Managing wounds in patients with epidermolysis bullosa

KEY WORDS
- Blistering
- Epidermolysis bullosa
- Gentic disorder
- Skin fragility
- Wounds

ABSTRACT Epidermolysis Bullosa (EB) is a group of rare genetic skin fragility disorders. This article focuses mainly on patients affected by the most severe subtype, recessive dystrophic EB. Wound management can be challenging and frequently needs to be adapted due to non- cutaneous and other factors which impact wound healing in this cohort of patients.

Epidermolysis bullosa (EB) is a group of rare, genetically inherited skin fragility disorders. There is a broad spectrum of severity but the common feature is blistering in response to mild mechanical trauma or friction (*Figure 1*). Blister sites go on to form wounds, which may become chronic lasting many years. In severe forms EB is a complex multisystem disease with wounds extending across the entire body and fragility extending to internal mucosa and eyes. EB is lifelong and there is no cure currently available.

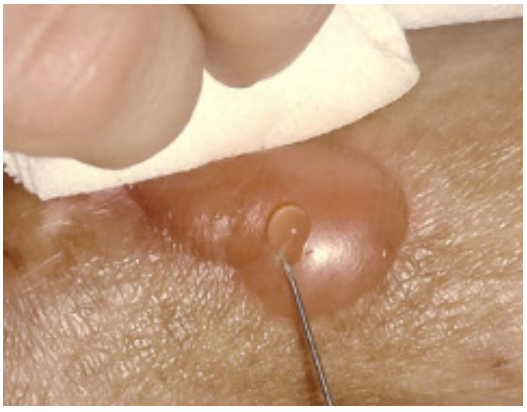

Figure 1. Epidermolysis bullosa blisters are not self-limiting and for this reason the recommendation is to burst the blister using a sterile needle.

Types of epidermolysis bullosa

There are four main subtypes of EB. (Has et al, 2020), EB simplex, junctional EB, dystrophic EB

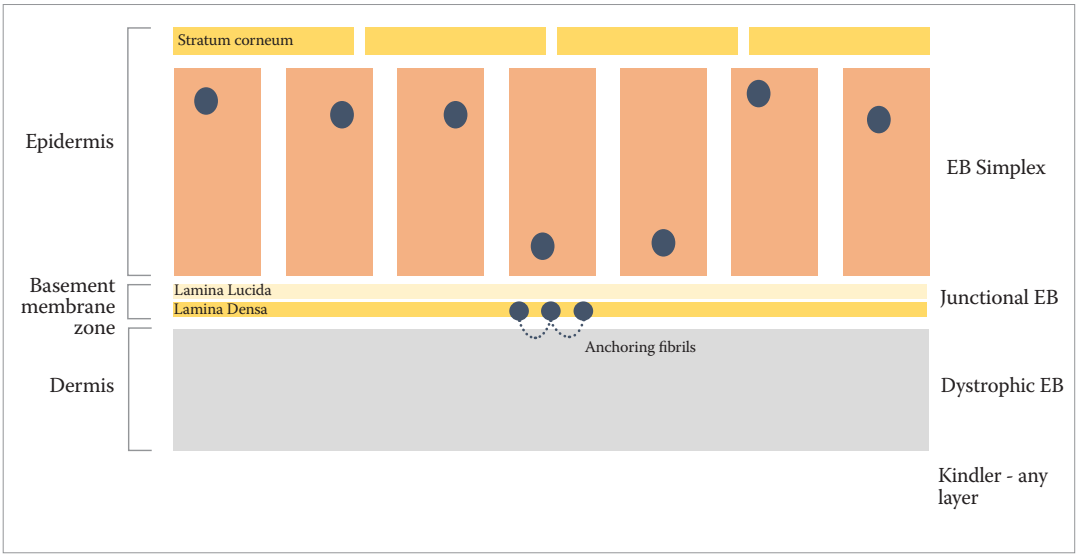

Figure 2. Structure of skin and level of skin separation in epidermolysis bullosa.

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Table 1. Types and characteristics of epidermolysis bullosa

Type of EB	Level of cleavage in skin and key genetic mutations	Common features
EB Simplex – localised and generalised (EBS) (*Figure 3*)	Epidermis Keratin 5 or 14	Mild blistering, usually localised to hands or feet.
Junctional EB (JEB) (*Figure 4*)	Basement membrane zone laminin or Type XVII collagen	Death in early infancy (severe JEB) or a relatively normal life span with chronic ulceration a common feature
Dystrophic EB (DEB) (*Figure 5*)	Dermis Collagen VII	**Dominant dystrophic EB (DDEB):** mild with blistering predominantly to lower legs **Recessive dystrophic EB (RDEB):** severe with extreme fragility, blistering and wounds all over the body with fragility extending to some of the internal mucosa (RDEB)
Kindler EB	Any level	Variable presentation

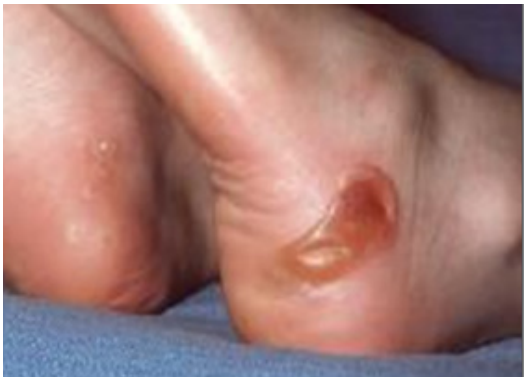

Figure 3. Typical blister seen in epidermolysis bullosa simplex

and kindler. The type is determined by the level of the skin where separation or cleavage occurs and there are common features seen within each subtype illustrated in *Figure 2*. These are summarised in *Table 1*.

Wound assessment and management

In recessive dystrophic EB (RDEB) in particular, it is very common to see multiple wounds of varying duration, extending over large surface areas and in different stages of healing. (Denyer et al, 2017)

To achieve optimal wound management the following factors should influence wound assessment and guide choice of dressing and topical treatments that are introduced to promote healing:

- Aetiology of wound or causative factors
- Systemic factors affecting healing

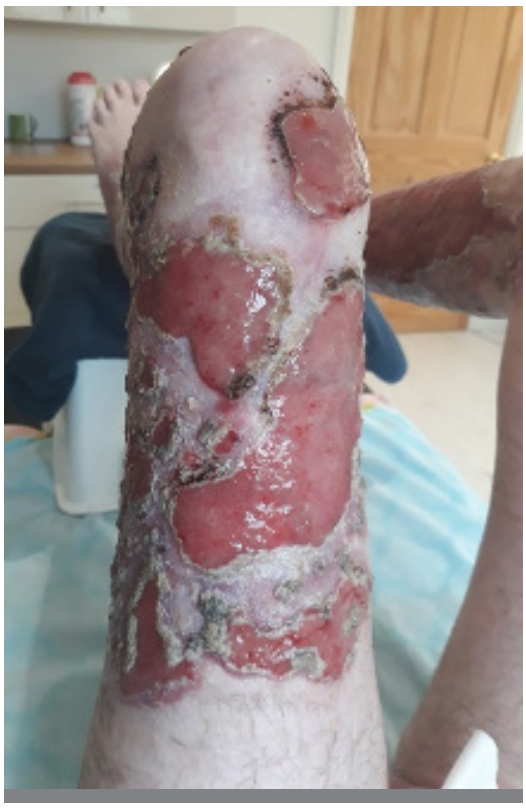

Figure 4. Chronic ulceration seen in junctional epidermolysis bullosa.

- Assessment of wound (WUWHS, 2016).

The TIME framework (Dowsett and Ayello, 2004) is a widely accepted standard for wound assessment. The Triangle of Wound Assessment tool is a useful

Table 2. Non-cutaneous factors affecting ability to heal in severe subtypes of epidermolysis bullosa.

Non-cutaneous factors	Impact on healing
Anaemia of chronic disease	Wound healing process requires oxygenation. Iron's primary role is to carry oxygen rich haemoglobin to cells in the body, including the wound site. Low oxygen levels as a result of anaemia may halt or slow down the wound healing process.
Nutrition may be compromised due to poor appetite, poor dentition, oral blistering and swallowing difficulties (oesophageal strictures) Patients in constant calorie deficit - there is increased calorie requirement to compensate for losses of proteins/fluids through large wounds (Hubbard et al, 2011)	Malnutrition is known to have a detrimental impact on wound healing. Macro-, micro-nutrient and fluid requirements need to be met in order promote good skin condition and maintain and repair tissues (Acton 2013)
Pruritis	Patients can fall into an 'itch/scratch cycle' Scratching may damage fragile skin and lead to further wound breakdown (Denyer et al, 2017)
Pain	Pain is a stressor which can have a detrimental impact on wound healing (Wynn & Holloway, 2019) Adequate analgesia essential for pain associated with dressing changes (Denyer et al, 2017)
Physical exhaustion 2-4 hours spent on dressing change daily	Detrimental impact on quality of life. Additional psychological stressor which can affect wound healing
Gene defect	Underlying gene defect affects integrity of skin and may affect ability to heal. Patient is trapped in a constant wound/heal cycle

Table 3. Factors influencing dressing choice in epidermolysis bullosa.

Non-cutaneous factors	Impact on healing
Fragile skin	Sticky dressings contraindicated
Large surface area wounds (*Figure 6*)	Dressing sizes may be too small
Exudate management	Patient may be unable to tolerate heavy dressings and instead opt for lighter dressings and change regularly
Patient choice	Expert patient with lifelong condition. Choice may be influenced by previous experiences Aim to work in partnership with patient to find acceptable dressings
Conformability	How will dressings be safely secured?
Access to dressings	Some CCG formularies restrict access to dressings
Suitability for long term use	Dressings required for months-years

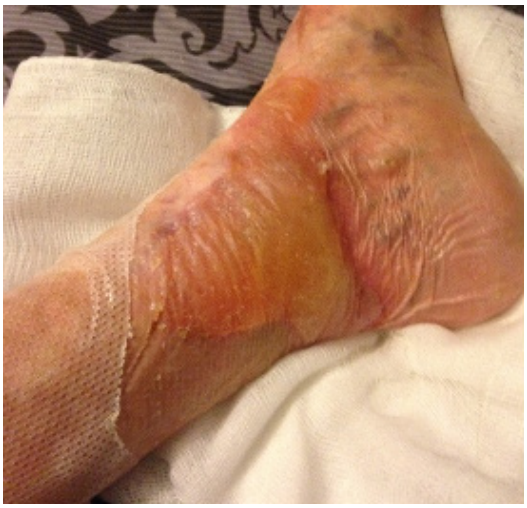

Figure 5. Extensive epidermolysis bullosa blister on the lower leg.

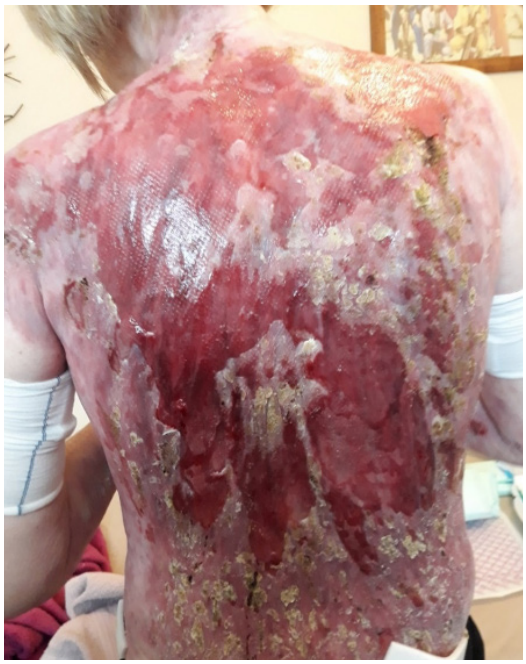

Figure 6. Large surface area, wound in various stages healing and evidence of hyperkeratosis. Typically in recessive dystrophic epidermolysis bullosa .

supplementary approach that extends the concept of wound bed preparation and TIME as it divides assessment of the wound into three areas: the wound bed, the wound edge, and the periwound skin. (Dowsett and von Hallern, 2017)

Wound assessment in EB is guided by the application of these frameworks, however when considering aetiology of the wound and other systemic factors affecting healing there is additional

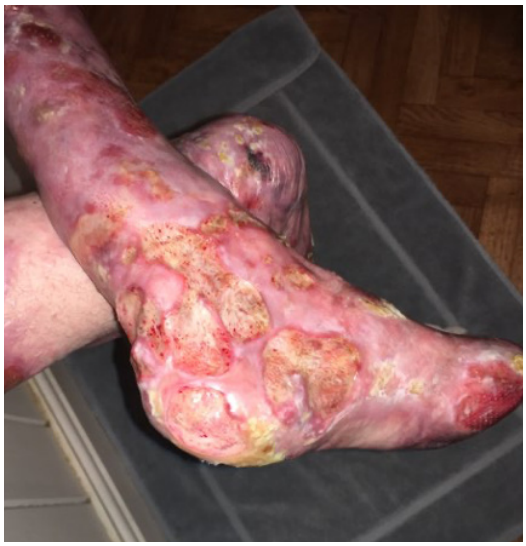

Figure 7. Chronic, deeply ulcerated and sloughy wounds on foot and lower leg in recessive dystrophic epidermolysis bullosa.

complexity to be considered in this patient group. These include non-cutaneous systemic factors which have a detrimental effect on wound healing and limitations on dressing choice. These factors are summarised in *Tables 2 and 3*

Chronic wounds in epidermolysis bullosa

In common with other chronic wounds complications seen in EB wounds, illustrated in *Figure 6* and *Figure 7*, can include:

- High wound bioburden
- Hyperkeratotic wound margins
- Presence of necrotic tissue/slough
- Poorly controlled exudate
- Presence of biofilm
- Infection due to loss of protective function of the skin.

Standard approaches to management, including cleansing and debridement and the management of wound infection, need to be adapted.

Cleansing and debridement

Use of effective cleansing and debriding agents to reduce the wound bioburden and disrupt any biofilm that may be present is a core principle in wound management. For many patients with severe EB, bathing or showering is too painful on fragile or broken skin and they may therefore choose not to bathe at all. Most will choose to cleanse the skin

with antimicrobial solutions or cloths. Many patients use a polymeric membrane dressing with an integral cleansing surfactant to aid wound cleansing.

For those who are able to bathe the use of a mild, well diluted cleanser may be recommended. This may be bleach (Milton) or ≤0.25% salt added to the bath water following dilution guidelines provided by health professionals. (Guy's and St Thomas' NHS Foundation Trust, 2019). Many patients may find that this is too uncomfortable to tolerate on open wounds or broken skin.

Wound debridement must be approached with caution in EB wounds due to extreme skin fragility, which further complicates the ability to effectively cleanse the skin and wounds and successfully prepare the wound bed for healing. (Denyer et al, 2017) Debridement using single filament fibre pads can be helpful in gently removing hyperkeratotic crust (especially in those who do bathe) but should always be used with extreme caution as blistering can occur as a result of even mild friction.

Wound infection in epidermolysis bullosa

EB patients are thought to be at increased risk of sepsis due to the constant presence of chronic, large surface area wounds, their nutritionally compromised status and other non-cutaneous factors. Patients have very little reserve leaving them more susceptible to overwhelming infection and sepsis. Effective prevention of infection is an essential part of wound management in EB.

Concern about antibiotic resistance through over use of topical and systemic antibiotics guide best practice in EB wound management and use of systemic antibiotics is restricted to clinically significant infections only. This would include; swelling, redness and increasing pain in affected area, inceased discharge or pus in or around the wound and feeling systemically unwell with a high temperature of 38ºC or higher. The only exception being treatment of Group A beta haemolytic streptococci. Topical antimicrobial therapies are used to prevent infection in this group of patients who are identified as being at increasedrisk of wound infection and to manage localised wound infection.

Use of antimicrobials

Once the wound had been cleansed and biofilm disrupted it is recommended that an antimicrobial product is used to prevent the biofilm reforming (WUWHS 2016). The type of antimicrobial used by EB patients tends to be rotated on a regular basis. Rotation of antimicrobial every 1–2 months is common practice in the EB patient group who are using topical antimicrobials on a long term basis, however there have been no formal studies to evaluate effectiveness. Topical gels and creams are generally better tolerated than dressings. Due to concern about silver toxicity associated with long term use in EB, silver dressings are used with caution and for duration of 7–14 days only.

A range of antimicrobial agents are well tolerated in patients with EB. These include Honey, Polyhexamethylene biguanide (PHMB), Enzyme Alginogel, Octenidine range, Kytocel, Polymeric membrane dressing and 1% stabilised Hydrogen Peroxide Cream (Denyer et al 2017).

CONCLUSION

Epidermolysis bullosa is a complex, multisystem condition characterised by skin fragility. Chronic and challenging wounds are frequently seen in the more severe subtypes. Principles of wound bed preparation and management including application of the TIME and Triangle of Wound Assessment need to be adapted due to aetiology and systemic, non-cutaneous factors which have an impact on potential for wound healing in this patient group. WUK

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