

Visual Outcome after Implantation of Intracorneal Ring Segments for Pellucid Marginal Degeneration

Mahmood Jabbarvand, MD^{1,2} • Kazem Amanzadeh, MD^{1,2} • Firoozeh Rahimi, MD³
Ahmad Kheirkhah, MD⁴ • Hassan Hashemi, MD^{1,2}

Abstract

Purpose: To assess the results of intracorneal ring (ICR) segments for treatment of patients with pellucid marginal degeneration (PMD)

Methods: In this prospective interventional case series study, eight eyes of 5 patients with PMD who were contact lens intolerant received one or two Intacs (Intacs, Addition Technology) segments. Preoperative and postoperative evaluations included slit-lamp examination, uncorrected visual acuity (UCVA) and best corrected visual acuity (BCVA) and keratometry by Pentacam scheimpflug camera (oculus opticerate GmbH). Patients were followed for 3 months and all parameters were reviewed at the end of follow-up time.

Results: Three months postoperatively, UCVA significantly improved from preoperative values after Intacs implantation (mean 1.07 ± 0.27 [SD] and 0.5 ± 0.27 respectively). The BCVA also improved from preoperative state compared to 3 months after surgery (Mean 0.40 ± 0.31 and 0.18 ± 0.17 respectively). The BCVA was $\frac{5}{10}$ or more in all 8 eyes. Spherical equivalent (SE) and refractive cylinder decreased significantly compared to preoperative values (Mean -9.40 ± 4.98 to -6.53 ± 5.00 and mean -6.93 ± 3.79 to -3.06 ± 2.07 respectively). Topographic cylinder decreased postoperatively, but this decrement was not statistically significant (mean -5.42 ± 3.47 [SD] to -5.08 ± 3.20).

Conclusion: In our series of patients, ICR implantation proved to be a safe and effective method for correction of PMD and improving the vision in these patients.

Keywords: Pellucid Marginal Degeneration, Intracorneal Ring, Visual Acuity

Iranian Journal of Ophthalmology 2010;22(1):41-45 © 2010 by the Iranian Society of Ophthalmology

-
1. Associate Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences
 2. Noor Ophthalmology Research Center, Noor Eye Hospital
 3. Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences
 4. Assistant Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences

Received: July 15, 2009

Accepted: October 8, 2009

Correspondence to: Hassan Hashemi, MD

Eye Research Center, Farabi Eye Hospital, Tehran, Iran, Tel: +98 21 55414941-6, Email: hhashemi@noorvision.com

Noor Ophthalmology Research Center, Noor Eye Hospital, Tehran, Iran, Tel: +98 21 88651515

Introduction

Pellucid marginal degeneration (PMD) is an uncommon, bilateral, progressive, noninflammatory corneal condition. PMD is characterized by a peripheral band of inferior corneal thinning, extending from 4 to 8 o'clock positions with 1 to 2 mm of normal cornea between the thinning and the limbus.¹ In this disorder the area of thinning is typically epithelialized, clear, avascular, and without lipid deposits.²

The etiology of PMD is not clear; although in most cases it is not hereditary, there are reports regarding the involvement of other family members with keratoconus and PMD. It is not clear whether PMD, keratoconus, and keratoglobus are distinct diseases or phenotypic variations of a single disorder.³

The topographic appearances in PMD show a classical "butterfly" shape, or "claw pattern", which demonstrates low corneal power along the vertical axis, an increased power at the inferior peripheral cornea, and high corneal power in the mid-peripheral cornea along the inferior oblique meridians. This pattern usually produces large amounts of with-the-rule irregular astigmatism in advanced cases.⁴⁻⁶ In atypical cases, topographical patterns are similar to classic types except for the involvement of nasal, temporal or superior quadrants, or a combination of these.⁷

PMD manifests in the second to fourth decade of life with visual loss due to severe irregular astigmatism. In early stages, it can be managed with spectacles or rigid contact lenses, however, in severe cases, or in those intolerant to contact lenses surgical intervention is indicated.

Several surgical techniques to treat PMD have been tried. These include thermokeratoplasty, epikeratophakia crescentic lamellar or full thickness resection.

Intracorneal ring (ICR) segments (Intacs, Addition technology, Inc, Fremont, CA) were designed to achieve refractive adjustment by decreasing irregular astigmatism, displacement of the cone to central cornea and overall flattening of the central corneal curvature while central optical zone remains clear.

Several studies show the safety and efficacy of Intacs in correcting ectatic corneal disorder like keratoconus and PMD. In this

study, we report the outcomes of Intacs implantation in 8 eyes with PMD.

Methods

This study included 8 eyes of 5 consecutive patients, which all were males, with a mean age of 32.4±7.6 years (range, 26-41 years) who presented to Farabi Eye Hospital, Tehran, Iran. All these eyes had PMD with clear central cornea, contact lens intolerance and an inferior corneal thickness of more than 400 µm at the location where Intacs inserts were to be implanted. Patients with ocular condition like recurrent epithelial erosion and corneal dystrophy or collagen vascular diseases and pregnant or nursing woman were excluded from the study.

The diagnosis of PMD was made on the basis of slit-lamp findings of inferior corneal thinning band and ectasia above the area of maximum thinning. The diagnosis was confirmed by corneal topography which show typical claw pattern and against-the-rule astigmatism.

Preoperatively, all patients underwent a full ocular examination, including medical history, uncorrected and best spectacle-corrected visual acuity (UCVA and BCVA, respectively), manifest and cycloplegic refraction, applanation tonometry, slit-lamp microscopy, fundus examination, and corneal topography with the Pentacam (Oculus Opticgerate GmbH) including the 4-map and full pachymetry map. All patients had clear corneas and a minimum inferior corneal thickness of 400 µm at 7 mm. After obtaining informed consent, the patients underwent Intacs (Addition Technology, Inc, Fremont, CA) by a single surgeon. Follow-up visits were scheduled for 1 week, 1 month, and 3 months after surgery.

The surgical procedure was performed under local anesthesia with peribulbar injection of lidocaine. Based on the refraction, and the location and extension of corneal protrusion in respect to the horizontal meridians, 0.45 mm thickness Intacs (Addition Technology, Inc, Fremont, CA) were implanted. In 6 eyes, only one piece was placed in the inferior cornea, one eye received 2 pieces vertically, and the remaining eye was corrected with one 0.45 mm Intacs in the inferior cornea and one 150 µm Ferrara ring

(Ferrara Ophthalmics, Brazil) in the superior cornea (Table 1).

Table 1. Preoperative and 3 months postoperative data

No.	Age/Gender/Eye	UCVA		BCVA		Refractive cylinder		Topographic cylinder		SE		Ring no/Site
		Preop	Postop	Preop	Postop	Preop	Postop	Preop	Postop	Preop	Postop	
1	35/M/R	.08	10	.50	.70	3.25	1.50	5.40	7.90	8.75	6.75	1/inf
	35/M/L	.20	.30	.60	.70	2.00	1.50	1.40	1.30	9.00	4.75	1/inf
2	41/M/R	.04	.10	.10	.60	5.50	4.50	6.70	5.30	19.25	16.25	1/inf
	41/M/L	.05	.40	.20	.60	11.5	6.00	8.70	4.60	11.75	10.50	1/inf
3	26/M/L	.20	.30	.30	.50	2.00	2.00	3.12	2.70	7.50	2.50	2/vertical
4	37/M/R	.05	.50	.60	.60	12.00	4.50	9.60	3.90	2.00	1.25	1 inf/1 sup*
	37/M/L	.08	.40	.80	.90	8.00	3.00	4.90	3.30	2.00	1.00	1/inf
5	23/M/L	.10	.30	.60	.60	10.00	6.00	11.30	9.70	1.25	.00	1/inf

*: Ferrara ring inserted superiorly

UCVA: Uncorrected visual acuity, BCVA: Best corrected visual acuity, SE: Spherical equivalent, M: Male, R: Right, L: Left, inf: Inferior, sup: Superior. SE, refractive cylinder, and topographical cylinder are in minus

Based on the location of the rings a small corneal incision (1.8 mm in length) at 70% of corneal thickness was made at the edge of the 7-mm optical zone. Intrastromal tunnels (clockwise and counterclockwise) were created with special care in the inferior cornea which was relatively thinner. For insertion of the Ferrara ring, the tunnel was created manually at the 5-mm optical zone. After ring insertion, the incision was closed with a single 10-0 nylon suture.

After surgery, patients were instructed to use betamethasone drop (Sina Darou) 6 times daily for 2 weeks, chloramphenicol drop (Sina Darou) 4 times daily for 5 days, and nonpreserved artificial tear (Artelac) (Baush & Lomb, France) for 3 months. The sutures were removed in all patients 2 weeks after surgery.

Postoperative examinations included UCVA and BCVA tests at 1 week, and 1 and 3 months. Manifest and cycloplegic refractions, and Pentacam topography (oculus opticegrate GmbH) were performed every month postoperatively until 3 months.

For statistical analysis, preoperative and 3-month postoperative values of UCVA, BCVA, spherical equivalent (SE), and

refractive and topographical cylinder were compared using the paired *t* test.

Results

ICR segments were successfully implanted in all eyes without any intraoperative complication. Table 1 shows the preoperative and postoperative UCVA, BCVA, refractive cylinder, topographical cylinder, and SE data. Three months after surgery, UCVA improved in all eyes (100%), while BCVA improved in 6 eyes, and remained unchanged in 2 eyes.

Table 2 shows the mean preoperative and 3-month postoperative UCVA and BCVA, spherical and cylindrical refractions. Postoperatively, the improvements in UCVA and BCVA from preoperative values were significant (mean=1.07±0.27 to 0.57±0.27 and mean 0.40±0.31 to 0.18±0.17, respectively). The decreases in SE and refractive cylinder were significant (mean=-9.40±4.98 to -6.53±5.00 and mean=-6.93±3.79 to -3.06±2.07, respectively). The decrease in topographical cylinder from preoperatively to 3 months postoperatively was not statistically significant.

Table 3 shows the preoperative and postoperative manifest refractions in all 8 eyes.

Table 2. Analysis of preoperative and postoperative patients' data

Variable	Preoperatively	Postoperatively
UCVA (logMAR)	1.07±0.27	0.57±0.27
BCVA (logMAR)	0.40±0.31	0.18±0.17
Refractive cylinder (D)	-6.93±3.79	-3.06±2.07
Topographic cylinder (D)	-5.42±3.47	-5.08±3.20
Spherical equivalent (D)	-9.40±4.98	-6.53±5.00

UCVA: Uncorrected visual acuity
BCVA: Best corrected visual acuity
D: Diopter

Table 3. Preoperative and postoperative refraction of patients

No.	Eye	Preoperative	3 months postoperative
1	OD	-7.25- 3.25×160	-6.00- 1.50×165
	OS	-8.00-2.00×160	-4.00-1.50×90
2	OD	-16.5-5.5×63	-14.00-4.50×55
	OS	-6.00-11.5×95	-7.50-6.00×90
3	OD	-6.5-2.00×130	-1.5-2.00×100
4	OD	+4.00-12.00×95	+1.00-4.5×70
	OS	+2.00-8.00×110	+0.50-3.00×100
5	OD	+4.00-10.5×90	+3.00-6.00×95

Discussion

PMD is a challenging disease for surgeons, as well as contact lens intolerant patients. Several surgical options have been proposed. Thermokeratoplasty and epikeratoplasty were the first procedures suggested by Zuchini¹⁰ and Fronterre et al¹¹ respectively, with subsequent improvement of visual function. These techniques are now mostly of historical interest as their results are poor.

Large eccentric penetrating keratoplasty extending near to the limbus has been used to treat PMD; however, it was not popularly accepted because of increased risk of graft rejection.¹² Lamellar crescentic keratoplasty first described by Schanzlin et al¹³ is another surgical option for correcting PMD. In a study by Javadi et al¹⁴ on 15 eyes of 9 patients, lamellar crescentic resection was found to be a safe and effective method with relatively prolonged visual recovery time.

Full thickness crescentic wedge resection and a combination of lamellar crescentic keratoplasty and penetrating keratoplasty suggested by Rasheed et al¹⁵ are 2 procedures with limited success in the treatment of PMD. All of these techniques have several disadvantages, including unpredictability, irreversibility, long rehabilitation time, and significant complication rates.

The goal of using ICR in PMD is to reshape the cornea without removing any tissue; this is done by lifting the ectatic portion of the cornea, tissue flattening, and decreasing the asymmetrical astigmatism. Mularoni et al² showed improved UCVA in all eyes (100%), a BCVA of $\frac{20}{25}$ in 75%, and a decrease in SE and cylinder error. Ertan¹⁶ reported 9 eyes that underwent ICR implantation by application of femtosecond laser. In her study, there was an improvement of 3.5 ± 1.6 lines in UCVA and a significant decrease in cylinder values (-2.4 D preoperatively to -0.94 D postoperatively).

In our study on 8 eyes of 5 patients, 3 months after surgery UCVA increased in all patients; mean UCVA improved from 1.07 ± 0.27 logMAR preoperatively to 0.57 ± 0.27 logMAR postoperatively. BCVA improved in 6 eyes and remained unchanged in 2 eyes. All patients had postoperative BCVA of $\frac{5}{10}$ or more, and mean BCVA improved from 0.40 ± 0.31 to 0.18 ± 0.17 in the logMAR scale. Mean refractive cylinder changes were significant (-6.93 ± 3.79 D preoperatively and -3.06 ± 2.07 D postoperatively, however topographical cylinder did not change significantly. The above results of cylinder changes demonstrated the fact that there is not a linear correlation between refractive and topographic cylinder. This may be the consequence of the biomechanic changes of cornea in the ectatic diseases. More studies should be conducted to reveal this correlation. SE decreased from -9.40 ± 4.98 D to -6.53 ± 5 D.

Conclusion

Finally, findings of our study, further confirm the results of previous reports of using ICR in PMD, however, assessment of more cases with longer follow-ups are recommended to determine the effect of ICR implantation on the course of the disease.

References

1. Rabinowitz YS. Keratoconus. *Surv Ophthalmol* 1998;42(4):297-319.
2. Mularoni A, Torreggiani A, di Biase A, et al. Conservative treatment of early and moderate pellucid marginal degeneration: a new refractive approach with intracorneal rings. *Ophthalmology* 2005;112(4):660-6.
3. Santo RM, Bechara SJ, Kara-José N. Corneal topography in asymptomatic family members of a patient with pellucid marginal degeneration. *Am J Ophthalmol* 1999;127(2):205-7.
4. Maguire LJ, Klyce SD, McDonald ME, Kaufman HE. Corneal topography of pellucid marginal degeneration. *Ophthalmology* 1987;94(5):519-24.
5. Rowsey JJ, Reynolds AE, Brown R. Corneal topography. Corneoscope. *Arch Ophthalmol* 1981;99(6):1093-100.
6. Maeda N, Klyce SD, Tano Y. Detection and classification of mild irregular astigmatism in patients with good visual acuity. *Surv Ophthalmol* 1998;43(1):53-8.
7. Rao SK, Fogla R, Padmanabhan P, Sitalakshmi G. Corneal topography in atypical pellucid marginal degeneration. *Cornea* 1999;18(3):265-72.
8. Barbara A, Shehadeh-Masha'our R, Zvi F, Garzozzi HJ. Management of pellucid marginal degeneration with intracorneal ring segments. *J Refract Surg* 2005;21(3):296-8.
9. Rodriguez-Prats J, Galal A, Garcia-Lledo M, et al. Intracorneal rings for the correction of pellucid marginal degeneration. *J Cataract Refract Surg* 2003;29(7):1421-4.
10. Zucchini G. Su di un caso di degenerazione marginale della cornea-studio clinico ed istologico. *Ann Ottalmol Clin Oculist* 1962;88:47-56.
11. Fronterre A, Portesani GP. Epikeratoplasty for pellucid marginal corneal degeneration. *Cornea* 1991;10(5):450-3.
12. Varley GA, Macsai MS, Krachmer JH. The results of penetrating keratoplasty for pellucid marginal corneal degeneration. *Am J Ophthalmol* 1990;110(2):149-52.
13. Schanzlin DJ, Sarno EM, Robin JB. Crescentic lamellar keratoplasty for pellucid marginal degeneration. *Am J Ophthalmol* 1983;96(2):253-4.
14. Javadi MA, Karimian F, Hosseinzadeh A, et al. Lamellar crescentic resection for pellucid marginal corneal degeneration. *J Refract Surg* 2004; 20(2):162-5.
15. Rasheed K, Rabinowitz YS. Surgical treatment of advanced pellucid marginal degeneration. *Ophthalmology* 2000;107(10):1836-40.
16. Ertan A, Bahadir M. Intrastromal ring segment insertion using a femtosecond laser to correct pellucid marginal corneal degeneration. *J Cataract Refract Surg* 2006;32(10):1710-6.