

Keratoplasty for keratomalacia in preschool children

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Br J Ophthalmol 2003;87:538–542

Aim: To study the results of surgical management of keratomalacia in children.

Methods: A clinical case series of all children with keratomalacia, admitted to an Indian centre during the period from June 2000 to June 2001 is presented. The parameters evaluated were demographic data, systemic associations, and results of medical and surgical intervention.

Results: 29 children with keratomalacia ranging from 2 months to 5 years of age (mean 1.8 (SD 1.4) years) were included in the study. All children belonged to families of lower socioeconomic status. 27 patients (93.1%) had not been immunised at all. The systemic diseases precipitating the onset of keratomalacia included measles (41.37%), pneumonia (31.03%), and acute diarrhoea (37.93%). 36 eyes (66.7%) had total corneal melting and 11 (20.3%) eyes had paracentral corneal melting. In 15 eyes (27.8%) an emergency tectonic penetrating keratoplasty was performed of which only five grafts (33.3%) remained clear at a mean follow up of 7.3 (6.8) months (range 3–24 months). Seven eyes underwent optical penetrating keratoplasty, of which four grafts (57.14%) remained clear at a mean follow up of 6.4 (3.6) months (range 3–12 months). None of these could achieve a visual acuity better than 6/60.

Conclusions: Corneal grafting surgery in keratomalacia is associated with poor visual outcome.

Keratomalacia is the major cause of paediatric ocular morbidity and severe visual impairment in developing countries.^{1,2} Ocular surface changes include xerosis, keratinised plaques, stromal punched out ulcers, and focal or diffuse stromal melting.³

Keratomalacia due to vitamin A deficiency as an important cause of preventable corneal opacification has a reported percentage varying between 8% and 27.3%.^{3–6} It is one of the common indications for keratoplasty in the paediatric age group.⁵ In the present study we report the associations and success of various treatments in the management keratomalacia in children.

MATERIAL AND METHODS

Case records of all children with keratomalacia admitted to the cornea service of Rajendra Prasad Centre for Ophthalmic Sciences, New Delhi, India, during the period from June 2000 to June 2001 were reviewed. Children with corneal melting in one or both eyes were included in the study.

The parameters evaluated on a chart review were age, sex, socioeconomic status (based on composite socioeconomic status scale^{7,8}), systemic associations, immunisation status (whether partially or fully immunised for age according to the national immunisation schedules followed by the Government of India), extent of corneal involvement in both eyes, microbial investigations and details of the medical and surgical management, clinical outcome, and complications. We did

not estimate serum retinol in our patients as the clinical presentation was standard.

At the initial presentation, visual acuity was recorded in possible cases. Visual acuity for older children was determined with the Snellen visual acuity chart and with Teller's acuity cards, for the younger children. Anterior segment examination and corneal scraping was sent for culture in blood agar, chocolate agar and Sabouraud's agar. Paediatric consultation for systemic management had been done for all cases.

All children received vitamin A supplementation as recommended by the World Health Organization (WHO) at the time of presentation. Vitamin A supplementation was done in the form of oral or parenteral dosage as per WHO recommended protocol—that is, 200 000 IU on 2 consecutive days and one dose repeated after 2 weeks in patients over 1 year of age and with body weight over 8 kg. Infants less than 1 year of age and children less than 8 kg body weight were given half the dosage. Children less than 6 months were given 50 000 IU doses for 2 consecutive days. Those whose general condition was not fit for general anaesthesia were subjected to medical management and subsequent surgical intervention, in the form of optical iridectomy or penetrating keratoplasty, was done on stabilisation of the general systemic status. Topical medical management comprised instillation of 4 hourly ciprofloxacin 0.3% (Ciprobid eye drops, Zydus Cadilla) eye drops along with ocular lubricants.

Children with diffuse corneal melt who were fit for general anaesthesia underwent emergency tectonic keratoplasty. Fresh McCarey-Kaufman preserved donor corneal buttons punched out from the endothelial side on a Teflon block with disposable corneal trephines and oversized by 1 mm greater than the host trephination (as our experience had shown better results with a 1 mm oversized graft in paediatric eyes and eyes with irido-corneal adhesions^{9,10}) were used. Pupilloplasty, synechiolysis, anterior segment reconstruction, and cataract extraction were performed when necessary. Grafts were sutured with interrupted 10-0 Nylon sutures.

Children with healed keratomalacia underwent optical iridectomy or penetrating keratoplasty. Fresh McCarey-Kaufman preserved donor corneal buttons punched out from the endothelial side on a Teflon block with disposable corneal trephines and oversized by 1 mm greater than the host trephination were used. While cutting the host cornea, the trephine blade was used to make an approximately three quarter depth cut after proper centration. A debulking technique with lamellar dissection¹¹ was used initially to preserve as much iris tissue as possible and then the anterior chamber was entered. The host cut was completed with curved Vannas scissors. Pupilloplasty, synechiolysis, anterior segment reconstruction, and cataract extraction were performed when necessary. Grafts were sutured with interrupted 10-0 Nylon sutures.

All patients were treated with 2 hourly topical steroids (betamethazone sodium phosphate 0.1%) (Betnesol eye drops, Glaxo) and antibiotics (ciprofloxacin 0.3%) (Ciprobid eye drops, Zydus Cadilla) four times, ocular lubricants 2 hourly, and homatropine (2%) eye drops twice daily after surgery. Topical steroids were reduced from 2 hourly for the first week to six times daily from the second week to the third

Table 1 Data of patients with acute keratomalacia

No	Age (months) /sex	Clinical presentation	Management	Follow up (months)	Visual acuity at presentation/at final follow up	Clinical outcome
1	24/F	RE 3 mm corneal ulcer* LE total corneal melt	RE medical LE tectonic PK	18	4/60/6/36 PL/1/60	RE moderate scar LE graft failed
2	9/M	RE 85 mm melt LE anterior staphyloma	RE tectonic PK + lens asp+ AV	6	PL/2/60 No PL/-	RE graft failed LE phthisis
3	18/M	RE total corneal melt LE phthisis	RE tectonic PK	6	PL/2/60 -	RE graft failed
4	36/F	RE total corneal melt LE phthisis	Both eyes tectonic PK	2	PL/3/60 -	Both eyes pthisis
5	18/F	RE total corneal melt LE phthisis	RE tectonic PK twice	24	PL/2/60 -	RE pthisis
6	42/F	Both eyes total corneal necrosis	Both eyes tectonic PK	4	PL/1/60 PL/2/60	RE graft clear LE graft failed (rejection)
7	2/M	RE total corneal Melt	RE tectonic PK	4	PL/1/60	RE graft failed
8	24/M	LE xerosis Both eyes 8 mm corneal melt	LE medical RE medical	6	6/9/6/9 PL/2/60	LE corneal xerosis RE adherent leucoma LE graft clear
9	8/M	RE 85 mm corneal melt LE xerosis	LE tectonic PK RE tectonic PK	5	PL/2/60 6/12/6/12	RE graft clear LE corneal xerosis
10	18/F	RE 9 mm corneal melt LE corneal ulcer*	RE tectonic PK LE medical	3	RE tectonic PK 3/60/6/36	RE phthisis LE mild scar + corneal xerosis
11	18/F	RE 85 mm corneal melt LE 35 mm cornea Ulcer*	RE tectonic PK	3	PL/1/60	RE graft clear
12	36/M	Both eyes 9 mm corneal melt RE phthisis	LE medical RE tectonic PK	3	4/60/6/36 Both eyes PL/1/60	LE mild scar + corneal xerosis RE graft failed
13	24/F	LE inferior 4 mm melt with perforation*	LE tectonic PK	18	No PL 2/60/6/60	LE severe scar LE graft clear
14	24/F	RE 5 mm corneal melt LE total corneal melt	Both eyes medical	3	PL/6/36 PL/no PL	RE adherent leucoma LE phthisis
15	36/M	Both eyes total corneal melt	Both eyes medical	Lost to FU	Both eyes PL/-	-
16	6/M	RE 9 mm corneal melt LE 5 mm corneal ulcer*	Both eyes medical	Lost to FU	PL/-	-
17	36/M	RE 6 mm corneal melt* LE corneal xerosis	Both eyes medical	3	2/60/- PL/6/36	RE adherent leucoma LE normal
18	9/M	Both eyes 8 mm corneal melt*	Both eyes medical		6/24/6/12 Both eyes PL/6/36	RE adherent leucoma LE adherent leucoma →OI
19	3/M	RE 35 mm corneal melt* LE corneal xerosis	Both eyes medical	6	PL/6/36 4/60/6/60	RE mild scar RE normal
20	24/M	RE total corneal melt LE phthisis	Both eyes medical	7	PL/6/60 No PL	RE adherent leucoma →OI
21	5/M	Both eyes 6 mm corneal melt	Both eyes medical	8	Both eyes PL/6/60	Both eyes adherent leucoma →OI
22	3/M	RE 8 mm corneal melt LE 7 mm corneal melt with perforation	Both eyes medical	3	Both eyes PL/PL	Both eyes adherent leucoma → anterior staphyloma (irregular FU)
23	60/F	Both eyes 9 mm corneal melt	Both eyes medical	11	PL/2/60 PL/no PL	RE adherent leucoma → PK + lens asp +AV (graft clear) LE phthisis
24	12/F	RE 75 mm corneal melt LE 35 mm corneal melt*	Both eyes medical	12	PL/2/60 2/60/6/18	RE adherent leucoma → PK (graft clear) LE mild scar
25	12/M	RE 8 mm melt LE 3 mm corneal ulcer*	Both eyes medical	4	PL/PL	RE anterior staphyloma → PK + lens asp + AV (graft failed)
26	60/M	Both eyes 95 mm corneal melt	Both eyes medical	13	3/60/6/18 PL/PL PL/6/60	LE mild scar RE anterior staphyloma LE adherent leucoma →PK + anterior segment reconstruc + AV (graft clear)
27	42/F	RE 3 mm corneal melt* LE 8 mm corneal melt	Both eyes medical	9	3/60/6/18 PL/HMCF	RE mild scar LE adherent leucoma → PK (graft failed)
28	12/M	Both eyes 8 mm corneal melt	Both eyes medical	4	PL/PL	RE anterior staphyloma
29	9/M	RE 8 mm corneal melt LE phthisis	Both eyes medical	4	PL/HMCF PL/PL No PL/no PL	LE adherent leucoma → PK (graft failed) RE adherent leucoma →PK (graft clear)

VA = visual acuity ; BCVA = best corrected visual acuity ; M = Male, F = Female; PK = penetrating keratoplasty; PL = perception of light; MLCO = maculoleucomatous corneal opacity; Asp = aspiration; AV = anterior vitrectomy; PED = persistent epithelial defect; FU = follow up; NMCO = nebulomacular corneal opacity; OI = optical iridectomy; HMCF = hand movements close to face; reconstruc = reconstruction.

*Paracentral corneal ulcer.

Table 2 Initial clinical presentation

Surgical intervention	No of eyes (n=54)	%
Total corneal melt	36	66.7
Paracentral corneal melt	11	20.3
Anterior staphyloma	1	1.9
Phthisis	6	11.1

month after surgery. The ciprofloxacin 0.3% eye drops (Ciprobid eye drops, Zyclus Cadilla) were given four times daily for 1 month. The dosage of prednisolone acetate 1% was tapered to four times daily for the next 6 months.

Following surgery, all children had remained hospitalised during the first 2 weeks. The patients had undergone weekly follow up examination for at least 3 months after surgery followed by monthly examination. Patients had been examined more frequently if there were new symptoms reported by the parents or patients. Each examination had included an interim history of ocular symptoms, intraocular pressure recording with a Perkins tonometer, biomicroscopy for integrity of the corneal epithelium, subepithelial infiltrates, stromal inflammatory cells, grading of stromal vessels (if any), epithelial/endothelial rejection lines, graft clarity cells and flare in the anterior chamber, and keratic precipitates. Refraction and visual acuity testing were performed at 3 and 6 months after surgery and 3 months thereafter.

Graft rejection was diagnosed when there was graft oedema without any apparent clinical reason after an initial period of a clear graft for at least 10 days. Patients with signs of rejection were treated with prednisolone acetate 1% eye drops every hour during the first week and then tapered to every 4 hours by the end of 1 month. No systemic therapy was given. Vascularised and infiltrated sutures were removed. Amblyopia therapy with conventional occlusion was begun as early as possible.

Tectonic keratoplasty was considered successful if ocular structural integrity was preserved at 3 months of postoperative follow up. The optical graft was considered successful if it remained clear (clear grafts were defined as optically clear corneas with no Descemet's folds or oedema and full visibility of the iris details) at 3 months of follow up.

RESULTS

Of a total of 89 paediatric in-patients to the cornea service during the period June 2000 to June 2001, 29 children (32.6%) had keratomalacia (Table 1), of which 18 were male and 11 female, and ranged in age from 2 months to 5 years. The mean age of the patients was 1.8 (SD 1.4) years. Five children were below 6 months of age and 24 were above 6 months of age. Mean follow up was 7.22 (5.5) months (range 2–24 months).

All children (100%) belonged to the lower socioeconomic status and had severe protein energy malnutrition. Twenty seven patients (93.1%) were not immunised and the remaining two were partially immunised. The systemic diseases present at the onset of ocular pathology included pneumonia (9/29, 31.03%), acute diarrhoea (11/29, 37.93%), and measles (12/29, 41.37%).

Of 29 children presenting with keratomalacia, 25 patients (86.2%) had bilateral corneal melting. Four patients (13.7%) had unilateral corneal melting with xerosis in the other eye (Table 1). Of 54 eyes with keratomalacia, seven eyes (12.9%) were not salvageable at the time of presentation (phthisical six (11.1%), anterior staphyloma one (1.9%)). Thirty six eyes (66.7%) had total corneal melting and 11(20.3%) eyes had paracentral corneal melting (Table 2).

Of 13 children fit for anaesthesia at initial presentation corneal scrapings for microbiological investigations had been

Table 3 Surgical management in keratomalacia

Surgical intervention	No of eyes (n=26)	%
Tectonic keratoplasty	15	57.7
Optical iridectomy	4	15.4
Optical penetrating keratoplasty	7	26.9

sent in five eyes of five children, of which coagulase negative staphylococci were isolated in three eyes and no organisms were isolated in two eyes. In the remaining 10 eyes of eight children who underwent general anaesthesia, no corneal scraping had been sent because of the extensive limbus to limbus corneal melting. In these eyes the whole of the cornea was replaced with a thin fibrinous membrane.

In 15 eyes (15/54, 27.8%) of 13 patients with acute corneal melting, fit for anaesthesia at initial presentation, an emergency tectonic penetrating keratoplasty (Table 3) was performed. In two patients bilateral tectonic grafts was performed.

Thirty two eyes of 16 patients with keratomalacia, including those not fit for general anaesthesia and who could not undergo tectonic keratoplasty, were medically managed. Of these, 14 healed with adherent leucoma formation (43.75%), eight as corneal scar (25%), four (12.5%) went into anterior staphyloma (irregular follow up), two became phthisical (6.25%), and four (12.5%) were lost to follow up.

Of the 14 eyes with adherent leucoma, three eyes with visual acuity of 6/36 required no surgical intervention, as the lesions were peripheral with a clear central cornea. Subsequent surgical intervention in form of optical iridectomy was done in four eyes and optical penetrating keratoplasty in seven eyes (Table 3). In the seven eyes with healed keratomalacia, which underwent optical penetrating keratoplasty, additional procedures such as cataract removal (three eyes), anterior segment reconstruction (one eye), and anterior vitrectomy (three eyes) were performed (Table 1).

All donor grafts were 1 mm oversized and the donor graft size of the tectonic keratoplasties ranged from 5.5 mm to 11 mm. Donor grafts of the optical keratoplasties ranged from 7.5 mm to 9.0 mm. One patient with tectonic keratoplasty for acute keratomalacia underwent resuturing on the fourth postoperative day. One patient with emergency tectonic keratoplasty underwent repeat tectonic keratoplasty for corneal melt secondary to persistent epithelial defect. Graft rejection occurred in one eye and epithelial rejection in one eye in the tectonic keratoplasties.

Of 15 eyes, the globe of 11 eyes (73.33%) with total corneal sloughing was salvaged by tectonic keratoplasty. Four eyes (36.37%) went into phthisis. Only five tectonic grafts (33.3%) remained clear at a mean postoperative follow up of 7.33 (6.78) months (range 3–24 months). None of the tectonic grafts achieved visual acuity greater than 3/60 except one, which had undergone a tectonic patch graft and had a postoperative best corrected visual acuity of 6/60.

Eyes with corneal scar (eight eyes) had best corrected visual acuity less than 6/18 (ranging from 3/60 to 6/18). Four eyes with optical iridectomy achieved a best corrected visual acuity of 6/60. Four of the seven optical grafts (57.14%) remained clear at a mean follow up of 6.43 (3.64) months (range 3–12 months). Postoperative visual acuity of 6/60 was obtained in one eye and <3/60 in three eyes of optically clear grafts. Amblyopia was responsible for non-improvement in vision in cases with clear grafts. Poor ocular surface, vascularisation, and secondary glaucoma in the remaining three cases (3/7, 42.9%) accounted for graft failure in the optical grafts.

DISCUSSION

Keratomalacia due to severe vitamin A deficiency is common in developing countries. A major study of 162 children with

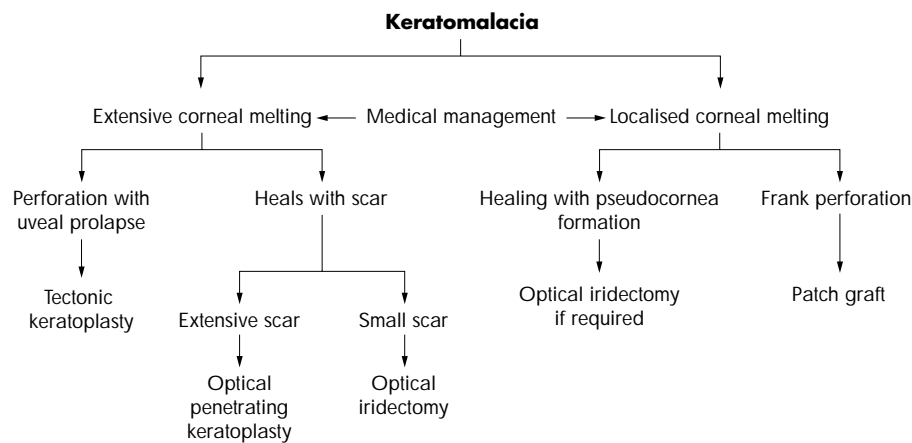


Figure 1 An algorithm for management of keratomalacia.

nutritional keratopathy analysed the pathology and clinical presentations of corneal xerophthalmia and keratomalacia.² In a follow up study of patients of severe xerophthalmia,¹² of 32 children with keratomalacia, nine died because of severe protein energy malnutrition. Of those remaining, five had become bilaterally blind, nine unilaterally blind, while nine retained adequate vision. No detail of visual acuity assessment has been cited in this study. The low prevalence of keratomalacia in community surveys is attributed to the high mortality in keratomalacia.¹²

Non-infectious keratomalacia in young children resulting from vitamin A deficiency is predisposed by multiple factors like malnutrition, systemic diseases, and lack of immunisation. Acute corneal melting result from the ocular pathological changes in severe vitamin A deficiency and is invariably accompanied by malnutrition^{5 13 14} and systemic disease.^{4 5 13}

Keratomalacia in children is hastened by protein-caloric malnutrition in addition to its association with measles.¹³ Malnutrition leads to multiple vitamin deficiencies including vitamin A deficient states and hence triggers keratomalacia. In our study, all the 29 children were malnourished. Pneumonia, diarrhoea and measles were the systemic conditions leading to the occurrence of keratomalacia in our study. Of 29 children, 27 (93.1%) were not immunised at all. The remaining two children (6.9%) had been partially immunised. Measles vaccination is a major preventive measure in malnourished children and can help to decrease nutritional keratomalacia in developing countries.¹³ Incomplete immunisation status predisposes to occurrence of systemic illnesses like measles and has been found to be associated with an increased risk for corneal ulceration.⁷ Given the rapidity of corneal necrosis and the potential blinding nature of the disease, the need for better preventive healthcare facilities to reach target populations is great.¹⁵ Our study has the limitation of being a small clinical case series; a larger community based case-control study is required to comment on the various associations and risk factors for keratomalacia.

Besides systemic management, specific medical measures in acute keratomalacia include vitamin A supplementation. Vitamin A supplementation can help reduce morbidity and mortality in affected children.^{14 16} The response to medical management is usually rapid and has good prognosis for visual improvement in less severe cases.² Our series, though small, also shows a similar result of good healing in cases of less severe corneal necrosis with preservation of relatively clear corneal areas, retaining ambulatory vision in these patients. Small perforated corneal ulcers are plugged by iris tissue and heal with adherent leucoma formation. In our series, all paracentral lesions healed as small adherent leucomas and corneal scars. Cases of severe necrosis of the cornea with or without perforation require surgical intervention in

the form of patch grafts¹⁷ and tectonic keratoplasty.¹⁸ Keratoplasty surgical techniques in paediatric eyes are difficult owing to the small anatomical configuration and decreased ocular rigidity. Large grafts are invariably required because of the total corneal involvement by the melting process. Also, special problems include difficulty in exact definition and demarcation of the unaffected host corneal tissue. This may cause annoying cheese wiring of the suture through the host rim during suturing and may necessitate resuturing in the immediate postoperative period as a result of the instability of recipient corneal stroma.

Our case series reports the clinical outcome of tectonic and optical keratoplasties in acute and healed keratomalacia in children. Singh and Malik¹⁸ have reported tectonic keratoplasty in eight eyes of six patients of keratomalacia and achieved therapeutic success in all. In our study, anatomical success rate of tectonic keratoplasty, in terms of restoring ocular structural integrity, in acute keratomalacia was 73.3% (11/15). Tectonic grafts remained clear in only 33.3% (5/15) of cases. Poor optical success rate of tectonic keratoplasty in acute keratomalacia in our series was due to ocular surface problems and graft rejection.

Keratoplasty for optical rehabilitation in healed keratomalacia had an anatomical success rate of 57.14% (4/7) in our series. Ocular surface problems and secondary glaucoma are responsible for failure of optical grafts. Poor visual outcome was due to the existence of dense amblyopia in these cases.

Despite aggressive and timely intervention, keratomalacia remains a preventable bilaterally blinding disorder in young children with poor visual recovery and suboptimal functional results. Based on our experience, we suggest an algorithm for management of keratomalacia cases (Fig 1).

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Accepted for publication 14 September 2002

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