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# Unmasking of subclinical keratoconus with Descemet membrane endothelial keratoplasty in Fuchs endothelial dystrophy

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## Abstract:

We report a case of Fuchs endothelial corneal dystrophy (FECD) with concurrent forme fruste keratoconus (KCN) that was unmasked with Descemet membrane endothelial keratoplasty (DMEK) in the right eye, but not with Descemet-stripping automated endothelial keratoplasty (DSAEK) in the left eye. The patient was a 65-year-old female with FECD who underwent uncomplicated combination cataract surgery and DMEK in the right eye. She subsequently developed intractable monocular diplopia associated with inferior displacement of the thinnest point of the cornea and subtle steepening noted on posterior corneal curvature on Scheimpflug tomography. The patient was diagnosed with forme fruste KCN. Altering the surgical plan to combine cataract surgery and DSAEK in the left eye successfully circumvented the development of symptomatic visual distortion. This is the first case providing comparable data from contralateral eyes in the same patient regarding the outcome of DMEK versus DSAEK in eyes with concurrent forme fruste KCN. DMEK appeared to unmask posterior corneal irregularities and resulted in visual distortion, whereas DSAEK did not. The additional stromal tissue in DSAEK grafts appears to help normalize alterations of the posterior corneal curvature and may be the preferred endothelial keratoplasty for patients with concurrent mild KCN.

## Keywords:

Descemet membrane endothelial keratoplasty, Descemet stripping endothelial keratoplasty, forme fruste keratoconus, Fuchs endothelial dystrophy

## Introduction

Keratoconus (KCN) is a bilateral, noninflammatory corneal ectasia, characterized by progressive thinning and steepening greatest at the corneal apex. A clinical diagnosis of KCN can be made based on the presence of characteristic findings, including high myopia, irregular astigmatism, anterior stromal scarring, and corneal hydrops (tears in Descemet's membrane).<sup>[1]</sup> However, advances in anterior segment imaging now allow the identification of subclinical stages of KCN, termed forme fruste KCN. Steepening of the posterior corneal curvature seen

on Scheimpflug tomography has been described as one of the earliest signs of KCN.<sup>[2]</sup>

Fuchs endothelial corneal dystrophy (FECD) is a late-onset, sporadic corneal dystrophy characterized by progressive corneal endothelial dysfunction, focal excrescences of Descemet's membrane called guttae, and corneal edema. In contrast to KCN, FECD results in central corneal thickening. Surgical treatment for FECD involves the replacement of the corneal endothelium, either by Descemet-stripping automated endothelial keratoplasty (DSAEK), which includes a layer of posterior stroma in addition to Descemet's membrane and

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endothelium, and Descemet membrane endothelial keratoplasty (DMEK), which repositions Descemet's membrane and endothelium only.<sup>[3,4]</sup> More recently, Descemet stripping only in which the Descemet membrane is removed without placement of a donor tissue has been demonstrated to be an option for some patients with FECD.<sup>[5]</sup>

Despite no known genetic associations, KCN and FECD both demonstrate a relatively high prevalence and may occur concurrently by chance, with estimated comorbidity of 1:100,000.<sup>[6]</sup> The corneal edema in FECD can mask the thinning and ectasia seen in KCN, and changes in the posterior corneal surface after endothelial keratoplasty may exacerbate the visual distortion from KCN. However, there have been no studies comparing the merits of DMEK versus DSAEK in the setting of underlying forme fruste KCN. We report a case of forme fruste KCN that became symptomatic after DMEK for FECD but was successfully treated with DSAEK in the contralateral eye.

## Case Report

A 65-year-old female with a history of FECD presented to the UC Davis Health Eye Center with worsening glare and blurry vision despite the use of sodium chloride 5% ointment twice daily in both eyes (OU). Her best-corrected visual acuity (BCVA) was 20/20 in the right eye (OD) and 20/25 in the left eye (OS). Glare testing reduced her vision to 20/50 OD and 20/50 OS. Her manifest refraction was Plano +0.75 × 160 OD and +0.50 + 0.50 × 135 OS. Ultrasound pachymetry demonstrated a central corneal thickness of 603 um OD and 607 um OS. Slit-lamp biomicroscopy revealed significant corneal guttae, mild corneal edema, and cortical cataract OU. There was no evidence of Vogt's striae, Fleischer rings, or other findings suggestive of KCN. Lenstar-LS900 (Haag-Streit AG, Koeniz, Switzerland) optical biometry was performed in preparation for combined cataract extraction with intraocular lens (CEIOL) implantation and DMEK OD. Keratometry from the Lenstar demonstrated Ks of 42.81 and 43.74 diopters (D) with the steep axis at 031.

The patient subsequently underwent uneventful combined CEIOL and DMEK in the right eye. Her postoperative eye drop regimen included ofloxacin 0.3% four times daily for 2 weeks and prednisolone acetate 1% four times daily, slowly tapered to once daily for 1 year. Her immediate postoperative course was without complications.

In postoperative month 3, however, the patient reported several weeks of intermittent monocular diplopia OD, described as a "trail of lights," "doubleness," and

"shadows on letters." The best spectacle-corrected visual acuity at that time in the right eye was 20/25 with a manifest refraction of -0.25 + 2.00 × 152. The cornea was clear and compact with a fully attached DMEK donor tissue, and there was no evidence of retinal pathology or cystoid macular edema. Scheimpflug tomography was obtained at postoperative month 6, which showed a stable anterior cornea with Ks of 43.3 D and 44.1 D, but an inferotemporal posterior elevation of + 22 um [Figure 1a-c]. The patient's visual symptoms persisted despite spectacle correction and removal of mild posterior capsular opacification with ND: YAG capsulotomy, but resolved with a scleral lens.

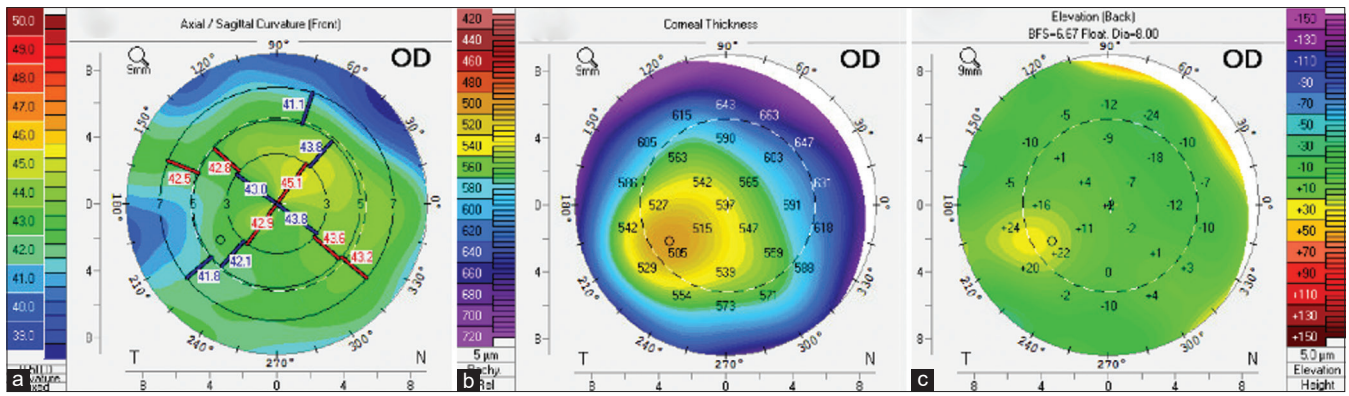
Given the evidence of forme fruste KCN OD, the decision was made to defer DMEK OS, and the patient underwent a combination of CEIOL and DSAEK OS. Preoperatively, tomography showed Ks of 42.4 D and 43.1 D, with minimal anterior or posterior astigmatism [Figure 2a-c]. Postoperatively, the patient received moxifloxacin 0.5% four times daily for 2 weeks, prednisolone acetate 1% four times daily, slowly tapered to once daily for 1 year, and sodium chloride 5% ointment nightly OS. The patient had an uncomplicated postoperative course after DSAEK surgery OS without the development of monocular diplopia. In postoperative year 1 after surgery in OS, her BCVA was 20/20 OD in a scleral lens and 20/25 OS with spectacles. Tomography OS showed Ks of 41.7 D and 42.9 D, with minimal changes to the anterior cornea. Evaluation of the posterior curvature revealed a uniform central contour without the development of focal elevation [Figure 3a-c]. The patient reported subjectively preferring the vision OS compared to OD.

## Discussion

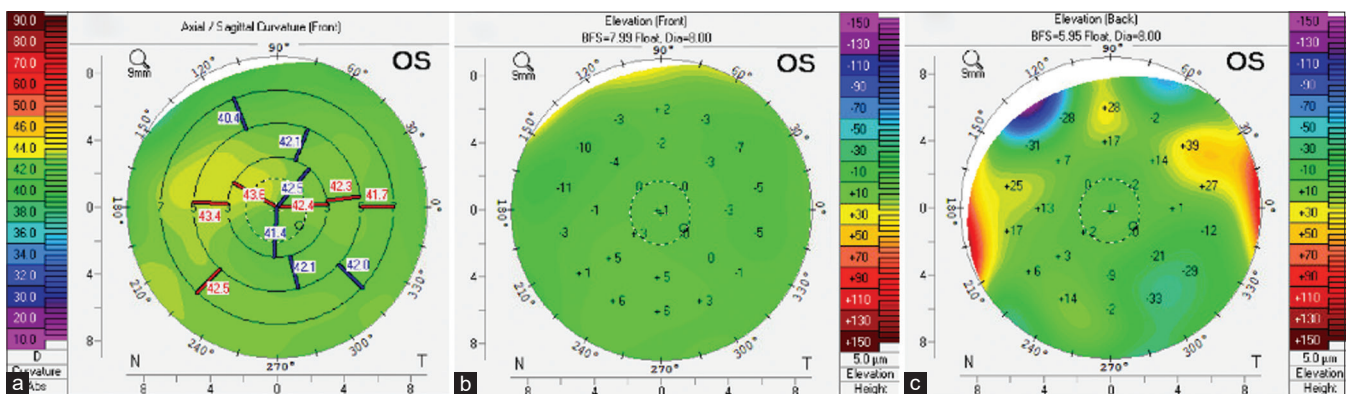
We have described a case of concurrent forme fruste KCN and FECD, where the patient developed symptomatic monocular diplopia with associated posterior corneal elevation after treatment of FECD with DMEK but reported excellent visual acuity with DSAEK in the contralateral eye.

The term forme fruste KCN has been used to refer to early, subclinical manifestations of KCN, often seen as subtle changes on topography, tomography, or epithelial thickness mapping, but without classic signs on manual keratometry, retinoscopy, or biomicroscopy.<sup>[7,8]</sup> Changes in the posterior cornea, including steepening, increased elevation, and irregularity, have been identified as early signs of KCN and correlate with decreased visual acuity.<sup>[9,10]</sup>

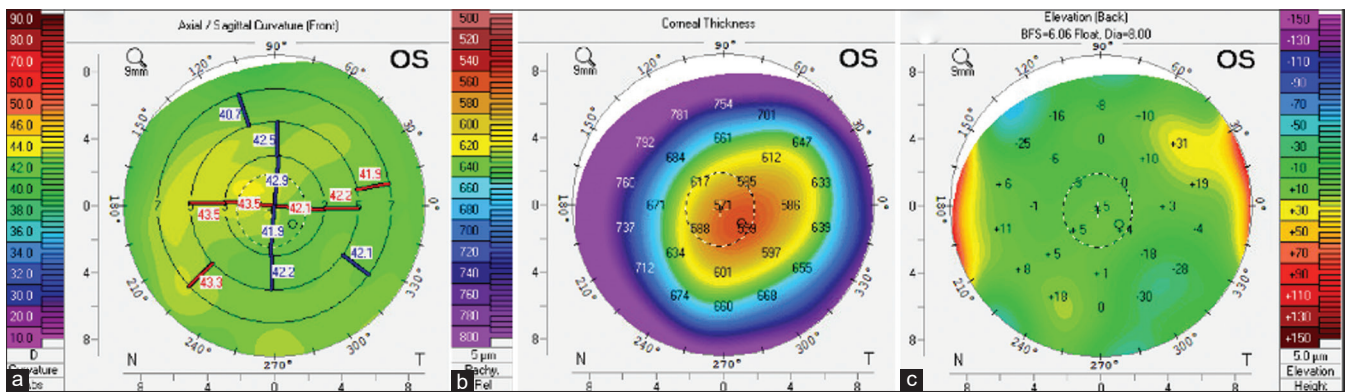
In contrast, corneal edema in FECD induces central thickening and flattening of the posterior corneal curvature. Examination of corneas with FECD before and



**Figure 1:** Postoperative month 6 status post-Descemet membrane endothelial keratoplasty tomography of the right eye shows minimal changes to the anterior curvature (a), but an inferotemporal displacement of thinning (b) associated with posterior elevation consistent with forme fruste keratoconus (c)



**Figure 2:** Preoperative tomography of the patient's left eye with Fuchs endothelial corneal dystrophy showing minimal anterior astigmatism (a), normal anterior elevation map (b), and posterior corneal flattening consistent with mild corneal edema associated with Fuchs (c)



**Figure 3:** Postoperative year 1 tomography after DSAEK OS demonstrates minimal changes in the anterior curvature (a), decreased corneal thickness (b), and a symmetric posterior curvature with mild peripheral steepening but without focal elevation (c)

after DMEK demonstrates that corneal deturgescence is accompanied by posterior corneal steepening, hyperopic shift, and forward displacement of the posterior surface by an average of 73  $\mu\text{m}$ .<sup>[11]</sup> Meanwhile, the anterior corneal curvature is minimally affected.<sup>[11]</sup>

In patients with concomitant KCN and FECD, the corneal edema from FECD may completely mask subtle cases of forme fruste KCN. Mild deviations of the posterior corneal curvature may remain asymptomatic and

undetectable until the resolution of the corneal edema. In this case, while the anterior corneal curvature remained stable, the development of a posterior cone after DMEK was associated with symptomatic monocular diplopia.

Price *et al.* reported a similar case of a 62-year-old male with FECD who underwent DMEK and subsequently requested evaluation for LASIK.<sup>[12]</sup> Examination revealed postoperative refraction of  $+0.75 + 2.50 \times 115$ , a central corneal thickness by ultrasound pachymetry of 481,

minimal anterior corneal changes, and focal posterior corneal elevation of 25 um concerning for forme fruste KCN not seen on preoperative imaging.<sup>[12]</sup> Cooper *et al.* also reported masking of anterior and posterior corneal steepening in a patient with KCN, which only became apparent after the resolution of corneal edema with DMEK.<sup>[13]</sup> These cases suggest that evaluation of the anterior or posterior corneal curvature by topography/tomography is an unreliable screen for forme fruste or early KCN in patients with FECD. Further study is required to determine the characteristic features and alternative screening methods for KCN in the setting of corneal edema.

Because DSAEK includes a layer of corneal stroma in addition to the Descemet membrane and endothelium, it may be the preferred endothelial keratoplasty for normalizing the posterior corneal curvature in patients with concomitant KCN. In this case, given the unsatisfactory outcome of DMEK in the right eye and the bilateral nature of KCN, the patient underwent DSAEK in the contralateral eye, recovered without the development of visual distortion postoperatively, and reports superior quality of vision with DSAEK compared to DMEK.

Vira *et al.* reported excellent outcomes in a case series of six eyes with concomitant FECD and mild-to-moderate KCN after DSAEK, with all patients improving to BCVA of 20/40 or better.<sup>[14]</sup> However, in this series, topography showed postoperative flattening of the anterior corneal curvature in these patients.<sup>[14,15]</sup> This discrepancy may be due to breaks in Bowman's membrane in more advanced KCN, which destabilize the anterior corneal curvature, allow for anterior steepening in the setting of corneal edema, and result in a greater than expected hyperopic shift after deturgescence.<sup>[15]</sup>

Concurrent FECD presents unique and complex challenges to the identification and management of KCN, especially forme fruste and early KCN. In these cases with minimal anterior corneal distortions, DSAEK may offer superior outcomes to DMEK by minimizing keratoconic deviations of the posterior cornea.

### Ethical statement

Due to the nature of case reports and the absence of identifiable information in the manuscript, this did not qualify as human subject research by the Institutional Review Board of University of California Davis indicating (1971253-1) and thus did not require a written consent form from the patient.

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Nil.

### Conflicts of interest

The authors declare that there are no conflicts of interest in this paper.

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