The use of HydroBalance Cellulose Based Dressings (HCBD) in the management of children with epidermolysis bullosa.

Jacqueline Denyer, EB Nurse Consultant (Paediatric), Great Ormond Street Hospital and DebRA UK, London.
email: jackie.denyer@debra.org.uk

Case Study 1

**Introduction**

This paper describes the use of HCBD for infants and children with the rare genetic skin fragility disorder, epidermolysis bullosa (EB). The term epidermolysis bullosa represents several disorders each having a wide range of severity (Fine JD 2008). The condition is characterised by an extreme fragility of the skin and mucus membranes and a susceptibility of these to blister or break down in response to minimal everyday friction and trauma. There are several types of EB, determined by the affected protein and the specific gene mutation. In its mildest form, EB causes painful blistering to the hands and feet, limiting function and mobility. Other types of EB can lead to death in early infancy or progressive disability resulting from contractual scarring of the skin and mucus membranes.

**Method**

Infants and children with all types of EB were selected for the study. Criteria included those suffering pain and itching and those with chronic wounds.

Children with EB simplex experience painful blistering of their hands and feet, especially during hot and humid conditions. The qualities we were looking for in the dressing were to be atraumatic to both the blister site and the surrounding skin, to reduce heat and to provide pain relief (Weiner M 2004). For these children a HCBD, Suprasorb® X was selected. Suprasorb® X has the ability to absorb and donate moisture according to the requirements of the wound.

Children and infants affected by the more severe forms (junctional and dystrophic EB) develop chronic wounds which are frequently critically colonised and infected. These children require an antimicrobial agent to control the bio burden and therefore Suprasorb® X+PHMB (Polyhexamethylene biguanide) was selected. In addition to the properties of Suprasorb® X, a HCBD dressing, Suprasorb® X+PHMB offers a safe antimicrobial for the treatment of critically colonised and infected wounds (Miano et al., 2007). Again the requirement for the dressing was to be atraumatic, comfortable, offer relief from pain and, in addition, to reduce pruritis. Pruritis is a major factor in these types of EB and leads to extensive damage from scratching. Traditional measures for control of pruritis such as use of antihistamines often have minimal effect.

**Case Study 1**

L. (EBS localised)

EBS localised results from missense mutations in the genes encoding keratines 5 and 14. Skin fragility is largely limited to the hands and feet, where painful blisters arise spontaneously and in response to friction. This type of EB is exacerbated by heat and humidity. Patients with the localised and generalised forms of EBS almost always have an extensive family history of the condition.

Blistering on the feet lead to pain and reduced mobility. Historically children with EBs localised find dressings can make the blistering worse, as heat can be generated under the dressing, especially when foams are used. The edges of dressings can also create blisters.

Many choose not to use dressings, leading to further problems with adherence to socks which need to be soaked free of debris accumulating in the wound.

L is 10 years old. She has EBSS inherited from her father. She has widespread blistering over her hands and feet. Her school attendance is poor and she is unable to wear shoes. In the house she crawls and outside she uses a wheelchair when walking more than a few metres. L is overweight, which may be partially due to her limited mobility.

L had used very few dressings. Her management was to lance the blisters with a hypodermic needle and to soak her feet in cold water to reduce the heat.

She had tried many dressings in the past but found them all to increase her discomfort - either by increasing the rate of blistering due to a rise in temperature, or movement causing the dressing to crease and be uncomfortable.

With great suspicion, L agreed to try Suprasorb® X on a blistered area over her heel. She complained of the cold feeling, but then almost immediately said it felt better. She then applied Suprasorb® X to all the blisters on her feet. The dressing was retained using a cohesive stretch bandage.

In order to maintain the soothing effect from the cool dressing, it needed to be changed once or twice a day depending on the environmental temperature.

L continues to use the dressings and her mobility is slightly improved. She is also able to wear slippers and soft shoes for short periods at a time.

**Case Study 2**

A. (Non-Herlitz JEB)

Junctional forms of EB are recessively inherited and are characterised by mechanically induced blistering occurring within the basement membrane level of the lamina lucida. Mutations in genes encoding components of the hemidesmosome anchoring filament complex affect dermal epidermal adhesion. Type XV11 collagen or laminin 332 proteins are defective in those with junctional EB.

A is 18 months old. She is severely affected by Non-Herlitz JEB and has extensive wounds mainly over her arms and legs; the majority of these have persisted since birth.

Her current dressings were offering protection from external trauma and providing a good environment to facilitate healing. Whilst we strive to heal wounds in all our patients, in some cases the overriding gene defect makes this a difficult challenge.

One of the main barriers to healing was repeated trauma caused by a scratching and rubbing her skin in response to pruritis. Many topical and systemic agents had been tried without much success.

Due to anticipatory fear and pain, dressings are changed only 2-3 times per week. To prevent adherence to the wound lipoidosiloid dressings were applied, followed by Suprasorb® X+PHMB.

This regime was followed by a foam for absorption and padding. The foam drew moisture from the Suprasorb® X+PHMB, which reduced its effectiveness and therefore an additional layer of lipoidosiloid was applied before the foam dressing. Although far from ideal, layering dressings is often necessary in patients with epidermolysis bullosa as the ideal dressing has yet to be manufactured. This method enabled the Suprasorb® X+PHMB to remain moist until the next dressing change.

As parents reported an immediate and sustained reduction in itching and a subsequent improvement in the wound has been observed.

**Discussion**

The use of Suprasorb® X and Suprasorb® X+PHMB in the management of infants and children with all types of EB has facilitated patient comfort and shown a significant reduction in both pain and pruritis.

**Conclusion**

Chronic wounds in epidermolysis bullosa patients are notoriously difficult to heal due to the underlying genetic defect. Reduction in pain and itch can lead to significant improvement in the quality of life of these patients, demonstrated by improvement of mood in those severely affected and greater mobility in those with milder forms.

**References**


Weiner M. Patch Management in Epidermolysis Bullosa on intractable problems. Poster/Podiatry Management, August 2004, vol 5, no 8, pp 1-14