Commentary: A perspective on pediatric keratoconus: One size does not fit all

Everyone knows "*one-size-fits-all*" is often a lie. Solutions might fit most, but the fit is usually really, poor for the few outliers. Pediatric keratoconus is no exception; in truth, it shows several distinctive features in clinical appearance, disease progression, and response to treatment in children compared with adults.[1] Therefore, children merit a customized therapeutic approach considering the cornea's structural and behavioral differences between children and adults.[2]

The causal association of keratoconus has remained elusive despite being the subject of global investigation over the past few decades.[2] Previously, it was thought that a noninflammatory process is involved in the pathogenesis of the keratoconus, however, recent studies have shown that the imbalance between pro- and anti-inflammatory cytokines that lead to altered corneal structure and function triggering

metalloproteinases and keratocyte apoptosis were responsible for the causation and disease progression.[3,4] This review on pediatric keratoconus have filled the much-needed gap by providing a comprehensive review primarily focusing on unique aspects of diagnosis, and gaps in the understanding of disease presentation and most appropriate management strategies based on the best available current evidence.

Compared to adults, keratoconus in children progresses more rapidly and is usually more severe at the time of diagnosis; therefore, early detection and treatment are paramount to prevent serious vision impairment, affecting the child's social and educational development, thus negatively impacting their quality of life. Studies on pediatric keratoconus suggest that at the time of diagnosis, 27.8% are at an advanced stage and 88% progress. Progressive thinning can lead to acute hydrops, a potentially blinding condition that results in blisters in the cornea, with scarring and significant diminution in vision. Once the diagnosis has been made, compliance with treatment recommendations is often poor.

Nonsurgical options such as spectacles and contact lenses in children are not always tolerated and often insufficient to obtain a satisfactory visual acuity. Furthermore, none of these conservative options halt the progression of the disease. Surgical interventions like intracorneal ring segments and penetrating keratoplasties have been used as a standard therapeutic modality in the pediatric population. The literature overwhelmingly shows higher rates of failure and progression despite these measures as compared to adults. Therefore, the current therapies used in adults may not be appropriate for the pediatric population.[3]

The application of Collagen Cross Linking (CXL) to help retard keratoconus marks a significant change in paradigm.[3] CXL induces and enhances cross-linking between collagen fibrils. Riboflavin causes photosensitization, and UV-A creates cross-linking by generating oxidative products. CXL improves the corneal biomechanical strength, thereby arresting the progression of ectasia, which is basically due to biomechanical weakening. However, because of the pediatric cornea's dynamic nature, stabilization with CXL has also been documented to be less efficient than in adults. It would be interesting to explore a therapeutic algorithm specific to the pediatric population to understand and treat pediatric keratoconus.[5-7]

CXL is quickly gaining popularity among clinicians as they now have an effective and safe intervention to offer. Several issues still need to be addressed. The timing of CXL is a million-dollar question. In contrast, several authors suggest using it in cases where progression in more than 1.5 D at least 6 months interval. It may be argued that since keratoconus does not follow a linear progression, such cutoffs may be all but arbitrary. Some may argue that CXL should be offered at the first diagnosis of keratoconus as the natural course of disease suggests likely progression; more so in children who have a family history of keratoconus, frequent eye rubbing, and associated allergy.[3,4]

Therefore, it is of utmost significance to understand the long-term role timely CXL and management of allergy play in pediatric keratoconus, the delay of which can cause irreversible long-term visual impairment. It has also been proven to be cost-effective for patients and healthcare providers compared to not cross-linking and eventually requiring keratoplasty.[2,6] Children who regress following CXL often have a more ocular allergy and eye rubbing, and it is essential to manage allergy long-term. This subgroup progresses rapidly when left untreated. For this reason, many groups strongly recommend CXL in children at first diagnosis of keratoconus without waiting for progression.[4] Worldwide documented research has shown that CXL has a role in the prevention of keratoconus progression. Therefore, as an option, CXL must be offered to every child presenting with progressive keratoconus so that an attempt at early treatment can be made. Furthermore, the association of pediatric keratoconus with inflammatory markers and hormonal etiologies should be explored further to see if they can be targeted for future therapy.

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