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Corneal Changes in Childhood Glaucoma

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Objective: To study the clinical features and topography of the cornea in eyes with childhood glaucoma.
Design: Cross-sectional, observational study.
Participants: Fifty-eight eyes with childhood glaucoma and 28 eyes of age-matched controls.
Methods: Clinical and topographic corneal changes were evaluated.
Main Outcome Measures: Corneal topographic changes were evaluated on Orbscan (Orbscan Topography System II; Bausch & Lomb, Salt Lake City, UT) in eyes with childhood glaucoma and those changes were compared with the control eyes.
Results: Fifty-eight eyes with childhood glaucoma and 28 eyes of age-matched controls were evaluated. Thirty-six eyes (62.1%) were classified as having primary childhood glaucoma and 22 eyes (37.94%) as having childhood glaucoma with associated ocular anomalies. The corneas in 18 of 58 eyes (31.0%) with childhood glaucoma were clear, whereas 24.1% of eyes (14/58 eyes) had some corneal opacification. Haab's striae were noted in 44.8% of eyes (26/58 eyes) and were most frequently present between 3 and 5 mm from the optical axis. The mean posterior elevation recorded in eyes with childhood glaucoma controlled with medication or surgery was significantly higher than that in control eyes: 0.043±0.027, 0.042±0.017, and 0.018±0.058 μm, respectively ($P < 0.0001$). The presence of Haab's striae was correlated significantly with a higher posterior elevation ($P = 0.0396$) and poor vision. The mean anterior elevation in eyes with childhood glaucoma (0.022±0.015 μm) and in control eyes (0.015±0.078 μm) was comparable ($P = 0.08$). Corneal astigmatism in eyes with childhood glaucoma was significantly higher and irregular compared with that in control eyes: 2.09±1.40 versus 0.93±0.60 diopter cylinder ($P = 0.0001$); the irregularity index was 2.8 (range, 1−18.1) and 2.3 (range, 0.6−2.3) at 3 mm ($P = 0.0005$) and 3.2 (range, 1.4−21.3) and 1.8 (range, 0.5−2.9) at 5 mm, respectively ($P = 0.0003$). Best-corrected visual acuity correlated significantly with cup-to-disc ratio, axial length, refractive error, astigmatism, and posterior corneal elevation. Multivariate analysis showed a significant correlation only with cup-to-disc ratio and axial length.
Conclusions: Childhood glaucoma causes a significant increase in posterior corneal elevation and irregular astigmatism, which contribute to visual disability in such eyes. *Ophthalmology 2015;122:87-92 © 2015 by the American Academy of Ophthalmology.*

Childhood glaucoma is associated with various corneal changes that include an increased corneal diameter, corneal edema, Haab's striae, and altered corneal thickness.[1−4] Despite controlling intraocular pressure (IOP) optimally, visual outcomes sometimes are suboptimal, and this is the result of glaucomatous optic neuropathy, but also could be the result of changes in the cornea such as nebulomacular corneal opacity, leukomatous corneal opacity, and Haab's striae. Corneal opacity or clouding and anisometropia may lead to stimulus deprivation amblyopia, which has been reported to be one of the major causes of vision loss in these children, up to almost 50%.[5]

Corneal abnormalities in childhood glaucoma have not been studied in detail or reported in the literature, to the best of our knowledge. This study was undertaken to evaluate corneal clinical features and topography in childhood glaucoma and to correlate these with best-corrected visual acuity (BCVA).

Methods

Consecutive patients with childhood glaucoma 5 years of age or older were screened at the Glaucoma Service, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India, for a period of 1 year. Only cooperative children with an IOP less than 15 mmHg after either medical therapy or a trabeculectomy with trabeculotomy performed at least 1 year earlier were included. Exclusion criteria were the presence of any other corneal pathologic features, childhood cataract, uveitis, any retinal disease or advanced optic disc cupping associated with a relative afferent pupillary defect, or the absence of vision on examination. It was deemed that children 5 years of age or younger would not be able to cooperate with the topographic testing techniques used in this study. Age-matched controls were consecutive patients reviewed for strabismus, with no significant refractive error, no amblyopia, nor any ocular pathologic features. Institutional ethics committee approval was obtained and the tenets of the Declaration of Helsinki were observed.

A detailed ocular examination, including slit-lamp biomicroscopy, applanation tonometry on slit lamp, pachymetry (performed on the Orbscan Topography System II; Bausch & Lomb, Salt Lake City, UT), and cycloplegic refraction, was performed. Orbscan topography was performed on Orbscan Topography System II. The parameters evaluated by the Orbscan topography system were anterior elevation, posterior elevation, simulated keratometry (SimK), central corneal thickness, thinnest area, white-to-white diameter, and astigmatism. Axial length of these eyes was recorded using the mean of 3 readings

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Table 1. Corneal Parameters in Childhood Glaucoma Eyes with and without Haab's Striae

Parameters	Haab's Striae Present (n = 24)	Haab's Striae Absent (n = 22)	P Value
BCVA*	0.477 (0−1.176)	0.477 (0−4)	0.111
Anterior corneal elevation*	0.015 (0.01−0.07)	0.02 (0.01−0.06)	0.2099
Posterior corneal elevation*	0.045 (0.023−0.069)	0.037 (0.01−0.08)	0.0396
Simulated keratometry*	1.5 (0.6−5.5)	1.7 (0.1−4.1)	0.7329
Maximum keratometry	42.66±3.18	42.30±2.34	0.6685
Minimum keratometry	40.51±3.12	40.12±1.42	0.3424
3-mm mean keratometry	41.38±3.18	41.23±2.56	0.8574
Astigmatism at 3 mm*	1.35 (0.1−7.9)	1.9 (0.3−8.6)	0.3848
5-mm mean keratometry	40.76±2.82	40.66±2.59	0.9019
Astigmatism at 5 mm*	2.75 (0.4−9.4)	1.9 (0.4−9.9)	0.1980
White-to-white diameter	12.54±0.80	12.47±0.64	0.7752
Thinnest central corneal thickness	456.45±139.17	499.92±129.18	0.2780
Central corneal thickness	533.72±83.0	548.47±145.54	0.7468
Irregularity index at 3 mm*	1.7 (1−9.1)	5.1 (1.6−21.3)	0.6425
Irregularity index at 5 mm*	1.4 (1.1−18.1)	3.1 (1.4−16.5)	0.4204

BCVA = best-corrected visual acuity.
Data are mean ± standard deviation or *median (range).

on ultrasound A scans (Alcon UltraScan Imaging System, 2000; Alcon Laboratories, Fort Worth, TX).

Differences between childhood glaucoma and control eyes, as well as different types of childhood glaucoma, were assessed using the unpaired *t* test or the Wilcoxon rank-sum test, 1-way analysis of variance, Kruskal-Wallis test, chi-square test, or Fisher exact test, as applicable. Correlation of parameters with BCVA was performed using univariate and stepwise multiple regression. STATA software version 11.2 (StataCorp LP, College Station, TX) was used for data analysis. A *P* value less than 0.05 was considered statistically significant.

Results

Sixty-three patients older than 5 years of age with childhood glaucoma were examined, and 29 patients who met all criteria were studied. Primary childhood glaucoma with only a trabecular dysgenesis but normal iris was seen in 18 of 29 patients (62.1%), whereas 11 of 29 patients (37.9%) had childhood glaucoma with other anterior segment anomalies, including Axenfeld-Reiger syndrome (n = 8), Sturge-Weber syndrome (n = 2), and neurofibromatosis (n = 1). The mean age of patients was 159.2±3.7 months (range, 72−336 months) and that of controls was a 175.2±60.8 months (range, 72−330 months; $P = 0.48$). Primary childhood glaucoma patients sought treatment early (mean, 7.03±3.70 months) compared with patients with childhood glaucoma with associated anomalies (mean, 86.72±35.46 months; $P = 0.0001$). Fifty of 58 eyes (86.2%) had undergone combined trabeculectomy and trabeculotomy surgery for the control of IOP, whereas 8 eyes (13.8%) demonstrated control of glaucoma with antiglaucoma medications alone. The IOP in medically controlled eyes was 12.25±1.15 mmHg and that in surgically managed eyes was 12.51±2.46 mmHg. The mean BCVA in eyes with childhood glaucoma was 0.68±0.72 logarithm of the minimum angle of resolution (logMAR) units, whereas all control eyes had an uncorrected visual acuity of 0 logMAR units.

A completely clear cornea was seen in 18 of 58 eyes (31.0%) with childhood glaucoma, whereas 14 of 58 eyes (24.1%) had some corneal opacification without Haab's striae: nebulomacular opacity (iris details were visible through opacity) in 8 eyes (13.8%) and a leukomatous corneal opacity (unable to visualize iris details through opacity) in 6 eyes (10.3%). All corneal opacities were avascular.

Haab's striae were present in 26 of 58 eyes (44.8%) and were multiple and circumferentially located in 22 eyes. They were exclusively horizontal in 4 eyes. Only 3 of 26 eyes (11.5%) had Haab's striae located within the central optical zone of 3 mm, and these patients had a BCVA less than 0.30 logMAR. Twelve of 26 eyes (46.1%) had Haab's striae located in the paracentral zone, between 3 and 5 mm from the optical axis, and 11 of 26 eyes had Haab's striae in the periphery, with a mean BCVA of 0.632±0.5381 logMAR (median, 0.4515 logMAR; range, 0−1.778 logMAR) and 0.624±0.460 logMAR (median, 0.477 logMAR; range, 0−1.176 logMAR), respectively ($P = 0.9699$). Orbscan was possible in 46 of 58 childhood glaucoma eyes. There was a significantly increased posterior elevation in eyes with Haab's striae compared with those without Haab's striae: 0.047±0.015 μm (0.015 μm; range, 0.01−0.07 μm) versus 0.037±0.021 μm (median, 0.02 μm; range, 0.01−0.06 μm), respectively ($P = 0.0396$; Table 1). Posterior elevation changes on Orbscan were seen even in the absence of Haab's striae (Fig 1). Although Haab's striae were more common in eyes with primary childhood glaucoma, it was not statistically significant ($P = 0.135$); similarly, there was no significant difference in the frequency of corneal opacification in both types of childhood glaucoma.

Comparing medically controlled eyes with childhood glaucoma having no prior surgical intervention with control eyes, there was a significant difference in SimK readings: maximum SimK was 43.99±0.80 versus 43.18±0.83 ($P = 0.018$), and minimum SimK was 42.53±2.57 versus 41.42±2.43 ($P = 0.004$). The mean posterior elevation in medically controlled eyes with childhood glaucoma was 0.043±0.027 μm (median, 0.039 μm; range, 0.01−0.08 μm) and was significantly higher than that in control eyes: 0.018±0.058 μm (median, 0.018 μm; range, 0.011−0.033 μm; $P = 0.016$). The mean keratometry at 3 mm, 42.07±2.50, was significantly lower in eyes with childhood glaucoma compared with that of control eyes, 43.60±0.85 ($P = 0.013$; Table 2). All eyes with childhood glaucoma (medically or surgically controlled) had higher irregular astigmatism compared with control eyes (Table 2). Operated eyes had significant higher astigmatism of