Editorial

Keratoconus; a True Corneal Disease

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Keratoconus (KCN) is a non-inflammatory disease that leads to progressive corneal ectasia and visual impairment. It is a global disease affecting millions of people throughout the world. Notably, it is a heterogeneous disease with highly variable presentation and progression. A genetic predisposition to keratoconus has long been recognized, however studies have not identified a common genetic defect or variant that could explain the majority of cases. Recently, mutations in miR-184 were implicated in a subset of patients with familial keratoconus and cataract.[1] miR-184 is the most abundant miRNA expressed in the cornea and lens. In this issue of JOVR, Farzadfard et al have reported that they did not identify any mutations in miR-184 among 47 patients, concluding that it is not a major cause of KCN among Iranian patients.[2] It is noteworthy that a variation in the pri-miR-184 encoding region was detected in a patient with familial KCN. The same variation was also detected in the affected sister. Overall, these studies highlight the need for identifying novel genes and variants associated with KCN. This information will ultimately allow us to identify patients at greater risk for development or progression of KCN while also providing novel targets for potential therapeutic strategies.

While the diagnosis of KCN can often be made by biomicroscopy, additional testing, such as pachymetry, keratometry and corneal topography are often used to further confirm and stage the disease. In subclinical KCN, the cornea appears normal on slit lamp examination and therefore additional tests are critical for the diagnosis. Diagnosing subclinical KCN is particularly important in refractive corneal surgery where patients at risk of ectasia need to be identified and excluded from surgery. In this issue, Feizi et al have assessed the predictive ability of the Galilei corneal imaging system, which is a combination of Placido and Scheimpflug imaging technology, for distinguishing KCN and subclinical KCN from normal corneas.[3] They evaluated keratometric values, pachymetry, elevation parameters and surface indices, and concluded that elevation parameters and surface indices can distinguish KCN from normal eyes in 100% of cases. For subclinical KCN a 3-factor model consisting of keratometric value, elevation data and surface indices provided the highest predictive ability. These results highlight the fact that anterior surface topography alone is not sensitive enough to detect all subclinical cases of KCN and suggest that the best strategy for detecting such cases requires measuring both the anterior and posterior cornea. As experience grows with the Galilei system, it should provide a highly sensitive tool for diagnosing subclinical KCN.

Depending on the stage of the disease, management of KCN may include spectacles, contact lenses, collagen crosslinking, intracorneal rings and lamellar/penetrating keratoplasty. In this issue of JOVR, the outcomes of two ring based treatments for KCN have also been assessed. Zare et al have reported that uncorrected and corrected distance visual acuity, cylinder and spherical equivalent, and keratometry were all improved after Intacs SK implantation.[4] Likewise, corneal biomechanics as measured by the corneal resistance factor and corneal hysteresis, improved postoperatively. Similarly, Janani et al have evaluated a full ring intra-corneal implant (MyoRing) for the management of KCN.[5] MyoRing is a flexible, 5 to 6 mm full-ring polymethylmetacrylate (PMMA) intracorneal implant which is inserted into a corneal pocket. After MyoRing implantation a significant reduction in sphere and cylinder, and also a significant improvement in uncorrected and corrected distance visual acuity were observed. Overall, these studies highlight the fact that in selected patients, intracorneal rings can be very effective for improving visual acuity and potentially delaying or avoiding the need for keratoplasty. However, at this time, it is still not clear which patients will benefit the most from these procedures and the long term results (particularly with MyoRing) are unknown.

In approximately 15-20% of KCN patients, the disease will progress to a point where the only remaining option is corneal transplantation, namely lamellar or penetrating keratoplasty. In recent years, there has been an increasing interest in anterior lamellar (instead of penetrating) corneal grafting procedures for KCN patients, thereby

preserving the patient's own endothelium and minimizing the risk of graft failure due to endothelial rejection. One of the most popular techniques involves air injection to dissect the patient's Descemet's membrane from the stroma (i.e. the "big bubble"). Technically, this is not always successful and in some patients the surgery has to be converted to a manual dissection technique instead. In this issue, Javadi et al have presented their results on visual outcomes after successful versus failed big-bubble deep anterior lamellar keratoplasty (DALK) in KCN.[6] They found that post-operatively, best corrected distance visual acuity was better in the bare Descemet's membrane group (successful big-bubble DALK) than the manual dissection (pre-Descemet's) group. These results are in contrast to some previous reports which have generally found similar visual outcomes with either technique. One difference, as the authors have indicated, may be residual stromal thickness, which was not evaluated in this study. Nonetheless, despite having slightly lower visual acuity, pre-Descemet's DALK is still preferred over penetrating procedures given the long term advantage of avoiding endothelial rejection.

In summary, as these studies demonstrate, KCN is a "true" corneal disease that continues to challenge us in terms of etiology, diagnosis and management. Future research will not only improve our understanding of its etio-pathogenesis, but will also open the door to more novel treatments that can prevent the development and progression of this global disease.

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