Keratoplasty in congenital primary aphakia

Sir,

The management of pediatric corneal opacities needs several preoperative considerations before keratoplasty to ascertain associated comorbidities that directly influence the outcome. B-scan ultrasonography and/or ultrasound biomicroscopy should be routinely performed in those cases where corneal involvement precludes the assessment of the anterior and posterior segment. It is important to evaluate the status of lens as many corneal opacities may have an absent lens, a condition known as congenital primary aphakia.[1,2]

Congenital aphakia may be primary or secondary. Congenital primary aphakia can occur as an isolated abnormality or as part of a complex anterior segment abnormality, i.e., microphthalmia, absence of the iris, anterior segment aplasia, and/or sclerocornea, or it may occur in association with multiple somatic abnormalities. The secondary form is characterized by absorption of the lens after it is formed. Congenital primary aphakia is thought to result from an abnormality during the 4th or 5th week of fetal development, which prevents the formation of any lens structure in the eye.[3] This original failure leads, in turn, to complete aplasia of the anterior segment of the eye, which is the diagnostic histologic criterion for primary aphakia.[3,4] Homozygous mutations in *FOXE3* have been implicated in primary congenital aphakia.[4] The clinical suspicion is made in those eyes with congenital opacities with a bluish hue [Fig. 1a]. A B-scan ultrasound examination helps in confirming the absence of lens echo.

We reviewed the outcomes of keratoplasty performed in this condition at our institute. Out of 71 eyes with

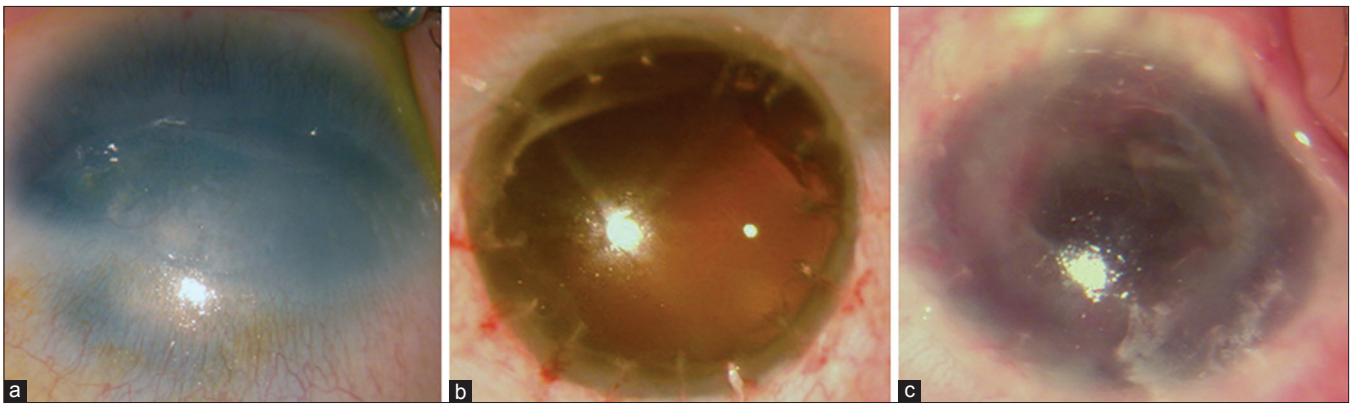

Figure 1: (a) Digital Photograph showing the typical bluish hue appearance of the cornea in a child with congenital primary aphakia; (b) digital photograph of the eye at 1 month after penetrating keratoplasty showing a clear graft; (c) photograph of the same eye at 6 weeks after penetrating keratoplasty showing an organized membrane at the ciliary body region and eye going into phthisis (pre-phthisis)

congenital primary aphakia with corneal involvement, 5 underwent penetrating keratoplasty. Even though the graft seemed to do well till 2–4 weeks postoperatively, all eyes progressed to phthisis bulbi at 6–8 weeks of the follow-up period [Fig. 1b and c]. In all other eyes where penetrating keratoplasty was deferred, the globe integrity was maintained, and the limited visual functions were preserved (Unpublished data).

While reviewing the literature on the outcomes of keratoplasty in this clinical condition, it was found that keratoplasty in these eyes is unsuccessful[5] and leads to a poor anatomical outcome owing to hypoplastic ciliary process, abnormal vitreous, and consequent hypotony ensues phthisis bulbi invariably.

Hence, keratoplasty in patients with congenital primary aphakia should be deferred as it has a disastrous outcome. It should be a mandate to confirm the lens status on B-scan ultrasonography in pediatric patients who present with a total corneal opacity and are considered for penetrating keratoplasty. Those patients where the lens echo is absent should be managed conservatively and referred for visual rehabilitation. Early recognition, counseling, and prompt visual training of the child from an early age are essential in the preserving the limited functional vision in this condition.

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Conflicts of interest

There are no conflicts of interest.

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