Response to comment on 'Unilateral corneal edema in young: A diagnostic dilemma'

Dear Sir,

We sincerely thank Singh *et al*.[1] for their interest in our article,[2] and hope to clarify their insightful queries in this reply.

First, the authors state that delayed onset congenital hereditary endothelial dystrophy (CHED) has been described in the first decade only. However, reports of genetically proven CHED with deterioration in later ages have been known.[3] Though CHED-1 has been removed from the recent International Classification of Corneal Dystrophies - Edition 2 (IC3D-2), Harboyan disease has been shown to be more closely related to CHED-2.[4]

A thorough evaluation by slit-lamp biomicroscopy, specular microscopy, and confocal microscopy was done to rule out guttae, vesicles, bands, rail-tracks, and corneal opacities; however, we are sorry that a retro-illumination photograph was not documented.

Posterior polymorphous dystrophy usually starts in early childhood with typical appearances, and corneal edema to the extent of requiring keratoplasty occurs in only 20–25%.[5] Our patient had never documented a high intraocular pressure, and his anterior segment optical coherence tomography did not reveal any peripheral anterior synechiae.

While genetic overlap between the various endothelial dystrophies is known to exist,[6] in the absence of molecular confirmation, diagnosis is based on contributory clinical evidences. We could not find significant guttae/vesicles/bands/opacities in the apparently normal eye. Also, the central corneal thickness (CCT) of apparently normal eye being 690µ and affected eye being 850µ with diffuse ground-glass appearance favoured CHED. This is in contrast to Fuch's endothelial dystrophy where eyes of early disease without edema showed mean CCT 576µ and advanced cases with edema showed mean CCT 649µ,[7] despite the Caucasian eyes in general having thicker corneas than Indians. However, these are studies on late-onset FECD, and reports confined to CCT of early-onset FECD are unavailable at present to the best of our knowledge.

Nonguttate corneal endothelial dystrophy with grossly reduced endothelial cells[8] appears to have a similar picture, but this dystrophy has been barely investigated and not categorised under IC3D-2.[5] Also, the sensorineural deafness at this young age remains unanswered.

Similar to Singh *et al*., we were also keen on the histopathological detailing of the patient's Descemet membrane, but sadly, the patient did not report back for the surgery.

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Conflicts of interest

There are no conflicts of interest.

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