

Repair of cicatricial ectropion in patients with Harlequin Ichthyosis

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Introduction

Harlequin ichthyosis is an extremely severe hereditary autosomal recessive skin disorder with remarkable hyperkeratosis and increased formation of epidermal scales giving the appearance of fish skin. Cicatricial ectropion due to excessive dryness of the skin is the most common ocular manifestation resulting in exposure keratopathy. The aim of this study is to present our experience with two children suffering from Harlequin ichthyosis that underwent correction of their cicatricial ectropia.

Methods

A retrospective case notes review of two patients with Harlequin ichthyosis. The first baby was examined immediately after birth and underwent surgical repair at the age of 6 weeks. The second one was assessed at the age of 6 months. Late surgical intervention was performed at the age of 18 months.

Literature

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Goldberg RA, Lee S, Jayasundera T, Tsirbas A, Douglas RS, McCann JD. Treatment of lower eyelid retraction by expansion of the lower eyelid with hyaluronic Acid gel. *Ophthal Plast Reconstr Surg.* 2007;23(5):343-8.

Results

The first patient (six weeks old) had bilateral upper eyelid cicatricial ectropia which were successfully treated in an early stage with posterior auricular skin grafts. The second one had severe bilateral upper and lower eyelid cicatricial ectropia with a risk of exposure keratopathy and was treated initially (at the age of 6 months) by two sessions of subcutaneous hyaluronic acid injections (Restylane subQ). After the initial correction of the lids' malposition, recurring bilateral ectropia necessitated repeated administration of hyaluronic acid under the eyelids at the age of 9 and 16 months resulting in improvement of ectropion. New recurrence was corrected by the use of full thickness supraclavicular and preauricular skin autografts at the age of 18 months which resulted in well positioned eyelids and full eyelid closure.



Fig 1. Severe bilateral upper and lower eyelid ectropia alongside with a left paracentral corneal opacity due to corneal exposure found on the second baby.



Fig 2. Surgical repair resulted in full lid closure with no evidence of rejection or contraction of the grafts.

Conclusion

In Harlequin Ichthyosis skin alterations and keratinisation cause shortening of the anterior lamella affecting more frequently the upper eyelids. Surgical repair requires lengthening of the anterior lamella and cicatricial ectropion can be effectively corrected with free skin grafts. Surgical correction remains a challenge in clinical practice as patient's skin condition makes the usual skin graft donor sites unavailable. However, injection of dermal fillers can provide adequate tissue expansion in the early period of a premature baby. This will allow the child to mature in order to make general anaesthesia safer and improve the quality and quantity of skin that should be harvested for grafting.

The necessity for surgery in ichthyosis has to be individualised. The widespread involvement of the skin in early infancy increases the risk of graft contraction and surgical treatment could be considered safer and more successful if postponed for a later stage when patients have better skin quality.