Ferrara intracorneal ring implantation and cataract surgery for the correction of pellucid marginal corneal degeneration

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We report a case of Ferrara intracorneal ring (Mediphacos) implantation and cataract surgery for the correction of pellucid marginal corneal degeneration. Preoperatively, the patient’s uncorrected visual acuity (UCVA) was 0.05 in the right eye and 0.10 in the left eye. The best spectacle-corrected visual acuity (BSCVA) was 0.50 with −2.00 −11.25 × 80 in the right eye and 0.60 with −2.00 −5.50 × 95 in the left eye. One month postoperatively, the UCVA was 0.50 in the right eye and 0.30 in the left eye. The BSCVA was 0.80 with −1.00 in the right eye and 0.60 with −0.75 −1.25 × 160 in the left eye. Ferrara intrastromal rings and cataract surgery resulted in a more regular corneal shape with a reduction in astigmatism for good visual recovery.


P ellucid marginal corneal degeneration (PMCD) is a rare, peripheral corneal ectatic disorder. Characteristics of this disease include a 1.0 to 2.0 mm wide arcuate band of stromal thinning, 1.0 to 2.0 mm from the limbus. The area of thinning is usually inferior, extending from 4 o’clock to 8 o’clock, although thinning may exist at other locations. The adjacent clear cornea protrudes markedly and results in high against-the-rule astigmatism.

Patients often present in the third to fifth decades with reduced visual acuity from increasing astigmatism. The correction of irregular astigmatism remains a challenge for eye-care practitioners. Comfortable spectacle correction or stable contact lens correction is difficult to attain and because of the paucity of case series, there is no consensus on the best form of surgical management. Surgery is often a last resort and several procedures have been attempted, including crescentic wedge resection, crescentic lamellar keratoplasty, central penetrating keratoplasty (PKP), oversized central PKP, and inferiorly decentrated PKP. The use of many different procedures implies that the most appropriate surgical option has not been devised.

We report the use of intracorneal rings and cataract surgery to manage PMCD. There is 1 report of intracorneal rings (Intacs®, Addition Technology) to manage PMCD and no report of Ferrara intrastromal rings combined with cataract surgery to manage it.

Case Report

A 70-year-old man presented to the Refractive and Cornea Surgery Unit at Brasilia Ophthalmic Hospital. He had no systemic illness and no surgical history. The patient had a complete ophthalmic examination including visual acuity, intraocular pressure (IOP), and posterior segment evaluation. The uncorrected distance visual acuity was 0.05 in the right eye and 0.10 in the left eye. The best spectacle-corrected visual acuity was 0.50 in the right eye and 0.30 in the left eye. The BSCVA was 0.80 with −1.00 in the right eye and 0.60 with −0.75 −1.25 × 160 in the left eye. Ferrara intrastromal rings and cataract surgery resulted in a more regular corneal shape with a reduction in astigmatism for good visual recovery.


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visual acuity (BSCVA) was 0.50 with $-2.00 -11.25 \times 80$ in the right eye and 0.60 with $-2.00 -5.50 \times 95$ in the left eye.

The patient had experienced a subjective decrease in visual function in both eyes because of cataract formation. The preoperative central corneal pachymetry was 465 $\mu$m in the right eye and 460 $\mu$m in the left eye (Tomey SP-2000), and the corneal keratometry was 40.80@80/50.75@170 and 42.20@90/48.25@180, respectively. The corneal topography (EyeSys Technologies) of both eyes is shown in Figure 1. The IOP was 12 mm Hg, and funduscopy was normal. Scotopic pupil size measured with a Colvard pupillometer (Oasis) was 5.00 mm in both eyes. The corneal endothelial cell count (noncontact specular microscope, Noncon Robo, Konan) was 2596 cells/mm$^2$ in the right eye and 2681 cells/mm$^2$ in the left eye. Pellucid marginal degeneration and cataract formation were diagnosed, and the patient was scheduled for Ferrara intracorneal ring implantation and cataract surgery.

Ferrara intrastromal corneal rings are made of polymethyl methacrylate (PMMA), with an external diameter of 6.2 mm, a triangular section, and a 600 $\mu$m base. In each eye, 2 160-degree segments were used. Selection of the appropriate ring was based on the refractive error in each eye; 200 $\mu$m rings were used superiorly and 250 $\mu$m rings were placed inferiorly in both eyes, aiming for maximal flattening of the inferior cornea.

The procedure was performed under topical anesthesia. A circular Ferrara marker centered on the corneal area was used to create 2 concentric circles. The difference in the radii of the 2 circles was equal to the width of the Ferrara rings; thus, the area between them corresponded to the desired position of the rings’ insertion channels. Two radial corneal incisions were created in the temporal periphery of the cornea with a diamond knife set to 80% of the thinnest corneal measurement, which were performed in the temporal periphery of the cornea. Corneal thickness, measured intraoperatively at the incision site, was 499 $\mu$m and 486 $\mu$m in the right eye and left eye, respectively. A corneal spreader was used to facilitate insertion of the double metallic arcuate guide (Ferrara spatula), which elevated the cornea and simultaneously dissected 2 intrastromal channels around the central area. The 2 PMMA segments were implanted superiorly and inferiorly. The procedure was uneventful.

One month after insertion of the Ferrara rings, the BSCVA was 0.50 with $-2.50 -1.75 \times 90$ in the right eye and 0.40 with $-0.75 -2.25 \times 170$ in the left eye. Two months after Ferrara ring implantation, cataract surgery was performed in both eyes. Corneal topography of both eyes after Ferrara intracorneal ring implantation and just before cataract surgery is shown in Figure 2. The SRK/T formula was used to calculate an intraocular lens (IOL) power of 18.0 diopters (D) and 16.0 D in the right eye and left eye, respectively. The postoperative target was $-1.00$ D. The bioconvex IOL (MA60AC, Alcon Laboratories) had a 6.0 mm optic, a 13.0 diameter, and 10-degree haptic angulation.

Uneventful temporal clear corneal phacoemulsification was performed under topical anesthesia (Figure 3). One month postoperatively, the best corrected distance acuity was 0.80 with $-1.00$ in the right eye and 0.60 with $-0.75 -1.25 \times 160$ in the left eye. Keratometry readings in the right eye and left eye were 43.5@2/45.0@90 and 39.8@130/
Figure 3. (Akaishi) Phacoemulsification after intracorneal ring implantation. A and B: A temporal clear corneal incision is made. C: An ophthalmic viscosurgical device is inserted. D: Continuous curvilinear capsulorhexis is performed. E and F: Emulsification is performed, and the nucleus is aspirated. G and H: Intraocular lens implantation is performed, and the incision is allowed to self-seal.
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42.3±55, respectively. No refractive or surgical complication was seen.

**Discussion**

Intrastromal corneal rings were first used by Fleming et al.⁹ to correct low myopia. Although the effect and mechanisms by which intracorneal rings can flatten the cornea in eyes with low myopia are well documented and thought to be secondary to an arc-shortening effect of corneal lamellae,¹⁰,¹¹ the structural changes responsible for the effect of the Ferrara intrastromal rings on keratoconic and PMCD eye topography are not known.

The topography in PMCD is characterized by a steep contour in the peripheral inferior cornea with high keratometric powers radiating toward the center from the inferior oblique meridians. There is typically an area of vertical flattening down the center of the cornea. This pattern usually generates a refraction with high against-the-rule astigmatism in advanced cases.

The main objective of using the Ferrara intrastromal ring for PMCD is to reshape the abnormal cornea without removing corneal tissue or touching the central cornea, flattening the soft ectatic corneal tissue to decrease asymmetric astigmatism. In addition, Ferrara ring implantation preserves an intact globe and the procedure is potentially reversible.

Good candidates for Ferrara rings are patients who are contact lens intolerant and have a clear central cornea. The complication rate for intrastromal corneal rings is low. Undercorrection, overcorrection, migration of 1 segment toward the wound, epithelial ingrowth, infection immediately postoperatively or later from contact lens wear, extrusion due to superficial implantation or eye rubbing, and glare or halos can occur with intrastromal corneal rings.¹²–¹⁴

Although more patients and a longer follow-up are necessary to confirm the efficacy and stability of the effect of Ferrara intrastromal rings for the treatment of PMCD, the rings could represent a safe procedure for contact-lens-intolerant patients with clear central corneas.

In some cases, combined surgical options such as a Ferrara ring and cataract surgery can be used successfully.

**References**