Rev Mex Oftalmol. 2015;89(4):237–240

Revista Mexicana de Oftalmología

www.elsevier.es/mexoftalmo

REVIEW ARTICLE

# What is the best surgical approach for ectopia lentis in Marfan syndrome?

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Received 13 March 2015; accepted 25 May 2015
Available online 31 August 2015

**KEYWORDS**
Marfan syndrome;
Ectopia lentis;
Surgical management;
Pars plana vitrectomy;
Intraocular lens

**Abstract** The surgical management of ectopia lentis (EL) in Marfan syndrome (MFS) represents a challenge to the ophthalmologist. We reviewed the literature on the surgical management of ectopia lentis in MFS patients from the classical pars plana lensectomy (PPL) to the most innovative scleral- and iris-fixated intraocular lens (IOL) surgical techniques. The results with the innovative approaches have been satisfactory but with a relatively short follow-up period and several complications associated, and the need of a highly experienced and skilled surgeon. We suggest that PPL approach with postsurgical aphakia is the safest surgical approach to ectopia lentis in MFS on a routinely basis.


**PALABRAS CLAVE**
Síndrome de Marfan;
Ectopia lentis;
Manejo quirúrgico;
Vitrectomia pars plana;
Lente intraocular

**¿Cual es el mejor manejo quirúrgico de ectopia lentis en síndrome de Marfan?**

**Resumen** El manejo quirúrgico de ectopia lentis en Síndrome de Marfan representa un reto para el oftalmólogo. Realizamos una revisión de la literatura sobre el tratamiento quirúrgico de ectopia lentis en pacientes con Síndrome de Marfan incluyendo el manejo clásico con lensectomía pars plana y afaquia postquirúrgica así como como el uso de lentes intraoculares fijados a iris o esclera. Los resultados con estos últimos han sido satisfactorios, pero con un periodo de seguimiento corto y diferentes complicaciones, además de que requieren de un cirujano con gran experiencia. Sugerimos que lensectomia pars plana es el abordaje quirúrgico más seguro de ectopia lentis en Síndrome de Marfan en la práctica diaria.


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http://dx.doi.org/10.1016/j.mexoft.2015.05.015

Marfan syndrome (MFS) is a connective tissue inherited disease associated with a decreased life expectancy.[1–4] It has and incidence of 2–3 per 10,000 individuals with no sex predilection.[1] It was first described by Antoine-Bernard Marfan in 1896.[1] It is caused by a mutation in *FBN1* (15q21.1) gene and is inherited in an autosomal dominant fashion.[2,4] This gene is involved in the production of the extracellular matrixprotein fibrillin, an essential glycoprotein for the formation of elastic fibers in connective tissue. It affects the ocular, skeletal and cardiovascular systems with great clinical variability.[5–7] Aortic dilatation and dissection are the most important and life threatening manifestations.[8,9] The diagnosis is based on clinical findings according to the revised Ghent nosology criteria that includes EL as one of its major criteria.[10,11] Ocular involvement in MFS is very common (>50%) and places a high burden on patients quality of life.[10–13] Although EL is the most common ocular manifestation, other ocular abnormalities can be found such as flat cornea, increased axial length (>3D), hypoplasia of the ciliary muscle or the iris, retinal detachment, cataracts, glaucoma, strabismus, and amblyopia.[14–20]

A stepwise approach is recommended for the management of EL. At first, when the visual axis is not compromised by the border of the dislocated lens causing diplopia or visual distortion, a conservatory management with optical refraction is preferable. However, surgical intervention is indicated when a functional best corrected visual acuity is not achieved, the refractive status is unstable because of lens mobility or posterior dislocation, or if anterior dislocation causes secondary ocular hypertension and risk of glaucomatous damage and/or risk of endothelial compromise.[20–22]

The surgical management of EL in MFS represents a challenge to the ophthalmologist. Fibrillin microfibrils are disrupted and fragmented in the lens capsule, iris and sclera, making the eye more susceptible to surgical complications.[23,24] For many years the preferable surgical approach to manage the dislocated lens in these patients has been standard lensectomy with or without anterior vitrectomy as well as pars plana vitrectomy (PPV) and lensectomy (PPL) with postoperative refractive correction including the use of aphakic glasses or contact lenses. With the advent of small-incision cataract surgery and better IOLs and capsular tension ring and segments, techniques and approaches for EL have evolved and various scleral- and iris-fixated surgical techniques have been proposed. However, the average patient follow-up in the majority of these published articles is 1 year and several complications using these proposed techniques have been found, including pupillary block, iris capture, lens decentration, and retinal detachment with fixation to the iris and/or sclera.[25–30]

We considered a classical surgical approach to manage ectopia lentis in MFS when a lensectomy is performed either through pars plana (PPV) or through a limbal approach with postoperative aphakia. Follow-up for these studies range from 1.5 to 102 months, with low incidence of complications.[31–39]

In the pars plana approach a standard vitrectomy technique is used. A 20G or 23G vitrectomy caliber can be used depending on surgeon's preference. The vitrector is used to engage the lens from a posterior approach removing the nuclear material utilizing the endoilluminator as a second instrument from the opposite port to stabilize the lens. In the limbal approach, as described by Plager[38] and Neely,[36] a peripheral corneal stab incision is made for an infusion cannula to maintain the anterior chamber throughout the procedure. A second anterior limbal incision is created with a MVR for the vitrector insertion. The MVR blade is advanced through the cornea to penetrate the peripheral anterior lens capsule creating 2–3 mm slit. The capsulorhexis can also be created using the vitrector which is then utilized to aspirate the lens followed by removal of most of the posterior and anterior capsule and a limited anterior vitrectomy.[40] These techniques minimize pulling forces and trauma to the zonules and iris and the vitreous base.

Capsular tension rings (CTRs) have provided the opportunity to perform small-incision phacoemulsification and in-the-bag implantation of a posterior chamber IOL (PCIOL). A capsular tension ring functions by exerting a centrifugal force at the capsular equator, expanding the capsular bag and redistributing tension from the weakened zonules to stronger, intact zonules.[40] However, the capsular tension ring cannot provide adequate support or correct decentration of the capsular bag in the presence of extensive zonular dialysis.[41] In 1998, the Cionni modified capsular tension ring (Morcher, FCI Ophthalmics, Marshfield Hills, MA, USA) was introduced to help manage profound zonular weakness.[42,43] The Cionni modified capsular tension ring can be fixated to the sclera without compromising capsular bag integrity with 1 or 2 sutures. In 2002, Ahmed designed the capsular tension segment (Morcher, FCI Ophthalmics, Marshfield Hills, MA, USA), of 120 degrees; it has an anteriorly positioned eyelet, which enables scleral suture fixation. Compared to the Cionni modified capsular tension ring, the capsular tension segment can be inserted into the capsule bag with greater ease and less trauma because a dialing technique is not necessary.[40] Bahar reported a series of intraoperative limitations difficulties using the Cionni ring during the creation of a central capsulorhexis in an unstable lens as well as during the implantation of the cionni ring with extensive subluxation, as it may be too large for the capsular bag, increasing the risk of a bag tear.[44]

Another option is an iris-fixated IOL. The iris-fixated IOL could be implanted in the posterior or the anterior chamber. Briefly, in the iris-fixated PCIOL a three-piece foldable PCIOLs is inserted with the haptics placed under the iris and the optic captured in the pupil. The haptics are sutured to the iris with a curved needle, as previously described.[28,45] For the iris-claw, the anterior chamber IOL (ACIOL) is introduced with the haptics at 3 and 9 o'clock centered on the pupil and an enclavation needle is used to fixate the IOL at the iris midperiphery.[28,46,47] The incidence of complications reported varies within the series. In a series of cases reported by Hirashima,[28] 33.3% of the patients treated with ACIOL developed iris atrophy at the enclavation site, and among the patients treated with PCIOL, 31.25% developed iris atrophy, 12.5% had a retinal detachment and 18.7% had IOL decentration. These complications were reported with a follow up of only 12 months. Also, the ACIOL may accelerate the endothelial cell count lose and potentially lead to bullous keratopathy.[48–50] Another concern when utilizing an ACIOL is the potential damage to the trabecular meshwork in an already predisposed glaucoma patient.[51,52]