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Primary congenital glaucoma.

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Abstract

Primary congenital glaucoma (PCG) is the most common nonsyndromic glaucoma in infancy, which can lead to blindness, or a lifetime of vision when diagnosed and treated properly. PCG is more common in populations with a higher prevalence of consanguinity and is associated with CYP1B1 gene mutations which show variable expressivity and phenotypes. The immature angle appearance of PCG likely results from arrested development of tissues of neural crest origin in the third trimester, with the severity of abnormality varying according to the stage at which arrested development occurred. Classic symptoms at presentation include tearing, photophobia, blepharospasm, eye rubbing, and irritability. Examination may reveal elevated intraocular pressure, corneal edema, increased corneal diameter, Haab striae, or enlarged axial length. Angle surgery remains the first line treatment for PCG with a recent advance being circumferential trabeculotomy with the potential to incise the whole angle during one operation as oppose to an incremental approach and the associated multiple anesthetics. Once angle surgery fails, either trabeculectomy or glaucoma drainage device surgery may be appropriate.

KEYWORDS:

Genetics; Goniotomy; Pediatric congenital glaucoma; Trabeculectomy; Trabeculotomy