

## Case Report

# Bilateral Circumscribed Posterior Keratoconus: Visualization by Ultrasound Biomicroscopy and Slit-Scanning Topography Analysis

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This paper documents a rare nonprogressive developmental disorder—bilateral circumscribed posterior keratoconus—in a 60-year-old man referred for a cataract surgery. For the first time ultrasound biomicroscopy was used to visualise the local anterior bulging of the posterior corneal surface with concomitant thinning of the stroma. The amount of localized posterior depression, corneal thickness and the refractive power of both the posterior and anterior corneal curvature were measured using slit-scanning topography analysis (Orbscan).

## 1. Introduction

Abnormal variation of the posterior corneal curvature may occur in two forms: the generalized posterior keratoconus, characterized by an regular increase of the curvature of the entire posterior corneal surface has, and the circumscribed posterior keratoconus, in which a localized paracentral or central posterior corneal indentation is seen [1]. In the generalized form, the corneal stroma typically remains clear. In contrast, the circumscribed posterior keratoconus shows stromal opacities overlying the localized anterior ectasia of the posterior surface, which may occupy the full stromal thickness [2]. The visual loss is not progressive and moderate [3]. Vision deterioration usually is caused by corneal scarring or amblyopia. Circumscribed posterior keratoconus is usually bilateral and sporadic, but familial cases have been also documented [4]. Despite the anterior protrusion in some cases, posterior keratoconus does not progress to anterior keratoconus and normally requires no treatment. Usually it is detected during routine ophthalmic examination. We describe a case of bilateral posterior circumscribed keratoconus.

## 2. Case Report

The 60-year-old white male of Mediterranean origin presented for a cataract extraction on his left eye. Visual acuity was 20/25 in the right eye and light perception in the left eye due to cataract formation. There was no amblyopia in the left eye before the onset of cataract. The patient denied history of injury, reporting only a bilateral ocular infection in childhood was reported. There were no systemic conditions.

Slitlamp examination revealed a bilateral paracentrally localized depression of the posterior curvature measuring 3 mm in diameter. There was scarring in the overlying corneal stroma (Figures 1, 2, and 3). An intraepithelial iron line was noted at the base of the lesion temporally. A few retrocorneal melanin granules were present (Figure 3). An irregular mosaic-like pattern was noted using retroillumination (Figure 4). The posterior depression was clearly detectable using ultrasound biomicroscopy (Humphrey, Zeiss, Oberkochen) (Figure 5) and slit-scanning topography analysis (Orbscan, Bausch and Lomb) (Figure 6). The amount of localized posterior depression was 75  $\mu\text{m}$  as indicated by topography. Corneal thickness measured 450  $\mu\text{m}$  within the lesion and 540  $\mu\text{m}$  in the adjacent healthy

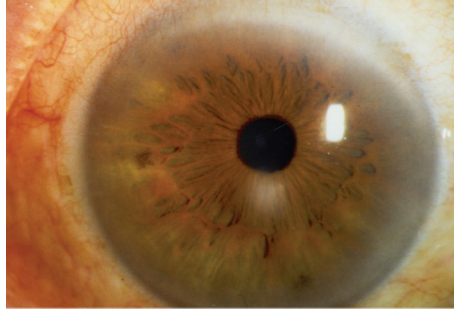


FIGURE 1: Right eye showing paracentrally inferiorly circumscribed corneal opacification.

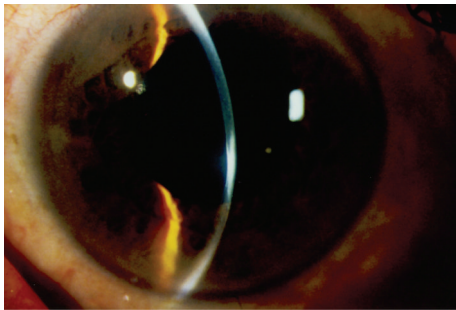


FIGURE 2: Slitlamp photograph showing circumscribed protrusion of the posterior corneal curvature with concomitant stromal thinning and an opacification of the overlying stroma.

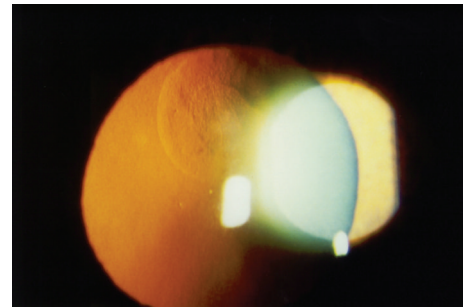


FIGURE 4: Retroillumination shows an irregularity with mosaic-like pattern. Note the sharp margin of the round lesion (arrow). There is a second sharp round line (arrowhead), forming a central and a peripheral zones.

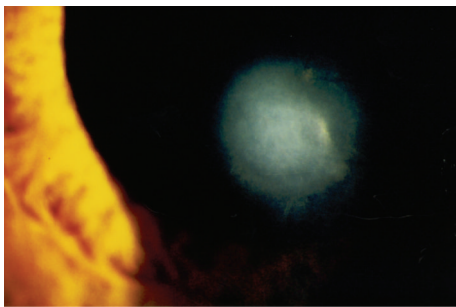


FIGURE 3: High magnification shows a relatively dense opacification of the cornea. Note the retrocorneal melanin granules at the edge of the stromal opacity.

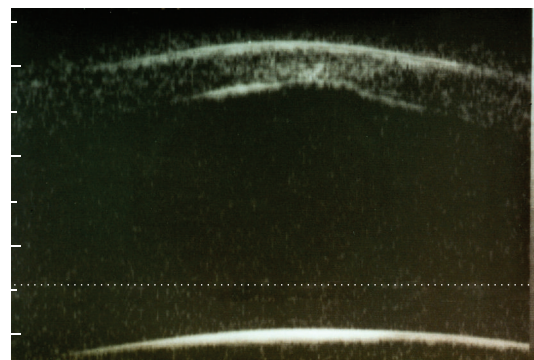


FIGURE 5: Ultrasound biomicroscopy shows the local anterior bulging of the posterior corneal surface with concomitant thinning of the stroma. Note the configuration of the enhanced stromal reflectivity (arrowhead) corresponding to the stromal opacity.

cornea using the Orbscan system. The refractive power of both the posterior and anterior corneal curvature was 50 to 56 diopters within the paracentral area. Otherwise, both eyes were unremarkable. Following phacoemulsification and posterior chamber lens implantation visual acuity increased to 20/50 in the left eye. The examination of the fundus and vitreous revealed no pathological findings.

### 3. Discussion

The clinical and topographic findings in this patient are consistent with the *paracentral keratoconus posterior circumscriptus* [5]. This is the first report on ultrasound

biomicroscopy to visualise the local anterior bulging of the posterior corneal surface with concomitant thinning of the stroma. Light microscopy of this abnormality has shown focal disorganization of basal epithelium and basement membrane, a replacement of Bowman's layer by fibrous tissue, a thinned stroma with an irregular arrangement of the central collagen lamellae, and a variable appearance of Descemet's membrane [6] with posterior excrescences indentating the vacuolated endothelium correspond to the corneal guttae seen in specular reflection [7]. Iron deposits



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