Recessive dystrophic epidermolysis bullosa. Part 2: care of the adult patient

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Abstract
This article is the second in a series of three focusing on the causes, clinical presentation, complications and care of adult patients affected by epidermolysis bullosa (EB) – a group of rare genetic skin fragility disorders. The emphasis of this article will be on the nursing care of adults with recessive dystrophic EB (RDEB). RDEB patients are never ‘wound free’ and the challenge to provide effective wound care is immense. This article details some of the basic practical wound care issues to be considered when caring for a patient with EB, particularly RDEB, emphasizing the need for a holistic approach, with strong patient involvement. Minimizing and preventing trauma, generating an optimal wound healing environment, and the management of pain and pruritis will also be discussed.

Key words: Adult nursing, DebRA, Epidermolysis bullosa, Severe skin fragility, Wound care

Epidermolysis bullosa (EB) is the ‘umbrella term’ for a group of rare genetic skin-blistering disorders characterized by the susceptibility of the skin and mucous membranes to blister or sheer away in response to minimal friction and trauma (Lin and Carter, 1992). This is due to reduction or absence of vital proteins, causing separation of the epidermis from the dermis or rupturing of the cytoskeleton of the cells (Lin and Carter, 1992).

EB is divided into three broad categories, determined by the level of skin cleavage – discussed in article one of this series (Pillay, 2008). Dystrophic EB is further classified by the mode of inheritance (dominant or recessive). Healing is abnormal leading to contractures and scarring (dystrophic).

The affected protein in RDEB is collagen VII and the variation seen in the disorder is determined by the causative genetic mutation and the amount of collagen VII the individual is able to make (Fine, 1999a). Collagen VII forms the anchoring fibrils, which bind the epidermis to the dermis. Weakened or absent anchoring fibrils lead to easy separation of the epidermis from the dermis and extensive skin involvement is likely with blisters and wounds in various stages of healing. However, it is important to remember that not all EB patients will present with extensive open wounds; for example, patients with EB simplex may appear to have ‘normal’ skin but will blister in response to heat, friction, shear or pressure. Due to the rarity of this condition, few healthcare professionals will ever care for someone with EB, and furthermore, will have little or no knowledge of the causative factors, clinical presentation and complications of the disorder. This knowledge is needed if healthcare professionals are to care for patients effectively and safely, and to ‘do no harm’.

In common with other genetic disorders, there is currently no cure for EB (Horn, 2003). As work progresses to an effective cure in the form of gene therapy, treatment remains symptomatic, with skin and wound management forming a large part of care. However, the extreme fragility of the skin severely limits the choice of products and methods of care.

Weller (2005) defines a wound as a cut or break in the continuity of any tissue resulting from injury or operation. In EB, minimal trauma to the skin in the form of friction, pressure or shear can cause blisters, which in turn become wounds (Atherton and Denyer, 2003). With appropriate management, many of these acute blister sites will heal; however, it may not be possible to heal all wounds due to the very nature of the condition, as the skin is likely to repeatedly break down (Uitto et al, 2000) (Figure 1). Mellerio et al (2007) highlights the issues that make EB wound management highly complex, with numerous factors negatively influencing the healing process (Box 1). Wound management objectives in EB are to provide the optimal healing environment, promote comfort and to protect from further trauma.

Patient assessment and care planning
Pressure relief
Whether caring for an EB patient in hospital or the community, it is important to carry out a full pressure risk assessment as soon as possible. Pressure areas can break down extremely rapidly. All EB in-patients will require a pressure-reducing

Box 1. Factors affecting wound healing in epidermolysis bullosa

- Trauma or friction to skin
- Anaemia
- Poor nutrition
- Infection
- Pruritus

From: Mellerio et al (2007)
mattress, both at home and in hospital. Repose™ mattresses (Frontier Therapeutics, Blackwood) have been found to be very effective in EB. These mattresses are both portable and 'low-tech', leading to ease of use for both patients and nurses.

Where the risk of skin damage is greater because of the severity of the condition, the need for an enforced period of bed-rest or at the end of life, a TheraKair Visio® low-air-loss (KCI Medical Ltd, Kidlington) mattress may be preferable (Pillay and Hon, 2007). Poor mobility and strictures of limbs are likely to increase the potential for pressure damage (Grey et al, 2006). A padded toilet seat, bath chair and manual handling aids, even if padded by soft towelling, may prevent tissue breakdown.

Nutrition
Poor nutritional status impacts on wound healing and is a common problem in RDEB. Many EB patients develop microstomia (small mouth opening) from repeated oral blistering and contracted scar tissue (Lin and Carter, 1992). This makes dental hygiene difficult and often leads to decay although primary enamel is good in this type of EB. A soft or puree diet is often enhanced with liquid nutritional supplements and where this is inadequate a gastrostomy may be placed (Mellerio et al, 2007). Blisters can also occur in the oesophagus and strictures may form. An oesophageal dilatation is offered to those experiencing dysphagia and this should be carried out with extreme caution and under sedation by a team experienced in the care of EB patients (Azizkhan et al, 2006). Some patients will require regular dilatations every few weeks, while for others an occasional one will suffice.

Patients with RDEB should be seen by a dietician, preferably one with understanding and knowledge of the condition, to assess their nutritional needs. This will certainly be increased to meet their wound-healing requirements and adjust for the loss of nutrients through constantly open wounds. If a patient’s nutritional status is compromised wounds are less likely to heal (McManus, 2007).

Blood chemistry
A comprehensive set of blood tests are recommended as most RDEB patients suffer from anaemia and other abnormalities of blood chemistry due to blood loss from the wounds, poor intake of oral iron and ‘anaemia of chronic disease’ (Mellerio, 2007). Iron supplements are recommended but where these cannot be taken orally an intravenous infusion can be given. In general these patients struggle to maintain their haemoglobin levels and regular infusions are necessary (Mellerio, 2007).

Infection
Infection and critical colonization of wounds is common and wounds which are painful, sloughy or producing malodorous exudate should be gently swabbed. Staphylococcus aureus and Pseudomonas aeruginosa can frequently present in the wounds of EB patients (Hartley, 2003). It is important that any signs of infection are acted on as soon as possible as any type of infection can delay wound healing (Mellerio et al, 2007) and compromise the general wellbeing of these vulnerable patients.

Antibiotics may be required if there is evidence of spreading wound infection, cellulitis or if the patient is systemically unwell, otherwise topical treatments are the treatment of choice for localized infection. Out of many topical products available, experience suggests that the following products are helpful: 1% hydrogen peroxide cream (Crystacide™, Derma UK Ltd, Bedfordshire), Dermol™ 500 Lotion (Dermal Laboratories, Hertfordshire), silver impregnated dressings,
including Urgotul® SSD (Urgo, Loughborough), Mepilex® Ag (Molnlycke, Bedfordshire), Aquacel® Ag (Convatec, Uxbridge) and medicated honey (Actixon Tulle® [Advancis Medical, Nottingham], Mestiran® [Unomedical, Worcestershire]), and Medihoney™ [Medihoney Ltd, Berkshire]). As the barrier function of the skin is severely compromised in these patients, it may be necessary to use antiseptics on a rotational basis in an effort to prevent infection.

All patients are advised to rotate the use of the products every 2–4 weeks to prevent resistance (European Wound Management Association [EWMA], 2006). Due to the possibility of systemic absorption, it is advised that silver products are used for a maximum of 4 weeks (Mellerio et al, 2007). Honey has been used successfully in EB, both to heal recalcitrant wounds and also for its antibacterial and anti-inflammatory properties (Hon, 2005). However, the pain caused by the strong osmotic ‘pull’ exerted by the honey and its acidity may make some patients reluctant to use these products (Molan, 2001). Dressings with antimicrobial properties have been available for many years, and an increasing range of adjunctive therapies in the form of clothing are becoming available. These include garments from 9–80% silver and woven silk, such as Dermasilk® (Espere Healthcare Ltd, Bedfordshire), which may assist in preventing bacterial colonization and pruritis in EB.

**Pain control**

The pain experience of people with EB varies enormously, as does the analgesia they require. Some manage with an occasional paracetamol while others take opioids regularly. Pain in EB emanates from a variety of sources. Wound pain emanates from:
- The wound
- Fear of the dressing change
- Infection
- Type of dressing used
- Dressing technique.

Non-wound pain emanates from:
- Oral ulcerations and erosions
- Dental decay
- Oesophageal blistering
- Osteoporosis and osteopaenia
- Contractures
- Corneal erosions and ulceration (Herod et al, 2002).

To implement effective therapy it is important to thoroughly investigate the source of the pain, and assess the type and severity of pain using a recognized pain assessment tool, such as the Numerical Rating Scale (Alexander et al, 1994).

Patients who are threatened by pain may suffer fear, anxiety and worry (Acton, 2007). Hollinworth (2000) states that the negative experience can remain with the patient for a long time afterwards. For EB patients who from childhood have suffered hours of painful dressing changes, often daily, the psychological pain is as real as the physical. Additional pain relief may be required before and during dressing changes. Pain may be exacerbated by the use of inappropriate dressings, infection and neuropathic pain. Allodynia – low intensity stimulus producing pain (White and Harding, 2006) in relation to chronic wounds – has also been noted in some EB patients.

It is often necessary to involve the pain team in the assessment and management of such complex pain issues. Wound pain may respond to oral analgesia, beginning with paracetamol and/or ibuprofen, with opiates being introduced as the pain requires, using the World Health Organization pain relief ladder (WHO, 1986) (see Figure 3). Opioids can also be used directly on the open wound by mixing morphine sulphate with a hydrogel (Mellerio et al, 2007). Gabapentin or amitriptyline is useful for neuropathic pain (British National Formulary, 2008).

Adequate pain relief prior to the dressing change is essential (Hollinworth, 2004), and must be given time to be effective before beginning the procedure. The nurse/carer must also plan and take into account how long the dressing procedure will take and ensure that additional analgesia is available for breakthrough pain. It is not unusual for severely affected RDEB patients to require 2–6 hours of nursing time to complete the procedure. The use of non-adherent dressings, silicone adhesive remover, or soaking dressings off, may also reduce the pain experienced (EWMA, 2002). Creating a calm, unhurried environment and ensuring the patient feels confident in the skill of the nurse is important; as is allowing the patient to control the speed of the dressing change, with rest periods when needed. Some patients may prefer to remove their own dressings and indeed replace some of them. The room should be warm as the extensive skin loss will mean that the patient cools rapidly once the dressings are removed. Allowing the patient to become cold not only enhances pain and discomfort, but also slows healing (Flanagan, 1998).

Other non-pharmacological strategies can include distraction or relaxation therapies, for example, getting the patient to watch a video, DVD or listen to music during the dressing change.

Dressings impregnated with ibuprofen (Biatain®-Ibu® Coloplast, Peterborough) are currently being evaluated in EB, and to date some patients have reported that the dressings do relieve wound pain within a very short time of application.

**Care of the wounds**

Unlike most other patients with wounds, EB patients and their families/carers have had to develop real expertise in dressing techniques and detailed knowledge of what dressings and topical treatments will work for them, and what won’t. This knowledge, combined with the knowledge of the healthcare practitioner, can be invaluable when making wound management decisions and carrying out the dressing change. It is therefore a lost opportunity when EB patients find that healthcare professionals do not listen to what they are saying about their dressing regime, or what dressings and topical products work best on their skin. Failure to listen can lead to patients feeling alienated from their treatment (Benner, 2001) and potentially cause both psychological and actual harm to the patient. When undertaking wound care the nurse should negotiate with the patient over what is acceptable to them and what the nurse feels able to undertake within the bounds of professional accountability, always remembering that these patients are experts in their own condition.

All EB patients are issued with ‘Handle with Care’ cards, giving basic information about EB, do’s and don’ts, and
**Figure 3. World Health Organization’s pain ladder (WHO, 1986).**

**STEP 1**
Non-opioid adjuvant, e.g. aspirin, paracetamol, or a non-steroidal anti-inflammatory drug

**STEP 2**
If pain relief not adequate, may need:
- Opioid for mild to moderate pain ± non-opioid adjuvant, e.g. codeine

**STEP 3**
If pain not resolved may need:
- Opioid for moderate to severe pain, e.g. morphine, oxycodone, fentanyl ± non-opioid ± adjuvant

contact numbers for DeBRA EB nurse specialists in the event of admission to hospital or emergency departments. These cards are a useful reminder to carers about the fragility of the patient’s skin (Hon, 2003).

Some patients choose to dress all of their wounds on a daily basis while others may find this too painful and too long a process. These patients may prefer to redress one or two wounds only each day. The decision to redress wounds will also be influenced by the type of dressing used, the stage of wound healing and whether infection is present. Wounds are treated as ‘clean wounds’, unless surgical, in which case a sterile dressing procedure is required.

It is important that adequate time and staffing is allowed for dressing changes. Two people may be needed to open packaging and hold dressings in place while bandaging, with one person being available to collect any needed items. As the dressing change may be a lengthy procedure in an overheated room with the possibility of the patient being distressed, nurses may need relief, both physical and emotional. Wound odour may also be a factor that makes the working environment difficult. Patients with extensive wounds easily lose their body heat (Demling and DeSanti, 2003), so a warm room, good preparation and timely redressing is essential. In an emergency or while waiting, for example, for a doctor on ward round, cling film may, for a short time, be applied to prevent heat and moisture loss. This also ensures the patient’s skin does not stick to clothes or bed linen and allows the use of a sheet to ensure the patient's dignity at all times. Many patients wish to assist in dressing removal. This can be a time-consuming process: unravelling the bandages without pulling on other dressings and skin that may have become stuck together with blood and exudates. Soaking dressings off in the bath may be a preferred method for some, and use of a medical adhesive remover to remove adherent and even low/non-adherent products. Over recent years many more non-adherent products have become available and while some have proved suitable, they still have the potential to damage the very fragile skin of those with severe forms of EB. A small patch test of a new product must be undertaken, before extensive use.

**Bathing and moisturization**
Water dries the skin and dry skin can become irritated, crack and harbour infection. The scratch/itch cycle is difficult to break, and can cause much damage to EB skin. Use of bath emollients and moisturizers on intact skin may assist. Non-perfumed products containing colloidal oatmeal have a natural antipruritic and moisturizing action (for example, Aveeno® [Johnson and Johnson, Maidenhead]). The Dermol® (Dermal Laboratories) range of products, including shampoo, contains antimicrobial and moisturizing properties. Anecdotal evidence suggests oil-based moisturizers are very effective, but can trap the heat and consequently cause irritation. So immediately after bathing or in warm weather a water-based product may be desirable due to the natural cooling effect as the water evaporates. Arguably good practice has always been thought to be the application of moisturisers to areas of intact skin in a stroking fashion in the direction of hair growth, to prevent folliculitis. However, there is no research evidence to support this common practice. A soap substitute should be used, and gentle patting dry the skin with clean towels or soft gauze. Non–biological washing powders for washing clothes and bandages is desirable. Following bathing, or before new dressings are applied, any blisters will need to be aspirated.

**Aspiration/drainage of blisters**
EB blisters are not self-limiting and will continue to enlarge unless punctured with a sterile needle or in some cases sterile scissors. A green gauge (21g) sterile needle is the preferred method of lancing a blister. It is important to make a big enough puncture in the blister so that the fluid can be successfully drained out. If a smaller gauge needle is used, the resultant hole is likely to re-seal quickly causing the blister to refill (Vonhof, 2006). Gently rolling sterile gauze across the blister can aid drainage. Once drained, it is important that the roof of the blister remains intact and is not removed as it acts as a natural ‘dressing’ protecting the raw skin underneath and as a barrier against infection. Once the blister has been successfully drained an appropriate dressing should be applied.

**Chronic and acute wounds**
Adult RDEB patients usually have a mixture of both chronic and acute wounds at any one time. Chronic wounds may be defined as those that take longer than 6 weeks to heal, or are not progressing along a steady trajectory to healing (McManus, 2007). These wounds may seem impervious to all therapies (Figure 4). Many EB patients in the community are self-managing, either dressing their own wounds or having this done by a carer whom they direct, thus leaving day-to-day decisions with them. Discussion with the patient as to current wound management, and careful explanation of the rationale of any proposed new therapies, is essential to gain concordance. If the therapy fails it may be helpful to again discuss with the...
patient the methods employed while using the product, and whether indeed the product is being used at all. Infection may be present so repeat swabs may be required, while necrotic tissue (commonly slough) in the wound bed may prevent healing. Overgranulation is a frequent problem preventing the migration of new epithelial cells and can usually be resolved with the application of a topical steroid cream. Any non-healing wound in the severer forms of EB should be examined in a specialist centre because of the possibility of squamous cell carcinoma. Any underlying health issues compromising wound healing should be identified (White and Harding, 2006), and where possible treated.

Chronic wounds are often referred to as being stagnant in the inflammatory stage of wound healing (Moore, 2004), often due to infection or critical bacterial colonization, altered growth factors, high levels of protease enzymes and changes in lymphocyte and macrophage populations (White and Harding, 2006). Advanced wound care technologies can potentially ‘kick start’ the healing phase and a protease inhibitor (Promogran® Prisma® Matrix [Johnson and Johnson, Maidenhead]) has recently been used successfully in chronic wounds in EB (Pillay and Mather, 2007). Another successful approach to the recalcitrant EB wound has been honey (Hon, 2005).

**Dressing selection**

While there is an abundance of dressings available on the market, many hospital and community practices are restricted to a limited range of products. Due to the severe fragility of the EB skin only a limited number of dressings are suitable for use – those which reduce the risk of further trauma. Part of the duty of healthcare professionals is to advocate for the use of such products where appropriate, while being mindful of cost (Foster, 1990). The use of inappropriate dressings can cause further damage and be very costly for the patient in terms of pain and distress. Healing time may be further extended and the overall cost in real terms is far higher than if the correct dressing had been selected initially. While some dressings may cause damage on removal, others can cause damage in situ by, for example, the depth of the edge of the dressing causing a ridge that then causes blistering. Some patients use dressings prophylactically on areas of intact skin and frequently patients layer their dressings to prevent damage. Through lifelong experience, patients do have an awareness of ‘what works for them’, and can be reluctant and fearful to change.

With ongoing advances in wound care it is often possible to meet the needs of the patient by using one dressing rather than layering several products. Mepitel® (Mölnlycke, Dunstable), one of the firstatraumatic (safe tac) dressings to be produced, has a flexible polyamide net, coated with soft silicone that stays in place and does not adhere to the wound or peri-wound skin. The dressing can be left in place for up to 7 days, with the secondary dressing being changed as required (Atherton and Denyer, 2003). White (2005) has found in his product review that this dressing is easily removed without causing pain or trauma to the wound or surrounding skin. It can however be difficult to work with, especially when wearing sterile gloves. Therefore, it is useful to remember to moisten the fingertips of the gloves with saline or 50:50 to prevent Mepitel® from sticking to the gloves and allow more flexibility in the application of this dressing.

Another primary dressing commonly used in EB is Urgotul® (Urgo Ltd, Loughborough), which is a net dressing coated with a non-adherent lipidocolloid (with or without silver) (Meaume et al, 2004). Urgotul® may need to be changed every 2–3 days. However, it can be left in situ for up to 6–7 days after careful consideration and evaluation of the wound exudates and infection status by the practitioner (Meaume et al, 2002). The secondary dressing can be changed if any strike-through exudates is observed. Both dressings are very beneficial to EB skin and the choice is often patient preference regarding ease of wear and comfort (Mellerio et al, 2007). Both Urgotul® and Mepitel® require the use of secondary dressings in the form of simple absorbent pads and this can be difficult and time-consuming to apply.

More recently, dressings have been produced which combine non-adhesive qualities with an absorbent foam. These include Mepilex® (Mölnlycke) which is suitable for medium to heavy exudates. Mepilex® Lite is thinner than the Mepilex® and is more conformable to curved body surfaces, but anecdotal evidence suggests that it does not offer quite as much protection from pressure/trauma. It is intended for use with lower levels of exudates than Mepilex® (White, 2005). The Urgo range also offers dressings which combine non-adherent qualities with the ability to absorb varying levels of exudates (Fays et al, 2005). Other commonly used dressings are the hydrofibres (Aquadex® [ConvaTec, Uxbridge]) with other dressings being used for antimicrobial properties, and Aquacel® Ag along with Algivon® (Advancis, Kirkby-in-Ashfield) and Activon Tulle®. A sheet hydrogel (Actiform Cool® [Activa, Burton-upon-Trent]) has also been used both for protection and its anti-pruritic properties (Figure 5).

**Adhesives and adherent products**

It is inadvisable in any form of EB to use a dressing that is adhesive in any form (Schober-Flores, 2003). However, White (2005) advocates the use of Mepilex® Border Lite (Mölnlycke) for blisters or skin tears, but adhesives may be beneficial in difficult to bandage areas and when a secure dressing is required. This includes such items as electrocardiogram electrodes and intravenous lines where clinical need will dictate their use, and the need for secure fixation. The development of...
Appeel® (CliniMed, High Wycombe), an advanced silicone-based medical adhesive remover, has revolutionized the process of removing adhesive products in EB patients (Mather and Denyer, 2007) (Figure 6). If Appeel® is unavailable the adherent dressing can be soaked in an oil-based product such as 50/50 (white soft paraffin and liquid paraffin). The method of removal is also important with the product being ‘rolled back on itself’ to reduce the ‘pulling’ forces exerted. The patient may prefer to remove adhesive or adherent products themselves, while it may be possible to leave other products to come off naturally. A patch test of a small piece of dressing/tape for ease of removal is preferable with non-adhesive products to be first choice, followed by low-adherence, then adherent. In all cases, the patient will have firm ideas about what their skin can tolerate. It is important to carefully consider the method of securing dressings so as not to produce more traumatic injury to the skin while preventing the embarrassment that patients fear when dressings become displaced and leak, or worse, actually dropping off when they are in public. The movement of dressings can also lead to wounds adhering to clothing and bedding, which is both painful and unhelpful to the wound healing process. Tubular bandages, soft retention bandages and fitted garments, including sports and underwear, are often used with good effect.

**Pruritus**

Dermatologists report that pruritus (itch) is the second most frequent symptom presented to dermatologists by patients (Murray, 2007), and certainly in the case of EB patients it is a very challenging part of their care. After pain, itch is probably the most troublesome element of the condition as it is very difficult to control. It is important to minimize patient discomfort in order to avoid further trauma to the skin caused by scratching. Treatments include moisturizers (as previously discussed), topical honey, corticosteroids and antihistamines which tend to provide short-term relief only. Some EB patients have successfully been treated for pruritis with topical tacrolimus, including one patient suffering from a form of EB, known as EB pruriginosa, where ‘itch’ is intense and the most troublesome symptom (Banky et al, 2004).

**Squamous cell carcinoma**

Patients with RDEB have a cumulative risk of 76.5% by the age of 60 years, of developing a squamous cell carcinoma.
(SCC) (Mallipedi, 2002). It is extremely important that patients, carers and healthcare professionals are aware of the risk and observe any wounds for suspicious changes. These may include the wound failing to make progress in healing or have increased pain, or in some cases no pain. It may exophytic or hyperkeratotic. Patients often report that an area ‘feels different’ before anything is noted by the observers. Always listen to the patient and arrange for them to be seen by a dermatologist experienced in assessing EB skin. Any suspicious area should be biopsied as soon as possible. If an SCC is diagnosed, then it should be excised and skin graft applied. SCCs occur mainly in RDEB patients, aged 30–40 years of age, although it can sometimes affect patients in the younger age group (Mallipedi, 2002). For reasons not yet understood these SCCs behave in a far more aggressive manner than an SCC on non-EB skin. A patient may develop several primary growths over a period of years, any one of which can become metastatic. Retrospective statistics show a survival rate of 5 years from diagnosis of squamous cell carcinoma (Fine et al, 1999b).

**Care at the end-stage of life**

At the end of life, which is frequently after a very foreshortened life-span, the care of the EB patient can become very complex. Many patients develop fungating malignant wounds causing pain, odour and exudate management problems. The development of such wounds, with their sensory impact, both on the patient and on those around them, leads almost inevitably to psychological difficulties and the potential for the patient to become isolated (Grocott, 2007). Alongside the visible malignancies, metastatic spread can affect a variety of organs. Metastases have been seen in the lungs, liver and bones. In common with other conditions, the aim in this phase of life is to maintain the patient as pain-free as possible with minimum disturbances to carry out nursing care. Syringe drivers for the delivery of analgesia and other symptom management medication can be used in EB patients, and secured with Mepitac®. As the ‘driver’ site will potentially have to be changed every 2–3 days, Appeel® medical adhesive remover can be used to prevent skin damage. As the patients condition deteriorates, so may the skin, and dressing changes can be very distressing. Some patients in this phase of life have benefited from being nursed on a low-air-loss mattress (TheraKair Visio®) which assists both with comfort and pressure re-distribution and also management of wound exudate (Pillay and Hon, 2007). Odour management may be required and includes the use of charcoal dressings, silver products and deodorizers, or essential oils applied to clothing/bedding or vaporizer. In the case of urinary incontinence a small soft-silicone catheter can be inserted without the usual concern (in EB) of the provocation of urethral damage.

**Psychological aspects**

While many people with even the most severe forms of EB remain incredibly positive in their approach to life, from time to time the unremitting nature of the condition causes even the most stoic to be low in mood or clinically depressed. The psychological welfare of the patient should always be part of an holistic assessment and help sought accordingly.

**Family/carer support**

RDEB has huge implications for the lives and livelihoods of immediate family members, who often become the main carers for the patient. Parents may have to give up work and as a consequence make financial sacrifices. The genetic inheritance of this condition can lead to parents carrying a psychological burden of guilt. As children grow to adolescence they have many issues to deal with in addition to the normal teenage ones and at this time they often begin to search for ways to become independent of their family. Many EB patients are very determined to live a full life, and with the appropriate family and multidisciplinary support can be assisted to go to university, travel and live independently in their own accommodation. While this can be a very positive experience for both patient and parents, it can also be very hard for the main carer to relinquish their role and they do require understanding and support while they adjust.

**DebRA**

DebRA is a registered charity dedicated to finding a cure for epidermolysis bullosa. It funds major research into this while providing information, advice and practical help to those with the condition through a nursing and social care team. The DebRA nursing teams offer a service throughout the UK, and are part of the specialist multidisciplinary clinics in London, Birmingham and Scotland. DebRA nurses strongly believe in holistic individualized care, and not only recognize that patients with EB are individuals, but also that they are the ‘real experts’ in their care. They offer patients support while encouraging independence and acting as advocates. Home visits, nurse-led clinics, multidisciplinary case conferences, telephone support, patient group meetings and healthcare professionals meetings, training sessions and conferences, are all essential aspects of the DebRA nurse service. Alongside this is the support offered to families and carers and information and education of local healthcare teams. Patient education and information play a
part in the work of the DebRA nurses by offering patients information on developments and wound care products thereby enabling the patient to make informed choices. More information on the adult nursing and allied services and request for information on specific aspects of EB care and nurse contact details can be found on the website (www.debra.org.uk).

Conclusion

It is hoped that this article will have given healthcare professionals an insight into the complexities of caring for patients with EB. It can be very challenging, but not impossible, and is very rewarding when patients who are as complex as this are treated to, and benefit from, the highest standards of nursing care. Until the day comes when a cure can be found for this terrible condition, all that can be offered is support in management of the condition through good multidisciplinary team work. Advances in gene therapy and advanced wound care continue to bring relief and hope to those with EB. The dedication of carers and informed healthcare teams can help in the process of enhancing comfort and patient autonomy.

The next article in this series on EB focuses on the psychological wellbeing and care of patients with EB and their families.


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