

Outcomes of keratoplasty in a cohort of Indian patients with xeroderma pigmentosum

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Purpose: To evaluate the outcomes of keratoplasty for xeroderma pigmentosum (XP) performed at a tertiary eye care center. **Methods:** A retrospective review of medical records of those patients who were clinically diagnosed to have XP (54 eyes of 36 patients) and underwent keratoplasty; either deep anterior lamellar keratoplasty (DALK, four eyes), endothelial keratoplasty (EK, eight eyes), or penetrating keratoplasty (PK, 42 eyes) from 1994 to 2018. **Results:** The median age at surgery was 20.6 years (interquartile range [IQR], 14.6–27.6 years) and 20 (55.6%) were males. Graft failure occurred in 15 eyes (35.7%) in the PK group and two eyes (50%) in the DALK group; none failed in the EK group. The probability of graft survival in the PK group was 97.2% ± 2.7% at 1 year, 74.0% ± 8.0% at 2 years, and 54.8% ± 11.7% at 5 years. In the PK group, 13 eyes needed antiglaucoma medications, 11 eyes developed graft infiltrate, and 13 eyes needed secondary interventions (cataract surgery, excision biopsy, and tarsorrhaphy). In the EK group, three eyes needed secondary interventions (excision biopsy). Median postoperative endothelial cell density at the last follow-up in the PK group was 1214 cells/mm² (IQR, 623–2277 cells/mm²). **Conclusion:** Despite the complexities of the ocular surface and adnexal issues in XP, keratoplasty had reasonably good outcomes. More than half of the PK grafts survived 5 years with no failures in the EK group. Regular follow-up and timely management of suture-related infections raised intraocular pressure, and suspicious ocular surface lesions, in addition to solar protection, are important for the success of keratoplasty in these eyes.

Key words: Deep anterior lamellar keratoplasty, endothelial keratoplasty, penetrating keratoplasty, xeroderma pigmentosum

Xeroderma pigmentosum (XP) is an autosomal recessive inherited disorder characterized by hypersensitivity to ultraviolet radiation as a result of an underlying defective DNA repair mechanism. Ocular involvement includes changes in the lid, conjunctiva, and cornea secondary to solar radiation-induced damage.^[1] The various corneal manifestations include calcific degeneration, corneal perforation, corneal edema, epithelial haze, recurrent corneal ulcerations, scarring, vascularization, and neoplasia.^[1-7]

Corneal opacification may necessitate keratoplasty. The type of keratoplasty depends upon the characteristic changes in the corneal stroma. Those with significant anterior stromal haze need a penetrating keratoplasty (PK) while those with predominant corneal edema relative to anterior stromal haze can be successfully rehabilitated with endothelial keratoplasty (EK). Corneal endothelial affliction in XP has been well reported and occurs due to irreversible attrition of corneal endothelium on chronic sun exposure. Hence, deep anterior lamellar keratoplasty (DALK) in eyes with corneal opacification can have unpredictable outcomes due to a likelihood of lower endothelial reserve.^[8-10]

There are a few isolated case reports or case series of keratoplasty in the management of corneal complications in XP.^[1-4,11,12] Herein, we report the outcomes of keratoplasty (DALK, EK, and PK) performed in XP in a large series of patients.

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Methods

This retrospective study was approved by the Institutional Ethics Committee and adhered to the tenets of the Declaration of Helsinki. Informed consent was obtained from all patients. Fifty-four eyes of 36 patients with XP who underwent DALK (four eyes), EK (eight eyes), Descemet's stripping automated endothelial keratoplasty (DSAEK; seven eyes), Descemet's membrane endothelial keratoplasty (DMEK; one eye), or PK (42 eyes) as primary keratoplasty procedure from 1994 to 2018 were studied. Eyes with a prior or previous keratoplasty (repeat keratoplasty) were not included in the primary outcome analysis. Data collected included demographics, clinical features, previous ocular surgeries, type of keratoplasty, donor and recipient details, endothelial cell density, survival, and outcomes of keratoplasty.

The surgical decision about the type of keratoplasty was decided based upon the clinical slit-lamp examination findings. Those eyes where the loss of corneal transparency was predominantly due to corneal edema with a minimal degree of anterior stromal scarring were subjected to EK. Eyes with significant scarring of the stroma with no visualization of the anterior segment details underwent PK. Four eyes of three patients with scarred corneas were undertaken for

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DALK. Further, with the understanding and recognition of endothelial involvement in these patients,^[8-10] DALK became the less favored approach thereafter.

The pre and postoperative visual acuities were documented either in the LogMAR scale or Snellen notation in the medical records and the Snellen notations were converted into LogMAR scales during analysis. The vision achieved following objective and subjective refraction was recorded as best-corrected visual acuity (BCVA). Graft failure was defined as loss of graft transparency and/or decrease in BCVA by two or more lines in the absence of any other ocular comorbidity that could lead to a drop in vision.

Post-PK and post-DALK treatments involved a prophylactic broad-spectrum antibiotic for 1–2 weeks, and prednisolone acetate 1% eye drops every 3–4 hourly for initial 1 week with tapering doses every week and maintenance of twice-daily regimen till sutures were intact, followed by at least once daily after all sutures were out. In DSEK cases, the early postoperative steroid regimen was the same with a maintenance dose of once-daily till the last follow-up. In those with raised intraocular pressure with or without disc changes, antiglaucoma medication was initiated and loteprednol etabonate 0.5% was used instead of prednisolone acetate.

The statistical analysis was performed using the software Origin v7.0 (Origin Lab Corporation, Northampton, MA, USA). Continuous data were checked for normality by the Shapiro-Wilk test and equality of variance by the Levene test. Median and interquartile range (IQR) described the continuous data. Categorical data were described in proportions. DALK group was excluded from comparisons owing to the small sample size. Continuous parametric data were compared between EK and PK groups by Student *t*-test and nonparametric data by the Mann-Whitney U test. Categorical data were compared between EK and PK groups by Chi-square test or Fisher's exact probability test. A paired *t*-test was used to compare donor and postoperative endothelial cell density (ECD). Kaplan-Meier survival analysis was performed to estimate the probability of graft survival in the PK group. A *P* value of <0.05 was considered statistically significant.

Results

Demographics and preoperative characteristics

Overall, the median age at presentation to the clinic was 20.2 years (IQR, 10.9–23.9 years). Twenty (55.6%) were males. Both eyes underwent keratoplasty in 18 patients (50%). Table 1 summarizes the demographics and clinical characteristics of the study patients. The age at presentation to the clinic, gender, history of parental consanguinity, and preoperative vision were comparable between EK and PK groups. All eyes had a variable degree of conjunctival hyperemia and 29 (53.7%) eyes additionally had corneal vascularization. PK group had a significantly higher proportion of preoperative vascularization compared to the EK group. None of the eyes had keratoconus in the study cohort.

Eighteen eyes had prior ocular surgery and five patients had prior systemic surgery in the PK group. Previous ocular surgeries in the PK group included alcohol keratoepitheliopathy (*n* = 8 eyes), electrolysis (*n* = 1 eye), amniotic membrane graft (AMG; *n* = 7 eyes), conjunctival limbal autograft (*n* = 1 eye), conjunctival tumor excision (*n* = 11 eyes; of the 11 eyes, four had carcinoma in situ, two had ocular surface squamous neoplasia [OSSN], two had conjunctival dysplasia, two had regenerative atypia, and one had actinic keratosis on histopathology examination), cryotherapy (*n* = 7 eyes), incision biopsy (*n* = 1 eye), pannus

resection (*n* = 1 eye), simple limbal epithelial transplant (*n* = 1 eye), and tarsorrhaphy (*n* = 1 eye), while in DALK group it was excision biopsy, cryotherapy, and AMG in two eyes for suspected OSSN. Prior systemic surgeries in the PK group were excision biopsy (involving the head/face), plaque brachytherapy, plaque removal, and removal of mass on the head.

Intraoperative features

Table 2 summarizes the intraoperative features during keratoplasty in the study groups. The mean age of the patients was comparable between EK and PK groups. The preoperative BCVA was better in the EK group compared to the PK group. Additional surgical procedures performed along with PK included amniotic membrane grafting (*n* = 7 eyes), conjunctival peritomy (*n* = 1 eye), conjunctival resection (*n* = 1 eye), extracapsular cataract extraction with posterior chamber intraocular lens implantation (*n* = 2 eyes), subconjunctival avastin (*n* = 2 eyes), and tarsorrhaphy (*n* = 8 eyes), while one eye underwent phacoemulsification with implantation of a posterior chamber intraocular lens along with DSAEK. The donor cornea characteristics were comparable between EK and PK groups.

Postoperative course and outcomes

Table 3 summarizes the postoperative outcomes after keratoplasty.

Penetrating keratoplasty

Eleven eyes (26.2%) in the PK group had graft infiltrate at a median interval of 4.7 months (IQR, 2.6 months to 3.4 years) following surgery. Among the species identified, 71.4% were gram-positive cocci (such as *Streptococcus pneumoniae*); 28.6% were not conclusive on smear or cultures. One eye developed graft infiltrate for the second time at 4.2 years (organism unidentified) after the surgery. Thirteen (31.0%) eyes needed AGM during the median follow-up duration of 2 years (IQR, 3 months to 6 years) following PK. The median cup-disc ratio in the PK group was 0.50 (IQR, 0.30–0.60) at a median duration of 2 years (IQR, 2 months to 6 years) following surgery.

Thirteen eyes (31%) in the PK group underwent additional interventions at variable periods in the follow-up period after keratoplasty (seven eyes [16.7%] had two secondary interventions and five eyes [11.9%] had three or more secondary interventions). The first interventions were cataract surgery (*n* = 6 eyes), excision biopsy (*n* = 3 eyes; one each for eyelid tumor, OSSN and actinic keratosis), graft-host junction resuturing (*n* = 1 eye), synechiolysis with anterior chamber reformation (*n* = 1 eye), tarsorrhaphy (*n* = 1 eye) and tarsorrhaphy release (*n* = 1 eye). The second interventions were cataract surgery (*n* = 2 eyes), excision biopsy (*n* = 2 eyes; one each for OSSN and conjunctival epithelial hyperplasia), fibrin glue application (*n* = 1 eye), tarsorrhaphy (*n* = 1 eye), and Yttrium-Aluminium-Garnet laser capsulotomy (*n* = 1 eye). The third interventions were excision biopsy (*n* = 3 eyes; one each for conjunctival epithelial hyperplasia, eyelid basal cell carcinoma [BCC] and actinic keratosis), and tarsorrhaphy (*n* = 2 eyes). Two eyes had additional eyelid skin grafts and another eye had multiple additional procedures such as excision biopsy for eyelid BCC and squamous cell carcinoma over zygoma and tarsorrhaphy.

Secondary graft failures occurred in 15 eyes (35.7%) in the PK group. The causes of secondary graft failure were graft rejection with vascularization in six eyes (40%), progressive scarring with vascularization in six eyes (40%), and central graft infiltrate in three eyes (20%). The probability of graft survival in PK group was 97.2% ± 2.7% at 1 year (number at risk = 32), 74.0% ± 8.0% at 2 years (number at risk = 22), and 54.8% ± 11.7% at 5 years (number at risk = 8; Fig. 1). The follow-up duration, graft infiltrate, usage of

Table 1: Demographics and preoperative features of xeroderma pigmentosum

This table summarizes the demographics and clinical characteristics of the study patients

Parameter	DALK 4 eyes 3 patients	EK 8 eyes 6 patients	PK 42 eyes 28 patients	P ^a
Age at presentation (years), median (IQR)	15.9 (13.9-19.1)	16.9 (8.9-23.9)	20.5 (10.2-24.5)	0.62
Males, <i>n</i> (%)	3 (100%)	2 (33.3%)	15 (53.6%)	0.67
Parental consanguinity (recorded), <i>n</i> (%)	2 (66.7%)	4 (100%)	14 (82.4%)	1.00
Previous ocular surgery, <i>n</i> (%)	2 (50%)	0 (0%)	15 (35.7%)	0.09
Previous systemic surgery, <i>n</i> (%)	0 (0%)	0 (0%)	5 (17.9%)	0.56
Vascularization	2 (50%)	1 (12.5%)	26 (61.9%)	0.02
Pseudo-pterygium	0 (0%)	0 (0%)	1 (2.4%)	1.00
BCVA at presentation (recorded)				
PL/PR, <i>n</i> (%)	0 (0%)	0 (0%)	4 (13.3%)	0.57
Rest in LogMAR, median (IQR)	1.14 (0.18-2.09)	1.00 (0.48-1.79)	1.74 (1.11-3.00)	0.09

^aDALK group was not included because of small sample size; BCVA: Best-corrected visual acuity; DALK: Deep anterior lamellar keratoplasty; EK: Endothelial keratoplasty; IQR: Interquartile range; LogMAR: Logarithm of the minimum angle of resolution; PK: Penetrating keratoplasty; PL/PR: Perception of light/projection of light

Table 2: Intraoperative characteristics

This table summarizes the intraoperative features during keratoplasty in the study groups

Feature	DALK	EK	PK	P ^a
Preoperative BCVA				
FFL/PL/PR, <i>n</i> (%)	0 (0%)	0 (0%)	5 (14.7%)	0.56
Rest in LogMAR, median (IQR)	1.39 (0.18-1.79)	1.05 (0.48-2.09)	2.09 (1.48-3.00)	0.04
Age at surgery (years), median (IQR)	17.3 (15.9-19.7)	10.7 (8.9-22.6)	21.3 (15.6-28.2)	0.03
Concomitant surgery, <i>n</i> (%)	0 (0%)	1 (12.5%)	13 (31.0%)	0.41
Donor age (years), median (IQR)	30.5 (21-40)	40.5 (19-50)	26 (22-36)	0.53
Donor ECD (cells/mm ²), median (IQR)	2991 (2924-3058)	3040.5 (2770-3125)	2881 (2709.5-3149)	0.34
Donor size (mm), median (IQR)	8.25 (<i>n</i> =1)	8.00 (8.00-9.00)	8.00 (7.50-8.25)	N/A ^b
Recipient size (mm), median (IQR)	8.00 (<i>n</i> =1)	7.75 (7.50-8.00)	7.50 (7.00-7.75)	N/A ^b

^aDALK group was not included because of small sample size; ^bN/A: Not applicable due to small sample size in EK group; BCVA: Best-corrected visual acuity; DALK: Deep anterior lamellar keratoplasty; ECD: Endothelial cell density; EK: Endothelial keratoplasty; FFL: Fixation and following of light; IQR: Interquartile range; logMAR: Logarithm of the minimum angle of resolution; PK: Penetrating keratoplasty; PL/PR: Perception of light/projection of light

Table 3: Postoperative course and outcomes

This table summarizes the postoperative outcomes after keratoplasty

Feature	DALK	EK	PK	P ^a
Follow-up duration (years), median (IQR)	7.2 (2.2-8.8)	3.6 (0.5-6.1)	4.0 (1.6-8.1)	0.55
Graft infiltrate, <i>n</i> (%)	0 (0%)	0 (0%)	11 (26.2%)	0.17
Anti-glaucoma medications, <i>n</i> (%)	0 (0%)	2 (25%)	13 (31.0%)	1.00
Cup-disc ratio, median (IQR)	Not available	0.40 (0.40-0.90)	0.50 (0.30-0.60)	N/A ^b
Ocular surface and adnexal interventions after keratoplasty, <i>n</i> (%)	0 (0%)	3 (37.5%)	13 (31.0%)	1.00
Graft failure, <i>n</i> (%)	2 (50%)	0 (0%)	15 (35.7%)	0.09
Best achieved BCVA (LogMAR), median (IQR)	0.18 (0.10-1.48)	0.29 (0.10-0.48)	0.40 (0.30-0.70)	0.13

^aDALK group was not included because of small sample size; ^bNot applicable due to small sample size in EK group; BCVA: Best-corrected visual acuity; EK: Endothelial keratoplasty; IQR: Interquartile range; logMAR: Logarithm of the minimum angle of resolution; N/A: Not applicable; PK: Penetrating keratoplasty

AGM, postoperative surgical intervention before failure, and the rate of graft failure were comparable between EK and PK groups.

In the PK group, 12 eyes (28.6%) underwent repeat keratoplasty procedures (PK: Six eyes, DSAEK: Five eyes; and Boston keratoprosthesis: One eye). Among the six eyes that underwent repeat PK, two developed graft infiltrates, two had a rejection, and two eyes failed due to progressive vascularization and scarring (one of these had a repeat PK

subsequently). Among the five eyes that underwent DSAEK after failed PK, one eye maintained corneal clarity till the last follow-up, one developed infiltrate, and three grafts failed. Repeat PK was performed in all these three failed grafts, which was clear in one eye till the last follow-up, failed in another and the third eye underwent enucleation at 3 years after the third keratoplasty due to rapid growth of invasive carcinoma. One eye that underwent a keratoprosthesis, developed intractable

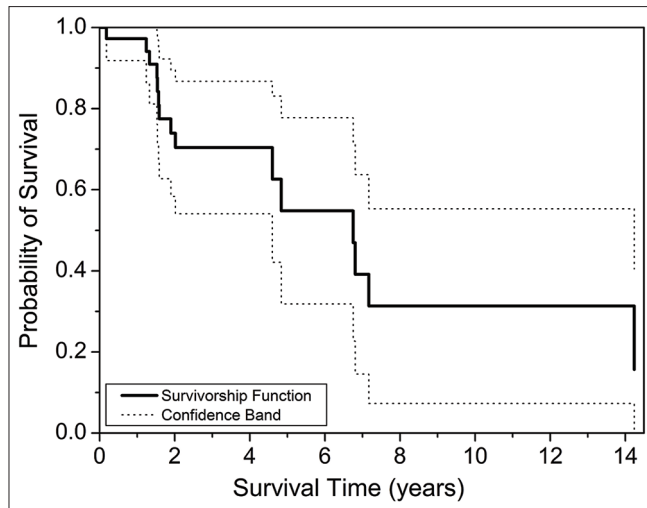


Figure 1: Kaplan-Meier survival analysis after penetrating keratoplasty in xeroderma pigmentosum eyes: This figure shows the probability of survival of grafts following penetrating keratoplasty in xeroderma pigmentosum

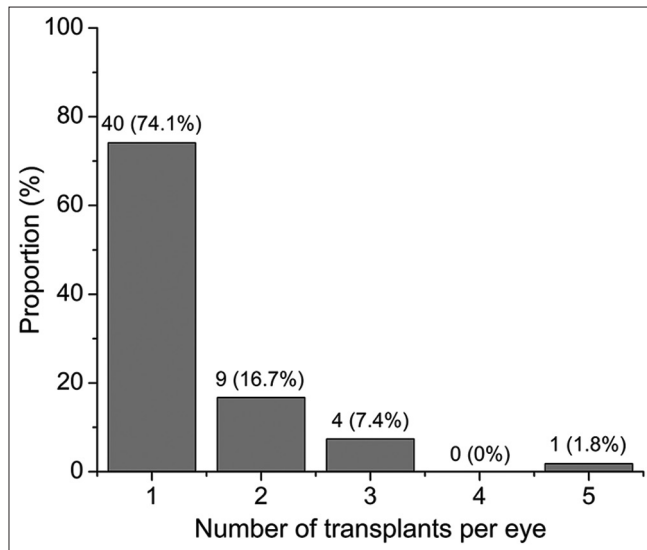


Figure 3: Number of corneal transplants per eye in xeroderma pigmentosum: This figure shows the number of transplants per eye in the study cohort

glaucoma and advanced glaucomatous optic neuropathy, for which filtering valve surgery was performed.

The specular microscopy was done in 11 eyes in the PK group at a median interval of 1.1 years (IQR, 0.9–5.2 years) after keratoplasty and the median ECD was 1214 cells/mm² (IQR, 623–2277 cells/mm²). There was a significant difference between the donor ECD and postoperative ECD ($P = 0.001$).

Endothelial keratoplasty

In the EK group, two eyes (25%) needed AGM at 7 months and 3.5 years, respectively following surgery. The median cup-disc ratio in the EK group was 0.40 (IQR, 0.40–0.90) at a median duration of 3.5 years (IQR, 3–3.5 years) following surgery. Three eyes (37.5%) in the EK group underwent excision biopsy following surgery (one each for conjunctival dysplasia, conjunctival regenerative atypia, and eyelid BCC), out of which two eyes underwent cryotherapy concomitantly and one eye (12.5%) underwent plaque brachytherapy. None of

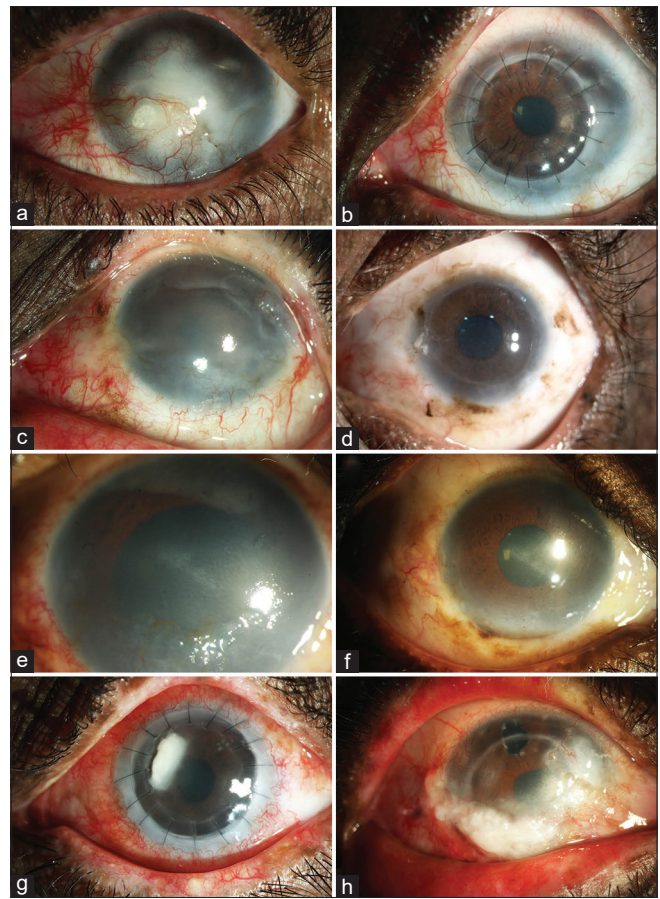


Figure 2: Representative images post-keratoplasty in xeroderma pigmentosum: (a) Preoperative photograph of the left eye of a patient shows scarring and vascularization for which penetrating keratoplasty (b) was performed; (c) Preoperative photograph of the left eye of a patient shows edema and mild stromal haze in central cornea for which Descemet's stripping automated endothelial keratoplasty (d) was performed; (e) Slit-lamp photograph of the right eye of a patient shows corneal edema involving the central cornea for which Descemet's membrane endothelial keratoplasty (f) was performed; (g) Suture-related infiltrate due to *Streptococcus pneumoniae*; (h) Ocular surface squamous neoplasia that developed a year after penetrating keratoplasty

the eyes failed in the EK group. Fig. 2 shows the representative images of some eyes that underwent PK and EK.

Deep anterior lamellar keratoplasty

There were two primary graft failures in the DALK group after uneventful surgery. The two cases of primary graft failure in the DALK group underwent optical PK and DSEK, respectively. Repeat DSEK remained clear until the last follow-up, whereas repeat optical PK underwent three repeats therapeutic PKs and the graft was failing until the last follow-up. Fig. 3 shows the number of transplants per eye in the study cohort. Nearly three-fourths of the eyes ($n = 40$) had one keratoplasty per eye, nine (16.7%) had two, and five (9.2%) had three or more keratoplasty per eye.

Discussion

XP is a disorder of impaired DNA repair mechanism that affects the sun-exposed organs. Ocular involvement has been reported in 30–40% of the patients with XP. The common ocular manifestations include lid abnormalities (ectropion, entropion, madarosis, trichiasis, lagophthalmos), conjunctival hyperemia, actinic keratosis, pterygium or pseudopterygium, corneal

opacification, haze and edema, and ocular surface neoplasia. Progressive changes in the cornea eventually lead to loss of vision necessitating keratoplasty.

The specific issues in XP that can potentially affect keratoplasty outcomes are eyelid and associated tear film abnormalities, corneal vascularization, risk of occurrence of ocular surface neoplasia, increased risk of graft rejection due to a vascularized bed, increased risk of suture-related complications, and difficulties in post-keratoplasty care (suture management, assessment of intraocular pressure, and disc evaluation) due to severe photophobia. In this study, we analyzed the visual outcomes and graft survival of keratoplasty (PK, EK, and DALK) in patients with XP seen at our tertiary eye care center. Some of the patients included in this study have a follow-up analysis of those reported earlier in literature with a shorter follow-up.

The ocular surface abnormalities often need attention either before keratoplasty or at the time of keratoplasty. Seventeen eyes (31.5%) needed an ocular surface surgery before keratoplasty and 14 eyes (25.9%) had concomitant ocular surface surgeries. These included amniotic membrane grafting, conjunctival peritomy, conjunctival resection, subconjunctival avastin, and tarsorrhaphy. The additional procedures were performed to prevent the anticipated ocular surface complications in the early postoperative period and make the ocular surface favorable for keratoplasty.

The postoperative management of XP patients' needs rigorous care and follow-up after keratoplasty, as the cornea is vascularized. More than 50% of the eyes had vascularization and conjunctival hyperemia was a universal clinical feature in all eyes. Approximately 70% of PK grafts developed microbial keratitis at some point in the follow-up period, which was bacterial. None of the patients in the DALK and EK groups had infections. Many of the XP patients are prone to develop ocular surface neoplasia. Twelve eyes had excision biopsy before keratoplasty (all were in the PK group) and eight eyes had excision biopsy after keratoplasty (five eyes after PK and three eyes after EK).

Although there were no graft failures after EK, 15 eyes had graft failure after PK. The various causes of graft failure were graft rejection with vascularization (40%), progressive scarring with vascularization (40%), and central graft infiltrate (20%). On account of corneal bed vascularization, suture loosening is likely to occur early and uncontrolled after a PK predisposing these eyes to infection. Considering the associated ocular surface comorbidities in these eyes, the probability of graft survival (~97% at 1 year, 74% at 2 years, and ~55% at 5 years after PK) seems to be a reasonably fair outcome. In our study, the median survival of PK grafts was 6.75 years or 81 months. This appears to be better than those reported by Joshi *et al.* for common indications of optical PK in the Indian population such as 27 months for corneal scars (post-infectious or posttraumatic) and corneal dystrophies or degenerations, 22 months for adherent leucomas, aphakic bullous keratopathies, pseudophakic bullous keratopathies, and phakic corneal decompensation, and 14 months for herpetic corneal scars, previously failed grafts, and congenital corneal scars or dystrophies.^[13] On comparing with the outcomes of optical PK for a homogenous indication lattice corneal dystrophy, the estimated median survival of grafts in XP seems to be lower (6.75 years versus 15.8 years).^[14]

There were no failures in those patients that had EK which reflects the inherent advantages of EK in these eyes. However, EK is limited to a smaller proportion of patients who have only a limited degree of anterior stromal scarring making EK a preferred option over PK in such cases. While two eyes of a patient maintained graft clarity after DALK, there were two

primary graft failures after uneventful DALK, one of which was managed with DSEK and the second with a repeat PK. The patient with clear grafts after DALK, however, had a remarkably low endothelial cell count.

Limitations of retrospective series with long look-back periods include the inability to examine patients directly, potential for missing data, patients with varying follow-up intervals, and changing surgical indications and techniques over time.

Conclusion

Despite the complexities of the ocular surface and adnexal issues in XP, keratoplasty had reasonably good outcomes. More than half of the PK grafts survived 5 years with no failures in the EK group. The outcomes of DALK could not be compared in this study due to a smaller number of eyes that had DALK. Given the evidence of endothelial affliction in XP, the outcomes are likely to be unpredictable. The ocular surface and suture-related events after DALK are likely to be similar to PK. Regular follow-up and timely management of suture-related infections raised intraocular pressure and suspicious ocular surface lesions, in addition to solar protection, are important for the success of keratoplasty in these eyes. Further, as XP is a genetic condition, these patients need constant monitoring and surveillance for a lifetime.

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Conflicts of interest

There are no conflicts of interest.

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