

Management of chronic facial wounds in infants and children with Herlitz junctional epidermolysis bullosa (HJEB)

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Introduction

Epidermolysis bullosa (EB) is an umbrella term for a group of genetically determined skin fragility disorders. Its effects vary between painful blistering of hands and feet, through increasing disability with a greatly increased risk of squamous cell carcinoma to, in its most severe form, death in early infancy. Whilst work is progressing towards stem cell and other therapies, at the present time management focuses on nutritional supplementation, pain control and skin and wound care. In severe forms of EB chronic wounds develop and healing is compromised by nutritional deficiencies, continual trauma, colonisation and infection and the underlying gene defect.

There are 4 main types of EB with Herlitz junctional EB (HJEB) being the most severe form. The prognosis of HJEB is poor with a predicted life expectancy of less than five years and the majority will die in infancy. Death results from the combination of failure to thrive and respiratory distress from repeated laryngeal blistering and subsequent scarring. One of the most distressing symptoms is the development of large areas of facial ulceration which are notoriously difficult to heal. These wounds result from continual trauma to the fragile skin and in combination with poor nutrition and chronic anaemia these are very difficult to manage. Exuberant over-granulation tissue which is friable and bleeds easily is a feature in those with junctional EB. It is difficult to manage these wounds with sheet dressings due to the risk of traumatic removal by the child and problems with safe fixation. Some of these children have tracheostomies which could become occluded by a displaced dressing leading to an obstructed airway.

This small study will demonstrate the combination of Flaminal® enzyme alginogel and a very potent topical steroid ointment leading to successful healing of chronic facial ulceration.

Method

Children and infants with HJEB who had chronic facial wounds were selected for the study. Thankfully HJEB is a rare disease and therefore numbers were small with four children selected for the initial study.

Factors considered were the requirements of the treatment to:

- Be pain-free with no stinging
- Be atraumatic
- Reduce critical colonisation and infection
- Reduce over-granulation tissue
- Have the potential to heal wounds

The wounds were initially treated by application of Flaminal® (Crawford Healthcare, UK). Flaminal® is an enzyme alginogel which has the ability to debride, balance moisture levels and is a very effective

antimicrobial agent. It contains the natural occurring enzymes glucose oxidase and lactoperoxidase which promote effective antimicrobial activity without the potential to damage healing cells. Flaminal® contains alginate which balances moisture levels within the wound. The product is easy to apply and does not sting which is a very important consideration in the management of infants and children.

Facial wounds were covered with a thick layer of Flaminal® once a day. Flaminal® Forte was used on moist areas and Flaminal® Hydro to areas of crusting and drier wounds. The wounds were left uncovered. After one week of this treatment clobetasol propionate ointment was applied in a thin layer 12 hours after the application of Flaminal®. It was necessary to continue daily treatment with Flaminal® in conjunction with the steroid ointment in order to control the bioburden as these wounds are continually contaminated by food and secretions.

Results

- 1 child progressed to full healing
- 1 infant demonstrated initial healing but died from complications of HJEB aged 13 months
- 2 children are receiving treatment and their wounds are healing
- Timescale to healing: 3 to 12 months

Neither Flaminal® or clobetasol caused any discomfort on application or during wear time.

Discussion

Using very potent topical steroid ointment to the face of infants and children is unusual and there is risk of absorption resulting in Cushing's disease. Care must be taken to avoid contact with the eyes which could induce glaucoma. The treatment needs to be continued for several months in order to achieve healing. However, as the wounds were left uncovered much of the cream was rubbed off leaving reduced capacity for absorption. In addition, as the wounds reduced in size a smaller amount of topical steroid was needed. Following success using this combination of treatments this method will be prescribed at the onset of the development of facial wounds.

It is the author's opinion that criticism of this off-licence treatment can be balanced by marked improvement in quality of life of both the child and their family.

Conclusion

The combination of Flaminal® and potent topical steroid ointment has proved to be a valuable tool in management of facial lesions in those with HJEB. Although life expectancy will not be improved, healing these wounds reduces pain and also reduces unkind comments and accusations of abuse from the general public.

Case study

James is a 3 year old boy with Herlitz junctional EB. He developed lesions on his napkin area and around his umbilicus shortly after birth. Over the following few weeks his finger and toe nails were shed and as expected with this type of EB the nail beds remained open. Aged one year, James developed acute respiratory obstruction and needed emergency surgery to create a permanent tracheostomy.

Figure 1. James aged 1 year



James had a large wound extending over his face which suffered continual trauma from rubbing and this was made worse when he resisted suction from his tracheostomy. The wound was encrusted with food, appeared critically colonised and had an offensive odour. *Staphylococcus aureus* and *Pseudomonas* were cultured on wound swabs but not treated with systemic antibiotic therapy. The extent of the wound meant it pulled on the skin below his eyes causing ectropion which resulted in reduced tear film production and subsequent repeated painful corneal abrasions.

Figure 2. 6 months post-treatment with Flaminal® and clobetasol



James is a bright inquisitive child who loved to go out but his parents found it difficult to cope with the unkind comments they received when taking him anywhere apart from hospital visits and attending respite at the local children's hospice. Cleansing of the wound was attempted but resisted greatly by James which led to further damage to his fragile skin and by crying he increased the secretions from his tracheostomy and required additional suction.

Flaminal® Forte was applied daily to the wound in a thick layer. Although initially fearful of his face being touched James allowed this to continue once he realised it did not sting. Areas of crusting received a daily application of Flaminal® Hydro which was equally well tolerated. After 7 days application of Flaminal® the wound appeared much cleaner and odour was no longer a problem.

Clobetasol propionate ointment was then commenced daily in a thin layer. Flaminal® was continued for several more weeks 12 hours after application of clobetasol and then stopped and clobetasol continued daily. However after a few days the wound showed signs of colonisation and Flaminal® was restarted.

The over-granulation tissue rapidly subsided and the wound slowly reduced in size. The distressing complication of ectropion was alleviated and James is able to close his eyes fully. He has not suffered any more corneal abrasions. Blood pressure was monitored due to the possible absorption of topical steroid but remained within normal limits.

Figure 3. James aged 3 years



After a year of treatment with Flaminal® and clobetasol James's facial wound healed completely and has remained so. James is now three years old and attends a nursery school for children with special needs where he communicates using an iPad® as he is unable to vocalise. His parents are able to take him to the park and other public places without receiving verbal abuse.