

# Epidermolysis bullosa simplex (EBS)

This leaflet explains more about EBS. If you have any questions or concerns, please speak to a doctor or nurse caring for you.

## What is EBS?

Epidermolysis bullosa (EB) is a rare, life-long, genetic skin-blistering disorder. Blistering can happen because of minor trauma (gently banging into something) or friction (rubbing). Sometimes they can just appear with no reason.

EBS is one of the less severe forms of EB, and is defined by the blistering in the top layer of the skin (epidermis). It usually does not cause scarring.

EBS is usually caused by a defect in keratin genes, specifically keratin 5 or 14. Keratin 5 and 14 produce proteins which form the scaffolding in the lowest layer of skin. If the protein is absent or defective, the scaffolding will collapse and the skin will separate, causing blistering. Minor friction to the skin, trauma or increased heat and humidity can cause skin blistering in EBS. The summer months can make symptoms worse.

## Inheritance patterns in EB

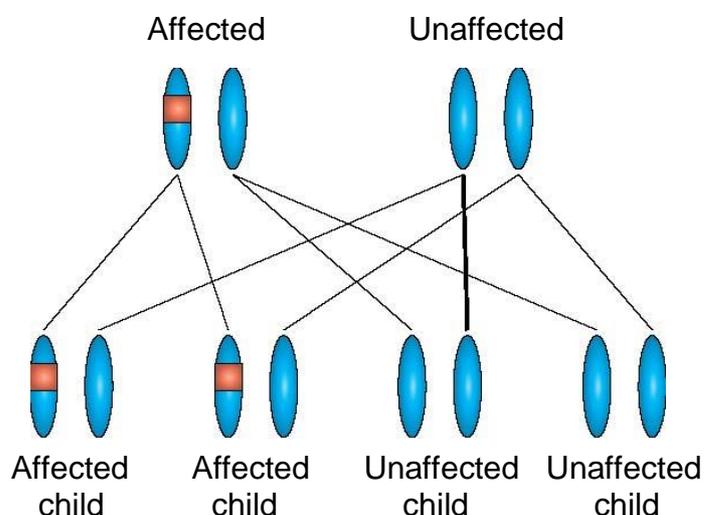
There are two types of genetic inheritance which are important in EB. These are called dominant and recessive. Most simplex cases are dominantly inherited.

Everyone has two copies of every gene - one inherited from their father and one from their mother. Sometimes, an affected child can be born to unaffected parents. This is what is known as a 'new mutation'.

### Autosomal dominant inheritance

The diagram below shows two gene pairs, one from a person affected with a dominantly inherited disease and the other is from an unaffected person. With a dominant inheritance pattern there is a one in 2 (50%) chance of passing EB on, in each pregnancy.

 = Altered gene



## Common types of EBS

There are four main types of EB: EB simplex, junctional EB, dystrophic EB, and Kindler EB. In each type of EB there are several sub-types.

The names given to the sub-types of EB have been re-named to make it easier to tell the differences in each type and sub-type of EB.

- Localised (previously Weber-Cockayne)
- Intermediate (previously Köbner)
- Severe (previously Dowling-Meara).

### EBS localised

This is the most common form of EBS. It usually starts in early childhood but sometimes it may not cause symptoms until later childhood or adulthood. Blistering generally occurs on the hands and feet, which are areas most prone to friction and trauma.

- Thick-walled blisters usually occur on the hands and feet after walking or physical activities. Increased sweating (hyperhidrosis) of the palms and soles is common and will increase the tendency to blister
- Thickening of the skin of the palms and soles is normal (callus or hyperkeratosis) and blisters occurring under this hard skin can be painful and hard to manage.

### EBS intermediate

This type of EBS causes more widespread blistering and can be present at birth or in early infancy. Blistering can happen in any areas where friction occurs, such as under waistbands, bra-straps, as well as on the hands and feet.

- It is unusual for nails, teeth and oral mucosa to be affected in this type of EB

### EBS severe

In this form of EBS, blistering is often present at birth, with some babies being severely affected. While it is often severe in infancy, this type of EBS usually improves with age. It can present as severe generalised blistering, which usually heals without scarring, but severe skin loss early in infancy can occasionally cause some scarring. This form of EBS is different for each patient.

- Typically, groups of blisters will occur at any sites of minor trauma, especially on the nails, hands and feet but can be on the face, trunk and limbs. Over time, blisters often become more localised to the hands and feet.
- The oral mucosa can be involved. Blisters and sores inside the mouth can be painful and make dental care difficult.
- Thickening of the nails is common.
- Frequently, blister sites on the palms and soles of the feet will result in a build-up of hard skin (hyperkeratosis).
- It is common for some blisters to heal leaving some inflammation and redness, and also increased or decreased pigmentation (colour) in the skin.

## Looking after EBS

- Burst (lance) blisters with a sterile needle or scissors. (Your GP can supply these for you, as well as a sharps bin for the used needles.) Make sure that you puncture the blister in two places (entry and exit point) to prevent the blister re-sealing.
- Apply a gauze pad to the blister and gently press to drain the fluid from the blister.
- Cover with a suitable dressing.

## Infections

It is unusual for people with EBS to get wound infections, but it does happen occasionally. We suggest using topical antimicrobial creams, or dressings that can reduce the bacteria and help the blister to heal.

## Dressings for EBS

Many people with EBS prefer not to cover their blisters because the dressings can cause further heat, or move and cause further blistering. However, if a blister leads to an open wound or there is a lot of leakage then dressings can be useful. Below is a table of suggested dressings that might help.

Product	Sizes/ordering information	Use/indications
Medical adhesive remover	Appeel® spray 50ml Niltac spray™	To gently remove dressings from the skin, to avoid trauma to the skin.
<b>All dressings listed are available in various sizes</b>		
Protective/preventative dressings	Spycra™ Protect	For areas that get a lot of friction or blistering. Superficial blisters, or healing superficial blisters or wounds.
Soft silicone foam dressings	Mepilex® Border Lite Biatain® Silicone Lite Kerrafoam™ Allevyn gentle border	For protection of vulnerable areas of skin and open blisters or wounds. Absorbs fluid.
Cooling dressings	KerraLite Cool™ (non-border) KerraLite Cool™ (border)	A soothing, debriding (removal of dead tissue) and moisturising dressing that provides the ideal environment for treating dry to lightly-moist wounds.
Pain relieving dressing	Biatain® Ibu	An absorbent foam that contains 0.5 mg/cm <sup>2</sup> of ibuprofen in the foam.
Antimicrobial (for infections)	Crystacide® cream 1%  Medihoney® gel 15g  Flaminal® Forte and Flaminal Hydro® 15g	Slow-release hydrogen peroxide. Apply 2-3 times a day or as needed. Do not use continually for more than three weeks.  Medical grade honey. It can sting.  Alginate gel that helps to clean wounds. Apply £1 coin thickness.  <b>Use a secondary dressing to cover the area</b>

## Preventing blistering

- Footwear is very important in helping to reduce blistering. There are many shoes available with 'gel cushioned' insoles, such as, Skechers®, trainers, etc. Alternating two pairs of comfortable shoes every other day may help to reduce or stop blisters.
- Coolsorb Insoles are advanced in-shoe temperature management insoles for cooling and comfort, designed to help people with EB. These can be bought through DEBRA UK.
- Silver-impregnated socks can be useful in helping to keep the feet cool. The silver has an antibacterial effect which can help to reduce infections. These are available from some pharmacies, online, or from DEBRA UK.
- DermaSilk® socks can help manage heat and comfort. Available to buy or on GP prescription.
- Reducing friction can help to prevent blisters. A light dusting of cornflour on your feet or in your socks can help to reduce friction.

## Management of callus

Callus build-up, or hard skin (hyperkeratosis) is a common problem for people with EBS. Calluses are thick areas of skin caused by pressure or friction on your feet. Blisters can form beneath the hard layer and can be painful and difficult to lance.

### Topical creams

Creams or balms high in urea can help reduce callus build-up. We recommend Dermatronics Once Heel Balm or Flexitol® Heel Balm (follow the instructions on the packet carefully).

### Podiatry/chiroprody

You may find the services of a podiatrist or chiroprody helpful to gently remove the callus. They can also give advice on insole selection and blister management. There is a podiatrist in the EB service at St Thomas' Hospital, so please ask your doctor or nurse about it if you would like more information.

## Pain in EBS

Blister sites in EBS can be very painful. This is because they are superficial (on the surface) wounds, and the nerve fibres are intact and exposed in the skin. Also, the blisters on the feet put pressure on the nerve fibres. Another cause of pain can be infection. To help reduce pain, try using:

- prescribed painkillers
- soothing and appropriate dressings (see table above)
- topical pain relief such as Biatain® Ibu foam dressing.

EBS blisters and pain can be debilitating and may restrict mobility and physical activity. Some people need a disability Blue Badge to reduce walking long distances. Letters of support with information about EBS can also be written to help with university or workplace parking permits if needed.

Living with a chronic condition, like EBS, can also have an effect on people's psychological well-being. Psychosocial guidelines have been written which highlight this and give information and advice to help with understanding and managing them. Please ask for help if you need support in this area.

Please discuss any issues or concerns with the EB team or your GP. DEBRA community support managers are also a useful contact to help with any social concerns.

## Useful sources of information

### DEBRA

Debra House, 13 Wellington Business Park, Dukes Ride, Crowthorne, Berks RG45 6LS,  
**t:** 01344 771961

The EB service runs in collaboration with the charity DEBRA. DEBRA is the national charity supporting those directly affected by, and working with, EB. A charity registered in England and Wales (1084958) and Scotland (SC039654). Company limited by guarantee registered in England and Wales (4118259).

### Best practice guidelines

Covers complex management of EB and includes lots of information about suitable dressings,  
**w:** [www.woundsinternational.com/best-practices/view/best-practice-guidelines-skin-and-wound-care-in-epidermolysis-bullosa](http://www.woundsinternational.com/best-practices/view/best-practice-guidelines-skin-and-wound-care-in-epidermolysis-bullosa)

Podiatry guidelines,

**w:** [www.debra.org.uk/downloads/community-support/podiatry-cpg-tariq-khan-et-al.pdf](http://www.debra.org.uk/downloads/community-support/podiatry-cpg-tariq-khan-et-al.pdf)

Psychosocial guidelines, **w:** [www.debra-international.org/eb-healthcare-resources](http://www.debra-international.org/eb-healthcare-resources)

## Contact us

### Adult nursing teams

St Thomas' Hospital, Lambeth Palace Road, London SE1 7EH  
EB Administrator, **t:** 020 7188 0843

Heartlands Hospital, Lode Lane, Solihull B91 2JL, **t:** 0121 424 2000

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit **w:** [www.guysandstthomas.nhs.uk/leaflets](http://www.guysandstthomas.nhs.uk/leaflets)

### Pharmacy Medicines Helpline

If you have any questions or concerns about your medicines, please speak to the staff caring for you or call our helpline. **t:** 020 7188 8748, Monday to Friday, 9am-5pm

### Your comments and concerns

For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.

**t:** 020 7188 8801 (PALS)      **e:** [pals@gstt.nhs.uk](mailto:pals@gstt.nhs.uk)

**t:** 020 7188 3514 (complaints)      **e:** [complaints2@gstt.nhs.uk](mailto:complaints2@gstt.nhs.uk)

### Language and accessible support services

If you need an interpreter or information about your care in a different language or format, please get in touch. **t:** 020 7188 8815      **e:** [languagesupport@gstt.nhs.uk](mailto:languagesupport@gstt.nhs.uk)



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