CASE REPORT

A Special Design of Intacs SK and Collagen Corneal Cross-linking for the Treatment of Pellucid Marginal Degeneration in a 74-Year-Old Male

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ABSTRACT

We present a case report of a 74-year-old male who suffers from advanced and progressive pellucid marginal degeneration in his right eye, his left eye is legally blind because of macular scar.

We performed a combined procedure of a special design Intacs SK insertion followed by epi-off collagen corneal cross-linking according to the Dresden protocol. The Intacs SK were inserted by the manual technique, no intra- or postoperative complications were noted. Two months postoperatively, his uncorrected and best spectacle corrected visual acuity improved, the astigmatism was reduced and regularization of the corneal topography was observed. The patient is very satisfied from the improvement of his vision by minimally invasive procedures.

Keywords: Collagen, Corneal rings, Corneal segments, Crosslinking, ICRS, Intacs, Intrastromal, Keratoconus, Pellucid marginal degeneration, SK, Topography.

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INTRODUCTION

Pellucid marginal degeneration (PMD) is a progressive, non-inflammatory, peripheral corneal ectatic disorder characterized by a band of thinning ranging from 1.0 to 2.0 mm in width. The area of thinning is typically found in the inferior cornea, extending from the 4 to the 8 o'clock positions.¹ Patients usually present in their fourth to fifth decades of life with reduced visual acuity (VA) and against-the-rule astigmatism.² Visual activity cannot be improved by glasses to a satisfactory level, and rigid glass permeable (RGP) contact lenses are rarely tolerated in advanced cases due to inferior decentration of the contact lens caused by the 'beer belly' shape of the cornea.

A number of surgical procedures have been suggested to improve VA in PMD, including thermokeratoplasty, epikeratophakia, total lamellar keratoplasty, crescentic lamellar or full thickness resection, large eccentric penetrating keratoplasty, central penetrating keratoplasty, crescentic lamellar keratoplasty, and a combination of the last two procedures. All these procedures require extensive surgery with unpredictable visual results.³⁻⁸

Decentered grafts have been tried in an attempt to remove as much of the affected lower cornea as possible; however, the proximity of the graft to the limbus and its blood vessels increase the rejection rate, and the inferior decentration of the graft causes a large degree of astigmatism.⁹ These procedures are not as efficient as penetrating keratoplasty (PKP) or deep anterior lamellar keratoplasty (DALK) are for the treatment of keratoconus, which are considered the gold standard surgical treatments for it.

Due to the complexity and unsatisfactory results with the available treatments for PMD, the intracorneal ring segments (ICRS) have a great value in treating this progressive frustrating disease. As expected, Intacs improved uncorrected visual acuity (UCVA) and best corrected visual acuity (BCVA) in patients suffering from PMD.¹⁰ Occasionally implanting one Intacs segment can be sufficient, as in our published case report, in which we implanted only one segment in the upper part. We chose this alternative because (after marking and pachymetry) the inferior segment was supposed to pass in a very thin area of the cornea, good results were obtained in terms of UCVA and BCVA.¹¹ Mularoni et al reported 1 year data of eight eyes with PMD that were treated with asymmetrical Intacs insertion, using the mechanical method, six eyes (75%) had BCVA of 20/25; the mean postoperative spherical equivalent (SE) was 1.52 D (3.01 SD).¹²

Ertan and Bahadır treated nine eyes of patients suffering from PMD by Intacs that were implanted using the femtosecond laser. Uncorrected visual acuity improved in all eyes, the BCVA improved in eight eyes, and remained unchanged in one eye.¹³

In a retrospective, consecutive case series, Pinero et al reported refractive and corneal aberrometric changes after ICRS implantation in corneas with PMD. This study included 21 eyes of 15 patients with a diagnosis of PMD,

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whose ages ranged from 21 to 73 years and were treated in four ophthalmologic centers. Intacs were implanted in only three eyes, whereas Kerarings in 18 eyes. Postoperative visits were scheduled for the first postoperative day and for months 1, 3 and 6 postoperatively. Incision was located on the steepest meridian of the anterior corneal surface in all patients.¹⁴ The incision was made along the steep anterior corneal meridian. No complications occurred intraoperatively. Segment ring explantation was performed in a total of four eyes (19.0%), due to significant visual deterioration during the follow-up; all of the explanted ring segments were Kerarings. A statistically significant reduction was found in sphere and cylinder after ICRS implantation. On an average, manifest astigmatism was reduced with the implants by 50%. Also, mean corneal curvature was significantly reduced with the ICRS. The authors assume that the flattening effect was due to the significant reduction of curvature that occurred in the steepest meridian. This could be explained by the combination of two flattening factors, the insertion of the midperipheral implants and the weakening effect induced by the corneal incision. Significant reduction was observed in the sphere and in the infero-superior asymmetry, along with relative centration of the peripheral corneal protrusion. A significant change was

also found in corneal asphericity, with a trend toward oblateness. The study reported anterior corneal aberrometric outcomes after ICRS implantation; astigmatism, higher order, and coma-like aberrations were significantly reduced with surgery. All of these changes in corneal aberrations were consistent with the improvement in BCVA (55% of eyes gained lines of BCVA). The authors used asymmetric Intacs and implanted the thicker one inferiorly and the thinner one superiorly. No extrusion occurred in the Intacs group. The study does not deal with Intacs and Kerarings separately, rather provided global results; nevertheless, it confirms the beneficial effect of Intacs and Kerarings in treating PMD.

Intracorneal ring segments have an important role in treating PMD and may be used as an alternative to the other available previously mentioned surgical options.

CASE REPORT

A 74-year-old male was referred to our medical center because of deterioration of his VA.

He had a deterioration of the VA in his left eye (LE) because of age related macular degeneration since 2008. Figure 1 shows the optical coherence tomography (OCT) of both eyes; he underwent cataract extraction in his right eye (RE) in 2010.

Fig. 1: Optical coherence tomography of the macula in both eyes, showing normal macula in the RE and scar in the LE

Fig. 2: Tomographic modelling system-5 topography (Tomey Japan) of the RE showing PMD

Eye examination: Uncorrected visual acuity 3 meters finger counting in the RE and 2 meter finger counting in the LE, BCVA 20/60 in the RE and 3 meters finger counting in the LE.

The refraction: +2.0 diopters sphere (DS), -19.0 diopters cylinder (DC) $\times$ 90° in the RE and +6.0 DS, -12.0 DC $\times$ 90° in the LE.

In the RE: Clear cornea, pseudophakia in the posterior chamber, clear intact posterior capsule, normal optic nerve, few small hemorrhages in the mid periphery and normal macula. The cornea topography showed PMD in both eyes (Figs 2 to 4). The central corneal thickness as measured by ultrasound pachymetry was 573 μ m.

No former topographies were available.

Because of his general health and social conditions, contact lenses are not an option in this case.

Intracorneal ring segments implantation with collagen cross-linking (CXL) was discussed with the patient and his family and informed consent was signed by the patient.

On the 14th of April 2015, a pair of Intacs SK, 130° length, 450 μ m thick inferiorly and 400 μ m thick superiorly (a special design of Intacs SK produced for me by AJL, Spain) were implanted in his RE using the manual technique. The incision was made along the 180° meridian which is the steep axis. The depth of the incision was set at 400 μ m because the thinnest point through which the ICRS were supposed to pass was 480 μ m (as measured by ultrasound pachymetry after marking the incision and the ICRS positions). Immediately after the creation of the tunnels,

the epithelium was removed from the central 9 mm of the cornea, the Intacs SK were inserted and riboflavin 0.1% with dextran was instilled every 5 minutes for half an hour until a strong yellow flare was observed in the anterior chamber. The eye was then exposed to ultraviolet A (UVA) 365 $\text{\AA}$, 15 mW for 5 minutes, a contact lens was applied and moxifloxacin 0.5% (Vigamox) eye drops were prescribed every 2 hours. Three days later, the contact lens was removed and antibiotics reduced to six times a day for 2 weeks and stopped; no intra- or postoperative complications were noted.

Two months after the combined Intacs SK and CXL the RE was quite and the patient very satisfied from his VA; UCVA in the RE 20/50, with +3.0 DS and -3.0 DC $\times$ 90, his vision improves to 20/40 partial. The cornea is clear, the Intacs SK are in the programmed position, the corneal topographies shows flattening and regularization of the cornea (Figs 5 to 8), the astigmatism was reduced dramatically from 18 to 3 DC, the UCVA improved from 3 meters finger counting to 20/15 and the BCVA improved from 20/60 to 20/40 partial.

DISCUSSION

Intracorneal ring segments proved to be useful in improving UCVA and BCVA in patients suffering from PMD as mentioned before. Collagen cross-linking proved to be efficient in arresting the progression of keratoconus and ectatic corneal diseases.¹⁵⁻¹⁷

Fig. 3: Tomographic modelling system-5 topography (Tomey Japan) of the LE showing PMD

Fig. 4: Tomographic modelling system-4 topography (Tomey Japan) of the RE showing PMD

Fig. 5: Tomographic modelling system-5 topography (Tomey Japan), postoperative showing the flattening and the regularization of the cornea in the RE

Fig. 6: Tomographic modelling system-4 topography (Tomey Japan), postoperative showing the flattening and the regularization of the cornea in the RE, on the left upper side showing the ICRS

Additive effect of the two procedures was reported, the mechanism of the additive effect is thought to be due to flattening of the cornea by CXL in addition to the flattening induced by the ICRS, biomechanical coupling due to local collagen changes around the ICRS and pooling of the riboflavin into the tunnels which may increase the cross-linking in that area.¹⁸⁻²²

What is Special in this Case?

- The age of the patient (74 years), and the fact that his vision is deteriorating with no clear cause for the deterioration except the PMD, the patient is pseudophakic, has no secondary cataract and normal macula as seen biomicroscopically and by OCT.

Fig. 7: Tomographic modelling system-4 topography (Tomey Japan), postoperative map, showing the flattening and regularization of the cornea in the RE in the upper right side; in the lower right side showing the preoperative topography, on the left side shows the difference map

Fig. 8: Colored photograph of the Intacs SK in the cornea

- The only eye that can give him a functional VA is his RE, his LE is legally blind because of a macular scar.
- The general health and the social conditions of the patient do not allow the use of contact lenses.

So, surgery had to be done and we chose non-invasive means to treat the patient.

A special design of Intacs SK, which is similar to the original design produced by AJL Spain (oval shape, 6 mm optical zone, 160° long, 400–450 µ thick), was used. We chose to perform CXL because we were convinced and so was his referring ophthalmologist that the deterioration of vision is the result of the PMD. Although we could not obtain serial corneal topographies to prove our claim, we decided to perform epi-off CXL according to the Dresden

protocol in order to try to arrest the progression of the PMD and to obtain additive effect of ICRS and CXL.

CONCLUSION

Intracorneal ring segments and CXL were effective in improving UCVA and BCVA in a 74-year-old male suffering from PMD; legally blind in his other eye and contact lens 'intolerant', the combined non-invasive treatment spared the patient invasive surgeries.

These two procedures should be offered to patients suffering from progressive PMD before other invasive surgeries.

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