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, London.

The term epidermolysis bullosa represents several disorders each having a wide range of severity.

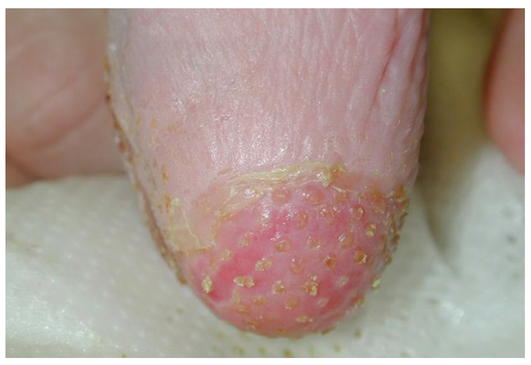
The common factor is an extreme fragility of the skin and mucous membranes and a susceptibility of these to blister or break down in response to minimal everyday friction and trauma. There are several types of EB, determined by the affected protein and the specific gene mutation.

Severe types of EB can lead to death in early infancy (Herlitz junctional EB) or progressive disability resulting from contractured scarring of the skin and mucous membranes (severe generalised dystrophic EB).

In its mildest form, localised epidermolysis bullosa simplex (EBS) painful blistering is largely limited to the hands and feet, causing pain and reduction in mobility and hand function. The development of chronic wounds is uncommon in this group. EBS localised results from missense mutations in the genes encoding keratins 5 and 14. These keratins provide the scaffolding to the cells within the skin, and reduction or absence of these causes the cell wall to rupture in response to minimal friction and spontaneously in the heat. Skin fragility is largely limited to the hands and feet where painful blisters arise both 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 often have extensive family histories of the condition, which can make it difficult for new dressing materials to be accepted as there is often a stoic approach towards the painful blisters and the attitude that “nothing can help”.

Many dressings are suitable for those with more severe types of EB, and trials of new products are constantly in place.



However, children with localised EBS have a hidden disability and historically have not benefitted from many products suitable for those with other forms of EB. They frequently find that dressings can make the blistering worse, as heat can be generated under the dressing particularly when foams are used. The edges of dressings can also create blisters. Dressings with an open mesh can lead to a blister forming in each hole of the mesh.

Many children have abandoned dressing use altogether, leading to further problems when the unprotected blisters rupture and adhere to clothing and bedding causing additional pain and trauma.

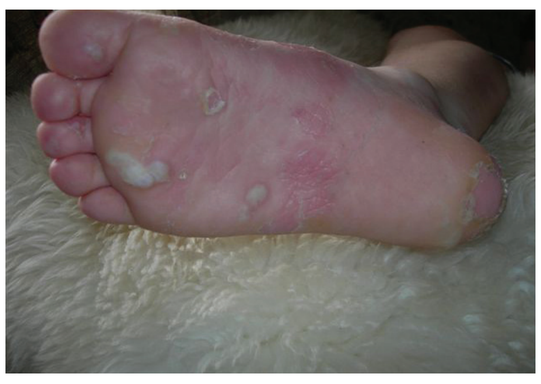
Method

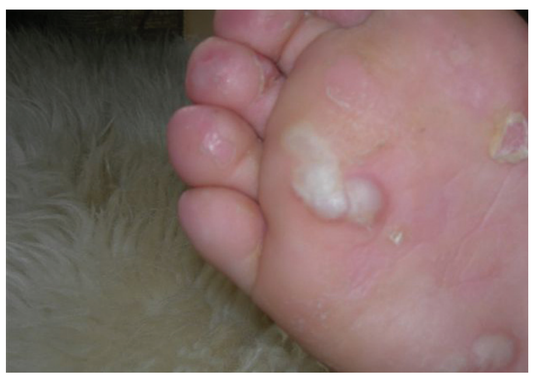
Children with localised EBS who had blistering to their feet were selected for the study. Localised EBS is generally more troublesome in children who are walking; therefore infants under one year of age were excluded from this initial study.

The qualities we were looking for in the dressing were to be atraumatic to both the blister site and the surrounding skin, to be easy to apply and remove, to reduce heat and to provide comfort and pain relief.

Case Study

L (EBS localised)

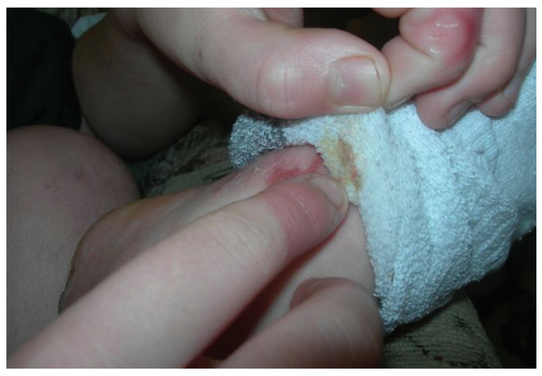


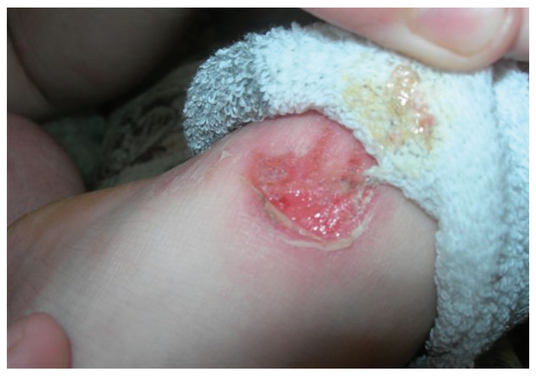


L is 10 years old. She has EBS inherited from her father. She has widespread blistering over her hands and feet. Her school attendance is poor as her sleep pattern is often disturbed by pain.

She is unable to wear shoes and walks in 2 pairs of socks even when outside. 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 father uses no dressings at all as he had been given unsuitable products as a child and is now wary of trying anything.

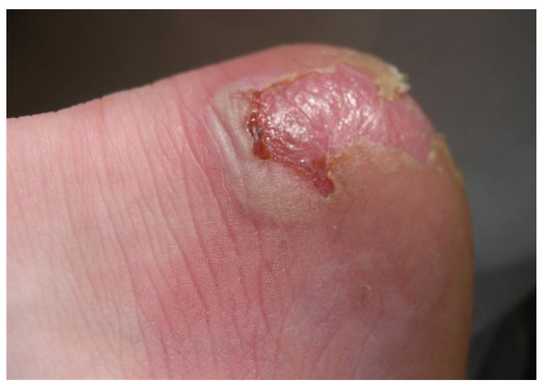


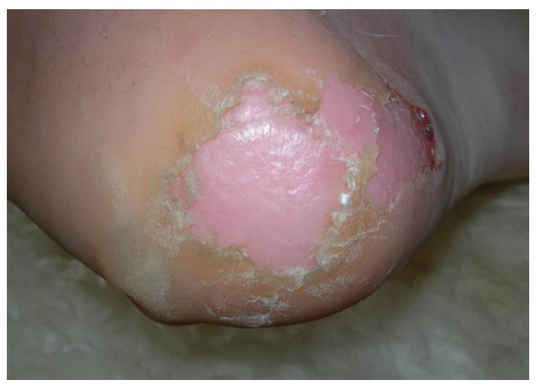


L’s management was to lance the blisters with a hypodermic needle and to soak her feet in cold water to reduce the heat. She often woke during the night due to pain from the blisters and needed to cool her feet in a bowl of water. Her pain management consisted of paracetamol and ibuprofen “on bad days” which was sub optimal, but L and her parents are reluctant to try analgesic interventions for moderate or severe pain or to address neuropathic pain.

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.

After great suspicion L agreed to try HCBD on a blistered area over her heel. She complained of the cold feeling but then almost immediately said it felt better. Prior to applying the dressing her pain on the Wong & Baker scale was recorded as 8 and this reduced to 4 after 20 minutes from the dressing being applied.





The pain relieving effect continued until the dressing became dry and was replaced. L experienced some adherence of the dressing and it was soaked off. No damage was sustained to the blister site or surrounding skin, but L was anxious and afraid. Lipidocolloid dressings were subsequently used as a wound contact layer and the HCBD applied over the top of this. This enabled easy and atraumatic removal of both dressings and L was confident with this combination.

She then applied Lipidocolloid and HCBD to all the blisters on her feet. The dressings were retained using a cohesive stretch bandage. In order to maintain the soothing effect from the cool dressing L needs to change these once or twice a day, depending on the environmental temperature.

L continues to use the dressings and her mobility is slightly improved. When the dressings are in place she is able to wear slippers and soft shoes for short periods at a time. At the first sign of any infection she uses HCBD containing PHMB which has avoided the need for oral antibiotics on the majority of occasions. She is also able to wear slippers and soft shoes for short periods at a time.

Discussion

The use of HCBD in the management of children with EBS has facilitated patient comfort and shown a significant reduction in pain with a corresponding improvement in mobility.

Conclusion

EBS is a difficult condition to manage and one which causes a hidden disability as the most troublesome blisters occur on the feet.

Reduction in pain can lead to significant improvement in the quality of life of these patients, demonstrated by improvement of mood and greater mobility. HCBD has proved to be effective management for pain in children with EBS.

References:

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* Suprasorb® X - HydroBalance dressing - Activa Healthcare, UK.
Suprasorb® X+PHMB - HydroBalance dressing with PHMB - Activa Healthcare, UK.

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