

Classifying Keratoconus

Surgeons discuss how they define mild keratoconus, keratoconus, and forme fruste keratoconus, and they share their top indices for distinguishing between the three corneal states.

BY STEPHEN G. SLADE, MD, FACS; CLARK SPRINGS, MD; WILLIAM B. TRATTLER, MD;
AND TREVOR WOODHAMS, MD

STEPHEN G. SLADE, MD, FACS

Keratoconus is corneal degeneration that is characterized by ectasia, a thinning and bowing forward of the cornea. Keratoconus is typically a bilateral, progressive disease. Keratoconus is thought by ophthalmologists to be partially genetic and is associated with several risk factors, including contact lens wear, eye rubbing, connective tissue disorders, ocular allergy, and Down's syndrome.

Forme fruste keratoconus is a subclinical disease and not a variant of keratoconus. Although clinicians use many other terms such as *mild keratoconus*, *early keratoconus*, and *subclinical keratoconus*, their exact meaning and application is less certain. These terms are not universally accepted. The diagnosis of keratoconus is a clinical one that is aided by topography. The diagnosis of forme fruste keratoconus is topographic. I use patterns of topography that have been described by Rabinowitz¹ and Binder et al² to diagnose forme fruste keratoconus. These patterns include inferior steepening and asymmetric bow ties with a skewed radial axis.

Importantly, there are no specific laboratory tests or numbers generated by diagnostic instruments that can make the diagnosis of forme fruste keratoconus or keratoconus. No specific indices or combination of any numerical level of any indices has been scientifically validated to diagnose keratoconus. All numbers such as steepest K reading, original pachymetry, and even the inferior/superior ratio must be considered in the context of the clinical picture. There are no magic numbers. Indeed, there are patients with the "genetics" for keratoconus whose disease will not be detectable at the time of

refractive surgery. Conversely, other patients with single or several signs of keratoconus may undergo refractive surgery and do quite well.

CLARK SPRINGS, MD

As refractive surgeons, we are experts in discriminating between normal and keratoconic corneas. We rely on our clinical judgment after carefully evaluating patients' manifest refraction, streak retinoscopy, pachymetry, corneal topography, wavefront analysis, and biomicroscopic examination. The daily clinical challenge is the cornea with borderline metrics. Specifically, where does one draw the line between normal variance and forme fruste keratoconus?

"No specific indices or combination of any numerical level of indices has been scientifically validated to diagnose keratoconus."

—Stephen G. Slade, MD, FACS

Because keratoconus by definition is a biomechanical disease process, placido disc corneal topography, in my opinion, is currently the best validated tool to screen for for early disease. Several screening algorithms exist that evaluate the cornea for asymmetry, which is one of the earliest signs of keratoconus.

Recently, technologies capable of imaging the posterior surface of the cornea have become available such as the

Orbscan topographer (Bausch & Lomb, Rochester, NY), Pentacam (Oculus, Inc., Lynwood, WA), and Visante (Carl Zeiss Meditec Inc., Dublin, CA). Although these systems have yet to be widely validated, it is known that the posterior surface of the cornea bulges forward in keratoconus. My colleagues and I recently evaluated 100 keratoconic corneas and 100 controls with the Orbscan topographer.³ We found that a posterior corneal protrusion was a very sensitive and specific indicator of keratoconus, along with the well-established anterior indicators such as steep keratometry and corneal asymmetry.

Many questions remain. Where do keratoconic changes appear first? Do they appear on the anterior or posterior surface of the cornea, or is it a combination of anterior and posterior changes? Furthermore, is the currently available technology sensitive enough to make such a distinction? In the interim, the clinician must continue to weigh all available data carefully.

WILLIAM B. TRATTLER, MD

Ectasia following LASIK was first described 8 years ago.⁴ We ophthalmologists have learned that patients who develop the disease in many cases have reduced corneal strength, either because the residual stromal bed is too thin or the cornea itself has decreased durability. We still do not have any tests to measure corneal strength directly, so we must rely on topographic changes that typically occur in patients with reduced corneal stability. The challenge is that some patients may not manifest changes in their topographic or Orbscan maps until their late 20s or 30s.

Screening criteria for forme fruste versus frank keratoconus were developed by Rabinowitz and Maeda et al.^{1,5} When forme fruste keratoconus or keratoconus is present, patients are at increased risk of developing post-LASIK ectasia. In my practice, I have found that using a combination of the Humphrey Atlas Topography System (Carl Zeiss Meditec Inc.) and Orbscan topographer allows me to identify patients with corneas that may be at increased risk for ectasia.

With the Humphrey topographer, I first make sure that both eyes of a patient are symmetrical, because asymmetry is a potential sign of forme fruste keratoconus. I identify the direction and degree of astigmatism. If the astigmatism is horizontal (against the rule), I am very suspicious for pellucid marginal degeneration and look carefully for any sign of a lobster claw pattern. If the astigmatism is vertical (with the rule), forme fruste keratoconus can be a concern. A helpful screening tool is the degree of inferior steepening, or the inferior/superior ratio. A value of 1.40D or more is suspicious for forme fruste keratoconus. Steep corneas (above 48 μ m) also raise the sus-

picion for forme fruste keratoconus. With the Orbscan topographer, a useful but not yet validated test is the posterior float. I become suspicious when the posterior float is greater than 0.04mm. The pachymetry maps on Orbscan topography can also be helpful. Both thin corneas and those with inferior pachymetry that is significantly thinner than superior pachymetry can also be a risk factor for forme fruste keratoconus.

For frank keratoconus, topography can reveal a number of findings, including steep K readings (often above 50) and an inferior/superior ratio greater than 2.00D. There are typically corneal signs (Fleischer ring, Vogt striae, apical scarring) and a reduction in BCVA.

TREVOR WOODHAMS, MD

With the tremendous success of keratorefractive laser surgery, it is certainly appropriate to try to provide its benefits to as many patients who seek it as possible. What are my colleagues and I to do, however, with the several patients who have atypical corneas and want to be free of optical prostheses?

First do no harm. Although the original contraindication labeling by the FDA, "signs of keratoconus," was based on a concern for corneal stability, it became apparent that compromised corneas often responded poorly enough to rule out routine LASIK.

From the increasing reports in clinical practice of cases of ectasia, the search for a reliably predictive analysis of atypical corneas has begun to assume a degree of importance it never had previously. Because there is no clear test for unstable corneas, we as surgeons have had to rely primarily on Placido-based topography supplemented by generalized patterns of pachymetry and topography (eg, Orbscan II [Bausch & Lomb]).

Unfortunately, several inherent aspects of the technology limit all Placido-based keratometry. First, it identifies only a relative (rather than an absolute) centration of corneal mapping, which occurs because the visual axis' intersection with the corneal surface determines the apparent corneal center. If the center of the cornea does not coincide with the corneal apex, it will appear that there is asymmetrical curvature (which is not necessarily the case). Second, the tear layer can affect accuracy. When the eye is dry, areas of unstable surface irregularities will appear. Conversely, in the presence of a very thick tear film, some abnormalities will be obscured. Last, the use of a best-fit sphere as a reference for posterior float, because the natural shape of the cornea is a prolate ellipsoid, the float will be exaggerated. All of these aspects will generate a high false-positive "abnormal" pattern. Furthermore, although a number of suspicious topographic patterns have been dubbed red flags by consult-

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ants for Bausch & Lomb in trying to identify the various “keratoconoid” conditions (forme fruste keratoconus, mild keratoconus, and pellucid marginal degeneration), none of these has actually been validated.

After 2 years of familiarizing myself with the technology, I have made the switch to what I consider true elevation corneal analysis in the form of the Pentacam. With this device, I no longer need to pay as much attention to superior/inferior steepness indices, because they can now be measured directly relative to the actual corneal apex rather than the corneal point of intersection between the visual axis and Placido plane.

In assessing corneas for possible keratoconoid conditions, I look for thin and/or ectatic areas that correspond to abnormally high posterior and anterior floats relative to a best fit ellipsoid (these can be central but are usually eccentric). ■

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