

# History

- A five-month-old baby was seen in clinic with bilateral congenital corneal opacities. The right cornea is shown in Figure 1.
- Examination revealed corneal oedema and opacities of Descemet's membrane and endothelium with a few vesicular lesions.
- Corneal transplantation was performed and specimen sent for ophthalmic histopathological and ultrastructural assessment.



Figure 1

# Questions

1. What are the clinical differential diagnoses for congenital diffuse corneal haze?
2. Figure 2 represents PAS and H&E stained sections of the posterior aspect of the cornea. What are they showing?
3. In Figure 3 the top image is an immunohistochemistry section for AE1/AE3. What is it demonstrating and which alternative markers could also be used?
4. Figure 4 represents EM sections of the posterior corneal surface. What are the most relevant features?
5. Considering the morphological features, what is the most likely diagnosis?

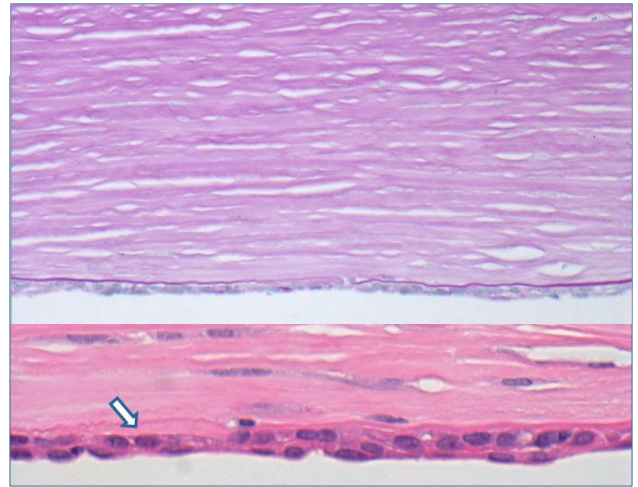


Figure 2

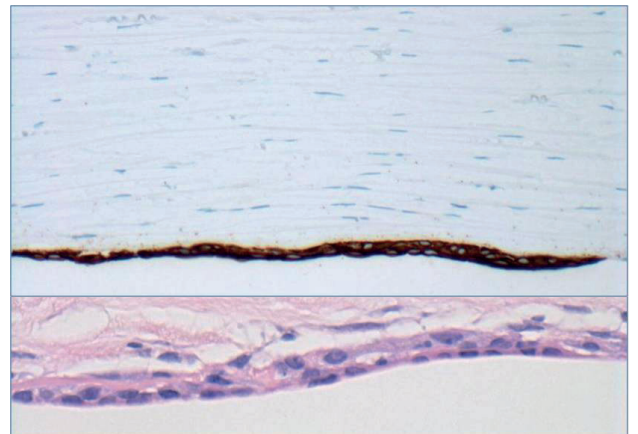


Figure 3

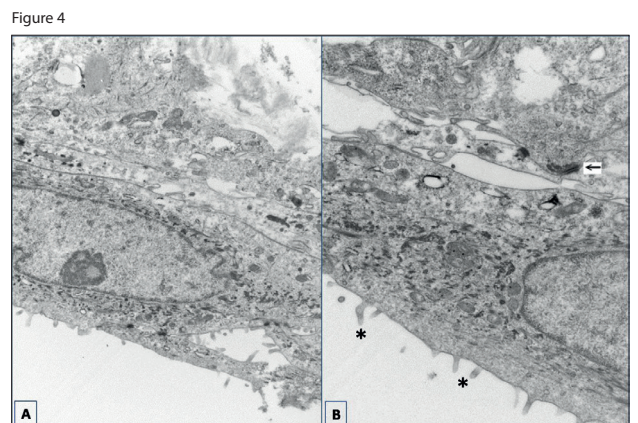


Figure 4

SECTION EDITOR



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1. Complete stromal haze is seen in congenital hereditary endothelial dystrophy (CHED), posterior polymorphous corneal dystrophy (PPCD), X-linked endothelial corneal dystrophy (XECD) and congenital glaucoma.
2. The PAS stained top image highlights a thin and discontinuous Descemet's membrane. The H&E stained bottom image shows multilayering of the corneal endothelium. Note focal collagen deposition underneath Descemet's membrane indicated by the arrow.
3. The brown staining shows the expression of this broad epithelial marker by the abnormal endothelium. Corneal stroma is negative as usual. Normal corneal endothelial cells do not express epithelial markers. Other antibodies also expected to be positive are anti-CK7, CK8, CK18, and CK19. The H&E stained section at the bottom shows another area of the multilayered endothelium.
4. Figure 4A demonstrates multilayered endothelium. In Figure 4B the arrow is pointing to a desmosome and the stars indicate microvilli. These are epithelial characteristics in the abnormal corneal endothelial cells.
5. Posterior polymorphous corneal dystrophy.

# Answers