



SKIN AND WOUND CARE

**Guidance for adults with epidermolysis bullosa (EB)
and their carers**

**This is how life feels
to people with EB.**



WHAT IS EPIDERMOLYSIS BULLOSA?

EB is a group of rare genetic disorders characterised by fragility of the skin and mucous membranes and mechanically induced blistering. EB comprises four main types - EB simplex (EBS), junctional EB (JEB), dystrophic EB (DEB), and Kindler EB (KEB), with more than 30 subtypes. EB is clinically heterogeneous including a broad spectrum of severity.

**Cláudia Lucien Cavalcante de Souza, living with recessive dystrophic EB, aged 21, Brazil
(photo credit: Guilherme Gobbi)**



Emanuel Kluge, living with recessive dystrophic EB, aged 18, Brazil (photo credit: Suelen Szymanski)

ABOUT THIS BOOKLET.....06

BLISTER MANAGEMENT08

WOUND TYPES10

DRESSINGS.....12

FACTORS AFFECTING WOUND HEALING13

PREVENTING INFECTION14

TREATING INFECTION17

ODOUR.....20

FACTORS AFFECTING HEALING & ACTIONS22

ITCH/PRURITUS26

SQUAMOUS CELL CARCINOMA28

GLOSSARY30

GET INVOLVED32

GENERAL INFORMATION34

CREDITS INFORMATION36

CONTACT INFORMATION38

NOTES.....39

NATIONAL DEBRA GROUPS42



Tauani Vieira (centre), living with recessive dystrophic EB, aged 24, Brazil (photo credit: Suelen Szymanski)

Foreword

For most adults living with EB, wound care becomes a way of life and they and their carers rapidly become experts in wound management. Professionals who are involved in looking after people with EB recognise that they work in partnership with expert patients and carers.

This booklet is a guide to general principles recognising that each person with EB is unique. The aim of this booklet is to equip people with EB with some basic knowledge so they can fully participate in managing their own condition.

This booklet is directed at adults (16+ years old) with more severe forms of EB, or who have one or two problematic wounds, and their carers. However, adults with less severe EB may find the general advice and instructions regarding dressings helpful. This is not intended to be a substitute for professional advice, but rather to ensure you are well informed.

Who is this booklet for?

This booklet is for adults (16+ years old) living with any of the following types of EB and their carers:

- ▶ EB simplex (EBS)
- ▶ dystrophic EB (DEB)
- ▶ junctional EB (JEB)
- ▶ Kindler EB (KEB)
- ▶ EB acquisita (EBA) - autoimmune, not genetic*
- ▶ *There was no evidence found in this population group for the CPG. However, it is assumed that they would require the same support.

What is this booklet about?

Topics covered in this booklet include:

- ▶ the fundamentals of wound care management
- ▶ dealing with infections
- ▶ factors impacting healing
- ▶ itch/pruritus
- ▶ squamous cell carcinoma (SCC)

Where does the information in this booklet come from?

The information and recommendations in this booklet are derived from the “International Consensus Best Practice Guidelines Skin and wound care in epidermolysis bullosa, an expert working group consensus”. The guidelines were written in 2017 by three EB clinical nurse specialists. The information and recommendations in the guidelines come from a variety of sources including clinical research and mainly expert opinion.

There are two different types of recommendations in this booklet:



RECOMMENDATION
based on research
evidence

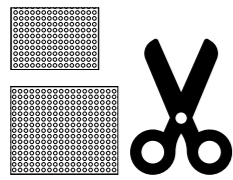


RECOMMENDATION
based on clinical
experience

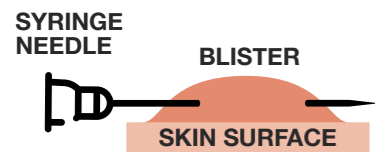


This icon signposts to recommendations that overlap with those from other EB clinical practice guidelines (CPGs). These are referenced to on page 35.

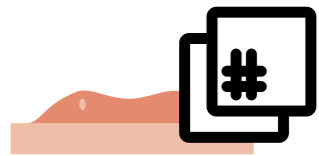
Bigger blisters = bigger wounds! Pop a blister as soon as you see it. Cleaning and removing debris/dead tissue helps to reduce bacteria in wounds and promote wound healing.



01 Prepare all dressing material in suitable sizes and amounts in advance to keep dressing changing times shorter



02 Use a clean needle to create an entry and exit hole so the fluid can escape



03 Roll a soft clean cloth over the blister to expel fluid



04 The roof should be left on the blister to protect the wound



05 Remove dead skin around the open wound

Managing blisters is one of the key areas of treating EB. It is recommended that blisters are lanced (burst) using a sterile needle or clean scissors. This should be at the lowest point of the blister so that gravity can help drain the fluid away. Pressure is then gently applied to the blister with clean gauze to drain the fluid. The aim is to prevent the blister from spreading and to leave an opening large enough to stop the blister from re-sealing and re-forming. If possible, the roof of the blister should be left intact in order to protect the skin underneath – this provides the best possible dressing for the wound. The roof should not be removed as this will cause additional pain and leave an open area exposed to infection. Some people prefer to use a syringe to remove fluid from blisters and this is perfectly acceptable.

Recommendation

Blisters are not self-limiting and intact blisters should be lanced and drained.



There are two types of wounds:

Acute wounds are uncomplicated and heal quickly.

Chronic wounds are more problematic and can last for months or years and need special attention.

There are many reasons why wounds don't heal. Ideally, chronic wounds should be reviewed by your EB team/healthcare provider as soon as you feel there is a problem. The sooner a problem wound is assessed, the greater the chance there is of it healing. Chronic wounds are also wounds that heal and break down again.

Non-healing wounds

A non-healing wound is one that does not heal as would normally be the case for you. Talk to your EB team/healthcare provider to try to find the reason why the wound isn't healing. It may be that your current dressing regime is not providing the best healing environment for the wound or infection is present.

Recommendation

Try dressings that can assist with healing:

- ▶ polyurethane foam
- ▶ medical grade honey dressings
- ▶ dressings that change the biochemistry, such as protease inhibitors

Be aware that a non-healing wound in the more severe forms of EB may be a squamous cell carcinoma (SCC). See page 29 for information on SCC.

Painful wounds

The most important thing is to work out why the wound is so painful; it may be infected or you may be using an inappropriate dressing.

- ▶ Wound pain can be managed with oral pain killers. Seek advice from your EB team/healthcare provider where possible.
- ▶ There is a dressing available that contains ibuprofen. Care needs to be taken to ensure that the anti-inflammatory effects of the ibuprofen do not mask signs of infection. It is also important to calculate your total daily dose if you are also taking an oral anti-inflammatory drug to ensure you are not overdosing.
- ▶ Small doses of topical morphine can have a good effect on wound pain without the side effects of oral morphine. Morphine mixed with a hydrogel is placed on the dressing or wound. Discuss this with your EB team/healthcare professional as this must be prescribed and monitored.

Heavy exuding wounds

These are wounds that produce exudate (an excessive amount of fluid). Discuss with your EB team/healthcare provider how to treat the cause and to find suitable dressing options. Super absorbers are available for extremely heavy exudate, which must be used over a non-adherent (non-stick) primary dressing.

Crust around wounds

Crust can slow down healing and should be removed where possible.

- ▶ A greasy moisturiser applied to the crust during a dressing change can soften it and make it easy to remove at the next dressing change.
- ▶ A soft debridement pad can help remove the crust.
- ▶ Some topical wound gels can help lift the crust.

Sloughy wounds

Slough is a sticky, thick, gluey coating of the wound bed and is not easily removed. Slough on wounds will slow down healing and should be removed.

Recommendation

There are a range of dressings and topical treatments that can be used to help break down slough:

- ▶ occlusive dressings
- ▶ foam dressings
- ▶ topical gels that liquefy the slough
- ▶ medical grade honey
- ▶ larvae (sterile medical grade maggots) therapy

A dressing on its own will not heal a wound. However, if it is the right dressing, it will be invaluable in providing the best environment to enable your body to do its work of healing. There are certain common characteristics that make some dressings more suitable than others.



Top tips for the ideal dressing in EB

The ideal dressing should:

- ▶ be acceptable to you as the person with EB and your carer(s)
- ▶ provide a suitable environment to encourage wound healing
- ▶ be non-adherent to the wound bed and surrounding skin
- ▶ be pain free on removal
- ▶ reduce the need for frequent dressing changes
- ▶ be able to absorb exudate
- ▶ be comfortable and remain in place
- ▶ be easy to use
- ▶ be easily available
- ▶ be available in a variety of sizes.

Dressing selection is based on the state of the wound and what you need to achieve. You will likely have dressings that you use regularly. However, if you have a wound complication, you will need to have the wound reviewed by your EB team/healthcare provider. Always ask for information about any new wound treatment so that you understand what it is and why it is recommended.

We recognise that some people will have limited access to wound care products or products that change rapidly in the market, therefore advice should be sought locally.

What to do when using a new dressing

- ▶ Discuss thoroughly with your EB team/healthcare provider what to expect with a new dressing/wound treatment.
- ▶ Get written instructions.
- ▶ Be clear how often to expect to change the dressing.
- ▶ Be clear when the new treatment should be reviewed.
- ▶ Some dressings may make things worse before they get better, such as if the aim is to remove slough, the wound may initially appear to get bigger. Ask your EB team/healthcare provider if this is likely to happen.

No dressing will make an immediate impact on a wound and generally you should expect to use it for two weeks before you see any improvement. Please talk to the person who recommended the dressing before consigning it to the back of the cupboard or the bin!

There are many factors that affect wound healing:



Nutrition

Wound healing depends on you being able to follow a well-balanced diet. This is particularly true in EB as there are many wounds to be healed. Nutrients including protein and some vitamins are lost in wound fluid and need to be replaced with supplements. Where possible, you should be regularly assessed by a dietitian.



Anaemia

Many people with EB are anaemic, which can cause poor wound healing. A diet rich in iron is recommended but additional supplements are often required. Your EB dietician or healthcare provider can help with this.



Other factors

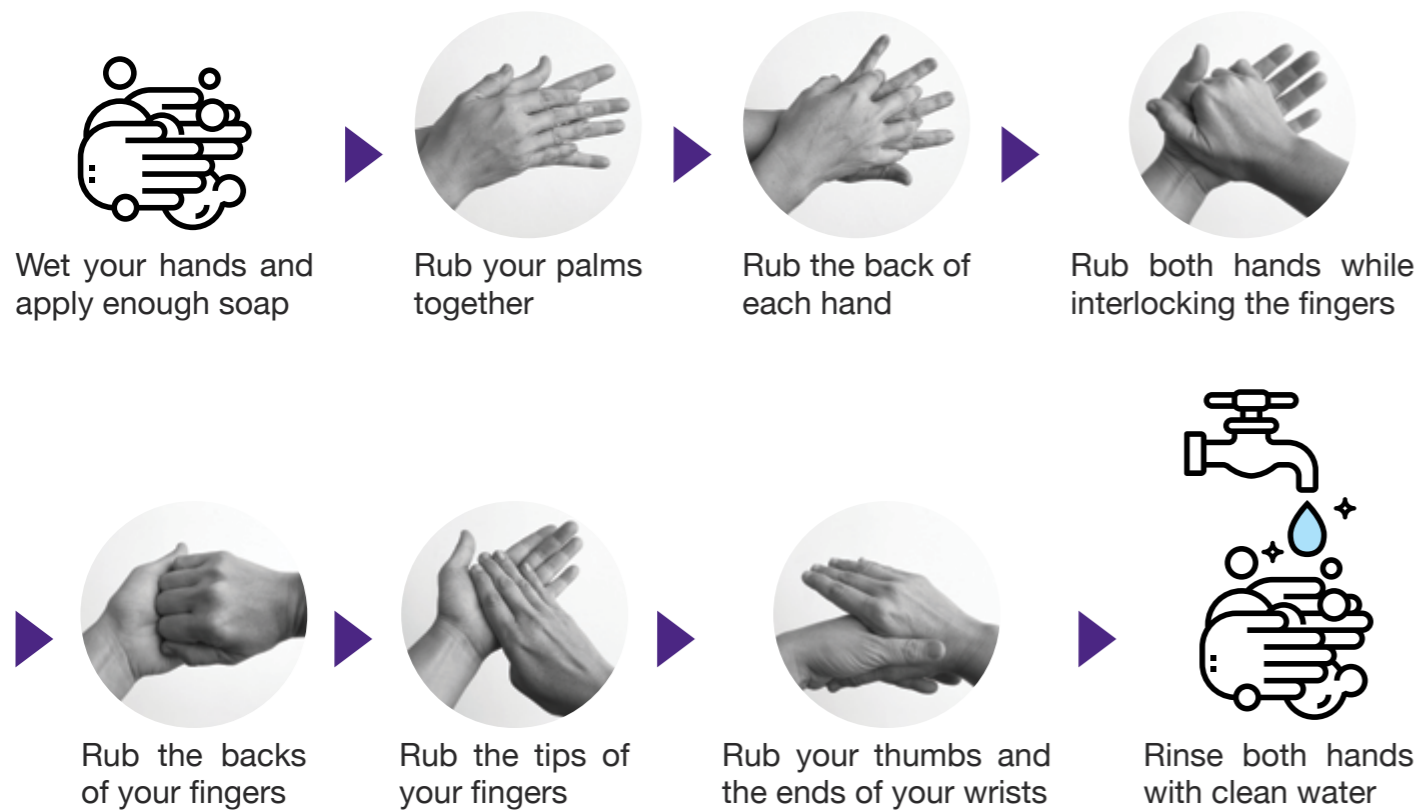
Stress, pain, and lack of sleep can all result from a non-healing wound (or wounds) and impact negatively on your body's attempt to heal itself. These symptoms may be improved so talk to your EB team/healthcare provider about this where possible.



Other infections

If you have an illness, such as flu you may notice that your blister sites do not heal as quickly. The rise in body temperature may also lead to new blisters forming more readily so keep as cool as possible.

Hands should be washed thoroughly before and after dressing changes.



You should aim to make dressing changes as clean as possible. This is particularly important when you are in hospital where cross infection is a constant hazard.

Before dealing with blisters and wounds, wash your hands and encourage others to do the same (healthcare staff included). Hands should be re-washed between removal of used dressings and applying new ones. One of the most common causes of infection is poor hand washing. Caregivers may choose to use gloves in order to reduce the risk of infection, but they should still wash their hands.

Try to prepare a clean 'field' to lay out everything you will need for a dressing change, such as a clean or sterile towel from a dressings pack.

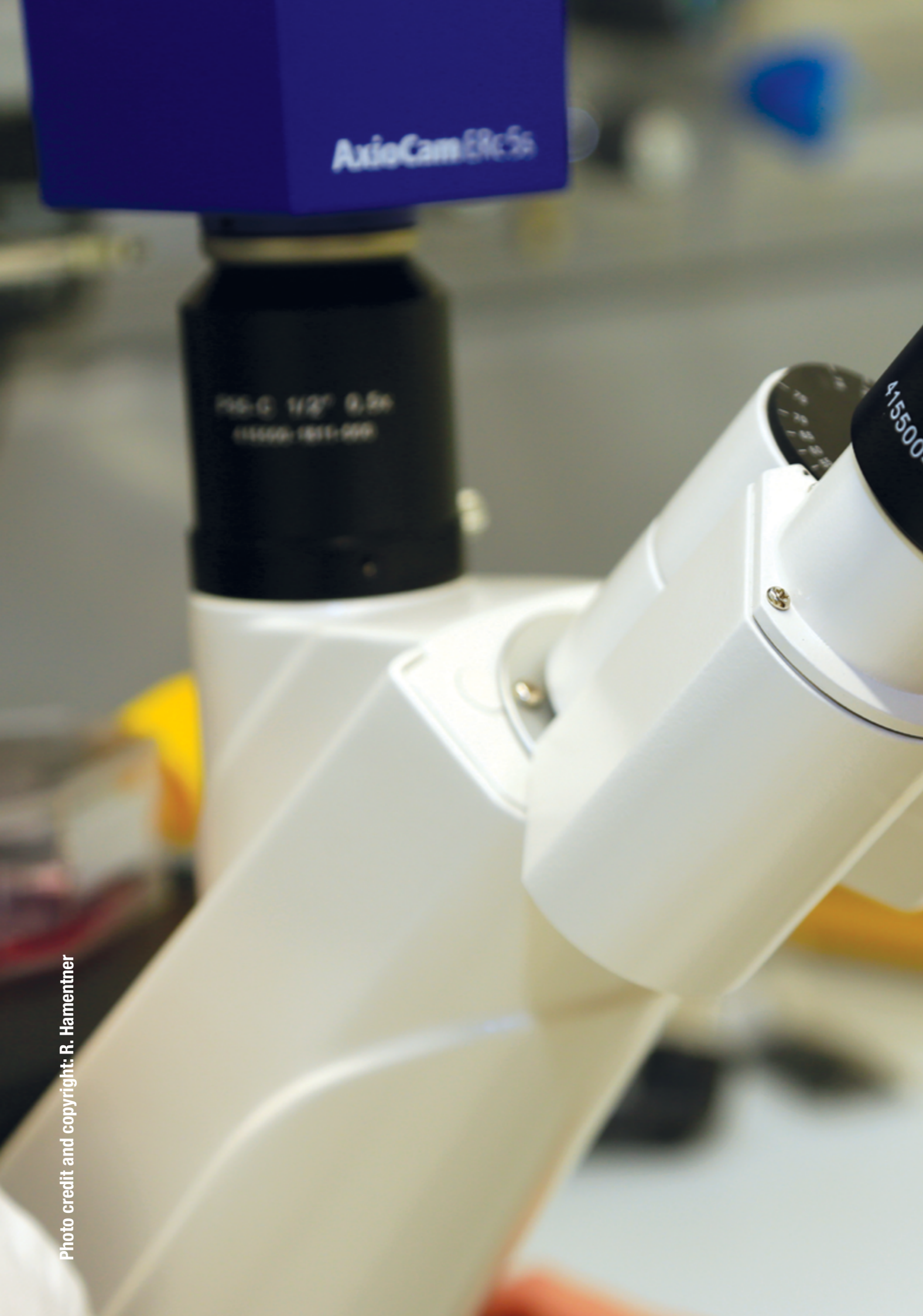
If you bathe or shower at dressing changes, please ensure that the bath/shower is clean; this is particularly important whilst you are in hospital where the bath/shower should be cleaned before and after you use it. This also applies to equipment such as bath seats. Cracks in equipment and baths will harbour bacteria and will pose an infection risk. Equipment in this state should be repaired or replaced.

If dressings are removed from their sterile pack and not fully used, you should store any remainder in a clean container; this also applies to dressings that you prepare in advance.

If you are using creams, ointments, etc. transfer what you need into a small pot to avoid transferring bacteria from the wound to the entire pot of cream.

Immediately dispose of any 'dirty' dressings into a disposal bag.

Change dressings on infected wounds separately and wash hands again before changing any further dressings.



This is a common problem, particularly in chronic wounds. Not all wounds will present with the same symptoms; however, it is important to seek diagnosis and treatment for a suspected wound infection. This is particularly important if you suspect the infection is spreading as antibiotics may be required. Due to the increased occurrence of antibiotic resistant bacteria, use of antibiotic therapy should only be taken on the advice of a certified health professional as recommended by the World Health Organisation (WHO). Antibacterial resistance happens if the bacteria are regularly exposed to antibiotics. The bacteria ‘learns’ how the antibiotic works and develops defence mechanisms, this is why we have so called ‘superbugs’ such as MRSA. Very few new antibiotics are being developed and we need to work hard to preserve the effectiveness of the antibiotics we currently have, using them only when really absolutely necessary.

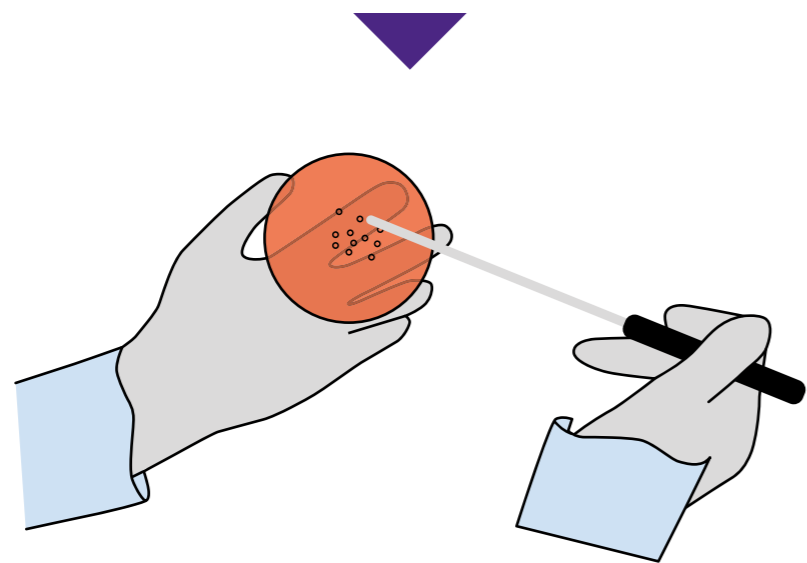
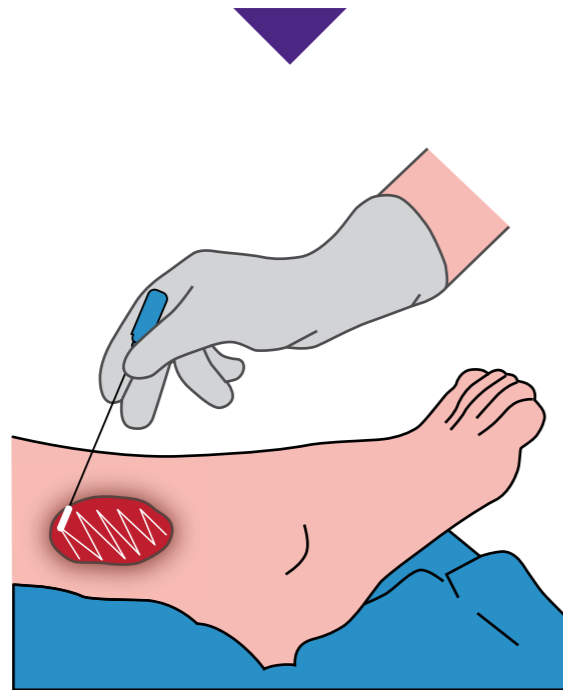
Local infection

This happens when your natural defences have been overcome and the bacteria are multiplying quickly in the wound, and symptoms begin to appear. In some chronic wounds the symptoms can be hard to detect.

Symptoms of infection can be some or all of the following:

- ▶ increased exudate which may be thicker than normal and/or bloodstained
- ▶ increased pain or change in the type of pain e.g. aching becomes ‘burning’ pain
- ▶ fiery red, fragile granulation tissue
- ▶ abnormal odour
- ▶ heat
- ▶ swelling
- ▶ delayed healing
- ▶ redness around the wound

A wound swab to identify the type of infection may be indicated (see page 19). If infection is present you may need to treat this with topical antimicrobials, dressings with antimicrobial properties, and/or antibiotics.



Spreading infection (Cellulitis)

This happens when the infection starts to spread into the tissues around the wound. This needs immediate treatment with antibiotics. Wound swabs should always be taken in order that the most effective treatment can be given.

Symptoms can include:

- ▶ redness spreading into the local tissues more than 2cms from the wound margins
- ▶ heat and swelling
- ▶ wound getting bigger and deeper
- ▶ increased exudate, which is infected
- ▶ feeling unwell with a raised temperature.

Wound swabs

Wound swabs taken in the absence of symptoms of infection are not helpful. If you have the symptoms outlined above, then you should have a wound swab correctly taken in order to identify the bacteria that are causing the infection. The correct way to take a swab is to clean the wound bed of exudate and debris and then gently roll the swab across the wound bed in a zig zag pattern.

Treatment of infection

Products generally recommended are:

- ▶ topical antimicrobials (gels and creams) in discussion with your healthcare professional:
 - alginate gel
 - octenidine wound irrigation and gel
 - PHMB irrigation and gel
 - medical grade honey
- ▶ antimicrobial dressings:
 - polymeric membrane foam
 - dialkylcarbamoil chloride (DACC)
 - silver

SILVER PRODUCTS SHOULD BE USED FOR A PERIOD OF 2 WEEKS ONLY BECAUSE OF THE RISK OF ABSORPTION WHICH CAN BE HARMFUL.

- ▶ Antibiotics should **only** be taken if the infection is causing a clinical problem or the infection is spreading from the wound.
- ▶ Antibiotics should **only** be taken on the advice of a health care professional.



“Effective wound care is of course mandatory. It is really important to learn to do it well and adapt it to EB type and age (of the individual person).”

Anja Diem, Senior Physician, EB-Haus, Austria

Odour is a common wound care problem. This can be increased if the wound is infected or if you are unable to change your dressings as frequently as needed. Sometimes wounds can have an odour even when there is no infection and the dressings not been left in place for too long. This is particularly so in hot weather and in small confined spaces and is due to the nature of wound exudate.

There are a number of measures to tackle this problem:

- ▶ Change dressings and cleanse wounds by showering/bathing if you can tolerate this. If not, pour or gently squirt water onto the wound using a syringe, or gently place a moist cloth on the wound. It is also important to cleanse the skin around the wound.
- ▶ Change dressings as required and those where exudate is leaking.
- ▶ Use a topical product such as a honey ointment, which has proven effects against odour.
- ▶ Dressings containing charcoal can reduce odour for a time.
- ▶ Metronidazole gel can be used on wounds where a type of bacteria called an anaerobe is present. These types of bacteria produce particularly smelly compounds in your wounds and should be treated. Metronidazole should only be used on the advice of your healthcare professional.
- ▶ Treat all infected wounds if they are causing symptoms i.e. odour, pain etc. (see section on infection above).

Wound-related factors that affect healing and what to do when things go wrong

Factors affecting wound healing	Description of problem	Actions to take
Wet or dry wound bed	Wounds heal more quickly if moist.	<ul style="list-style-type: none"> ▶ Use a dressing that promotes moist wound healing. ▶ Ensure dressing does not absorb so much exudate that the wound becomes dry, or does not absorb enough so the wound is left 'soggy'.
Poor blood supply	Poor blood supply to the wound bed will slow healing.	<ul style="list-style-type: none"> ▶ Gentle exercise will increase blood flow. For wounds on the lower legs sit with legs elevated where possible. ▶ Avoid tight bandages. ▶ Reducing or stopping smoking will help to improve the efficiency of the circulation.
Temperature	<ul style="list-style-type: none"> ▶ Cooling of the wound bed will affect the activity of healing cells. ▶ During dressing changes, the temperature of the wound falls which could delay healing for several hours until the wound returns to normal body temperature. 	<ul style="list-style-type: none"> ▶ Change dressings in a warm, but not overly hot, room. ▶ Change dressings as quickly as possible by ensuring all equipment and dressings are prepared in advance. Instruct person helping you what is required before starting dressing change.
Wound location	<ul style="list-style-type: none"> ▶ Trauma - wounds on joints or those subject to repeated trauma, such as on the hands take longer to heal. ▶ Pressure - areas subject to pressure e.g. back, buttocks, and backs of the legs may take longer to heal. ▶ Clothing or tight bandaging can cause pressure on wounds which may cause further damage. 	<ul style="list-style-type: none"> ▶ Pad areas as necessary. ▶ Move regularly. ▶ Use pressure relieving mattress or cushion.

Factors affecting wound healing	Description of problem	Actions to take
Over-granulation	<ul style="list-style-type: none"> ▶ This is very common in EB and can be recognised when the red granulation tissue of the wound bed rises above the surrounding healthy skin. ▶ Over granulated wounds will not heal, as new skin cells cannot grow across the surface of the wound (they don't climb hills!) 	<ul style="list-style-type: none"> ▶ A fine mesh or foam dressing may limit the overgrowth of granulation tissue through the holes in the dressing. ▶ Apply a steroid cream to the wound for a few days only. ▶ If these actions do not resolve the problem, further investigation may be necessary. ▶ Poorly controlled exudate or infection may contribute to over-granulation in a wound and should be treated.
Foreign bodies	<ul style="list-style-type: none"> ▶ Any foreign body in the wound will provoke an inflammatory response. This will prolong the healing time and foreign bodies should be removed wherever possible. ▶ A common foreign body in EB is hair, particularly in wounds around the neck region. ▶ Dressing residue will also provoke the same response. 	<ul style="list-style-type: none"> ▶ Avoid dressings that shed loose threads into the wound and gently remove any dressing residue by irrigating the wound with water. ▶ If you have long hair pin it up when attending to wounds in the neck, shoulder and back region.
Dead tissue in the wound	<ul style="list-style-type: none"> ▶ This can be black necrotic tissue, or more commonly in EB is a yellow-creamy coating (slough) of the wound bed that is not easily removed from the wound. ▶ Slough will need to be removed before the wound can heal. ▶ Slough can also act as a focus for infection. 	<ul style="list-style-type: none"> ▶ Certain dressings, or topical products such as a hydrogel or those containing medical grade honey are used to remove slough from the wound. ▶ Slough can also be gently removed using a soft gauze or debridement pad.



Factors affecting wound healing	Description of problem	Actions to take
Substances toxic to new cells	These can be strong antiseptic solutions, some over the counter treatments and creams or long-term use of steroid cream.	<ul style="list-style-type: none"> ▶ Only use new products after you have discussed with your healthcare professional where possible. ▶ These products should be used with caution.
Dressings that 'stick' to the wound	<ul style="list-style-type: none"> ▶ This will remove healing cells. ▶ Dressings should be easy to remove. If they are stuck it may be necessary to soak them prior to removal. 	<ul style="list-style-type: none"> ▶ Use non-adherent wound care products. ▶ Use water, 50/50 (50% white soft paraffin 50% liquid paraffin) or medical adhesive removal spray (MARS) to remove dressings that are stuck. ▶ Lift the edge of the dressing gently, roll back on itself and apply the water, 50/50 or MARS.
Excess exudate	<ul style="list-style-type: none"> ▶ Excess exudate is a common problem in wounds that are chronic and/or infected. ▶ Chronic wound exudate is extremely alkaline and can be destructive to the surrounding skin. Extremely alkaline solutions have a similar destructive effect to very acid solutions. 	<ul style="list-style-type: none"> ▶ Use an absorbent dressing and change as frequently as required. ▶ Protect the fragile surrounding skin using a barrier cream or spray. ▶ See section on maceration/excoriation.
Wound infection	See section above on infection	
Maceration	<ul style="list-style-type: none"> ▶ Maceration is due to the effect of excess wound exudate on the good skin around a wound. ▶ The skin becomes white (a bit like when you have spent too long in the bath) and will eventually break down thereby extending the wound. 	<ul style="list-style-type: none"> ▶ Change the secondary absorbent dressing to one which is capable of holding more exudate. ▶ Change the dressing more frequently. ▶ Protect the good skin with a barrier product. ▶ Correct the cause of the extra exudate e.g. infection.



Geanyne Ferreira Stüpp, living with recessive dystrophic EB, aged 22, Brazil (photo credit: Suelen Szymanski)

This is one of the biggest problems in EB as severe itch followed by scratching causes many new blisters and skin trauma, and leads to the breakdown of nearly healed wounds. The most severe itching tends to be found in EB Pruriginosa, which means 'itchy EB'. Some of the most useful advice is adapted from the UK National Eczema Society.

What makes itching worse?

Temperature and sweating

Any change in temperature can induce itch. Some people with severe EB do not sweat effectively due to scarring of the sweat glands, and the body's ability to cool itself is therefore reduced. Suggestions to help you keep cool include:

- ▶ Consider lightweight adjustable bedding.
- ▶ Wear light, loose clothing made of natural fibres where possible. Dress in layers so that clothes can be taken on and off as needed.
- ▶ Avoid hot baths.
- ▶ A portable air conditioning unit may be a worthwhile investment for the hot summer months.

Healing wounds

All wounds tend to be itchier as they heal.

Opiate/opioid analgesia

Some opiates/opioids, such as products containing morphine may increase itch.

Washing clothes

Consider using a hypoallergenic washing product and avoid strongly perfumed items. Be aware that it can take some time to be sensitised to a product, and it might be a while before your itch increases after you introduce a new product.

Soaps and skin creams

Use gentle, fragrance free products wherever possible.

Emotional factors

Stress may increase itch. Stress management strategies such as relaxation exercises or meditation may help.



Practical tips

- ▶ Moisturise your skin frequently. Many people with EB say they have to find a balance between soft skin and skin so soft it blisters more easily.
- ▶ Where possible apply moisturisers in smooth downward strokes in the direction of hair growth.
- ▶ After bathing pat the skin dry with a soft towel. Some people find it helpful to apply moisturisers when the skin is still damp, to 'seal the moisture in'.
- ▶ A substance contained in oats reduces itching and you can add this to your bath by putting porridge oats in a sock and tying the end – an 'oaty' bath bomb.
- ▶ Keep your nails (if you have them) short and smooth.
- ▶ If the urge to scratch is overwhelming, try and distract yourself.
- ▶ You may be able to substitute scratching by pressing on the skin or blowing on it.
- ▶ If you absolutely have to scratch, then try using one of the massage devices which have rounded ends which go on the skin.
- ▶ Cooling the skin, with something like a bag of frozen peas may help. Place the peas in fabric such as a pillowcase to prevent freezer 'burn'.
- ▶ Apply moisturisers that have been kept in the fridge.
- ▶ Hydrogel sheet dressings on very itchy areas can be helpful.
- ▶ Try to dress in layers so that clothing can be removed if you are hot.
- ▶ If your iron levels are low (as is common in severe EB) you may be more 'itchy'. A diet rich in iron and oral iron supplements may help. Regular blood tests will diagnose if your iron levels are very low which may need treating with an intravenous infusion.



“ In November 2014, a small wound between my right thumb and index finger would not heal. Given that I have dystrophic epidermolysis bullosa (DEB), this obviously happens frequently. However, the wound was also causing a stabbing pain. That worried me. I went to the hospital, but the treatment using creams, special bandages, and drugs did not help. Quite the contrary, the wound and the pain got worse. I returned to the hospital in late December. The dermatologist took a tissue sample and had it examined under a microscope. The result of the biopsy was not good: the tissue contained malignant cells, I had squamous cell cancer. ”

Stief Dirckx, born with recessive dystrophic EB, (1978-2018), Belgium

This is a common complication of severe EB. SCCs can develop anywhere but are most commonly found in areas where the skin is frequently damaged e.g. ankles, knees, hands, and on sites of chronic blistering and scarring.

Many people with EB who have had a cancer have recognised that there was a problem and brought it to the attention of their healthcare professionals. Things to watch out for are:

- ▶ wounds that don't heal as they would normally
- ▶ wounds that 'feel' different
- ▶ wounds that behave differently, or are unusual for you
- ▶ a rapidly growing wound especially if it is raised or cauliflower like
- ▶ wounds that are deeply 'punched out' especially with raised or rolled edges
- ▶ wounds that have a thick raised crust or crusts that do not lift
- ▶ wounds that have altered sensation different to your normal EB wounds. This may be a feeling such as tingling, increased pain or no pain at all
- ▶ wounds that have no feeling.

If you have an area you are concerned about, contact your healthcare professional and they will be able to organise an urgent appointment for you. A number of small biopsies may be taken. If they prove to be cancer it will be surgically removed. As EB skin looks so 'different' under the microscope, you need to be seen and treated in a centre that offers a specialist service.

These are some of the terms you may hear a healthcare professional using.

Biofilms

Biofilms can be found in chronic wounds but are common throughout nature. A biofilm we are probably all familiar with is dental plaque. The biofilm contains many types of bacteria living as a community which have encased themselves in a type of protective slime. This makes them difficult to identify with a wound swab and also to treat effectively. It is not possible to see a biofilm but one should be suspected if the wounds are not responding to appropriate treatment.

Effective treatments for biofilms can include:

- ▶ PHMB products
- ▶ octenidine products
- ▶ debridement pads (this can be difficult in EB wounds because of pain)

Epithelialisation

This is good news as it occurs when new skin is being formed either around the margins of the wound or as islands – both are usually visible in a healing wound. Epithelialisation can be recognised as either small white islands on the surface of the red granulation tissue, or as a pinky/white margin to the wound. This new skin is very fragile and you should care for it accordingly. Itching can occur at this point and you should talk to your health care providers if it is an overwhelming problem.

Excoriation

This refers to the red dry area that can develop around a wound and can be caused by the effects of exudate on good skin. The effects of dressing adhesives and some creams and gels (topical) can also cause excoriation. Soft silicone and lipidocolloid wound care products can protect the good skin around the wound, as can barrier products. If wound exudate is improperly controlled, you will need to consider if you are using the right dressing (see section on maceration for more advice).

Exudate

This is the fluid that comes out of the wound. In a healthy wound it is normally straw coloured clear fluid. Generally, exudate encourages wound healing as it provides nutrients and growth factors. It also allows for the migration of new cells across the wound. However, it is important to get the balance of moisture in the wound correct. It should neither be too wet nor too dry, but moist. Good dressing choice is the primary factor in achieving this balance. Exudate can also tell us something of the state of the wound; offensive exudate or sudden increase of exudate can indicate the wound is infected, along with other symptoms (see section on wound infection).

Granulation

This refers to the red tissue base to deeper wounds. It will not normally be seen in superficial blister sites. The granulation tissue should be red and can be seen as the ‘factory’ that manufactures new tissue to fill the wound. This provides a base over which epithelialisation can occur. Over-granulation is a common problem in EB and can be recognised when the red base of the wound is raised above the surrounding skin. Over-granulation will need to be reduced as new skin cells will be unable to migrate across the wound. If the granulation tissue becomes dark red or bluish, bleeds easily, or is very fragile it can indicate a wound infection. Contact your healthcare professional for advice.

Necrotic tissue

This is dead tissue that must be removed from the wound bed to allow healing to take place. It will often have a black appearance.

Over-granulation

See above. Discuss using a steroid cream with your healthcare professional.

Unhealthy granulation tissue

Unhealthy granulation tissue can suggest infection and the possibility of a squamous cell carcinoma may need to be ruled out.

Slough

This is a sticky, thick, gluey coating of the wound bed. Unlike pus this can be difficult to remove. It is made up of dead tissue and will need to be removed before the wound can heal. Slough can also act as a focus for infection.

Tell us what you think

Have your say in the future patient versions of clinical practice guidelines (CPGs) for epidermolysis bullosa (EB)

The purposes of this survey are to:

- ▶ assess the quality of the information, presentation, and delivery of the patient versions
- ▶ help us to develop a standard for all patient versions now and in the future.

The data collected will help us to improve the information provided and experience of the user in all future CPG patient versions. The data may be used to report the development steps taken to improve their quality; this may be done through conference presentations, posters, abstracts, or studies.

We want to make sure that all patient information provided meets the needs of everyone living with EB.

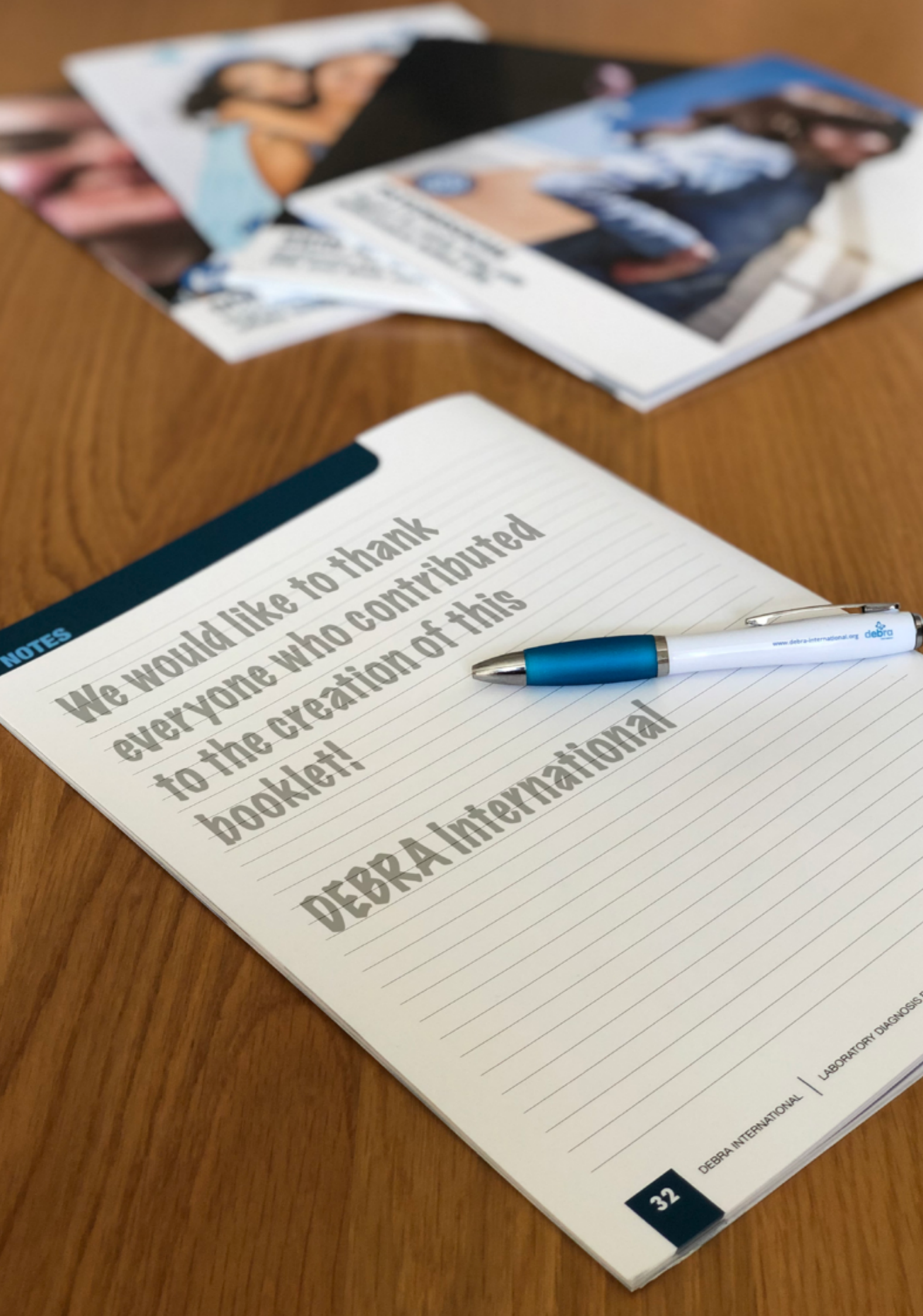
Help us create new CPGs and patient versions

All responses to the above survey are confidential unless you decide to join the DEBRA International CPG network. Please consider joining the network if you are interested in becoming involved in the development of CPGs and patient versions in the future. To join the network, please complete question 1 on page 7. If you do not complete this question, we will not receive any of your personal details and you will remain anonymous. Joining the CPG network is entirely voluntary and you may choose to opt out at any time by contacting DEBRA International.

If you have any questions when completing this survey or about joining the CPG network, please contact the DEBRA International CPG Coordinator, Kattya Mayre-Chilton by email at: kattya.mayre-chilton@debra-international.org

TELL US WHAT YOU THINK!

Answer the survey to help us improve the patient versions of the CPGs:
www.surveymhero.com/c/PatientVersionsSurvey





Emanuel Kluge, living with recessive dystrophic EB, aged 18, Brazil (photo credit: Suelen Szymanski)

Disclaimer

The information contained in this booklet does not indicate an exclusive course of action or serve as a standard of medical care. Variations, taking individual circumstances into account, may be appropriate. The authors of this booklet have made considerable effort to ensure that the information contained within accurately reflects the content of the guidelines on which it is based. The authors, DEBRA Sweden, and DEBRA International accept no responsibility for any inaccuracies, information perceived as misleading, or the success of any recommendations, advice, or suggestions detailed in this booklet. The information provided on the following pages is subject to change without notice. For the most up-to-date information on available clinical practice guidelines, booklets, and contact information, please visit: www.debra-international.org

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Development source

International Consensus Best Practice Guidelines for Skin and Wound Care in Epidermolysis Bullosa 2017

This is not a DEBRA International guideline but was developed in close association with DEBRA International.

This patient version was approved by DEBRA International and funded by DEBRA Sweden.

To access the following CPGs and patient version booklets, please visit: www.debra-international.org

Other CPG topics

- ▶ Clinical Practice Guidelines for Epidermolysis Bullosa Laboratory Diagnosis
- ▶ Foot care in Epidermolysis bullosa: Evidence-based Guideline
- ▶ Management of Cutaneous Squamous Cell Carcinoma in Patients with Epidermolysis Bullosa: Best Clinical Practice Guidelines
- ▶ Occupational therapy for epidermolysis bullosa: clinical practice guidelines
- ▶ Oral Health Care for Patients with Epidermolysis Bullosa - Best Clinical Practice Guidelines
- ▶ Pain care for patients with epidermolysis bullosa: Best care practice guidelines
- ▶ Psychosocial recommendations for the care of children and adults with epidermolysis bullosa and their family: evidence based guidelines

Other languages

We are happy to consider requests for this booklet to be provided in other languages. Please send all enquiries to: office@debra-international.org

How was the Skin and wound care guideline produced?

- ▶ The CPG development was led by specialist EB nurses and reviewed by an expert panel of clinicians from Denmark, the United States, India, and the United Kingdom; guideline methodologists from the United Kingdom and Ireland; and international EB teams from Great Ormond Street Hospital, London; St Thomas' Hospital, London; Birmingham Children's Hospital, Heartlands Hospital, Birmingham; DEBRA Ireland; and DEBRA International.
- ▶ A systematic literature search was undertaken to provide further evidence for recommendations.
- ▶ The information was supplemented by day-to-day experience of people living with EB and their carers' testimonials.
- ▶ A total of 601 articles were identified. Finally, 36 papers were appraised, each by two panel members, according to the Critical Appraisal Skills Programme (CASP) and Scottish Intercollegiate Guidelines Network (SIGN) quality rating.
- ▶ The majority of the papers were graded level 3, being small-scale case studies with many others being level 4, i.e. expert opinions.
- ▶ Knowledge of good wound care practice, the evidence presented here, and experience among professionals, patients, and carers provides a substantial body of evidence to support current wound care practice in EB.

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