Combined Wedge Resection and Beveled Penetrating Relaxing Incisions for the Treatment of Pellucid Marginal Corneal Degeneration

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Purpose: To evaluate a new surgical approach for the treatment of pellucid marginal corneal degeneration (PMCD).

Methods: Prospective, noncomparative, interventional case series. Ten eyes of 10 consecutive patients were included in the series. Inclusion criteria were a clinical diagnosis of PMCD, topographic astigmatism >10 D, and contact lens intolerance. The patients underwent an inferior arcuate wedge resection of the thinned cornea (0.5 mm wide, 6.0 mm long) combined with 2 penetrating, sutureless, clear corneal tunnel relaxing incisions (3.2 mm wide) at the steep meridians.

Results: Visual acuity, refraction, and corneal topography were assessed preoperatively and at 1, 3, 6, 12, and 24 months postoperatively. Suture removal was complete in all patients by 4 months. Spectacle-corrected visual acuity was 20/40 or better in 8 of 10 patients at 6 months, with no substantial change at later examinations. Mean keratometric astigmatism was reduced from 15.1 D preoperatively to 4.6 D at 6 months postoperatively and was stable at further follow-ups.

Conclusions: Corneal wedge resection combined with paired, opposed clear corneal penetrating relaxing incisions is a suitable surgical option for the treatment of PMCD, providing early adequate astigmatic control with long-term stability.

Key Words: pellucid marginal corneal degeneration, wedge resection, relaxing incisions

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Pellucid marginal corneal degeneration (PMCD) is a rare, progressive, idiopathic ectatic condition of the cornea. It is characterized by a crescent-shaped, noninflammatory, nonulcerative thinned area of the inferior peripheral cornea, although up to 15% may occur superiorly.[1] It is often misdiagnosed as keratoconus, although it may also coexist with it. Vision is affected by high, often irregular, against-the-rule astigmatism,[2] but potentially also by episodes of acute hydrops and perforation.[3]

Treatment of PMCD is initially conservative, with spectacles and contact lenses maintaining satisfactory functional levels of vision in 55%–67% of patients.[4,5] When these methods fail, surgical options ranging from thermokeratoplasty[3,5] to crescentic lamellar keratoplasty, possibly combined with penetrating keratoplasty,[6–8] have been suggested. None of these techniques reliably achieve early visual rehabilitation with long-term stability while avoiding the risks of penetrating allograft procedures. Specifically, penetrating grafts carry the long-term risks of endothelial rejection,[8] glaucoma, and astigmatism relating to graft decentration. These risks, together with the facts that the endothelium is unaffected and central corneal thickness in PMCD is typically within the normal range,[9] make the use of these procedures conceptually inappropriate.

Attempts at surgically correcting the astigmatism while conserving unaffected corneal tissue have led to the development of the wedge resection procedure.[10] However, this is associated with long-term astigmatic drift,[11] probably as a result of unstable, persistent tension at the sutured wound. MacLean et al[11] suggested that a more stable long-term refraction might be achieved by modifying the wedge resection technique.

Many studies have been published to assess the mechanism and effectiveness of a peripheral clear corneal relaxing incision in reducing astigmatism.[12,13] An incision depth of 90%–100% provides a maximal astigmatic effect and that 2 opposite, long incisions have a more regular effect on the topography.[13] Determination of the corneal relaxing incision (CRI) axis from preoperative topography is thought to aid the refractive outcome.[14]

Thus, 2 beveled clear corneal penetrating incisions, positioned at the steep axes, may be the most effective and reliable augmentation procedure for wedge resections performed in patients with PMCD. We propose that the combined procedure of beveled CRIs and wedge resection will release the tension placed on the latter, thereby optimizing refractive results and long-term stability of surgical treatment of PMCD.

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MATERIALS AND METHODS

Patient selection was on the basis of the criteria of a clinical diagnosis of PMCD (peripheral thinning of the inferior cornea with typical "knee" at a thin slit examination), a topographic against-the-rule astigmatism of ≥10 D, and intolerance to contact lenses. Ten eyes of 10 patients who consecutively presented to Villa Serena Hospital, a tertiary referral center in Forlí, Italy, during the period January 2002 to December 2003, were enrolled. All patients provided informed consent for the procedures performed. All surgical procedures (described in detail below) were performed by 1 surgeon (M.B.), and the assessments were carried out by 1 of 3 investigators (M.B., V.S., S.M.).

Data extraction was performed by patient chart review, and analysis was performed and graphed by using a standard spreadsheet program. Snellen best spectacle-corrected visual acuity (BSCVA), manifest refraction, and corneal topography (EyeSys 2000; EyeSys Technologies, Houston, TX) were documented preoperatively and postoperatively at 1, 3, 6, and 12 months. All patients attended all follow-up visits, and 24-month data are also presented for 4 of the 10 patients.

Topographic astigmatism values used are the simulated keratometric values obtained from topography. Astigmatism analysis was performed as discussed by Morlet et al[15] by vector trigonometry into with-the-rule and against-the-rule components. The 2-tailed paired *t* test was used to determine astigmatic stability (values of $P \leq 0.05$ were considered statistically significant).

All procedures were performed under peribulbar anesthesia, according to the scheme in Figure 1. A 15° metal blade was used to delineate the arcuate peripheral border of the wedge resection (immediately before the beginning of the area of normal thickness), and the incision was deepened to at least 80% of corneal thickness through its length (Fig. 2A). The central border of the wedge was similarly prepared, aiming for resection of a full-thickness piece of cornea 0.5 mm wide and 6 mm long (Fig. 2B). Although in some cases the actual area of ectasia may have been >0.5 mm, suturing of larger resections would have needed much higher tension and caused a too pronounced corneal distortion.

After resection of the tissue wedge (Fig. 2C), the anterior chamber was filled with viscoelastic substance, and a 3.2-mm metal keratome was used to create 2 beveled, penetrating clear-corneal keratotomies, 1 at each of the topographic steep axes, immediately anterior to the limbus (Fig. 2D, E). The wound resulting from the wedge resection was sutured with 1 continuous running 10-0 nylon suture (Fig. 2F). Postoperatively, gentamicin and dexamethasone eyedrops were administered every 2 hours for the first week and tapered off over a 6-week period.

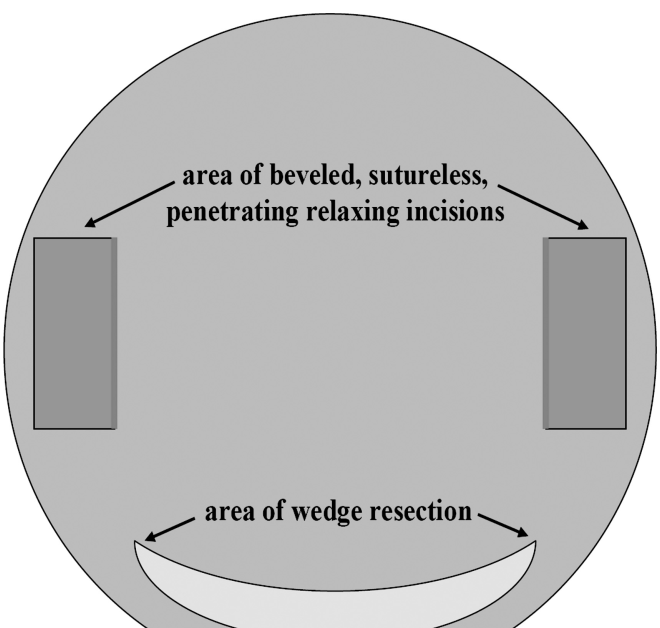

FIGURE 1. Diagrammatic representation of the surgical procedure, showing the inferior crescent-shaped wedge excision and 2 opposed clear corneal tunnels anterior to the limbus.

RESULTS

The patient group consisted of 7 men and 3 women. The average age of the patients was 57.7 years (range, 34–72 years). Mean preoperative topographic astigmatism was 15.1 ± 4.59 (SD) D (range, 10.4–26.6 D). Surgery was uneventful in all patients. The mean follow-up was 20.4 months (range, 18–24 months). No cheese-wiring or vascularization of the wedge sutures was observed. No patients returned to the operating theatre for resuturing or repeat astigmatic procedures. On the basis of the amount of overcorrection achieved, suture removal was completed between 2 and 4 months postoperatively in all patients. Slit-lamp examination (Fig. 3) showed the postoperative absence of an area of peripheral inferior thinning (Fig. 3B), clearly visible before surgery (Fig. 3A).

Figure 4 summarizes the improvement of BSCVA over time. At 6 months, BSCVA was ≥20/40 in 8 of 10 cases, and the mean topographic cylinder was 4.6 D. No substantial difference was recorded at the 12- and 24-month examinations. In the only patient with a persistent vision of 20/200, a macular scar was present.

The amplitude of the refractive astigmatism is shown in Figure 5 and can be seen to parallel the improvement in visual acuity.

Figures 6–8 show the pattern of progression of the astigmatism magnitude over time and of its vector components. The desired initial overcorrection, as exemplified by the topographic maps A and B in Figure 9 can be seen as a with-the-rule peak in Figure 7. This component decreases with the removal of sutures in some of the patients by the third postoperative month. As shown by map C in Figure 9, by 6 months, there is partial return of an against-the-rule component, consistent with removal of sutures in all patients. With respect to long-term stability, the mean magnitudes of astigmatism at 6, 12, and 24 months were 4.56 ± 2.39, 4.89 ± 2.37 (n = 10), and 5.08 ± 2.72 D (n = 4). Between 6 and 12 months, the average increase in the with-the-rule component was 0.13 D and that of the against-the-rule components was 0.31 D. Between 6 and 24 months, the average with-the-rule component increase was 0.55 D and the against-the-rule

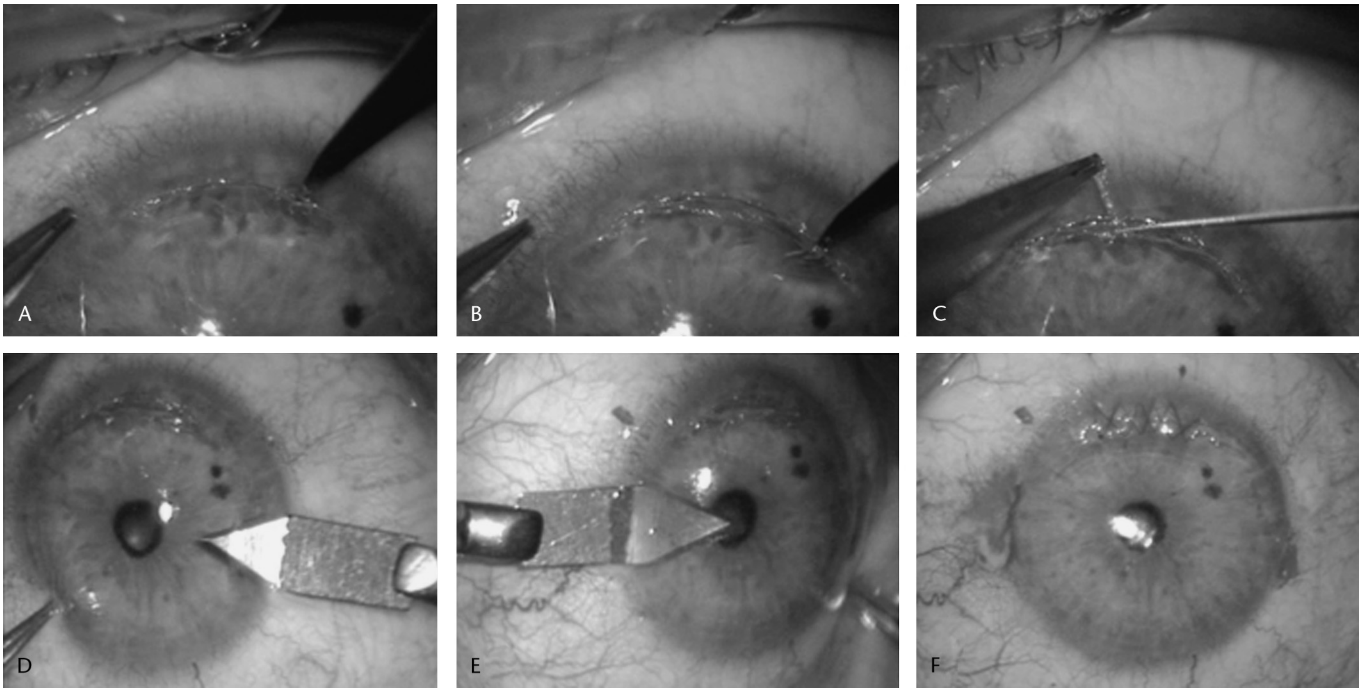

FIGURE 2. Photo essay of the surgical technique used. The (A) peripheral and (B) inner border of the tissue to be excised is marked with a 15° blade. (C) The wedge of tissue is excised. (D) Nasal and (E) temporal relaxing, clear corneal beveled tunnel incisions are made, and (F) the inferior wound is sutured with 1 continuous suture.

component increase was 0.59 D (n = 4). These changes were not found to be significant ($P = 0.28$ at 12 months, $P = 0.35$ at 24 months). By vector recomposition, the overall mean astigmatic drift from the sixth postoperative month until the end of the follow-up period (mean observation time, 14.4 months) was calculated to be 0.78 D.

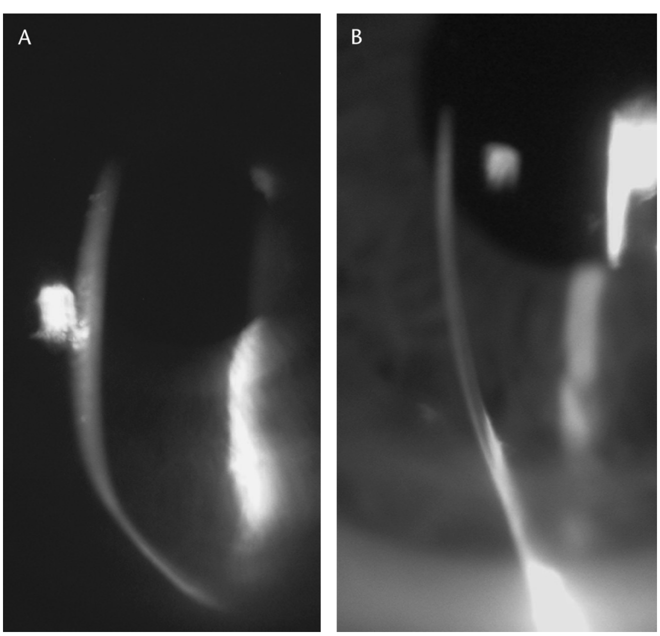

FIGURE 3. Slit-lamp photographs showing (A) the preoperative appearance of the thinned, inferior cornea, and (B) the postoperative appearance of the same cornea, after removal of sutures, showing successful excision of the thinned area of cornea.

DISCUSSION

The surgical treatment of PMCD described in this study aims at minimizing postoperative refractive error and improving its stability while preserving normal corneal tissue. This prospective, noncomparative, interventional case series evaluated the technique, with respect to reduction of keratometric cylinder and improvement of visual acuity.

Resection of a wedge-shaped piece of thinned tissue in PMCD, either partial or full thickness, has been described by

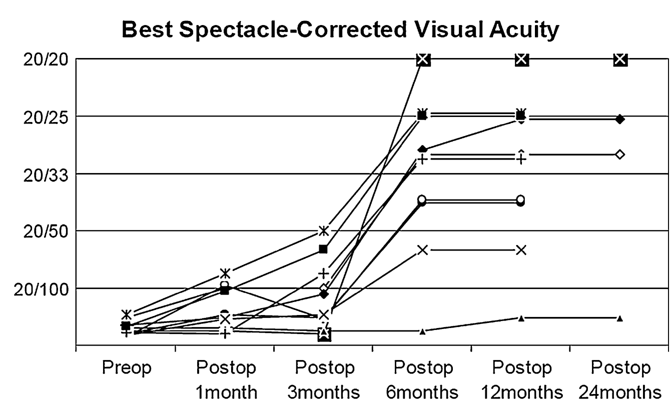

FIGURE 4. Best Snellen spectacle-corrected visual acuity of patients, before and after the wedge excision, for the 10 patients over the follow-up period.

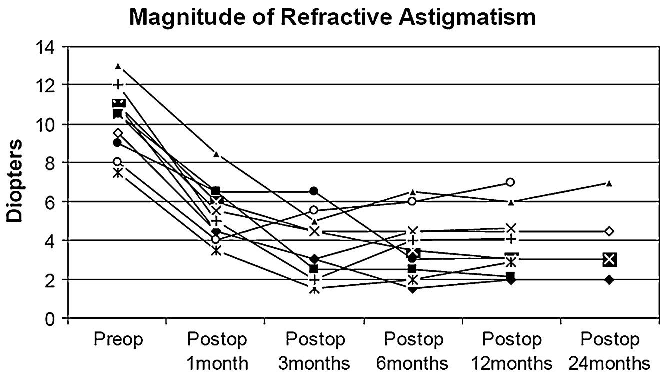

FIGURE 5. Absolute magnitude of astigmatism, obtained by subjective refraction, for the 10 patients over the follow-up period. This represents the outcome of most clinical interest and shows marked improvement compared with the preoperative values and good stability after the 6-month visit.

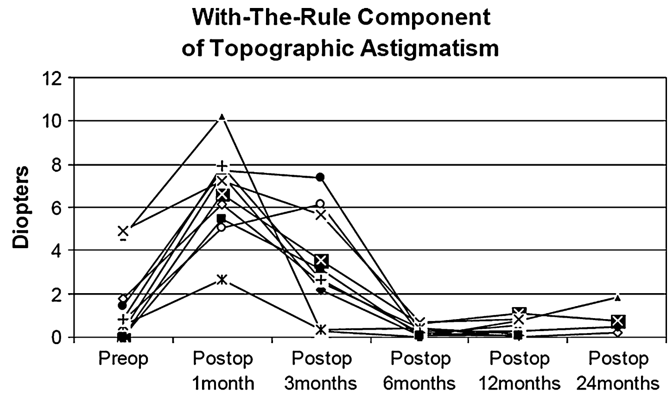

FIGURE 7. With-the-rule component of topographic astigmatism, obtained by trigonometric vector decomposition of the topographic values shown in Figure 6, for the 10 patients over the follow-up period. This shows the initial overcorrection at 1 month, which decreases markedly on removal of sutures.

several authors.[5,10,11] This technique presents several advantages over the use of a penetrating grafting procedure, including the preservation of normal central cornea, a mechanically stronger wound, shorter visual rehabilitation period (5.4 vs. 11 months),[8,11] and the absence of steroid-related adverse effects or immunologic rejection.[8]

To the best of our knowledge, CRIs have not previously been described in combination with wedge resection. The use of paired, opposed beveled clear corneal tunnel incisions to decrease astigmatism has been extensively studied.[16–19] They have been found to be an effective way of reducing astigmatism with an effect ranging between 0.50 and 2.25 D, and refractive stability reported within 1 week.[16] In addition, as opposed to vertical incisions, the architecture of beveled, self-sealing incisions reduces the risk of postoperative dehiscence, and wound contamination is less likely in the absence of exposed stroma.

Relaxing incisions performed in corneas with structural abnormalities, such as keratoconus and PMCD, may result in less predictable incisions than made in normal corneas. The rationale behind our combined approach is that incisions centered around the steeper meridian would potentiate the effect of a wedge resection performed in the inferior, ectatic cornea. As a consequence, a smaller excision of corneal tissue would be sufficient to obtain the desired effect, whereas the reduced tension in the corneal structure would permit less postoperative regression and more stable refractive results. At the same time, the effect of relaxing incisions would be maximized on a cornea with increased structural tension induced by the wedge resection.

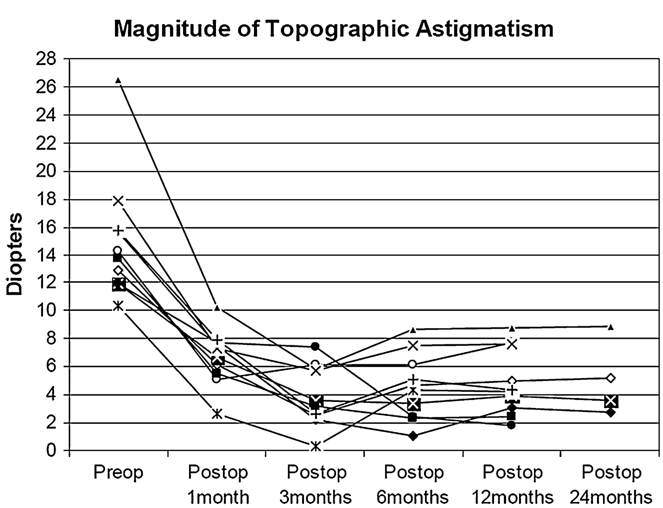

FIGURE 6. Absolute magnitude of astigmatism, obtained from the difference between the topographic Sim-K values, for the 10 patients over the follow-up period. The initial postoperative reduction in astigmatism is partly reversed when the sutures are removed, but there is astigmatic stability after the 6-month review.

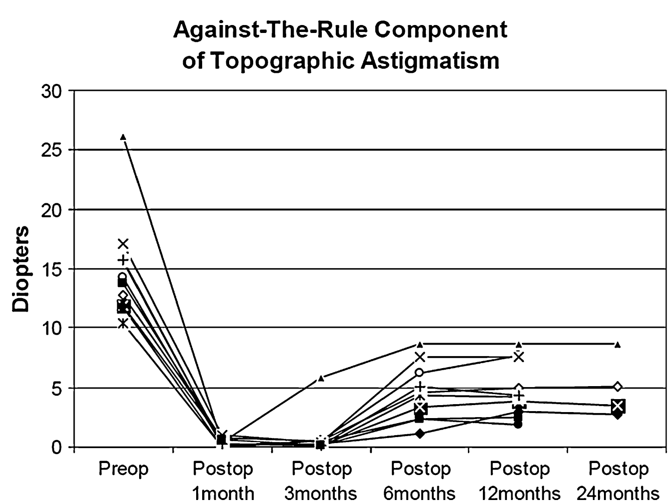

FIGURE 8. Against-the-rule component of topographic astigmatism, obtained by trigonometric vector decomposition of the topographic values shown in Figure 6, for the 10 patients over the follow-up period. The initial postoperative reduction in this component is partly lost on removal of sutures, but there is good stability over the 12- and 24-month follow-up period. Note the different vertical scale.

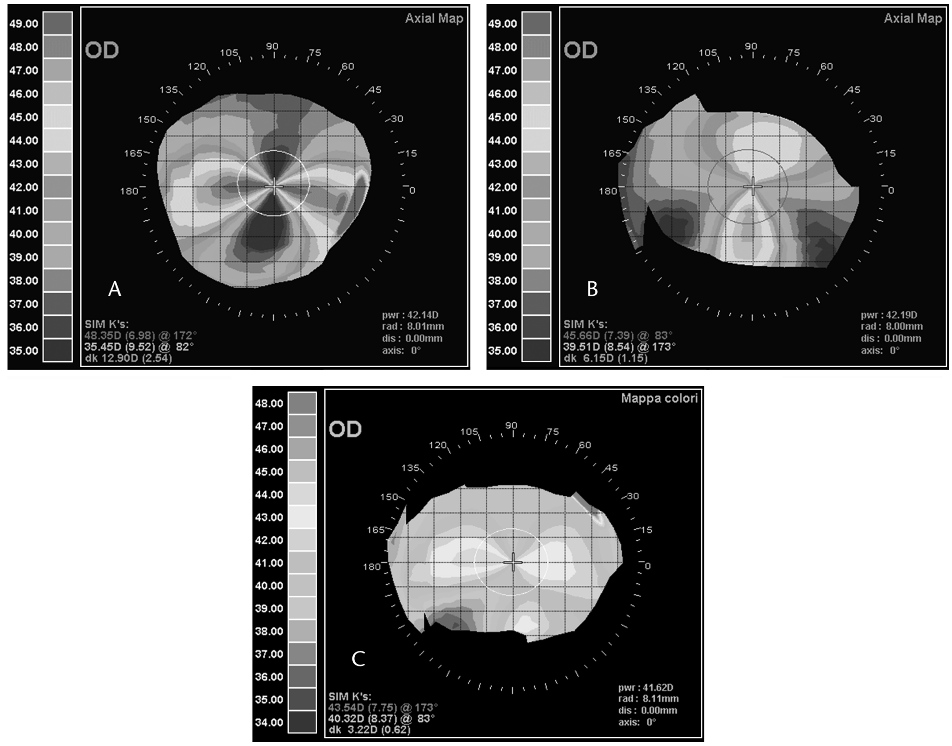

FIGURE 9. Topography images of (A) preoperative PMCD, showing advanced against-the-rule astigmatism, and (B) the same cornea, in the immediate postoperative period, showing overcorrection of the astigmatism. Six months after surgery (C), the map shows limited regression of effect, with low-degree against-the-rule astigmatism.

In our case series, a reduction in astigmatism from 15.1 to 4.6 D was obtained as a result of the procedure. The amount and axis of the cylinder stabilized between 3 and 6 months postoperatively, coinciding with the period of suture removal. There was minimal long-term astigmatic drift for the follow-up period. Accordingly, the visual acuity improved and stabilized over the same period.

Biswas et al,[5] who did not describe their wedge resection technique in detail, reported a mean postoperative astigmatism of 8.9 D after a 57-month follow-up period. MacLean et al,[11] on the other hand, obtained a mean postoperative keratometric astigmatism of 1.4 D, which is considerably less than the average astigmatism in our series. This is probably because of the more aggressive tissue resection used by MacLean et al (2- vs. our 0.5-mm-wide wedge) and a resulting greater dependence on suture tension. Selective suture removal performed by these authors during the postoperative period could successfully minimize the astigmatic error. However, the significant long-term astigmatic drift reported (2.1 D; range, 0.5–5.5 D; average follow-up, 59 months) suggests that the ongoing presence of tight sutures prevents stabilization of the result. Similarly, Cameron[7] reports a mean postoperative astigmatism of 2.6 D after a lamellar technique of wedge resection of undocumented width, but again with high dependence on suture tension. (n = 3; long-term astigmatic drift not evaluated). This study reports a much lower amount of astigmatic drift (0.78 D) over the 12- to 18-month period of observation. Although this is a shorter period of follow-up compared with MacLean et al, it can be reasonably expected to include the greater part of total long-term astigmatic drift, which is known to take place mainly early after surgery.[20] With respect to the time required to obtain a stable postoperative refraction, this was noted between 3 and 6 months in all patients of this study, comparing favorably with the range of 3–12 months reported by MacLean et al,[11] and 5.5–11 months reported by Cameron.[7] However, a more pronounced regression of effect may represent a trade off for increased stability of the refractive result, as obtained in the patients of our series as a consequence of early suture removal.

Eight of 10 patients achieved a BSCVA of 20/40 or better by 6 months. All patients were used to wearing spectacles with high cylinders (between 4 and 6 D) preoperatively, and none chose to wear hard contact lenses after surgery. Both retinal disease and persistent high astigmatism contributed to the poor visual outcome of 1 patient. Similar results have been obtained by Cameron[7] and MacLean et al,[11] but in 2 of the cases reported by the former, hard contact lenses were used, whereas the latter did not specify the type of correction. These statistics may therefore overestimate the effectiveness on the regularity of the corneal contour obtained by these authors, because hard contact lenses mask irregular and high-degree regular astigmatism.

Penetrating keratoplasty performed for PMCD yields varying astigmatic results. Varley et al,[8] who reviewed the keratometric outcomes of 12 large, eccentric grafts, obtained a mean astigmatism of 2.46 D, measured 6–21 months postoperatively. Sixty-four percent of grafts had at least 1 episode of rejection, with other complications also reported. Rasheed and Rabinowitz[6] reviewed the outcomes of 5 crescentic lamellar grafts performed in combination with central penetrating keratoplasties. After the 5–20 months needed for visual rehabilitation, a mean of 3.2 D astigmatism was obtained, with a further astigmatic drift over long-term follow-up. Two of the 5 patients developed steroid-related complications. Although the astigmatic outcomes are adequate in these 2 studies, the time necessary for visual rehabilitation and graft-related complications make penetrating keratoplasty a poor surgical option in comparison with wedge resection.

The results presented in this study suggest that the concomitant use of CRIs with corneal wedge resection in PMCD may reduce the dependence on suture tension, thereby obtaining more stable astigmatic results at an earlier postoperative point in time. These results could be further validated by evaluating a control group of patients undergoing first the wedge resection and then adding the beveled CRI at a later time. The authors suggest that the moderate residual astigmatism reported here could be prevented by further overcorrecting the against-the-rule astigmatism, such as by resection of a wider wedge of thinned tissue, longer CRIs (extending to 90° arcs),[13] and/or more centrally located CRIs.[21]

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