Wound management in epidermolysis bullosa

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INTRODUCTION

Epidermolysis Bullosa (EB) is the generic term for a large complex group of inherited blistering and skin fragility disorders. There are 4 main types of EB and many additional subtypes. The common factor is fragility of the skin and mucous membranes with a tendency for blisters and wounds to develop following minimal everyday friction and trauma. Fragility results from reduced or absent vital proteins, which give the skin its tensile strength. EB can be inherited either dominantly or recessively, with the recessive types generally being more severe.

The 4 main types of EB are EB simplex; junctional EB; dystrophic EB and Kindler syndrome. Whilst experienced clinicians may be able to make a provisional diagnosis, this is difficult in the presentation of affected new-borns and diagnosis must be made from analysis of a skin biopsy using the key diagnostic tools of positive immunofluorescence, antigen mapping and electron microscopy. Re-categorisation in 2014 removed many of the eponyms and included more variants.[1]

The severity of the condition varies between painful blistering of the hands and feet in EB simplex localised, to death in early infancy resulting from laryngeal disease and faltering growth in those with generalised severe junctional EB. Scarring in those with severe forms of dystrophic EB leads to development of contractures, microstomia, oesophageal strictures and pseudosyndactyly.[2]

PRINCIPLES OF WOUND CARE

Wound healing is compromised by the underlying genetic defect, poor nutritional status, anaemia (both resulting from chronic disease and iron deficiency), pain and pruritus. The general guidance of selection of the correct dressing, protecting the peri-wound skin, avoiding skin stripping, lancing blisters to limit their spread, addressing the bio-burden and exudate management apply. In addition, the type of EB further dictates skin and wound care. Whilst this article will briefly describe these categories and recommendations, more detailed information is available in Best Practice Guidelines for Skin and Wound Care in Epidermolysis Bullosa.[3]

Although there is a plethora of wound care products available, the selection suitable for those with fragile skin is more limited. Adhesive dressings and even those coated with soft silicone may result in skin stripping in the most fragile patients. Using a Silicone Medical Adhesive Remover (SMAR) can eliminate this risk. SMARs are also essential in removing adhesive products such as fixation for cannulas and essential monitoring devices.[4]

Due to the increase in antibiotic resistant organisms, oral or intravenous antibiotics are reserved for systemic infection with a preference to use topical antimicrobial therapies as first line treatment.[5]

NUTRITIONAL SUPPORT

Oral blistering and dysphagia compromise intake in severe forms of EB. Long term enteral feeding is often necessary to increase nutritional requirements and aid wound healing.[6]

CARE OF THE NEWBORN WITH SEVERE EB

Severely affected infants often present with widespread skin loss resulting from inter-uterine movements and compounded by the trauma of delivery. In addition to complex wound management, all screening and handling procedures require modification to reduce additional damage to the fragile skin and mucosa.[7]

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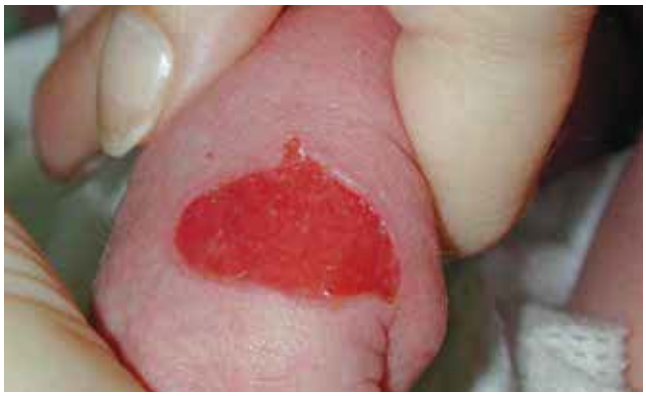
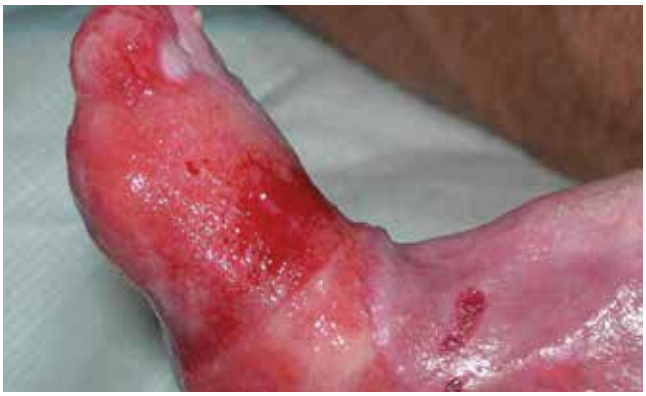
Conflicts of interest: None



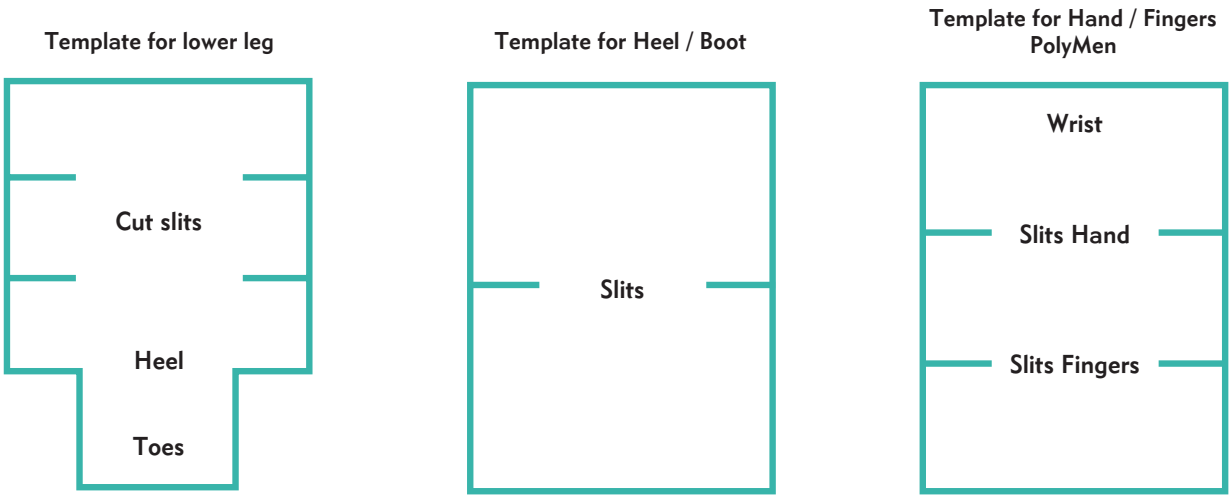
Skin stripping following removal of adhesive tape



Birth damage in a severely affected new born infant

Figure 1: Foot and lower leg dressing template

Remove cord clamp and replace with a ligature	To protect peri-umbilical skin
Nurse in cot/bassinette unless incubator required for reasons such as prematurity	Heat and humidity may exacerbate blistering
Give regular analgesia and additional pre-procedure doses using a validated neonatal pain score for adjustment	To provide adequate pain relief
Lance all blisters with a hypodermic needle	Blisters are not self- limited and left unchecked will spread, resulting in pain and further tissue damage
Leave the blister roof in situ	The roof will act as a protective dressing and reduce pain and tissue damage
Cover open wounds with a non- adherent absorbent foam dressing e.g. polymeric membrane	To provide an optimal wound healing environment, continual cleansing of the wound and management of exudate To reduce duration of dressing changes
Use a template to pre-cut dressing shapes. Cover entire limb, overlap dressing and secure by taping to itself	To avoid skin stripping from adhesive tapes and to protect undamaged skin from normal baby movements
Dress digits individually	To avoid early pseudosyndactyly
Cleanse napkin area with 50% liquid paraffin, 50% white soft paraffin in ointment or aerosol form	Water may cause stinging to open wounds and blister sites
Line napkin with soft material such as commercial liner	To reduce trauma from the edges of the napkin
Protect intact skin with a barrier product	To promote skin integrity
Cover wounds and blister sites with hydrogel impregnated gauze dressings	To provide a moist wound environment and protect from faecal contamination
Nurse on neonatal mattress	To protect skin, offer comfort and for ease of handling
Avoid bathing until inter-uterine and birth damage have healed	To avoid further trauma from handling
Dress in front fastening baby suit	For ease of handling and for added protection from normal baby movements
Always use a Silicone Medical Adhesive Remover to safely take off adherent dressings, adhesive tapes or adherent clothing	To avoid skin stripping



WOUND MANAGEMENT SPECIFIC TO THE TYPE OF EB

EB SIMPLEX (EBS)

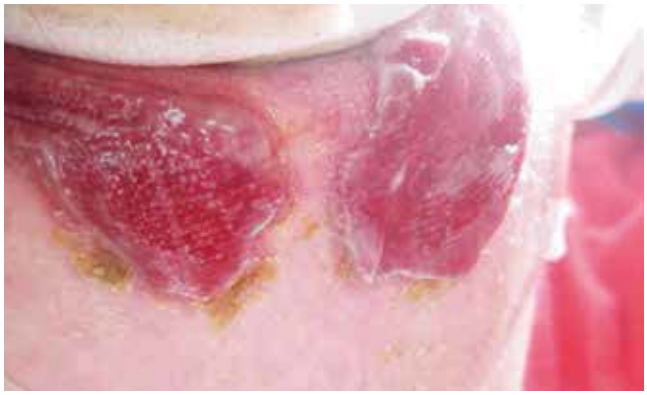
Most forms of EBS are dominantly inherited and result from a disorder of keratin proteins, which provide scaffolding for basal epidermal cells. Defects of keratin result in blisters forming following minimal friction and trauma.[8]

With the exception of neonates with generalised severe EB simplex who are frequently born with extensive wounds, the localised and generalised types of EB simplex are characterised by blistering caused by friction. EB simplex is affected by heat and humidity, with blistering being much worse in the summer months.

Management of EBS is by lancing blisters, reducing friction and use of measures to keep cool such as using socks containing silver thread, cooling insoles and shoes offering ventilation.[9]

Suitable dressings offer comfort, reduce heat and promote healing of blister sites. Appropriate dressings include sheet hydrogels, bi-stretch silicone dressings and bordered soft silicone dressings. However, many affected individuals prefer not to use any dressings at all.

Care must be taken to ensure blistering does not result from trauma caused by the edges of dressings. This can be minimised by rounding off the dressings and padding beneath the edges with dressings such as lipidocolloid or hydrofiber.



Over granulation tissue in a child with junctional EB

JUNCTIONAL EB (JEB)

JEB is a recessively inherited condition. Blistering occurs within the lamina lucida. Mechanical integrity of the hemi-desmosomes or anchoring fibrils is compromised by gene mutations.

In its most severe form, generalised severe junctional EB infants rarely survive beyond the first two years of life.[10] Less severe forms of junctional EB (JEB Intermediate) predispose to the development of chronic wounds, alopecia and with an increased risk of squamous cell carcinoma in mid life.

Over-granulation tissue features in wounds of all types of severe EB, but it is most common in those with junctional EB. Measures to deter this florid tissue include selecting a primary dressing with a very fine mesh such as a lipidocolloid, or using hydrogel impregnated gauze dressings. Hypergranulation is most troublesome on the face of longer-term survivors, causing chronic wounding, pain and disfigurement. Treatment with a very potent topical steroid in combination with an antimicrobial agent destroys the over- granulation tissue and encourages healing to take place.

DYSTROPHIC EB (DEB)

Dystrophic EB can be inherited either dominantly or recessively. In DEB, collagen VII is reduced and, in severe recessive forms, completely absent.

Collagen VII forms the basis of anchoring fibrils, which attach the epidermis to the dermis leading to formation of traumatic wounds following minimal shearing forces. Those with markedly reduced or absent collagen VII heal with atrophic scarring leading to development of progressively disabling contractures.

Wound management in dystrophic EB is complex, with a need to protect from trauma, manage the bio-burden and ▶

exudate, and attempt to delay formation of contractures and pseudosyndactyly.

There is a high risk of development of aggressive forms of squamous cell carcinoma in young adults with severe dystrophic EB.[11] Patients and carers need to be made aware of the signs and encouraged to report any change in appearance of a chronic wound, an abnormal sensation or increase in pain or exuberant tissue growth within an existing wound.

Suitable dressings in the management of dystrophic EB are plentiful and can be selected according to need, such as exudate management, critical colonisation, infection, odour or protection. Recommended dressings include soft silicone, lipidocolloid, foams, honeys, enzymatic gels and super-absorbers.

KINDLER SYNDROME

Kindler syndrome is recessively inherited and results from mutations in the gene FERMT1.[12]

This is a rare type of EB, which is difficult to diagnose as it may demonstrate features of other types of EB due to the unique feature of variable level of cleavage with blister formation occurring within the epidermis, lamina lucida or sub lamina densa.

Neonatal skin loss and blisters are common but reduce during infancy. Later changes include photosensitivity requiring sun protection, atrophic and pigmentary skin changes.

Dressing selection is indicated by level of blister formation with epidermal blistering following recommendation for those with EB simplex and sub lamina densa that of dystrophic EB.

CHRONIC WOUNDS

Those with severe forms of EB are predisposed to the development of chronic wounds. Common causes include infection and critical levels of colonisation, poorly controlled exudate, presence of slough and necrotic material and destruction from scratching to relieve the discomfort resulting from pruritus.

Management of these wounds is complex, and due to the underlying gene defect and multiple co-morbidities, not always successful.[13]

Appropriate management of exudate is crucial if attempts are to be made to protect the peri-wound skin from maceration.

Bathing may be difficult for individuals who have multiple wounds and contractures due to pain, risk of damage from handling and duration of a full dressing change.[14] In the absence of bathing, wound cleansing with an antiseptic agent is encouraged as is removal of slough and dried exudate using debridement cloths and pads. Medical grade honey preparations, hydrogels, enzymatic and larval debridement have also been successfully employed.

Advanced therapies for management of chronic wounds using bioengineered skin grafts, protease modulators, collagen and keratin dressings should be considered if regular recommended dressings are unsuccessful.

INFECTED WOUNDS

Infected wounds should be managed using topical antimicrobial products unless the patient is systemically unwell. Recommended products include medical grade honey, enzyme alginogel, polymeric membrane, polyhexamethylene biguanide and silver dressings. Silver products should be used with caution in infants under one year. When used in large areas there is a potential risk of raised plasma silver levels, so restrict usage to 14 days.

MANAGEMENT OF FUNGATING WOUNDS

Patients with severe forms of EB, in particular those with generalised severe dystrophic EB, have a very high risk of developing squamous cell carcinoma. Towards the end of life these may result in an inoperable tumour, which develops into a fungating wound. Careful management to address pain, exudate, bleeding and odour may require multiple layers of dressings. Dressing changes should be kept to a minimum in order to reduce the risk of bleeding and additional pain.[15]

PRURITUS

Pruritus is a challenge for affected individuals, carers and health professionals. Scratching leads to extensive skin damage and wounding. Additionally intense pruritus forms part of the pain spectrum and can result in depression and insomnia.[16]

Management of pruritus is by simple measures such as applying emollients or products containing menthol to cool the skin, avoiding warm environments if possible or using air conditioning or non–buffeting fans to circulate the air.

Loose cotton clothing or specialised silk garments have anti-pruritic properties and can be helpful.

MANAGEMENT OF PAIN

Pain in severe EB is multi-factorial and requires input from a specialised pain team with an age-appropriate validated pain tool used at each dressing change in order to achieve optimal pain management.

Management of wound pain must consider the presence of both nociceptive and neuropathic pain and may require regular and procedural opioid treatment together with agents such as gabapentin and pregabalin. Topical agents including morphine gels and dressings containing ibuprofen are helpful for individual painful wounds.

Non-pharmacological treatments such as guided imagery and psychotherapy are used in conjunction with pharmacological treatments.[17]

CONCLUSION

Wound management in EB is complex as it is influenced by multiple co morbidities and the fragility of the skin.

Dressing management is specific to the type of EB, presence of infection or critical colonisation, levels of exudate, availability of products and personal preference. Families with a long history of EB, such as those with EB simplex localised, may shun modern technologies and prefer to use less suitable products as dictated by family members, which is frustrating for the professional attempting to help. ■

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