

Classical signs of Keratoconus

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Abstract

Keratoconus is a bilateral progressive disease of unknown etiology characterized by gradual corneal thinning, steepening and ectasia. Though corneal topography plays a significant role in its diagnosis, early identification with clinical signs can greatly improve the visual outcome with appropriate management. This PG snippet elaborates on the classical signs of keratoconus by clear demonstration with slit lamp pictures.

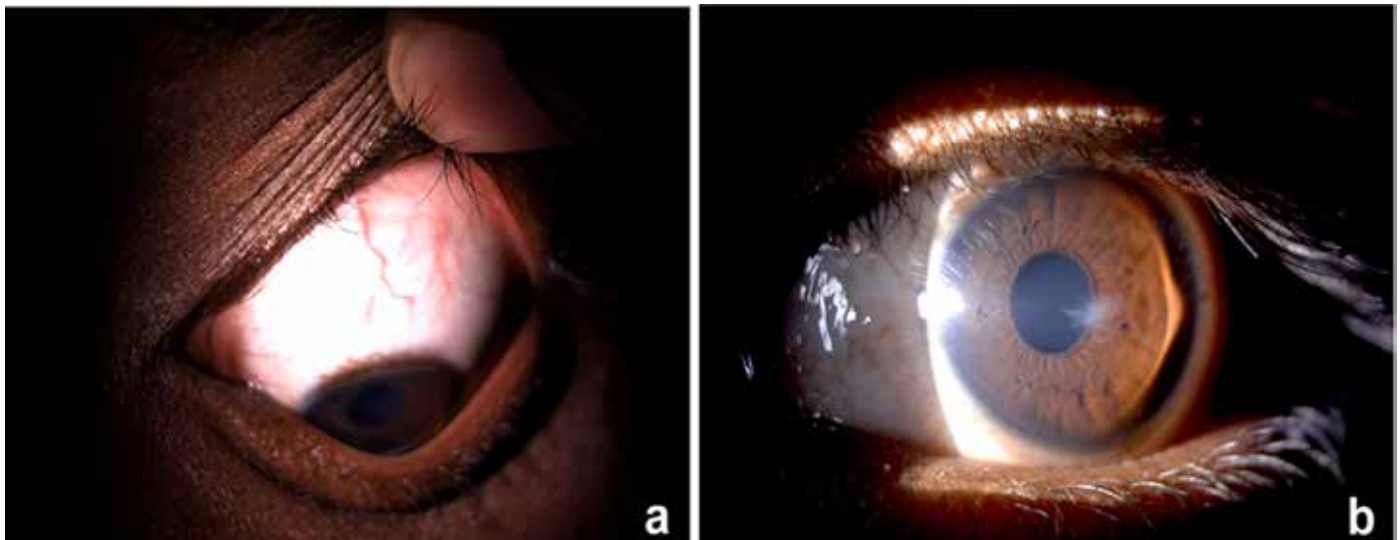
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External Signs

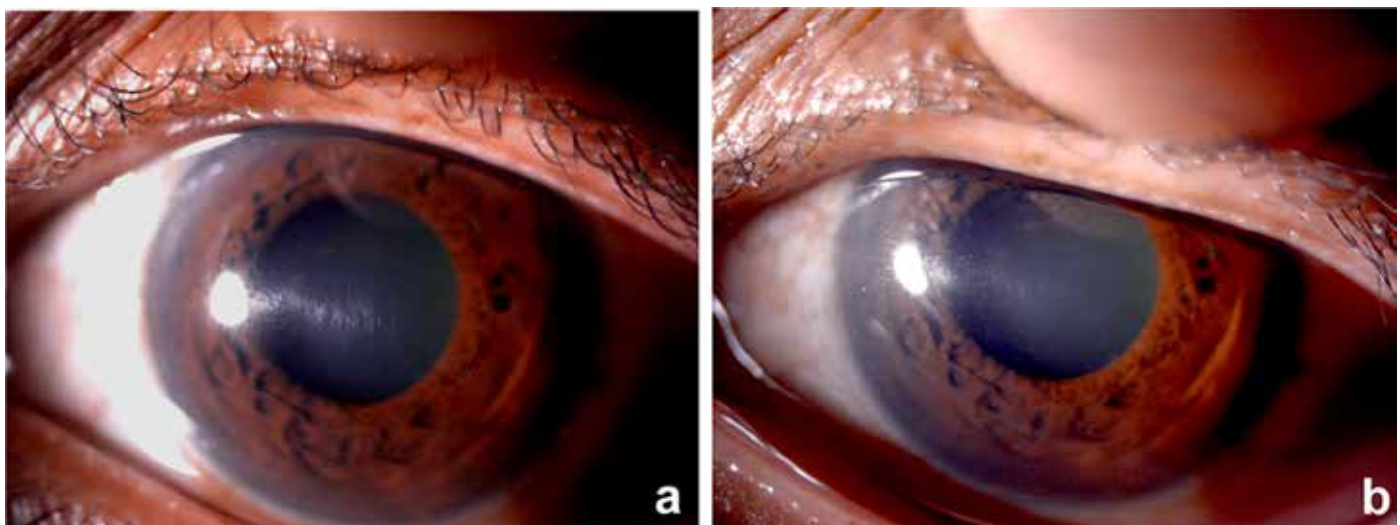
1. **Munson's sign:** V-shaped conformation of the lower lid produced by the ectatic cornea in down gaze (Figure 1a).
2. **Rizzuti's sign:** Sharply focused conical beam of light near the nasal limbus, produced by lateral illumination of the cornea from temporal side, in patients with advanced keratoconus (Figure 1b).

Slit Lamp Signs

1. **Vogt's striae:** Fine vertical stress lines in the deep stroma and descemet's membrane that are parallel to the steep axis of cone (Figure 2a). These lines disappear transiently on gentle digital pressure (Figure 2b).
2. **Fleischer ring:** Deposition of iron in the basal epithelial cells in a ring shape at the base of the conical protrusion



Figures 1: 1(a) Munson's sign 1(b) Rizzuti's sign



Figures 2: (2a) Vogt's striae (2b) Disappearance of striae on digital pressure

(Figure 3a). This ring is faint and broad in early keratoconus and becomes thinner and more discrete as the condition advances. It is best appreciated in cobalt blue or green filter (Figure 3b).

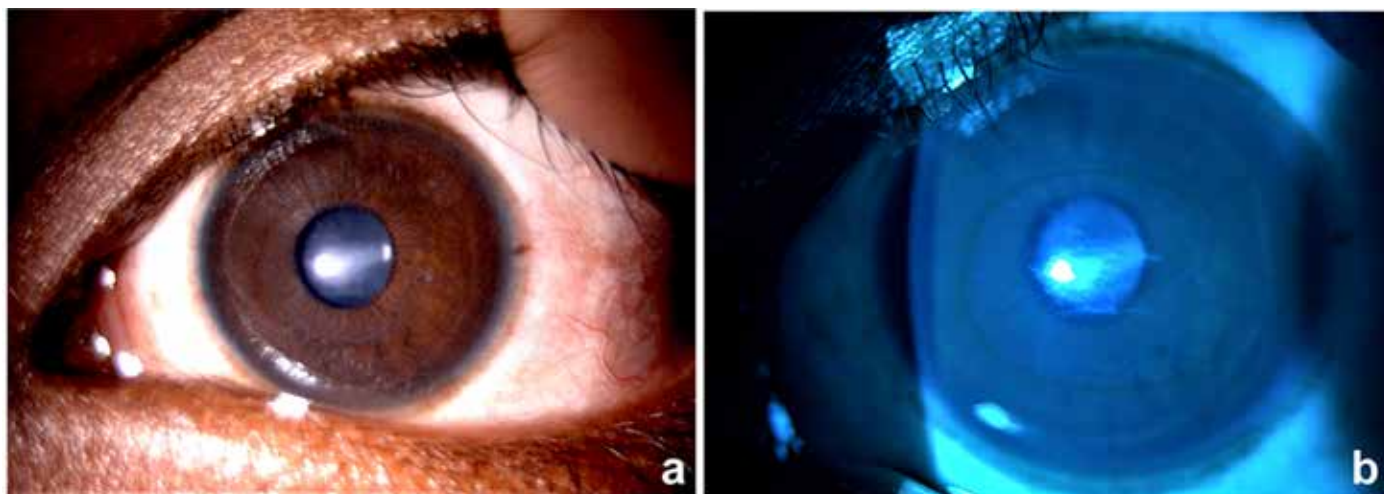
3. **Corneal thinning:** Slit lamp examination in keratoconic eyes show central or inferior corneal thinning. Maximum corneal thinning corresponds to the site of maximum steepening. (Mild Keratoconus: Figure 4a) (Severe Keratoconus) (Figure 4b).
4. **Corneal apical scarring:** Approximately 20 percent of eyes with moderate or severe keratoconus develop corneal scarring. It occurs as a part of natural progression of the disease but is worsened by the wearing of rigid contact lenses. It usually appears at the apex of the cone, starting as fine lines initially and then develop into nebular scarring as it progresses (Figure 5a).
5. **Hydrops:** An acute rupture in Descemet's membrane causing imbibition of aqueous into corneal stroma causing it to swell. It is a complication usually seen in patients with advanced keratoconus with associated allergy or eye rubbing (Figure 5b).
6. **Prominent corneal nerves:** It is not likely that the nerve

fibers are more numerous in keratoconic patients, but only that they are more easily seen due to changes in density. An increased visibility of the corneal nerve fibers cannot be considered a singular distinction of keratoconus. (Figure 6a)

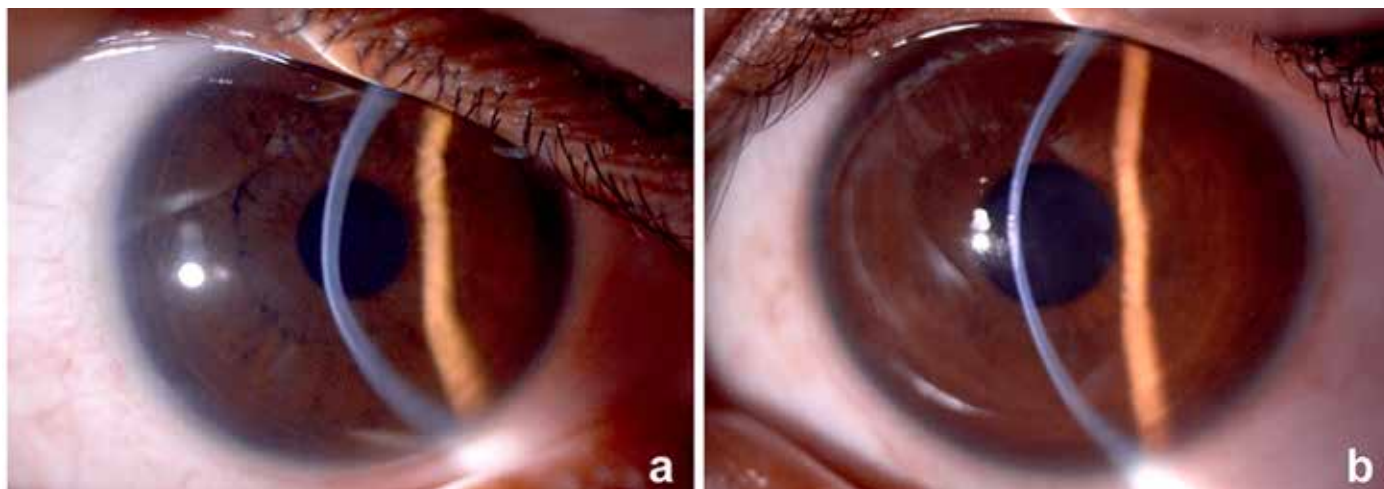
7. **Rupture in Bowman's layer:** These are irregular superficial opacities that develop due to breaks in Bowman's layer. They can cause significant visual loss due to scarring in advanced cases. (Figure 6b)

Retroillumination Signs

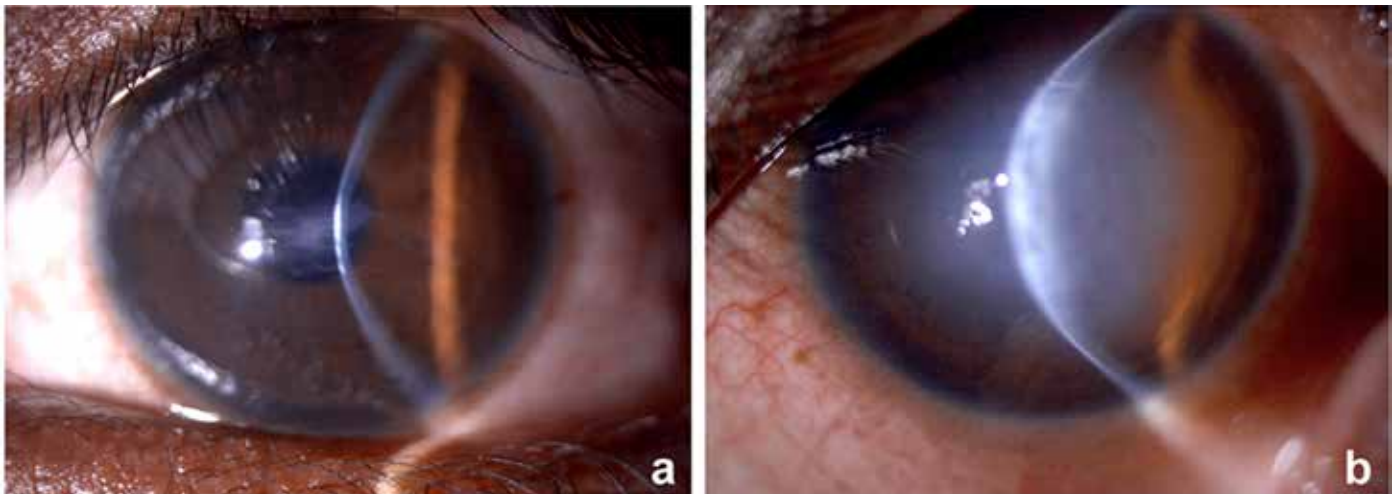
1. **Scissoring reflex:** During retinoscopy, two bands of the light beam reflexed from the retina, moves like the blades of the pair of scissors, towards and away from each other. This sign is diagnostic of keratoconus.
2. **Oil droplet sign ("Charleaux" sign):** Dark Reflex in the area of the cone on observation of cornea in dilated pupil by distant direct ophthalmoscopy. The total internal reflection of light due to conical cornea produces a dark, round shadow in corneal mid periphery, separating the central bright red fundus reflex from a red reflex in corneal periphery.



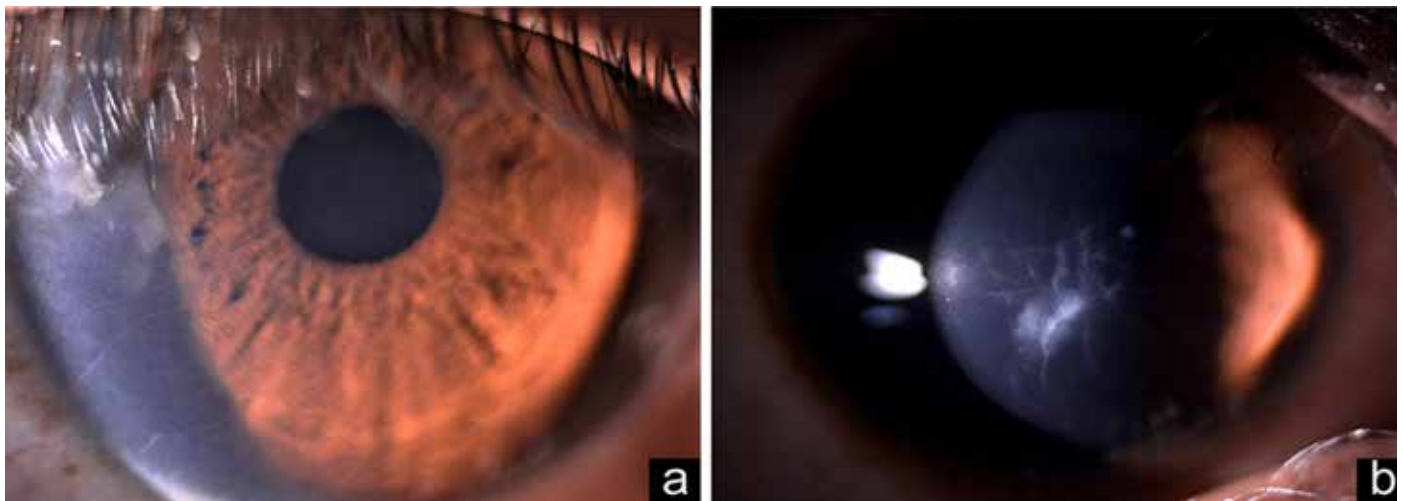
Figures 3: (3a) Fleischer's ring (3b) Brown ring in Green filter



Figures 4: (4a) Thinning in Mild Keratoconus (4b) Thinning in Severe Keratoconus



Figures 5: (5a) Apical corneal scarring (5b) Acute Hydrops

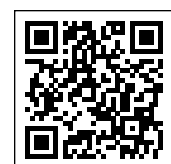


Figures 6: (6a) Prominent corneal nerves (6b) Bowmans rupture

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