Original Article

Keratoglobus: An experience at a tertiary eye care center in India

Varsha M Rathi, Somasheila I Murthy[1], Bhupesh Bagga[1], Mukesh Taneja[1], Sunita Chaurasia[1], Virender S Sangwan[1]

Context: This study was carried out as a part of an internal audit and is the largest series of patients having keratoglobus, published in the literature. Poor visual acuity of the patients indicates the blinding nature of the disease. **Aims:** We report our experience with patients having keratoglobus at a tertiary eye care center in India. **Settings and Design**: Retrospective study. **Materials and Methods:** We analyzed adults and pediatric patients (<16 years) with keratoglobus, seen during 2008–2012. The age, gender, consanguinity, presenting ocular signs, ocular and systemic associations, visual acuity, corneal topography, and surgeries were documented. **Results:** Forty-eight patients (mean age 22 ± 15 years, 31 males) having keratoglobus were analyzed. 21 patients (42 eyes) were <16 years. Twelve eyes (16 events) had positive history of trauma. The presenting clinical signs were corneal scars/scars of tear repair (15 eyes), hydrops, healed and acute (14 eyes) and corneal or globe rupture (9 eyes). Best-corrected visual acuity was >20/40 in 6/42 (14.3%) pediatric eyes and 15/53 (28.30%) adults. Visual acuity ranging from counting of fingers to no light perception was noted in 20/53 (37.74%) adults and 21/42 (50%) pediatric patients; 13/20 (65%) with blue sclera and 8/22 eyes (36.37%) without blue sclera. Vernal keratoconjunctivitis was present in one pediatric patient. Choroidal osteoma, retinitis pigmentosa, and retinal detachment were present in adults. Surgeries performed were corneal tear repair (5 eyes), tissue adhesive application (2 eyes), descematopexy (4 eyes) and penetrating keratoplasty (PK - 8 eyes: Three had post-PK glaucoma, graft failure-one eye, 4 patients wore scleral lens - prosthetic replacement of the ocular surface ecosystem). **Conclusions:** About 50% of pediatric eyes (65% having blue sclera) had no functional vision. Trivial trauma was responsible for corneal rupture indicating need for protective glasses. About 50% patients had post-PK glaucoma though grafts were clear.

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Keratoglobus is a noninflammatory ectatic disorder of the cornea characterized by thinning from limbus to limbus resulting in globular protrusion of the cornea.[1,2] Acquired keratoglobus has been described in association with vernal keratoconjunctivitis (VKC), chronic marginal blepharitis, dysthyroid ophthalmopathy, Ehlers–Danlos syndrome and Marfan syndrome.[1,2] Congenital keratoglobus has been described with Leber's amaurosis and blue sclera syndrome.[1,3]

Management usually involves conservative therapy in the form of spectacles or contact lenses. Corneal rupture following trivial trauma has been reported in patients with keratoglobus. Spectacles provide functional vision and protection from trauma.[2] Contact lenses are generally considered after balancing the risk of trauma.[2,4] Surgeries such as lamellar graft, epikeratoplasty, partial thickness corneoscleroplasty followed by smaller diameter penetrating keratoplasty (PK) are performed when necessary to improve the chances of survival of graft as well as to maintain the integrity of the globe.[3,5-7]

In this study, we report presenting signs, visual acuity and management of keratoglobus in a cohort of patients seen at our center.

Cornea Services, [1]L V Prasad Eye Institute, Kallam Anji Reddy Camus, Hyderabad, Telangana, India

Correspondence to: Dr. Varsha M Rathi, L. V. Prasad Eye Institute, Kallam Anji Reddy Campus, L V Prasad Marg, Banjara Hills, Hyderabad - 500 034, Telangana, India. E-mail: varsharathi@lvpei.org

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Materials and Methods

We audited the medical charts of those patients diagnosed with keratoglobus and seen at the Cornea Services at a Tertiary Eye Care Centre in India between January 2008 and December 2012. The Local Ethics Committee approved the study. The diagnosis of keratoglobus was based on the clinical findings of corneal ectasia from limbus to limbus with thinning. Corneal topography was done with Orbscan IIz (Bausch + Lomb) where possible.

Age, gender, laterality, family associations, associated ocular and systemic diseases were documented. The presenting signs and a positive history of trauma were noted. Acute hydrops was diagnosed by the sudden onset of corneal edema. Healed hydrops was diagnosed by the presence of the corneal scar with a Descemet tear or split [Fig. 1]. Best-corrected visual acuity (BCVA) and various surgeries performed were documented. The refractive correction in the form of spectacles and contact lenses was noted. The literature on keratoglobus is scarce. Cameron *et al*. had described the results in two age groups of pediatric patients and adults.[2,3] We divided the patients into two groups; ≤16 years of age at the time of presentation and >16 years of age. The results of PK are different in these groups. The pediatric group was sub-divided into two groups by noting the presence or absence of blue sclera on examination.

Statistical analysis was done using software R version 2.14.1 (open source, available from http://www.r-project.org/). Shapiro–Wilk test was used to assess the normality of data. Two-sample *t*-test was used for parametric data, and Wilcoxan

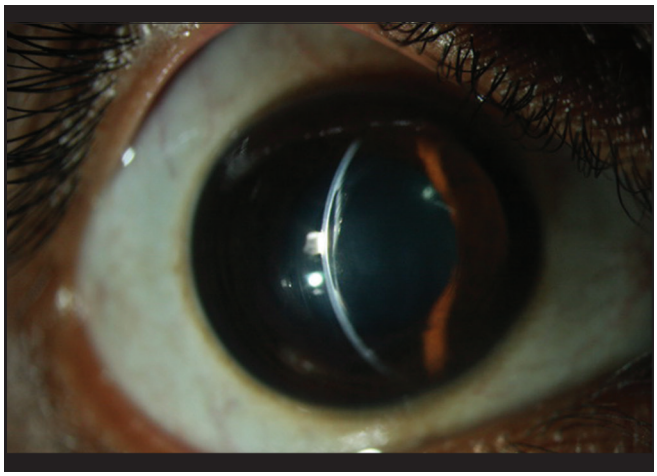

Figure 1: Keratoglobus with healed hydrops the tear in the Descemet's membrane is visible as vertical line

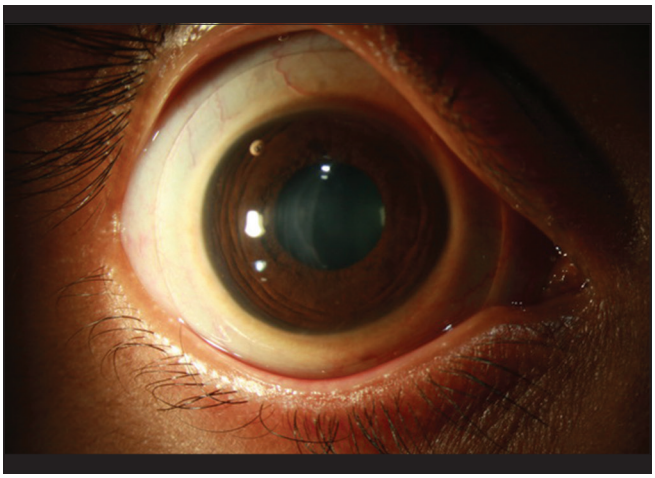

Figure 2: The same patient in Figure 1 wearing prosthetic replacement of the ocular surface ecosystem (PROSE) that is, scleral contact lens. Visual acuity with PROSE wear was 20/30 with a 3 years follow-up

signed rank sum test was used for nonparametric data. Fisher's exact test was used to compare the proportion of data.

Results

A total of 48 patients (95 eyes) with the diagnosis of keratoglobus were identified. The mean age of these patients at the time of presentation was 22 ± 15 years (range: 4–62 years). A total of 21 patients (42 eyes) were <16 years of age. The average follow-up was 24.7 ± 20.5 months with a range of 0–60 months. The mean age of the patients having blue sclera was 7.5 ± 3.81 years and without blue sclera was 10.1 ± 3.07 years. Tables 1 and 2 show the demographics and the presenting signs for both adult and pediatric patients, with and without the presence of blue sclera.

Table 3 shows the clinical findings at the time of presentation in these patients for both adult and pediatric age groups. The adult patients had bilateral choroidal osteoma (n = 1 patient), retinitis pigmentosa (2 patients - 4 eyes), retinal detachment (3 patients - 3 eyes) and pigment epithelial detachment in one eye of a patient. Two adult patients had joint hyperextensibility and deafness, and 1 patient was mentally challenged.

On corneal topography with OrbscanIIz, the average steep and flat sim k values in pediatric patients were 57.95D ± 6.85D and 52.15D ± 6.33D respectively. In adult eyes, the steep and flat sim k values were 54.7D ± 8.29D and 49.37D ± 7.68D respectively. The P values were 0.3 and 0.26 for flat and steep sim k respectively. The average central/thinnest pachymetry values on Orbscan IIz examination were 343.7 ± 72.98/319.3 ± 66.52 microns in pediatric patients and 393.82 ± 117/360 ± 132.65 microns in adult patients. Though the average values were lesser in pediatric age, the P value of 0.18 was not considered as statistically significant.

Glasses were prescribed for 21 pediatric eyes (11 patients) for visual improvement. Contact lenses were dispensed for use for four eyes. Two patients wore scleral contact lens Prosthetic replacement of the ocular surface ecosystem (PROSE, Boston Foundation for Sight, Needham Heights, MA, USA); one patient had healed hydrops prior to lens wear and visual acuity with PROSE wear was 20/30, over 3 years of follow-up [Fig. 2]. The

Table 1: Demographics in pediatric and adult patients

	Pediatric age group <16 years	Adult age >16 years
Number of patients	21 patients (42 eyes)	27 patients (53 eyes)
Mean age (years)	8.86±3.6	32.79±12.9
Range (months)	4-15	17-62
Male: Female	13:8	18:9*
Blue sclera	20 eyes (10 patients)	5 eyes
History of consanguineous marriage	3 patients (2 had blue sclera)	0
Average sim k	55.05D±7.1D	52.03D±8.32D#

P value was calculated with Fisher's exact test. **P* value was 0.76: Not statistically significant, #*P* value was 0.15: Not statistically significant

Table 2: Demographics of pediatric patients subdivided by the presence or absence of bluish appearance of sclera with ocular and systemic associations

	Presence of blue sclera	Absence of blue sclera
Number	n=20 eyes	n=22 eyes
Median age at the time of presentation	6.5 (4, 9)	10 (7.5, 11.75)
Gender male: female	5:5	8:3
History of consanguineous marriage	2	1
Siblings affected	2	2
Associations	Connective tissue disorder 1 patient	Hyperextensible joints-1 patient Mentally challenged-1 Ehlers-Danlos syndrome-1 VKC-1

VKC: Vernal keratoconjunctivitis. p=0.01 and was statistically significant for age. P value was not statistically significant for gender (p=0.38) or history of consanguinity (p=0.59)

second patient who wore PROSE developed hydrops following 3 months of lens wear, and BCVA reduced from 20/20 to 20/399. The hydrops resolved at 1-month with conservative management. Two patients underwent PK; one patient underwent PK combined with cataract surgery at the age of 4 years. He had a surgical scar of tear repair; the other eye of this patient was phthisical. The patient was lost for follow-up after postoperative day 1. The second patient underwent PK for corneal scar at the age of 12 years. The follow-up after PK was for only 4 months.

Glasses were prescribed to 20 adult eyes (11 patients) for visual improvement. Six eyes of four adult patients wore contact lenses; rigid gas permeable lenses for four eyes and PROSE was prescribed for two eyes: One patient after hydrops healed and the other patient for post-PK high astigmatism. Fifteen adult patients (27 eyes) were not prescribed any refractive correction for visual improvement. Table 4 shows BCVA in pediatric and adult patients. Table 5 shows the various surgeries performed in adult and pediatric patients [Figs. 3-6]. In our analysis, only one adult patient presented with corneal tear and underwent repair. The visual acuity was 20/80 at 18 months of follow-up.

Two adult patients underwent cataract surgery, and BCVA was 6/30 and 6/18 with spectacles. PK was done for six adult eyes, of which three eyes had post-PK glaucoma. Of these three eyes with glaucoma, one graft failed, and the patient underwent Descemet's stripping endothelial keratoplasty with trabeculectomy. Two patients without glaucoma were doing well after PK, one post-PK patient had 20/30 vision with PROSE and the other had 20/50 with his spectacles. All the grafts were clear.

Table 3: The clinical findings at the time of presentation

	Pediatric age group			Adult age	*P*
	Presence of blue sclera (eyes)	Absence of blue sclera (eyes)	Total (eyes)	Total (eyes)	
Disfigured eye					
Phthisis bulbi	2	1	3	1	0.16
Auto evisceration	0	1	1	0	
Scars					
Scar	5	4	9	5	0.05
Scar of healed hydrops	4	0	4	3	
Scar of tear repair	1	0	1	0	
Acute hydrops	2	1	3	4	1.00
Perforation or tear					
Corneal perforation	1	1	2	1	0.13
Sealed corneal perforation	1	0	1	0	
Corneal tear	1	2	3	1	
PMD	0	1	1	6 (1 superior, 4 inferior, 1 superior and inferior in same eye)	0.12
Other					
Absorbed lens	1	0	1		
Cataract			1	3	
Nystagmus	2 eyes of 1 patient	2 eyes of 1 patient	4	0	
H/o trauma	6	6	12 eyes (16 events)	1	<0.001

PMD: Pellucid marginal degeneration. Trauma with own finger, fist, elbow was reported by 12 patients with repeated events resulting in 16 events. *p* value was significant for scars and positive history of trauma

Table 4: BCVA in pediatric and adult patients

Visual acuity	Pediatric group			Adult group		
	With blue sclera (20 eyes)	Without bluish sclera (22 eyes)	Total (eyes)	With blue sclera *n*=5 eyes	Without bluish sclera *n*=48 eyes	Total (eyes)
No PL	3	2	5	0	8	8
PL, HM, CF	10	6	16	3	8	11
20/200-20/80	1 eye	7	8	0	12	12
20/70-20/50	3	2	5	0	7	7
20/40-20/20	3	3	6	2	13	15
Unable to assess		2	2			

BCVA: Best-corrected visual acuity, PL: Perception of light, HM: Hand movements, CF: Counting fingers close to face. *p* value was not statistically significant between pediatric and adult patients. But, *p* value was 0.03 when compared for keratoglobus with and without having blue sclera

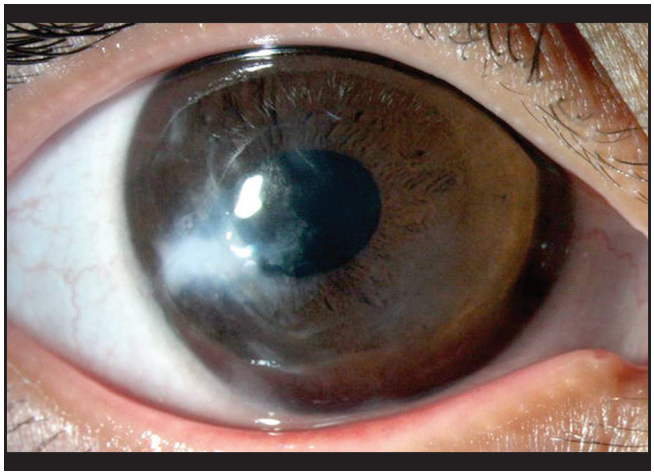

Figure 3: Scar in the right eye following tear repair in a 4-year-old boy; he sustained trauma to his right eye with elbow and underwent tear repair and tissue adhesive application. The visual acuity was 20/80 in the presence of scar at last follow-up

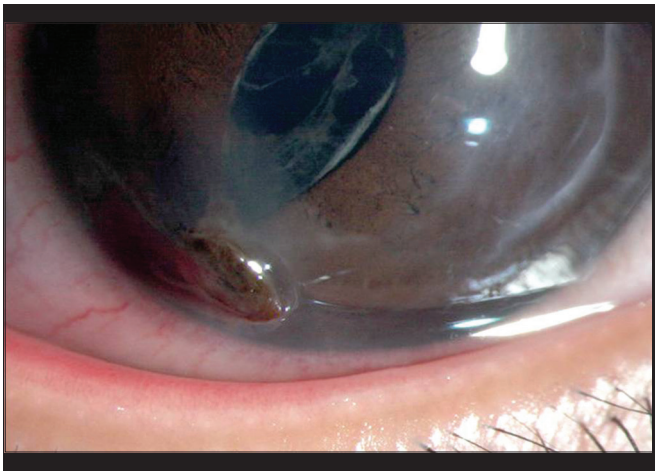

Figure 4: The same patient of Figure 3 sustained trauma to his left eye with first 4 years after the tear repair of his right eye. Iris prolapse is visible. He underwent tear repair and had best-corrected visual acuity of 20/200 at 2 years follow-up

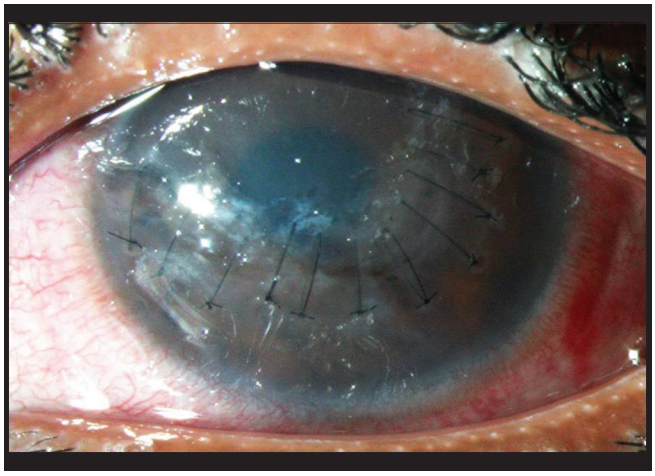

Figure 5: Sutured corneal tear with a bandage contact lens in the immediate postoperative period

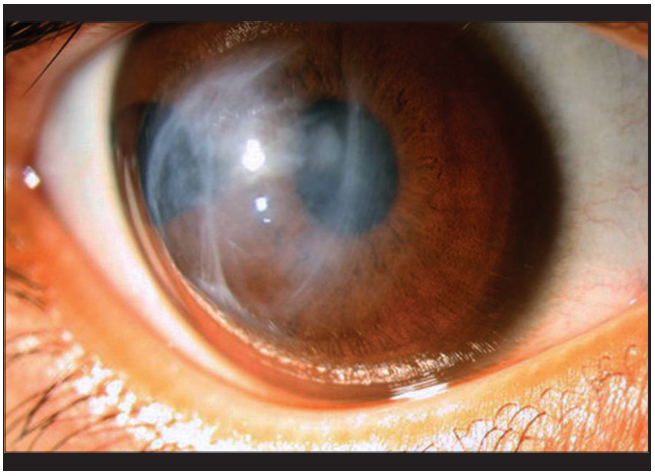

Figure 6: Scar after descemtopexy was performed for acute hydrops in a 4-year-old boy. Visual acuity was counting fingers close to face

Table 5: Various surgeries performed in both adult and pediatric patients

Surgeries done	Pediatric patients (eyes)	Adult patients (eyes)
Corneal tear repair	4 (5 events)	1
Globe repair	1	
Tissue adhesive and BCL	2 (3 events)	
Descematopexy	3	1
PK	2	6
Regraft		1
Cataract surgery	1	3
Vitrectomy		2
Retinal detachment surgery		3
Enucleation		1
Evisceration	1	
Glaucoma surgery		1

PK: Penetrating keratoplasty, BCL: Bandage contact lens

Discussion

Ours is the largest series in the published literature of 48 patients having keratoglobus with 21 patients in the pediatric age group and 27 in adult age group. Cameron *et al*. had reported 12 adult patients and 11 pediatric patients in their series.[2,3] Males were twice in number compared to females in our series similar to Cameron[2] Bilaterality was noted in all except one of our patients; we assume phthisis bulbi in three pediatric patients and one adult patient might have occurred secondary to trivial trauma in these patients with the presence of keratoglobus. Cameron have reported 3/12 adult patients to have unilateral keratoglobus.[2]

Best-corrected visual acuity was 20/40 or better in 6/42 (14.3%) pediatric eyes and 15 out of 53 (28.30%) adult eyes with keratoglobus. Visual acuity ranging from counting of fingers close to the face to no perception of light (PL) was noted in 21/42 (50%) pediatric patients and 20/53 (37.74%) adults.

Associated ocular conditions in our series were VKC in pediatric age group which may be due to the rubbing of

eyes similar Cameroon *et al*.[8] Choroidal osteoma, Retinitis pigmentosa, and pigment epithelial detachment were noted in adult patients, which have not been reported earlier.

Corneal tear or globe rupture in keratoglobus can occur either spontaneously or following trivial trauma.[3] Cameron *et al*. had reported that 7/11 patients (9 eyes) having keratoglobus with blue sclera with connective tissue disorder, had a rupture either spontaneously or secondary to trivial trauma.[3] Biglan *et al*. had reported corneal perforation in 15/20 eyes in patients of <18 years of age.[9] In our analysis, 12 eyes of pediatric patients had corneal perforation or tear secondary to trauma; six eyes had blue sclera, and four eyes had repeated corneal ruptures secondary to trivial trauma. The positive history of trauma was significant in the pediatric group as compared to adult group.

Cameron had reported that none of their 12 adult patients had corneal tear except one patient who had adherent leukoma suggestive of the prior event.[2] In our analysis, one adult patient presented with corneal tear and underwent repair with improved visual acuity postoperatively.

Descematopexy was performed for acute hydrops in three pediatric patients and had poor visual outcome. One adult patient underwent descematopexy, and visual acuity improved to 20/30. The remaining four adult eyes with acute hydrops had poor visual acuity to start with; one patient had RP, one was difficult to assess, one patient underwent enucleation for painful eye and one had PL. Basu *et al*. have reported that resolution time for clearing of corneal edema in acute hydrops is similar in keratoglobus for both descematopexy and with conservative management.[10] Patients presented with breaks in Descemet's membrane in seven eyes each in both pediatric and adult patients. Hydrops in our series is lesser (7/53 eyes – 13.20%) as compared to Cameron where they reported occurrence of hydrops in 10 out of 12 adult patients that is, 19 out of 21 eyes (90.47%).[2]

The average sim *k* value in pediatric patients was higher (55.05D ± 7.1D) than in adults in our study. However, this was not statistically significant. The adult sim *k* values were lesser (52.03D ± 8.32D) in our study compared to Cameron (>55D).[2] The corneal pachymetry values were lesser in pediatric patients as compared to adults.

Management of keratoglobus is challenging. Conservative management in the form of spectacles and contact lenses is described though trivial trauma such as contact lens insertion and removal can cause rupture of the globe.[2,4] One of the 4 patients dispensed with PROSE developed hydrops, and we attribute this to the suction which is created when the lens is removed.[11] Though the fitting of the PROSE was optimal when dispensed after performing a 4 h challenge, the suction generated with daily lens removal might be responsible for the occurrence of hydrops over follow-up. The other patient continued to wear PROSE with a follow-up of 3 years [Fig. 2].

Poor visual acuity was noted in 65% of pediatric patients with blue sclera-from no PL to counting fingers close to face indicating the blinding nature of the disease. This may be due to corneal tear secondary to trivial trauma or spontaneously with recurrent events of corneal rupture though the patients were using protective glasses. Multiple events can occur in the same eye or the other eye even when patients are using protective glasses.

The surgical management in our series was mainly for corneal tear or perforation and hydrops. When doing a surgical repair for a corneal tear, the thin cornea during suturing is a sign to examine the other eye carefully for the presence of keratoglobus. Large diameter PK can be performed in these patients to improve vision.[2] None of the patients during the study period had lamellar keratoplasty performed. In this study, PK was performed in two pediatric eyes and six adult eyes. The follow-up in pediatric patients was less and could not be analyzed. The visual acuity in adult patients ranged from 20/100 to 20/30. Three eyes developed glaucoma following keratoplasty; visual acuity was 20/100 in one patient in both eyes. The graft was clear in all but one adult eye that developed graft failure secondary to rejection. Subsequently, the patient underwent Descemet's stripping endothelial keratoplasty with trabeculectomy for post-PK glaucoma and the graft failed. Cameron had one graft failure secondary to glaucoma in five adult eyes that underwent PK.[2] Various surgical procedures such as epikeratoplasty, deep lamellar keratoplasty or tuck in keratoplasty are described in literature as case series or reports.[1,3,12] There are no standard surgical procedures described and the management is usually tailor made to suit individual patient based on the mode of presentation.

To summarize, this is the largest series of patients having keratoglobus. About 50% of pediatric eyes had no functional vision; 65% of pediatric patients with blue sclera had visual acuity ranging between no PL to counting fingers close to face indicating the blinding nature of the disease. Visual acuity of 20/40 or better was more in adult as compared to pediatric eyes. Visual acuity of counting fingers close to face to no PL was noted in 21/42 (50%) pediatric patients and 20/53 (37.74%) adults. Trivial trauma was responsible for corneal rupture with multiple events indicating the need for protective glasses. Descematopexy for acute hydrops had poor visual outcome and should be considered case wise. Keratoplasty should be performed with caution as 50% of patients (3 out of 6 eyes) had post-PK glaucoma. Surgery should be customized as per individual case as there are no clear guidelines.

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