Transplant Prevention: Management of PMD with Ampleye® Scleral Lenses

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Abstract: Specialty contact lenses and corneal transplantation are both treatment options that can restore vision and improve quality of life for patients with corneal ectasias, such as pellucid marginal degeneration (PMD). PMD is often grouped together with other ectasias such as keratoconus (KCN), however its distinguishing features have important clinical implications. This case report summarizes the distinctions between PMD and KCN and outlines why scleral lenses should be considered superior to RGP lenses in the management of PMD and attempted prior to cornea transplantation.

Key Words: *Pellucid Marginal Degeneration, Corneal Degeneration, Scleral Lenses, Contact Lenses, Corneal Scarring, Contact Lens Intolerance, Gas Permeable Lenses*

Introduction

Pellucid marginal degeneration (PMD) is a bilateral, non-inflammatory, ectatic disease characterized by thinning of the peripheral cornea[1]. PMD typically presents with an area of inferior thinning separated from the limbus by 1-2mm[1]. The incidence or prevalence of PMD is not clearly reported as it is often misdiagnosed as keratoconus due the similarities in clinical presentation[2]. The Collaborative Longitudinal Evaluation of Keratoconus (CLEK) study established that those with corneal ectasias have a significantly impaired vision-related quality of life[3]. Given the significant impact of PMD and other ectasias, an increasing number of patients and practitioners have turned to specialty contact lenses for visual rehabilitation.

PMD results in irregular astigmatism, causing aberrations that are not correctable by standard glasses or contact lenses[3]. Scleral lenses are large diameter gas permeable lenses that mask optical distortion by allowing a tear lens to form between the posterior surface of the contact lens and the irregular surface of the cornea. Advantages of scleral lenses include their ability to clear the cornea and provide vision free from distortion.

The fitting of contact lenses in PMD is in general more challenging than in other corneal degenerations due to the large inferior protrusion of the cornea[4]. Traditional rigid gas permeable (RGP) lenses can be difficult to fit in PMD as the inferior location of the apex of the cone and large area of corneal involvement cause inferior decentration of the lens[4] Because scleral lenses vault the cornea entirely, they can eliminate decentration challenges and are useful in the management of PMD.

The following case report discusses a successful scleral lens fitting for a ‘contact lens intolerant’ patient with advanced PMD, delaying and possibly eliminating the need for a corneal transplant. This report will highlight considerations for optimal management of PMD and its commonly associated scleral lens complications.

Case Report

Initial Visit (11/29/2022)

A 55-year-old Caucasian male was referred by his optometrist for consideration of corneal transplantation due to contact lens intolerance, corneal scarring, and thinning OS>OD secondary to PMD. He reported that his habitual RGP contact lenses, fit by another practitioner 1 year prior, were causing significant discomfort and poor vision. The patient had failed two scleral lens fittings in the past two years due to persistent issues with fogging and discomfort, and felt he was unable to tolerate contact lenses.

Ocular history was remarkable for bilateral PMD OS>OD. BCVA in the habitual RGP lenses was 20/20 OD, 20/40 OS. On examination, it was noted that both habitual RGP lenses displayed excessive corneal touch, and <0.10 mm movement. OS K exam was

remarkable for extensive scarring corresponding to the area of lens touch. Notable bowing of the inferior cornea was noted OU.

Corneal topography from the initial visit revealed keratometry of 54.88D/57.01D @ 155 with a K_{max} of 58.0D OD in the right eye, and 50.30D/60.27D @ 120 with a K_{max} of 60.52 OS. Corneal astigmatism was 2.13D OD and 9.97D OS. OD topography revealed a classic “kissing dove” or “crab claw” topographic pattern characteristically seen in PMD. OS topography was limited in quality due to the extensive corneal scarring noted on examination. Topography from Nov 29, 2022, can be seen in **Figure 1** and **Figure 2** respectively.

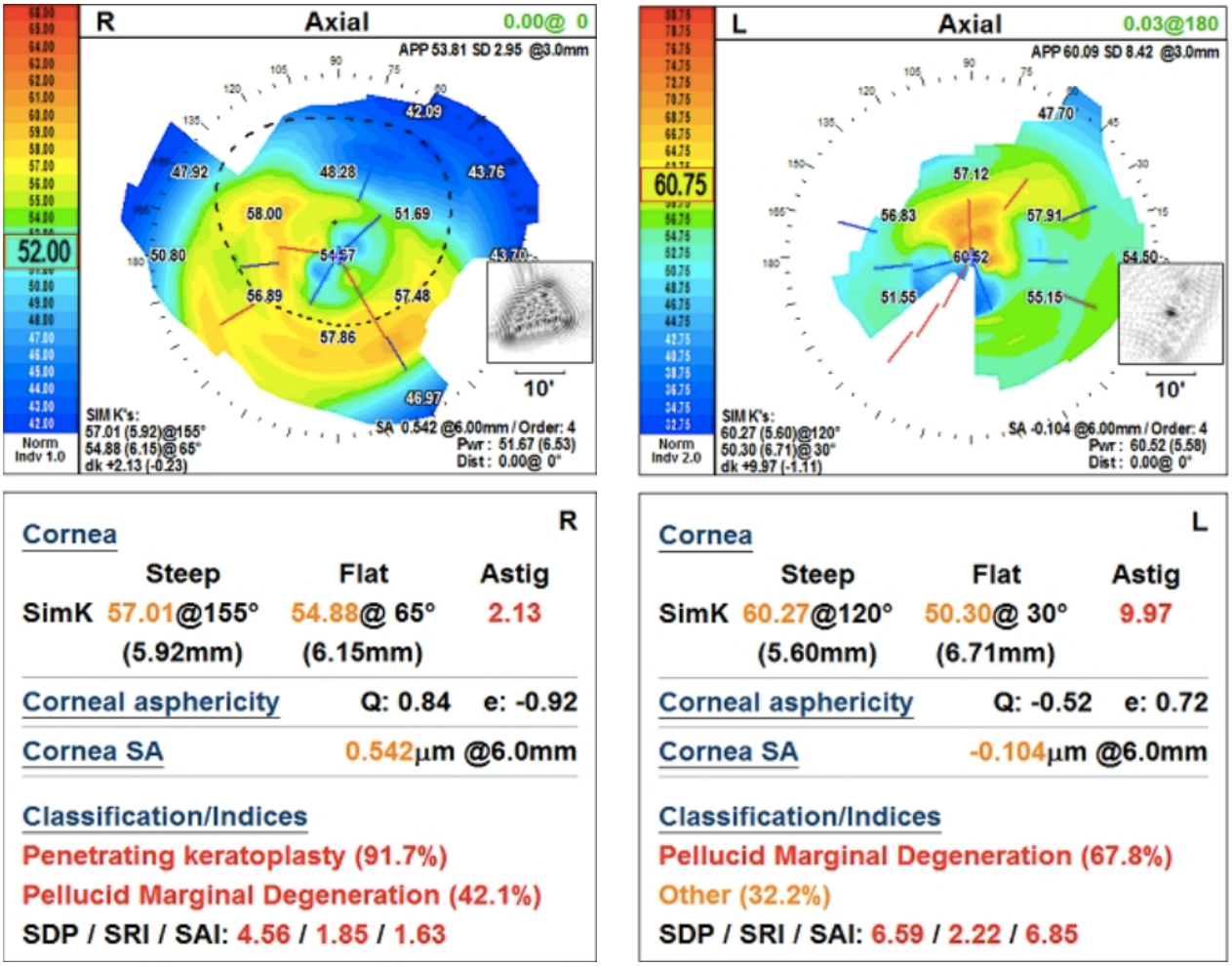

Figure 1. OPD® corneal topography of right and left eye from Nov 29, 2022. K_{max} 58.0 D OD and 60.52D OS with classic “crab claw” topographic pattern OD. OS topography quality compromised by extensive corneal scarring.

The cornea specialist the patient was referred to requested one final trial in scleral lenses prior to proceeding with deep anterior lamellar keratoplasty (DALK) or Penetrating Keratoplasty (PKP). The patient was then referred for a final diagnostic scleral lens fitting and told to discontinue wear of habitual RGP lenses due to excessive corneal touch.

Diagnostic Scleral Lens Fitting (03/17/2023)

In March 2023, the patient returned for diagnostic scleral lens fitting. The parameters of the diagnostic Ampleye® scleral lenses can be found in **Table 1**. Initial visual acuity was 20/30-2, 20/40+2. The right lens had 200 µm of clearance at apex, adequate limbal clearance, the sag indicator at 12 o'clock. The left lens had 200 µm of clearance at apex, adequate limbal clearance, the sag indicator at 11 o'clock. There was adequate alignment of the periphery with no vessel impingement and no obvious edge lift OU. Both lenses were centered and displayed no movement. The surface of each scleral lens had adequate wettability. An over-refraction of -0.50 -0.25 x 135 OD and -0.50 -1.00 x 030 resulted in visual acuities of 20/20 and 20/30+2.

Table 1: Diagnostic Ampleye® Lens Parameters.

OD	Parameter	OS
16.5mm	Diameter	16.5mm
42.00D/8.04mm	Base Curve	42.00D/8.04mm
4400	Sagittal Depth	4600
-4.00 DS	Power	-6.00 DS
Standard	PCZ	Standard
Standard	LCZ	Standard
+6 (150µm)	Toric Haptic	+6 (150µm)
Optimum Extra	Material	Optimum Extra
0.3	Center Thickness	0.3
Clear	Color	Clear
None	Coating	None

The following lenses were ordered as the initial pair:

OD 8.04/4600/-4.50
OS 8.04/4800/-6.75

Given that the diagnostic lenses had 200 µm of clearance at apex with less than 10 minutes of settling time, 200 µm of extra clearance was added to avoid corneal touch. Over-refraction was incorporated into each lens, and the lenses were ordered in Optimum Infinite® material to optimize oxygen transmission.

Scleral Lens Dispense (04/21/2023)

At scleral lens dispense, visual acuity was 20/20-2 OD, 20/50-1 OS with over refraction of plano OD, +2.75 -0.75 x 180 OS. Clearance was 400 μm at apex OU, with sag indicators at 5 o'clock and 7 o'clock in the right and left eye respectively. Periphery remained aligned OU.

Even though the OS lens was clearly overminused, lenses were deemed dispensable. Insertion and removal technique and scleral lens hygiene was reviewed. Initial lenses were sent home at this visit and a 1 week follow up was scheduled.

Scleral Lens Follow Up #1 (04/28/2023)

The patient returned for follow up 1 week later, reporting wearing the lenses for 16+ hours per day, with excellent comfort. He complained of clouding of vision OD>>OS after 2-3 hours of wear. He also expressed dissatisfaction with his reliance on reading glasses, as his habitual RGP lenses had been a multifocal design. At the time of examination, the patient had been wearing the lenses for 6 hours. Visual acuity was 20/20-2 OD, and 20/40 OS.

On examination, the right lens had 400μm of clearance over the apex, 0.25mm movement, and the sag indicator at 5 o'clock. The NaFl tap test was conducted and revealed instant and significant tear exchange at 6 & 12 o'clock, corresponding with the steep meridian of the lens. **Figure 2** demonstrates the tear exchange occurring along the steep meridian during this follow up.

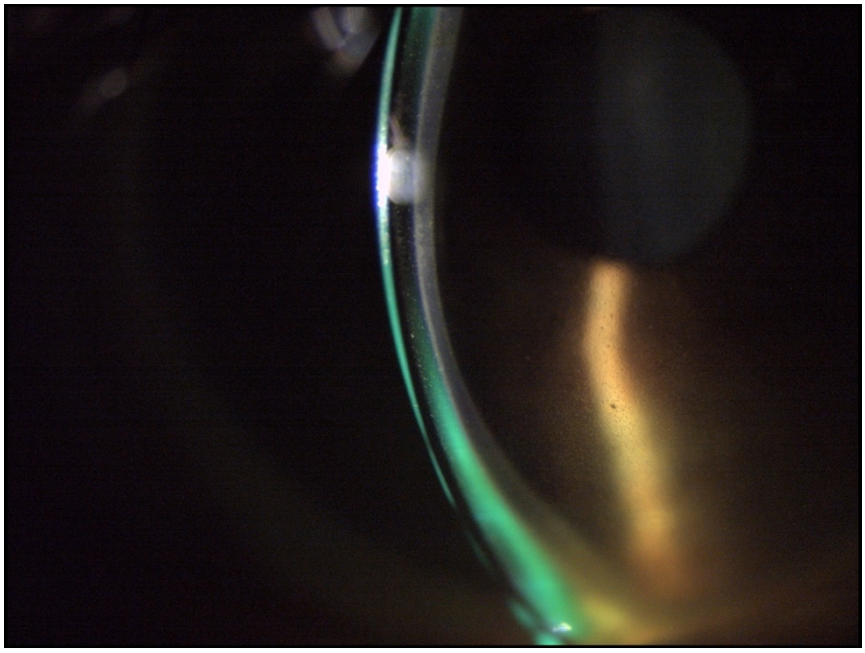

Figure 2. OD lens NaFl exchange along inferior steep meridian during a NaFl tap test, consistent with the patient's complaints of foggy vision after 2-3 hours of wear.

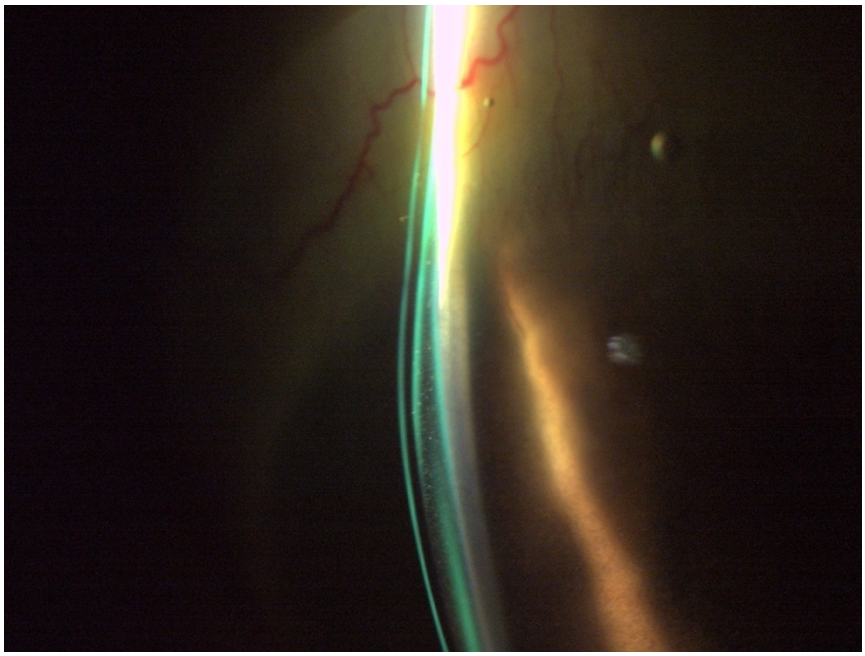

Figure 3. OD lens NaFl exchange along superior steep meridian during a NaFl tap test, consistent with the patient's complaints of foggy vision after 2-3 hours of wear.

The left lens had only 200µm of clearance over the apex, no movement, and the sag indicator at 7 o'clock. The NaFl tap test was conducted and revealed slight tear exchange at 6 & 12 o'clock, also corresponding with the steep meridian of the lens.

Overefraction was -0.25D OD, +3.00 -0.50 x 180 OS, consistent with the previous finding that the left lens had been overminused, with visual acuities of 20/20 and 20/25.

To address the tear exchange, the right lens periphery was tightened 50µm 360 degrees by changing the SLZ to +2. The toric haptic was increased to +8, adding an additional 50µm of edge tightening in the vertical meridian only. The overall central clearance was reduced by 75µm by changing the PCZ to -3. In the left eye, the toric haptic was increased to +8, adding an additional 50µm of edge tightening in the vertical meridian only to address the slight tear exchange. +2.75D was added to the power of the left lens.

It was decided that mini-monovision would be incorporated at the next visit to address the patient's complaints of poor vision at near.

Table 2: New Lens Parameters - Ampleye® Scleral Lenses with increased toricity

OD	Parameter	OS
16.5mm	Diameter	16.5mm
42.00D/8.04mm	Base Curve	42.00D/8.04mm
4600	Sagittal Depth	4800
-4.50 DS	Power	-4.00 DS
-3.00	PCZ	Standard
Standard	LCZ	Standard
+2.00	SLZ	Standard
+8	Toric Haptic	+8
Optimum Extra	Material	Optimum Extra
0.3	Center Thickness	0.3
Clear	Color	Blue
None	Coating	None

Follow-up #2 (05/26/2023)

The patient returned to clinic one month later, reporting improved vision and comfort with the new scleral lenses, with complete resolution of fogging of vision. He noted he was able to wear the lenses 16+ hours per day, without needing to remove lenses due to fogging. Visual acuity in the new scleral lenses was 20/20, 20/25 OD/OS. The patient continued to express frustration with his dependence on reading glasses at near.

Evaluation of the right lens revealed 325 μm of clearance over the apex after 6 hours of wear, with adequate limbal clearance, no movement, the sag indicator at 5 o'clock and an aligned periphery. Evaluation of the left lens revealed 200 μm of clearance over the apex after 6 hours of wear, with adequate limbal clearance, no movement, the sag indicator at 7 o'clock and an aligned periphery. There was slight tear exchange along the vertical meridian noted OD during NaFl tap test, however it was minimal, and the patient indicated he was no longer noticing any fogging in either eye. Over refraction was -0.25D OU, with no change in visual acuity. The lens surface was wetting adequately.

Eye dominance was tested, and the patient was determined to be OD dominant. After educating the patient on possible symptoms of monovision, including impaired depth perception, difficulty focusing and headaches, it was mutually decided to proceed with +1.75D monovision, OS lens as the near eye.

The following OS lens was ordered to incorporate +1.75D monovision:

OS 8.04/4800/-2.50

No other fitting parameters were changed at this time.

Follow-up #3 (06/09/2023)

The patient returned 2 weeks later for his final follow up visit. He reported excellent near vision, and denied symptoms of dizziness, impaired depth perception or difficulty focusing. He continued to be able to wear his lenses for 16+ hours a day comfortably without fogging.

Final BCVA was 20/20 OD, 20/25 OS, with a -0.25D/-1.75D overrefraction in the right and left eye respectively.

The lenses had 325 μm/200 μm of clearance over the apex after settling and aligned peripheral landings OU (aligned OS periphery seen in **Figure 4**).

The scleral lenses were then finalized. A note was sent to the referring cornea specialist indicating that DALK/PKP was not indicated given the success, and a follow up visit was scheduled for 1 year.

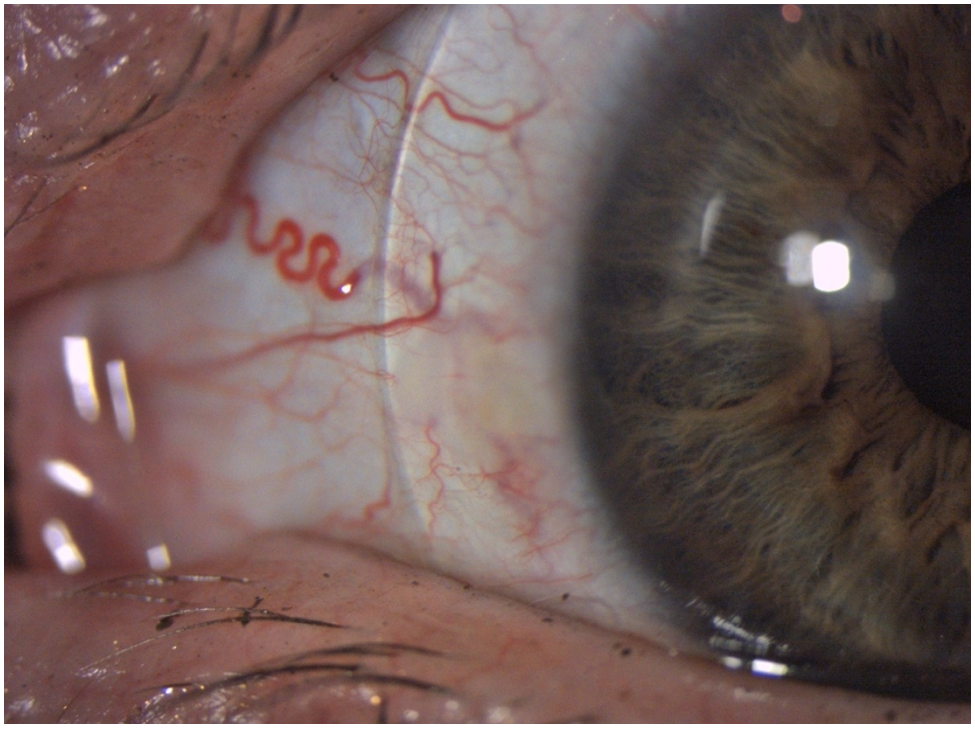

Figure 4. Aligned nasal periphery OS. Final lens.

Discussion

PMD is a non-inflammatory, non-hereditary corneal ectasia that is often misdiagnosed as keratoconus[1]. While PMD and keratoconus share many clinical features, there are important distinctions. Differentiating between the two ectasias is of potential clinical

importance since their distinguishing characteristics mean different optimal treatment and management.

In keratoconus, the central or paracentral cornea undergoes progressive thinning and steepening causing irregular astigmatism. The location of the apex of the cone can be central or inferior[5]. Keratoconus is generally well managed with RGP lenses, particularly in cases with central cones where the lens can “sit on” the cone and remain centered[6]. In PMD, the peripheral band of corneal thinning occurs in the inferior cornea between 4 to 8 o’clock[6]. The area of thinning is much larger than in keratoconus and can reach up to 20% of the cornea[7]. Unlike in keratoconus, the steepest protrusion in PMD is above the area of stromal thinning, creating high magnitude against the rule astigmatism[6]. Because the area of steepening is larger than that seen in keratoconus, it presents challenges for optimal contact lens fitting[6]. In PMD, a larger diameter lens is needed to achieve adequate vision and comfort[6]. Smaller RGP lenses are very unstable on a pellucid cone and almost always pose decentration issues[6]. It is also important to note that PMD does not present with an iron ring, cone, or apical scarring, meaning the scarring noted on anterior segment examination in this case was from the RGP lens rather than disease progression, further demonstrating the superiority of scleral lenses in the management of this case.

Another important distinction between KCN and PMD is how amenable they are to corneal transplantation. While KCN can be successfully managed by routine DALK/PKP, PMD transplantation is technically difficult and associated with poorer outcomes. Large diameter PK is the preferred technique for PMD but is associated with a higher rate of graft rejection due to 1) large differences in corneal thickness between the donor and host cornea, 2) progressive thinning of the corneal periphery after transplantation and 3) the larger diameter graft being near the limbal vasculature and stem cells[8]. In fact, Speaker et al reported a rejection rate of 100% with the use of large-diameter (>9.5 mm) penetrating grafts compared with 36% in smaller grafts (8.5/9 mm)[9]. “Tuck in” lamellar keratoplasty (TILK) has demonstrated better success with PMD transplantation[8]. However, there are many risks of lamellar transplantation procedures, including corneal perforation through the thinnest portion, wrinkles in the lamellar graft, graft dehiscence, and stromal rejection[8]. Given that the extreme thinness of the cornea in PMD makes transplantation difficult, and transplantation procedures often do not achieve optimum visual results, scleral lenses should be considered as first line treatment option in the management of PMD, superior to RGP lenses and attempted prior to cornea transplantation.

Conclusion

PMD and KCN are often grouped together but have several important distinctions, including the location of steepest protrusion and the percentage of cornea involved. Because of these distinguishing features, PMD is notoriously more difficult to fit in contact lenses and is associated with a higher rate of graft rejection compared with KCN[9]. As demonstrated in this report, scleral lenses should be considered superior to RGP lenses in the management of PMD and attempted prior to cornea transplantation.

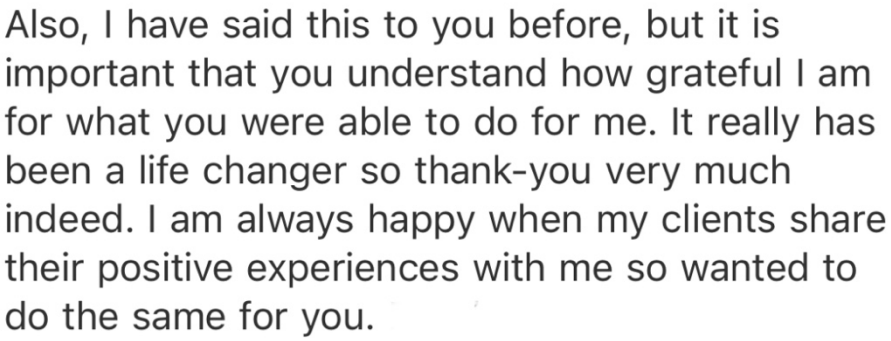

Figure 5. Email from PMD patient in case report at conclusion of scleral lens fitting :)

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