

The Use of Atraumatic Wound Dressings in Epidermolysis Bullosa

Authors: Jacqueline Denyer, Nurse Consultant, Epidermolysis Bullosa (Paediatric), Great Ormond Street Hospital, London & DeBRA UK

INTRODUCTION.

Epidermolysis bullosa (EB) is a rare, genetically determined skin disorder in which the skin and mucous membranes are extraordinarily fragile. Blisters and wounds arise readily from minimal everyday friction and trauma. There are several distinct types of EB, each with a different prognosis varying from minor blistering on the feet to death in early infancy (Fine et al. 2000). Work continues towards gene therapy, but at present treatment is symptomatic. Severely affected children benefit from multi disciplinary care at a specialised centre.

Neonates are particularly vulnerable and transportation is hazardous often resulting in extensive skin loss (Denyer 2000). An outreach service by specialist nurses is provided to neonatal units, with the infant then being seen at the specialised centre once inter uterine and birth damage have healed. Careful selection of dressing materials and specialised handling techniques together with modification of monitoring and screening should be implemented as soon as possible after the birth of an affected infant. Dressing technique must be done in a way to minimise deformity from contractural scarring in those with dystrophic EB.

A lack of widespread knowledge of the condition and its management lead to well-meant, but inappropriate care resulting in distress to the child, family and medical practitioners.

Case Study

Baby C was born via a normal delivery following an uncomplicated pregnancy, she is the second child of unrelated parents.

This baby was noted to have absence of skin over both hands and both feet, (Figs 1 & 2) these were wrapped in Vaseline gauze. Gentle handling resulted in small skin tears on her thighs (Fig 3) and drying her face with a soft cloth caused her to develop painful corneal abrasions (Fig 4). A cannula was sited into her foot to administer intra venous antibiotic therapy. The cannula was secured with adhesive tape and film (Fig 5). A layer of Vaseline gauze was added and the foot wrapped in an open weave bandage. The skin over her heel was intact and this was left exposed, but the skin was lost almost immediately when she kicked and degloved the area on the soft sheet (Fig 6). Sucking on a soft teat resulted in extensive blistering to her lips and oral mucosa.

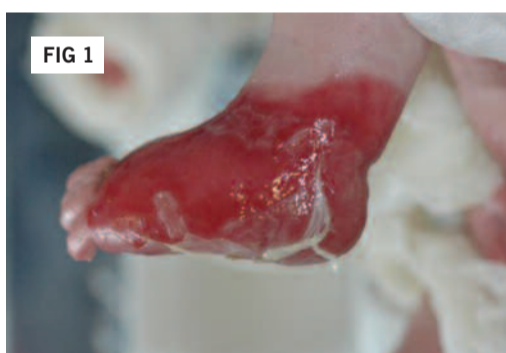
A clinical diagnosis of dystrophic epidermolysis bullosa was made and confirmed by skin biopsy. Unfortunately during her treatment the Vaseline gauze had dried out and firmly adhered to the wounds as had the open weave bandage (Fig 7). Oral morphine was given and removal of the gauze facilitated by liberal application of 50% white soft paraffin, 50% liquid paraffin (50/50), however, some skin tearing still occurred. Although the use of 50/50 was essential it made redressing the wounds extremely difficult.

Wounds were then dressed using soft silicone dressings. Her feet were dressed with Mepitel and covered with Mepilex, to provide protection from kicking (in our experience Mepilex used as a sole wound dressing can sometimes adhere to wounds in those with epidermolysis bullosa). Her hands were dressed with Mepitel, the fingers wrapped individually to prevent fusion and covered with Mepilex Transfer.

Dressings were changed twice weekly and removal of all the soft silicone products were atraumatic. Community nurses attended each dressing change to ensure competency following discharge. Healing was achieved after 21 days following which she was discharged home (Figs 8, 9, 10), although her skin will remain fragile and will need to be protected. Mepilex Transfer is used to protect healed areas or areas of skin adjacent to scabbed lesions which can rub on the affected skin (Figs 11, 12).

Pathology – Dystrophic EB may be inherited as an autosomal dominant or autosomal recessive trait. Carrier frequency for recessive DEB is estimated at 1 in 350 (Pfindner E 2001). Mutations are present in the gene for type V11 collagen, the major component of anchoring fibrils (Burgeson 1993). The most severe form of recessive DEB, Hallopeau-Siemens have little or none of this essential protein leaving the skin deficient in anchoring fibrils. Friction, a knock or shearing forces cause the skin to blister or shear away leaving an open wound. Healing results with mutilating scarring leading to loss of mobility and pseudosyndactyl of hands and feet. Internal blistering and subsequent scarring lead to microstomia and dysphagia. A later complication is the development of squamous cell carcinomas on areas of recurrent ulceration and scarring. Screening for malignancy should begin in the teenage years (Ayman 2002).

Protection of vulnerable areas is essential to minimise wounds and subsequent scarring.



CONCLUSION

Inappropriate dressing materials caused skin tearing which exacerbated skin loss caused by inter uterine movements and delivery. Use of atraumatic dressings resulted in rapid healing of all wounds. All scarred areas remain fragile and require protection.

Dressings with Safetac soft silicone products have proved to be atraumatic on removal for the majority of those with EB, and remain our initial choice of dressing for neonates with dystrophic epidermolysis bullosa. Safe fixation of cannulae can be achieved using Mepitac in place of adhesive tape.

References

- Fine J., Eady R., Bauer E., et al. revised classification system for inherited epidermolysis bullosa; report of the second international Consensus Meeting on diagnosis and classification of epidermolysis bullosa. J Am Acad Dermatol 2000;42:1051-66
- Denyer JE., management of severe blistering disorders. Semin Neonatal 2000; 5: 321-4
- Pfindner E., Utito J., Fine JD., Epidermolysis bullosa carrier frequencies in the US population. J Invest Dermatol 2001; 116:483-4
- Burgeson RE., type V11 collagen, anchoring fibrils and epidermolysis bullosa. J invest dermatol 1993; 101:252-
- Ayman T., Yerebakan O., Ciftcioglu MA. Et al. A 13 year old girl with recessive dystrophic epidermolysis bullosa presenting with squamous cell carcinoma. Pediatr Dermatol 2002; 19:436