

## Late-Onset Keratoconus Prior to Cataract Surgery?

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Careful preoperative evaluation prior to cataract surgery is integral in delivering improved refractive outcomes. Corneal topography, in particular, has become an invaluable tool in preoperative planning. With this in mind, a 62-year-old Caucasian male presented for cataract evaluation OD. Preoperative corneal topography identified mild inferior steepening OU (Figure 1). Cataract surgery was performed OD with an excellent postoperative course.

Two years later, the patient returned with a visually-significant cataract in the fellow eye. Corneal topography was repeated (Figure 2). At this time, the I-S ratio – a metric used to evaluate the severity of keratoconus – was measured at 2.67. This confirmed progressive inferior steepening OS when compared to the initial topography from two years prior, which measured an I-S ratio of 2.24 (Figure 2). Furthermore, repeat biometry showed a 1.5-D reduction in the preferred intraocular lens (IOL) power from 19.5 D to 18.0 D due to an increase in corneal power over two years (Figure 3).

At this juncture, the differential diagnoses of unilateral irregular astigmatism were considered. These differentials included ocular surface disease (OSD), epithelial basement membrane dystrophy (EBMD), development of a Salzmann nodule, corneal scarring, and keratoconus. Our patient's history was negative for most of the aforementioned diagnoses, and the anterior and posterior segment exams were entirely normal, with the exception of a decreased tear breakup time and trace inferior punctate epithelial erosions, consistent with OSD.

The diagnosis of progressive unilateral keratoconus was entertained, given the worsening inferior steepening and significant I-S ratio demonstrated on topography. While progressive keratoconus is less common in patients in their 60s, keratoconus itself does occur with relative frequency. Risk factors for progressive keratoconus include vigorous eye rubbing and a significant family history; however, neither of these were present.

Following treatment for ocular surface disease, the patient successfully underwent cataract surgery OS with implantation of a monofocal IOL. The choice of IOL power was decreased by 1.5 D when compared to IOL calculations from two years prior to compensate for the increased corneal curvature, and to attain the desired postoperative outcome of slight myopia to emmetropia. The lens power choice highlights the importance of repeat biometry if there exists any doubt as to the accuracy of initial measurements, or if there might have been an extended period of time between measurement and surgery, as was the case here.

It is of note that the choice to place a monofocal IOL was made given the irregular astigmatism and the concern for the possibility of other underlying corneal pathology. Placing a multifocal IOL in such cases increases the risk of decreased patient satisfaction and reduced quality of vision.

The best corrected visual acuity (BCVA) immediately post-cataract extraction was 20/25 OS. Two months later, the patient returned complaining of worsening blurry vision OS with a BCVA of 20/30. Anterior and posterior segment exams appeared normal; however, following instillation of fluorescein, the patient demonstrated a small area of negative staining inferiorly (Figure 4), which was not evident preoperatively. The area of negative staining was consistent with the diagnosis of map-dot-fingerprint dystrophy or EBMD. Coincidentally, the area of EBMD observed on slit lamp exam corresponded to the worsening inferior steepening demonstrated on corneal topography. Because of this, a diagnosis of EBMD masquerading as a late-onset unilateral

keratoconus was made. Since the slit lamp exam of the fellow eye was entirely within normal limits, without evidence of EBMD, we also considered that OS findings might simply represent localized epithelial hyperplasia.

EBMD is found to be present in 6-18% of the population, predominantly in females greater than 50 years old.<sup>1</sup> The typical presentation may include blurry vision, monocular diplopia, ghost images, and recurrent corneal erosions. There have been reported cases of EBMD falsely masquerading as keratoconus. Stein et al retrospectively reviewed 1000 cases of presumed corneal ectasia (keratoconus, keratoglobus, pellucid marginal degeneration) referred for possible corneal crosslinking (CXL), PRK, or intrastromal ring placement.<sup>2</sup> Twenty-six eyes of 20 patients were falsely diagnosed with an ectatic disorder. Of these misdiagnosed cases, the true underlying pathology in nine eyes was EBMD. Other etiologies included superficial punctate keratitis in seven eyes, high astigmatism in three eyes, amiodarone keratopathy in two eyes, corneal warpage in two eyes, corneal scars in one eye, and measurement error in two eyes. Furthermore, Reinstein et al found utility in the use of epithelial thickness profiling (Artemis VHF ultrasound) to confirm non-keratoconic pathology in eyes that would have normally been classified as keratoconic on topography.<sup>3</sup>

Treatment of EBMD is not necessary if the patient is asymptomatic; however, lubricants, hypertonic solutions, and contact lenses may be trialed as initial conservative management. Surgical treatments for visually-significant EBMD include simple epithelial debridement, with or without diamond burr polishing, and phototherapeutic keratectomy (PTK). In a study by Itty et al, 74 eyes in 55 patients underwent simple mechanical debridement with average BCVA improvement from 20/44 to 20/33.<sup>4</sup> In this study, EBMD recurred in 18 of 74 eyes. There were no infections or persistent epithelial defects reported, but subepithelial haze was present in 19 eyes. A study by Lee et al sought to examine the efficacy of PTK in the management of EBMD.<sup>5</sup> Fifty-eight eyes in 51 patients who failed conservative management for EBMD experienced an average BCVA improvement from 20/32 to 20/25 with PTK. In eyes with recurrent erosions (n=29), the average BCVA improvement was from 20/25 to 20/20. They concluded that PTK was safe and effective over the long term with comparable results to other surgical modalities.

After providing informed consent, our patient decided to undergo PTK. Following the procedure, the topography normalized (Figure 5) and vision reached a BCVA of 20/20. The power of the cornea (keratometry) returned to the value as that of the original biometry readings performed at the time of initial presentation (from 3 years prior). The now flatter keratometry resulted in a hyperopic shift to +1.50 + 1.00 x 180. Although the patient's BCVA was 20/20, the patient desired to be spectacle-free, and the patient underwent a piggyback IOL procedure to improve distance vision.

In summary, this case brings to light a unique manner in which EBMD may present, as well as the medical and surgical implications of this presentation. Specifically, the case demonstrates the importance of a thorough slit lamp exam with the use of fluorescein, repeated pre- and postoperative testing to ensure accurate assessments of keratometry, and the use of monofocal IOLs in cases where other underlying corneal pathology might be present.

## References

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4. Itty S, Hamilton SS, Baratz KH, Diehl NN, Maguire LJ. Outcomes of epithelial debridement for anterior basement membrane dystrophy. *Am J Ophthalmol*. 2007;144(2):217-221.
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## Disclosures

Jonathan Go, DO: No financial Interest

Alice Wong, DO: No financial Interest

William Trattler, MD: Consultant JNJ

Steven Litinsky, MD: No financial Interest

# 2011

OD

OS

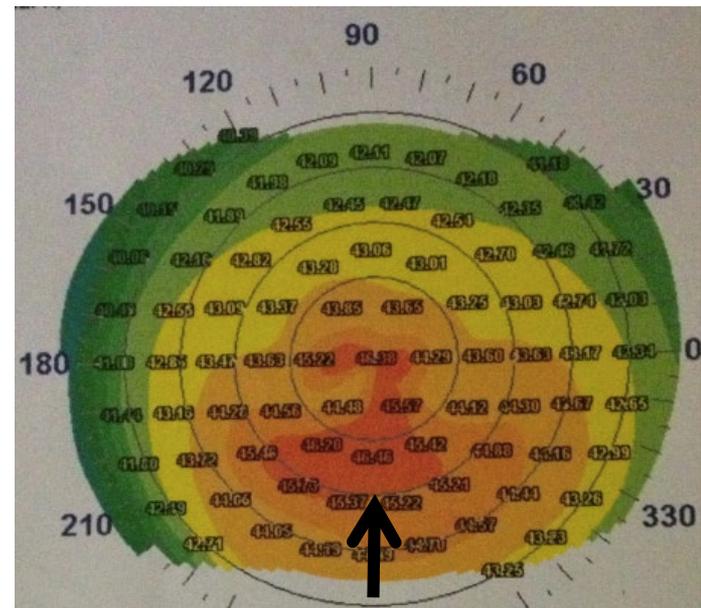
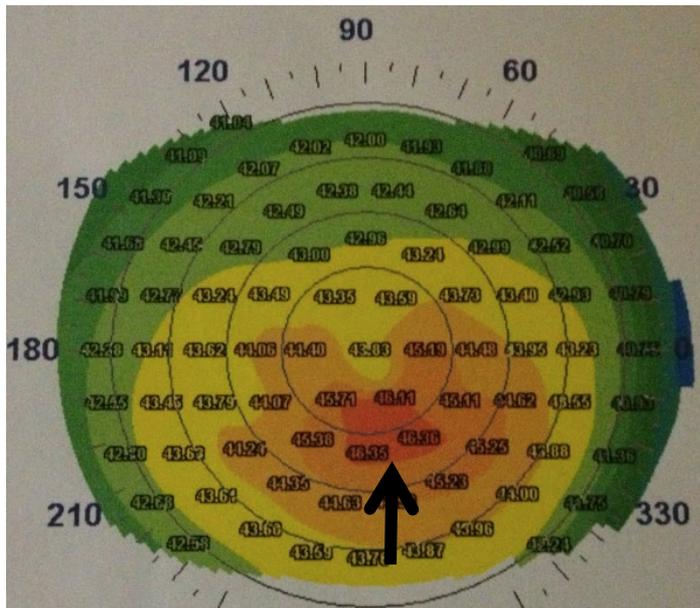
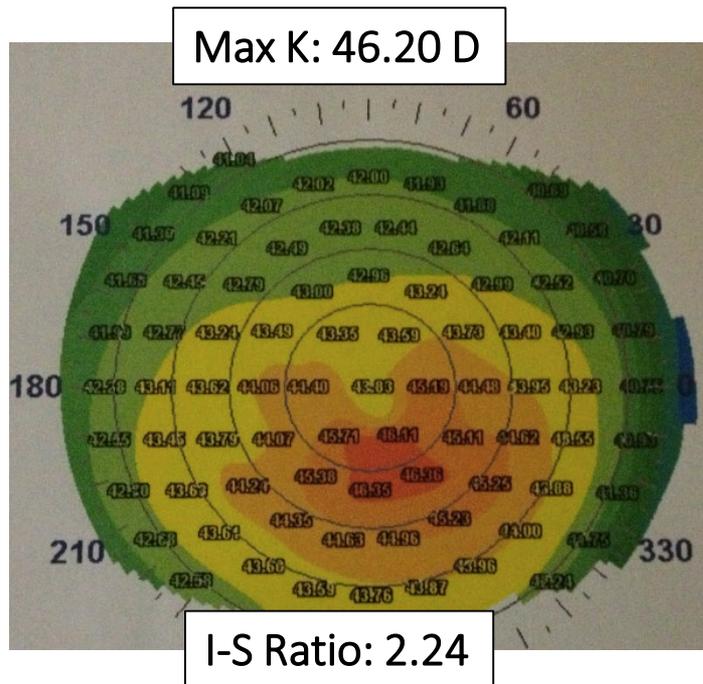


Figure 1 Corneal topography OU from 2011 demonstrated mild inferior steepening OS>OD (*black arrows*).

# OS

2011



SEPTEMBER 2013

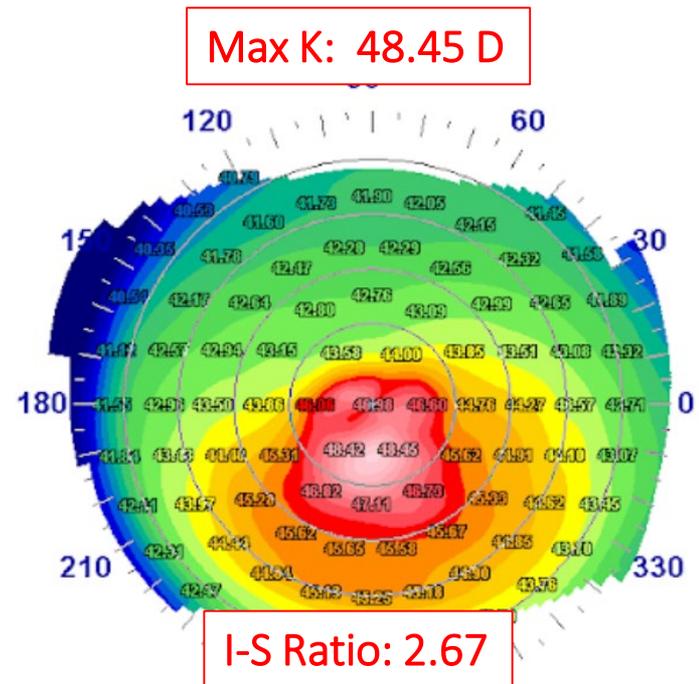


Figure 2 A comparison of corneal topography OS showed progressive inferior steepening with an increased I-S ratio over two years, suggesting keratoconus.

# Biometry OS

2011

SEPTEMBER 2013

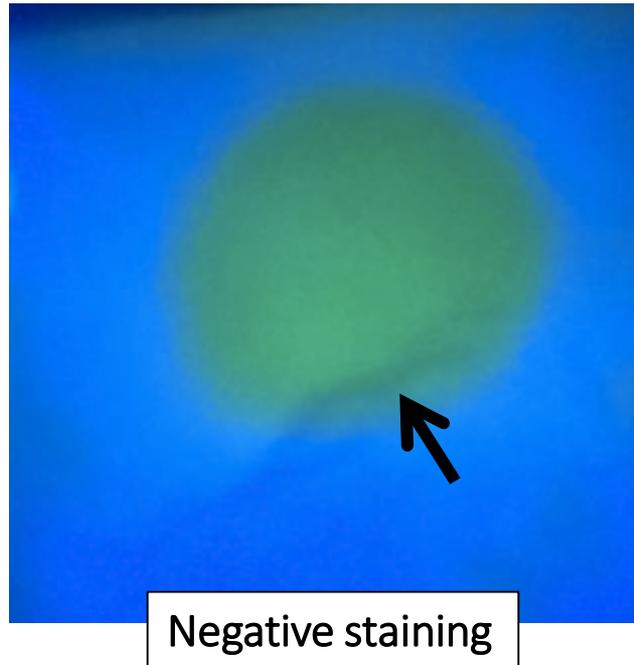
Significant change in keratometry over 2 years

2011				2013			
OS left				OS left			
AL: 24.04 mm (SNR = 127.5) K1: 43.83 D / 7.70 mm @ 53° K2: 44.76 D / 7.54 mm @ 143° R / SL: 7.62 mm / 44.30 D Cyl.: 0.93 D @ 143° ACD: 3.37 mm Status: Phakic				AL: 24.07 mm (SNR = 162.4) K1: 45.36 D / 7.44 mm @ 0° K2: 45.73 D / 7.38 mm @ 90° R / SL: 7.41 mm / 43.33 D Cyl.: 0.37 D @ 90° ACD: 3.36 mm Status: Phakic			
TECNIS MF1 ONE ZMB00		SOFTEC-HD		Tecnis MF1 ONE ZMB00		SOFTEC HD	
A const: 119.3		A const: 118.4		A const: 119.30		A const: 118.40	
IOL (D)	REF (D)	IOL (D)	REF (D)	IOL (D)	REF (D)	IOL (D)	REF (D)
21.5	-1.58	19.5	-0.94	20.0	-1.50	19.0	-1.54
21.0	-1.24	19.25	-0.77	19.5	-1.18	18.5	-1.20
20.5	-0.91	19.0	-0.60	19.0	-0.86	18.0	-0.86
20.0	-0.58	18.75	-0.43	18.5	-0.50	17.5	-0.53
19.5	-0.25	18.5	-0.26	18.0	-0.23	17.0	-0.21
19.0	0.07	18.25	-0.09	17.5	0.00	16.5	0.12
18.5				17.0			
18.0				16.5			

1.5-D reduction in IOL power from 19.5 D to 18 D over 2 years

Figure 3 Biometry OS showed significant change in keratometry between 2011 and 2013 that contributed to a 1.5-D reduction in IOL power.

# Fluorescein Stain OS



**Figure 4** Slit lamp exam of cornea with fluorescein dye revealed an inferior epithelial ridge with negative staining (*black arrow*) suggestive of EBMD.

# Topography OS January 2014

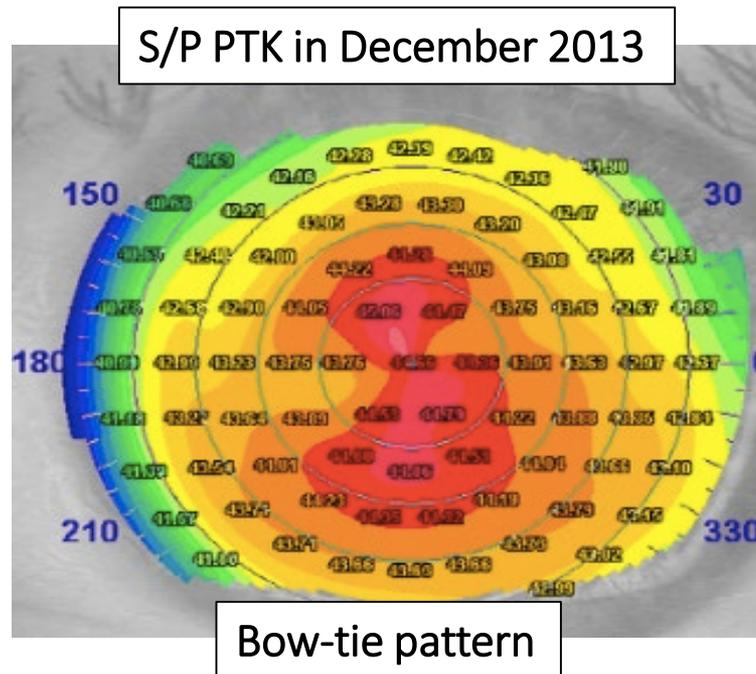


Figure 5 Topography OS from January 2014 following PTK in December 2013 demonstrates the bow-tie pattern of regular astigmatism.