

Congenital entropion with progeria: 4 going 40

Himika Gupta, Suhas Pawar

Key words: Entropion, maternal skin graft, mucus membrane graft, progeria

A 4-year-old male, with progeria, presented with pain, redness, and watering in the right eye since birth [Fig. 1a].

He had a biological age of 40 years, pinched nose facies, fat atrophy, senile and fragile skin. Right eye upper lid was inturned and shortened, inducing a 3 mm lagophthalmos. Posterior lamellar shortening was more than anterior lamellar shortening. The eyelashes rubbed on the cornea, causing superior corneal keratinization. Due to renal compromise, atherosclerosis, failure to thrive, as well as difficult intubation, he was considered high risk for anesthesia and surgery.

Surgery involved strategic placement of a double maternal donor graft as follows:

1. A mucus membrane graft^[1] for posterior lamellar lengthening was placed just inside the upper lid margin [Fig. 2a and b].
2. A full-thickness skin graft was placed just below the brow for anterior lamella lengthening, ensuring that both grafts were not placed one over the other [Fig. 2a-c].

Corneal scraping was done and bandage contact lens was placed. On subsequent follow-up, both grafts integrated well and entropion was corrected [Fig. 1b and c]. At 2 years follow-up, his lid position and ocular surface were well maintained.

Progeria, also known as Hutchinson–Gilford progeria syndrome (HGPS), is a rare genetic condition (1 in 4 million births) that causes accelerated aging. Average life expectancy is 13 years. The commonest eye finding is nocturnal lagophthalmos and exposure keratopathy.^[2] Only two cases of progeroid entropion have been reported.^[3,4] Approach to such patients can be a challenge as they are pediatric patients with geriatric problems.

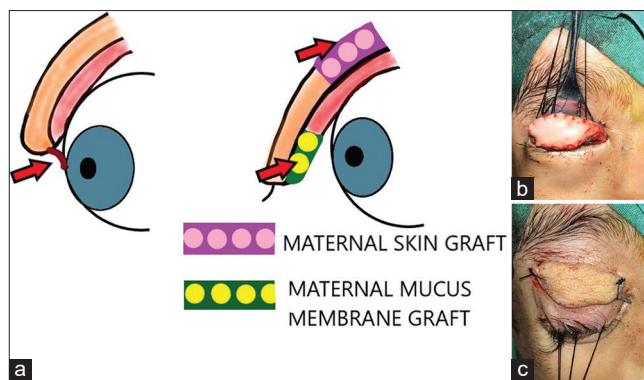


Figure 1: (a) Preoperative presentation with upper lid entropion. (b) Postoperative appearance with ocular surface improvement, at 1 month follow-up. (c) Postoperative outcome at 3 months follow-up



Figure 2: (a) Diagrammatic presentation of strategic placement of grafts. (b) Maternal mucus membrane graft placed just inside the lid margin (upper lid everted). (c) Maternal skin graft positioned along the upper lid crease

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Department of Pediatric Ophthalmology, B J Wadia Children's Hospital, Mumbai, Maharashtra, India

Correspondence to: Dr Himika Gupta, Spectra Eye Care Clinic, F/4-2 Sector 5 CBD Belapur, Navi Mumbai - 400 614, Maharashtra, India. E-mail: himika.gupta@gmail.com

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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