

# Topographic Interpretation of Posterior Keratoconus

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We present a case of a sixty years old Asian male who presented to us with gradual decrease in vision of both eyes. Slit lamp examination revealed paracentral thinning with a dome-shaped excavation in the posterior corneal surface in each eye. Other than the early lens changes, rest of the ocular examination was normal. A diagnosis of bilateral Posterior Keratoconus was made. Corneal topography was done to confirm the diagnosis. Findings of the Galilei scan of the patient are discussed in this case report in relation to normal corneas.

**Key words:** Posterior keratoconus, corneal curvature, Galilei scan, Pachymetry, corneal topography.

Posterior Keratoconus is a rare sporadic condition which is characterized by non progressive increase in curvature of posterior surface of cornea.<sup>1</sup> Anterior corneal surface remains normal, so visual acuity is unimpaired. Posterior keratoconus is a non progressive, non inflammatory, unilateral (rarely bilateral) disorder which, is characterized by increase in the curvature of posterior corneal surface<sup>2,3</sup>.

Corneal involvement can be diffuse or localized. Diffuse form is called Keratoconus posticus generalis in which cornea typically remains clear. Localized form is called keratoconus posticus circumscriptus, in which, central or paracentral areas of posterior excavation is seen<sup>4</sup>.

Decrease in visual acuity occurs because of irregular astigmatism but it is not as severe as seen in anterior keratoconus. As the refractive indices of cornea and aqueous humour are similar i.e. 1.376 and 1.336 respectively, there is minimal effect on refraction at aqueous-posterior corneal surface interface. Usually diagnosis occurs by chance when the patient comes for other reason. A similar patient who came to our out-patient department for decreased vision was found to have posterior keratoconus.

A sixty years old Asian male presented, in a

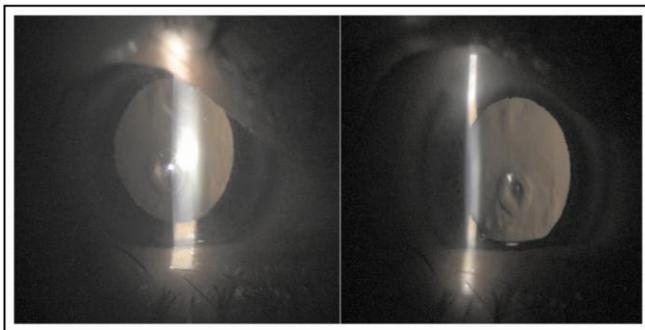
tertiary care hospital of Lahore, with gradual decrease of vision in both eyes for previous six months. Visual complaint was not associated with redness, watering or pain in the eyes. There were no aggravating or relieving factors. He was non-diabetic, non-hypertensive and non-smoker. On examination, the patient was an average stature, average built male and had no systemic abnormality. He was orthotropic and had uncorrected visual acuity of 6/18 in each eye. There was no improvement with pinhole. His color vision was normal. Slit lamp examination revealed normal conjunctiva and sclera. Cornea showed paracentral thinning with a dome - shaped excavation in the posterior corneal surface in each eye (Figure 1). Anterior chamber was deep and quiet. Cortical cataract and nuclear changes were seen in the crystalline lens. Pupillary reactions were normal and dilated fundus examination showed normal discs and macula.

Keratometry was performed which revealed vertical Keratometry ( $K_V$ ) of 45 D and horizontal K ( $K_H$ ) of 47 D in right eye. In the left eye,  $K_V$  was 43D and  $K_H$  was 45.5 D.

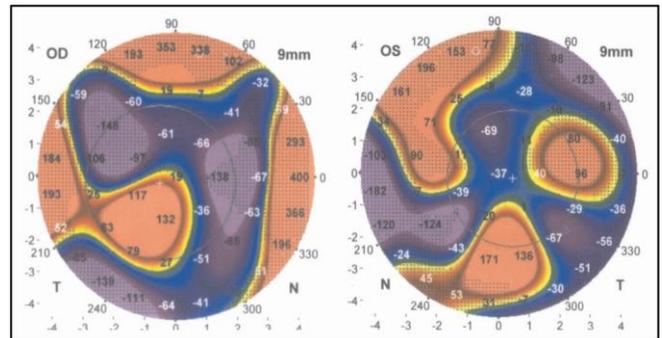
Galilei scan was performed. On topographic scan, anterior axial curvature of a normal eye shows Enantiomorphism (the maps of both eyes are mirror

**Table 1:** Pachymetry comparison of normal and patient's eyes.

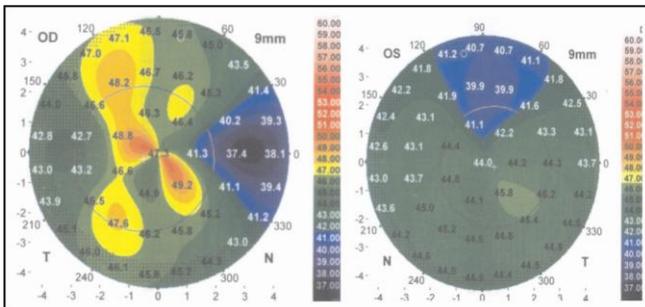
Parameters	Normal	Right eye	Left eye
Thickness at the thinnest location	500 microns	-52 microns	97 microns
Y co-ordinate of thinnest location	< - 500 microns	3.83 mm	3.83
Thickness at Corneal apex minus thickness at thinnest location	< 10 $\mu$ m	570 $\mu$ m	452 $\mu$ m
S-I difference on central 5mm	< 30 mm.	39 mm	284 mm



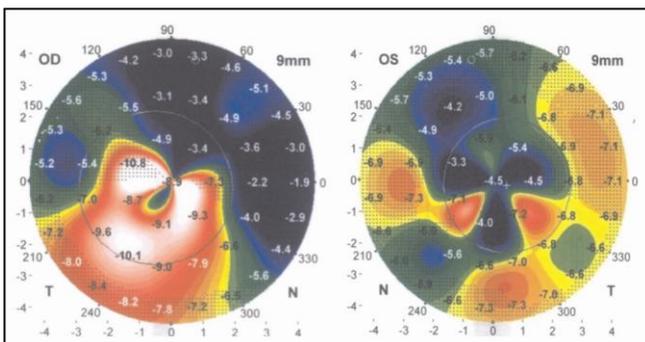
**Fig. 1:** Slit lamp examination showing excavation of the posterior corneal surface.



**Fig. 4:** Posterior elevation maps.



**Fig. 2:** Anterior Axial Curvature map.



**Fig. 3:** Kissing bird sign in right eye and trefoil pattern in left posterior curvature map.

images of each other), symmetric Bow tie pattern and steep K of less than 48 D. In our patient, the normal Enantiomorphism was absent, there was loss of symmetric bow tie pattern and Steep K was 48.77 D in right eye and 44.66 D in left eye (Figure 2). Findings of Pachymetry are shown in table 1. Posterior corneal curvatures were -9.96 D in right eye and -5.41D in the left eye (Figure 3).

In our patient, isolated island patterns were seen in Posterior elevation maps of both eyes. (Normal eyes have Symmetric sandy watch pattern). Best Fit Toric elevation (BFTE) in central 5mm was 132 mm in right eye and 40 mm in the left eye (Figure 4), which were higher than the normal value of less than 15mm.

Based on these findings a diagnosis of posterior Keratoconus was made. Treatment is not required in posterior keratoconus with clear corneas. Glasses were prescribed and the patient was suggested follow up after six months.

**DISCUSSION**

Posterior keratoconus is a rare, non-progressive corneal condition, which was first described by T. Harrison Butler in 1930<sup>5</sup>. This condition is classified

into Diffuse and Localized forms depending on the area of cornea involved. However, Srinivas has further classified Localized form into central, paracentral and peripheral types<sup>6</sup>. Our patient had paracentral type of localized posterior keratoconus.

Posterior keratoconus is also divided into congenital and acquired forms. Electron microscopy of the congenital type has shown an abnormal anterior banded layer of Descemet's membrane which corresponds to an abnormality in 6 to 8 months of gestation<sup>7</sup>. Later, Krachmer suggested that the abnormality would have originated in the fifth or sixth month of gestation<sup>8</sup>.

Acquired type is reported to have occurred secondary to trauma and interstitial keratitis<sup>9,10</sup>.

It should be kept in mind that Posterior keratoconus is not the same as keratoconus. They are entirely different entities. There is only one case, reported in literature in which, a patient had anterior keratoconus in one eye and posterior keratoconus in the other eye<sup>11</sup>. However, clinical similarity does exist between Peter's anomaly and posterior keratoconus. In Peters' anomaly, contrary to posterior keratoconus, the corneal endothelium and Descemet's membrane are absent or thinned out.

Histologically, epithelium is disorganized, Bowman's layer is replaced by fibrous tissue, stroma becomes thinned and structural changes are seen in Descemet's membrane. In some cases absence of Bowman's membrane in generalized posterior keratoconus is also seen. In the same patient, amyloid deposits were seen in the stroma<sup>13</sup>. Histology of our case was not done and amyloid deposits were also not seen. Recently, slit-scanning topography analysis (Orbscan) and Anterior segment OCT is used to identify the structural and histopathological abnormalities in cases of posterior keratoconus.

Although many ocular and systemic associations of posterior keratoconus are described in literature but our patient had no such associations. Ocular anomalies include anterior cleavage syndrome. Other anterior segment abnormalities include aniridia, ectropion uvea, iris atrophy, glaucoma, anterior lenticonus, ectopia lentis, and anterior lens opacities.

Systemic abnormalities include cleft lip and palate, genitourinary abnormalities, short stature, and mental retardation. None of these changes were present in our case.

## CONCLUSION

Posterior keratoconus is a rare corneal disorder in which visual drop is not marked because of the normal anterior curvature. Visual disturbance usually occurs as a result of stromal scarring, anterior segment anomalies or amblyopia. Corneal Topographic maps are helpful in understanding the basic pathology of this disease.

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