

Keratoconus

- Is bilateral non inflammatory corneal thinning disorder leading to protrusion, distortion, and scarring of the cornea.
- First described in 1854

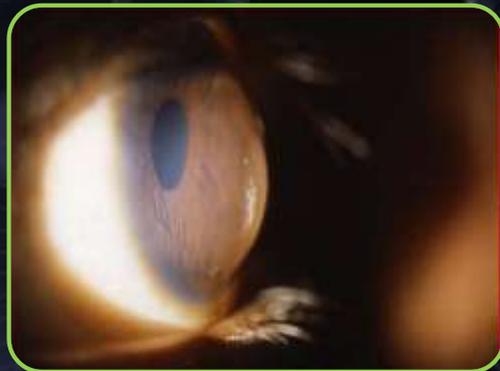
Systemic association:

- Down syndrom
- Turner syndrome
- Marfan syndrome
- Atopy
- Osteogenesis imperfecta
- Mitral valve prolapse



Ocular association

- Vernal diseases
- Retinitis pigmentosa
- Blue sclera
- Aniridia
- Ectopia lentis



Course

- Usually bilateral
- Onset often during puberty
- Occure in all races
- More in females
- 6-8% with family history

Pathology of Keratoconus

- Loss of Bowman's Layer.
- Stromal Thinning.
- Apoptosis.
- Increased Enzyme Activity.
- Enlarged Prominent Corneal Nerves.

Causes of Keratoconus

- **Heredity vs. Mechanical**
- **Cellular**
- **Tissue**
- **Genetic**

Progression and Prognosis

- **Age is a big factor.**
- **The younger the diagnosis, the poorer the prognosis.**
- **Less likely to progress to the point of a transplant if diagnosed in the 30's.**
- **20% of Keratoconus patients result in corneal transplants.**
- **35 to 45% of all transplants are due to Keratoconus.**

Diagnosis of keratoconus

Clinical Findings

Corneal Hysteresis

Corneal topography

Specular Microscopy

Confocal Microscopy

Clinical Findings

External signs

Retinoscopy signs

Keratometry signs

Slit lamp biomicroscopy signs

External signs

Munsons sign

When the patient is asked to look downward toward the floor V-shaped profile of the lower lid margin can be seen



Rizzuti sign

This sign is observed by seeing light on the nasal anterior sclera when the light is directed on the cornea from the temporal direction

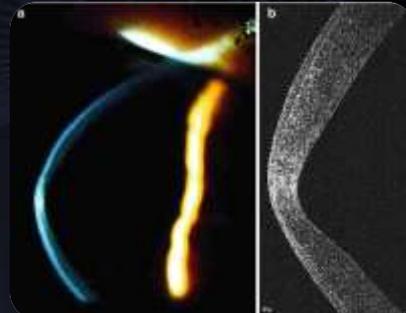
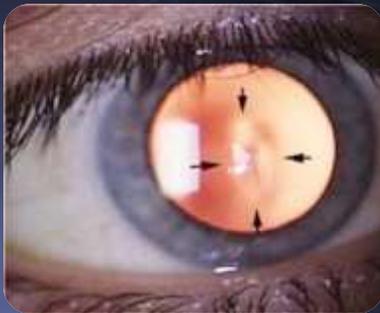


Retinoscopy signs

- The scissoring effect of the retinal reflex seen with retinoscopy is high diagnostic
- Best seen when the pupil is dilated
- Occure even in mild cases of Keratoconus

Slit lamp biomicroscopy signs

1-Focal thinning



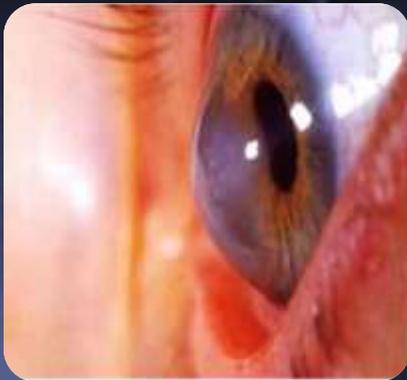
2-Fleischer iron ring

3-vogets stria

4-scar



5-corneal hydrops



Keratometry and Photokeratometry

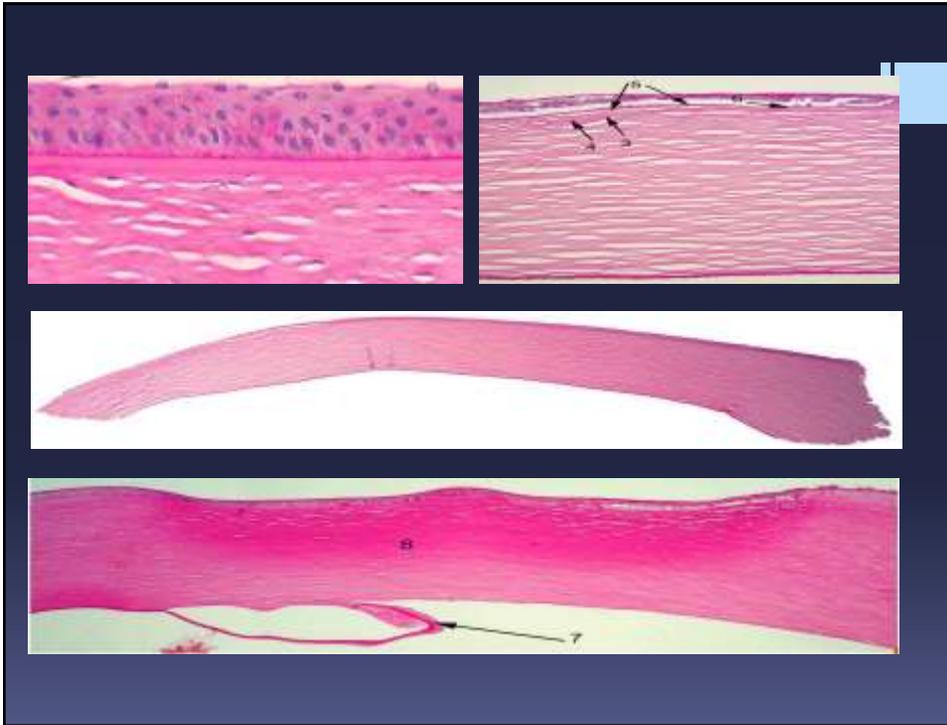


- When the curved surface of the cornea is viewed by keratoscope or photokeratoscope the rings appear to be thin and tightly squeezed together in the regions where the curvature is steep and broadly dispersed wherever the curvature is flat
- In normal spherical cornea the rings are spherical.
- In corneal astigmatism the rings are oval with the short axis corresponding to the steep meridian.
- In keratoconus the rings are distorted and grouped more closely in the region of the cone.



Confocal Microscopy

- Elongated exfoliating superficial epithelial cells
- Enlarged wing and basal epithelial cells
- Bright reflective material deposited within basal epithelial cells
- Prominent thickened subbasal nerves
- Thinning of the stroma
- Irregular arrangement of the stromal keratocytes
- Z shaped folds in the anterior, mid and posterior stroma
- Folds in Descemet's membrane
- Pleomorphism and enlargement of endothelial cells
- Increased endothelial cell density



Specular Microscopy

- As seen in focal microscopy
- Altered endothelial cell morphology
- Significant increase in polymegathism
- Significant decrease in hexagonality
- Higher pleomorphism

Corneal topography

Topographic Criteria of Keratoconus

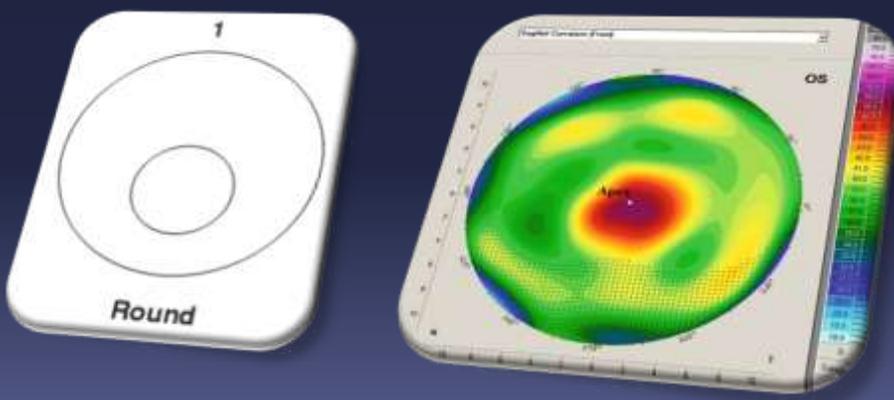
When more than one of the following criteria are found frank KC, Forme Fruste KC (FFKC), early stage KC, or at least a case of suspicion (according to the severity and amount of signs) is considered:

According to curvature

- There are several patterns of corneal curvature some can be accepted others are risky for lasik or even indicators for keratoconus .
- Corneal irregularities may appear as one of the following patterns :

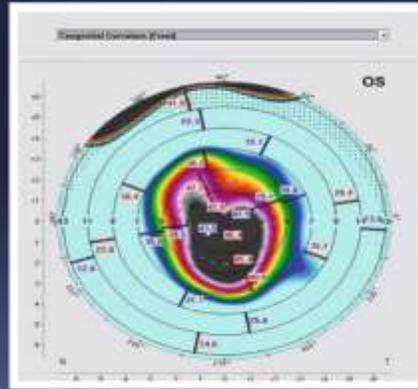
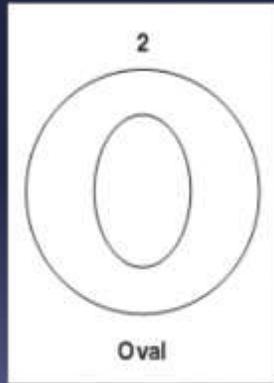
Pattern 1

Round



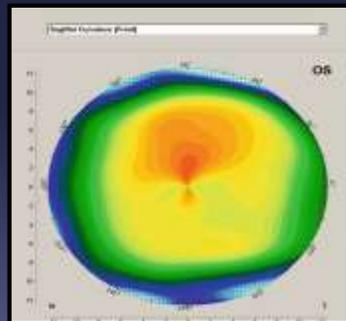
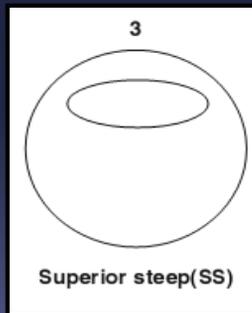
Pattern 2

Oval



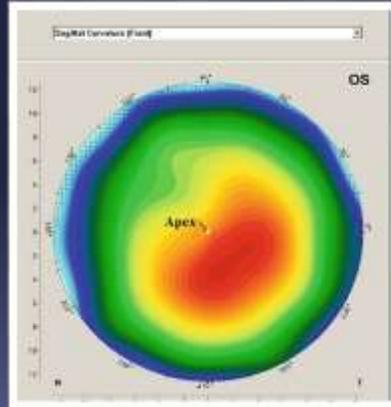
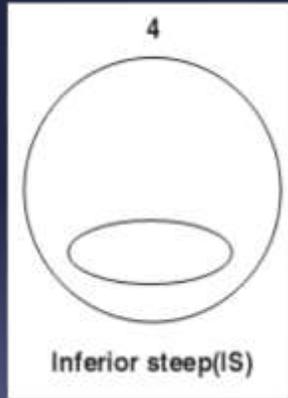
Pattern 3

Superior Steep(SS)



Pattern 4

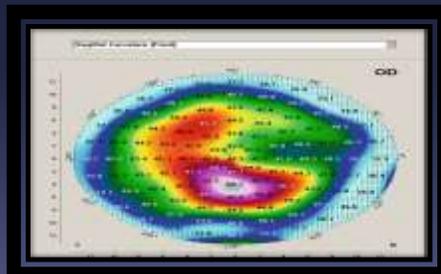
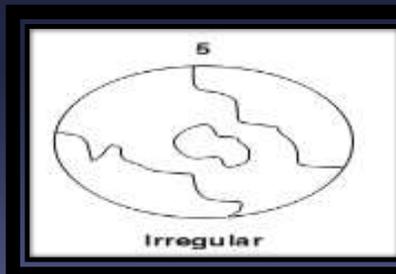
Inferior Steep (IS)



Pattern 5

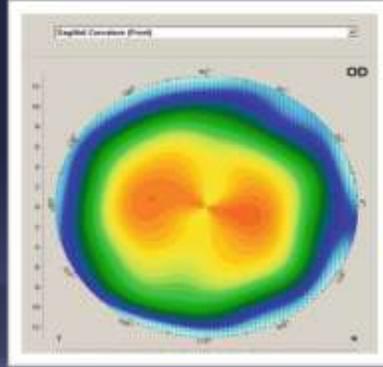
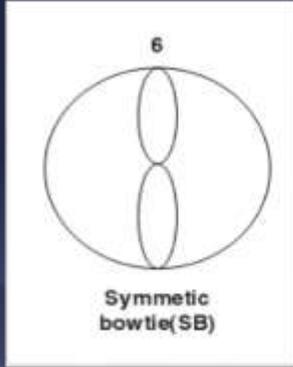
Superior Steep (SS)

Irregular



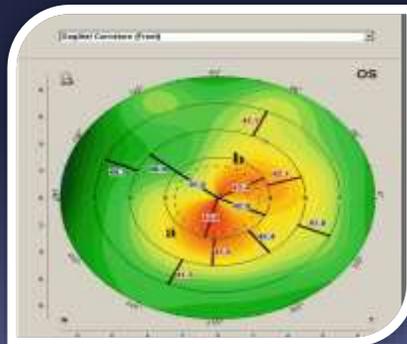
Pattern 6

Symmetric Bowtie



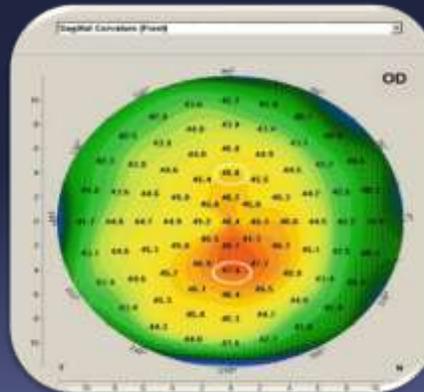
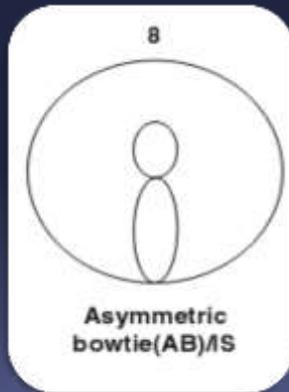
Pattern7

Symmetric Bowtie SB /Skewed Steepest Radial Axis Index SRAX



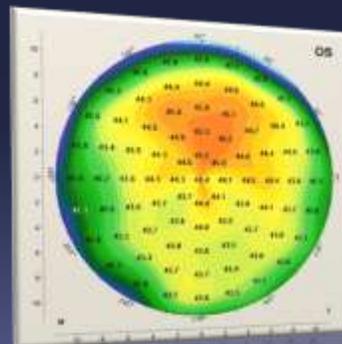
Pattern 8

Asymmetric Bowtie AB /IS



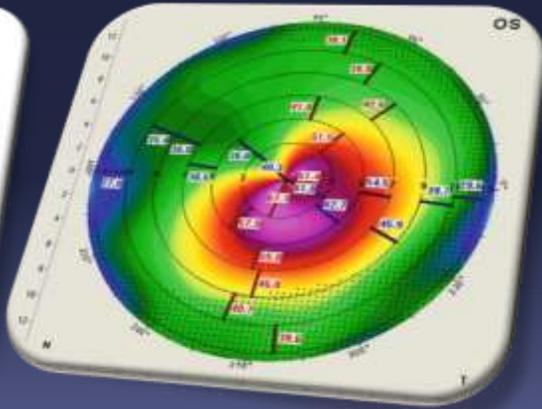
Pattern 9

**Asymmetric Bowtie AB/
Superior Steep**



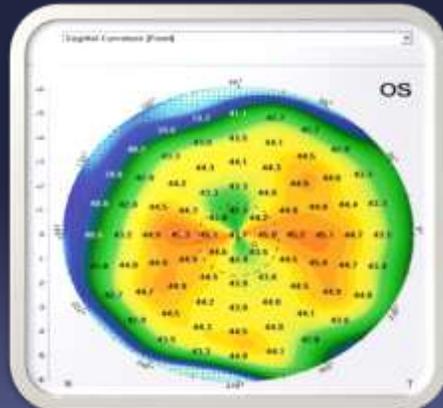
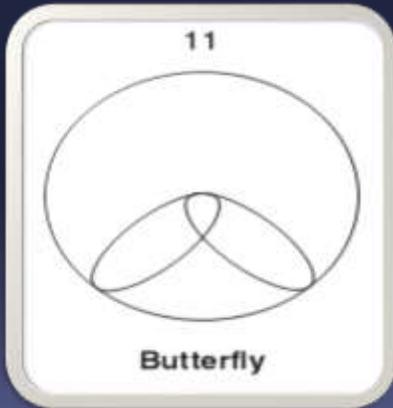
Pattern 10

AB/SRAX



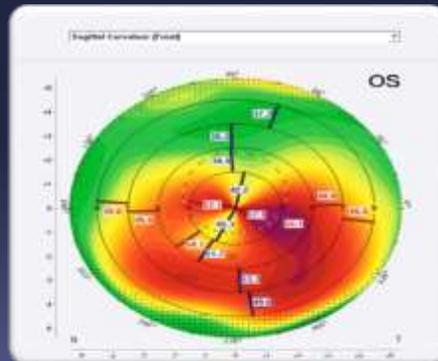
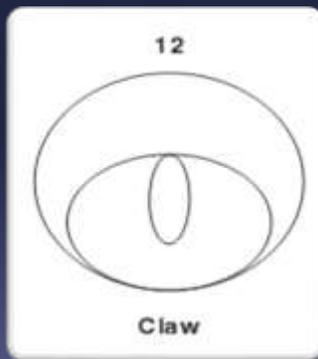
Pattern 11

Butterfly



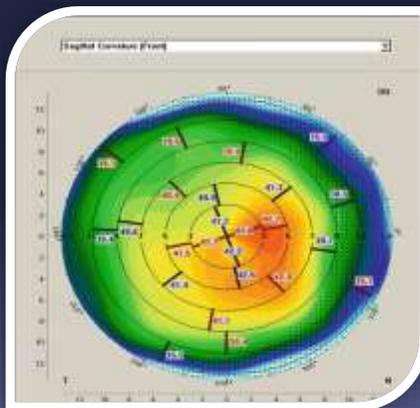
Pattern 12

Clow Pattern



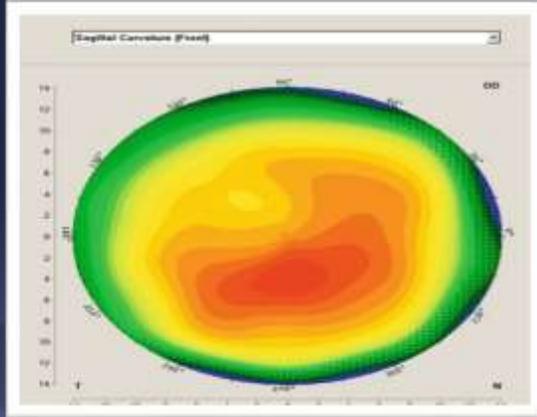
Pattern 13

Junctional Pattern



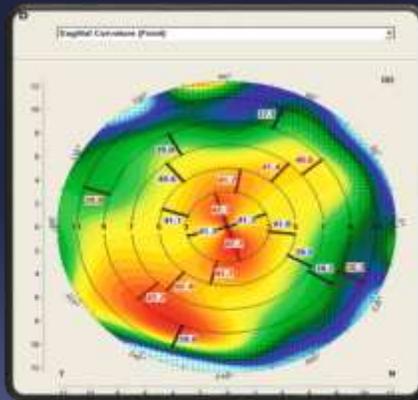
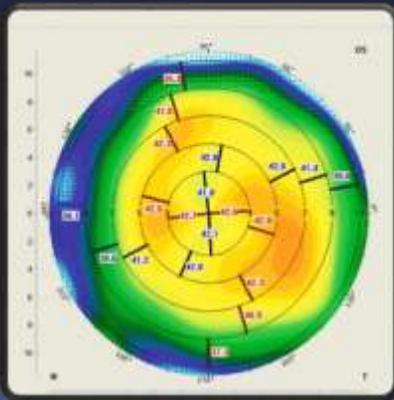
Pattern 14

Smiling Face



Pattern 15

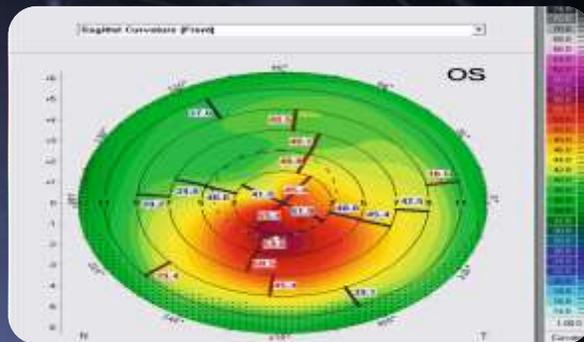
Vortex Pattern





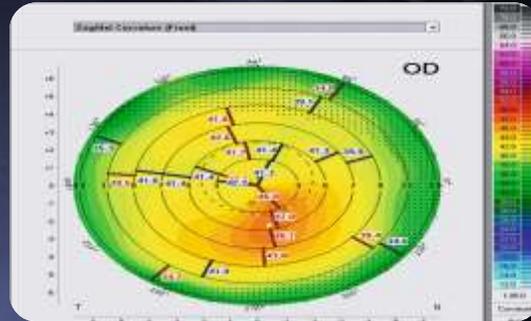
Pattern 1

The inferior step 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



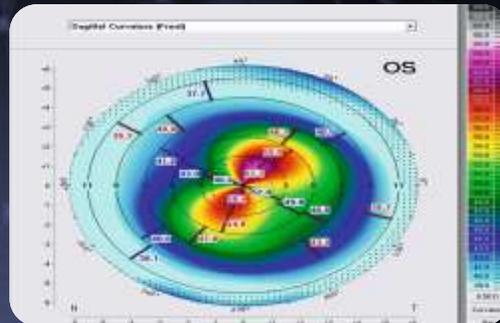
Pattern 2

The inferior step pattern, it is like pattern1 except that there is a more than 22° of skew between the two axes



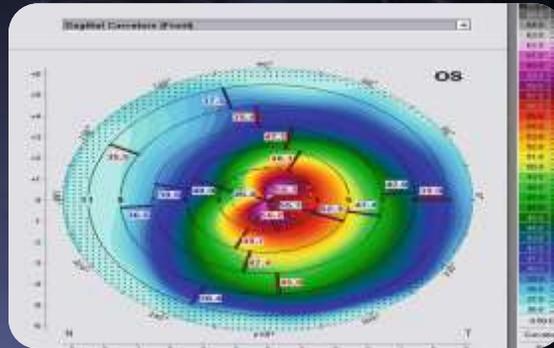
Pattern 3

Both segments of the bow tie are equal in size and have straight and aligned axes



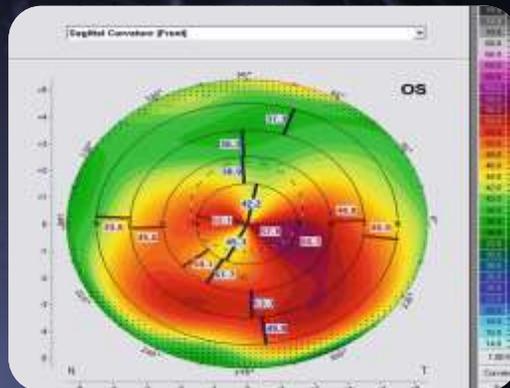
Pattern 4

The two segments are equal in size but there is more than 22° of skew between the two axes



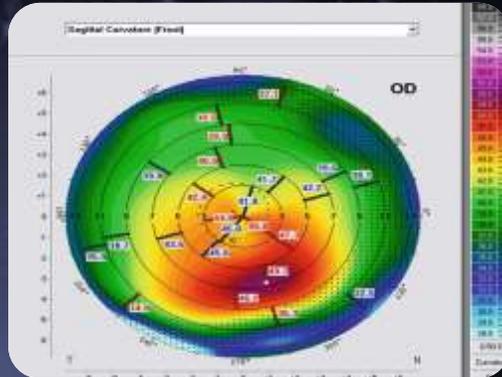
Pattern 5

It is PMD or Pellucid-like KC (PLK) with straight axis



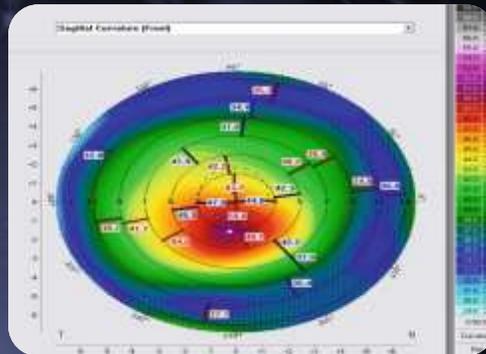
Pattern 6

It is PMD or PLK with more than 22° of skew between the two axes



Pattern 7

Where the cone is eccentric and the steep and flat axes are difficult to identify



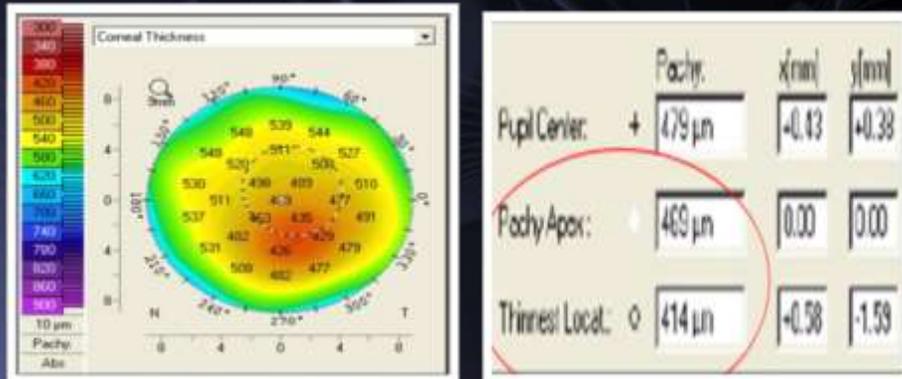
On the sagittal curvature map

- ❖ K-readings > 48 dpt.
- ❖ SRAX $> 22^\circ$.
- ❖ Superior–inferior difference (S-I) on the 4 mm circle > 2.5 dpt.
- ❖ Inferior–Superior difference (I-S) > 1.5 dpt.
- ❖ Corneal astigmatism on either surface should not be higher than 6D; otherwise, it is a risk factor.
- ❖ Against the rule, astigmatism is considered suspicious.
- ❖ Calculate I-S Rabinovich ratio, it should be $< +2$.

On the thickness map

- ❖ Cone-like shape.
- ❖ Superior–inferior at 4 mm circle $> 30 \mu$.
- ❖ Thinnest location $< 470 \mu$.
- ❖ Thickness at pachy apex -thickness at thinnest location $> 10 \mu$.
- ❖ Y coordinate value of the thinnest location $> -500 \mu$.
- ❖ Difference in thickness between both eyes at thinnest locations $> 30 \mu$.

Look at the thickness map: Compare thickness values between the apex and the thinnest point in each eye and those points between both eyes.

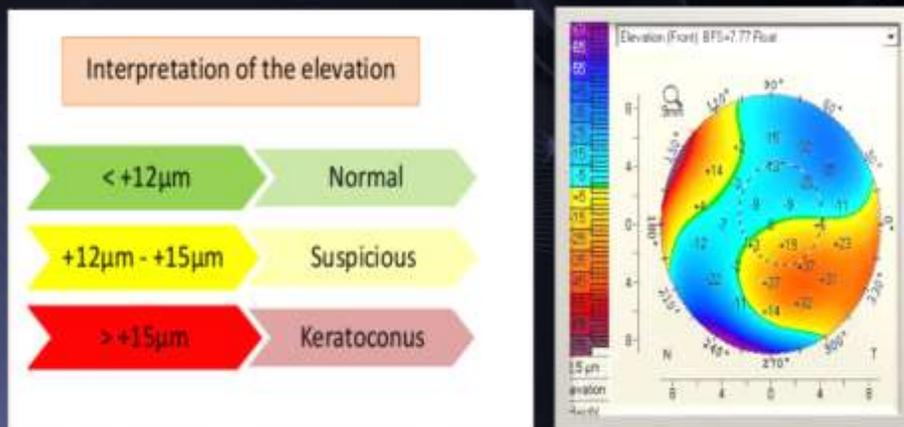


On the elevation maps

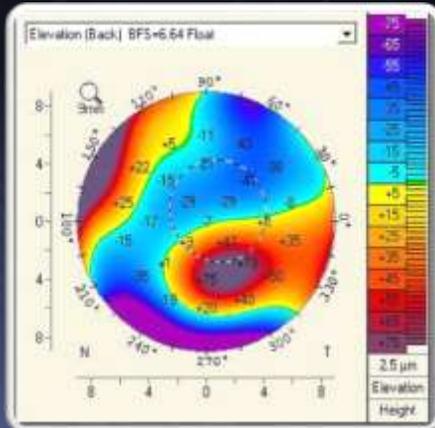
- ❖ The elevation values on the front surface map should not exceed +12 μ . Values between +13 μ and +15 μ are suspected, and any value > +15 μ is considered a risk factor
- ❖ The elevation values on the back surface map should not exceed +17 μ . Values between +18 μ and +20 μ are suspected, and any value > +20 μ is considered a risk factor

- ❖ The difference between the back and front surfaces (back-front) should not exceed $+5 \mu$ at the same point.
- ❖ If there is any isolated island on either front or back surfaces, it would be suspected, even with values within the normal limits

Look at anterior elevation map:



Look at posterior elevation map



Interpretation of the elevation

< +17 μm Normal

+17 μm - +20 μm Suspicious

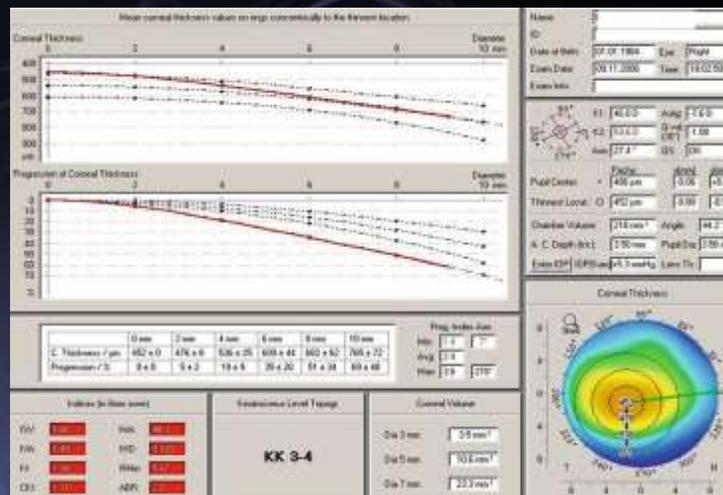
> +20 μm Keratoconus

Anterior
elevation >
+15 μm

posterior
elevation >
+20 μm

Thickness
difference >
10 μm

A case of
keratoconus



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

Table 9.1: Topographic indices

- ISV = Index of surface variance
 - Gives the deviation of the individual corneal radii from the mean value
 - Elevated with irregular corneas
- IVA = Index of vertical asymmetry
 - Gives the degree of symmetry of the corneal radii with respect to the 180° meridian as axis of reflection.
 - Elevated in cases of oblique axes, KC or limbal ectasias.
- IHA = Index of height asymmetry
 - Degree of symmetry of height data with respect to the horizontal meridian as axis of reflection
 - Analogous to IVA but sometimes more sensitive
- ABR = Aberration coefficient
 - Calculated on basis of Zernicke analysis
 - 0.0 is perfect system, becomes 1.0 or greater with aberrations
- KI = Keratoconus index
 - Elevated in KC
- CKI = Center keratoconus index
 - Elevated in central keratoconus
- RMin = Radii minimum
 - Smallest radius of curvature in entire field measurement
 - Elevated in KC
- IHD = Index of height decentration
 - Calculated from Fourier analysis of height
 - Gives degree of vertical decentration
 - Steeper in KC

Table 9.5: KC index levels

Index	Abnormal (Yellow)	Pathological (Red)
Index of Surface Variance (ISV)	≥ 37	≥ 41
Index of Vertical Asymmetry (IVA)	≥ 0.28	≥ 0.32
Keratoconus Index (KI)	>1.07	>1.07
Center Keratoconus Index (CKI)	≥ 1.03	≥ 1.03
Radii Minimum (R_{min})	<6.71	<6.71
Index of height asymmetry (IHA)	≥ 19	≥ 21
Index of height decentration (IHD)	≥ 0.014	≥ 0.016
Aberration coefficient (ABR)	≥ 1	≥ 1

Indices (in 8 mm zone)

ISV:	27	IHA:	7.6
IVA:	0.31	IHD:	0.019
KI:	1.09	RMin:	6.98
CKI:	1.01	ABR:	1.1

Various indices, described in, are used to determine the level of KC risk, and suspect values appear in red

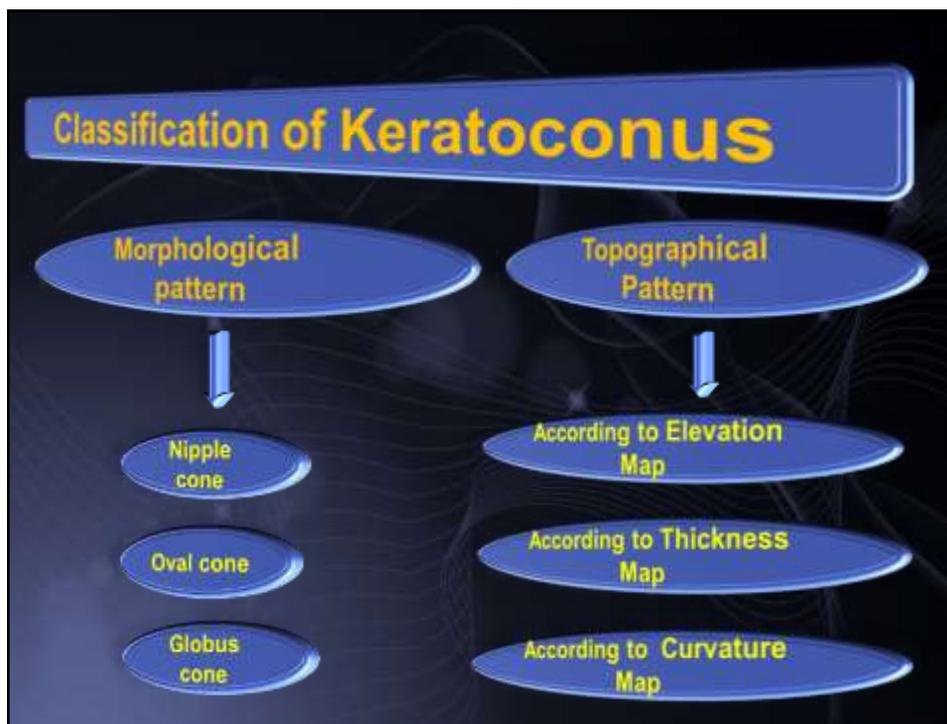
Table 3.1: The normal, suspected and abnormal findings in corneal topography

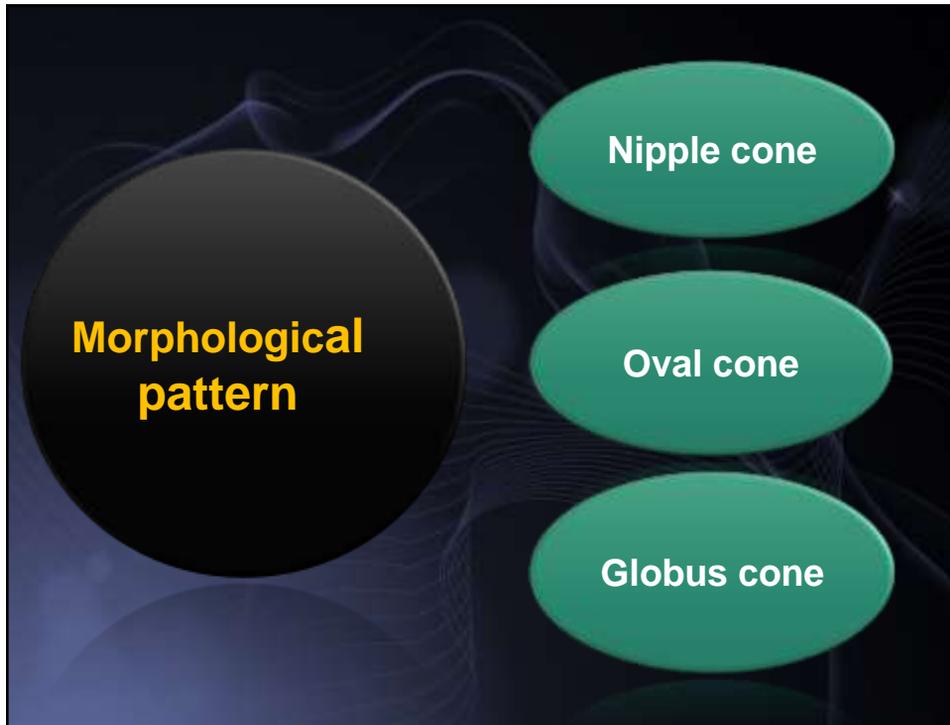
	<i>Normal value(s)</i>	<i>Suspected value(s)</i>	<i>Abnormal value(s)</i>
Quality specification (QS)	white	yellow	red
K-readings:	look at flat K for myopic treatment, look at steep K for hyperopic treatment		>48
Corneal astigmatism	compare with manifest astigmatism		> 6
Average Q-value	0 to -1		0,-1
Topometric map (vertical and inferior Q-values)		-0.5 to -0.55	>-0.55
Thinnest location			
Thickness	> 500	470 – 500	< 470
Difference in thickness between patchy apex and thinnest location	< 5 μ	5 – 10 μ	>10 μ
Coordinates	< 500 μ	500 – 1000 μ	>1000 μ
Pupil center coordinates	important for treating hyperopia and >3D astigmatism		

Contd...

	<i>Normal value(s)</i>	<i>Suspected value(s)</i>	<i>Abnormal value(s)</i>
KPD	<+0.75	+0.75 to +1.5	>+1.5
Anterior curvature map			
Maximal K	important when treating hyperopia		
Pattern	refer to topographical patterns		
I-S Rabinovich ratio			>+2
Skewed Steepest Radial Axis Index (SRAX)			>22°
Superior-inferior difference on the 4 mm circle			>1.5D when the inferior is steeper >2.5 when the superior is steeper

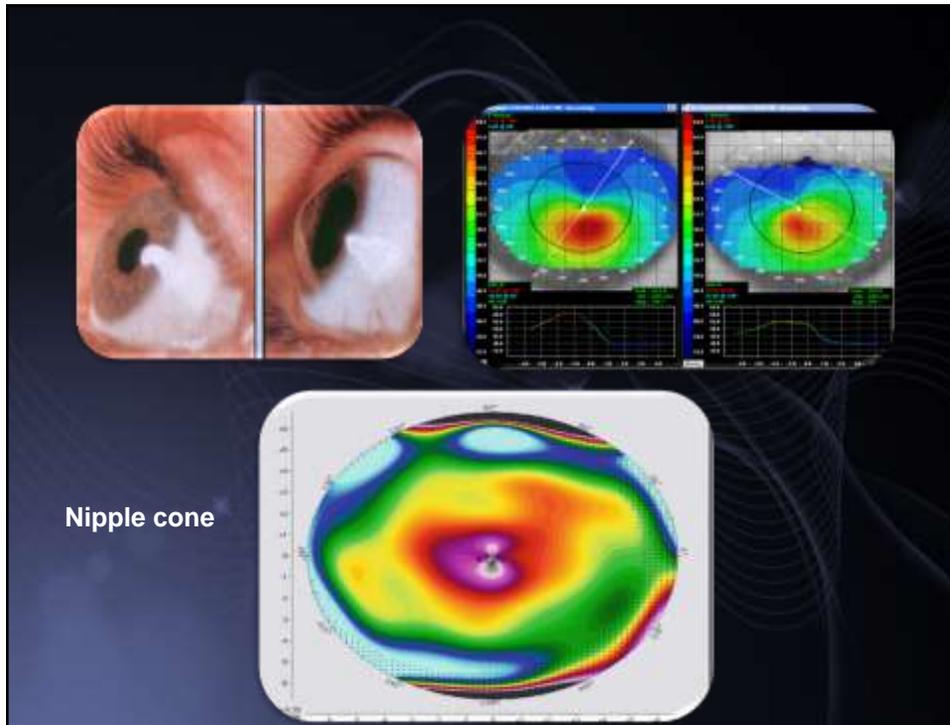
Elevation maps (within the 4 mm central circle)			
Anterior	$\leq 12 \mu$	13 - 15 μ	$> 15 \mu$
Posterior	$\leq 17 \mu$	18 - 20 μ	$> 20 \mu$
Anterior-posterior difference	$< 5 \mu$	$> 5 \mu$	
Isolated island (or tongue like extension)	might be an indicator for FFKC or subclinical keratoconus		
Corneal thickness map			
Shape		cone like	
Superior-inferior difference	$< 30 \mu$	$> 30 \mu$	
Thinnest location difference between both eyes	$< 30 \mu$	$> 30 \mu$	
Keratoconus diagram			
Shape and location of the curve		- out of normative range - deviation before the 6 mm circle	
Average	1	1.1 - 1.2	> 1.2
Indices of irregularity	white	yellow	red





Nipple Cone

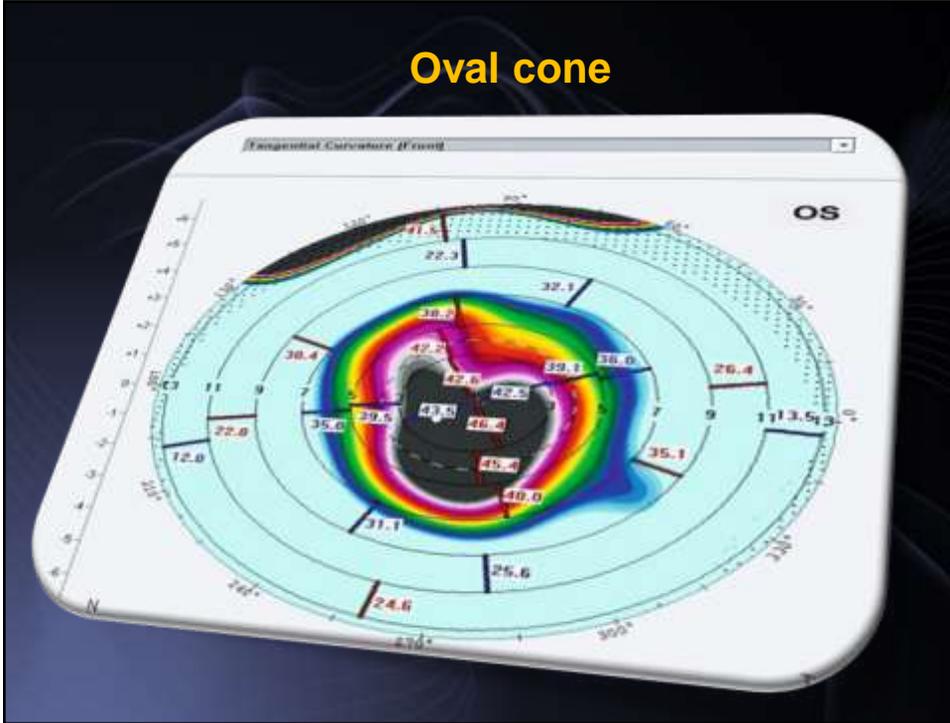
- Small size(5mm)
- Steep curvature
- The apical center is either central or para Central
- Commonly displaced inferonasally



Oval cone

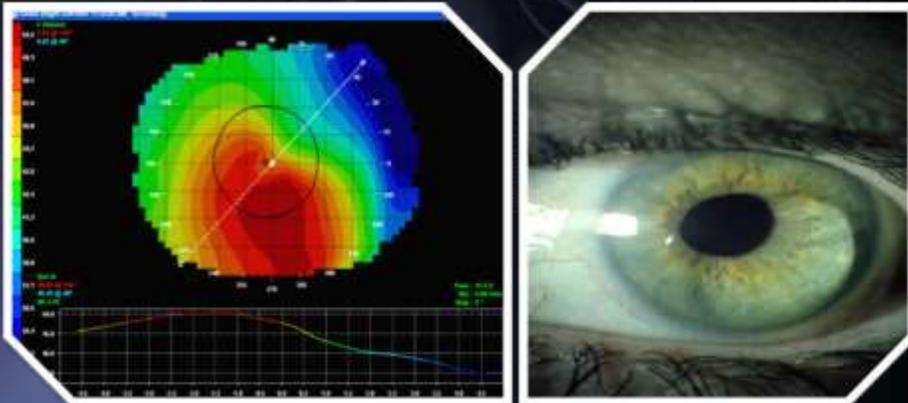
- Larger (5-6)mm
- Ellipsoid
- Commonly displaced inferotemporally

Oval cone

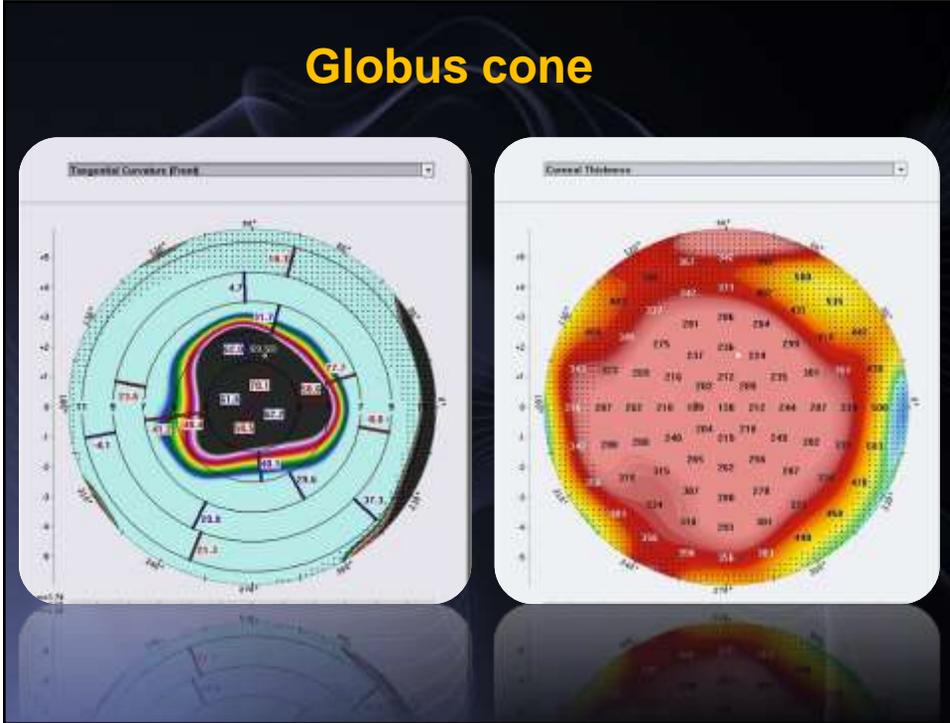


Globus cone

- Largest more than 6mm
- May involve more than 75% of the cornea



Globus cone



Topographical pattern

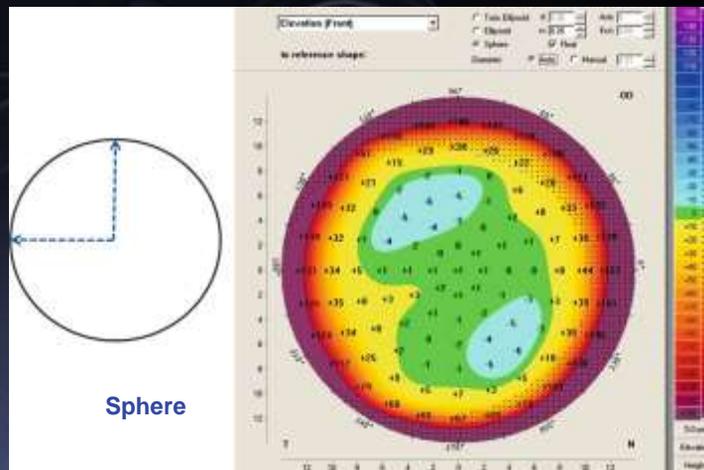
Elevation
Map

Thickness
map

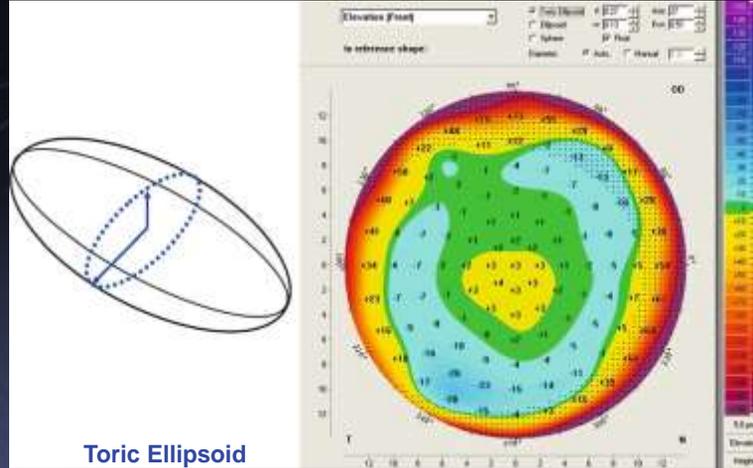
Curvature
map

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) or by best fit toric ellipsoid mode (BFTE)
- The best to locate the cone is the BFS, and the best to evaluate the real height of the cone is the BFTE.



The elevation map displayed in the best fit sphere float mode



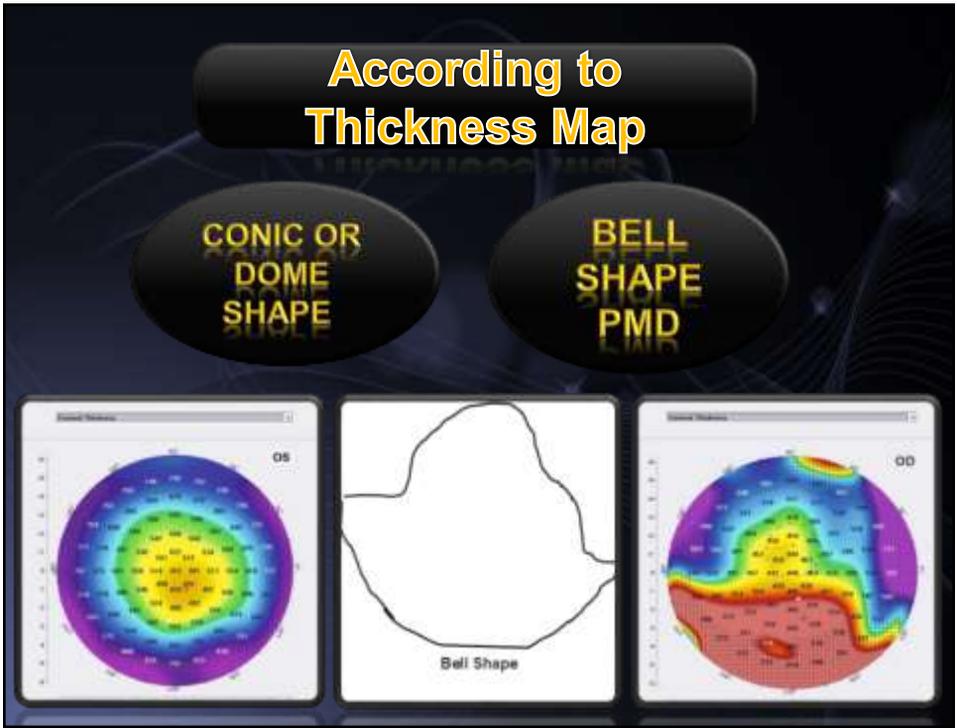
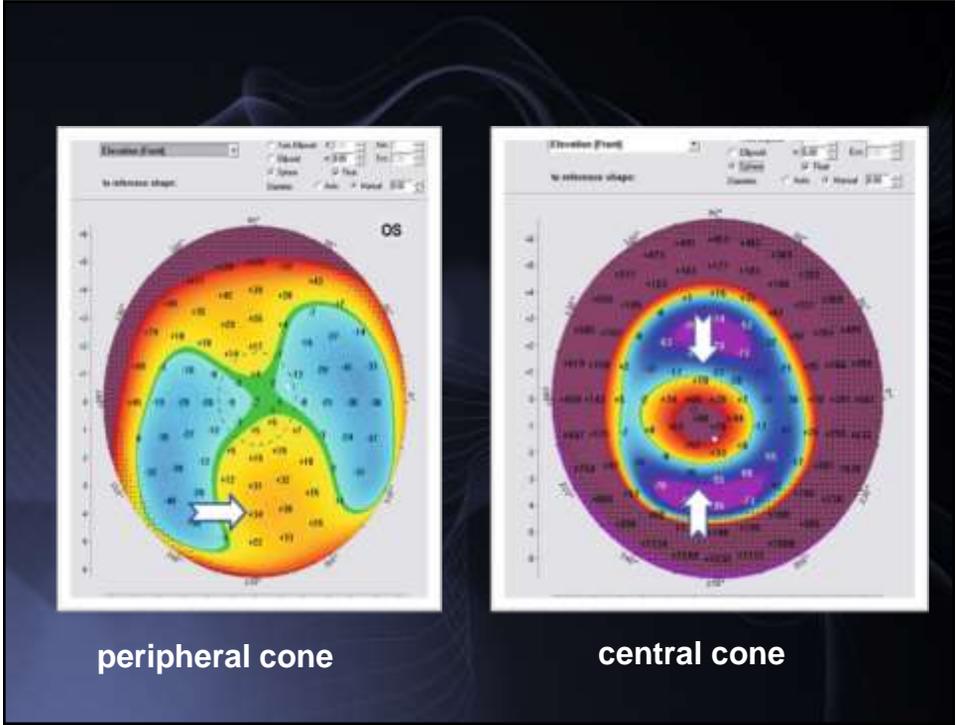
The elevation map displayed in the best fit toric ellipsoid float mode

According to elevation map

Central

Paracentral

Peripheral



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

Forme Fruste Keratoconus

- Forme Fruste Keratoconus (FFKC) is a subclinical disease
- The diagnosis of KC is a clinical one that is aided by topography, while the diagnosis of FFKC is topographic
- 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

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.

Differential Diagnosis

Pellucid Marginal Degeneration (PMD)

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

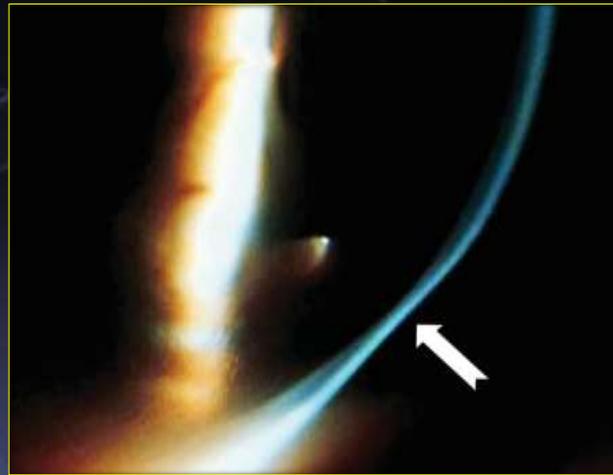
Clinical Findings

- ❖ 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.

Slit lamp examination

- ❖ 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
- ❖ 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.

- ❖ The area of thinning typically is epithelialized, clear, avascular, and without lipid deposits.
- ❖ On careful slitlamp evaluation, prominent lymphatics often are detected at the inferior limbus parallel to the area of thinning.
- ❖ Vertical striations at the level of the Descemet's membrane (similar to the Vogt striae) may be seen in rare instances.
- ❖ The light slit becomes very narrow abruptly in the inferior part of the cornea which is the hallmark of the disease

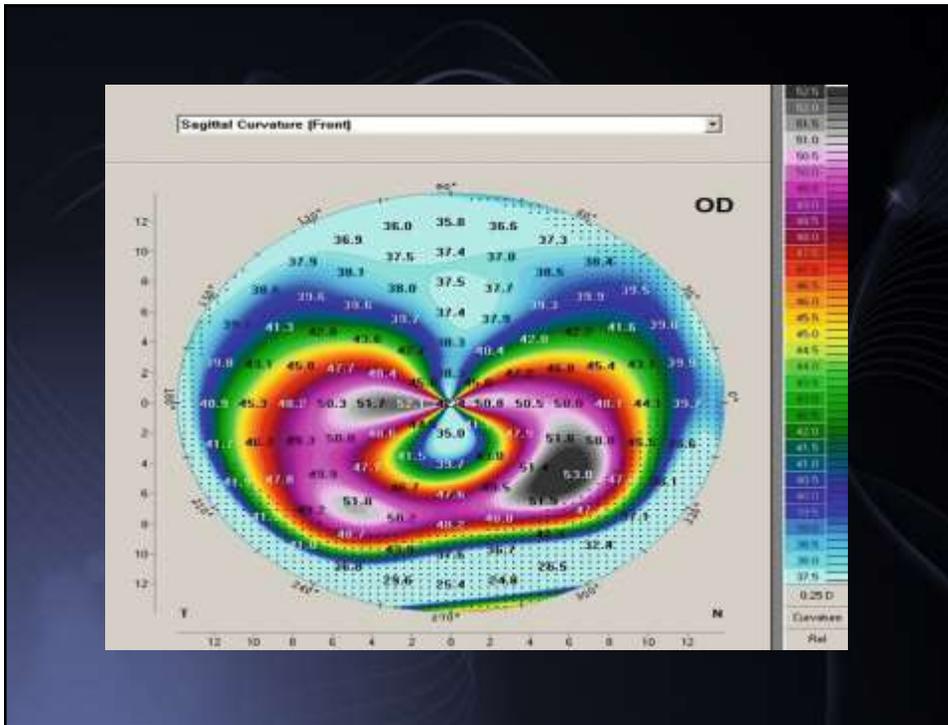


Slit lamp view of PMD, the light slit becomes very narrow abruptly in the inferior part of the cornea (white arrow) which is the hallmark of the disease

Topographical Findings

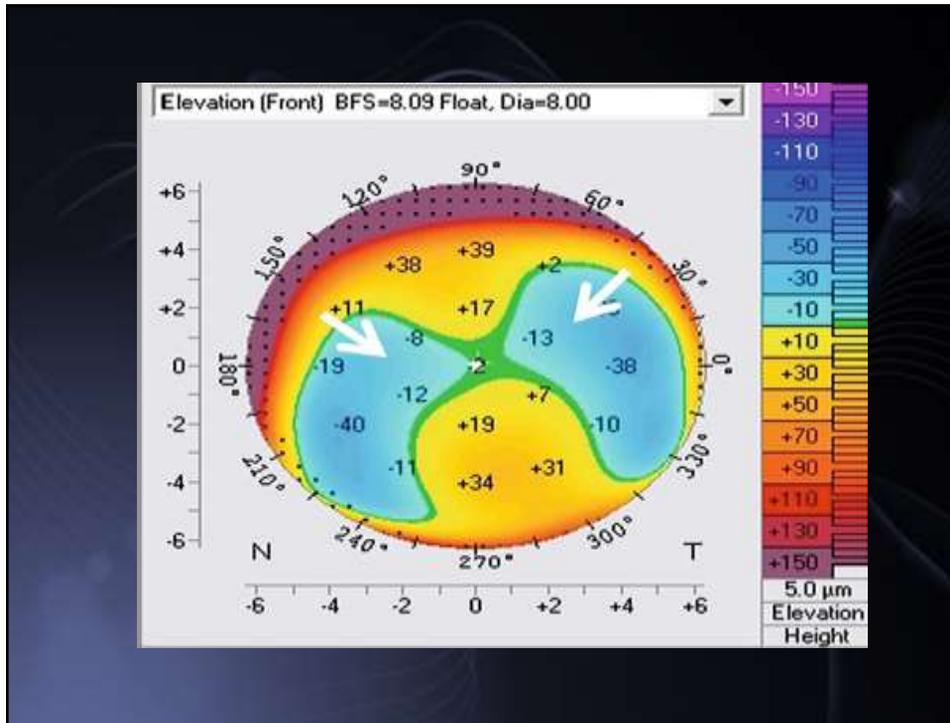
Curvature map:

- ❖ low corneal power along the central vertical axis.
- ❖ increased power as the inferior cornea is approached.
- ❖ high corneal power along the inferior oblique meridians.
- ❖ This topographical pattern takes a butterfly appearance.



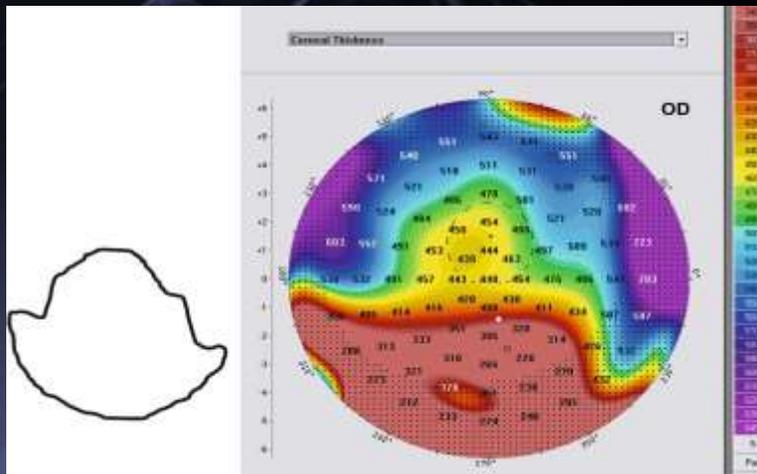
Elevation maps

- ❖ kissing birds sign: when the cone is peripheral, this sign is present; when the cone is central or paracentral, this sign is Absent
- ❖ This sign appears when the BFS float mode is used while it disappears when switching to the BFTE float mode



Corneal thickness map:

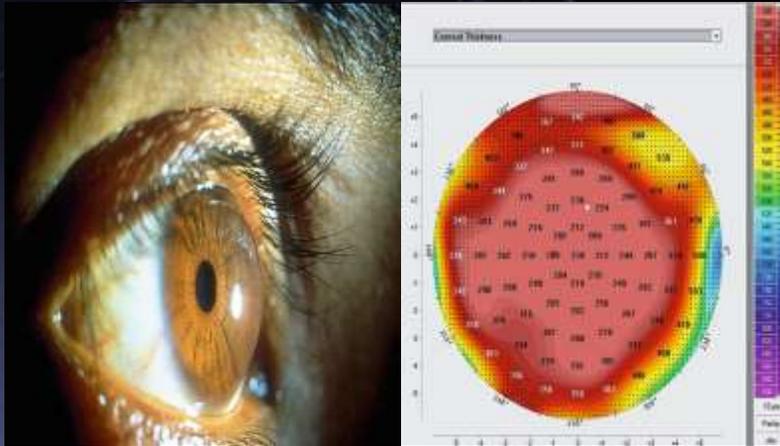
- ❖ Corneal thinning is a reversal of the typical pattern in which the cornea thickens from the center to the periphery.
- ❖ the corneal thickness map reveals a thinning of the inferior cornea. This thinning is characterized by a special sign that can be called “bell” shape



the bell sign.

Keratoglobus

- ❖ It is a generalized thinning of the cornea
- ❖ 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.



Keratoglobus

Terrien marginal degeneration

- ❖ It affects an age group similar to that affected by PMD.
- ❖ 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



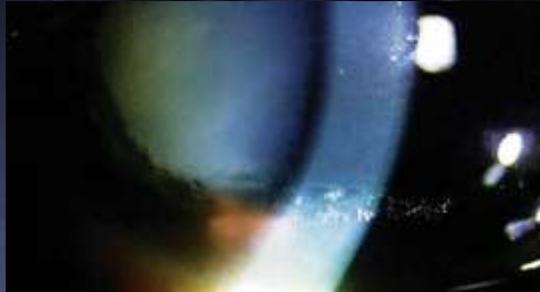
Terrien marginal degeneration. Note the lipid deposition

Furrow degeneration

- ❖ 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.



Furrow degeneration

