

Bathing in epidermolysis bullosa: benefit over trauma?

Epidermolysis bullosa (EB) is the term given to a large group of genetically determined skin disorders in which the common factor is fragility of the skin and mucous membranes (Lin and Carter, 1992). There is a wide range of severity, varying between mild discomfort and death in early infancy. The presence of multiple wounds of varying gestation make management difficult and care must be regularly modified. Non-cutaneous complications further compromise well-being. The aim of this paper is to discuss the practice of bathing for severely affected infants and children and whether this is always appropriate.

Jacqueline Denyer, Louise Stevens

KEY WORDS

Epidermolysis bullosa (EB)
Bathing
Pain management
Quality of life

Epidermolysis bullosa (EB) is defined as a rare group of inherited skin disorders (Denyer, 2006). The most recent classification (Fine et al, 2008) describes four categories of EB depending on the level of cleavage within the dermal/epidermal junction:

▶ EB simplex (EBS). This has several subtypes, the vast majority of which are inherited in an autosomal dominant pattern. Most of the subtypes are caused by mutations in the genes encoding for keratins 5 and 14. In the most common type of EBS (localised EBS), blistering is

limited largely to the hands and feet causing pain and reduced mobility and function, but non-cutaneous complications are rare. EBS Dowling Meara and Generalised EBS cause widespread blistering and in Dowling Meara blistering of the larynx can lead to morbidity, or, in rare cases, mortality in infancy

▶ Junctional EB (JEB) — this has two major subtypes, both of which are recessively inherited. Defects for junctional EB lie within laminin-322. Herlitz JEB is the more serious type, and blistering of the larynx together with poor nutrient absorption generally leads to death within the first two years. Non-Herlitz JEB is a milder phenotype, but complications include chronic wound formation, alopecia and urinary tract anomalies

▶ Dystrophic EB (DEB) — this is caused by mutations in the gene COL7A which encodes for type VII-collagen. It may be dominant or recessively inherited. There is a wide scale of severity within the spectrum of DEB. In its severe form (severe generalised dystrophic EB), there is complete loss of type VII collagen with reduced or absent anchoring fibrils. These patients develop atrophic scarring leading to pseudosyndactyly, oesophageal strictures, growth retardation, eye involvement, anaemia and a greatly increased risk of squamous cell carcinoma in adulthood

▶ Kindler syndrome — this is a newer addition to the EB classification. This is characterised by loss of kindlin-1. The level of blister formation may involve multiple levels of the skin and is an inherited mechano-bullous disease.

Within each of these categories there are subtypes that differ in their clinical effects. Mutations have now been found in more than 10 genes that encode the structural proteins within keratinocytes or muco-cutaneous basement membranes (Lanschuetzer, 2009).

The common factor in all types of EB is the tendency for skin and mucous membranes to blister, or shear away in response to minimal everyday friction and trauma. EB varies in its effects between simple blistering to the hands and feet, particularly in warm weather; to death in early infancy from a combination of laryngeal disease and failure to thrive (Denyer, 2007).

Those severely affected suffer recurrent blistering and skin loss. The tendency to develop chronic wounds results from the underlying gene defect, nutritional deficiencies and repeated infection and trauma (Denyer, 2010). The underlying principle for the management of lesions is to apply an atraumatic dressing to prevent pain and bleeding on removal (Dystrophic Epidermolysis

Jacqueline Denyer is a Clinical Nurse Specialist for children with EB, Great Ormond Street Hospital, London; Louise Stevens is an EB Clinical Nurse Consultant (wound care), Brightsky, Sydney, Australia. Brightsky Australia has been selected by the Australian Government to administer and supply dressings on the inaugural Australian Epidermolysis Bullosa Dressing Scheme which commenced in January 2010. Louise previously worked at Sydney Children's Hospital as Clinical Nurse Consultant for EB

Bullosa Research Association [DebRA], 2002). However, management must be tailored to both the type of EB and to the specific characteristics of the wound. Personal preference, lifestyle and availability of both dressing materials and carer time also play a part in selection.

Approximately 1000 people suffer from EB in Australia (DebRA, Australia 2002). Estimated numbers in the UK exceed 5000. Those severely affected require specialised management throughout life, as EB can involve multiple organ systems including gastrointestinal, musculoskeletal, ophthalmological, respiratory and genitourinary.

This pathology has important implications on the psychological, physical and social well-being of the child and the family. Good wound care is an essential part of the management of EB, but no single approach to managing wounds has proved totally effective (Caldwell et al, 1992). Indeed, management of the various types of EB requires a different approach, as wounds may be specific to the type of EB and experience of a large caseload will identify the most appropriate type of dressing. Dressings provide a barrier between the patient and the environment, help to prevent infection, support wound healing and relieve pain. There are a wide variety of primary and secondary wound dressings, which are described as non-adherent and effective in promoting healing. Dressings are also constantly evolving and so management of patients with EB changes. Cleansing and dressing patients in the management of EB, however, is a contentious issue and differs both worldwide and according to local policy.

A widely recognised practice is the containment of blisters by lancing them to limit their spread and therefore reduce the amount of tissue damage. This is generally done by lancing the blister at its lowest point with a hypodermic needle and gently expelling the fluid. In the authors' experience, application of simple cornflour will also help to dry up the area and reduce additional blister formation.

Cornflour is hygroscopic and therefore safe if in contact with mucous

membranes. This practice has been employed for over 10 years without any reported adverse effects.

EB is a very individual disease and as the patients grow older and more autonomous, they often come to their own decisions as to what cleansing and dressing regimen suits them. However, at any age there is no doubt that the cleansing and bathing of EB-related wounds can be a painful and time-consuming process. There is a wide difference of opinion as to the best management of these wounds, and how to combat critical colonisation.

In the authors' experience, without the use of an antimicrobial agent, wounds become recalcitrant and repeated infection requires treatment with antibiotic therapy, predisposing to the development of resistant organisms.

There are some recommendations for short-term Hibiclens baths in an attempt to reduce Gram-positive organisms, or vinegar soaks for control of Gram-negative organisms such as pseudomonas (Schober-Flores, 2009). Others believe that caution should be exercised with ongoing use of Hibiclens, as chlorhexidine may be potentially neurotoxic (DebRA UK, 2002).

A recent clinical study suggested that bleach combined with mucipin reduced rates of infection in atopic dermatitis (Huang et al, 2009). This theory has been advocated in some centres for the management of patients with EB. Salt baths have proved effective and popular with other patients; presumably the osmotic effect prevents pain.

Recommendations for antimicrobial agents for the authors' patients include dressings using the principle of hydrophobic interaction (Cutimed® Sorbact®, BSN Medical), with bacteria becoming irreversibly bound to the coating of a fatty acid derivative (dialkylcarbamoylechloride [DACC]) (Denyer, 2009). Honey ointments and impregnated dressings are successful in reducing the bacterial load, although the osmotic pull makes these too painful to use for some children (Hon, 2005).

Dressings with polyhexamide biguanide (PHMB) are also successful in reducing infective episodes (Kingsley et al, 2009).

Good dressing technique is probably one of the most important factors in the pain management of patients with EB (Herod et al, 2002; Meaume et al, 2004).

In the UK, DebRA nurses do not advocate bathing in the neonatal period until prenatal and birth-induced wounds have healed. The authors have found the practice of bathing neonates with extensive wounds to be traumatic both in terms of pain and the additional damage that results from being unable to protect the infant in the absence of dressings.

Assessing the need for pain relief before any dressing change is a priority. The paediatric DebRA UK nurses have found the best dressing change practice for infants with EB is to attend to their limbs one by one. This avoids pain and trauma and further damage to the skin from handling and kicking during a full bath. It has been carried out for many years by experienced EB nurses, as in line with best practice.

Some practitioners advocate that in hotter climates bathing should be practiced more regularly to avoid the risk of a culture medium under the wound dressing (Kopecki et al, 2009). However, in the authors' experience, using dressings with suitable antimicrobial properties is sufficient to control the level of bacteria. Many modern non-adherent dressings recommended for use in EB are designed to remain in place for up to seven days. Leaving the wound undisturbed allows re-epithelialisation to occur and avoids re-igniting the inflammatory phase, which causes bleeding and trauma and delays healing. However, in the authors' experience, leaving dressings *in situ* for that length of time results in malodorous wounds which quickly reach levels of critical colonisation.

An important consideration in neonates and young infants is the bonding between the child and parents. Bonding and attachment is critical for any infant, but perhaps more so in the case of a child whose quality of life is largely dependent

on excellent parenting. Bonding may already be compromised by the minimal handling restrictions imposed upon fragile infants, and malodorous wounds can only further inhibit the process.

If tolerated, honey dressings can successfully combat malodour (Hampton, 2008).

Polymeric membrane dressings (PolyMem, Ferris) contain a mild non-toxic cleanser of F68 surfactant which reduces the bioburden and optimises conditions for wound healing. In the authors' experience, the dressing becomes wet and strikethrough occurs rapidly, necessitating the need for frequent dressing changes. This can be reduced by additionally using PolyMem® Wic (Aspen Medical) underneath to prevent maceration. Although a polymeric membrane requires more frequent changes initially, the duration of dressing change is reduced by the avoidance of an additional primary and secondary dressing. This dressing also contains glycerin which is reported to reduce overgranulation. Another quality of glycerin is its ability to prevent adhesion to the wound bed and the fragile surrounding skin. Studies have shown that this dressing is an effective way of cleansing the wound while improving the healing process (Rodeheaver, 1989; Denyer et al, 2009).

Case report I

Baby A was born by normal vaginal delivery at 38 weeks' gestation. Skin loss was noted at birth to the arms, hands, lower legs and feet. Erosions on the face, abdomen and buttocks were also present. EB was suspected and confirmed on skin biopsy to be non-Herlitz junctional. Baby A was transferred to Sydney Children's Hospital at 18 days of age.

His main problems were pain management, traumatic dressing changes, difficulty in handling, feeding/nutritional concerns and developmental issues.

At this centre wound care for babies with EB traditionally centres on daily bathing and dressing changes. A non-adherent primary dressing of soft silicone mesh and a secondary



Figure 1. Baby A's hand at eight weeks old when daily baths and dressing changes were being carried out.

dressing of soft silicone foam were the dressings of choice at this time. These were used for all infants regardless of type of EB. Despite oral analgesia of morphine and sedation with midazolam for dressing changes (Weiner, 2004), the baby was distressed during the 2–3-hour procedure, using scoring 8 and above on the FLACC (face, legs, activity, cry, consolability) scale (Lawrence et al, 1993; Merkel et al, 1997; available online at: www.childcancerpain.org/content/cfm?content=assess08).

The FLACC scale is a behaviour assessment tool for measurement of pain in infant and paediatric patients. It quantifies behaviours with scores ranging from 0 (no pain response behaviours) to 10 (most possible pain behaviours). Exposed wounds are more painful and the baby's discomfort was distressing to all those who cared for him. Studies highlight that inter-utero traumatised patients show a heightened sensitivity to pain (Jonsdottire and Kristjandottir, 2005).

It has been suggested that children's memories of early pain experiences may be related to exaggerated memories of pain over time (Noel et al, 2009).

Figure 1 demonstrates baby A's hand at eight weeks old when daily baths and

dressing changes were being carried out. Although there were some areas of healing, these were offset by areas of new skin loss and bleeding which occurred due to kicking and friction from handling and movement.

Research has demonstrated that maintaining a constant wound temperature of 37 degrees Celsius increases mitotic and phagocytic activity (Lock, 1980). It has also been suggested that long periods of exposure of wounds during dressing changes can reduce the temperature of the wound (Turner, 1995). Mitotic and phagocytic activity is reduced during this time and it can take up to three hours for activity to return to normal.

An infant's inability to thermoregulate is widely recognised. Exposure during lengthy dressing changes predisposes the infant to high hypothermia or places them at risk of the deleterious effects of cold stress (Johnston et al, 2003). Overhead heaters are not advocated as direct heat can encourage blister formation.

Baby A's trauma when being bathed was clear and this was also upsetting for the nursing staff. Dressings were time-consuming, which was posing a huge demand on the ward staff as the parents



Figure 2. Baby A's hand wound at three months old showing epithelial edging and minimal hypergranulation.



Figure 3. Baby A's hand wound at five months. The wound has shrunk in size, with epithelial migration.

had chosen not to be involved (Fine et al, 2005).

'Non-adherent' dressings did sometimes adhere to the skin, and daily changes meant disruption of re-epithelialisation. Due to the effects of the daily narcotic pain relief and sedative, baby A remained sleepy for long periods of time which made feeding problematic. Nutritional requirements are high in babies with EB as they are catabolic (Haynes, 2006). Supplementing him with nasogastric feeds became necessary due

to the low volume taken orally, imposing an additional challenge from the difficulty in securing the tube and the resulting skin trauma and blistering.

The clinical nurse consultant began to look at what was being practiced in specialised EB areas. The DebRA UK literature and their practice and Great Ormond Street Hospital for Children in the UK recommend that bathing is avoided in the neonatal period to allow intrauterine and birth damage to heal (Denyer, 2009). This practice is also

appropriate to avoid further trauma of the vulnerable skin in the exposed neonate. It is suggested that the secondary dressing can be changed if there is ooze or strikethrough, which also allows the opportunity to examine for any blistering.

Limb-by-limb dressing changes are also advocated by the nursing team at Great Ormond Street Hospital, London. This is a more controlled method that reduces trauma and there is less chance of the baby damaging themselves from friction through movement.

With initial reluctant consent from the dermatology team at Sydney Children's Hospital, it was agreed to change the practice from daily baths to limb-by-limb dressing changes every three days. If there were sloughy areas or hypergranulation that needed a daily change or steroid application, these areas were irrigated and changed daily. When improved, this area would revert to third-daily changes again. The cessation of daily baths meant that baby A's psychological trauma was reduced and pain management improved. Self-damage was minimised, as well as handling-related trauma. Figure 2 shows his hand wound healing well with epithelial edging and minimal hypergranulation at three months old. Bathing was reintroduced as his birth trauma improved.

The change in dressing practice reduced analgesia and sedation requirements. Re-epithelialisation occurred and the baby's increased energy reserves resulted in weight gain. Baby A's quality of life improved and nursing staff reported a greater satisfaction in caring for him. At three and a half months old, baby A was successfully fostered to a family who maintained the regimen of dressing changes every three days. A few weeks after discharge his nasogastric tube was no longer required.

Figure 3 shows his hand at five months old — a healthy wound that has shrunk in size, with epithelial migration.

Despite his EB and living with chronic wounds he continues to thrive with the same foster parents. Now, with the use of polymeric membrane dressings, he only

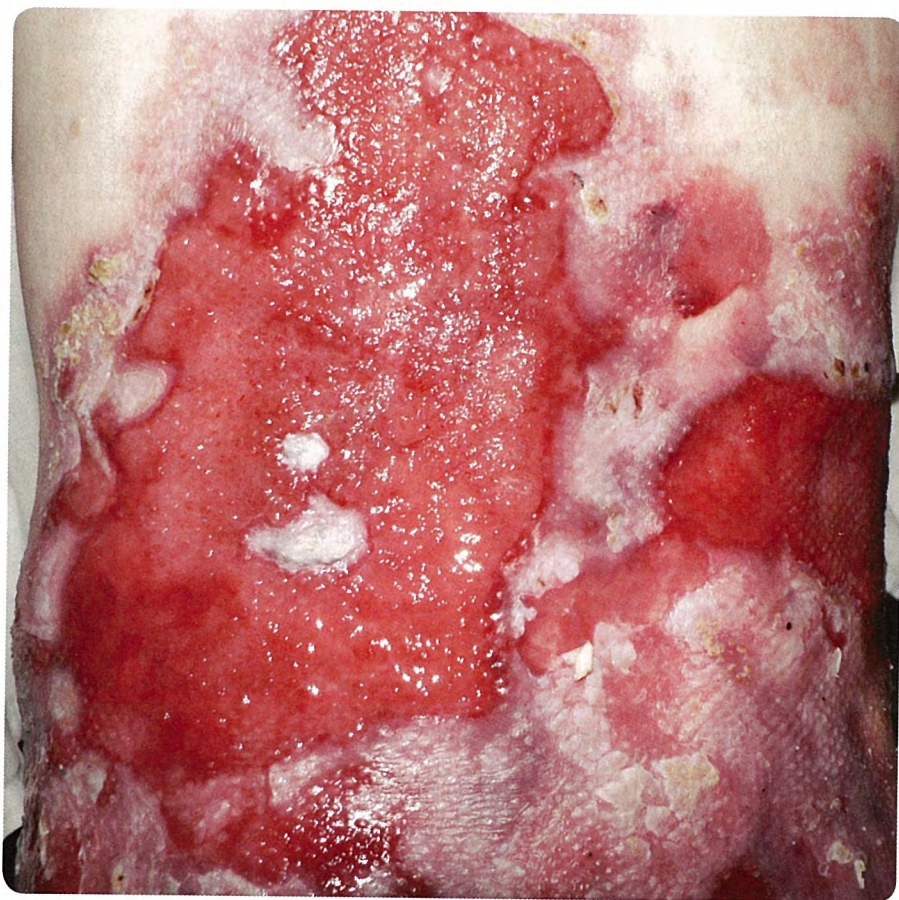


Figure 4. Chronic back wound at the start of the trial.



Figure 5. Back wound eight weeks later

needs dressing changes twice a week, which has improved the quality of life for the whole family.

Case report 2

Patient B was a seven-year-old boy, who suffered from severe generalised dystrophic EB and multiple chronic wounds. A large wound had been present over his back for five years and was increasing in size. In common with many EB patients over the world, patient B was reluctant to bathe. Bathing 3–4 times per week was a painful, traumatic, time-consuming procedure, often resulting in being late for school. A recent trial using polymeric membrane dressings showed a reduction of 43cm² in his back wound, the improvement being noted over an eight-week period (Figures 4 and 5) (Stevens, 2010).

After this trial his parents chose to continue using polymeric membrane dressings. They were able to go on holiday, and during this time they did not bathe him for one week, but just gave him two full dressing changes. As this polymeric membrane dressing reduces the need for primary and secondary dressings, the process is quicker. If blisters are suspected between dressing changes, the dressing can be temporarily lifted, the blister lanced and the dressing fixed in place again.

Figure 6 demonstrates that despite one week without a bath, the wound was clean, with no maceration, less hypergranulation tissue and clear epithelial migration at the wound edges. The surrounding skin is also healthier.



Figure 6. Back wound after one week without a bath.

Key points

- ▶▶ Epidermolysis bullosa is a rare genetic skin condition.
- ▶▶ Handling results in trauma and skin loss.
- ▶▶ Dressing regimens are complex and time-consuming.
- ▶▶ Pain is multifactorial and difficult to control.

The family expressed an improvement in their quality of life by reducing bathing at home and being able to enjoy a family holiday without the trauma of bathing before dressing changes.

Conclusion

These two case reports demonstrate that bathing does not need to be a regular occurrence for those living with EB. Advances in modern dressings, wound cleansing and dressing change practices managed in line with the needs of the individual and their family, all help to improve healing of chronic wounds and, most importantly, quality of life. The development of dressings containing a safe cleanser provides an alternative method of reducing the bioburden and has a subsequent effect on reducing odour and colonisation. **WUK**

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