# TOPOGRAPHIC ANALYSIS IN PELLUCID MARGINAL CORNEAL DEGENERATION AND KERATOGLOBUS

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## SUMMARY

**Pellucid marginal corneal degeneration (PMCD) is an uncommon cause of inferior peripheral corneal ectasia, affecting patients between the ages of 20 and 40 years. Although histopathologically it is considered a variant of keratoconus, it differs in that the marked corneal steepening occurs more inferiorly, above a narrow band of corneal stromal thinning concentric to the inferior limbus. Here we present two cases. The first case is a clinically typical bilateral PMCD with a characteristic pattern of irregular against-the-rule astigmatism on corneal topography. The second case had an uncommon presentation of hydrops in a clinically keratoglobic eye which showed a marked steepening of the inferior corneal periphery on corneal topography. The other eye showed both clinically and topographically the features of PMCD. Corneal topography suggested that in the second patient, PMCD may have preceded the development of keratoglobus. Keratoconus, PMCD and keratoglobus are considered to be associated as part of the spectrum of non-inflammatory corneal thinning disorders. However, although the finding of PMCD and keratoconus in fellow eyes has been reported, to the best of our knowledge progression from PMCD to keratoglobus has not previously been shown.**

Pellucid marginal corneal degeneration (PMCD) is a non-inflammatory disorder showing a typical crescent-shaped area of thinning in the inferior portion of the cornea separated from the limbus by 1–2 mm of normal-thickness cornea.[1] Irregular astigmatism is very common in this condition and the fitting of hard contact lenses is often difficult due to inferior decentration. Different surgical approaches, in isolated case reports and small series, have been described for the treatment of this uncommon condition.[2-4]

Keratoglobus is an extremely rare bilateral corneal ectasia which is characterised by a globoid protrusion of a clear, diffusely thin cornea of normal to moderately increased diameter.[5] The corneal stromal thickness in this condition is more attenuated near the limbus – in contrast to the thinning in PMCD.

This report describes and illustrates the topographic appearance of PMCD in two patients, one of whom also has keratoglobus. Corneal topographic analysis suggests that PMCD may have preceded the keratoglobus.

## PATIENTS AND METHODS

Computer-assisted corneal topographic maps were obtained from four eyes of two patients with a working slit lamp diagnosis of bilateral PMCD in one patient, and keratoglobus with PMCD in both eyes of the second patient. The TMS-1 videokeratoscope (Computed Anatomy, New York) with a 25 ring Placido cone was used for the capture of the images, and the 1.61 software version for the analysis. The absolute colour scale, with dioptric power intervals of 1.5 dioptres (D) in the midrange (from 35.5 to 50.5 D) and 5.0 D above or below this range, was used for the construction of the colour-coded maps.

## CASE REPORTS

*Case 1*

A 40-year-old Caucasian man was referred to Bristol Eye Hospital by his local optician with a progressive bilateral increase in astigmatism from 2 DC to 8 DC over a period of 3 years. The patient gave a history of progressive deterioration of vision in both eyes. The provisional slit lamp diagnosis was bilateral PMCD, and topographic pictures were obtained with the

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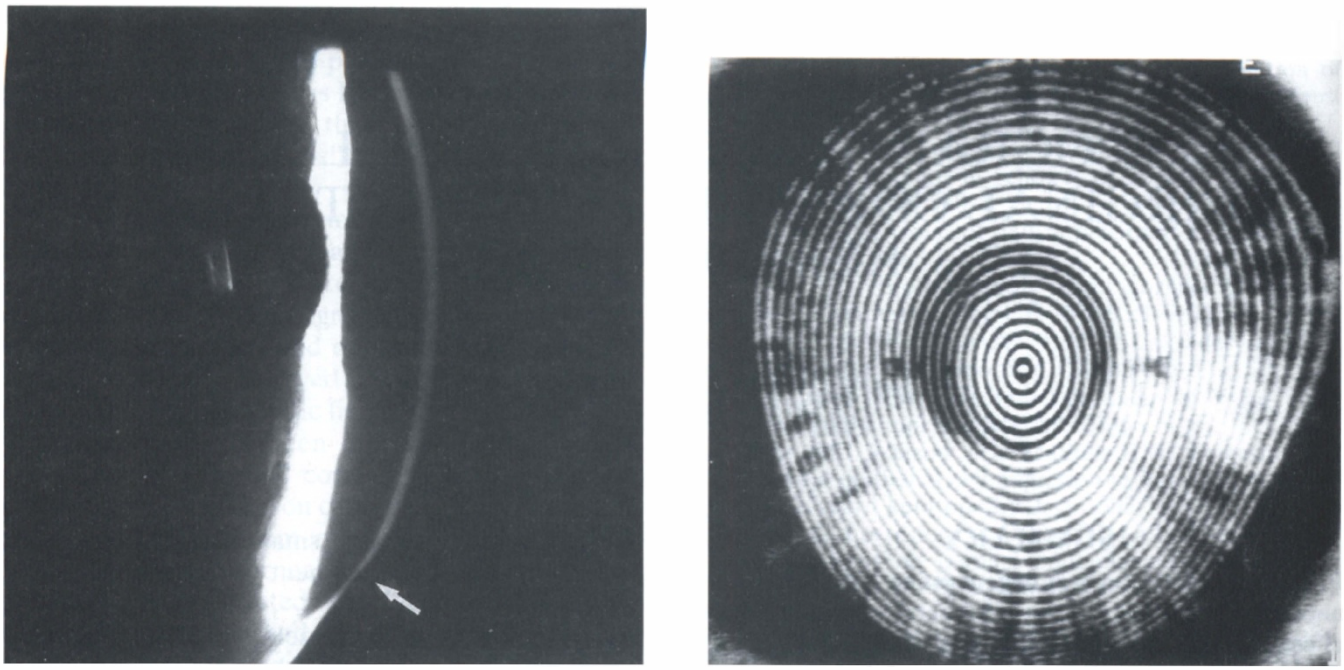

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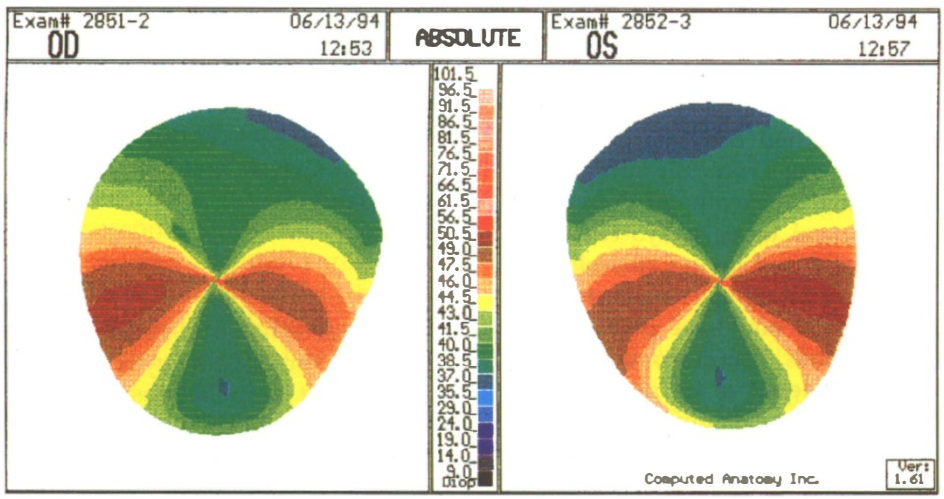


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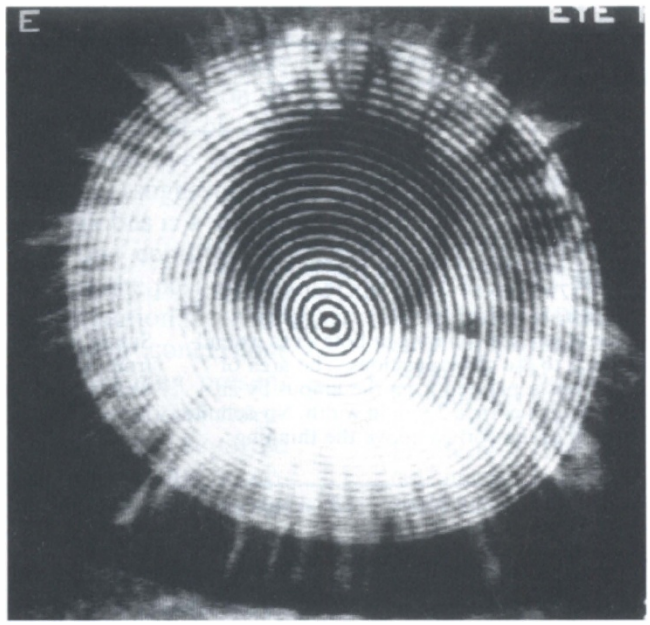
**Fig. 1.** *Case 1. (a) Slit photography of the right eye (pellucid marginal corneal degeneration). Inferior corneal thinning was observed (arrow) 1–2 mm from the limbus, extending from the 5 to 8 o'clock positions in both eyes of case 1. (b) Videokeratoscopic image of the right eye shows a typical pear-shaped image with compression of the inferior rings. The left eye of the same patient had an identical mirror videokeratoscopic image. It can also be seen that the keratoscopic mires are hardly covering the area where the peripheral corneal thinning occurs. (c) The corneal topographic maps (absolute scale). The right eye shows against-the-rule astigmatism of 10.6 DC. The left eye shows enantiomorphic symmetry (mirror image) to the right eye, with 11.9 DC of against-the-rule astigmatism. In early PMCD the power of the cornea is lowest at a vertical axis very close to 90 degrees. The area of greater power is presented in a 'bow-tie' configuration of two semimeridians inferior and oblique to the horizontal axis. In both eyes the topographic maps failed to identify any areas of increased corneal power inferiorly.*

TMS-1 system. Investigations and clinical data of the case are shown in Table I and Fig. 1. The topographic maps indicated a 'bow-tie' configuration of against-the-rule astigmatism in both eyes.

During the follow-up period the patient has been fitted with different sizes of gas-permeable contact lenses with reasonable success. Sequential topography maps (6 months intervals) remain identical, showing fairly regular against-the-rule astigmatism, but a trend towards an increase in simulated keratometry (simk) readings.

*Case 2*

A 44-year-old Caucasian man was referred from another Ophthalmic Unit where he initially presented with an acute onset of redness, pain, watering



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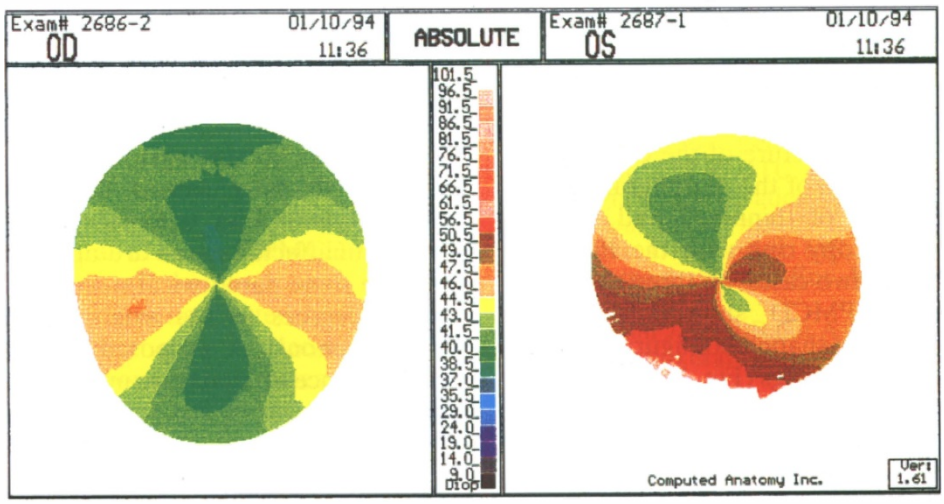


(b)

**Fig. 2.** *Case 2. (a) Videokeratoscopic image of the left eye. Compared with Fig. 1b, this shows inferonasal narrowing of the rings, indicating steepening but without the typical pear-shaped configuration seen in the other eyes. The patient is fixating to the target. The videokeratoscopic image of the right eye was similar to the pear-shaped image demonstrated in case 1. (b) The corneal topographic maps (absolute scale). The right eye shows marked against-the-rule astigmatism of 6.3 DC. The left eye shows irregular astigmatism with quite irregular power distribution. The axis of lowest corneal power is shifted about 35 degrees from the vertical axis, with a very asymmetrical 'bow-tie' configuration and with the inferior low-power semimeridian positioned above an area of high power at the inferior peripheral cornea. This area of peripheral inferior corneal steepening is extending to the steep oblique semimeridians.*

and significant deterioration of his left eye vision. On that occasion he was found to have a 'keratoglobus type of condition' in that eye, with some stromal oedema. The intraocular pressure was raised at 30 mmHg and the patient was started on topical g. fluorometholone q.d.s., g. timolol maleate b.d. and acetazolamide p.o. He was subsequently referred to Bristol Eye Hospital for a second opinion. The previous ophthalmic history was of progressive myopia in both eyes since his twenties, with the left eye the worse of the two.

Examination data of this case are shown in Table I. The clinical picture in the right eye was consistent with PMCD, whereas that of the left eye was consistent with keratoglobus with additional signs compatible with a resolved acute hydrops. Videokeratography and corneal topographic maps were obtained from both eyes and are shown in Fig. 2.

The patient was given a new spectacles prescrip-

**Table I.** Refractive, keratometric, topographic and clinical data of the two cases

| | Case 1 | | Case 2 | |
|---|---|---|---|---|
| | Right eye | Left eye | Right eye | Left eye |
| Diagnosis | PMCD | PMCD | PMCD | Keratoglobus |
| VA unaided | 6/18 | 6/60 | 6/60 | 6/60 |
| Refraction | −5.00/+10.00×180 | −4.50/+10.00×2 | −3.75/+5.25×2 | −6.25/+6.25×32 |
| VA aided | 6/6 | 6/6 | 6/5 | 6/7.5 |
| Correction | Spectacles/RGPCL | Spectacles/RGPCL | Spectacles | Spectacles |
| Keratometry | 47.29@3/38.55@93 | 49.11@3/38.29@94 | NA | NA |
| simk readings | 49.3×179/38.7×89 | 50×176/38.2×86 | 45.7×14/39.4×104 | 50.2×34/42.9×124 |
| **Figures** | **1a, 1b, 1c** (left) | **1c** (right) | **2b** (left) | **2a, 2b** (right) |
| Clinical appearance | Peripheral inferior thinning in both eyes extending from 5 to 8 o'clock positions. The area of thinning was separated from the limbus by an area of normal cornea 2 mm in width. No definite protrusion of the cornea above the thinning | | Subtle inferior thinning from about 5 to 7 o'clock position | Circumferential zone of peripheral corneal thinning. Central cornea bulging with resolved hydrops in the inferotemporal quadrant |

VA, visual acuity; RGPCL, rigid gas-permeable contact lens; simk, simulated keratometry.

tion and has been reviewed annually. No further action is planned for his condition at present.

## DISCUSSION

### *Corneal Topography of PMCD*

The clinical picture of PMCD results in a flattening of the vertical meridian and the appearance of a marked against-the-rule irregular astigmatism. The typical photokeratoscopic picture of the condition is a very close approximation of the peripheral keratoscopy rings in the area of thinning,[6] and a pear-shaped central keratoscopy ring with the Placido's disc-like target.[7] In our two cases, all three eyes with clinical diagnosis of PMCD showed a typical videokeratograph of pear-shaped configuration with compression of the inferior rings (Fig. 1b), which is in accordance with the previously reported findings. However, it must be pointed out that this appearance, although typical, is not diagnostic of the condition, as a similar videokeratoscopic image can be seen in patients with keratoconus.

The typical topographic map of the condition has been reported to show marked flattening of the cornea along a vertical axis and a steepening of the inferior cornea peripheral to the site of the lesion. This area of steepening extends into the areas of associated horizontal steep semimeridians.[7] However, a similar picture can be seen in other cases of peripheral thinning such as in Terrien's marginal degeneration. In this condition also a flattening of the cornea over the thinned area is noted, associated with a relative steepening of the surface approximately 90 degrees away; it is thought that the two conditions can be distinguished topographically by the steepening of the inferior corneal periphery extending into the mid-peripheral inferior oblique meridians that occurs in PMCD but not in Terrien's.[8]

Our observations suggest that the so far typically described topographic picture of PMCD may not always present as an inferior peripheral corneal steepening, at least when using a 25 ring photokeratoscope. This target has a limited area of coverage, not extending up to the limbus (Fig. 1b), and covers an even smaller area in steep corneas. In our cases a characteristic 'bow-tie' configuration of marked against-the-rule astigmatism slightly oblique inferiorly can be seen, without peripheral steepening (Figs. 1c and 2b left). In this respect the topographic pictures are not diagnostic, but rather similar to those obtained in other peripheral ectasias. Typically, stromal thinning causes flattening overlying the area of tissue loss, with steepening as the borders of the unaffected tissue are approached. There may also be some paradoxical steepening of the meridian at 90 degrees to the axis of flattening ('coupling phenomenon'). A proposed explanation for that is the presence of intact circumferential rings of stromal lamellae.[9]

### *Corneal Topography of Keratoglobus*

To the best of our knowledge the topographic appearance of keratoglobus has not been reported previously. It is our own observation that obtaining reliable and reproducible images in keratoglobus patients is very difficult. This may reflect a limitation of videokeratography in measuring the topography of highly irregular corneas, either due to inaccuracy of the currently used algorithms for very steep corneas, or due to the disturbances of the tear film in these corneas. The latter could cause disturbance of the mire pattern and poor quality of topographic information.

The left eye of case 2 presented clinically as early keratoglobus. Reliable topographic pictures were obtained which showed an arc of peripheral increase in corneal power (steepening) (Fig. 2b left). This suggests that it is probably a case of PMCD which advanced to a keratoglobus condition by circumferential extension of the peripheral thinning as noted on slit lamp examination. When a peripheral gutter

extends circumferentially, more extensive stromal tissue disruption occurs and the flattening produces an 'arching' effect in the 'bow-tie' configuration of corneal topography.[9] Considering this kind of corneal topography progression in the left eye of our case 2 (from Fig. 2b left to Fig. 2b right) suggests that progression from PMCD to keratoglobus is a strong possibility. The progression from one condition to the other has only been advocated in the past in one case, but without topographic confirmation.[10]

In summary, there are four main conclusions arising from this study. First, PMCD may present topographically without an area of peripheral steepening. In contrast to what has been reported previously,[5,8] the picture is not always pathognomonic of the condition. Second, given the known inaccuracies of the currently used algorithms in producing reliable data from the periphery of irregular aspherical surfaces,[11] and the limited area of corneal coverage with the 25 ring target, corneal topography systems may fail to identify early cases. Other methods of corneal topography that can provide information on the entire corneal and limbal surfaces, such as rasterstereography,[12] might be a more useful alternative in such cases. We also think that the use of a 32 ring cone should be more appropriate for examining peripheral corneal diseases. Third, our second case suggests that PMCD may precede the development of keratoglobus in some cases. The natural history of PMCD may be towards development of keratoglobus with a mechanism of circumferential extension of the peripheral gutter. Fourth, there is a place for routine corneal topographic analysis in these cases, mainly to monitor progress of the disease, as the topographic changes *per se* are not always diagnostic and may resemble other ectatic conditions. Careful clinical examination remains valuable, but probably decisions on further management rely upon sequential topographic information.

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Key words: Pellucid marginal corneal degeneration, Corneal ectasia, Keratoglobus, Corneal topography.

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