Cataract surgery after lamellar wedge resection in advanced pellucid marginal degeneration: A case report

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ABSTRACT

In advanced pellucid marginal degeneration (PMD) cases, Lamellar Wedge Resection (LWR) is a method that is used to postpone or eliminate the necessity for allogenic lamellar or penetrating keratoplasty.In this study, we presented the results of LWR and cataract surgeries in two cases, 62 years old with bilateral PMD (case 1) and 83 years old with unilateral PMD (case 2) presenting with acute hydrops.In Case 1, topographic astigmatism (TA) decreased from 20.2 D to 6.3 D, and visual acuity (VA) increased from 1.80 LogMAR to 0.2 LogMAR Snellen chart after all sutures have been removed.In the second year TA increased to 15.2 D and did not change until the end of the 3rd year. In Case 2, TA decreased from 11.6 D to 3.3 D after 28 months of follow-up, and VA increased from 2.1 LogMAR to 0.10 LogMAR level. Monofocal IOL was performed due to the continuous changes in astigmatism in the early period after LWR, resulting in an increase in VA in both cases.

Keywords: Pellucid marginal degeneration, wedge resection, phacoemulsification.

INTRODUCTION

Pellucid Marginal Degeneration (PMD) is a rare, progressive, non-inflammatory corneal ectasia characterized by slow progression of vision loss between the 3rd and 5th decades of life, which most commonly involves the lower quadrant of the cornea. It is characterized by an area of thinning in the 4-8 o'clock positions, separated from the limbus by a 1-2 mm intact area and the anterior bulging of cornea just superior to the thinning area.[1] The vision loss results from progressive irregular astigmatism while irregular astigmatism may develop. The vertical flattening of the cornea and corneal ectasia just superior to thinning area form the typical topographic appearance of a 'butterfly wing' or 'crab claw'.[1] Rare cases have also been reported in the upper and temporal quadrants.[2,3] The visual acuity can be improved with glasses and contact lenses in cases at early phases; however, surgical treatment is often necessary in advanced cases. In advanced PMD cases, Lamellar Wedge Resection (LWR) is a method used to reduce or delay the need for allogeneic lamellar or penetrating keratoplasty in surgical treatment.[1,4]

In this study, we presented two cases with advanced Pellucid Marginal Degeneration (one case with unilateral PMD and other with bilateral advanced PMD) which were treated with Lamellar Wedge Resection and subsequent cataract surgery.

Case 1: A 62-year-old female patient presented with complaint of progressively reduced vision acuity (VA) over the past 10 years. In the initial examination at presentation, visual acuity (VA) was 0.7 logMAR in the right eye and 1.80 logMAR in the left eye. Biomicroscopic examination revealed that the central cornea was clear in both eyes, and that there was a crescent-shaped area of thinning 3 mm above the limbus in the lower quadrant, as being more pronounced in the left eye (Figure 1a). No iron lines, vascularization, or lipid deposits were observed. There was corticonuclear cataract in the left eye. Intraocular pressure

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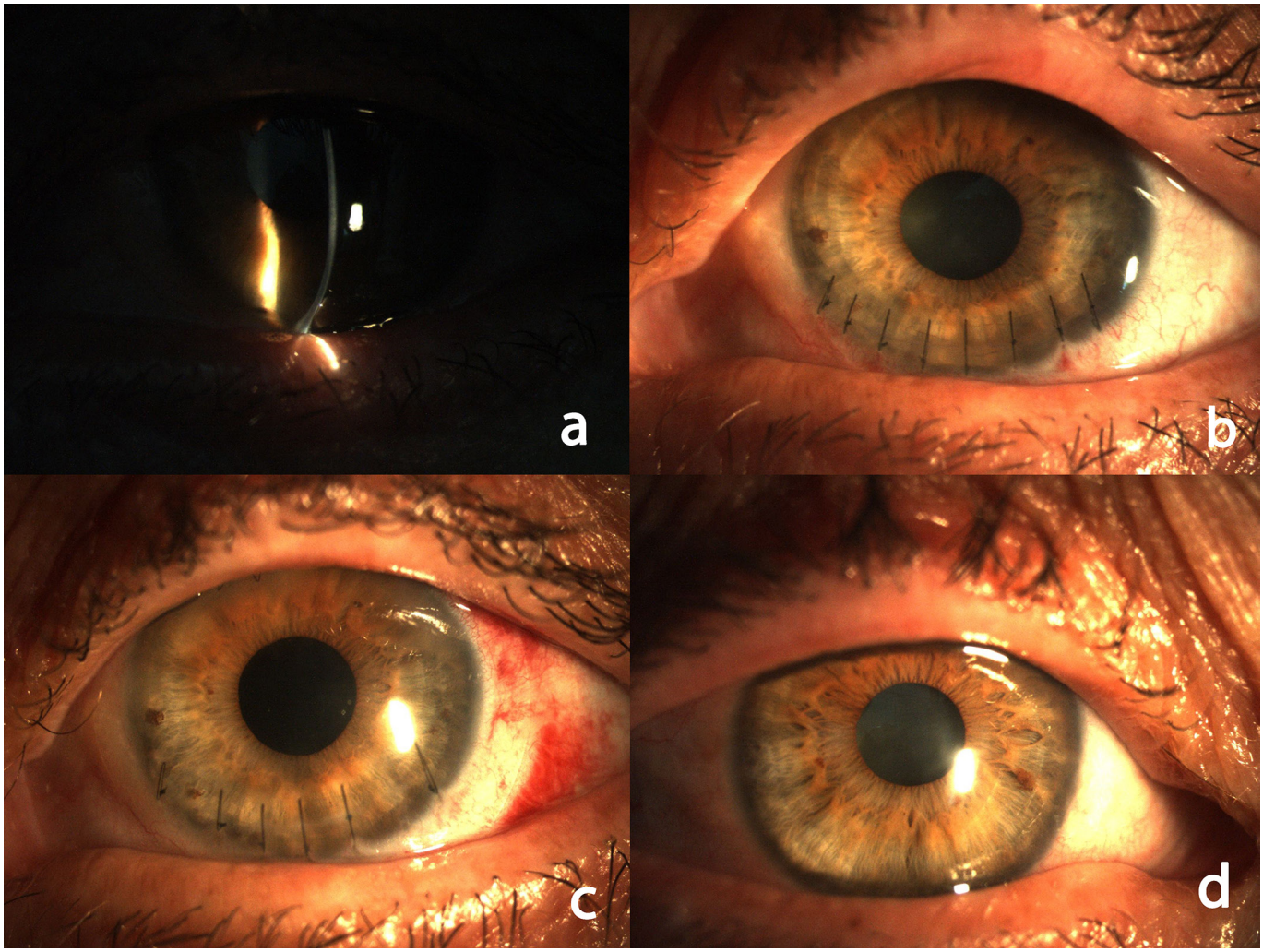

Figure 1: *Case 1 Anterior segment image of left eye a:-pre LWR , b; post LWR week 1 c; Day 2 after cataract surgery d; post-lamellar wedge resection year 3.*

was measured as normotensive in both eyes. Topographic examination (Pentacam, Oculus Optikgeräte GmbH, Wetzlar, Germany) revealed high "against-the-rule" astigmatism and the typical 'crab claw' appearance for PMD in both eyes (Figure 2a). Central corneal thickness was 484 microns (µ) in the right eye and 477 µ in the left eye while total anterior and posterior topographic astigmatism (TA) was 20.6 diopters (D) in the right eye and 20.2 D in the left eye. Surgery was planned for the left eye with lower visual acuity. LWR was performed on the left eye as the central cornea was clear and the central corneal thickness was 477 µ.

Under the surgical microscope, the edges of the crescent-shaped area (1.5-2 mm in width) to be excised were marked using gentian violet. Lamellar corneal tissue was excised to a depth of approximately 80% using a crescent knife. Anterior chamber paracentesis was performed through a 1.2 mm side port incision to achieve ocular hypotony. The wound created by LWR was closed with 10-0 monofilament

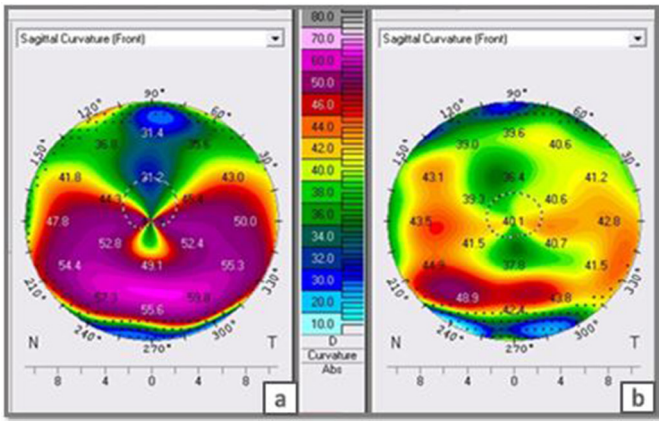

Figure 2: *Case 1 Topographic change a; post-lamellar wedge resection, b; Year 1 after ctaract surgery.*

nylon sutures (n=8) where the first suture placed at central, followed by stepwise addition of single sutures to both edges (Figure 1b). At postoperative period, topical antibiotic drops were administered 5 times daily over 2 weeks while topical corticosteroids were tapered over 2

months. On post-LWR week 1, VA was 0.7 logMAR while TA was 10 D and central corneal thickness was 583 µ. Due to the progressive nature of the disease, collagen cross-linking (CxL) was performed 4 months after LWR. After CxL, TA was 10.3 D with mild central corneal haze and thickness of 457 µ. After 7 months, phacoemulsification surgery (PE) plus a three-piece intraocular lens (IOL) (AcrySof MA60AC) implantation were performed due to corticonuclear cataract through a 3.0 mm superior clear corneal incision under subtenon anesthesia (Figure 1c). The IOL power was calculated using the SRK II formula based on optical coherence biometry (AL-Scan, Nidek Co., Aichi, Japan).

Selective suture removal started at month 9 and was completed by month 15 . After 15-months follow-up, TA decreased gradually from 20.2 D to 6.3 D (Figure 2b), and VA improved from 1.8 logMAR to 0.3 logMAR (-2.50 axis 180°). In the patient, the minimum topographic astigmatism was observed in this period where suture removal was completed. TA was gradually increased in the year following suture removal, reaching up to 15.2 D. In the subsequent 12 months, TA remained at 15.2 D (Table 1). At the end of the 36-months follow-up, VA was maintained between 0.52 logMAR (-5.00 axis 90°) 0.30 logMAR (Figure 2d). Although the patient was informed about the progressive nature of the disease, she decline CxL treatment for her right eye; thus, no intervention was performed on the right eye.

Case 2: An 83-year-old female patient presented with complaints of watery eye, light sensitivity, and stinging in her left eye. The visual acuity (VA) was 2.10 logMAR in the left eye. Ophthalmological examination revealed crescent-shaped corneal edema extending temporally in the lower quadrant, anterior bulging, Descemet membrane breaks, and corticonuclear cataract in the left eye (Figure 3a). She has no history of immunological disease, trauma, or prior ocular surgery. The patient was diagnosed with acute hydrops in the left eye and received conservative treatment. After treatment for acute hydrops, biomicroscopic examination showed a crescent-shaped thinning in the 4-8 o'clock positions, separated from the limbus by a 2 mm intact zone (Figure 3b, c); in addition, a topographic 'crab claw' appearance was present (Figure 4a). Biomicroscopic and topographic findings led to a diagnosis of PMD in the patient. The total astigmatism (TA) was 11.6 D, and the central corneal thickness was 453 µm. No pathological

Table 1: *Keratometric and corneal thickness changes.*

	K1 (D)	K2 (D)	Apex (µ)	Astigmatism anterior surface (D)	Astigmatism posterior surface (D)	Axis (degree)	K1 (D)	K2 (D)	Apex (µ)	Astigmatism anterior surface (D)	Astigmatism posterior surface (D)	Axis (degree)
	Case 1											
	Right Eye						**Left Eye (LWR)**					
Preop	36.3	53.6	484	17.3	3.3	90.2	36.2	53.4	477	17.2	3.0	89.7
Postop Week 1	36.6	51.0	503	14.3	3.3	90.2	32.8	40.6	583	7.8	2.2	1.5
Postop Month 1	37.2	52.5	494	15.3	2.9	84.7	35.3	49.3	489	14.0	2.1	177.6
Postop Month 6	36.4	53.3	483	16.8	3.2	93.6	37.8	46.8	457	9.0	1.3	6.1
Postop Year 1	35.8	53.8	486	17.6	3.4	95.2	36.5	42.2	394	5.7	0.6	100
Postop Year 2	38.0	53.8	484	15.8	3.2	92.4	31	44.7	384	13.7	1.5	93.2
Postop Year 3	37.2	53.9	491	16.7	3.2	86.7	31.6	45.2	329	13.7	1.5	91.5
	Case 2											
	Right Eye						**Left Eye(LWR)**					
Preop	47.0	47.8	497	0.8	0.3	2	42.2	52.6	453	10.4	1.2	98
Postop Week 1	47.1	47.6	490	0.5	0.2	19	42.8	53.2	469	10.4	1.2	24.7
Postop Month 1	46.6	48.3	483	1.8	0.3		42.9	55.3	459	12.4	1.2	28.5
Postop Month 6	46.9	47.6	497	0.6	0.1	38	44.8	54	446	9.2	1.0	27.4
Postop Year 1	47.0	48.0	513	1.0	0.6	23	39.0	47.0	299	7.4	1.0	52.5
Postop Year 2	46.5	47.5	504	0.9	0.1	21.4	39.1	41.8	260	2.7	1.2	65.0

Preop: Preopretaive, Postop: Postoperative

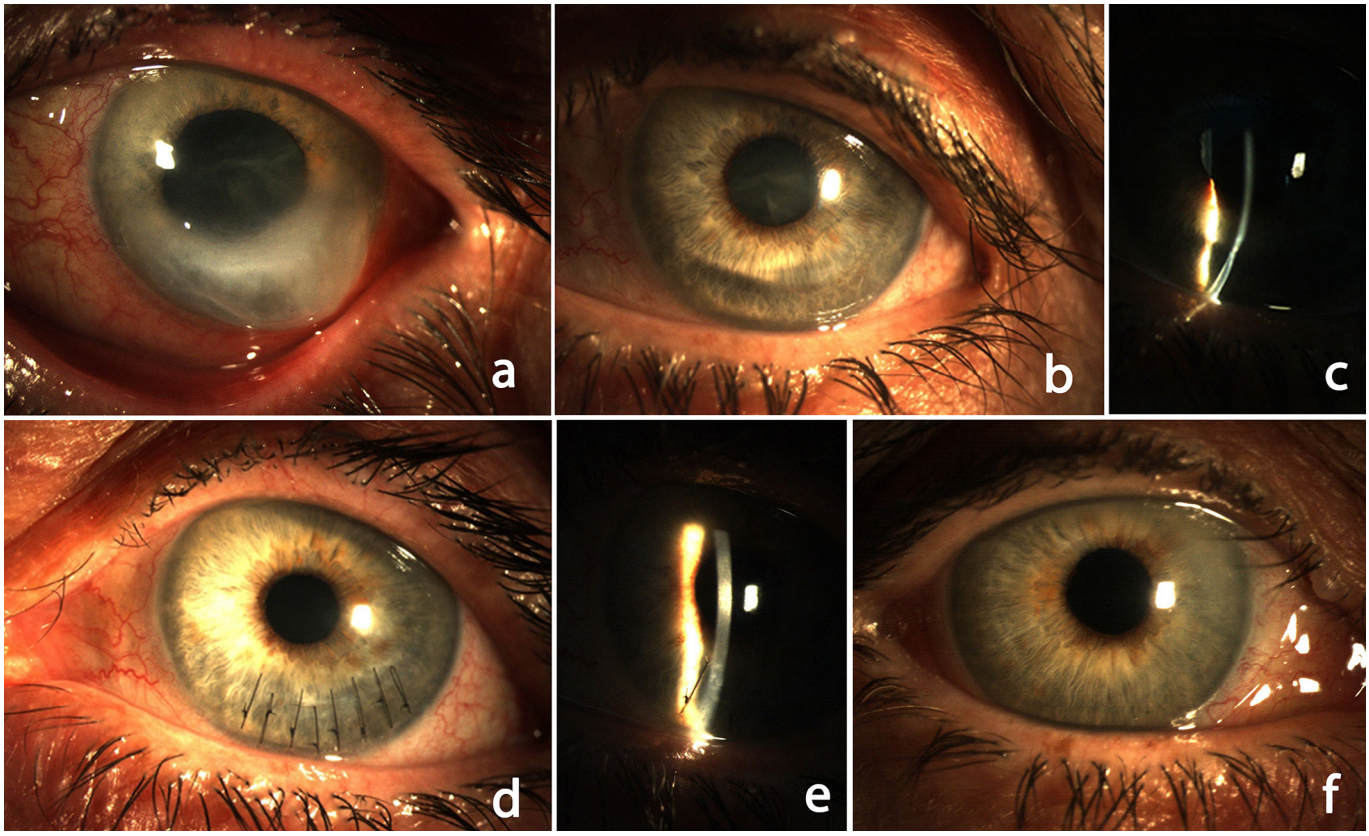

Figure 3: *Case 2 Anterior segment image a: presentation, b: After treatment of hydrops but before lamellar wedge resection , c: Slit lamp image before LWR d: Week 1 after cataract surgery e; Year 1 after cataract surgery partial suture removal, f: final examination of right eye.*

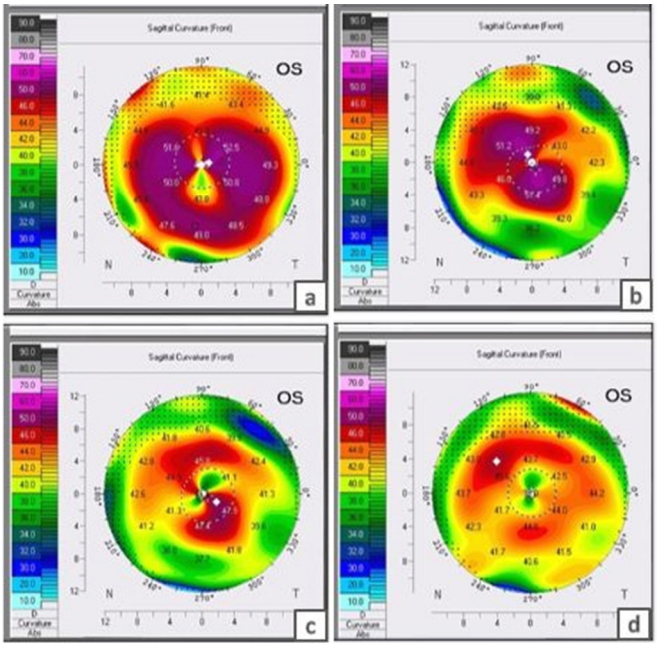

Figure 4: *Case 2 Topographic change a: pre-lamellar wedge resection, b: Month 6 after cataract surgery, c: postoperative year 1, suture removal was completed, d: postoperative year 2.*

findings suggestive of PMD or keratoectatic disease were observed in the right eye (Figure 4b). The TA was 1.1 D, and the corneal thickness was 497 µm in the right eye.

Six months after the treatment for hydrops , LWR was performed as described in the first case, followed by phacoemulsification surgery (PE) and subsequent three-piece intraocular lens (IOL) (AcrySof MA60AC) implantation into the capsule one week later (Figure 3d). The TA decreased from 11.6 D to 8.4 D at 12-months follow-up whereas 3.3 D at the 28-months follow-up (Table 1). The VA improved from 2.10 logMAR to 0.52 logMAR and further to 0.1 logMAR with a correction of +1.00 (+2.00 axis 170°) during follow-up (Figure 3e). At the final examination, the right eye also showed a TA of 1.0 D, a corneal thickness of 515 µm, and a VA of 0.05 logMAR, corrected to 0.00 logMAR (-0.50 axis 130°) (Figure 3f). Intraocular pressure remained within normal limits throughout the follow-up period.

DISCUSSION

Typically, PMD is a bilateral peripheral corneal ectasia affecting the 4-8 o'clock positions of the cornea in older individuals. Albeit rare, cases with unilateral and superior quadrant PMD have been reported in the literature.[2,3] Acute corneal hydrops is also highly rare in PMD, seen in only 2.5-6% of cases.[5] In case 2, there was an unusual presentation of unilateral PMD with acute hydrops. Over

the 28-month follow-up period, no pathological finding suggestive of PMD or keratoectatic disease were observed in the right eye of the patient.

Since PMD is a progressive ectasia, collagen cross-linking (CxL) treatment with riboflavin and ultraviolet A (UVA) light is used to prevent the PMD progression although it typically occurs in older individuals. In previous studies, it was shown that disease progression can be stopped with CxL treatment, either alone or in combination with photorefractive keratectomy.[6, 7] In case 1, we performed CxL treatment 4 months after LWR. Following CxL treatment, TA decreased from 20.2 D to 6.3 D during the approximately 15-months period where selective suture removal was completed, . In the subsequent 12 months, TA increased to 15.2 D and remained stable at 15.2 D over 12 months.

In cases with early PMD, visual improvement can be achieved with glasses and contact lenses, while advanced cases often require surgical treatment. Due to the progressive nature of the disease and its frequent localization in the lower peripheral cornea, several surgical methods including intracorneal rings, full-thickness or lamellar crescent-shaped wedge resection, phakic intraocular lenses, lamellar keratoplasty, and penetrating keratoplasty have been described to improve visual acuity.[1, 8]

Lamellar wedge resection offers several advantages in the treatment of advanced pellucid marginal degeneration (PMD). This technique preserves the nearly normal corneal thickness of the clear corneal tissue, allowing for rapid visual rehabilitation by reshaping the cornea.[4] Since no allogeneic tissue is used, no long-term steroid use is required, avoiding immunological rejection reactions and the side effects of corticosteroids. By excision of the ectatic corneal tissue and suturing the wound, a mechanically stronger corneal structure is formed. Additionally, it avoids the complications associated with open sky surgery because no full-thickness resection is performed.

There are studies reporting successful outcomes in correcting high refractive errors and astigmatism through lens extraction combined with phakic or toric intraocular lens (IOL) implantation in pellucid marginal degeneration (PMD).[9,10] Toric IOL implantation can be an good option to correct astigmatism in ectatic corneal diseases with concurrent cataract. Before planning the surgery, it is crucial to ensure that the ectasia progression is stabilized. Studies indicate that current toric IOL implants correcting up to 6 D and custom-made toric IOLs for higher astigmatism cases yield good visual outcomes in stable PMD cases.[10-13]

In cataract surgery for ectatic diseases, a significant challenge is calculation of the IOL power due to high astigmatism and myopia. The location of the ectasia is a crucial parameter that affects the accuracy of the surgical outcome. In PMD, it was reported the peripheral location of the ectasia had a lesser impact on keratometric measurements, resulting in a more accurate IOL power calculation.[11,14] Thebpatiphat et al. compared the SRK, SRK II, and SRK/T formulas in keratoconus patients. Authors found that the most accurate results were obtained in patients with moderate keratoconus. They also reported that the SRK II formula provided more reliable results, while , the calculations were less precise in eyes with advanced and severe keratoconus, and there were no significant differences between the formulas.[13,14]

If toric IOL implantation would be planned, it is important to ensure that the astigmatism is relatively regular and that the manifest refraction is compatible with topographic and biometric measurements, especially in cases of high astigmatism.[15,16] In the recent literature review (2018-2020) Smith et al. suggest a hybrid approach once stability is achieved by taking stage of keratoconus into account. Authors suggest the comparative use of the Barrett II formula and the SRK-T formula for moderate keratoconus patients as a more reliable method. For stage 3 keratoconus patients, authors suggest using the SRK-T formula if the corneal power is below 55 D; however, if the corneal power exceeds 55 D, they suggest using a keratometric value of 43.25 D rather than the actual corneal power, aiming for a target refraction of (-0.75) to (-1.25) D.[15]

Some authors recommend avoiding toric IOL implantation if patients achieve good preoperative visual acuity with soft or hard contact lenses.[17,18] Patients should be informed that they might experience refractive surprises following toric IOL implantation and may require more complex contact lens systems for optical correction.

Ideally, cataract surgery should be planned once corneal stability is achieved in patients with keratoectatic diseases. However, in our two cases, cataract surgery had to be performed shortly after LWR due to low visual acuity caused by cataracts and unwillingness to wait for corneal stability. Astigmatic shift is an anticipated situation in the early period after LWR. Therefore, monofocal IOL implantation was preferred in these two cases where cataract surgery was required early after LWR. In both cases, significant changes in astigmatism were observed due to LWR wound healing and selective suture removal post-cataract surgery. At the end of year 1 after selective

suture removal, the extent of change in astigmatism, with stabilization of refraction in both cases.

In conclusion, LWR achieved regular corneal astigmatism in two cases of Pellucid Marginal Degeneration. Ideally, after achieving refractive stability post-LWR, cataract surgery and toric IOL implantation should be performed in suitable cases to correct corneal astigmatism. However, in cases where cataract surgery is required in the early period before refractive stability is achieved, monofocal IOL implantation can be used to enhance visual acuity and avoid postoperative refractive surprises due to changes in astigmatism. Given the progressive nature of the disease, large patient series and long-term follow-up are necessary to evaluate the effectiveness of these methods.

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