

Classification of KC is the first step in approaching the disease because the severity of the disease and the stage at which the patient is diagnosed and treated affect treatment results. KC can be classified according to the morphology of the cone and the pattern of corneal topography.

2.1 Morphological Patterns

Morphologically, KC has three types of cones:

- (a) Nipple cones, characterized by their small size (5 mm) and steep curvature. The apical center is often either central or paracentral and commonly displaced inferonasally (Fig. 2.1).
- (b) Oval cones, which are larger (5–6 mm), ellipsoid, and commonly displaced inferotemporally (Fig. 2.2).
- (c) Globus cones, which are the largest (>6 mm) and may involve over 75% of the cornea. Figures 2.3 and 2.4 are the tangential curvature map and the thickness map respectively of a globus cone, note the generalized corneal thinning.

Morphology of the cone is determined according to its size on corneal topography. The best map to evaluate the cone is the tangential map since it is the best to highlight corneal irregularities. In mild cases, cone morphology may be indeterminate.

2.2 Topographical Patterns

Topographically, KC can be classified according to elevation maps, to thickness map or to curvature maps.

2.2.1 Classification According to Elevation Map

Cone location is determined only by the elevation maps. The elevation maps can be displayed either by best fit sphere mode (BFS) as shown in Fig. 2.5, or by best fit toric ellipsoid mode (BFTE) as shown in Fig. 2.6. The best to locate the cone is the BFS, and the best to evaluate the real height of the cone is the BFTE. On the BFS, the cone can be central, paracentral, or peripheral as shown in Figs. 2.7 and 2.8. This classification is important for differentiating KC from PMD and for treatment as will be discussed later in details.

2.2.2 Classification According to Thickness Map

There are two patterns of the thickness map in KC, the conic or dome-like and the “bell” shape. The conic or dome-like shape (Fig. 2.9) is encountered in KC, while the bell shape is encountered in PMD (Fig. 2.10). The bell shape comes from the inferior wide thinning of the cornea found with PMD. When the bell shape is seen, PMD is to be suspected and inserting intracorneal rings carries the risk of perforation, this will be discussed later in details.

2.2.3 Classification According to Curvature Map

Upon studying corneal topography, special attention should be paid to the anterior sagittal curvature map. There are several abnormal signs on these maps that we

Fig. 2.1 Nipple cone. A small steep central or paracentral cone

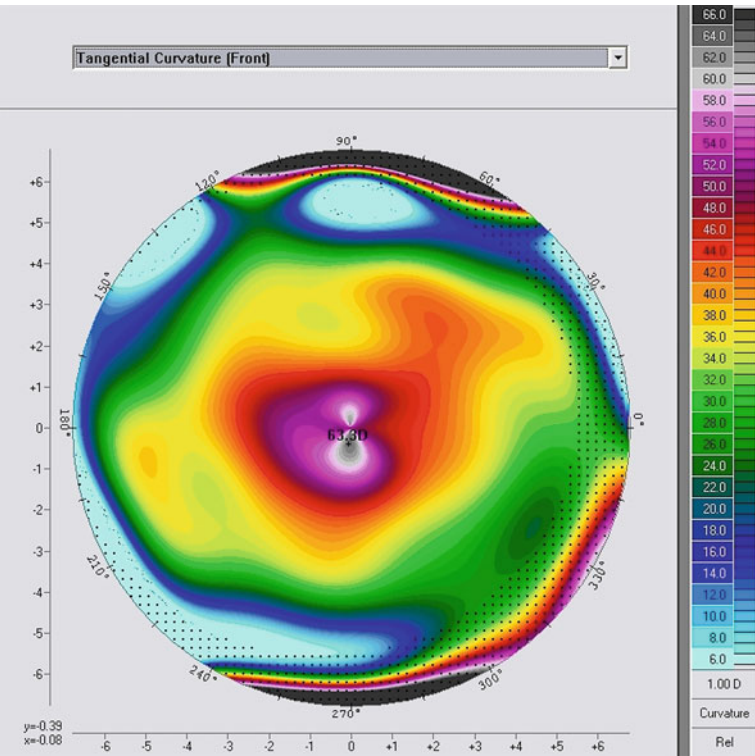


Fig. 2.2 Oval cone. A steep elliptical cone that is commonly displaced inferotemporally

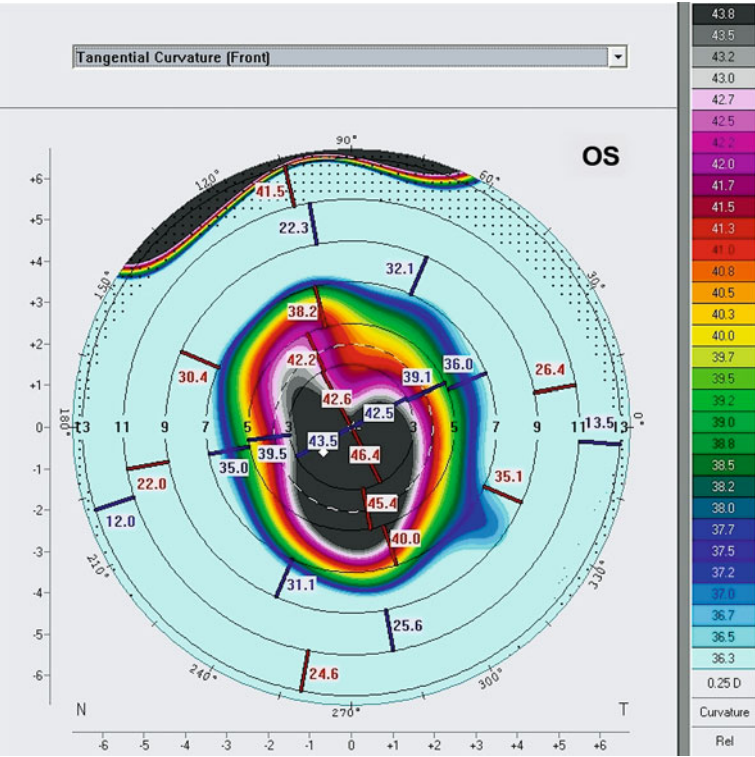
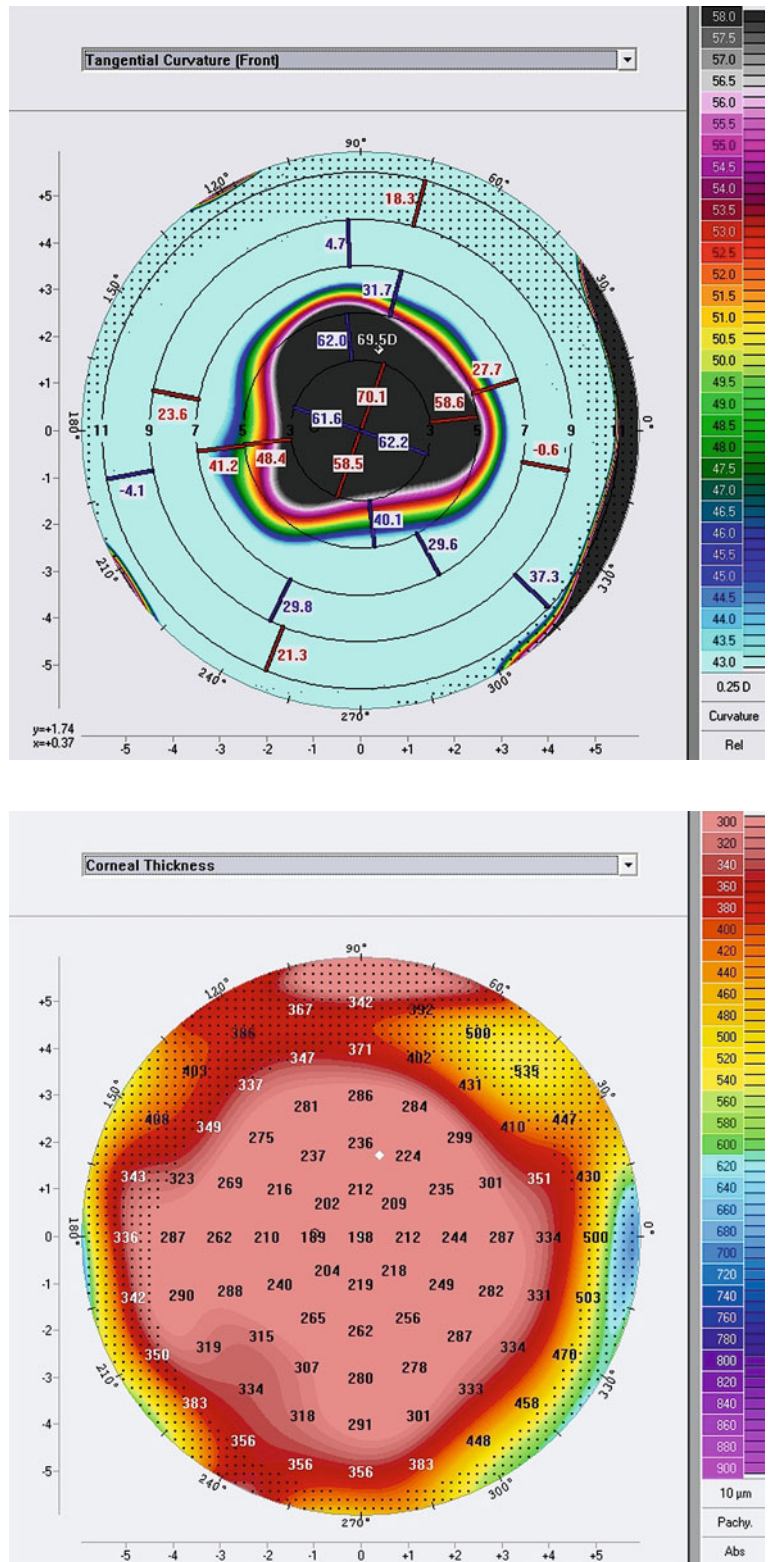


Fig. 2.4 Globus cone. Corneal thickness map: generalized corneal thinning



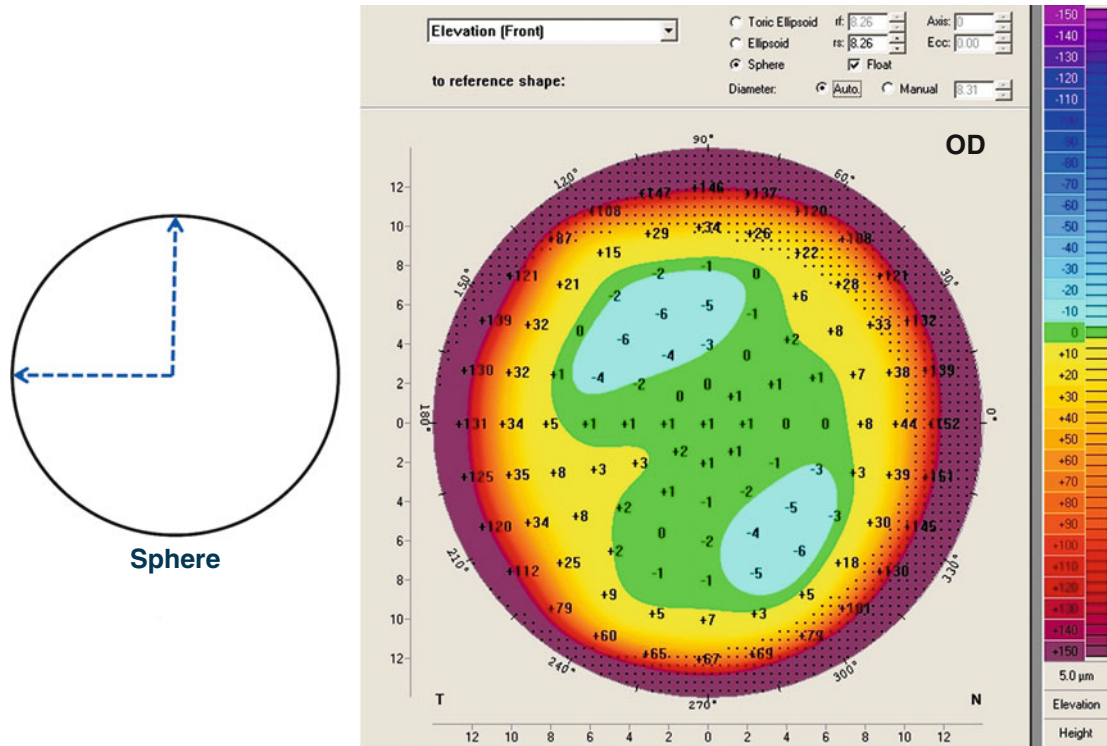


Fig. 2.5 The elevation map displayed in the best fit sphere float mode

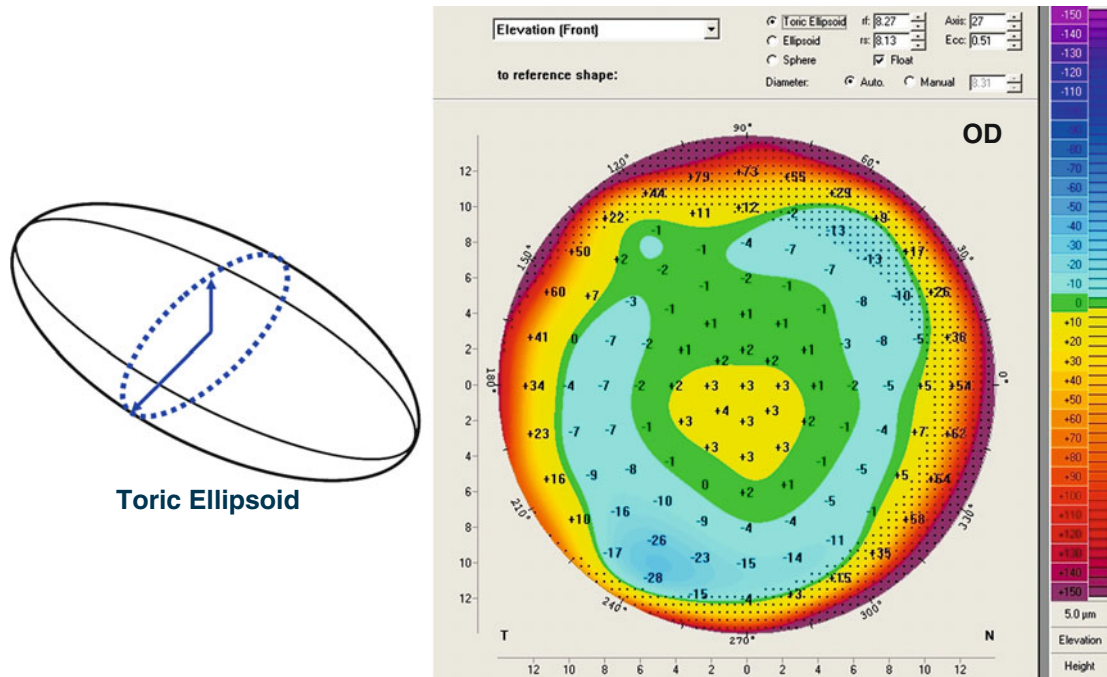


Fig. 2.6 The elevation map displayed in the best fit toric ellipsoid float mode

Fig. 2.7 A central cone as shown on the elevation map with the best fit sphere float mode; the white arrows point at the cone

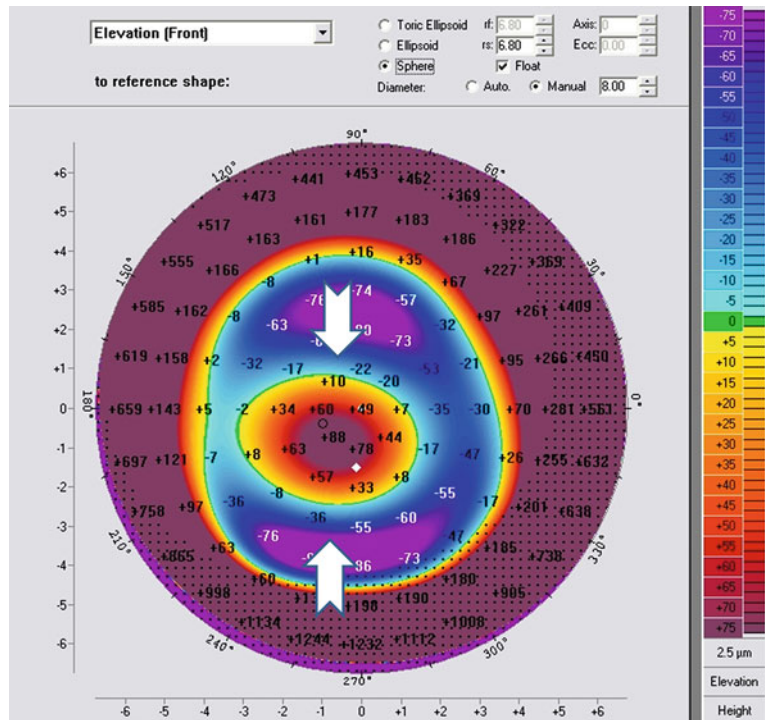


Fig. 2.8 A peripheral cone as shown on the elevation map with the best fit sphere float mode; the white arrow points at the cone

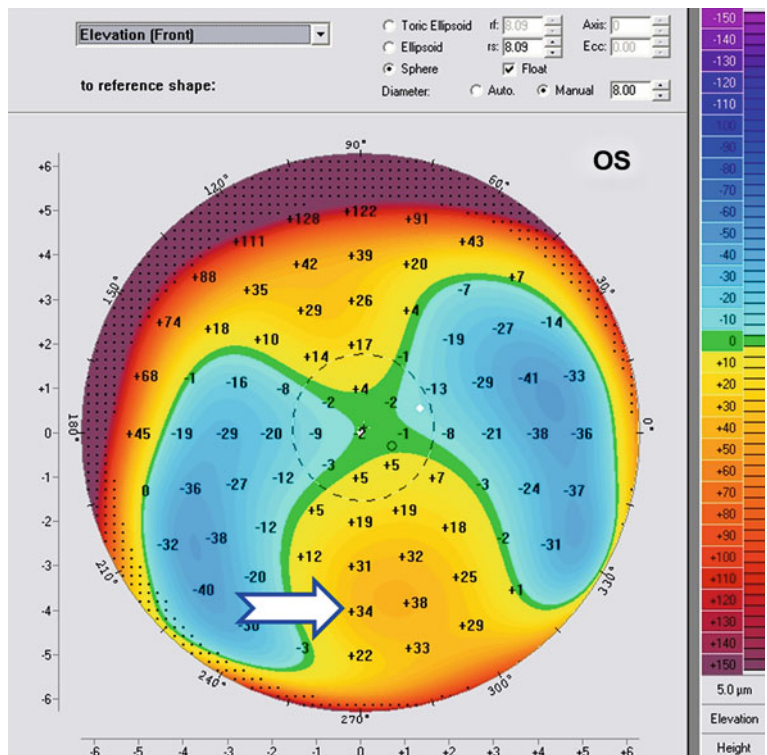


Fig. 2.9 Dome shape of the cone in KC on the corneal thickness map

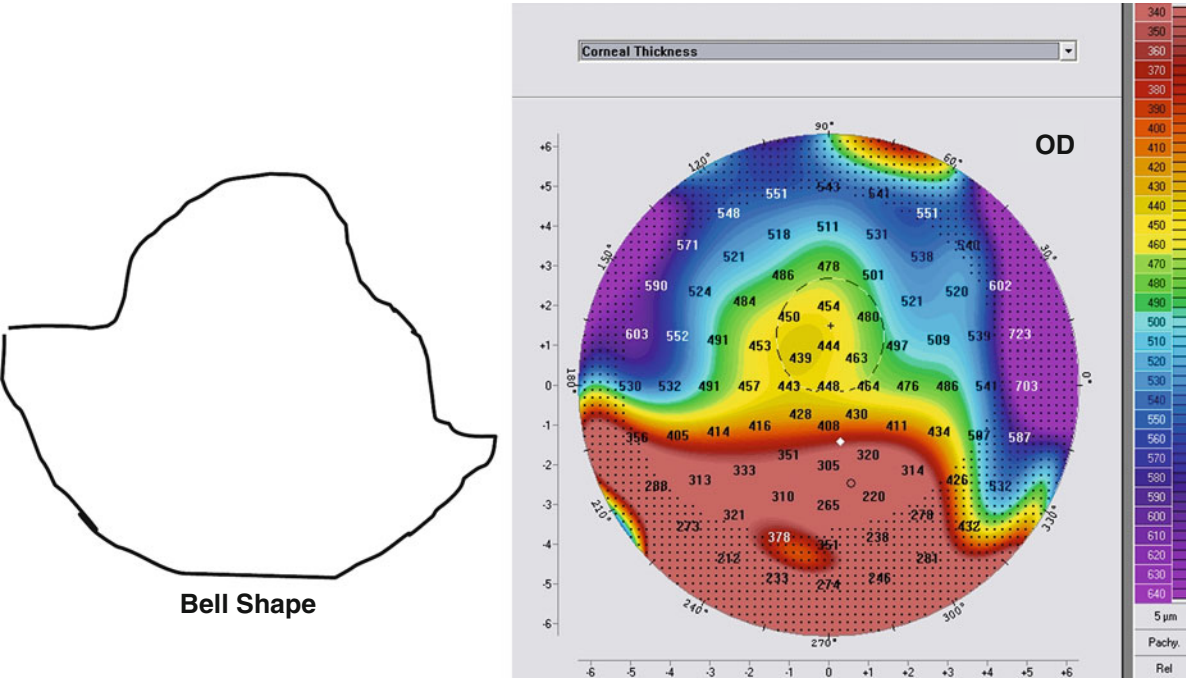
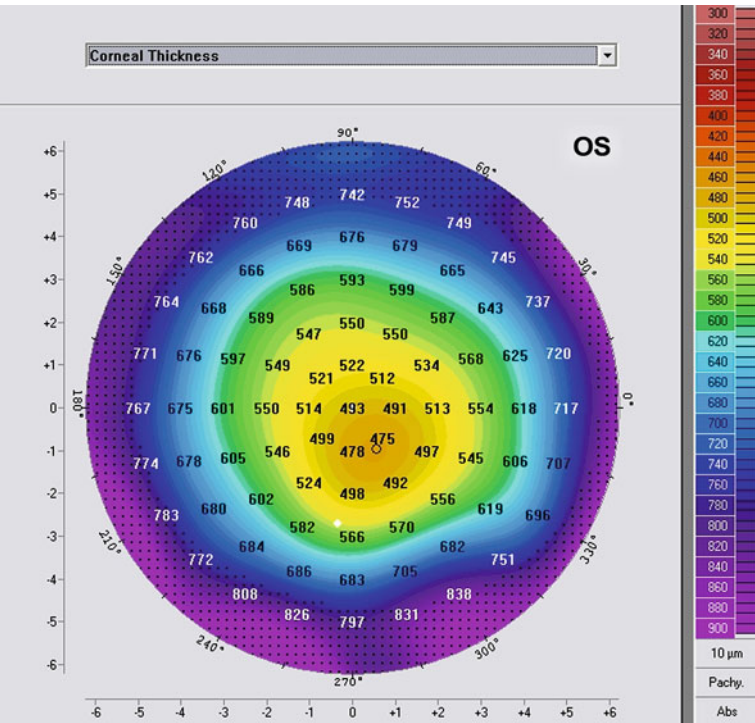
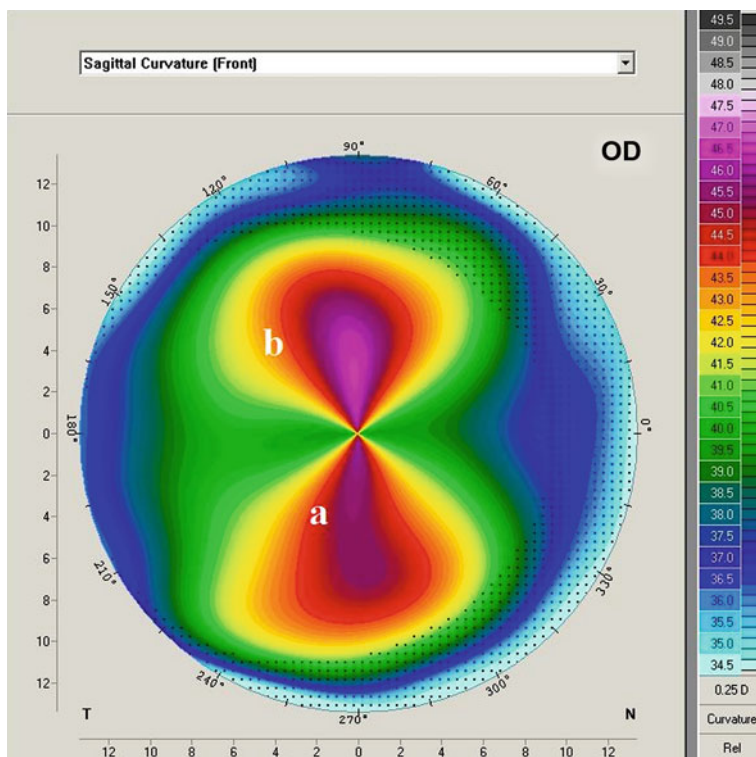


Fig. 2.10 Bell sign on the corneal thickness map in PMD

Fig. 2.11 Symmetric bowtie (SB). It has two equal and aligned segments “a” and “b.” When the SB is aligned vertically, it represents with-the-rule astigmatism



should be aware of; some of these signs indicate KC and some indicate corneal irregularities, i.e. KC is always considered as corneal irregularity but not every corneal irregularity is a KC! To understand the curvature classification, the normal cornea should be studied.

2.2.3.1 The Normal Cornea

When considering the topography of a normal cornea, we feel the need to remember that there is a wide spectrum of normality. No human cornea demonstrates the kinds of regularity found in the calibration spheres of the topographer: The eye is not molded glass-made. Normal corneal topography can take one of the followings:

Regular astigmatism: Every human being has a certain amount of astigmatism, though minimal. The rule is that the vertical meridian of the cornea is slightly steeper than the horizontal. This is known as with-the-rule astigmatism. Figure 2.11 shows the symmetry between segments “a” and “b.” They are also equal in size. That is the normal pattern, it is known as “Symmetric Bowtie (SB)”;

see also Fig. 2.16. If the SB is horizontal, it represents an against-the-rule astigmatism, 90° rotated when compared with a with-the-rule astigmatism (Fig. 2.12).

When the bow tie is diagonal, it represents a cornea having an oblique astigmatism (Fig. 2.13).

In the normal eye, nasal cornea is flatter than temporal. The nasal side of a healthy corneal map becomes blue more quickly, indicating that the nasal cornea is flatter than temporal.

Generally, the two eyes of the same subject are very similar, and present a mirror image of each other (Fig. 2.14). This phenomenon is called enantiomorphism. The knowledge of this fact is useful to decide whether a cornea is normal or not, by comparing with the map of contralateral eye.

P.S. When studying the pattern of corneal curvature, it is important to study the single enlarged map choosing the option of projected circles and the two major axes of curvature; in order to easily compare values in the same eye and between both eyes (Fig. 2.15).

Fig. 2.12 Symmetric bowtie (SB) aligned horizontally representing against-the-rule astigmatism

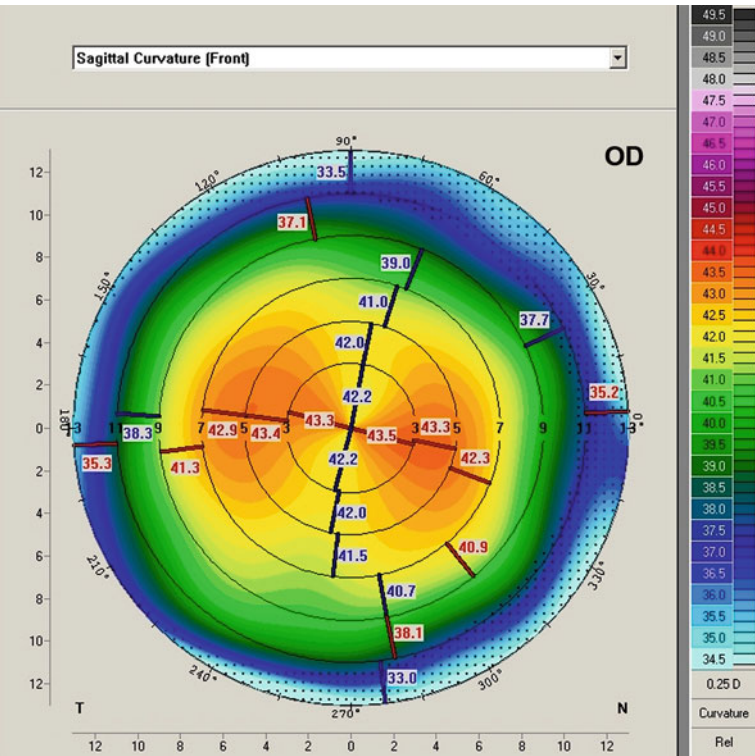
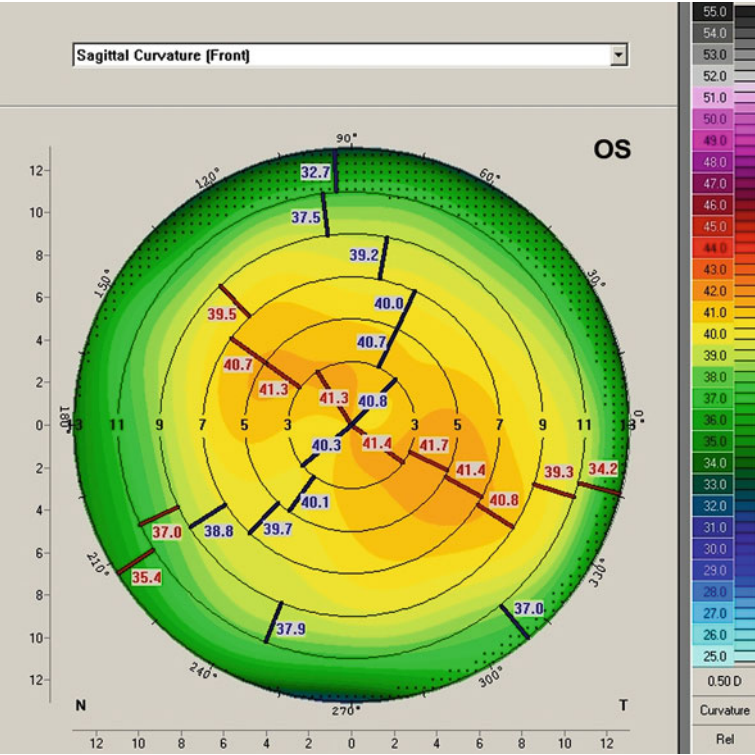


Fig. 2.13 Symmetric bowtie (SB) aligned obliquely representing oblique astigmatism



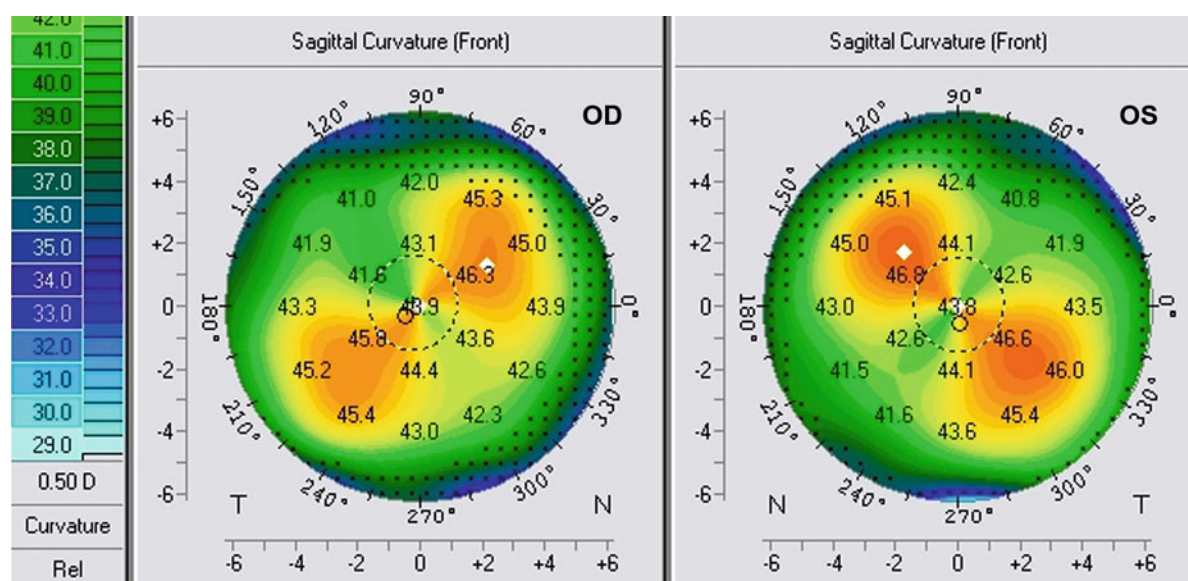


Fig. 2.14 Enantiomorphism. The two eyes of the same subject are very similar, and present a mirror image of each other. The knowledge of this fact is useful to decide whether a cornea is normal or not, by comparing with the map of the contralateral eye

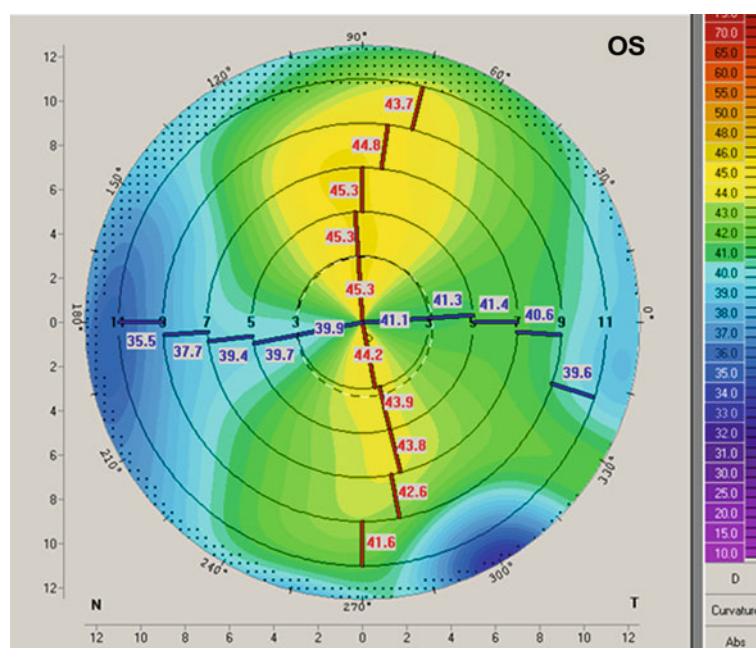


Fig. 2.15 The curvature map as a single enlarged map with projection of circles and the two major axes of curvature. This is important for comparing values in the same eye and between both eyes

2.2.3.2 Topographic Shape Patterns

Characterizing Irregularity (Fig. 2.16)

There are several patterns of corneal curvature; some can be accepted, others are considered risky for lasik surgery or even indicators for KC. Corneal irregularity may appear as one of the following patterns:

Pattern 1: Round. The steepest part of the cornea (hot spot) is round (Fig. 2.17) and often decentered.

Pattern 2: Oval. The steepest part of the cornea (hot spot) is oval and may be centered or decentered (Fig. 2.18).

Pattern 3: Superior Steep (SS). The steepest part of the cornea is localized in the upper part of the cornea (Fig. 2.19).

Pattern 4: Inferior Steep (IS). The steepest part of the cornea is localized inferior to the apex of the cornea (Fig. 2.20).

Pattern 5: Irregular. The corneal surface takes no particular shape; in this pattern, steep areas are mixed with flat areas (Fig. 2.21).

Pattern 6: Symmetric bowtie (SB). This pattern may be an indicative of normal astigmatism or occasionally symmetric type of KC (Fig. 2.22).

Pattern 7: Symmetric bowtie (SB)/Skewed Steepest Radial Axis Index (SRAX). That is a SB with angulation (skew) between the axes of segments “b” and “a.” In this case, corneal astigmatism is called “non-orthogonal astigmatism,” or the “lazy 8” pattern. Angulation is considered clinically significant when it exceeds 22° (Fig. 2.23).

Pattern 8: Asymmetric Bowtie (AB)/IS. That is an AB which is inferiorly steep. The curvature power of segment “a” is higher than that of segment “b.” If the difference is more than 1.5D on the 4-mm circle, it is

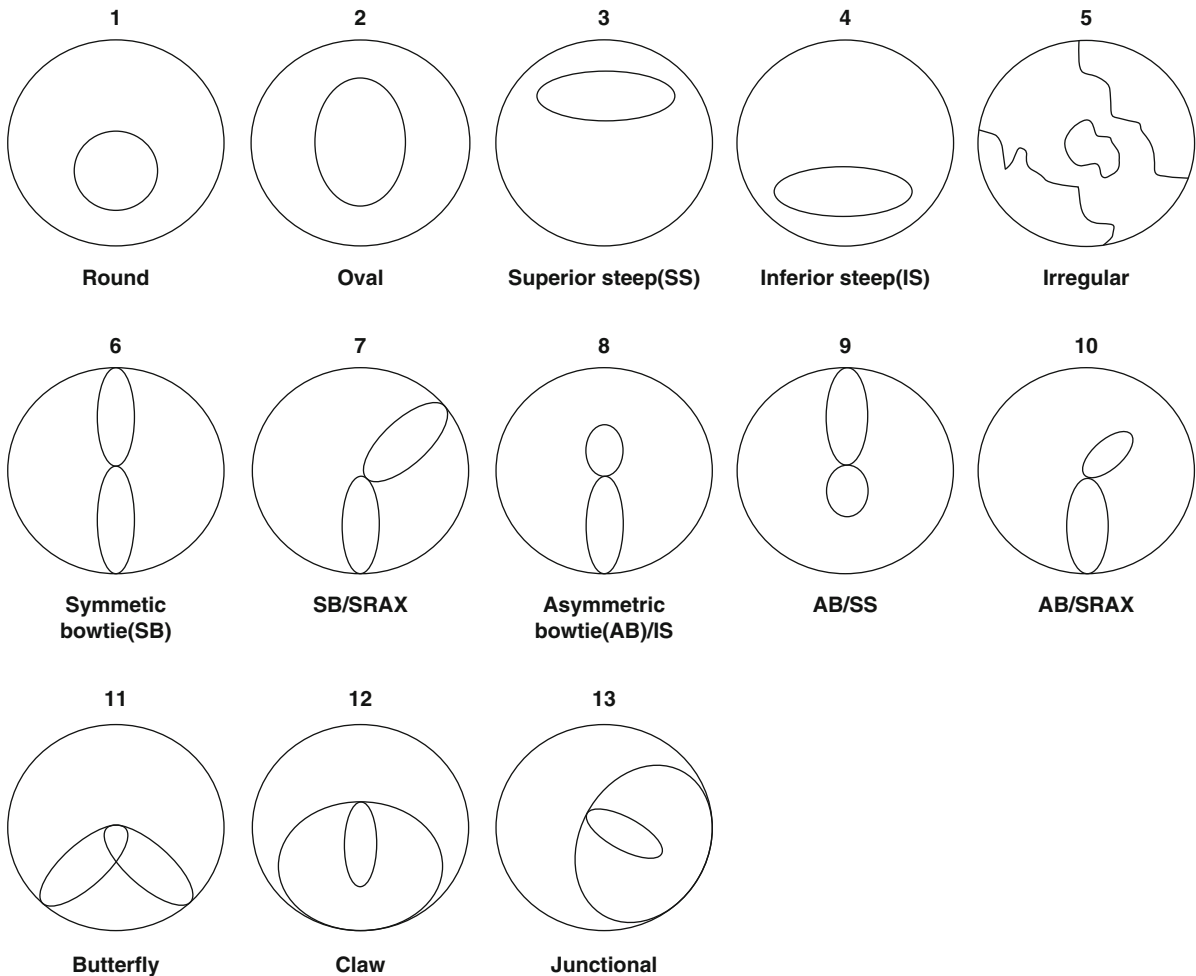


Fig. 2.16 Patterns of the anterior curvature map. The steep part of the curvature map may take a bowtie shape, a hot spot shape, or an irregular shape

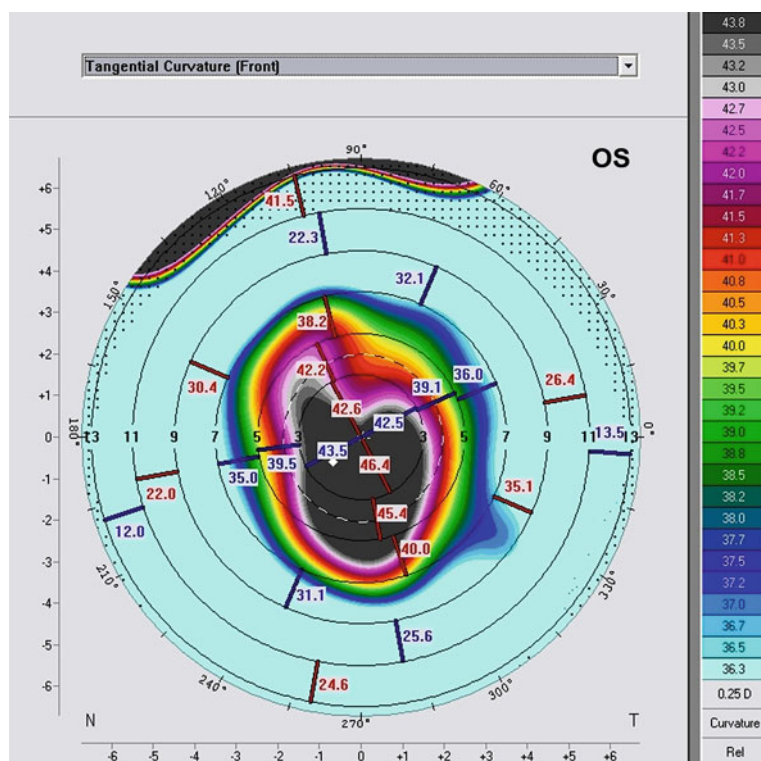
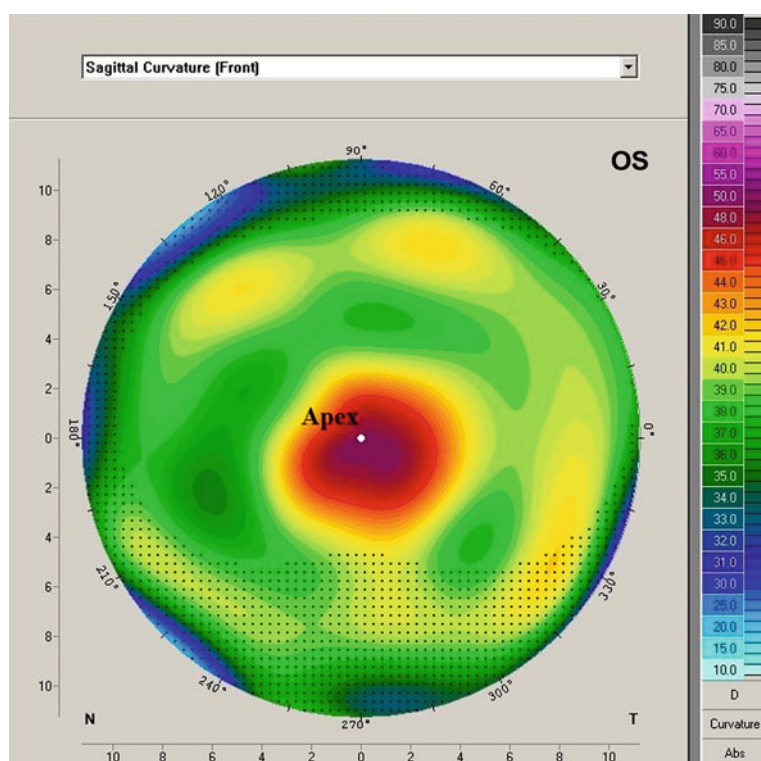
Fig. 2.17 Round hot spot**Fig. 2.18** Oval hot spot

Fig. 2.19 Superior hot spot: This pattern is called superior steep (SS)

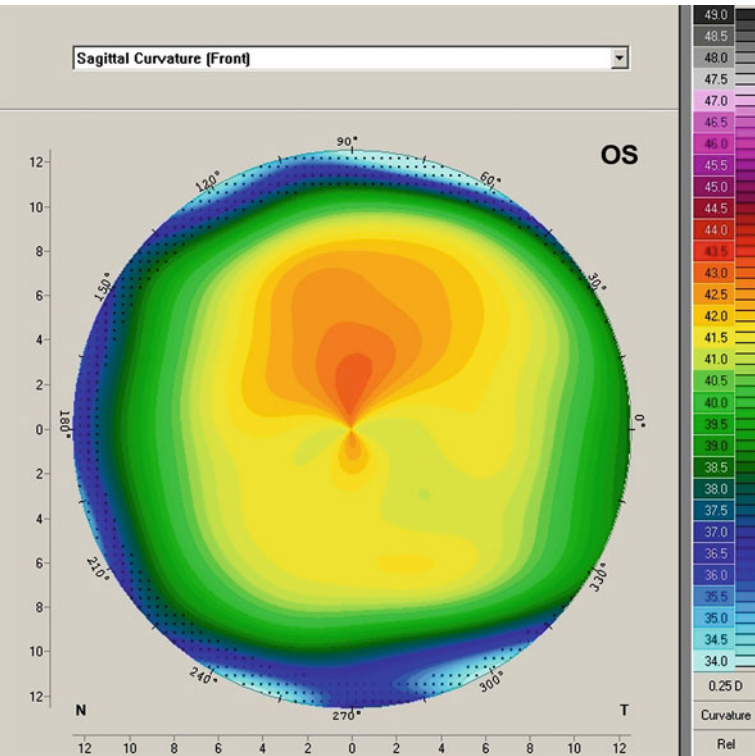


Fig. 2.20 Inferior hot spot: This pattern is called inferior steep (IS)

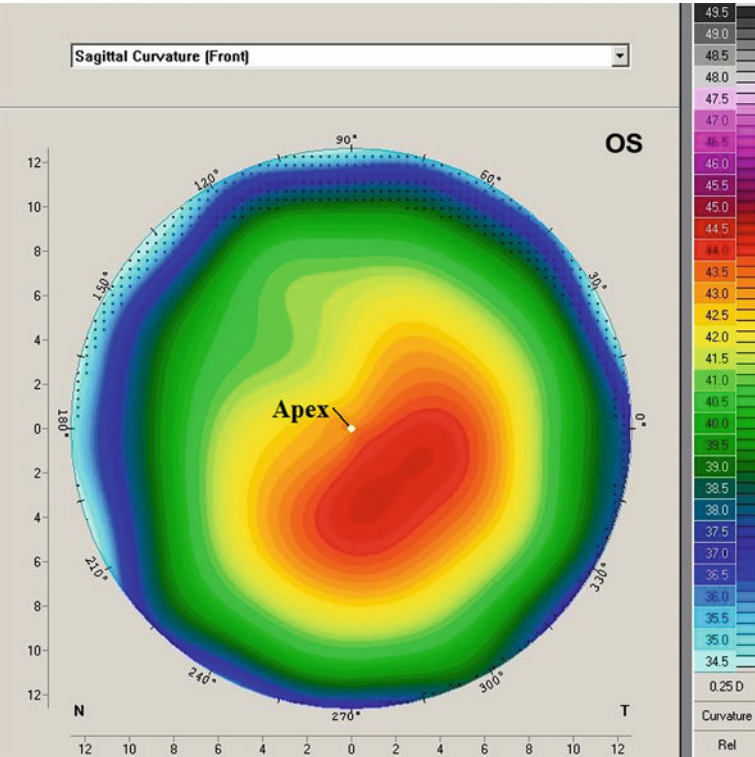


Fig. 2.21 Irregular shape: There is no particular shape where steep areas are mixed with flat areas

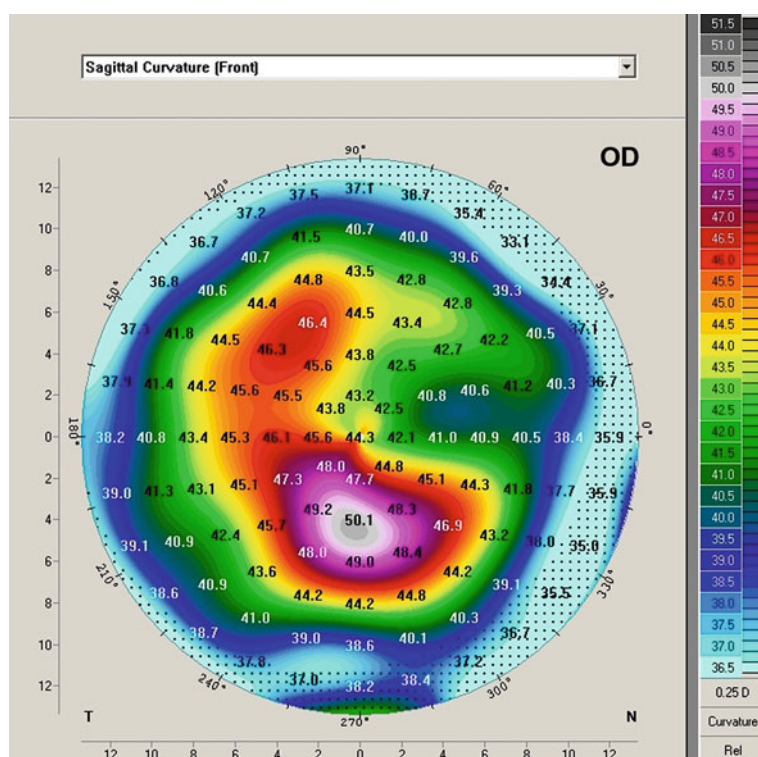


Fig. 2.22 Symmetric bowtie: It may be an indicative of normal astigmatism or occasionally symmetric type of KC

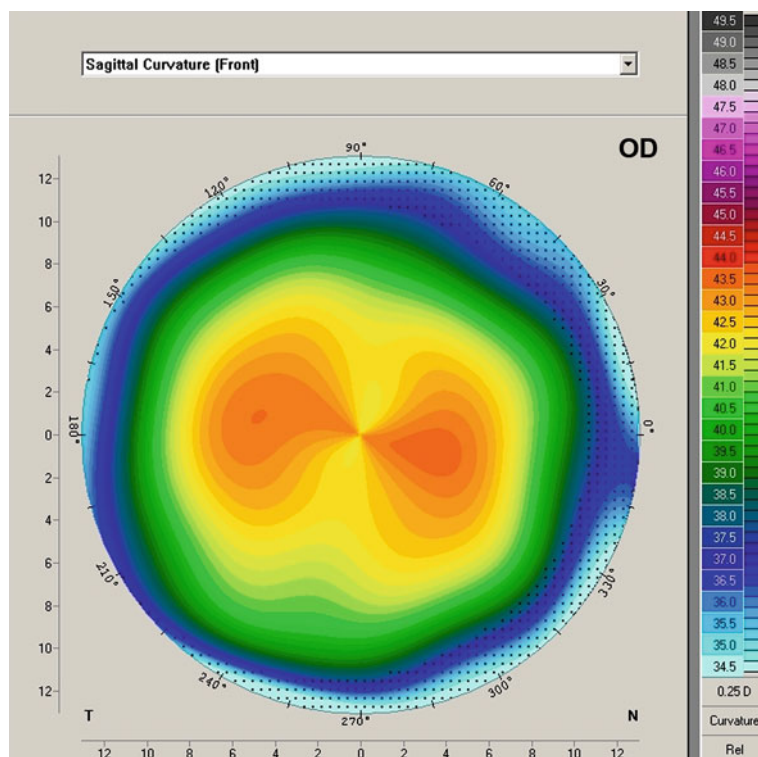
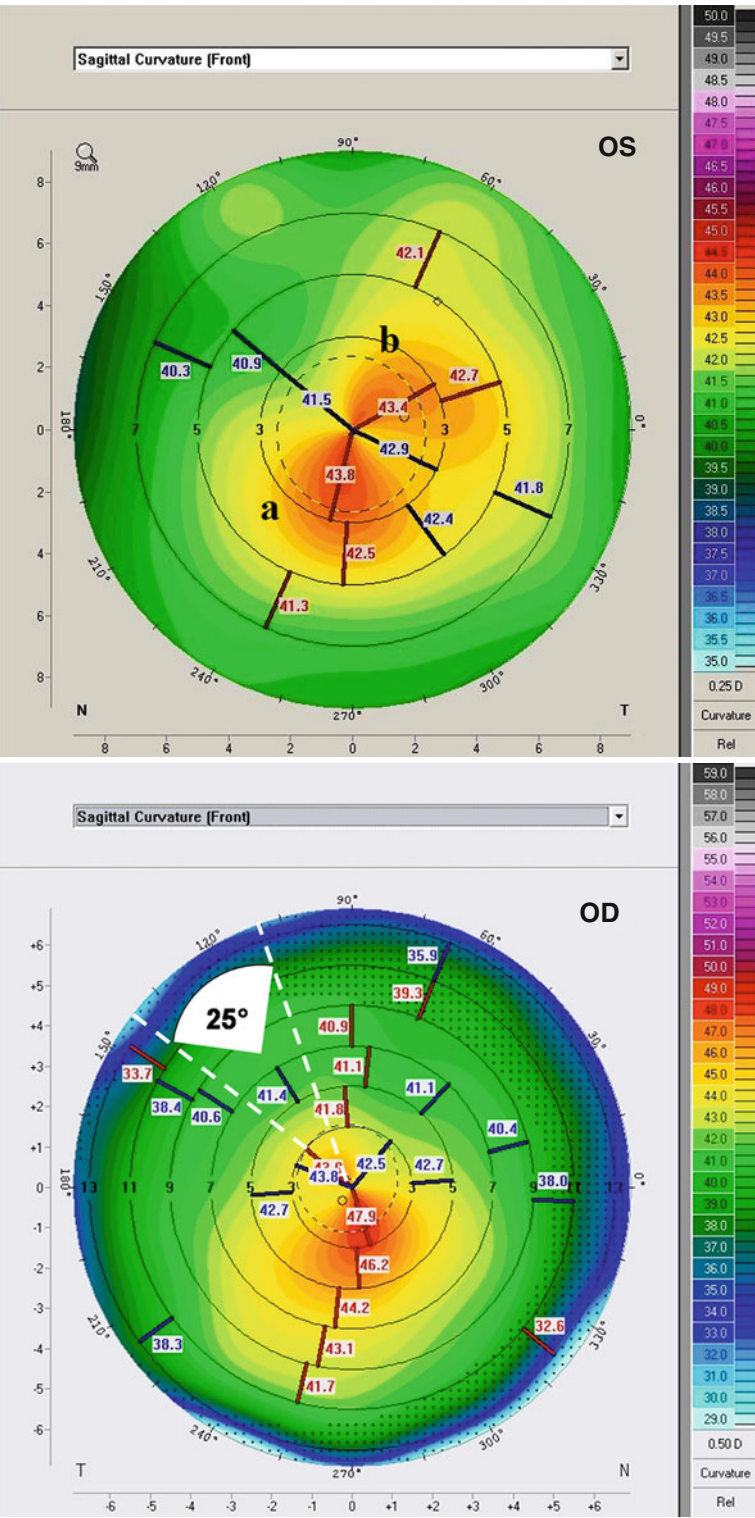


Fig. 2.23 Symmetric bowtie (SB) with Skewed Steepest Radial Axis Index (SRAX): SB/SRAX. There is an angulation between segments' axes. This angulation is clinically significant when it is $>22^\circ$



considered significant (Fig. 2.24). Corneal apex is located in this type in the direction of the steeper (inferior) segment.

Pattern 9: AB/SS. That is an AB superiorly steep. This pattern is the reverse of pattern 8. If the difference is more than 2.5D, precaution should be taken when taking the decision (Fig. 2.25). In this type, the corneal apex is located in the direction of the steeper (superior) segment.

Pattern 10: AB/SRAX. That is an AB with angulation between the axes of the two segments (Fig. 2.26a, b).

Pattern 11: Butterfly: The bow-tie is horizontally aligned with the lobes inferiorly angulated or wing-like spread just like the wings of a butterfly. The lobes are not necessarily equal in size, therefore it can be considered as horizontal sisterhoods of patterns 7 and 10 (Fig. 2.27).

Pattern 12: Claw pattern: it is also known as “kissing birds”: it is like the butterfly pattern added that the wings are joined inferiorly with a central flat area (Fig. 2.28). The vertical meridian of the cornea is flat, on which the K-readings are low, and usually the patient has a hyperopic component. This pattern is encountered in PMD or in PLK.

Pattern 13: Junctional: It is a circular shape, where the two lobes are laterally connected. It can be considered a vertical or oblique type of pattern 12. It is a subject of suspicion (Fig. 2.29a and b).

Pattern 14: Smiling Face: This is certainly risky because it often leads to postoperative ectasia, and might be an indicator of KC (Fig. 2.30).

Pattern 15: Vortex pattern: It is also known as the “Nazi Logo” (Figures 2.31a to d). This is an indicator of corneal instability, and it may precede KC, (notice the vortex distribution of the red and blue lines).

P.S. The most concerning of the previous parameters are steep K-readings, inferior-superior asymmetry, and skewing of the steep axis.

2.2.4 Summary of Topographic Criteria of Keratoconus

When more than one of the following criteria are found, any of the above mentioned patterns is considered as frank KC, Forme Fruste KC (FFKC), early

stage KC, or at least a case of suspicion (according to the severity and amount of signs):

On the sagittal map:

- (a) K-readings > 48 dpt.
- (b) SRAX > 22°.
- (c) Superior–inferior difference (S-I) on the 5 mm circle > 2.5 dpt.
- (d) Inferior–Superior difference (I-S) > 1.5 dpt.
- (e) Corneal astigmatism on either surface should not be higher than 6D; otherwise, it is a risk factor.
- (f) Against the rule, astigmatism is considered suspicious.

On the thickness map:

- (a) Cone-like shape.
- (b) Superior–inferior at 5 mm circle > 30 μ .
- (c) Thinnest location < 470 μ .
- (d) Thickness at pachy apex – thickness at thinnest location > 10 μ .
- (e) Y coordinate value of the thinnest location > –500 μ .
- (f) Difference in thickness between both eyes at thinnest locations > 30 μ .

On the elevation maps:

- (a) Isolated island or tongue-like extension (BFS mode) on either surface.
- (b) Values > 12 μ within the central 5 mm on the anterior elevation map (BFTE mode).
- (c) Values > 15 μ within the central 5 mm on the posterior elevation map (BFTE mode).

For further information, please refer to my book: “Corneal Topography in Clinical Practice, Jaypee Brothers 2009.”

2.2.5 The Author’s New Classification of Topographical Patterns of Keratoconus

Upon revision of the results of the author’s first 400 cases of intracorneal rings with at least 6-month follow-up, he could recognize some factors affecting the results. From a topographical point of view, studying the changes that occurred in the sagittal curvature map revealed that there are three factors affecting the response to the rings: (1) the skew between the axes of the bowtie segments, (2) the size of the bowtie segments, and (3) the shape of the map. To understand this, the steep and flat segments should be projected on

Fig. 2.24 Asymmetric bowtie inferiorly steep: AB/IS. The inferior segment has higher values than the superior one. As shown in *white circles*, the inferior value is higher than the superior by more than 1.5 dpt, which is clinically significant

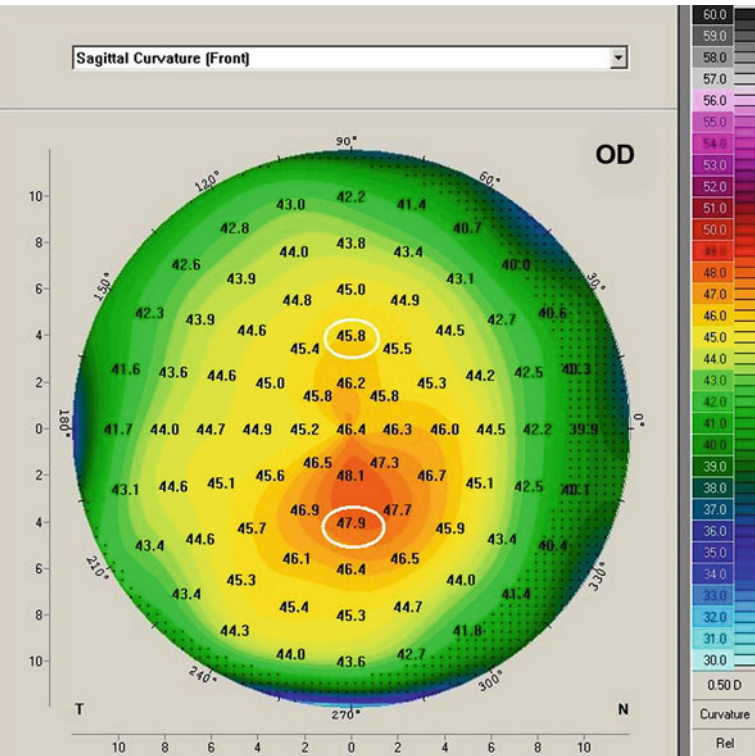


Fig. 2.25 Asymmetric bowtie superiorly steep: AB/SS. It is opposite to the pattern ion Fig. 2.24

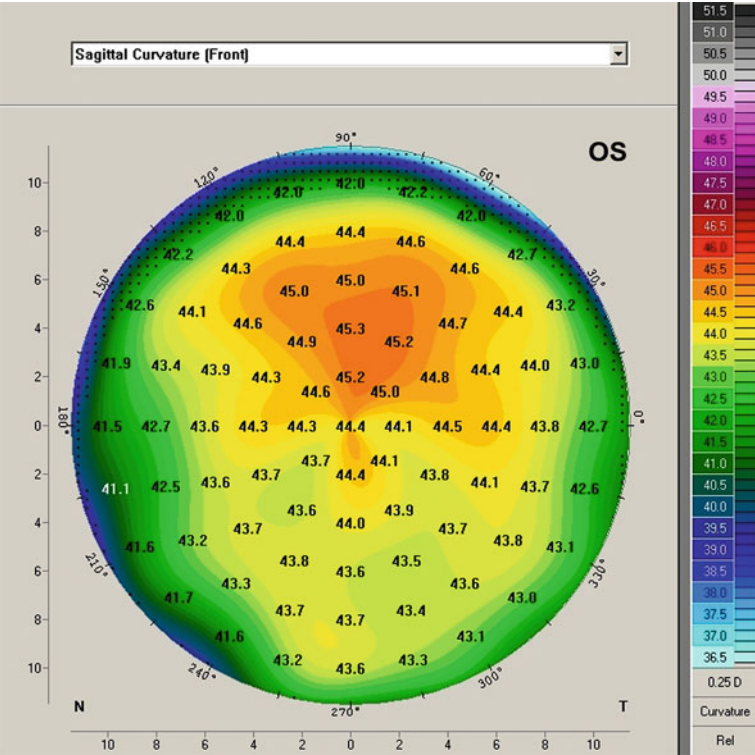


Fig. 2.26 Asymmetric bowtie with Skewed Steepest Radial Axis Index: AB/SRAX. There is an angulation between asymmetric segments. This angulation is clinically significant when it is $>22^\circ$. (a) is the anterior sagittal map without projected meridians; (b) is the same map with projected meridians to show the skew

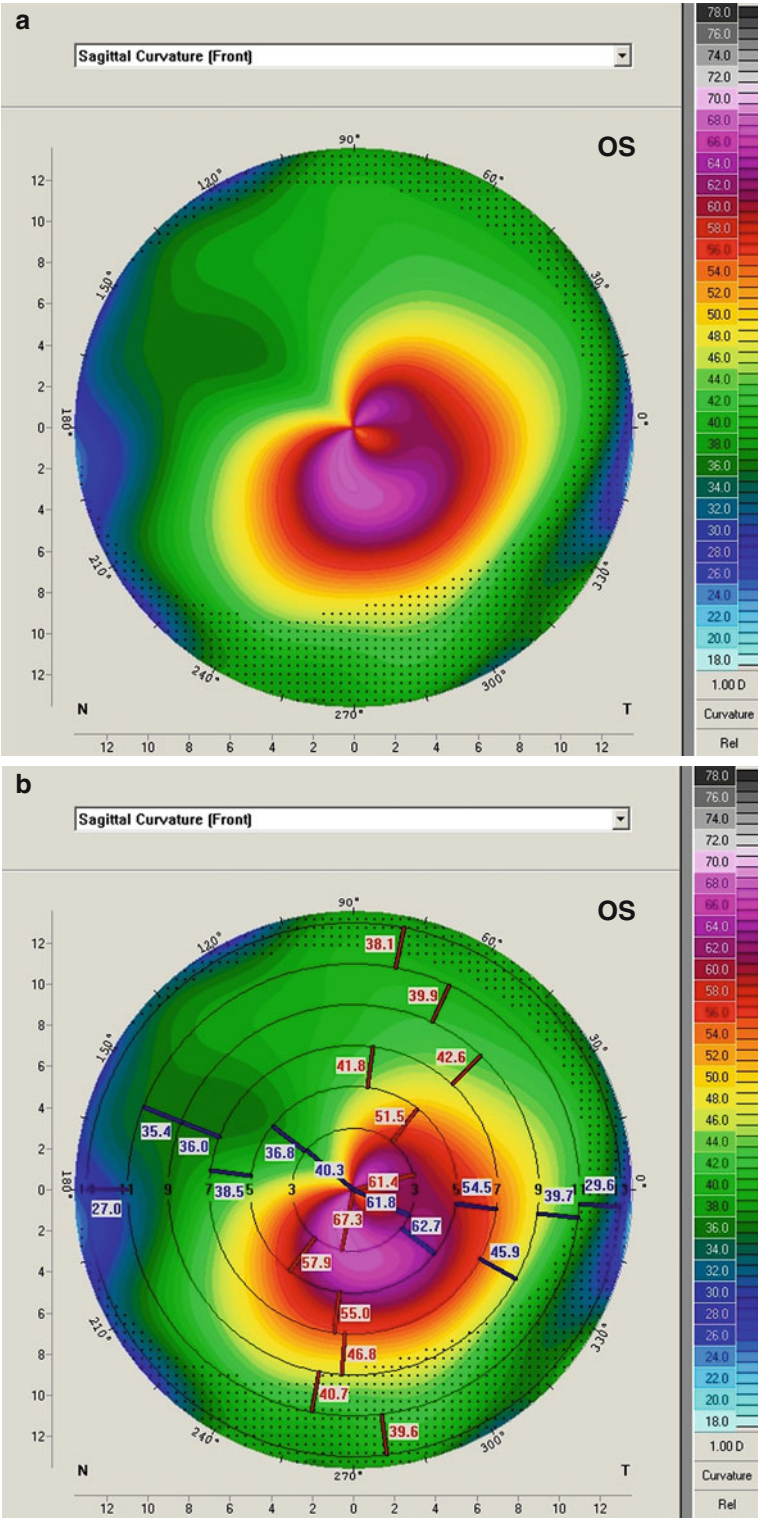


Fig. 2.27 Butterfly. The Bow-tie is horizontally aligned with wing-like spread of the lobes

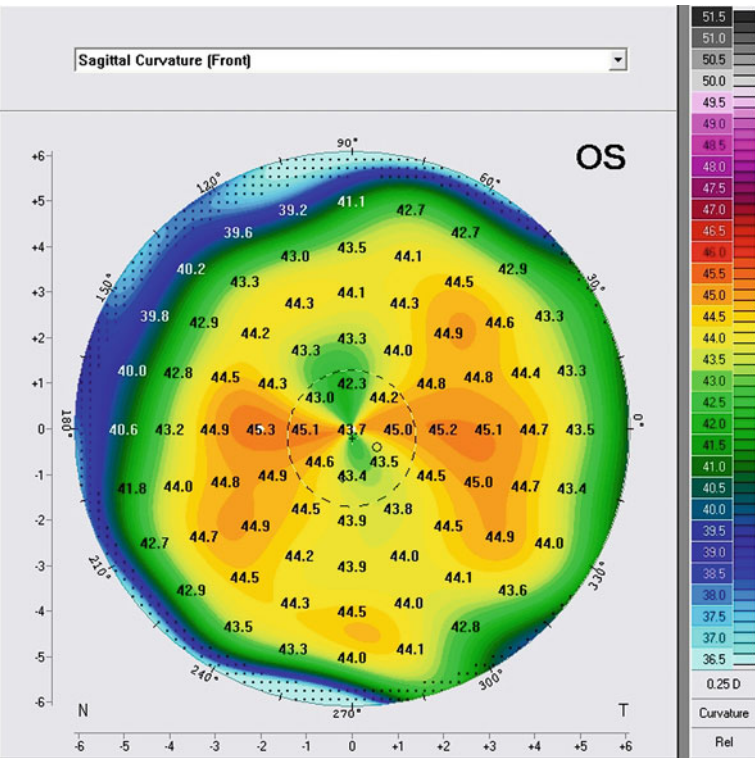


Fig. 2.28 Claw pattern or the kissing birds pattern. The lobes of the bow-tie or the wings of the butterfly are inferiorly joined

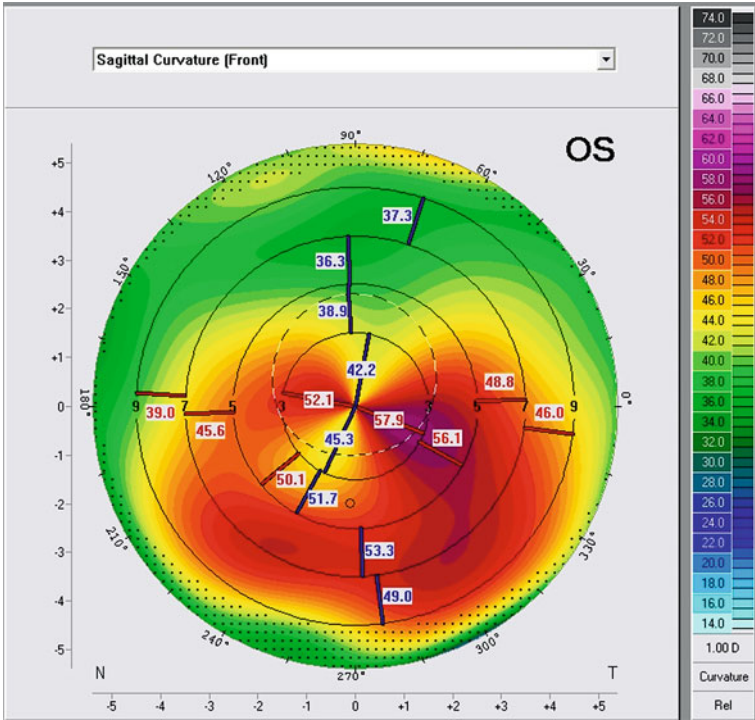


Fig. 2.29 Junctional pattern. KC may present with this pattern. Junctional pattern is better seen with the projected circles and curvature segments off. (a) With and (b) without

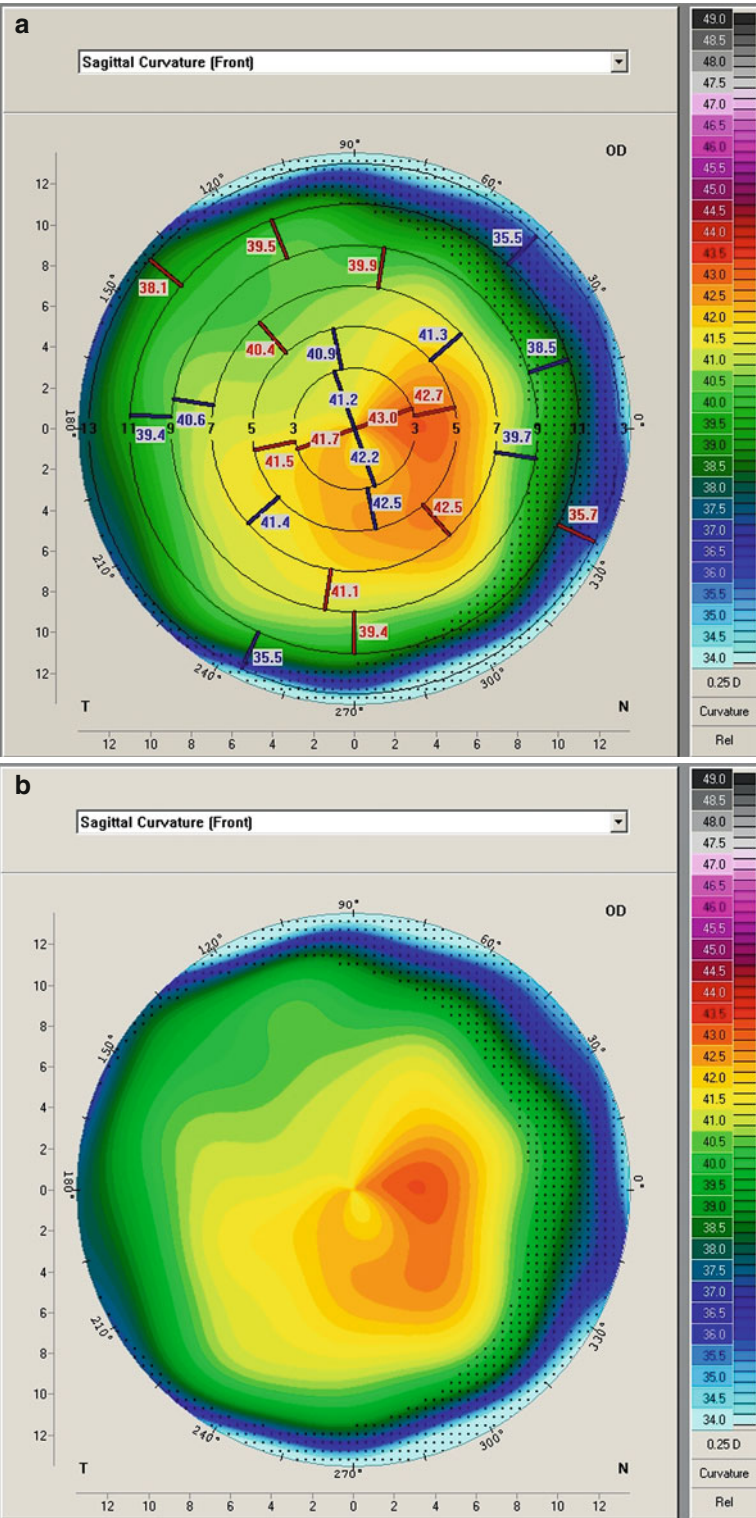


Fig. 2.30 Smiling face. KC may present with this pattern

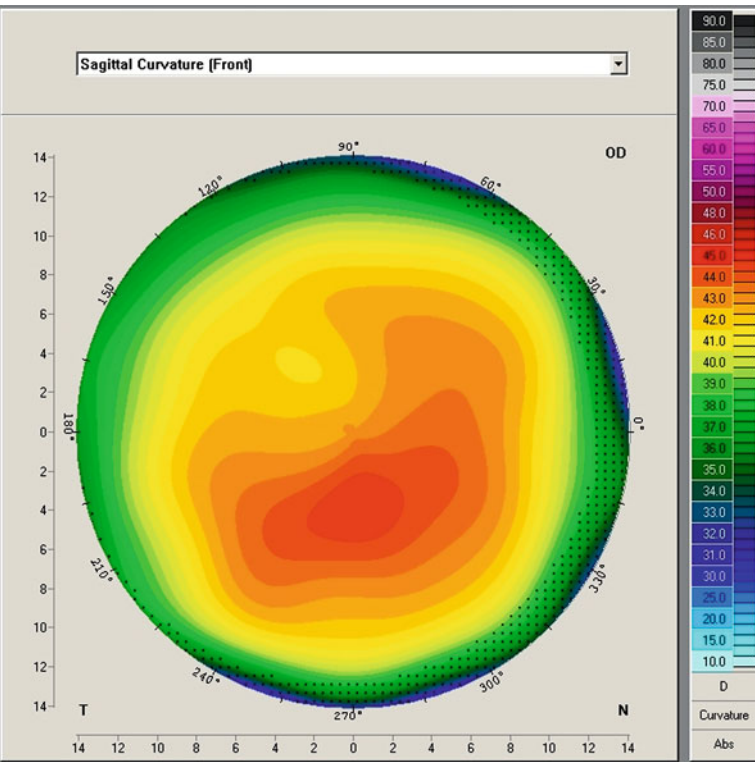


Fig. 2.31 Vortex pattern. The projected curvature (red and blue) segments take a vortex distribution. (a–c) Different shapes of the vortex pattern. Unlike the junctional pattern, the vortex pattern is better recognized with the projected curvature segments on, this can be seen when comparing (d) with (a–c)

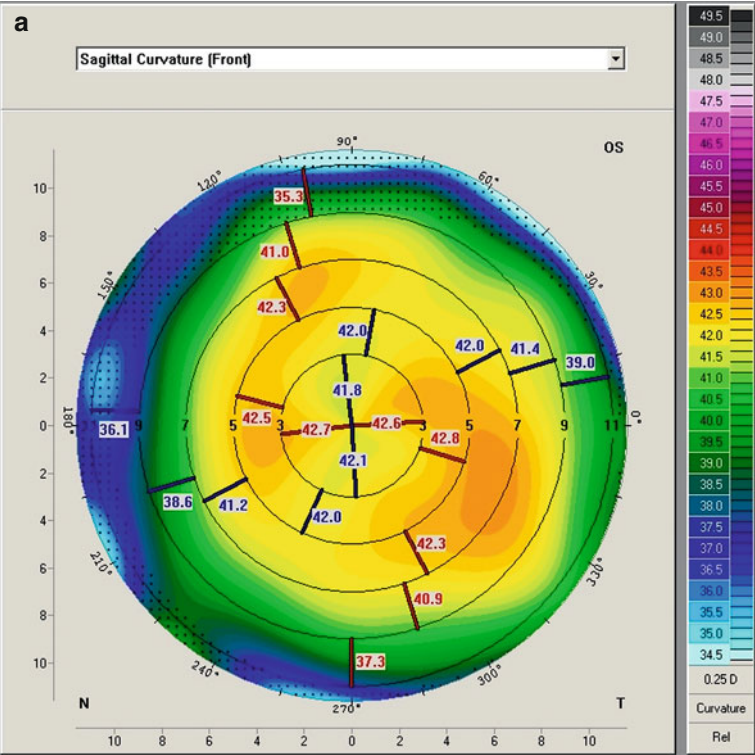


Fig. 2.31 (continued)

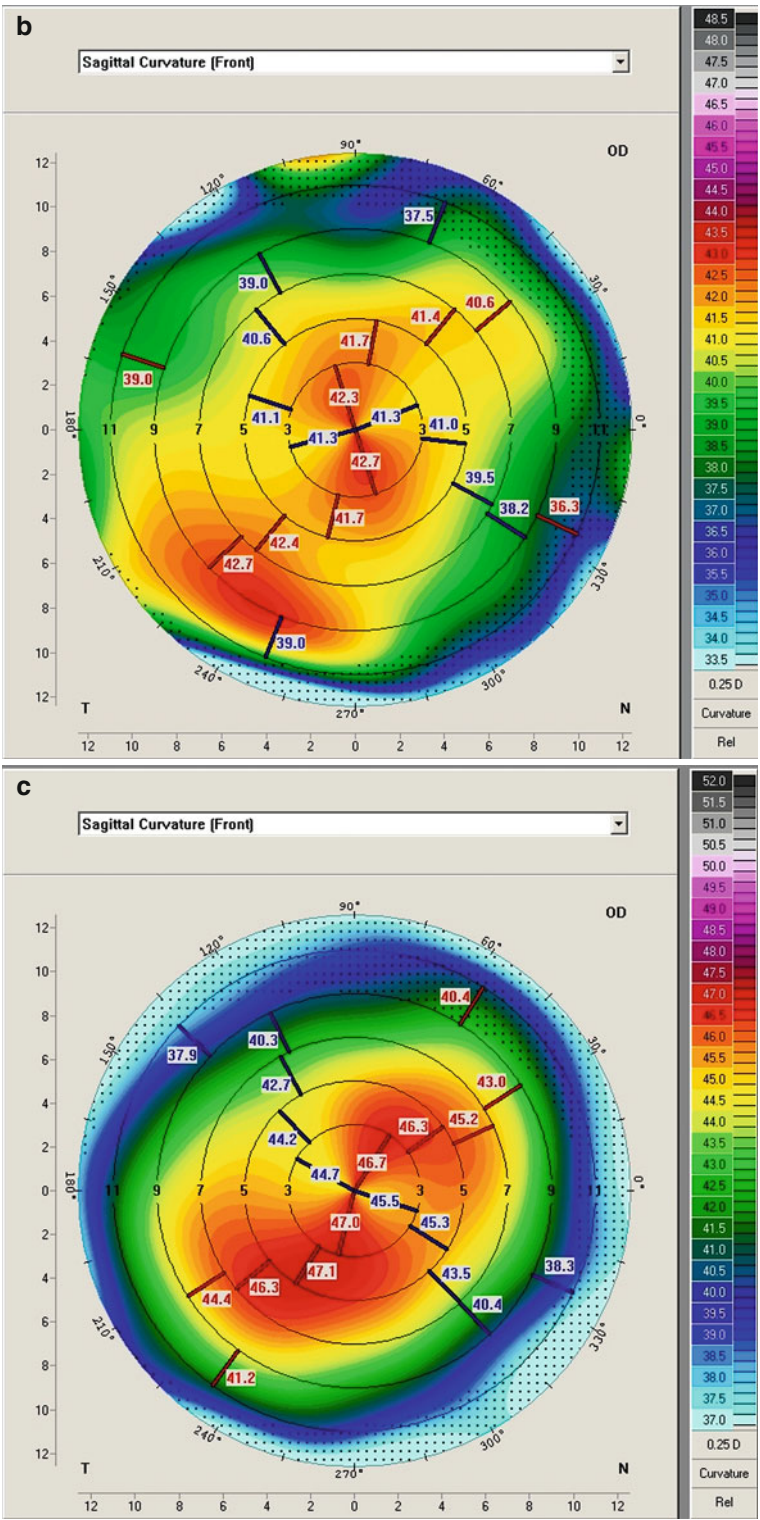
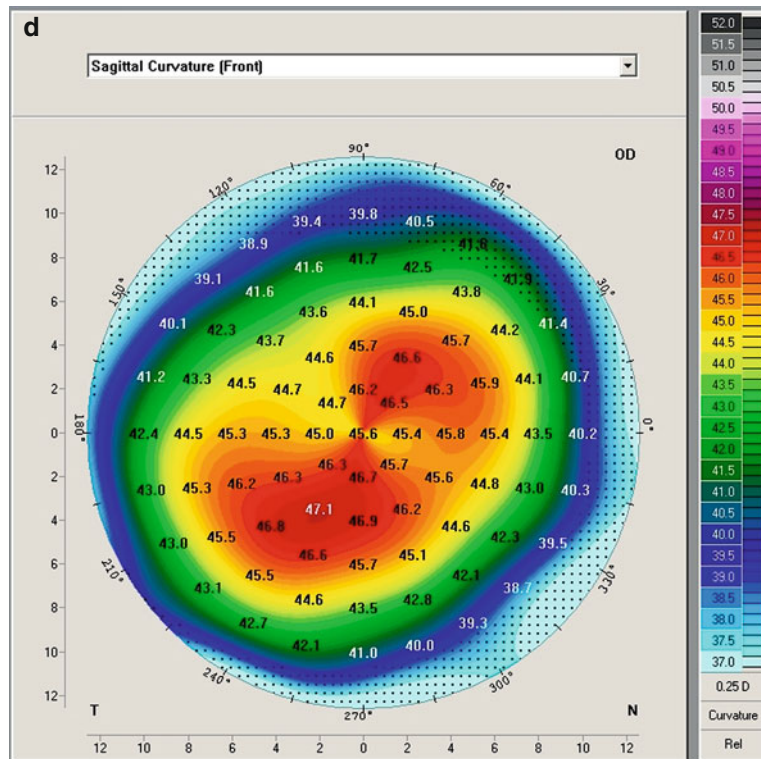


Fig. 2.31 (continued)

the map, then the size and the axis of the upper and the lower segments of the bowtie are studied.

The author finds it useful to classify the curvature map of KC into seven patterns as follows:

1. Pattern 1: The inferior steep pattern, where the inferior segment of the bowtie is steeper (larger) than the superior segment, with the axes of the central parts of these segments straight (Fig. 2.32).
2. Pattern 2: The inferior steep pattern, it is like pattern 1 except that there is a more than 22° of skew between the two axes (Fig. 2.33).
3. Pattern 3: Both segments of the bowtie are equal in size and have straight and aligned axes (Fig. 2.34).
4. Pattern 4: The two segments are equal in size but there is more than 22° of skew between the two axes (Fig. 2.35).
5. Pattern 5: It is PMD or Pellucid-like KC (PLK) with straight axis (Fig. 2.36). PLK will be discussed later in details.
6. Pattern 6: It is PMD or PLK with more than 22° of skew between the two axes (Fig. 2.37).
7. Pattern 7: Where the cone is eccentric and the steep and flat axes are difficult to identify (Fig. 2.38a, b).

The importance of this classification will be clear when talking about the intracorneal rings.

2.3 Krumeich Classification of Keratoconus

Severity of KC is also classified by Krumeich. This classification depends on mean K-readings on the anterior curvature sagittal map, thickness at the thinnest location, and the refractive error of the patient. Table 2.1 demonstrates grading of KC severity, where grade 4 is the worst. This classification is useful; it helps choosing the best approach for KC.

There might be some intersection between the categories, such as >55 dpt of Km (grade 4) and 400 μ thickness (grade 2). In such cases, a full judgment should be followed, which is the aim of this book.

2.4 Forme Fruste Keratoconus

Forme Fruste Keratoconus (FFKC) is a subclinical disease and is not a variant of KC. Although clinicians use many other terms such as mild KC, early KC, and subclinical KC, their exact meanings and applications are less certain. These terms are not universally accepted. The diagnosis of KC is a clinical one that is aided by topography, while the diagnosis of FFKC is topographic.

Fig. 2.32 Pattern 1 (author's classification). Inferior steep with straight *central red line* (steep axis)

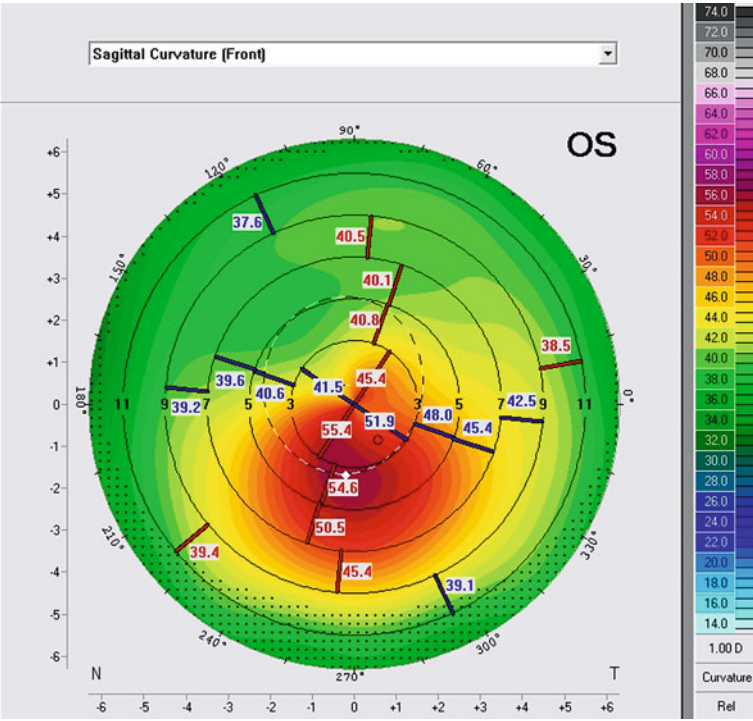


Fig. 2.33 Pattern 2 (author's classification). Inferior steep with skewed *central red line* (steep axis)

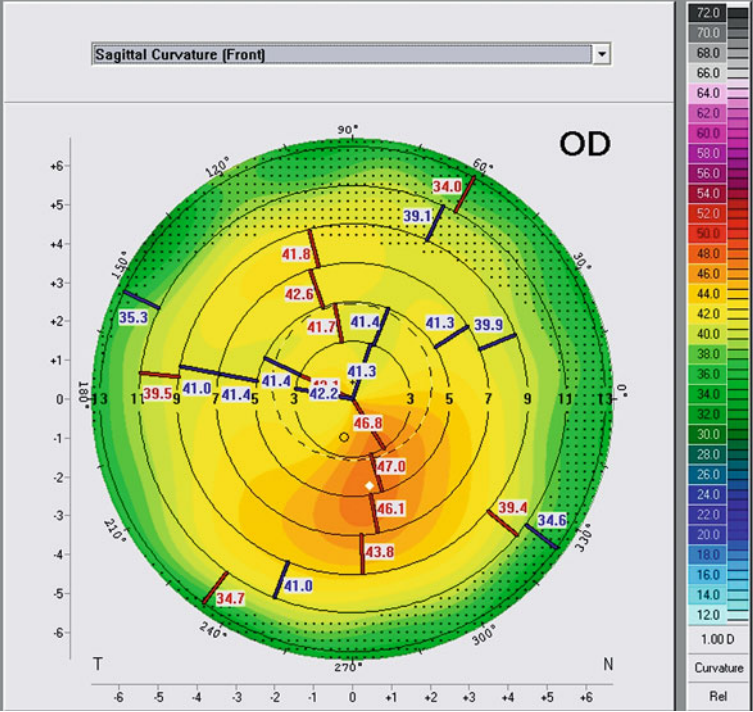


Fig. 2.34 Pattern 3 (author’s classification). Symmetric bowtie with straight central red line (steep axis)

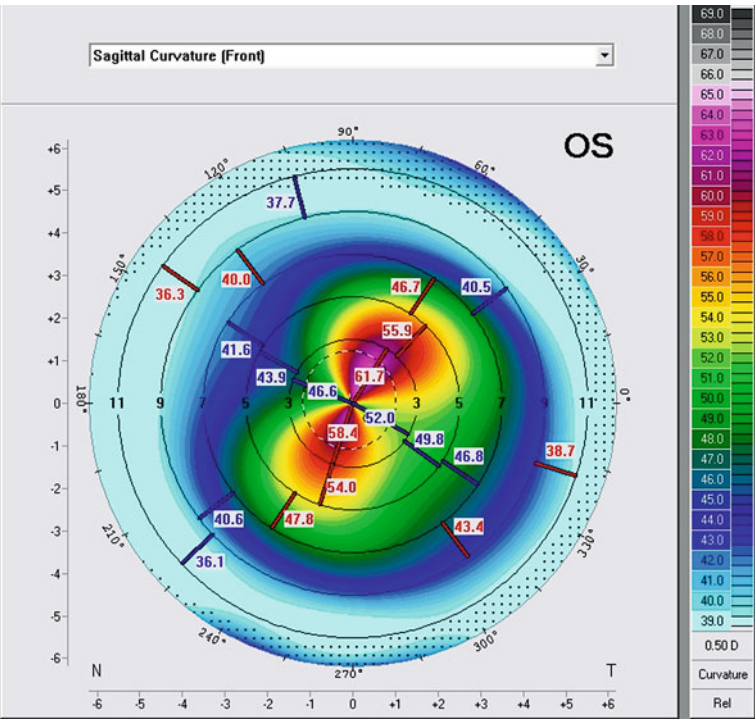


Fig. 2.35 Pattern 4 (author’s classification). Symmetric bowtie with skewed central red line (steep axis)

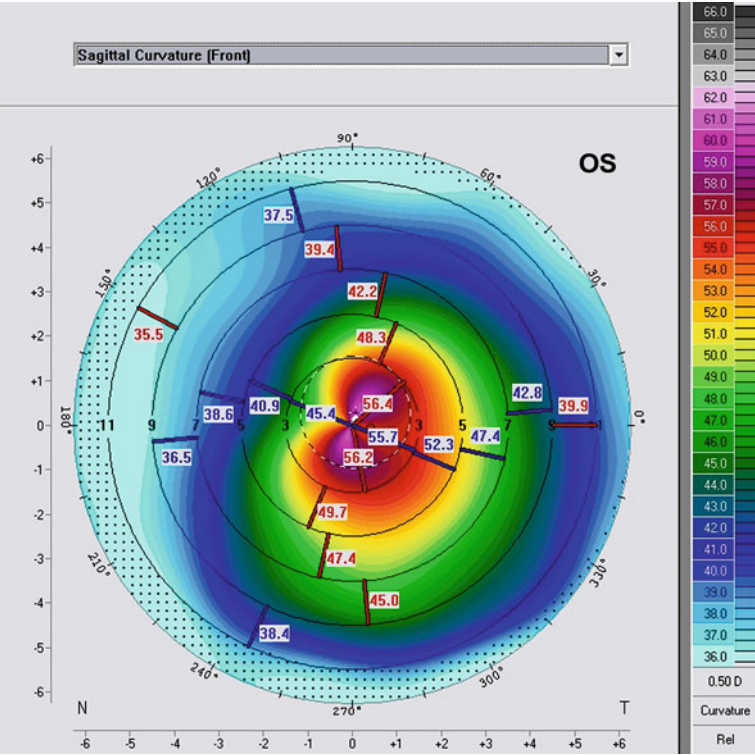


Fig. 2.36 Pattern 5 (author's classification).
PMD or Pellucid-like keratoconus with
straight central red line (steep axis)

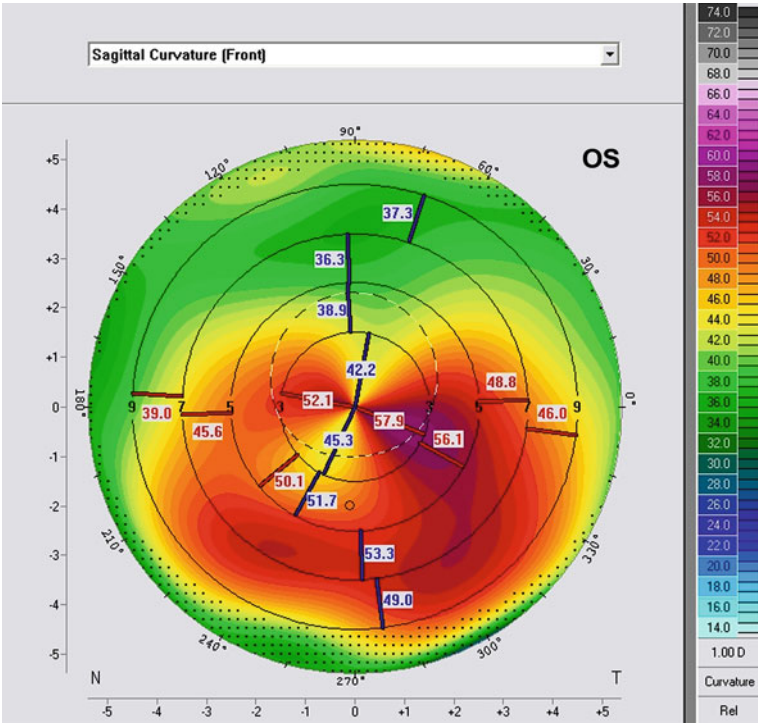


Fig. 2.37 Pattern 6 (author's classification).
PMD or Pellucid-like keratoconus with
skewed central red line (steep axis)

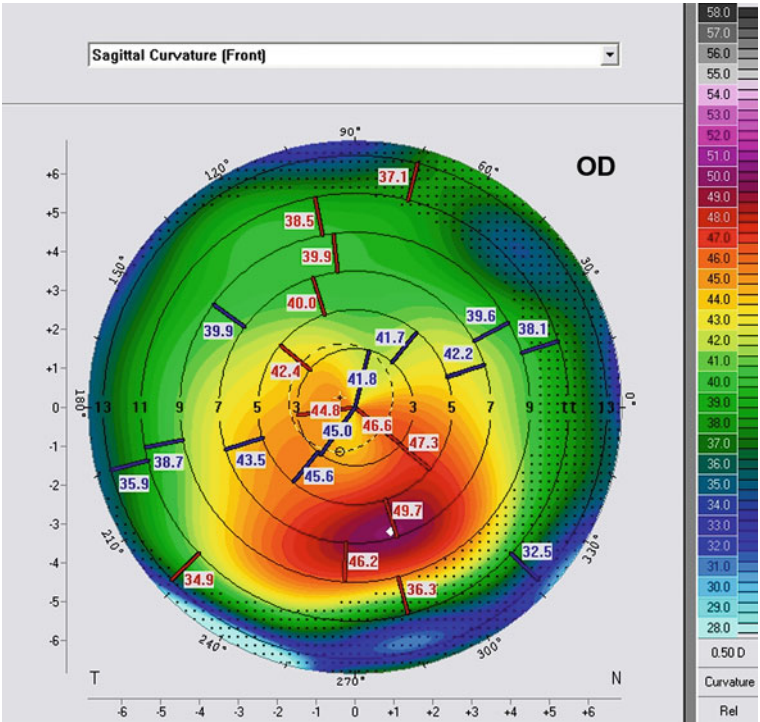


Fig. 2.38 (a, b) Pattern 7 (author's classification). Eccentric cone with the steep and flat axes difficult to identify

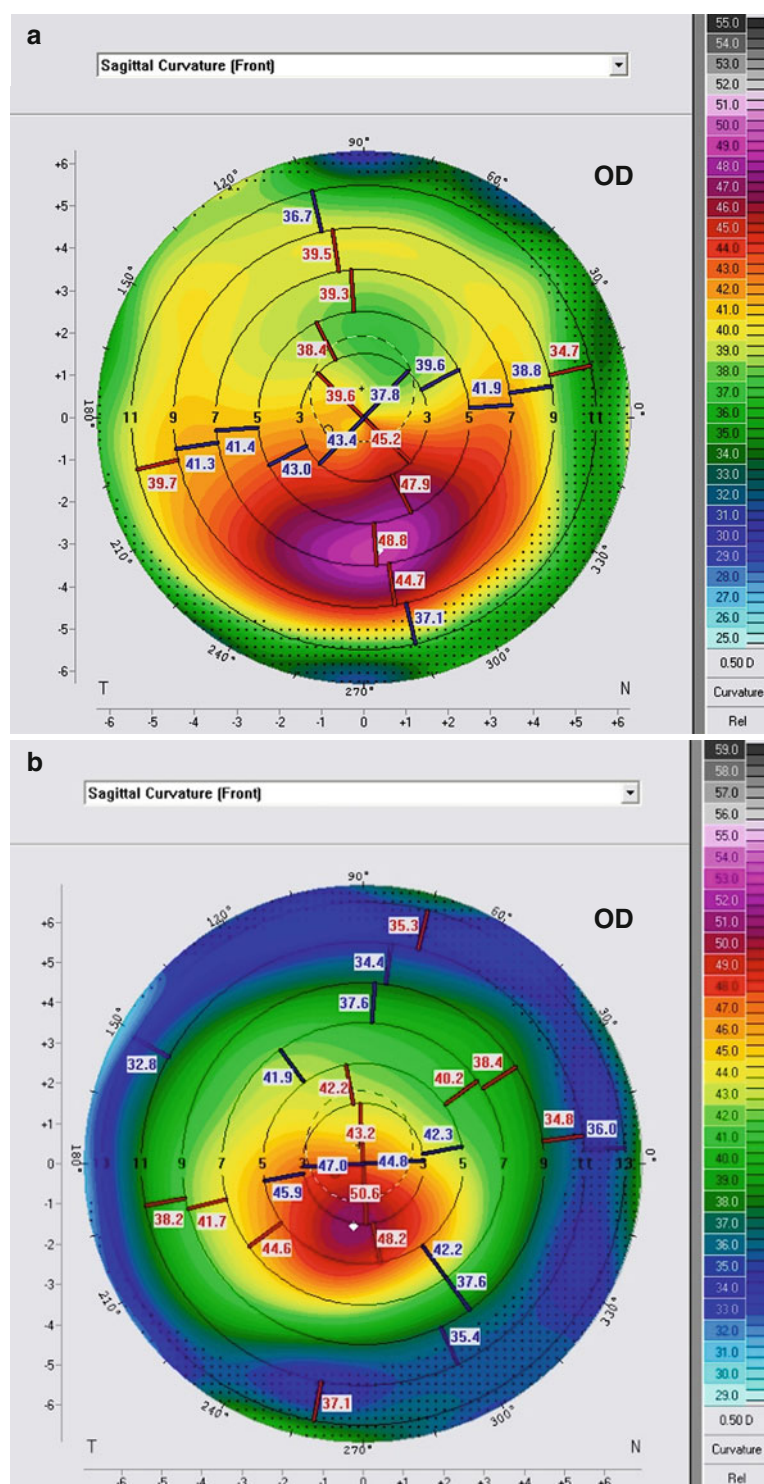


Table 2.1 Krumeich classification of keratoconus

Severity	Km (sim K)	Thickness (μ)	Spherical equivalent	Cornea
4	>55	<200	Not measurable	Central scars
3	54–55	200–400	>–8D	No central scars
2	48–53	400–500	[–5, –8]D	No central scars
1	<48	>500	<–5	No central scars

Recently, there are two opinions regarding the definition of this disease:

1. FFKC is a completely normal cornea with neither clinical nor topographical risk factors, but this cornea is able to develop KC when treated by laser. The fellow eye may be keratoconic or there may be a family history of KC as shown in Fig. 2.39a–c. Figure 2.39a represents a relatively normal corneal topography of the left eye. Figure 2.39b represents

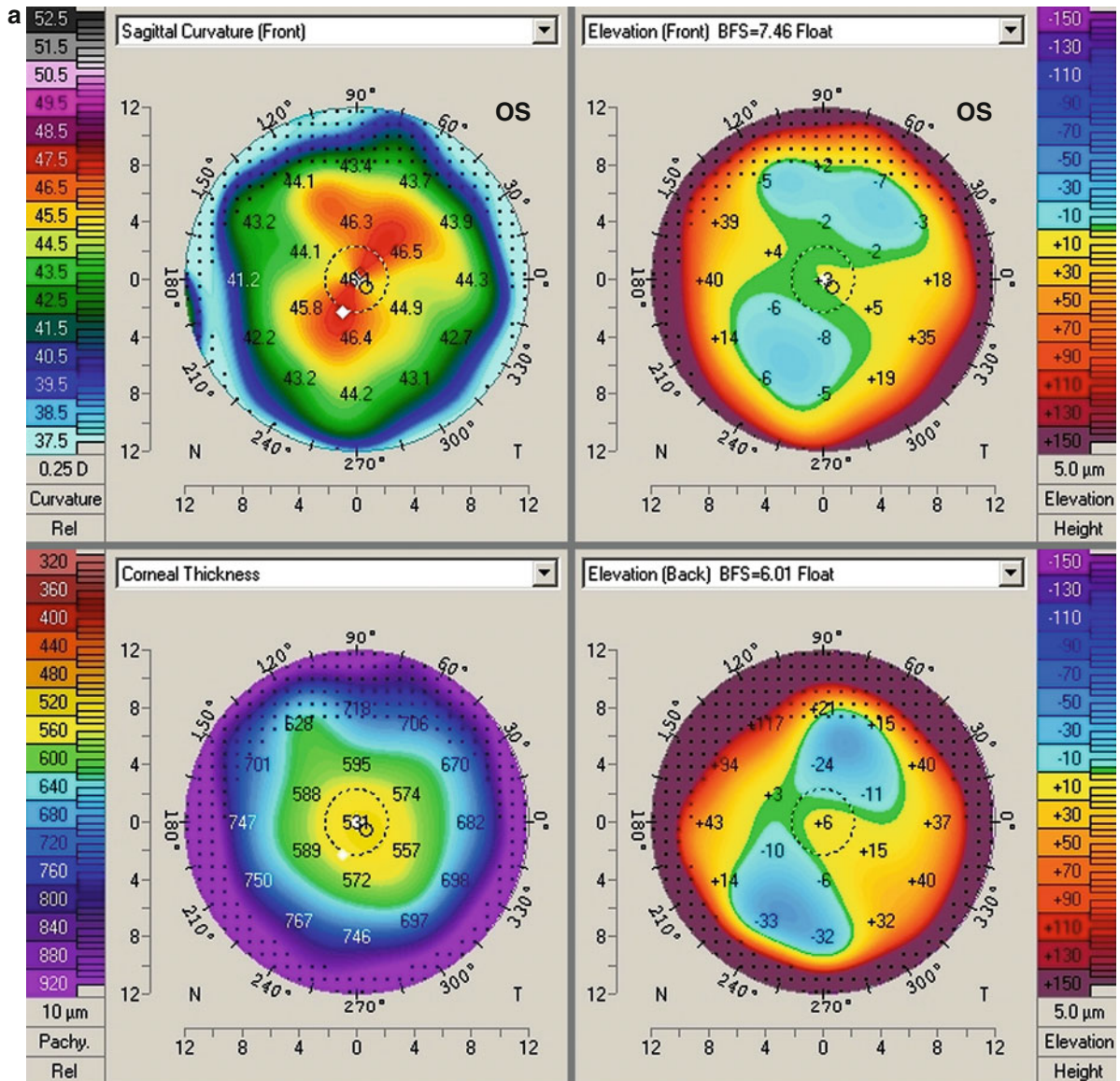


Fig. 2.39 Corneal topography of FFKC. (a) corneal topography of the left eye which is defined as having FFKC, (b) corneal topography of the right eye of the same patient, it is very irregular

and can be considered KC, (c) corneal topography of the patient's brother who has frank KC

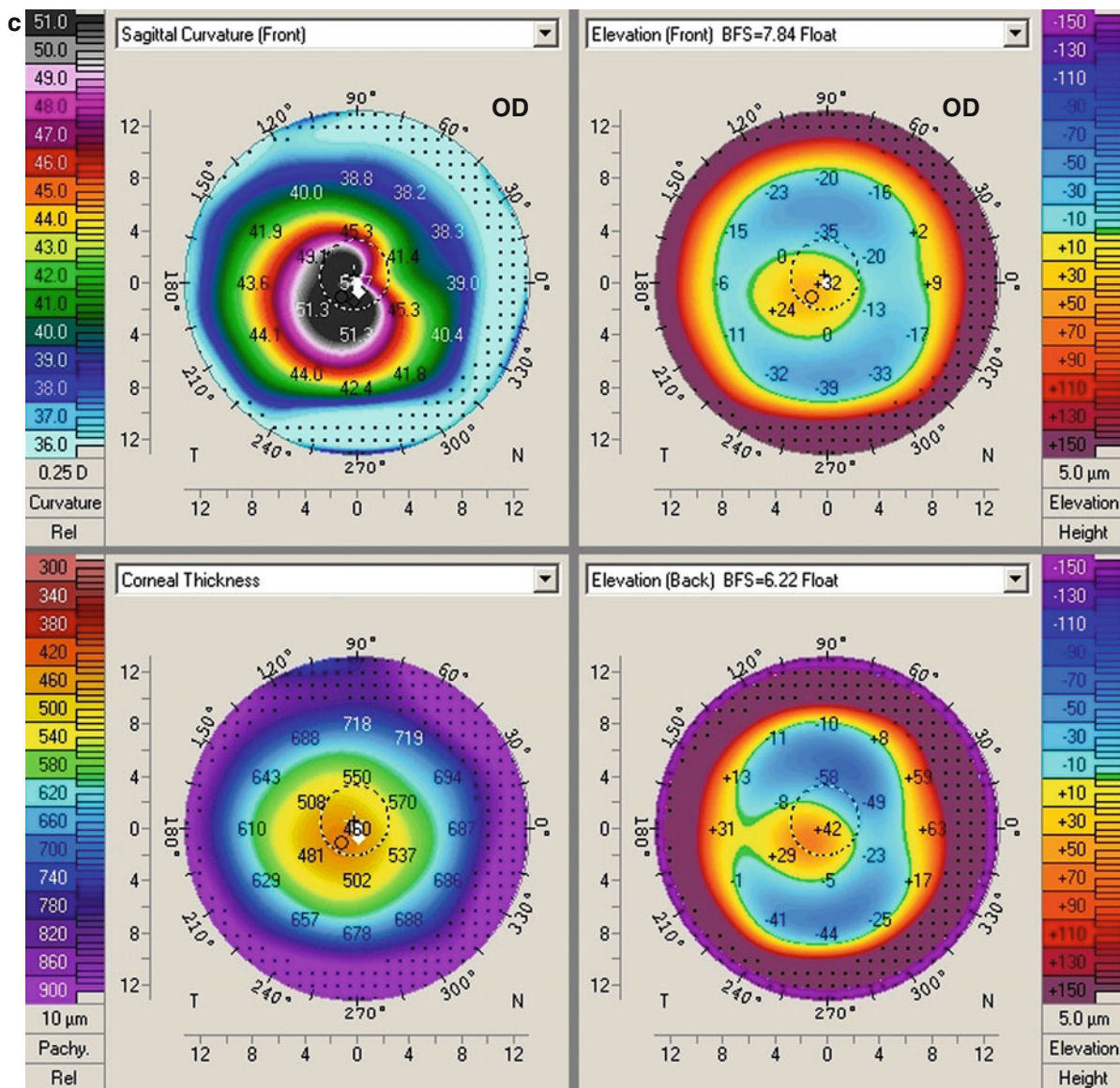


Fig. 2.39 (continued)

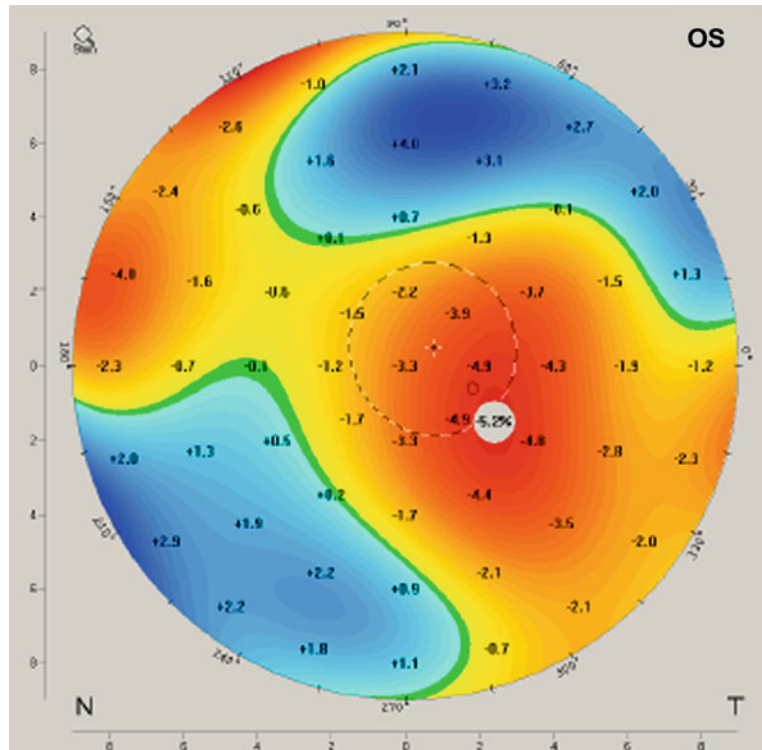
the right eye of the same patient, please note the abnormal and irregular cornea which can be considered as KC. Figure 2.39c is corneal topography of the right eye of the patient's brother, it is a frank KC. According to this definition, the left eye of the patient is FFKC.

2. FFKC is an abnormal cornea. Corneal topography or corneal hysteresis or both are abnormal; i.e., there are risk factors but the case is still not a clinically obvious KC. There are also two opinions regarding this definition:
 - (a) The numerical values of topography and/or corneal hysteresis are in the suspected field

(neither normal nor risky). Readers can refer to author's two books in corneal topography by Jaypee Highlight.

- (b) The shape of corneal topography is taken into consideration. Anterior tangential curvature map, relative pachymetry map, and posterior elevation maps are the key. The tangential map is the most sensitive for determining the geometry of the cornea. Unlike the sagittal map, curvatures on the tangential map are relative to the surface, not to the axial center of the surface. Thus, it shows the exact location of the nipple of the cone. The relative pachymetry map gives the thickness of the

Fig. 2.40 The relative thickness map. Zero values mean that such points match those in the reference normative map. Positive values mean that such points are thicker than those in the reference normative map. Negative values mean that such points are thinner than those in the reference normative map. When the most negative point corresponds with the K-max point on the anterior tangential map and the highest point on the posterior elevation map (BFTE mode), it is FFKC



cornea at a given point relative to the normal thickness at that point as a percentage. For example: +5 and +8 points, i.e., these two points are thicker than the zero point by 5% and 8% respectively, and vice versa when the values are negative. A standard value study was carried out on untreated corneas to adopt the normal average values as a standard reference map. So, when the computer matches the relative map of an abnormal cornea with the standard reference map, the abnormal locations can be immediately noticed, especially when the cornea is keratoconic. A normal map would be 0% at all points. When there is an abnormal point, it appears with minus value on this map as shown in Fig. 2.40. The back elevation map values are more accurate when using the BFTE reference body rather than the BFS. It is of interest to note that the epithelium of the corneal anterior surface thins over the nipple, reducing the sensitivity of the diagnosis from the front surface alone. When the hot spot on the anterior tangential curvature map, relative pachymetry, and posterior elevation map using the toric ellipsoid are all at the same point, the diagnosis of FFKC is confirmed.

2.5 Pellucid Marginal Degeneration (PMD) and Pellucid-like Keratoconus (PLK)

PMD is a bilateral, non-inflammatory, peripheral corneal thinning disorder characterized by a peripheral band of thinning of the inferior cornea. The cornea in and adjacent to the thinned area is ectatic.

The etiology of PMD has not been clearly established, but collagen abnormalities, as seen in KC, have been reported. No racial preponderance has been identified. An equal distribution exists between the sexes. Patients usually are aged 20–40 years at the time of clinical presentation.

2.5.1 Clinical Findings

PMD is usually asymptomatic, except for the progressive deterioration in uncorrected and spectacle-corrected visual acuity caused by the irregular astigmatism induced by the corneal ectasia.

The thinned and presumably weakened cornea may protrude as a result of the positive intraocular pressure.

Episodes of corneal hydrops with resultant pain, as seen in KC, have been reported, but they occur only rarely.

Uncorrected visual acuity is often severely reduced. Visual acuity, as measured by using a pinhole, is close to normal. Refraction and keratometry show against-the-rule astigmatism. Visual acuity typically cannot be restored by using a spherocylindrical combination of lenses. Using rigid gas permeable (RGP) contact lenses are often required to achieve good Snellen acuity, but patients often experience glare and decreased contrast sensitivity despite this. It is unclear if this condition is due to the corneal disease or the contact lens wear.

Both eyes are usually affected, but the degree of involvement may be asymmetric.

Results of slitlamp examination are characterized by:

1. A peripheral band of thinning of the inferior cornea from the 4 o'clock position to the 8 o'clock position. This thinning is accompanied by 1–2 mm of normal cornea between the limbus and the area of thinning.
2. Corneal ectasia is most marked just central to the band of thinning. The central cornea is usually of normal thickness, and the epithelium overlying the area of thinning is intact.
3. The area of thinning typically is epithelialized, clear, avascular, and without lipid deposits.
4. On careful slitlamp evaluation, prominent lymphatics often are detected at the inferior limbus parallel to the area of thinning.
5. Vertical striations at the level of the Descemet's membrane (similar to the Vogt striae) may be seen in rare instances.
6. The light slit becomes very narrow abruptly in the inferior part of the cornea which is the hallmark of the disease (Fig. 2.41).
7. Fluorescein pattern with the RGP lens: there is an inferior touch between the cornea and the lens as shown in Fig. 2.42. In the same figure, the Placido rings are distributed in a vertically oriented oval due to the against-the-rule astigmatism. Note that the rings become very thin and close to each other in the inferior cornea while they are relatively broader and not crowded in the superior part of the cornea, this is due to the inferiorly displaced cone characterizing PMD.

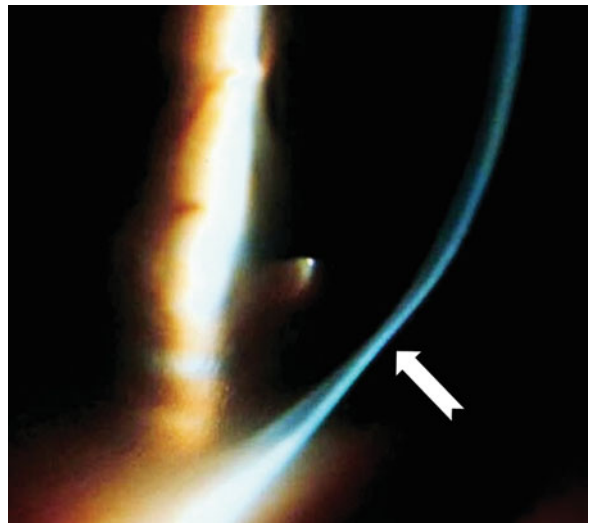


Fig. 2.41 Slitlamp view of PMD, the light slit becomes very narrow abruptly in the inferior part of the cornea (white arrow) which is the hallmark of the disease

2.5.2 Topographical Findings

Identifying features of PMD on corneal topography is very important. That is because there is some similarity between PMD and PLK on corneal topography especially in early stages of PMD. This similarity leads doctors to misinterpret PLK as PMD. Careful studying of the topography reveals many differences between these two entities. Features are mainly seen on the curvature, elevation and thickness maps, and on the keratoconus curve diagram.

- **Curvature map:** The anterior sagittal curvature map is extremely useful in detecting and diagnosing early disease, which may not be readily detectable on slitlamp evaluation. It shows low corneal power along the central vertical axis, increased power as the inferior cornea is approached, and high corneal power along the inferior oblique meridians. This topographical pattern takes a butterfly appearance, as shown in Fig. 2.43. These features are seen in both PMD and PLK.
- **Elevation maps:** There are two important things related to each others that can be identified on the elevation maps, mainly the anterior elevation map, the location of the cone and the “kissing birds” sign. Figure 2.44 shows the “kissing birds” sign. When the cone is peripheral, this sign is present; when the cone is central or paracentral, this sign is absent (Figs 2.44 and 2.45). This sign appears when the

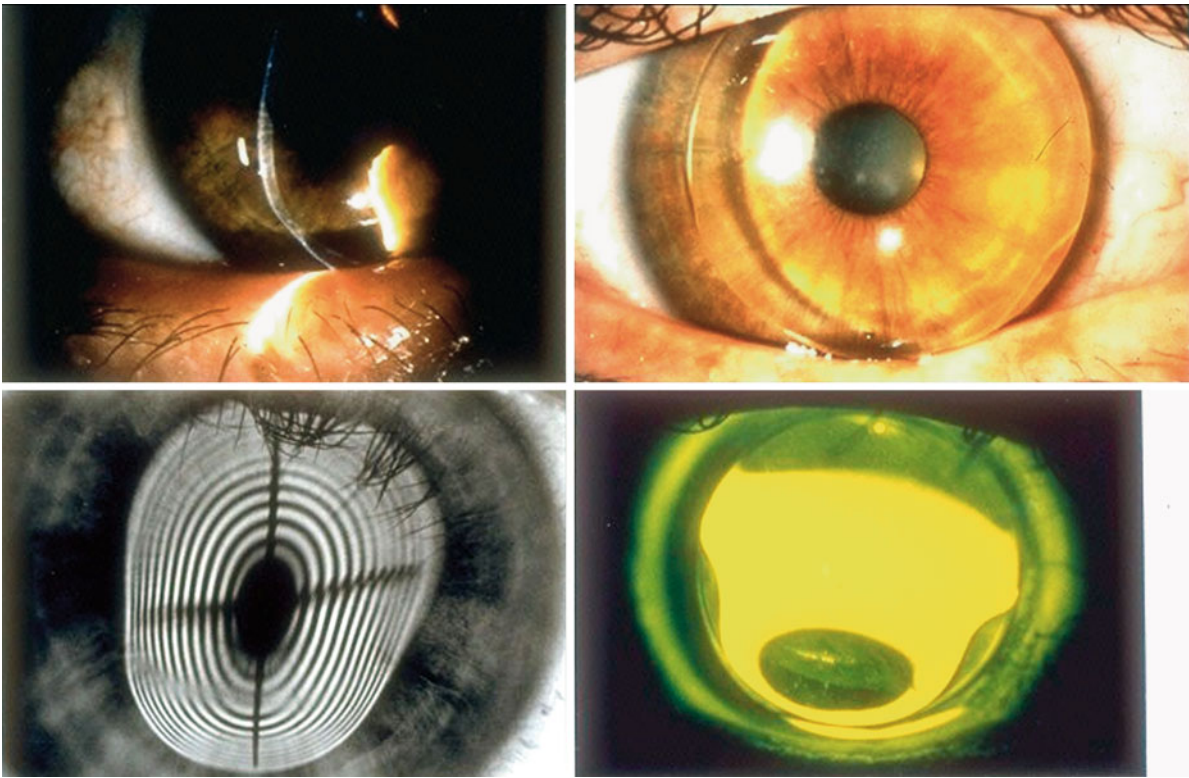
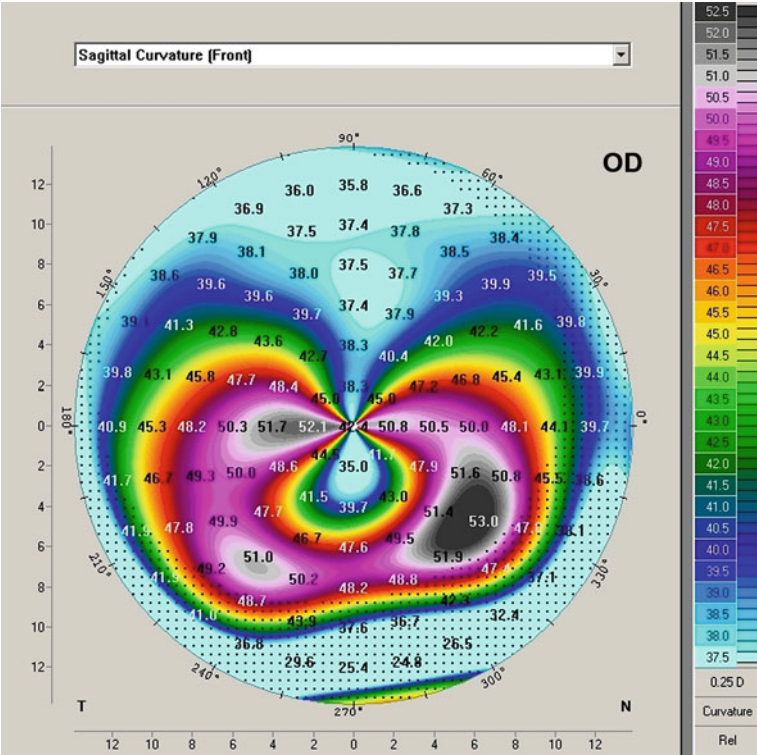


Fig. 2.42 PMD. The *upper left image* is the slitlamp view demonstrating the inferior thinning. The *upper right image* is an RGP lens. The *lower left image* is Placido image; note the vertical oval distribution of the mires the inferior touch. The *lower right image* is the fluorescein pattern, note

Fig. 2.43 Butterfly appearance of PMD and PLK on the anterior sagittal curvature map. Corneal power is low along the central vertical axis, but it increases as the inferior cornea is approached



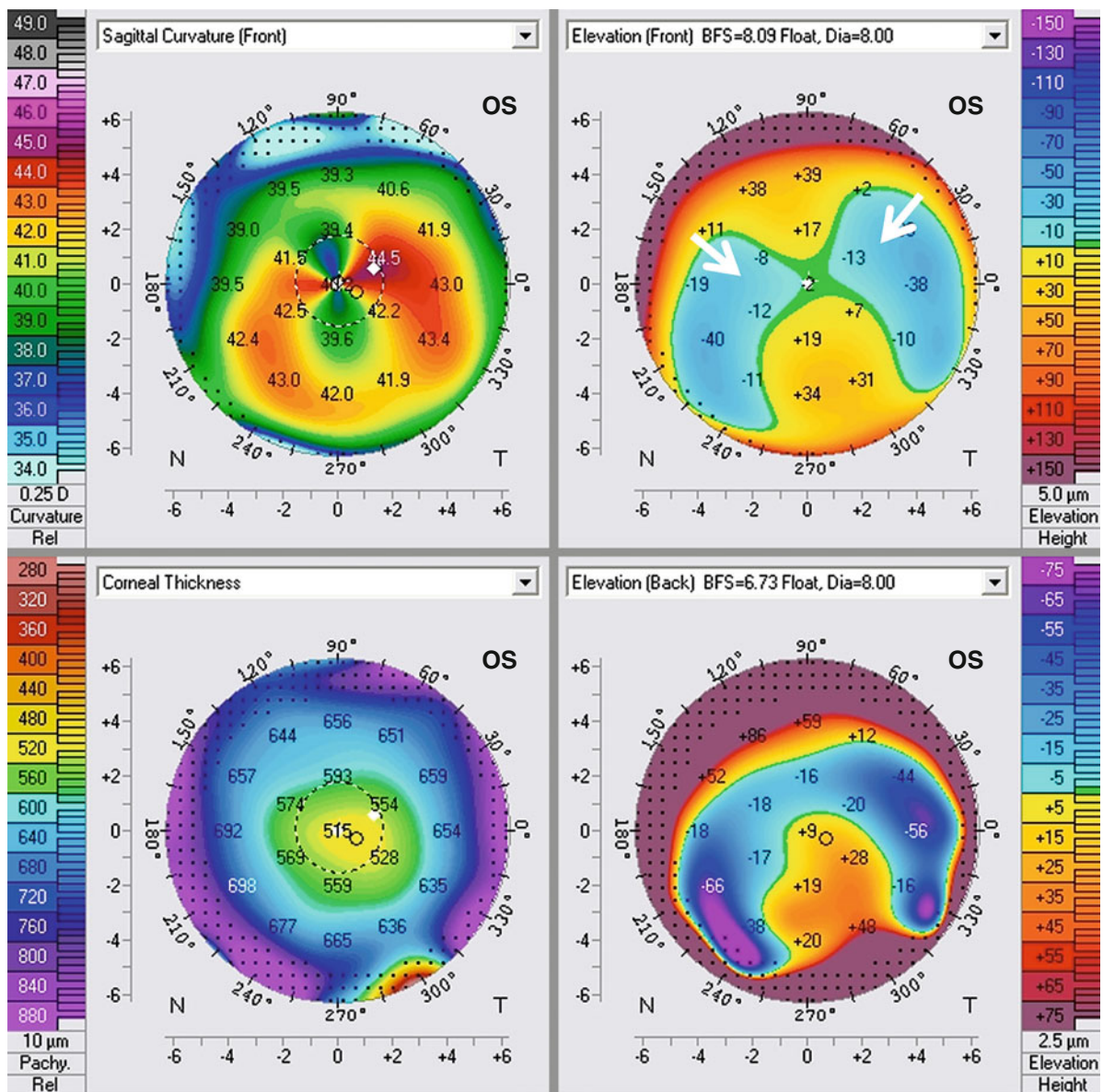


Fig. 2.44 The kissing birds sign on the anterior elevation map (arrow heads). This sign exists when the cone is peripheral

BFS float mode is used (Fig. 2.46), while it disappears when switching to the BFTE float mode (Fig. 2.47). On the other hand, there is no correlation between the cone location and the differentiation between PMD and PLK, i.e., the cone can be central, paracentral, or peripheral in both entities. Therefore, neither the kissing birds sign nor the peripheral cone is a hallmark of PMD. Figure 2.48 is a PMD case without the kissing birds sign; Fig. 2.49 is a PLK case with this sign.

- Corneal thickness map: Corneal thinning is a reversal of the typical pattern in which the cornea

thickens from the center to the periphery. If any part of the peripheral cornea is thinner than the center, this is a cause for concern. In PMD, the corneal thickness map reveals a thinning of the inferior cornea. This thinning is characterized by a special sign that can be called “bell” shape (Fig. 2.50). This sign is a hallmark of PMD; it is absent in PLK. In regard with the location of the thinnest point of the cornea, theoretically, the thinnest point should be inferiorly displaced in PMD and in KC, but the amount of displacement on Y coordinate is much bigger in PMD especially in advanced cases (Figs. 2.51 and

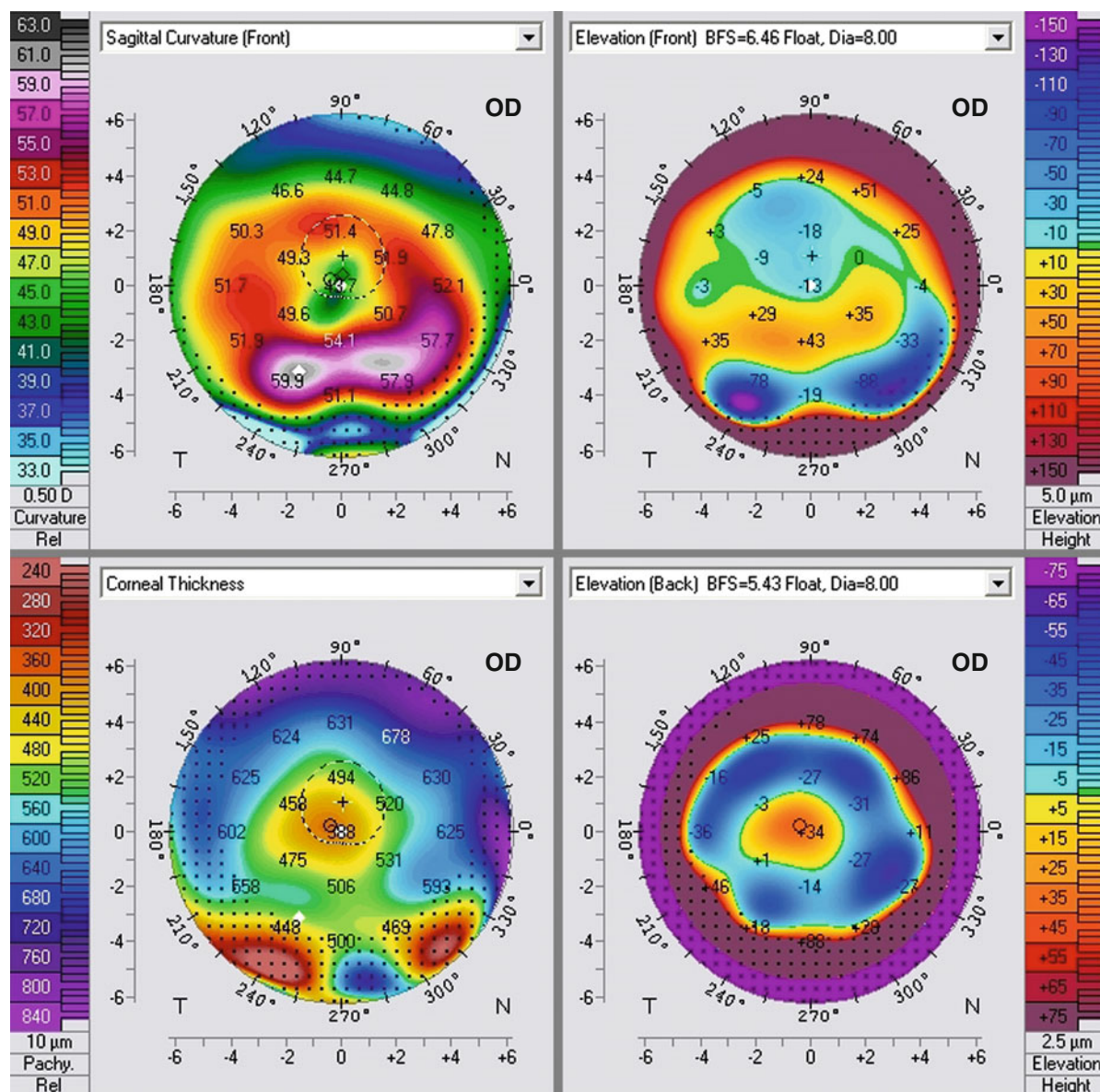


Fig. 2.45 The kissing birds sign. It does not exist when the cone is central or paracentral

2.52: white arrows). This is not always true. In very advanced PMD, the cornea is severely distorted, and the topographers may miss many data from the inferior part of the cornea and may display a proposed plotting with dark dots indicating that an important part of the data is missing in this area. Figure 2.53 is a PMD case as it appears on the slit lamp; notice the very thin area in the inferior cornea and the bulging above. Figure 2.54a and b are the vertical sector of the Scheimpflug image of the

same cornea; notice the very thin inferior part (white arrow). Figure 2.55 is the topography of the case; surprisingly, the thinnest location is relatively not displaced (red arrows), but if we look at the inferior part of the thickness map (black arrow), relatively black dots are present. This point is very important when concerning intracorneal rings or corneal grafts as will be discussed later in details.

- **Keratoconus Curve Diagram:** This curve is an indicator of the gradual change in thickness beginning

Fig. 2.46 The kissing birds sign. It can be seen when the BFS mode is used

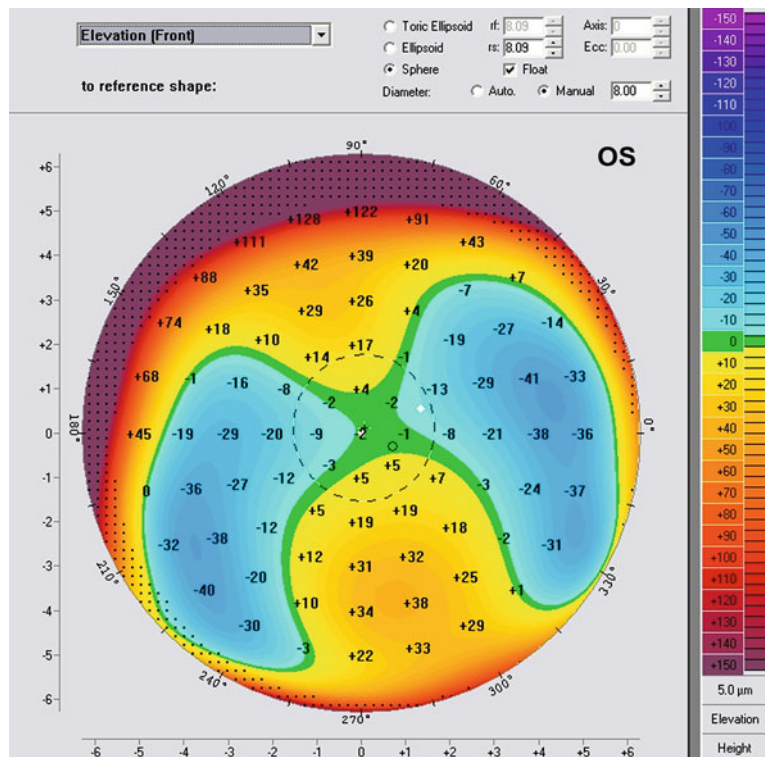
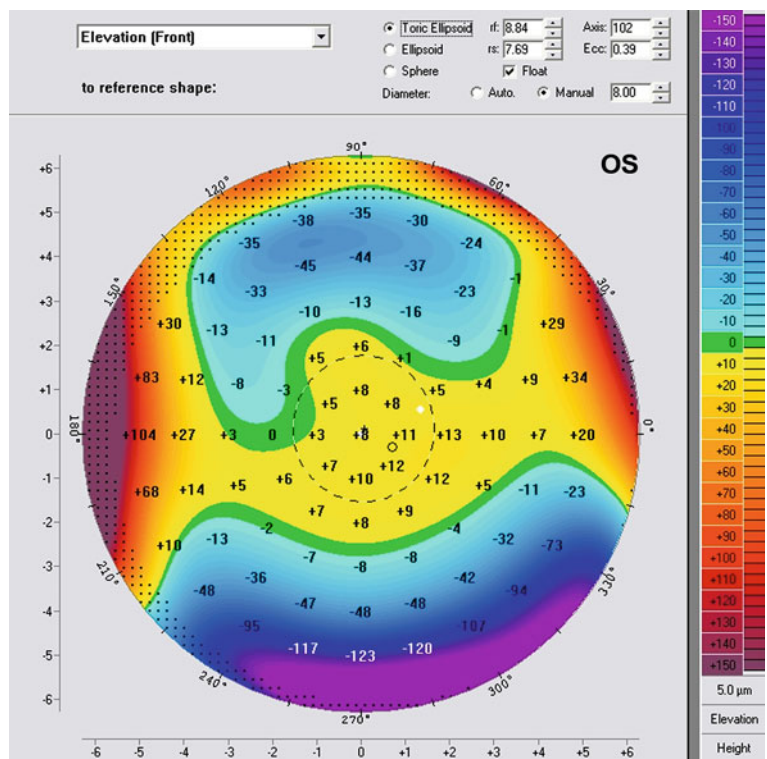


Fig. 2.47 The kissing birds sign. It cannot be seen when the BFTE mode is used



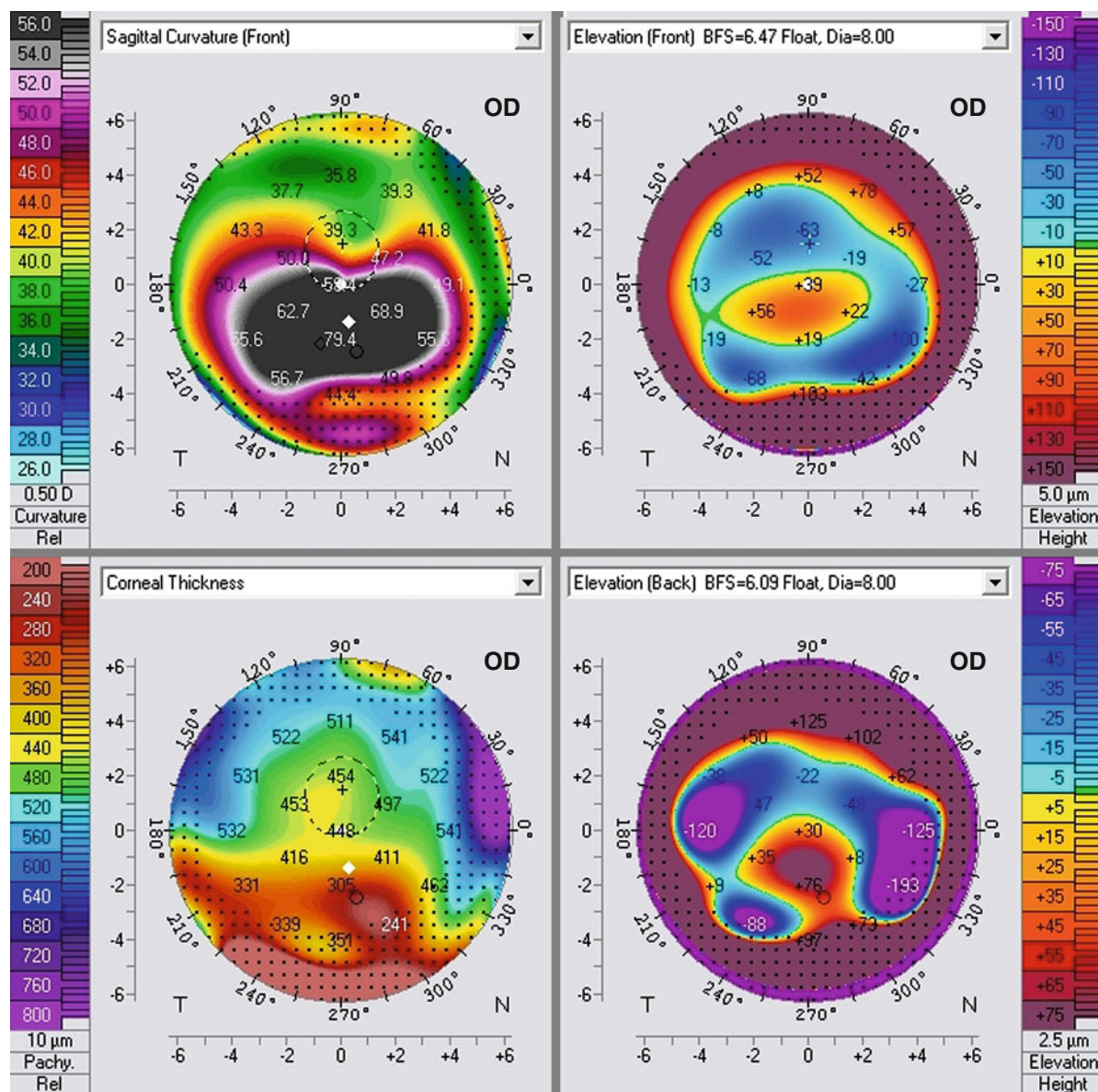


Fig. 2.48 PMD without the kissing birds sign

from the thinnest point toward corneal periphery (for more details about this curve please refer to the book: “Corneal Topography in Clinical Practice, Jaypee Brothers, 2009). In normal corneas, the red line takes the same slope of and lies within the normal range (the black dotted lines) as shown in Fig. 2.56. In KC, PLK and in PMD, this curve deviates from the normal range rapidly (Fig. 2.57). In advanced cases of PMD, the curve usually takes an inverted passage (Fig. 2.58).

Table 2.2 summarizes the difference between PLK and PMD.

2.5.3 Complications

A rare complication in PMD is corneal hydrops similar to that seen in KC, with disruption of the endothelial basement membrane and hydration of the adjacent corneal stroma.

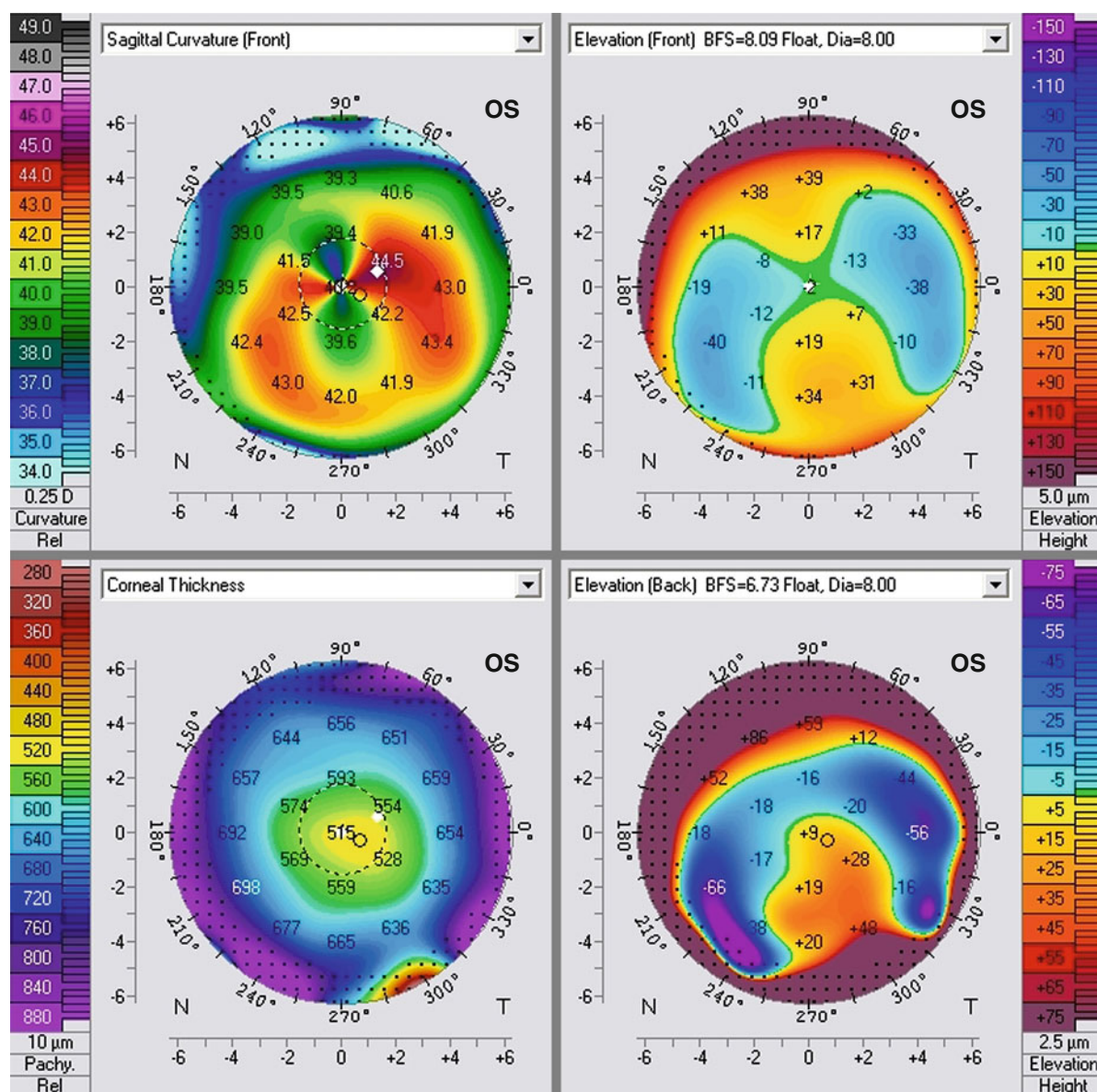


Fig. 2.49 PLK with the kissing birds sign

2.5.4 Differential Diagnosis

1. Keratoglobus: It is a generalized thinning of the cornea (Figs. 2.59 and 2.60).

- The thinning is most marked at the limbus, extending circumferentially for 360°; this makes it different from the globus morphological pattern of the KC.
- The whole cornea protrudes, in contrast to the regional thinning seen in KC and the inferior paralimbal thinning in PMD.

2. Terrien marginal degeneration: It affects an age group similar to that affected by PMD (Fig. 2.61).

- Terrien marginal degeneration can be bilateral.
- Although this condition can be associated with large amounts of astigmatism, it can be differentiated from PMD because the superior cornea is predominantly affected and because the area of thinning is often associated with vascularization and lipid deposition.

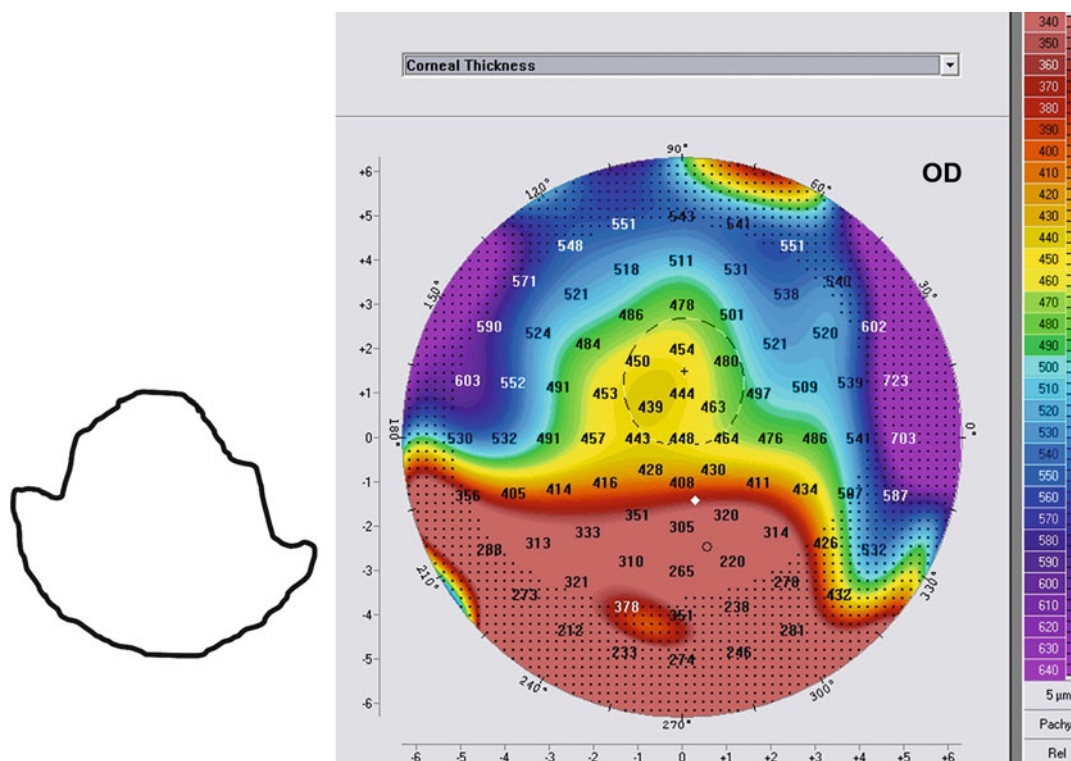


Fig. 2.50 The hallmark of PMD on the corneal thickness map: the bell sign. This sign is due to inferior corneal thinning encountered in PMD

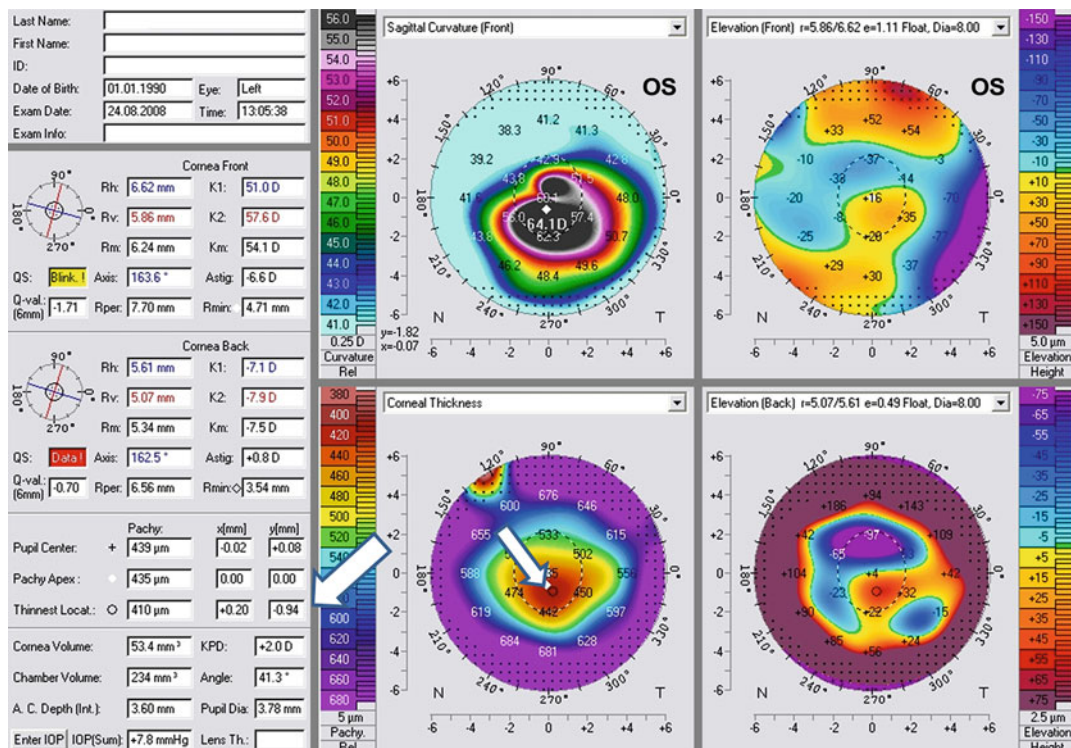


Fig. 2.51 Vertical displacement of the thinnest point on the corneal thickness map in KC. The white arrows point at the symbol of the thinnest location and at the value of displacement on the Y axis

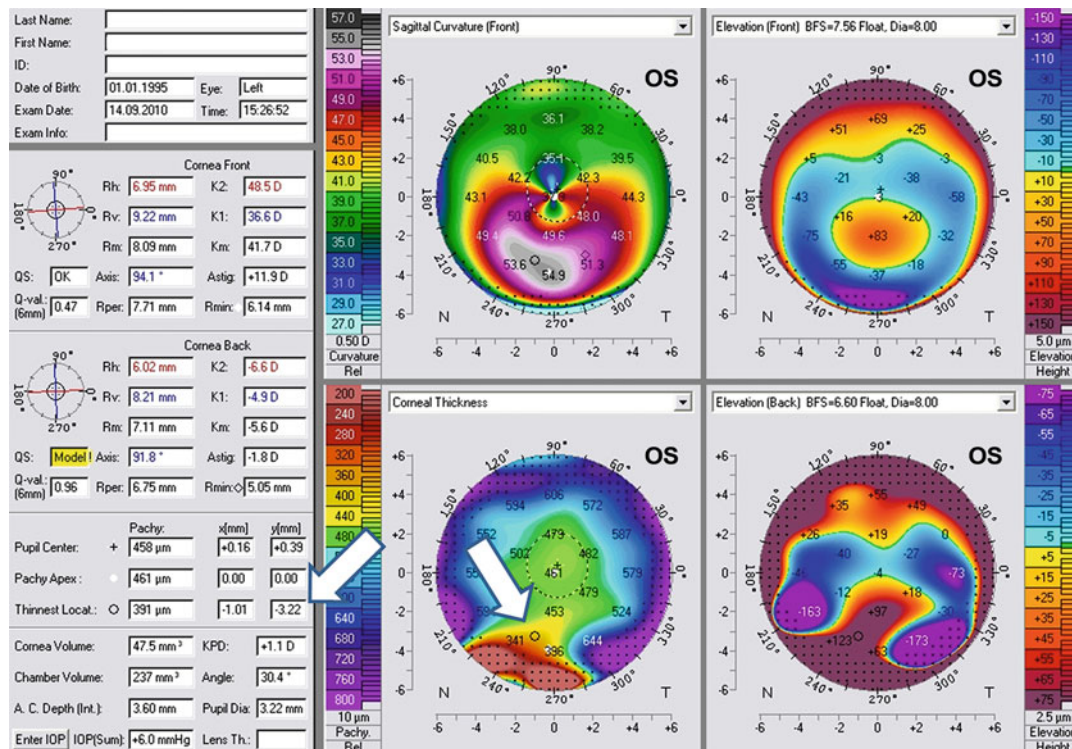


Fig. 2.52 Vertical displacement of the thinnest point on the corneal thickness map in PMD. The *white arrows* point at the symbol of the thinnest location and at the value of displacement on the Y axis

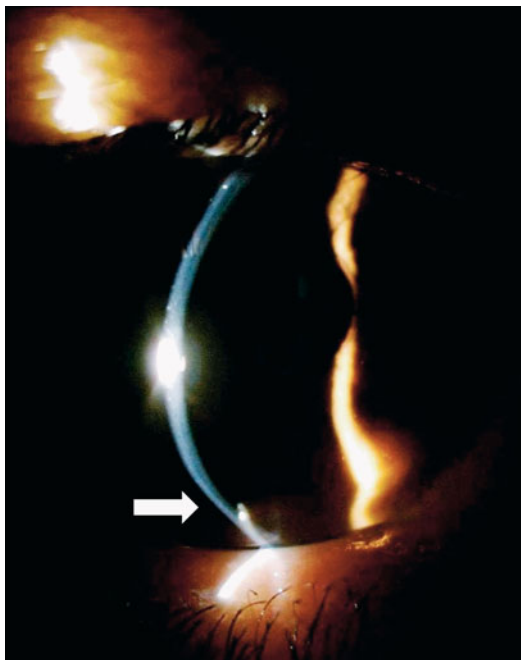
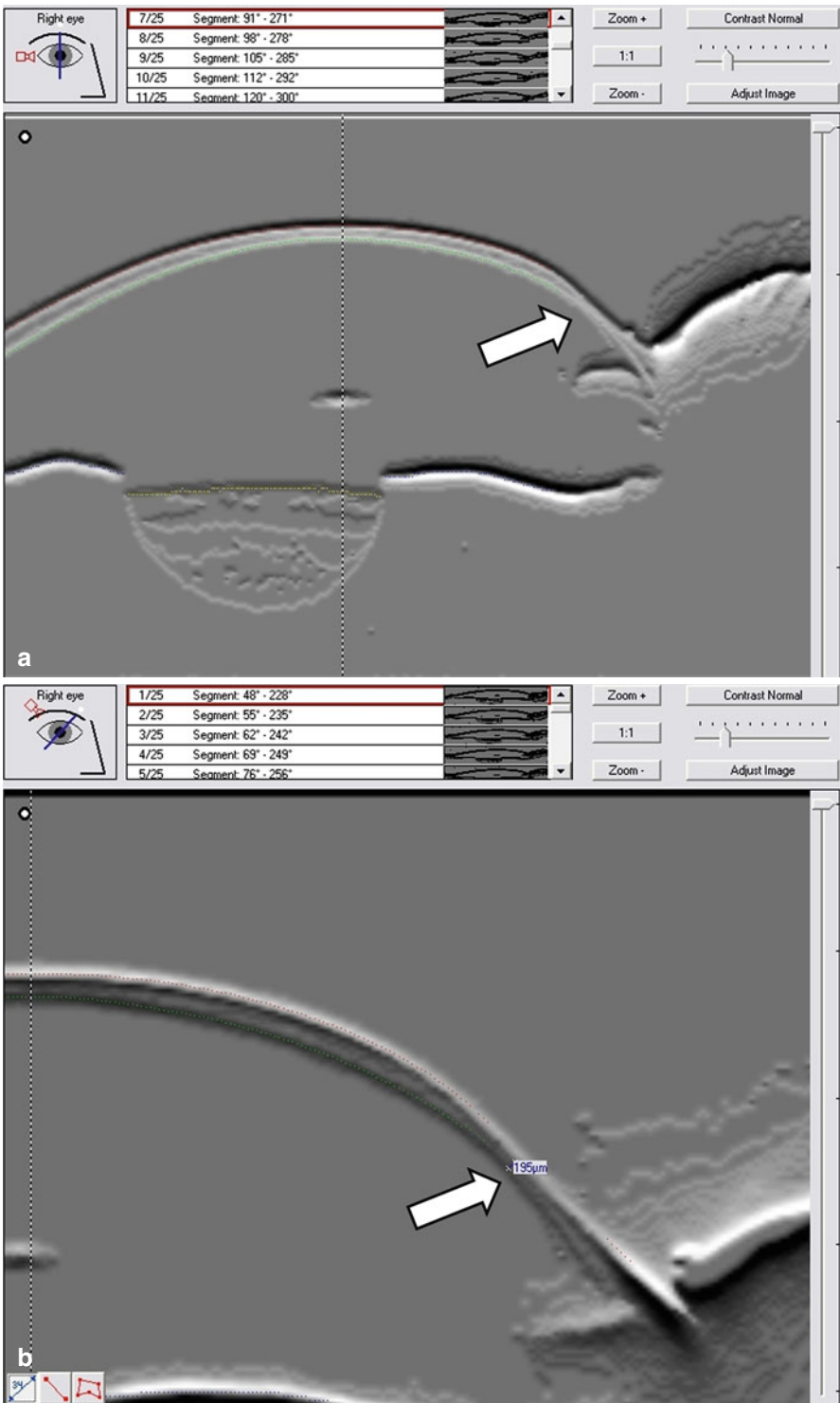


Fig. 2.53 Slitlamp view of a case of PMD. The *white arrow* points at the inferior thin area

3. Furrow degeneration: It has some features of PMD (Fig. 2.62).

- An intact epithelium is present, and the area of corneal thinning is not vascularized, at least in the acute phase.
- The differentiating feature is that the area of thinning is closer to the limbus with virtually no intervening zone of normal cornea, unlike the findings in PMD.
- Furrow degeneration occasionally involves the superior cornea, and an associated adjacent area of scleritis may be present.
- Edges of the furrow are steeper than the gradual attenuation seen in PMD.
- Furrow degeneration occurs adjacent to the lipid deposition in arcus senilis, which is typically observed in elderly patients.
- May be associated with systemic diseases such as rheumatoid arthritis, systemic lupus erythematosus, polyarteritis nodosa, or other collagen vascular diseases.

Fig. 2.54 (a, b) Scheimpflug images of the same case of PMD in the previous figure. The white arrow points at the inferior thin area



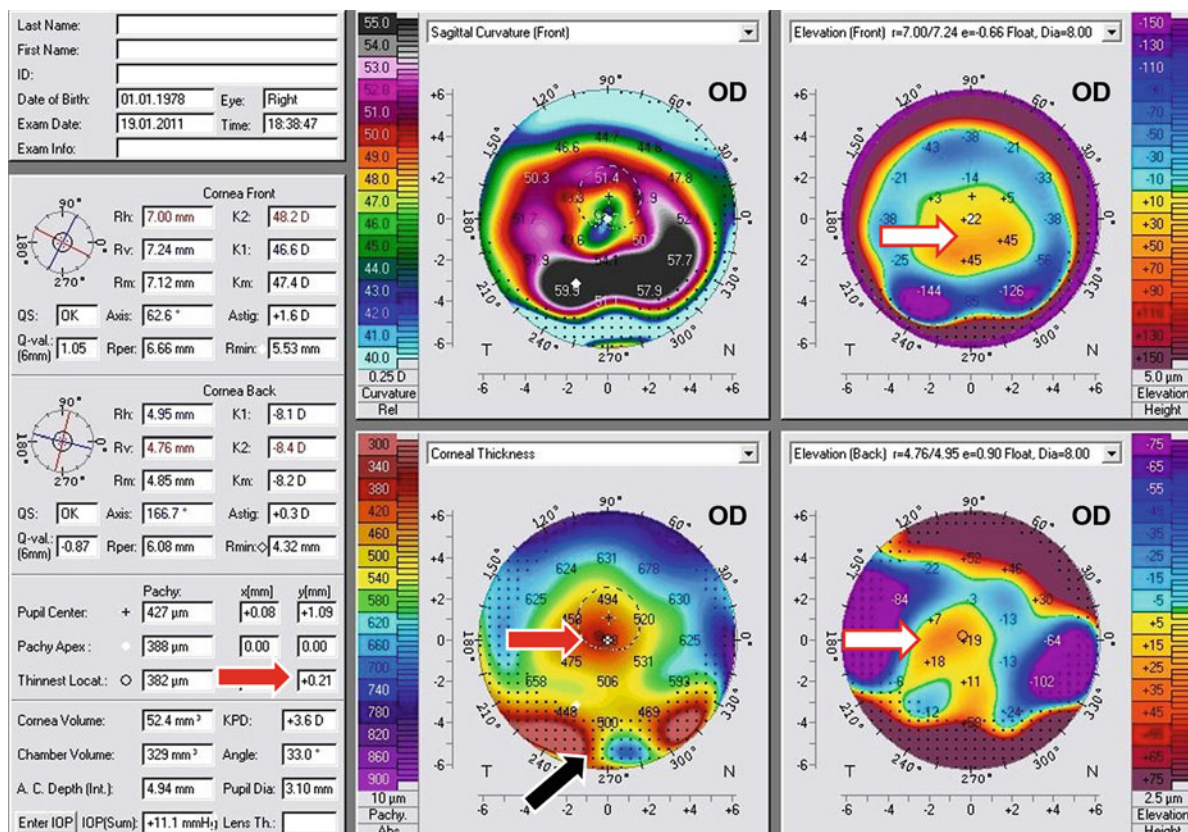


Fig. 2.55 Corneal topography of the same case in the previous figures. Although there is an inferior thin area, the thinnest point is plotted centrally (red arrows). The black arrow points at the proposed dotted inferior area. Note the bell sign

4. Peripheral corneal melting disorders, such as Mooren ulcer, or peripheral melting secondary to rheumatologic disorders are characterized by pain.
 - This pain may be severe in cases of Mooren ulcer.
 - Associated findings include an epithelial defect over the area of thinning and corneal vascularization adjacent to the area of thinning in the acute phase.
5. Contact lens-induced warping of the cornea can mimic the appearance of PMD and PLK on corneal topography. History of prolonged usage of contact lenses and recapturing corneal topography after proper stop of usage will retain corneal regularity and hence differentiates the case from PMD and PLK.

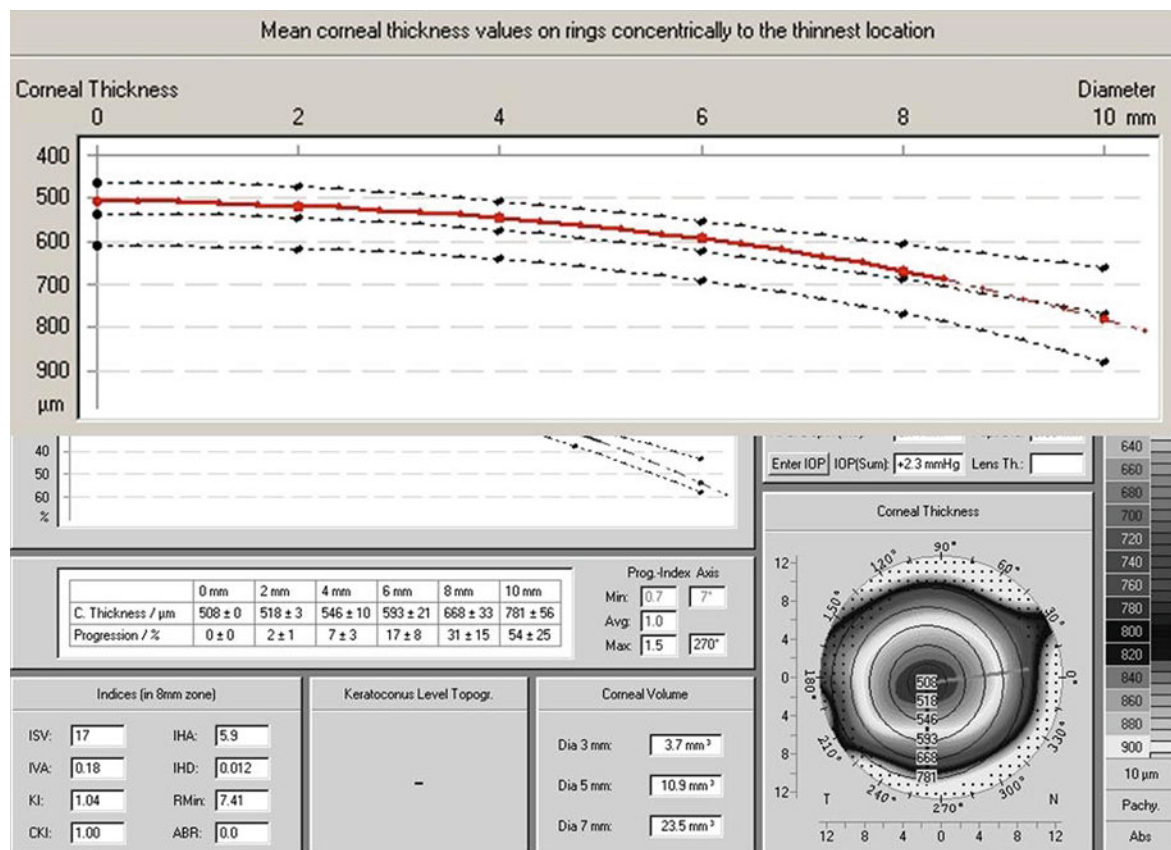


Fig. 2.56 Normal keratoconus curve diagram. The red line (patient's data) is consistent with and lies within the normative data range

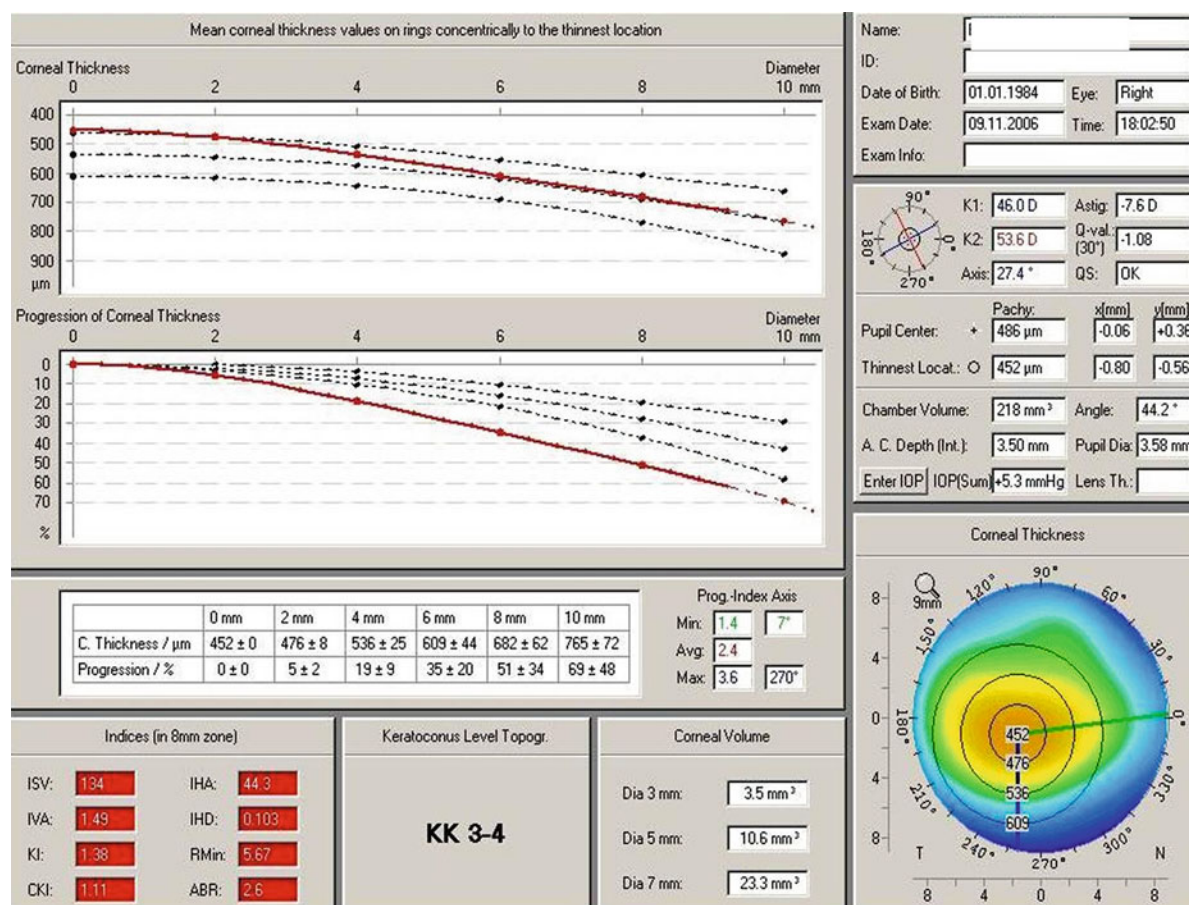


Fig. 2.57 Keratoconus curve diagram in a KC case. The *red line* is out of the normal range and does not take the same slope of it

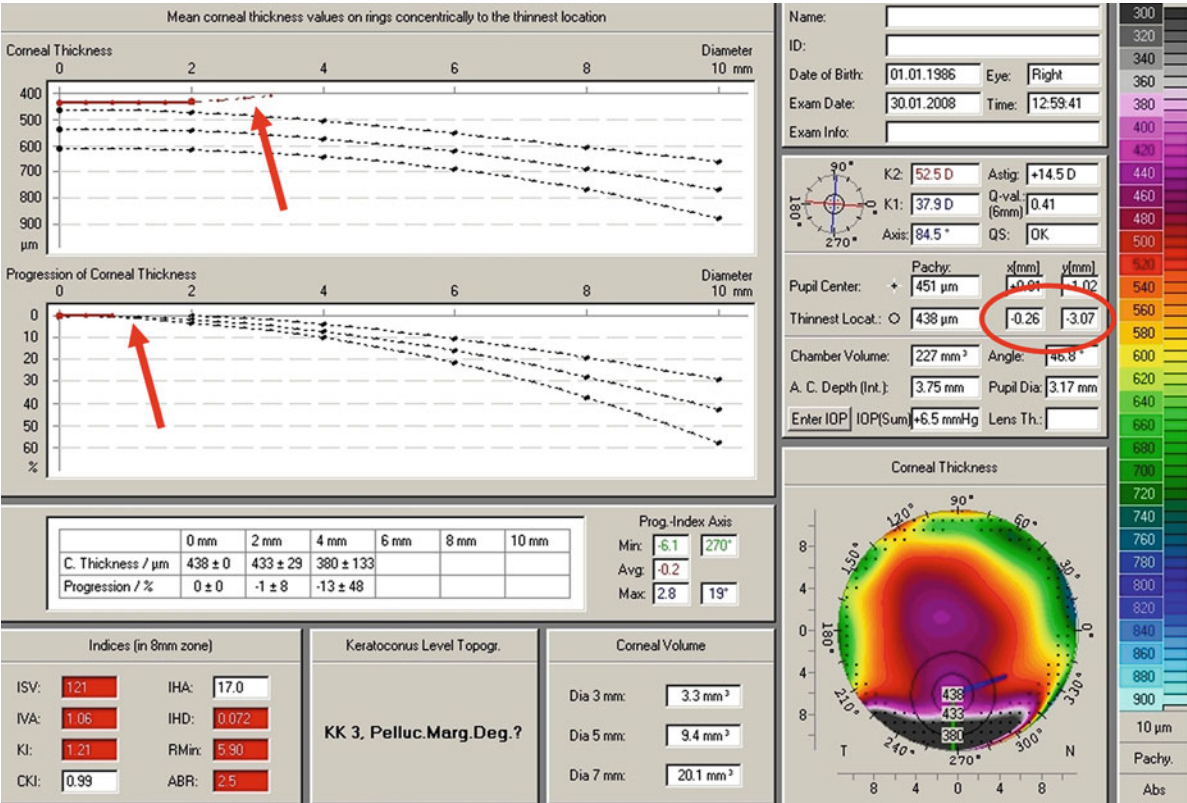


Fig. 2.58 Keratoconus curve diagram in an advanced case of PMD. The red line is out of the normal range and inverted superiorly (red arrows). Notice the big displacement of the thinnest location especially on the Y axis (red circle)

Table 2.2 Comparison between PLK and PMD

	PLK	PMD
Nature	Central or paracentral ectasia	Peripheral ectasia
Age of presentation	Early teens	20–40
Slitlamp and Scheimpflug image	Central or paracentral thinning	Inferior peripheral thinning
Curvature map	Butterfly	Butterfly
Elevation maps	Cone	Central, paracentral, or inferior peripheral cone
	Kissing birds	Present in early and moderate cases
Thickness map	“Bell” sign	Present in moderate and advanced cases
	Thinnest location	Usually largely displaced
Keratoconus curve diagram	Deviated	Deviated and usually inverted in advanced cases

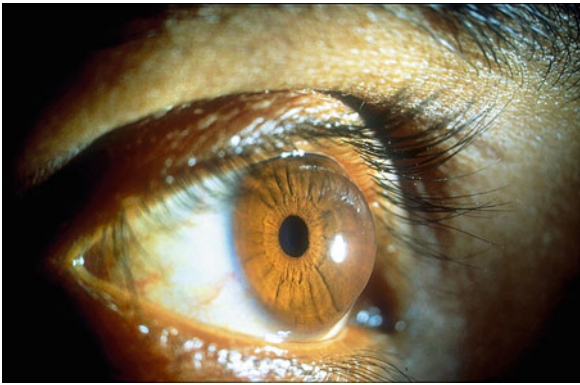


Fig. 2.59 Keratoglobus. A generalized bulging and thinning of the cornea

Fig. 2.60 The corneal thickness map in keratoglobus. Note the generalized thinning including the limbal part of the cornea, which differentiates between keratoglobus and the globus morphological pattern of keratoconus

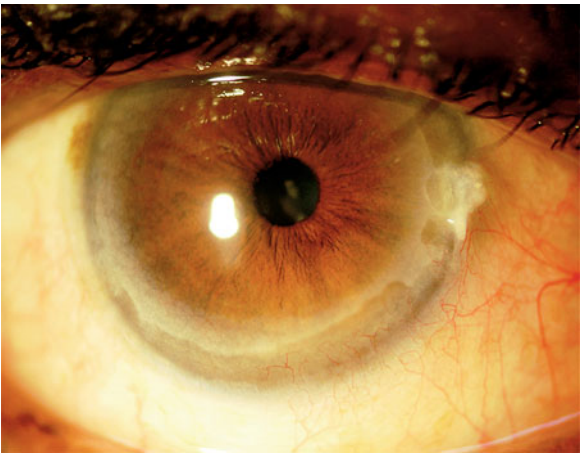
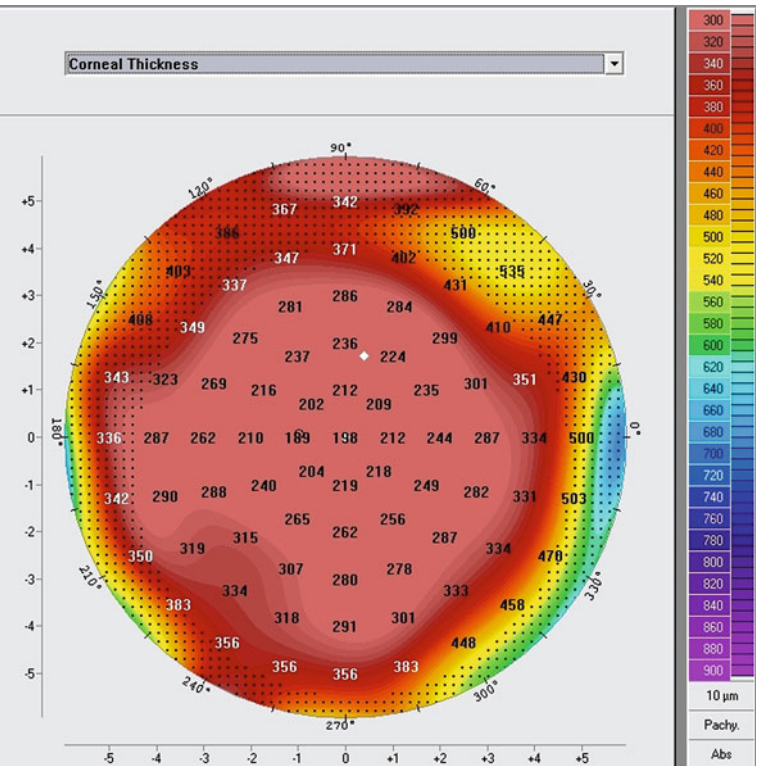


Fig. 2.61 Terrien marginal degeneration. Note the lipid deposition



Fig. 2.62 Furrow degeneration

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