Pellucid Marginal Degeneration Challenges Overcome with Scleral Contact Lenses

A Case Report on Avoiding a Penetrating Keratoplasty

Nicholas Wolf, O.D.

-Abstract-

Introduction: Pellucid marginal degeneration is a largely bilateral corneal disease characterized by clear, circumlimbal peripheral thinning of the inferior cornea. This thinning causes outward bulging of the cornea resulting in high, irregular astigmatism, increased corneal aberrations, and decreased vision with spectacles or traditional soft contact lenses. **Case Report:** 61 year old Caucasian female presented for examination with a history of being diagnosed with keratoconus, but previous attempts with contact lenses had been unsuccessful. She was recently told by another eyecare provider that she no longer was legal to drive with glasses and that the next step would be a corneal transplant. Her goal of therapy was to retain her driver's license and hopefully achieve better vision, but with lenses that she was comfortable using. Upon testing she was diagnosed with pellucid marginal degeneration and fit successfully with scleral contact lenses. To this point, she continues to be more than satisfied with the comfort and vision that scleral lenses provide. **Discussion:** Pellucid marginal degeneration and keratoconus are often conflated; however, most literature and professionals believe they are two unique but similar diseases. This is more than a distinction without a difference as the two conditions can have very different clinical arcs which necessitate proper diagnosis to facilitate proper care. Many contact lens options exist to correct for pellucid marginal degeneration, but large diameter corneal RGPs as well as scleral lenses are most appropriate for moderate to severe corneal disease. As with any contact lens, hypoxia is always a concern, but doubly so with scleral contact lenses due to the deep tear reservoir these lenses create. Additionally, these lenses produce some complications unique to scleral lenses such as mid-day fogging, epithelial bogging, and conjunctival prolapse. If contact lenses fail to provide adequate vision or comfort, intrastromal ring segments are helpful to stabilize the cornea, and corneal transplants utilizing either full thickness (penetrating keratoplasty) or partial thickness (deep anterior lamellar keratoplasty) are proven effective. However, with the recent FDA approval of corneal crosslinking, more invasive surgeries and transplants should hopefully become far less common. **Conclusions:** Pellucid marginal degeneration is an uncommon cornea ectasia that can be differentiated from keratoconus through the history, corneal topography, and clinical presentation. Because of the often severe inferior corneal thinning and steepening, scleral contact lenses are an excellent choice. While small diameter RPGs and hybrids are possible with mild PMD, larger diameter RPGs and scleral contact lenses should be the first choice for patients with more significant disease.

Pellucid Marginal Degeneration Challenges Overcome with Scleral Contact Lenses

A Case Report on Avoiding a Penetrating Keratoplasty

Nicholas Wolf, O.D.

Introduction

Pellucid marginal degeneration is a largely bilateral corneal disease characterized by clear, circumlimbal, peripheral thinning of the inferior cornea. This thinning causes outward bulging of the adjacent cornea resulting in high, irregular astigmatism, increased corneal aberrations, and decreased vision with spectacles or traditional soft contact lenses. Oftentimes this condition is confused with keratoconus; however, most professionals view pellucid marginal degeneration as a unique entity with a unique clinical presentation and unique challenges when fitting medically necessary contact lenses.

The following case report showcases the management of a 61 year old female with moderate to advanced pellucid marginal degeneration looking for an alternative to undergoing a corneal transplant.

Case Report

History and Visual Needs

61 year old Caucasian female CM presented to our office for a second opinion on 09/06/2016. She reported good general health aside from hypertension (treated with enalapril) and sleep apnea. She had no known medical allergies. Family ocular history was positive for cataract removal on both parents.

CM entered this process knowing that she had *keratoconus* which was diagnosed approximately 20-25 years ago. However, it had been at least 5 years since her last eye exam and old records and testing could not be located. She was a full-time glasses wearer but had noticed her vision declining, particularly while driving at night and at work repairing jewelry. She recently failed her driver's license vision screening which prompted her to seek care. At that visit (several months before our exam), she was told by a doctor that she could not continue to drive with her current glasses. Given that new glasses wouldn't help due to her keratoconus, she was told she might be a good candidate for a corneal transplant. She further explained that several doctors, many years ago, had attempted to correct her vision with hard contact lenses, but the vision and comfort were not acceptable and she returned to glasses wear.

Her main goal was simply to avoid surgery and retain her driver's license to continue working and visiting her family and friends. Being a jeweler, she also needed to have the visual capacity to make sales, minor repairs, and judge the clarity/quality of gemstones.

Initial Exam and Testing

At her initial visit, we recorded vision at 20/400 in each eye without correction and 20/50 in each eye with her current glasses (below).

Habitual Rx

OD:	+1.00 – 4.50 x 083	ADD: +2.00
OS:	+1.50 – 3.50 x 092	ADD: +2.00

Pupils, ocular motility, confrontation fields, and ocular alignment were normal. Intraocular pressure was obtained at 11:45 am and measured 16 mmHg in both eyes with iCare® tonometry. Retinoscopy showed a characteristic scissor reflex. Manifest refraction was performed and very difficult for both patient and practitioner.

Manifest Refraction

OD:	+2.25 – 8.50 x 080	BVA: 20/60
OS:	+1.50 – 6.50 x 095	BVA: 20/40-2

Anterior segment evaluation showed mild blepharitis with lash scurf as well as eyelid margin telangiectasia. Epilation and light microscopy did not reveal demodesis. There was mild conjunctivochalasis in both eyes, but an otherwise clear and quiet anterior surface. Her corneas were also clear in both eyes with no scarring, striae, or Fleischer's ring noted. Both corneas, however, had significant inferior bulging and

sagging as evident with slit lamp and gross examination although no clear Munson sign was present.

CM was dilated with 1 drop of Paremyd® in each eye (1% hydroxyamphetamine hydrobromide/ 0.25% tropicamide ophth. sln.). Internal evaluation showed grade 1+ nuclear sclerotic cataracts in both eyes. Optic nerves were healthy and well perfused with a cup to disk ratio of 0.3 in each eye. The macula in both eyes looked normal with slit lamp and OCT testing, and the peripheral retinas in both eyes were unremarkable.

Corneal topography was ordered and performed at this visit with the Medmont™ E300 topographer. The results (figures 1 and 2) revealed significant peripheral, inferior steepening in the classic *crab-claw*, *butterfly*, or *kissing-doves* presentation. This was present in both eyes, but more severe in the right eye than the left. Steep keratometry readings in the right and left eyes were 53.8 D and 51.5 D, respectively. All screening criteria for ectatic disease built into the Medmont™ Studio software were also flagged as abnormal. Via topography, the HVID was measured at 11.6 mm in the right eye and 11.7 mm in the left eye. Testing was reliable and of good quality.

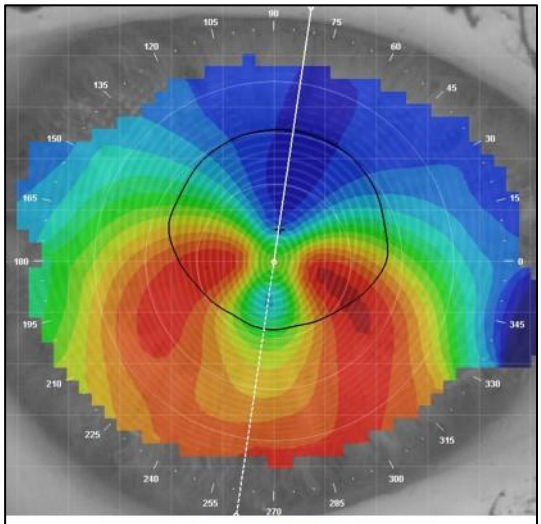

Figure 1 – OD Topography

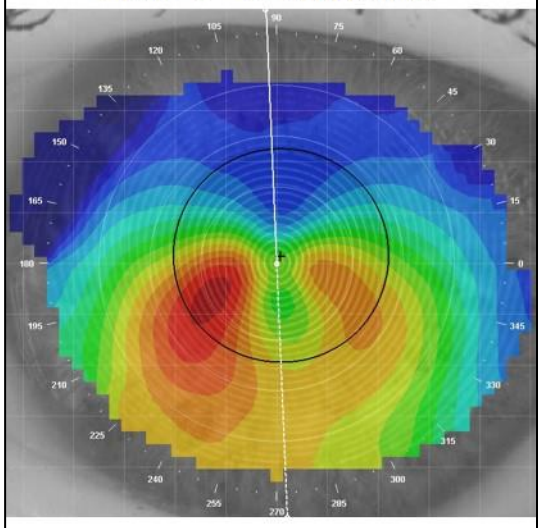

Figure 2 – OS Topography

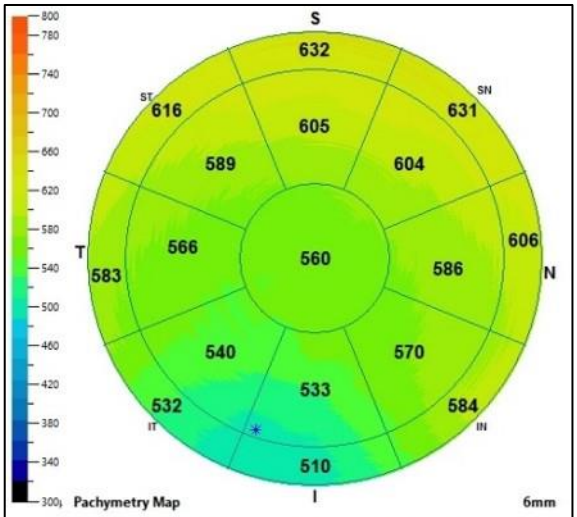

Figure 3 – OD Baseline Pachy Map

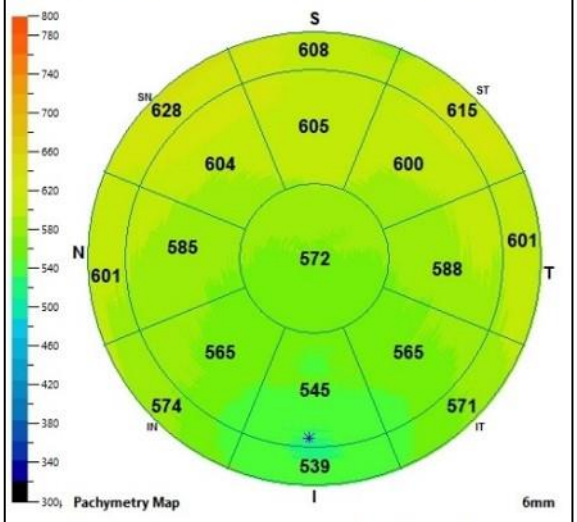

Figure 4 – OS Baseline Pachy Map

Pachymetry mapping with anterior segment optical coherence tomography (iVue® by Optovue™) was also ordered and performed (figures 3 and 4). The pachymetry map shows some general thinning inferior in both eyes (OD>OS) but the thinning was not substantial. The baseline pachymetry map will allow us to monitor subtle, hypoxia-related edema from scleral lens wear and to make fitting revisions if needed.

Differential and Final Diagnosis

With the topography showing clear ectasia without a history of corneal surgery or contact lens wear, the differential diagnoses on this patient was rather small. Her previous diagnosis of keratoconus was possible, but the lack of any corneal signs (striae, scarring, Munson's sign, or Fleischer's ring) is uncommon for keratoconus this advanced. In addition, the classic *crab-claw/kissing-doves/butterfly* topography is not pathognomonic for, but highly suggestive of, pellucid marginal degeneration[1]. Due to the combination of clinical signs and the topography, CM was diagnosed with moderate to advanced pellucid marginal degeneration in both eyes, more severe in the right eye than the left eye.

Lens Selection and Initial Fitting

As glasses hadn't been successful, we reviewed either a surgical or a medically necessary contact lens approach to reach the patient's treatment goals. As had been recommended prior, a penetrating keratoplasty, or more conservatively ring implants, would likely have helped. However, with her optically clear, minimally thinned corneas, coupled with an otherwise healthy eye, contact lenses were recommended as the next logical step to return functional vision, with surgery as a last resort.

All lens options where discussed with CM, including specialty soft lenses, small diameter hard lenses, hybrid contact lenses, as well as semi-scleral and full scleral contact lenses. The patient had already tried, and was intolerant to, small diameter GP lenses. Additionally, the far peripheral inferior steepening made it unlikely that a hybrid, or even a large diameter hard lens, would properly center on the corneal topography. In the end, we decided on a full scleral to allow clearance of the inferior bulging and better centration.

We decided on the Europa Scleral™ contact lens by Visionary Optics©. While this lens is not a true oblate design, the reverse geometry in the mid-peripheral curve was deemed likely sufficient. The initial lens was selected based on the Medmont™ Studio calculated sagittal depth from the composite testing at a chord of 16 mm with an additional 400 µm added for clearance. One drop of anesthetic was administered prior to lens insertion to facilitate initial application. Trial lenses were filled with sterile, non-preserved inhalation solution (Addipak®) along with sodium fluorescein dye and inserted by a staff member with a classic DMV™ inserter. In this instance, we started with the same diagnostic lens (below).

Europa: -2.50 DS, 7.18 mm, 16 mm diameter, Standard landing, Standard peripheral curves

After 5-10 minutes of settling, the diagnostic lens was evaluated for fit, vault, and over-refraction utilizing both slit lamp and AS-OCT (iVue® by Optovue™).

Right Eye – Figure 5

The diagnostic lens in the right eye showed good centration and minimal movement. The landing cleared the limbus as shown on anterior segment OCT and with analyzing the sodium fluorescein pattern with a wratten filter at the slit lamp. Mild inferior conjunctival compression was noted, but an overall appropriate scleral landing and apposition. Initial sagittal depth was 349 µm centrally with no cornea touch inferiorly. This was deemed in the acceptable range of 300-400 µm to allow for settling after initial dispensing. Sphero-cylinder over-fraction was -1.00 DS with best corrected visual acuity of 20/20.

Left Eye – Figure 6

The same diagnostic lens in the left eye also exhibited proper centration and movement. Proper limbal clearance was shown with OCT and fluorescein. In the left eye, there was significant conjunctival compression and blanching supra-temporally (between 1-2 o'clock), and infero-nasally (between 7-8 o'clock). The patient noted some lens awareness and mild fogging after just 15 minutes of wear. The fogging is visible on the AS-OCT. Initial sagittal depth was 380 µm centrally (again within the acceptable range) with closer, but no corneal touch inferiorly at the apical 'belly' of the cornea. Sphero-cylinder overfraction was -0.75 – 0.75 x 165 with best corrected visual acuity of 20/25.

The following lenses were ordered and the patient was scheduled for a scleral lens training and dispense in two weeks.

Europa Scleral™ Lenses – Boston XO2™
OD: BC 7.18, Sag. Depth 4751 µm, Diameter 16.0 mm, Power: -3.50, Standard Landing Zone, Standard Peripheral Curve
OS: BC 7.18, Sag. Depth 4751 µm, Diameter 16.0 mm, Power: -3.50, Standard Landing Zone, 2 D Scleral Toric Haptic

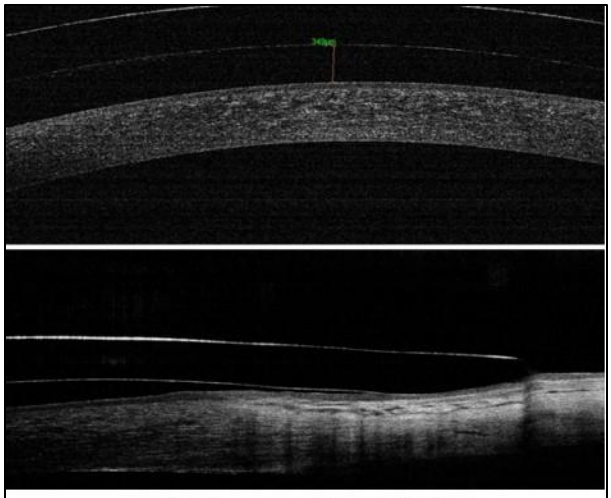

Figure 5 – OD Initial Fitting

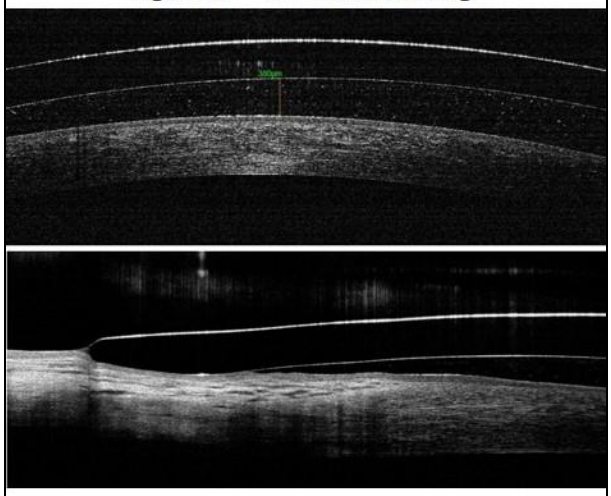

Figure 6 – OS Initial Fitting

Dispense, Initial Evaluation, and Training

CM was seen for dispensing and I&R training on 09/20/2016. At this visit she was again anesthetized with topical anesthetic and a staff member used inhalation solution with sodium fluorescence for initial insertion of the lenses. The initially dispensed lenses were allowed to sit for 15 minutes before the evaluation was performed.

Slit lamp examination revealed good centration of both lenses with minimal movement. Limbal clearance was present 360 degrees and no corneal touch was noted over the inferior corneas. Mild compression but little blanching was noted inferiorly in both eyes, but deemed acceptable. The toric haptic on the left scleral lens nicely resolved the heavy 2 and 8 o'clock compression noted on the initial fitting. Sagittal depth was confirmed acceptable with AS-OCT at 350 µm in the right eye and 370 µm in the left eye. Over-refraction with the scleral lenses was plano with best corrected vision of 20/20 in the right eye and -0.25 – 0.75 x 120 with best corrected vision of 20/15- in the left eye.

CM noted immediate improvement in vision and was pleasantly surprised with the comfort of her new lenses. She was educated on the insertion, removal, and care of her scleral contact lenses. Clear Care® hydrogen peroxide based cleaning solution was recommended for daily cleaning as well as an enzymatic cleaner for as needed use. For insertion solution, CM was prescribed 0.9% NaCl inhalation solution (Addipak®) for off-label use. In addition to samples of these solutions, the patient was also supplied with a classic DMV™ for insertion, a 45 degree DMV™ for removal, detailed written instructions, as well as our after-hours contact information. She was also instructed to purchase over-the-counter reading glasses for her near task needs.

Finally, given the known association with ocular surface disease and scleral fogging, CM was instructed to perform lid cleanings twice daily with Ocusoft™ foaming eyelid cleanser and begin a fish oil regimen. CM left wearing her new lenses and follow up was set for 2 weeks.

One Month Follow Up

CM returned to our office about 1 month after initial dispensing on 10/25/2016. She reported excellent results with her scleral contact lenses, was proficient at insertion and removal, and was thrilled overall with both the vision and comfort. She reported wearing them daily for about 12-14 hours and at evaluation she had been wearing her lenses for 10 hours. On occasion, she did report mild to moderate fogging in both eyes, but that was easily corrected with removal and reinsertion and did not prevent her from daily wear.

Vision was measured at 20/20 in the right eye and 20/25- in the left. Over refraction was again Plano in the right eye and -0.25 – 0.50 x 125 in the left eye which improved the vision in that eye to 20/20. Both lenses exhibited good positioning and minimal movement with equal scleral apposition and minimal compression. Fluorescein dye was applied over top of the contact lenses and then evaluated. No large veins of fluorescein bleeding were noted. After some perfusion under the lens, proper limbal clearance and no peripheral corneal touch was confirmed with a wratten filter. Sagittal depth was measured with AS-OCT at 314 µm in the right eye and 411 µm in the left eye. Peripheral clearance was also evaluated with AS-OCT and showed plenty of clearance. This confirmed that the reverse

geometry of the Europa Scleral™ lens peripheral curve was adequate for this case and assuaged the fear of needing a true oblate design.

The lenses were then removed and no corneal staining or epithelial bogging was noted nor was there any rebound conjunctival redness.

To combat the mild fogging complaint, and facilitate better oxygen bio-availability we decreased the sagittal depth in both eyes to approach the recommended 250 µm vault and had her continue her lid hygiene. Additionally, a slight prescription revision was made for the left lens. Here were the parameters ordered.

Europa Scleral™ Lenses – Boston XO2™
OD: BC 7.26, Sag. Depth 4702 µm, Diameter 16.0 mm, Power: -3.00, Standard Landing Zone, Standard Peripheral Curve
OS: BC 7.38, Sag. Depth 4630 µm, Diameter 16.0 mm, Power: -2.75, Standard Landing Zone, 2D Scleral Toric Haptic

These lenses were ordered and our staff did a brief AS-OCT 1 week later to confirm the goal sagittal depth at lens dispense. CM was then scheduled for a 3 month follow up.

Three Month Follow Up

CM presented back to our office as instructed on 01/26/2017. She continued to report great vision and comfort and also noted that her issues with fogging had decreased, but on occasion she still needed to remove and reinsert her lenses. Vision was 20/20 in both the right and left eyes and sagittal depth was measured with AS-OCT at 235 µm in the right eye and 260 µm in the left eye. Corneal health was stable and no additional revisions were deemed necessary. CM was scheduled for evaluation in 6 months for repeat topography to monitor for progression as well as a pachymetry map to assess hypoxia-related corneal swelling.

6 Month Follow Up

CM returned for her six month follow up on 08/29/2017. One year out from starting scleral lens wear and she was still happy with the vision and comfort. Lens evaluation was similar to testing six month prior and slit lamp evaluation showed healthy corneas without sequelae. Repeated corneal topography confirmed no progression in the pellucid marginal degeneration. AS-OCT pachymetry serial mapping of the central cornea showed similar values to initial evaluation. The difference mapping (figures 7 and 8) suggests no subclinical hypoxia-related corneal edema and confirms adequate central corneal oxygen bio-availability. Given the stable testing and lens appearance, a backup pair of scleral lenses was ordered and the patient was scheduled to follow up at our office yearly.

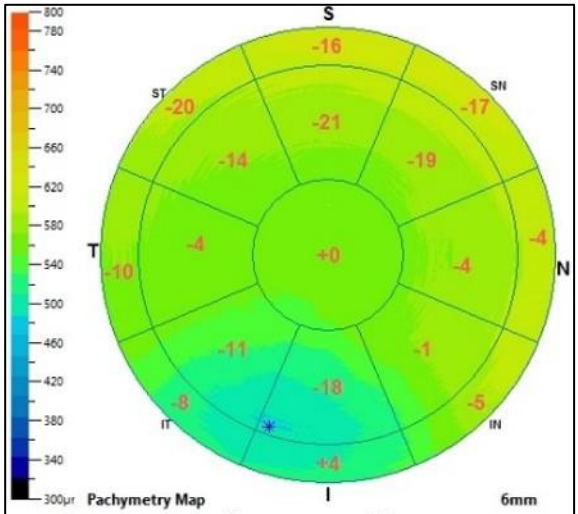

Figure 7 – Pachymetry Difference OD

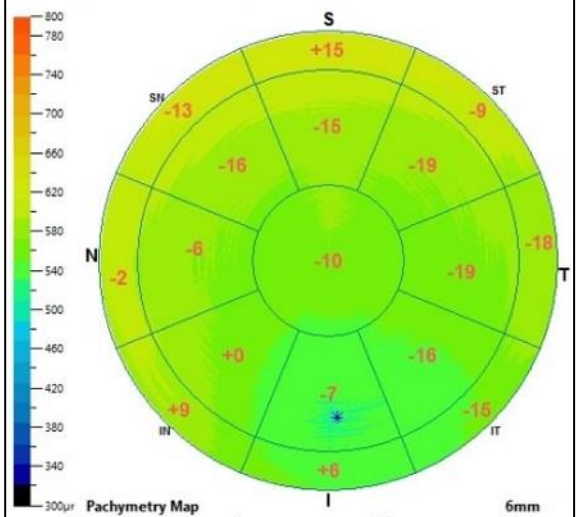

Figure 8 – Pachymetry Difference OS

Discussion

Pellucid Marginal Degeneration Versus Keratoconus

Pellucid marginal degeneration (henceforth PMD) is a corneal ectasia oftentimes seen as, or confused with, the more common ectasia, Keratoconus (henceforth KCN). There are unique characteristics between the two that distinguish the conditions. This is

not just a distinction without a difference as proper diagnosis is important to understanding the likely course of the disease, as well as choosing the best treatment route, whether surgically or with medically necessary contact lenses.

There are many similarities between the conditions. Both PMD and KCN cause non-inflammatory breakdown of the collagen matrix of the cornea that leads to thinning and subsequent bulging of the cornea. Both conditions are usually bilateral although cases of unilaterality are not rare[2,3] and both conditions are linked to allergic and vigorous eye rubbing[2,4]. Additionally, both conditions have a mild male predilection[2,5,11] but have no other ethnic, racial, socioeconomically, or geographical predisposition[6].

KCN, however, is relatively common affecting from 1:2000 to as high as 1:375 individuals[5]. PMD on the other hand is quite rare, no concrete incidence or prevalence data exists. This however, may be somewhat misleading, as PMD is often misdiagnosed at KCN or the conditions may concomitantly occur[9].

Initial diagnosis of KCN usually occurs at the end of the first and into second decade of life and worsens for a decade or two before stabilizing[5,8]. Often, earlier diagnosis coincides with a more severe course of the disease[7,8]. By contrast, PMD typically develops in the 2nd – 5th decade of life. In studies, the spread of the average diagnosis age was 34 to 48 years old[10,11]. This is followed by slow but methodical thinning and progression before eventually stabilizing.

Pathologically, the main difference between PMD and KCN is where the thinning occurs. KCN begins as an isolated area of corneal thinning, typically slightly inferior or infero-temporal. By contrast, PMD causes thinning in a circumlimbal band between 4 and 8 o'clock in the inferior cornea, and normal cornea centrally as well as a 1-2 mm band of normal cornea adjacent to the limbus[6]. Corneal thinning in this presentation results in the classic topographical *crab-claw*, *butterfly*, or *kissing doves* appearance. However, severe KCN can also result in a similar corneal topography so this presentation is not pathognomonic for PMD[1], just highly suggestive.

The term *Pellucid* is defined as *translucently clear*, and this may be a simpler method than topography at properly diagnosing this condition clinically. KCN has a specific location of greatest thinning accompanied by the expected apical scarring, endothelial striae, and Fleischer's ring. While scarring and even hydrops can occur with PMD, the classic KCN corneal signs are not typically noted with PMD[10]. Clinically, PMD ectasia causes an overall sagging appearance (termed *beer belly*) as opposed to KCN where the cornea takes on a conical shape and results in Munson sign, which is also absent in PMD.

Refractive Correction for PMD

Thinning from PMD typically occurs inferiorly, but there are case reports of it occurring elsewhere on the cornea[12,13]. As the inferior cornea thins, the normal cornea above bulges and results in large amounts of irregular, against-the-rule astigmatism. Initially, glasses and traditional contact lenses can oftentimes provide satisfactory vision. However, as the disease progresses, more irregular astigmatism develops along with larger amounts of higher order aberrations eventually reaching a point that spectacles can no longer compensate. Medically necessary contact lenses for PMD often will include small and large diameter RGP lenses, hybrid lenses, and scleral lenses.

Hard lenses have been, and continue to be, a mainstay in the treatment for patients with corneal ectasia. They help because the rigid nature of the contact lens holds its shape and forms a new, smooth, refracting surface. Tears fill in the area between the lens and the diseased cornea resulting in decreased higher order aberrations and improved vision. When fitting a cornea with PMD, the main problem with small diameter cornea RGP lenses is centration. Ever increasing amounts of irregular, against-the-rule astigmatism causes the lens to decenter downward with inferior lift. This leads to several possible complications. First, the decentering lens also cause the optic zone to drop leading to halos, glare, and visual fluctuations. Secondly, inferior edge lift leads to increased lens awareness, discomfort, and could causes the lens to decenter off the cornea or fall out completely. Thirdly, fitting a corneal RGP on such irregular topography will cause excessive rubbing on the areas of most aggressive touch which results in at the least lens awareness and surface staining, and at the worst corneal scarring[14]. Some of these centering challenges can be overcome by moving to larger diameter corneal RGP lenses with a reverse geometry design to compensate for the peripheral corneal topography[15].

Hybrid lenses offer additional benefits over small diameter RGPs due to the soft lens skirt. These include improved comfort with less lens-corneal interaction and decreased inferior decentration. While these options have a place in PMD care, for moderate to advanced disease, the literature and our own clinical experience suggest scleral contact lenses are a better choice[16,17].

Scleral contact lenses work by vaulting completely over the compromised cornea and resting only on the sclera with tears and saline accounting for the space between. This offers several advantages for severe ectasia and PMD in particular. First, a lens resting on the sclera is more comfortable compared to a lens resting on the cornea, as the sclera is a much less sensitive tissue. Secondly, the topographical severity is not as critical, as the corneal-contact lens interface does not exist. This vastly improves centration by landing on the normal (by contrast) curvature of the sclera. Additionally, corneal touch, discomfort, and mechanical scarring are reduced because the lens-cornea interface is absent. This fact is especially important with the broader area of thinning found in PMD patients, which makes traditional RGP fitting difficult.

Scleral contact lenses are not, however, a panacea, and there are some potential pitfalls that are either unique to, or more a concern with, scleral contact lenses. Three unique complications with scleral lenses are epithelial bogging, mid-day fogging, and conjunctival prolapse[18].

Epithelial bogging presents as non-staining but very irregular corneal epithelium. This is thought to occur from constant saturation from the fluid reservoir, similar to "pruned fingers" after swimming. While visually striking, this complication is transient and disappears within 1-2 months after starting routine scleral lens wear[31,32].

Mid-day fogging is a common complication and is visually significant in about 30% of scleral contact lens wearers. Since there is minimal turnover of the fluid reservoir, mucus, debris, and epithelial cells build up under the lens and at high concentrations cause blurring and fogging of vision. This occurs after several hours of wear and requires removal and reinsertion of the lens with fresh solution. The exact etiology of this is not known, and a multipronged approach to decrease fogging is best.

1. **Ocular Surface Disease:** It is imperative to treat any ocular surface disease, especially lid disease like blepharitis, preferably before scleral lenses are prescribed.
2. **Scleral Landing:** Make sure the landing zone of the lens has good apposition 360 degrees. Sodium fluorescence applied over top of the lens with a wratten filter is useful to isolate any areas of fluorescein bleeding that would suggest a toric haptic is needed for better alignment.
3. **Sagittal Depth:** If applicable, a lower sagittal depth also decreases the volume of clouded tears and reduces the symptoms.
4. **Lens Size:** A smaller diameter scleral lens will cause less mechanical stress of the conjunctiva and trap lens mucin-secreting goblet cells under the lens.

Even with an optimally fit scleral lens, fogging may still occur but typically improves over time, with most patients noting far less fogging after the first couple months of wear. For added protection against fogging, NPATs can be used to completely or partially fill the scleral bowl before insertion. Non-preserved Refresh Optive® and Systane Ultra® work well and Refresh Celluvisc® could also be used for severe fogging cases.

Conjunctival prolapse or conjunctival hooding occurs when the suction forces of the scleral lens, in conjunction with the pressure of the blink, pull loose conjunctiva up into the bowl of the scleral lens. This usually occurs inferiorly and is secondary to the slight inferior displacement of the scleral lens causing increased limbal clearance. This is largely considered a benign finding unless synechia forms between the cornea and conjunctiva or neovascularization is noted. Nonetheless, it is best practice to avoid this complication by decreasing the limbal clearance to reduce the space for prolapse to occur.

While there are some unique challenges with scleral contact lenses, one important factor to keep in mind with all lenses, including scleral lenses, is hypoxia. This is doubly true for scleral lenses as oxygen must travel not only through the RGP material, but then diffuse through the tear lens to supply oxygen to the cornea. Research has shown that in order for corneal tissue to avoid edema, the minimum Dk/t required for the central cornea is 24[19]. Increasingly higher vaults are often required with severe or peripheral ectasia, including cases of PMD. With these deeper sagittal depths, it is useful to calculate what oxygen bioavailability exists at the corneal surface. The calculation for central Dk/t is as follows[21]:

Dk/t = 1/[(Thickness of Lens/Dk of Lens Material) + (Thickness of Post Lens Tear Layer/Dk of Tears)]

In the case of CM, the central thickness of her Europa Scleral™ lenses was 430 µm, the material used was Boston XO2™ with a Dk of 141, and the final sagittal depths readings were 235 µm and 260 µm in the right and left eye respectively. The only value not taken from the fitting process is the Dk of the tears which is 80[20]. Our calculations are as follows:

OD: Dk/t = 1 / [(430/141) + (235/80)] = 16.7
OS: Dk/t = 1 / [(430/141) + (260/80)] = 15.8

As shown, our patient does not meet the Dk/t of 24 to avoid hypoxia and resultant corneal edema. This is quite common when fitting these lenses to the scleral lens manufacturer's fitting recommendations. Furthermore, both studies and theoretical models have shown that mild cornea swelling from hypoxia does occur commonly[21,22]. In practice, however, severe corneal edema and other sequelae from hypoxia are thankfully uncommon and corneal neovascularization often regresses when scleral lenses are employed. Nonetheless, it is good practice to maximize the corneal oxygen. This is accomplished by fitting the highest Dk materials, with low sagittal depths (200-250 μm), and thinner lens thicknesses, as well as routinely monitoring scleral lens wearers for hypoxia and edema. While hypoxia unquestionably exists with scleral contact lens wear, the long-term effects of this mild but chronic oxygen deprivation is not known and fitters should be cognizant of potential complications.

Surgical Options for PMD

In studies, 88% of PMD patients achieve acceptable vision and quality of life with glasses and medically necessary contact lenses[23]. However, there are several surgical options that can be useful to either stop the progression of PMD or return more useful vision secondary to severe thinning, scarring, or contact lens failure.

The recent FDA approval of corneal crosslinking (CXL) has greatly expanded patient access to a straightforward procedure that has a proven track record of arresting the progression of corneal ectasia. CXL for KCN has been heavily studied and halts the progression in as high as 93% of cases[24]. PMD has not been as extensively studied, but several publications show that this condition does benefit by arresting the corneal steepening and in some cases causing beneficial corneal flattening[25]. This procedure involves epithelial removal and saturation with Riboflavin (vitamin B2) followed by exposure to a precise wavelength of ultraviolet light (365 nm). This process causes additional bonds to form between the corneal collagen fibers resulting in the strengthening and increased hysteresis of the cornea[26].

Another useful procedure to shore up the structural integrity with PMD is through the use of intrastromal segmented ring implants. With this procedure, a femtosecond laser is used to cut pockets around the area of ectasia. Small semi-circular plastic pieces are inserted into these pockets. The rigid plastic pieces help to normalize the corneal curvature to reduce irregular astigmatism and aberrations. While this procedure is better known for KCN, ring implants have also been proven quite effective in normalizing keratometry readings and improving spectacle BCVA in PMD patients[27].

PMD can reach the level where contact lenses and more conservative treatments are not effective. At that point, corneal transplants are very successful to restore useable vision. There are two types of transplantation that are useful for PMD: penetrating keratoplasty (PK) and deep anterior lamellar keratoplasty (DALK). PK is a full-thickness transplant that has been proven successful in ectasia patients including those with PMD[28]. Due to the inferior displacement of the ectasia, surgeons often opt for an inferiorly displaced graft or even a large diameter penetrating keratoplasty (LDPK)[29]. The chief concerns with PK is the risk of infection and potential for graft rejection. DALK is a newer surgery that sought to decrease the amount of rejection by transplanting down to, but preserving Descemet's membrane and the endothelium. This surgery maintains the closed system of the eye which decreases infection postoperatively. Additionally, retaining an intact endothelium lessens the chance of rejection, all while creating visual outcomes that are comparable to a PK[30].

Conclusions

Pellucid marginal degeneration is an uncommon cornea ectasia that can be differentiated from keratoconus by the history, topography, and clinical presentation. This distinction is important as these two very similar conditions can have a very different clinical arc. Due to the often severe inferior corneal thinning and resultant steepening with PMD, scleral contact lenses are an excellent choice. While small diameter RPGs and hybrids are an option with milder PMD, larger diameter RPGs and scleral lenses should be the first choice for patients with more significant disease.

CM was an interesting case in that her first successful contact lens experience was with a scleral contact lens at the age of 61 and while correctly diagnosed with an ectasia, PMD was the more appropriate diagnosis and better matches her clinical presentation. Her continued success, both clinically and in her day to day life, underscores that scleral contact lenses are an effective and user-friendly lens modality for pellucid marginal degeneration patients.

References

1. Lee BW, Jurkunas UV, et al. “Ectatic disorders associated with a claw-shaped pattern on corneal topography.” AM J Ophthmol. 2007 Jul;144(1):154-156.
2. Shimazaki J, Maeda N, Hieda O, et al. “National survey of pellucid marginal corneal degeneration in Japan.” Jpn J Ophthalmol. 2016 Sep;60(5):341-8.
3. Imbornoni LM, Padmanabhan P, Belin MW, Deepa M. “Long-Term Tomographic Evaluation of Unilateral Keratoconus.” Cornea. 2017 Jul 24
4. Gordon-Shaag A, Millodot M, Kaiserman I, et al. “Risk factors for keratoconus in Israel: a case-control study.” Ophthalmic Physiol Opt. 2015
5. Godefrooij DA, de Wit GA, Uiterwaal CS, Imhof SM, Wisse RP. “Age-specific Incidence and Prevalence of Keratoconus: A Nationwide Registration Study.” Am J Ophthalmol. 2017 Mar;175:169-172.
6. Junabhai A, et al. “Pellucid corneal marginal degeneration: A review.” Contact Lens Anterior Eye (2011),dol:101016/j.ciae.2010.11.007
7. Barr JT, Wilson BS, Gordon MO, et al. “Estimation of the incidence and factors predictive of corneal scarring in the Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study. Cornea 2006;25:16-25.
8. Romero-Jiménez M[1], Santodomingo-Rubido J, Wolffsohn JS. “Keratoconus: a review.” Cont Lens Anterior Eye. 2010 Aug;33(4):157-66.
9. Kayazawa F, Nishimura K, Kodama Y, Tsuji T, Itoi M. “Keratoconus with pellucid marginal corneal degeneration.” Arch Ophthalmol. 1984;102(6):895–896.
10. Krachmer JH, Feder RS, Belin MW. “Keratoconus and related non inflammatory corneal thinning disorders.” Surv Opthalmol 1984:28;293-322
11. Tzelikis P, Cohen EJ, Rapuano C, Hammersmith K, Labson PR. “Management of pellucid marginal corneal degeneration.” Cornea 2005;24:555-60
12. Puy P, Stoica BT, Alejandre N, Toledano N. “Temporal pellucid marginal degeneration displaying high "with-the-rule" astigmatism.” Can J Ophthalmol. 2013 Dec;48(6):e142-4.
13. Taglia DP, Sugar J. “Superior pellucid marginal corneal degeneration with hydrops.” Arch Ophthalmol. 1997;115:274-5.
14. Zadnik K, Barr JT, Steger-May K, et al. “Comparison of lat and steep rigid contact lens fitting methods in keratoconus.” Optom Vis Sci. 2005 Dec;82(12):1014-21.
15. Ozbek Z, Cohen EJ. “Use of intralimbal rigid gas-permeable lenses for pellucid marginal degeneration, keratoconus, and after penetrating keratoplasty.” Eye Contact Lens. 2006 Jan;32(1):33-6.
16. Rathi VM, Dumpati S, Mandathara PS, Taneja MM, Sangwan VS. “Scleral contact lenses in the management of pellucid marginal degeneration.” Cont Lens Anterior Eye. 2016 Jun;39(3):217-20.
17. Asena L, Altınörs DD. “Clinical outcomes of scleral Misa lenses for visual rehabilitation in patients with pellucid marginal degeneration.” Cont Lens Anterior Eye. 2016 Dec;39(6):420-424.
18. Walker MK, Bergmanson JP, Miller WL, Marsack JD, Johnson LA. “Complications and fitting challenges associated with scleral contact lenses: A review.” Cont Lens Anterior Eye. 2016 Apr;39(2):88-96.
19. Holden BA, Mertz GW. “Critical oxygen levels to avoid corneal edema for daily and extended wear contact lenses.” Invest Ophthalmol Vis Sci. 1984 Oct;25(10):1161-7.
20. Benjamin WJ. “Oxygen transport through contact lenses.” In: Guillon M, Ruben M., editors. Contact lens practice. Chapman Hall Medical Publishers; 1994: p 47-69.
21. Michaud L, van der Worp E, Brazeau D, Warde R, Giasson CJ. “Predicting estimates of oxygen transmissibility for scleral lenses.” Cont Lens Anterior Eye. 2012 Dec;35(6):266-71.
22. Vincent SJ, Alonso-Caneiro D, Collins MJ, Beanland A, Lam L, et al. “Hypoxic Corneal Changes following Eight Hours of ScleralContact Lens Wear.” Optom Vis Sci. 2016 Mar;93(3):293-9.
23. Kompella VF, Assuri MK, Rao GN. “Management of pellucid marginal corneal degeneration with rigid gas permeable contact lenses.” CLAOJ 2002;28:140-5
24. Shalchi Z, Wang X, Nanavaty MA. “Safety and efficacy of epithelium removal and transepithelial corneal collagen crosslinking for keratoconus.” Eye (Lond). 2015 Jan;29(1):15-29.
25. Mamoosa B, Razmjoo H, Peyman A, Ashtari A, Ghafouri I, Moghaddam AG. “Short-term result of collagen crosslinking in pellucid marginal degeneration.” Adv Biomed Res. 2016 Dec 27;5:194.
26. Beckman Rehnman J, Behndig A, Hallberg P, Lindén C. “Increased Corneal Hysteresis After Corneal Collagen CrosslinkingA Study Based on Applanation Resonance Technology.” JAMA Ophthalmol. 2014;132(12):1426–1432. doi:10.1001/jamaophthalmol.2014.3029
27. Tognon, Taíse et al. “Indications and Visual Outcomes of Intrastromal Corneal Ring Segment Implantation in a Large Patient Series.” Clinics 72.6 (2017): 370–377. PMC. Web. 13 Aug. 2017.
28. Varley G, Macsai M, Krachmer J. “The results of penetrating keratoplasty for pellucid marginal corneal degeneration.” AM J Ophthmol. 1990 Aug;110(2):149-152.
29. Skeens HM[1], Holland EJ. “Large-diameter penetrating keratoplasty: indications and outcomes.” Cornea. 2010 Mar;29(3):296-301.
30. Moshirfar M, Edmonds JN, Behunin NL, Christiansen SM. “Current options in the management of pellucid marginal degeneration.” J Refract Surg. 2014 Jul;30(7):474-85.
31. Bennett E, “Scleral Lens Troubleshooting FAQs” by GPLI and SLES. Published on GPLI.info, 2017
32. Caroline PJ, Andre MP. “’Life’ beneath a scleral lens… epithelial bogging.” Contact Lens Spectrum 2015;30(3):56.