet, and eye dysgenesis. These patients need careful ophthalmologic follow-up at all ages. Some of the causes of progressive vision deterioration include myopia, retinochoroidal dystrophy, and earlier lens opacities, interfering with daily activities [1].

We described a white female 29-year-old with Cohen Syndrome (CS) with a subluxated posterior polar cataract and zonula ciliaris was absent. The capsular support was also subluxated.

Biometry measurements (SRK/T formula, IOL MASTER V.4) for the right eye were: The ultrasonic axial length (ALu): 22.11 mm, the optical axial length (ALo): 22.41 mm, K1/K2: 45.42 /48.28D, Anterior Chamber Deph (ACD): 2.41 mm; for the left eye: ALu: 22.43 mm, ALo: 22.63 mm, K1/K2: 45.61/48.35D, ACD: 2.50 mm. Iris fixation of posterior chamber intraocular lens (PCIOL) was chosen (*Verisyse®*, VRSA54, PMMA, 5.4mm optic w/overall diameter of 8.5 mm, Abbott Medical Optics, Neth-

Citation: Guerriero S, Albano V, Alessio G. Retro Pupillary Posterior Iris Fixation of the Iris-Claw Lens Implantation for the Management of *Ectopia Lentis* in Cohen Syndrome. J Clin Med Surgery. 2022; 2(2): 1023.

erlands). Power of the implanted intraocular lens (IOL): +20,0 D right eye (ELP=115.0); +18,5 D left eye (ELP= 115.0). Postoperative VA increased in both eyes (from 20/200 at baseline to 20/32, Snellen) and it remained stable for the 1-year follow-up.

Surgical technique

The surgical procedure was bilateral. The sight defect was corrected in both eyes during a single operating session by the same surgeon. The surgery has been performed under general anesthesia. After a superior corneal tunnel was made, phacoemulsification was performed on the previous insertion of iris retractors. Two limbal scleral incisions were made, at 3 o'clock and 9 o'clock. We introduced viscoelastic fluid into the anterior chamber. Then we implanted the *Verisyse® IOL* from the superior incision. One haptic is guided below the iris by an IOL fixation hook and enclaved for half part of the iris. The same procedure has been repeated for the other haptic. Peripheral iridectomy was not performed in any case. Once the retropupillary iris lens was placed, the posterior capsule was removed using forceps. The two scleral incisions were sutureless. Finally, a criss-cross tunnel suture was performed.

Discussion

In an epidemiological study reported [2] a review of systemic disorders associated to lens opacification. Patients with neurologic disorders might present decreased vision and cataract. Mc Kibbin *et al* [3]. Described an early mortality of some of these before undergoing cataract surgery. As Summanen P *et al* [4]. Explained in 2002 about Finnish patients relatively to the emmetropic eye of a young adult with CS, they have multiple ocular dysgenesis which may affect cornea, ciliary body, and frequently showed spherophakia. These features determine that myopia is a predominant refractive type in CS, to be attributed to high corneal and lenticular power rather than an axial length.

Retropupillary iris-fixation intraocular lens is a new take on an old technique. As we know, the concept of iris suture fixation for posterior chamber intraocular lenses (PCIOL) dates back to 1976 and it was an surgical choice for aphakia [5]. This technique may be preferable to anterior chamber implantation or scleral fixation of a posterior chamber IOL in cases of absence of the zonular or capsular support [6]. Furthermore, iris-fixated IOL prevents later subluxation or dislocation [7]. Jare *et al* [8]. Studied 108 aphakic eyes and later functional and morphological outcomes after PCIOL implantation. Chronic inflammation, poor lens stability, significant central endothelial or presence of Central Macular Edema (CME) not observed during the 6-month follow-up period.

Moreover, several studies described an increased risk of posterior capsule opacification (PCO) in young patients. Shetai *et al* [9]. capsule posterior removal is a very advantageous procedure that prevents PCO complication.

Value statement

As we know, retropupillary iris-claw implantation is considered a good choice for surgical management in all cases where zonula ciliaris or posterior capsules were absent. This practice seems to be a low profile risk procedure and allows a less rate of complications.

To our knowledge, implantation of the iris-claw lens in Cohen Syndrome has not been studied yet. This paper adds that this surgical technique appears to be a safe and effective approach when treating Cohen Syndrome patients with *ectopia lentis* and subluxated capsules. Endothelium integrity was preserved. A peripheral iridectomy was not required. It was a repeatable and quicksurgical tecnique.

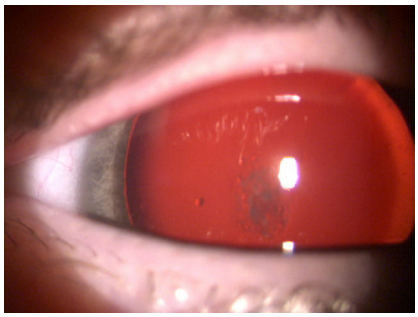

Figure 1: The subluxated posterior polar cataract.

Disclosure: None of the authors have received grant support or research funding and do not have any proprietary interests in the materials described in the article.

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