Keratoconus is a bilateral conical ectasia of the cornea with well-described clinical and slit-lamp microscopic signs. Subclinical forms of this ectasia are known to exist in individuals with normal corneas on slit-lamp microscopic examination. The incidence of keratoconus is approximately 1 in 2000 in the general population and may be much higher if subclinical forms are taken into account.

These patients experience early spectacle blur and multiple contact lens changes and might present themselves as candidates for myopic refractive surgical procedures. Poor results in individual patients who have had myopic refractive keratoplasty could in some instances be attributed to undetected subclinical keratoconus. Computer-assisted videokeratography systems are sensitive devices for detecting subclinical keratoconus. They cost from $25,000 to $35,000.

We solicited the opinions of researchers, clinicians, and refractive surgeons on two questions relating to the early detection of keratoconus: 1) Is computer-assisted videokeratography the most sensitive means of detecting early keratoconus? and 2) Should every candidate for myopic refractive surgery be screened with this device? The following are their responses:

Is computer-assisted videokeratography the most sensitive means of detecting early keratoconus?

Given the current generation of computerized corneal topography systems (CCTS), it surprises me that there is any question as to the most sensitive method by which to diagnose keratoconus. No other clinical tool or diagnostic sign has the clear advantage afforded, in particular, by the color-coded contour map of corneal surface powers. This presentation method is now used routinely to detect early corneal ectasias in clinics around the world. With this technology, keratoconus appears as a region of surface power noticeably greater than that of the surround. This area of elevated power can be located anywhere on the corneal surface (including the central and superior regions), although there is a tendency for cones to emerge inferonasally. CCTS can detect keratoconus before the patient experiences the earliest symptoms such as spectacle blur. Even in the best hands, traditional methods have missed obvious (by CCTS) cases of keratoconus.

One caveat: contact lenses can warp corneas in such a way as to mimic keratoconus, particularly in patients with corneal cylinder who wear lenses that ride off center. Keratoconus and contact lens warp are differentiated by discontinuing contact lens wear until topography stabilizes, which may take several weeks.

STEVE KLYCE, PhD
Louisiana State University
New Orleans, La

The answer is a difficult one. A best spectacle corrected visual acuity of worse than 20/20 is a clue that something might be wrong with the cornea. Finding irregular and scissoring red retinoscopic reflexes indicates that something is wrong with the ocular media. After identifying either or both of these situations, the clinician might then look for objective signs of keratoconus. I find that inferior corneal steepening upon slight upgaze as measured by the keratometer is a very sensitive and reliable sign of early keratoconus. Careful examination with a broad oblique slit beam might reveal a diffuse brownish haze which is the beginning of the iron ring. Pachometry just below the visual axis often discloses thinning that is too subtle to be noticed using the slit-lamp microscope. We are just beginning to learn the value of computer analysis of corneal curvature. In the future, I believe that computerized imaging will be the most sensitive way to diagnose early keratoconus.

JAY KRACHMER, MD
University of Iowa
Iowa City, Iowa

Computer-assisted keratography is a valuable tool for assessing corneal topography since it gives an overall perspective of the corneal surface. Currently, this modality is more quantifiable when dealing with
Keratoconus & Videokeratography/Rabinowitz

a spherocylindrical corneal surface than when dealing with an aspheric surface, such as following refractive surgery, when it tends to be more qualitative. Such limitations are imposed by attempting to digitalize distorted keratography mires, even with sophisticated computer software. The manual keratometer, however, remains the most sensitive instrument for determining irregular astigmatism; the presence of which, even in the face of a normal corneal exam, indicates very early keratoconus.

LEE NORDAN, MD
La Jolla, Calif.

Videokeratography provides rapid information concerning the corneal asymmetry and progressive astigmatism in keratoconus. Although digitization of the photographs is required to determine the subtle inferior temporal steepening, which is characteristic of the early stages of keratoconus, recent documentation of the superior and inferior corneal asymmetry on digitized photokeratographs or videokeratographs emphasizes the value of screening by these techniques. This asymmetry cannot be visualized on undigitized photographs with vertical disparities under 2.00 diopters. However, the same disparity can be appreciated readily by an asymmetry on the retinoscopy reflex on dilated pupil exam during "radical retinoscopy" as described by Jack Copeland. In this technique, the ophthalmologist retinoscopes from a working distance of 4.00 inches from the patient's face.

The narrowing of the inferior side of the retina's myopic image proves that the inferior cornea is steepening more rapidly than the superior cornea. This subtle sign of keratoconus is rarely appreciated because this technique is a lost examination procedure which is rarely taught today. Retinoscopy can discriminate as little as 0.50 diopters of corneal asymmetry by this technique, and is, therefore, a valuable screening modality for keratoconus. The classical bull's eye red fundus reflex seen in keratoconus is a further accentuation of the same appearance.

JAMES ROWSEY, MD
University of South Florida
Tampa, Fla.

The question of the most sensitive way to diagnose keratoconus really depends on the minimal threshold criteria to define keratoconus. Such criteria are usually inadequately defined or not defined at all in much of the literature. Likewise, the criteria may change depending on the status of the patient's other eye, the family history, etc. Thus, video or photokeratography may be the most sensitive way to diagnose keratoconus in the contralateral, normal-appearing eye of a patient with keratoconus in the first eye. Identical changes, however, in an asymptomatic individual with no slit-lamp biomicroscopic evidence of progression since childhood are more difficult to call keratoconus. At the present time, a combination of history slit-lamp biomicroscopic evaluation, fundus reflex evaluation, and keratography are all important in confirming the diagnosis of keratoconus.

JOEL SUGAR, MD
University of Illinois
Chicago, Ill.

A major factor in the efficacy and safety of refractive surgical procedures is the preoperative detection of subclinical diseases of the cornea, the most common of which is early keratoconus. In my clinical experience, objective clinical signs are relatively late indicators of disease. Maguire, Rabinowitz, and our own group has shown that the earliest manifestations of keratoconus are frequently detectable only with computer-assisted topographic analysis. The disease may even escape detection by visual inspection of keratoseope mires when the computer digitized keratograph clearly shows the characteristic abnormalities. In addition, other preoperative abnormalities of the corneal topography that are common in candidates for refractive surgical procedures such as contact lens-induced corneal warpage may go undetected without preoperative keratography. I, personally, would not consider performing any refractive surgical procedure without excluding the possibility of keratoconus or other topographic abnormalities with keratography.

STEVEN WILSON, MD
University of Texas
Dallas, Tex.

Comments by Dr Rabinowitz: Techniques for detecting early keratoconus can be divided into five broad categories: slit-lamp microscopy, retinoulation techniques, pachometry, keratometry, and corneal topography. Slit-lamp microscopic signs include: early corneal thinning, vertical striae at the level of Descemet's membrane (Vogt's striae), and an incomplete epithelial circular iron ring (Fleischer ring).

Retinoulation techniques comprise the following: the "Charleaux oil droplet sign" with ophthalmoscopy, scissoring of the red reflex by retinoscopy, and "radical retinoscopy" (as described by Rowsey above). Distortion of the keratometry mires and regional keratometry are considered sensitive methods for detecting keratoconus. What is not commonly recognized is that thinning detectable by pachometry only is often present prior to the onset of clinically detectable disease.8

The most sensitive of the five modalities is the study of the paracentral corneal topography. Amsler2 in the early 1900s showed that early keratoconus in the absence of clinical signs was detectable by observing the deflection of a horizontal line on a
hand-held photographic placido disc; subsequently Rowsey and coworkers in the 1970s using a nine-ring photokeratoscope to study the topography of the cornea showed that an egg-shaped mire (a sign of inferotemporal steepening) was detectable in the absence of slit-lamp microscopic signs of the disease. To this day, the hand-held Klein keratoscope (made by Keeler, costing $400) remains an excellent inexpensive tool for detecting early keratoconus. The problem with all subjective interpretation methods is that interpretation of the abnormalities of the corneal curvature is dependent on the clinician's experience. Early keratoconus detected by one clinician may be interpreted as normal by another. In this respect, computer-assisted corneal topography systems represent a significant advance. Once the clinician is familiar with the technical use of the device and gets acquainted with the interpretation of digitized keratographs, these devices have been shown to be highly accurate and reproducible at least in the central two-thirds of the cornea. Over the past 3 years, I have studied several hundred normal and keratoconic eyes using this device and have detected at least a dozen patients whose topographic maps suggest early keratoconus in the fellow eyes of patients with unilateral keratoconus or "keratoconus fruste" in family members of patients with keratoconus (Figure). In none of these patients was I nor my colleagues (all of whom have subspecialty training in cornea and external disease) able to detect any of the clinical signs described above for the early detection of keratoconus. Additionally, no abnormalities were detectable on examining their anterior corneal topography with a nine-ring photokeratoscope. As clinicians gain more experience with these devices, I have little doubt that the consensus of opinion will be that computer-based corneal topography systems are the most accurate and reliable means of detecting early keratoconus.

Should every candidate for myopic refractive surgery be screened with computer-assisted videokeratography?

Since there have not been any prospective studies to compare the diagnosis of keratoconus using "standard" clinical diagnostic techniques with videokeratography, I don't think anyone can claim one technique is superior to the other. I have personally examined one patient in whom the computer-assisted topography map suggested focal corneal steepening in the inferior visual axis in a pattern consistent with that of keratoconus. With slit-lamp-microscopic examination, keratometry, and examination of the red reflex, I was unable to ascertain evidence of keratoconus in either eye. Although, I personally believe that topography is an additional tool in making this diagnosis, I think there is insufficient information at this time to require it for a screening device for all candidates for refractive surgical procedures.

PERRY BINDER, MD
La Jolla, Calif

There is no doubt that performing any refractive surgery on patients with keratoconus is less predictable. Although stable, "subclinical keratoconus" may not be an absolute contraindication, it
would still be extremely important to identify these patients preoperatively. The diagnosis of keratoconus is a clinical one that includes visual and family history, best spectacle corrected visual acuity, and the presence of objective findings. In our practice, the tear-drop reflex on retinoscopy (after the patient has been out of rigid contact lenses for at least 2 weeks), the Humphrey automated keratometer with its confirmation percentages, and the International Diagnostic Cone-/scope are all used to detect early cases of keratoconus. There are no data available that would indicate a significant increase in detection of “subclinical keratoconus” with computer-assisted corneal topography. Until this is determined, we would most likely not use this additional, more expensive testing in our corneal and refractive practice.

RICHARD DAMIANO, MD
Littleton, Colo

I firmly believe that topographic studies must be obtained before refractive surgery is performed on any patient. Although advanced cases of keratoconus, pellucid marginal degeneration, and other corneal abnormalities can be easily diagnosed by a well-trained clinician, early cases frequently cannot. Keratometry is not an adequate diagnostic tool in early cases. Keratometry readings measure only a small (approximately 3.00-millimeter diameter) central area of the cornea, and the results often appear to be spherical or nearly spherical when the earliest pathologic changes are developing just outside the zone. Even when photokeratoscopy is studied by an experienced cornea specialist, subtle and early anomalies are often missed. Such early changes can also precede any changes in the retinoscope reflex as well. Since patients with early keratoconus are likely to be unhappy with their glasses or contact lenses for reasons they find difficult to articulate, they are likely to end up in an ophthalmologist's office requesting refractive surgery. I think that many of these people have, in the past, been included in refractive surgical studies, such as the PERK study, and they may be responsible for some of the outliers seen when the results are presented graphically.

MARGUERITE MCDONALD
Louisiana State University
New Orleans, La

There is no question that corneal topography is helpful in evaluating subclinical corneal surface abnormalities. The red fundus reflection, hand-held and photokeratoscopes can be just as helpful as videokeratography, although the video devices are faster and easier to use and present more detailed information. However, the heart of your question deals with preventing refractive surgery on a subclinical keratoconus cornea. In my clinical experi-

ence with many thousands of astigmatic and radial keratotomies over the past 12 years, I have found only a small incidence of keratoconus and only a few cases where the photokeratoscope detected subclinical keratoconus. To my knowledge, no undetected keratoconus cornea that underwent radial keratotomy surgery has progressed to full keratoconus. Certainly, any objective evaluation of corneal topography is helpful in evaluation of refractive surgery candidates.

ALBERT NEUMANN, MD
Deland, Fla

Since late spring 1990, videokeratography has been part of our routine examination of all candidates for keratorefractive procedures to correct myopia (PRK), astigmatism (PRK or holmium treatment), or hyperopia (holmium treatment). In 9 out of approximately 500 patients, we found peripheral steepening zones making these eyes highly suspicious for keratoconus. Keratometry readings, photokeratography, and slit-lamp microscope inspection revealed no other signs of keratoconus. Clearly, we would have missed these subclinical cases of keratoconus without corneal topography. There is broad agreement in the field that keratoconus corneas should not be operated on for refractive purposes by keratotomies or keratectomies. Therefore, we recommend examination of all candidates for refractive surgery by computer-assisted videokeratography to rule out keratoconus at any stage.

THEO SEILER, MD, PhD
Freie University, Berlin

Patients who have early forms of keratoconus, Terrien's marginal degeneration, pellucid degeneration, or forme fruste of these or abnormalities of corneal thickness or shape may have a very unpredictable response to refractive corneal surgery. Although these conditions can usually be diagnosed by a careful slit-lamp microscope examination and keratography, early cases may be missed easily by these examination techniques. Computer-assisted videokeratography is the most sensitive technique for detecting abnormalities of corneal shape. The sensitivity of present video keratography systems—about 0.25 diopters—far exceeds our ability to interpret the mires of the keratometry photographs subjectively. In our refractive surgery practice, we have diagnosed a number of patients with early keratoconus who were referred for refractive surgery. We think that computer-assisted videokeratography should be performed on all patients considering refractive keratoplasty to rule out the presence of an occult disorder of corneal shape or thickness.

KEITH THOMPSON, MD
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Precise measurement of the cornea is of course desirable, but we do not believe that keratoconus is an absolute contraindication for radial keratotomy. In reviewing Dr O’Dell’s records we found five eyes with keratoconus which had been treated by radial keratotomy. Reductions in astigmatism were 1) 9.25 to 0.50, 2) 8.75 to 2.00, 3) 6.75 to 3.00, 4) 4.00 to 0.75, and 5) 4.75 to 1.00 diopters. Increases in uncorrected distance visual acuity were 1) 20/400 to 20/50, 2) 20/2000 to 20/60, 3) 20/2000 to 20/60, 4) 20/100 to 20/25, and 5) 20/300 to 20/70. Patients were pleased by the improvement in visual acuity; and none has required a cornea transplant to date. These anecdotal results obtained without computer-assisted keratography imply that radial keratotomy may eventually prove to be surprisingly useful in the treatment of keratoconus. For medicolegal reasons (at least) it would be well to diagnose all keratoconus patients preoperatively. Keratometer readings and slit-lamp microscopy along with retinoscopy should suffice.

PETER WYZINSKI, DSc, MD
Eugene and Beaverton, Ore

Comments by Dr Rabinowitz: Several interesting and conflicting viewpoints are presented in these responses. Questions which need to be resolved are: Is it at all necessary to screen for keratoconus patients? Drs O’Dell and Wyzinski believe that radial keratotomy might not be a bad procedure for keratoconus. Is subclinical keratoconus a contraindication to refractive surgery? Comments by Drs Damiano and Neuman suggest that this might not be a contraindication and that subclinical forms are more rare than might commonly be thought. Given the expense of computer-based corneal topography systems and the possible low cost-benefit ratio to the practitioner, is keratometry and clinical examination alone sufficient for screening patients prior to refractive surgery?

Reports in the scientific literature indicate poor refractive surgical results in patients with keratoconus. I personally, along with Drs McDonald, Seiler, and Thompson would never perform PRK or radial keratotomy on any patient whom I thought had clinical or subclinical keratoconus. We still know very little about the pathogenesis of this disorder and trigger mechanisms for hastening its progression. Some reports suggest that trauma such as eye rubbing and hard contact lens wear may hasten its progression. Could the trauma from the excimer laser represent a stimulus for enzymes leading to progressive thinning of the cornea? Currently, we do not know the answers to these questions. We do know, however, that keratoconus can progress at any age. We, therefore, have a responsibility to explain this to our patients and indicate to them that because of the natural history of the disease we are unable to predict what the ultimate refractive outcome might be. We are rapidly learning how the excimer affects normal corneal tissue; in one reported case, it appears that the effect on corneal tissue of patients with keratoconus might be different from other corneas that were exposed to excimer laser photoablation for removal of superficial corneal opacities.

I think the best course of action presently is to exclude any patient with clinical or subclinical keratoconus from refractive corneal surgery until we develop a better understanding of the long-term effect of refractive surgery on keratoconus. Certainly, some long-term follow up with reports in the scientific literature on cases quoted by O’Dell and Wyzinski would be most useful; additionally it would be interesting to review data on excimer laser photoablation done on the clinically “normal” fellow eye of patients with unilateral keratoconus, as our experience is that this condition is almost always a bilateral one.

REFERENCES