

# Pellucid marginal degeneration: postponing penetrating keratoplasty for over a decade

## *Degeneração marginal pelúcida: postergando o transplante de córnea em mais de uma década*

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### ABSTRACT

*Pellucid marginal degeneration (PMD) treatment can be challenging in moderate to advanced cases and penetrating keratoplasty should be avoided due to high risk of complications, such as graft rejection. We report a case of a 30-year-old female patient with PMD that was referred to our service with bilateral low visual acuity, worse in her right eye and contact lenses intolerance. We performed a corneal crescentic lamellar wedge resection that resulted in good useful vision and postponed the keratoplasty for eighteen years.*

**Keywords:** Cornea; Corneal topography; Corneal diseases; Corneal surgery, Ectasia; Corneal transplantation; Visual acuity; Case reports

### RESUMO

A degeneração marginal pelúcida (DMP) é uma ectasia corneana de difícil manejo em casos moderados e avançados. O transplante de córnea penetrante nesses pacientes deve ser evitado ao máximo tendo em vista que a descentração necessária do botão aumenta o risco de vascularização e consequentemente rejeição, além das dificuldades técnicas e possibilidade de deiscência de sutura devido ao afinamento corneano pronunciado característico desta ectasia. Nós relatamos o caso de uma paciente de 30 anos com DMP com queixa de baixa visual bilateral, pior no olho direito e intolerância a lentes de contato. Foi realizada ressecção lamelar em crescente neste olho, com excelente resultado, concedendo visão útil a paciente e postergando o transplante penetrante por dezoito anos.

**Descritores:** Córnea; Topografia da córnea; Doenças da córnea; Ectasia; Transplante de córnea; Acuidade visual; Relatos de casos

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**INTRODUCTION**

**P**ellucid marginal degeneration (PMD) is a bilateral cause of corneal ectasia and is characterized by inferior corneal thinning, approximately 1 - 2 mm in vertical length, typically between the 4 o'clock and 8 o'clock positions, crescent-shaped, with the point of steepest corneal protrusion occurring above the area of stromal thinning.<sup>(1)</sup> The gold standard diagnostic test for PMD detection is corneal topography, that will demonstrate flattening of the vertical meridian and higher curvature inferiorly, generating an “against-the-rule” astigmatism, with a “butterfly” or a “crab claw-like” appearance.<sup>(2,3)</sup> The treatment for PMD is difficult and challenging, due to poor results in visual acuity improvement with glasses, decreasing stability of contact lenses as the disease progresses and the need of a larger and inferiorly decentered graft, closer to the limbus, if penetrating keratoplasty is chosen, increasing the risk of corneal graft rejection.<sup>(4)</sup>

To improve visual acuity in advanced cases, avoiding or postponing a penetrating keratoplasty, several techniques have been described, such as lamellar crescentic wedge resection, exemplified in this case report. This technique made it possible in this case to postpone the corneal transplant for 18 years, permitting rehabilitation of useful vision to the patient in this period.

**CASE REPORT**

A 30-year-old female patient was referred to Hospital do Servidor Público Estadual - São Paulo in 1996 complaining of slowly and bilateral decreasing of visual acuity, worse in the right eye. Refraction was + 2.50<sup>Δ</sup> - 7.50 75° (20/200) in her right eye and -0.50<sup>Δ</sup> - 2.50 105° (20/40) in her left. Slit-lamp exam showed an inferior corneal thinning in both eyes, accentuated in the right eye. Corneal topography closed the diagnosis of Pellucid Marginal Degeneration as suggested by the clinical exam (Figure 1). Due to contact lenses intolerance, the patient was submitted to lamellar crescentic wedge resection in the right eye, according to the following surgical steps: 1) Mark of the thinning’s horizontal length at the slit-lamp 2) Inferior limbal peritomy 3) Manual lamellar resection of the thinning marked area 1mm wide and as deep as possible with a crescent format 4) Paracentesis 5) Suture of the wound with single knots using mononylon 10.0, leaving 10 dioptres with-the-rule astigmatism at the intraoperative keratotomy.

The patient acquired good vision with glasses in the right eye (20/40) and this scenario was stable until 2003, when she abandoned treatment.

In 2009, the patient returned with complain of low visual acuity in her left eye. Refraction was +1.00<sup>Δ</sup> -5.50 70° (20/50) and +0.50<sup>Δ</sup> -8.50 80° (counting fingers). The topography demonstrated increased irregular astigmatism in both eyes (Figure 2).

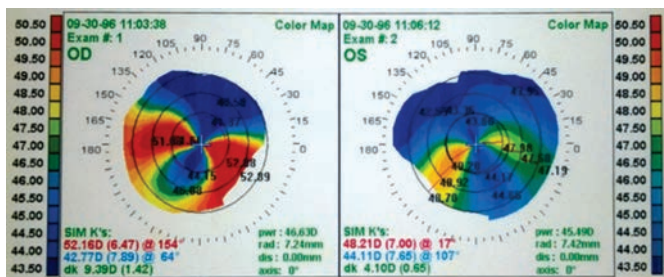


Figure 1: A: Preoperative corneal topography (September/1996).

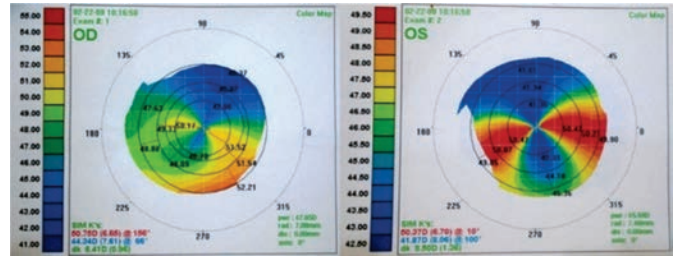


Figure 2: Topography in 2008 (OD = Right eye: 12 years after lamellar crescentic wedge resection; OS = Left eye: preoperative)

The patient was then submitted to a lamellar crescentic wedge resection in her left eye. Same surgery that was performed in her right eye in 1996. The final result is shown in figure 3.

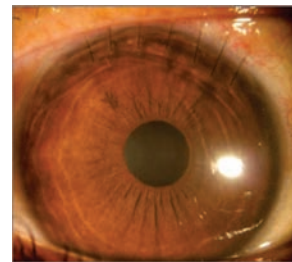


Figure 3: One month postoperative – left eye (2010).

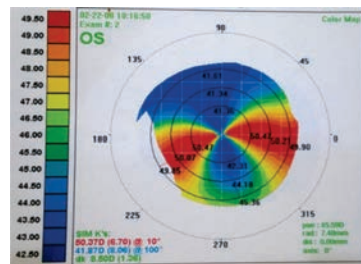


Figure 4: Left eye’s astigmatism preoperative (2008).

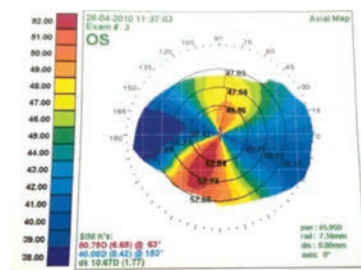


Figure 5: Left eye’s astigmatism in 2010 (postoperative).

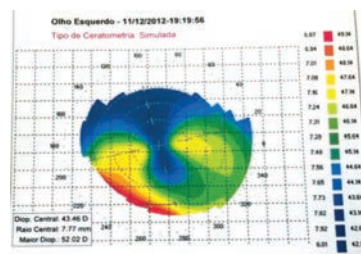


Figure 6: Left eye’s astigmatism in 2012.

The astigmatism evolution of the left eye is presented in table 1 and figures 4 to 6. Note the 2010 with-the-rule astigmat after the procedure and the slow return of the ectasia shown in 2012.

**Table 1**  
**Left eye's refraction evolution**

Date	Refraction	Visual acuity
Sept 02/2009	+0.50 – 8.50 80°	Counting fingers
Feb 24/2010	lamellar crescentic wedge resection	
April 08/2010	+1.50 – 4.50 165°	20/30
May 30/2012	+0.50 – 3.25 105°	20/30
May 18/2015	+1.50 – 4.75 105	20/25

Due to reduction in the right eye's visual acuity (20/100), penetrating keratoplasty was indicated in 2014. We were able to postpone the procedure for 18 years using the lamellar crescentic wedge resection technique.

### DISCUSSION

The non-surgical treatments available for PMD include spectacles and soft contact lenses for initial cases and rigid gas permeable (RGP) lenses for moderate cases. Hybrid contact lenses, with a rigid gas permeable center surrounded by a soft material also provide good results. As the ectasia and corneal thinning progresses, RGP lenses fitting becomes more difficult. In these cases, scleral contact lenses may represent a good alternative. When useful visual acuity can not be achieved by clinical methods, a surgical approach should be elaborated.<sup>(2,4)</sup>

From all the surgical options available, penetrating keratoplasty tends to be the most avoided one. The limbus proximity with the corneal thinning area demands larger and inferiorly decentered grafts, which raises the chances of corneal neovascularization, rejection, secondary glaucoma and also makes intraoperative suture more difficult.<sup>(2, 4, 5)</sup>

Several surgical techniques have been described as an alternative to penetrating keratoplasty, such as full-thickness crescentic wedge resection, crescentic lamellar wedge resection, crescentic lamellar keratoplasty, deep anterior lamellar ceratoplasty, intracorneal rings and also a combination of some of these procedures.<sup>(6-10)</sup>

Crescentic lamellar wedge resection, opposite to full-thickness crescentic wedge resection, does not penetrate the anterior chamber and preserves the posterior layers of corneal

stroma. Both techniques do not require donor tissue, excluding the risk of corneal rejection. The with-the-rule astigmatism induced by the surgery must be high, because with time, there is a reduction of it associated with corneal remodeling and return of the ectasia. It allows to postpone a penetrating keratoplasty and increases the thickness of the inferior cornea, making it possible to perform it later without decentering the corneal graft.<sup>(2,5,6)</sup>

In this case report, the patient had good visual acuity on her right eye over a decade after crescentic lamellar wedge resection and remains with good visual acuity on her left eye after the same procedure performed six years ago, which exemplifies the benefits of this surgical technique.

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