Megalocornea

**BY JONATHAN D. CHRISTENBURY, MD; LOUIS E. PROBST, MD;
AND BRIAN S. BOXER WACHLER, MD**

CASE PRESENTATION

A 33-year-old female presents with a desire for refractive surgery. Topography measurements taken serially over the past 6 months show stability. A review of systems and the patient's past medical and ocular history are unremarkable. Her pupils are 5 mm in diameter and normal (Figure 1). Her manifest refraction is -3.00 +0.50 X 100 = 20/20 OD and -3.25 +0.50 X 81 = 20/20 OS. According to her history, the patient's manifest refraction is stable and matches her current prescriptions for contact lenses and glasses.

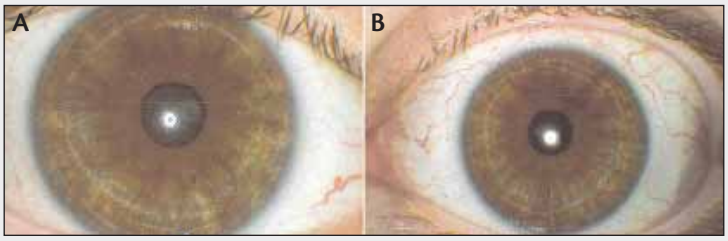

Figure 1. External photographs of the patient's right (A) and left (B) eyes.

Ultrasound pachymetry readings are 540 µm OD and 552 µm OS. The thinnest pachymetry measurements using the Pentacam Comprehensive Eye Scanner (Oculus, Inc., Lynnwood, WA) are 526 µm OD and 527 µm OS (Figures 2 and 3). The funduscopic examination is normal. The only abnormal finding is that her white-to-white measurement is 13.1 mm OD and 13.0 mm OS. The patient's ophthalmologist diagnosed megalocornea and referred her to you for a discussion of her options for refractive surgery. How would you proceed?

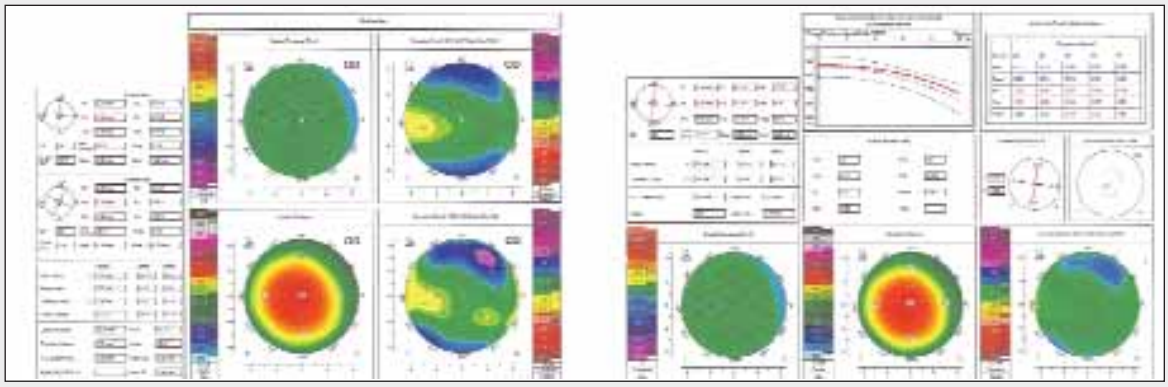

Figure 2. Analysis of the patient's right eye with the Pentacam.

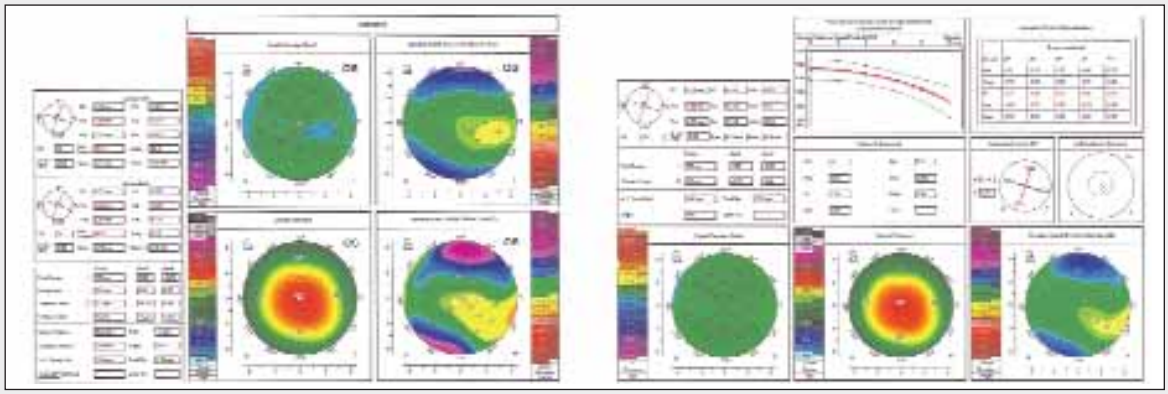

Figure 3. Analysis of the patient's left eye with the Pentacam.

JONATHAN D. CHRISTENBURY, MD

I have never seen a patient present for laser vision correction who had what I would diagnose as megalocornea. I understand that the histological appearance of the cornea is normal in this X-linked disorder. Due to its association with Marfan's syndrome and other ocular and systemic congenital anomalies, however, one may assume a possible abnormality of collagen synthesis.

If this patient is contact lens intolerant and is unable or unwilling to wear spectacles, laser vision correction might be an option. Normal pachymetry and topography readings, however, do not eliminate the possibility of abnormal collagen tissue/cross-linking and the risk of corneal ectasia. I would not perform LASIK and would be cautious to recommend PRK only after determining that corneal rigidity and hysteresis were normal on testing with the Ocular Response Analyzer (Reichert, Inc., Depew, NY). I would perform PRK on one at eye at a time and would repeat testing with the Ocular Response Analyzer after operating on the first eye. I would probably have the patient sign an amended informed consent and would emphasize to her the importance of taking a daily oral vitamin C supplement and wearing sunglasses that block blue light when she is outdoors during the first year after surgery.

If testing with the Ocular Response Analyzer is perfectly normal, then this patient may have been misdiagnosed. She may simply have a large white-to-white diameter.

> "I would decline to perform corneal refractive surgery in this case."
>
> —Louis E. Probst, MD

LOUIS E. PROBST, MD

I would decline to perform corneal refractive surgery in this case. Because megalocornea is extremely rare, the outcome of LASIK or PRK could be less predictable than in normal eyes. In the past 14 years, two patients with this congenital abnormality have presented to me with a request for refractive surgery, and I declined in both cases.

I would guess that PRK would be safer than LASIK for these eyes, but I would not offer either surgical option, because the previous diagnosis of megalocornea creates a special high-risk legal situation. Any problem in the future could be tied to this diagnosis so as to call the surgeon's judgment into question. Refractive procedures are elective, and patients do not tolerate complications well. I therefore do not take extra risks. Another way of looking at this situation is to ask what I would do if these were my eyes. I am sure that most ophthalmologists would not even consider a refractive option for themselves if they had megalocornea.

BRIAN S. BOXER WACHLER, MD

I am not aware of any patients with megalocornea undergoing keratorefractive surgery. Current diagnostic equipment allows further investigation in order to identify any known risk factors for such corneas undergoing keratorefractive surgery. The results of the Pentacam examinations are within a normal range in both eyes. In particular, the mean corneal thickness values relative to the thinnest location are normal, as are the anterior curvature and pachymetry readings. Analysis with the Pentacam appears to indicate that megalocornea is a normal variant, not a pathological state. Hence, there does not seem to be any risk factor for ectasia with LASIK or PRK in megalocornea. The caveat is, "never been done, so don't know." I would therefore propose either performing PRK on one eye at a time or implanting the Visian ICL (STAAR Surgical Company, Monrovia, CA) as conservative means of vision correction. ■

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