Editorial

Can You spot the Keratoconus Suspect?

We have come a long way since the Placido disk was first used to detect the corneal distortions characteristic of keratoconus. We learned that automatic computerized image analysis could be used to accurately determine the mire positions reflected from the corneal surface.[1] Although there were no exact mathematical formulas with which to use these data to reconstruct corneal curvature and shape, we learned that approximations could be devised that would faithfully reveal corneal topography.[2] We also learned how to avoid misalignment effects that could produce corneal topographies with the asymmetries characteristic of keratoconus.[3] But like most developing technologies, as alternative methods and instruments were developed, not all corneal topographers received passing grades on their accuracy and freedom from artifact.[4,5] On the contrary, Placido corneal topography today is a mature and standard diagnostic test that is essential for corneal care.

Because of its high sensitivity to subtle changes in corneal curvature, Placido corneal topography is the principle test used to detect the earliest surface changes associated with keratoconus. Ultrastructural studies have shown that keratoconus often develops as a localized subepithelial degenerative change in the organization of Bowman's layer. These subtle beginnings initiate biomechanical weakening thinning, and changes in surface curvature. At present, detection methods that are sensitive enough to measure these very early changes in patients are not available with the exception of anterior surface corneal topography.

As we review methods for keratoconus detection, it is important to point out criteria that must be met when evaluating these. First and foremost, the method should be able to detect the keratoconus suspect, apart from being able to recognize the signs of clinical keratoconus where changes in thickness may be present and measureable. The keratoconus suspect cornea will have subtle changes in its anterior curvature that may include inferior steepening with or without an asymmetric bowtie, a truncated or foreshortened bowtie, a bowtie that shows skewing or simply abnormal central steepening. Other subtle abnormalities may include the 'D' shape[6] or the arcuate fish mouth or 'C' shape characteristic of topographic pellucid marginal degeneration. The term forme fruste keratoconus should be reserved for post hoc observations describing keratoconus suspect corneas that progressed to clinical keratoconus or as a label for a normal-looking fellow eye of a 'unilateral' keratoconic patient. We should accept and, if our notion that keratoconus has genetic underpinnings is correct, consider that keratoconus is a bilateral condition and that use of 'unilateral' should refer to the unexpressed state in a fellow eye of a keratoconic.

Methods for keratoconus suspect detection have been developed,[7] but there are few implementations that have been validated in the literature. Often, tests claiming to include the keratoconus suspect group will be comprised of the undeveloped fellow eyes of a keratoconic. Such eyes are not suspect, but are forme fruste keratoconus as noted above. Other studies have included corneas with major amounts of inferior steepening asymmetry (I-S > 1.9 D[8]) beyond that associated with keratoconus suspect, but instead characteristic of clinical keratoconus. It seems clear that there are greatly differing opinions for grading keratoconus and some of the difficulties arising have been discussed by Gatinel and Saad.[9] In fact, variability in corneal topography interpretation by clinicians has been an ongoing issue that is under intense study currently (IC Ramos, R Ambrósio Jr, personal communication). Despite the availability of standards for the display of corneal topography,[10] not all clinicians use the same display methods. Hence, interpretation becomes an individual learning task that is not easily adaptable to additional or collective opinion in evaluating questionable cases.

The issue today is not how to detect clinical keratoconus; there are now ample means to view the corneal curvature changes with either topography or tomography and to examine pachymetric maps for localized thinning or radial anomalies. The greatest risk factor for the development of ectasia after refractive surgery is the finding of abnormal corneal topography,[11] and, after ruling out contact lens molding, the major cause of abnormal corneal topography among refractive surgical candidates is keratoconus. The most sensitive way to detect keratoconus suspect remains anterior surface curvature display supplemented with validated pachymetry analyses. Hope springs eternal that one day a sensitive enough methodology will be developed to catch the very early biomechanical weakening thought to accompany keratoconus suspect or that the genetic basis for keratoconus and screening method will be discovered to give us a better tool for diagnosis. Until then, we must proceed with caution and learn corneal topography interpretation well to deliver the safe management of refractive error to our patients.

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Stephen D Klyce PhD FARVO
Editorial Board Member