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Revised Calculation of the Prevalence of Keratoconus Based on Updated Scheimpflug Corneal Tomography Metrics in a Pediatric Population From a Chicago-Based School Age Vision Clinic

To the Editor:

We report an important update to the paper, Harthan JS, Gelles JD, Block SS, et al., Prevalence of Keratoconus Based on Scheimpflug Corneal Tomography Metrics in a Pediatric Population From a Chicago-Based School Age Vision Clinic. Published in *Eye and Contact Lens* in 2024.¹ The original paper studied school age children ages 3 to 18 years and is significant in that the diagnosis of keratoconus was made using modern tomographic parameters and stressed the importance of early diagnosis. Earlier papers on the prevalence of disease commonly used anterior surface topography, which while specific, lacks sensitivity as it identifies disease in the later stages when anterior surface changes occur. This often underestimates the prevalence and does not allow identification of early or subclinical disease where intervention (e.g., corneal crosslinking) can prevent vision loss.

The paper initially reported a prevalence of 1:334 for keratoconus and a prevalence of 1:223 when keratoconus suspects were included. Tomographic criteria for keratoconus included a final “D” from the Belin/Ambrósio display (Pentacam, OCULUS GmbH, Wetzlar, Germany) $\geq 3.0^2$ and a posterior elevation at the corneal thinnest point ≥ 18 μm for myopic individuals and ≥ 28 μm for hyperopic patients. Prevalence rates were reported by subject if one or both eyes met criteria. The same analysis was used in the revised tabulation.

The Belin/Ambrósio display was initially developed to screen patients for refractive surgery, and the final “D” represents variance from normal, but, in itself, is not specific for ectatic disease (e.g., corneal edema will result in an elevated final “D”). Typically, additional tomographic parameters, in addition to the final “D,” are included to improve specificity for keratoconus. These additional parameters may include an abnormal pachymetric progression and/or posterior elevation at the thinnest point (as in this study). Most of the confounding conditions, such as endothelial dysfunction, occur in adults and confounding conditions in children (e.g., congenital hereditary endothelial dystrophy) would be readily apparent. The goal of adding additional parameters is to improve specificity but should not be so restrictive as to decrease sensitivity. The values for the posterior elevation at the thinnest point chosen by the authors represent values 3.0 SD from the norm,

which when added to the final “D” ≥ 3.0 become overly restrictive.³

In collaboration with the original authors and after the original publication, it was agreed that the results should be retabulated using the same final “D” (≥ 3.0), but with values for the posterior elevation at the thinnest point of 13 and 23 μm (myopic/hyperopic) roughly corresponding to 2 SD.^{4,5} This change should not significantly alter the overall specificity but should decrease false negatives. Prior studies showed that a final “D” ≥ 2.69 had a 100% specificity for keratoconus in an adult population when other known pathologies were excluded. How this compares to a pediatric population is not known as most studies dealt with a refractive surgery population.⁴ The revised parameters defined keratoconus as a final “D” ≥ 3.0 with a posterior elevation at the thinnest point $\geq 13/23$ μm for myopic and hyperopic eyes, respectively. Suspect included eyes with a final “D” ≥ 2.0 but < 3.0 with the same posterior elevation parameters. Normal eyes had a final “D” < 2.0 . With these adjusted parameters, the revised prevalence of keratoconus in the study population of 2007 individuals is 1:251 (an increase from six to eight classified as keratoconus), the prevalence of keratoconus suspect is 1:100 (an increase from 3 to 20), and the prevalence when combining keratoconus and keratoconus suspects is 1:72 (an increase from 9 to 28).

Keratoconus is a visually disabling disorder that if left untreated or if treated late can lead to permanent, life-long debility. Using modern corneal imaging, the old historical teaching of keratoconus as a rare disease no longer applies. Early diagnosis is paramount if one wishes to intervene before significant visual loss. Since keratoconus typically first becomes evident in the pediatric age group knowledge of the disease, its diagnosis and treatment options need to be known not only by ophthalmologist and optometrists but by pediatricians, family practitioners, and school health screeners. The paper cited above, along with the even higher prevalence rates presented here, stresses the importance of keratoconus screening using modern diagnostic modalities. In addition to the use of Scheimpflug corneal tomography, advances in optical coherence tomography and biomechanical assessment may further enhance sensitivity and specificity in the detection of keratoconus and keratoconus suspects. We hope to increase awareness of the prevalence of the disease along with the goal of identifying disease early to limit future vision loss.

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