Managing pain in children with epidermolysis bullosa

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Children with this rare skin disorder often experience pain during dressing changes.

**In this article...**

- Different types of epidermolysis bullosa
- How pain affects children with EB
- Managing pain during dressing changes

**5 key points**

1. The term epidermolysis bullosa encompasses a group of genetically determined rare skin disorders.
2. There are four main categories of the condition.
3. People severely affected have many non-cutaneous complications.
4. Pain is a common complaint from children with all types of EB.
5. One of the greatest challenges is controlling pain during dressing changes.

The term epidermolysis bullosa (EB) encompasses a group of genetically determined rare skin disorders in which the skin and mucous membranes are fragile, leading to the ready development of blisters and wounds following relatively minimal friction and trauma.

A key structure in the skin is the dermal-epidermal basement membrane zone, which comprises intracellular and extracellular matrix proteins and structures that give the skin its tensile strength. Hereditary EB is a group of basement membrane zone disorders (Myung et al, 2010).

The condition is classified into four categories: EB simplex; junctional EB; dystrophic EB; and Kindler syndrome (Fine et al, 2008). Each has subtypes, and each of these has different effects and outcomes.

For example, those with localised EB simplex experience simple blistering affecting the hands and feet only, while children with severe generalised dystrophic EB have increasing disability as a result of contractural scarring. In Herlitz junctional EB, a very limited life expectancy is usual following respiratory distress from internal blistering and profound failure to thrive.

Appearance at birth is not an indication of the type of EB and diagnosis is made by analysing a shave skin biopsy (Fine, 1994).

Those severely affected have many non-cutaneous complications as EB can involve multiple systems including gastrointestinal, musculoskeletal, ophthalmological, respiratory and genitourinary (Goldschneider and Lucky, 2010; Van Scheppingen et al, 2008).

**Pain**

Pain is a common complaint from children with all types of EB and, in those with severe types, the sources of pain are multifactorial (Weiner, 2004). Children with localised EB simplex describe pain from blister sites and areas of skin loss; those with more severe types of EB have pain from wounds and blisters, with additional pain from lesions on mucous membranes, the cornea and from non-cutaneous complications.

Many children with severe generalised dystrophic EB develop oesophageal strictures which require regular dilatation (Azizkhan et al, 2006). Some also need supplementary nutrition via a gastrostomy (Haynes, 2007).

Corneal abrasions occur with all severe forms of EB following trauma or can result simply from the child opening their eyes in the morning and the eyelid tearing the cornea. Reduced tear film and blepharitis increase the risk of abrasions but lubricating drops can be difficult to administer as even minimal force used to open the eye can damage the skin.
Discussion

Non-cutaneous sources of pain include: osteoporosis and osteopenia with associated risk of fractures (Fewtrell et al, 2006); gastro-oesophageal reflux; colitis (Freeman et al, 2008); and contractures. Additionally, procedural, acute, chronic and neuropathic pain make managing severely affected children extremely complicated (Howard et al, 2008).

Pain during dressing changes
One of the greatest challenges is controlling pain during dressing changes (Price, 2005). Dressing changes continue throughout life and are a source of stress, with many children procrastinating and finding excuses to delay or avoid the procedure.

Undoubtedly, modern atraumatic wound products have alleviated much of the pain associated with adherent dressings but, even so, this remains a pain-filled experience. Dressing changes can take several hours and, despite dressings being designed to remain in place for several days, more frequent changes may be needed to control odour, reduce colonisation and prevent maceration (Mellerio et al, 2007). Severely affected infants often lose skin in utero, which is compounded by the trauma of delivery. Those born by Caesarean section may be less severely damaged at birth but often have stripping of the oral mucosa following oropharyngeal trauma of delivery. Those born by Caesarean section may be less severely damaged by their general health – premature infants require minimal handling so dressings placed between his toes in an attempt to prevent digital fusion and the wound was covered with polymeric membrane. The simplicty of this dressing regimen reduced distress as there was no adherence and the pre-cut dressing shapes were easy to apply. The time taken to complete dressing changes greatly reduced.

Analgesia was reduced to regular paracetamol and morphine before dressing changes and following any new injury. Henry’s parents were relieved that he was more comfortable and were able to hold and feed him more confidently and allow easier dressing changes.

Dressings
Selecting dressings in neonates is dictated by their general health – premature infants require minimal handling so dressings...
that can remain in place for several days should be selected wherever possible.

All additional care should be coordinated to coincide with dressing changes to allow infants to rest between procedures. Apnoeic spells can result from prolonged handling, so dressing changes for different parts of the body may need to be rotated on different days.

Using templates to cut dressings in advance will speed up the procedure, minimizing damage from the time the infant is exposed without protective dressings and ensuring dressings are the appropriate size and shape.

Dressings that have a cooling effect, such as hydrogels, may not be suitable for extensive use in small babies because of the risk of inducing hypothermia.

Bathing should be delayed until prenatal and birth damage has healed as it is not possible to prevent further damage when all dressings are removed at any one time (Denyer and Stevens, 2009). Limb-by-limb dressing changes should be carried out using polymeric membrane dressings containing the cleanser surfactant F68, which provides continual wound management. Polymeric membrane dressings can be placed directly onto both the wound and the surrounding intact skin, making dressing changes faster and less traumatic (Denyer et al, 2009).

Older children with severe forms of EB develop chronic wounds and dressing changes can take several hours each day. Managing procedural pain

Pain reduction at dressing changes is complex and requires constant evaluation and adjustment of pharmacological prescriptions and non-pharmacological recommendations (Power et al, 2007), together with changes in dressing management.

Unusual measures such as adding salt to the bath to make the water isotonic (isotonic saline is 9g of salt/l of water) has anecdotal support from many families of patients with EB (DEBRA, undated).

When children have very painful wounds and oral analgesia is ineffective, topical morphine has proved beneficial (Watterson et al, 2004).

Wounds in children with severe EB can be acute or chronic and most have a combination of both. Dressing management is tailored to each wound although the principles of non-adherence, comfort and control of exudate and odour apply to all.

Correct choice of anatraumatic primary wound dressing where necessary is essential; our department recommends soft silicone mesh or lipido-colloid. Other soft silicone dressings including foams are designed to be placed directly onto the wound bed but these can become adherent in those with severe forms of EB.

Dressings or clothing that have become adherent can be removed with minimal trauma by using silicone medical adhesive remover (Denyer, 2011). Non-silicone based removers may sting when in contact with broken skin. A sudden increase in pain together with increased skin breakdown may be caused by infection, with strep-tococcal infection being particularly painful.

Itch is an intractable problem and the resulting scratching can destroy intact skin rapidly. Oral medication such as omdansetron, antihistamines, tricyclic antidepressants and gabapentin may help to reduce itch (Goldschneider and Lucky, 2010). Dressings with a cooling effect such as sheet hydrogels or hydrocellulose may help reduce irritation.

Practitioners should follow this advice for dressing changes:

» Close windows and doors, and switch off fans to reduce pain to the wound bed from air currents;

» Use pre-cut dressings if possible to reduce the time of wound exposure and shorten the duration of dressing changes;

» Allow children some control, for example, by removing dressings themselves;

» Have silicone medical adhesive remover available in case of adherence;

» Take photographs (with permission) at dressing changes to avoid repeated removal of dressings when staff visit at other times.

Conclusion

Children with EB have wide variations in their need for pain management. This appears to be related to severity but is also influenced by lifestyle, personality and individual pain thresholds.

Pain from broken skin is one of the main sources of complaint with procedural pain being difficult to manage in view of the lifelong need for dressing changes.

DEBRA is the charity for families affected by EB. It provides information, practical help and professional advice through its nursing and social care teams. Visit the website at www.debra.org.uk.

References


DEBRA (undated) Pain-Free Washing. tinyurl.com/ DEBRA-washing


