MANAGEMENT OF CHRONIC FACIAL WOUNDS IN INFANTS AND CHILDREN WITH HERLITZ JUNCTIONAL EPIDERMOLYSIS BULLOSA (HJE'B)

Jackie Denyer

Clinical Nurse Specialist Epidermolysis Bullosa (Paediatric), Great Ormond Street Hospital for Children NHS Foundation Trust, London and Debra UK. jackie.denyer@gosh.nhs.uk

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 disabling 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 severe form, death in early infancy. Whilst work is progressing towards stem cell and other therapies, at the very least, factors considered were the requirements of the child and family as well as the need for therapy to:

- Be atraumatic
- Reduce over-granulation tissue
- 2 children are receiving treatment and their wounds are healing
- 1 child progressed to full healing
- 2 children are receiving treatment and their wounds are healing
- Timelapse to healing: 3-12 months
- Neither Flaminal® or cloabetasol 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 Cushings’ 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 washed off and the child was unable to wear glasses, and by crying he increased the secretions from his tracheostomy 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.

Case Study
James is a 3 year old boy with Herlitz junctional EB. He developed lesions on his lower body and around his umbilicus shortly after birth. Over the following few weeks his fingers 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.

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. S. aureus and Pseudomonas were cultured on wound swabs but not treated with systemic antibiotic therapy. The extent of the wound meant it pulled on his skin below his eyes causing ectropion which resulted in reduced tear film production and subsequent dry eye symptoms.

After a year of treatment with Flaminal® and cloabetasol James’ 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.