



Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Epidermolysis bullosa simplex: localised and generalised types

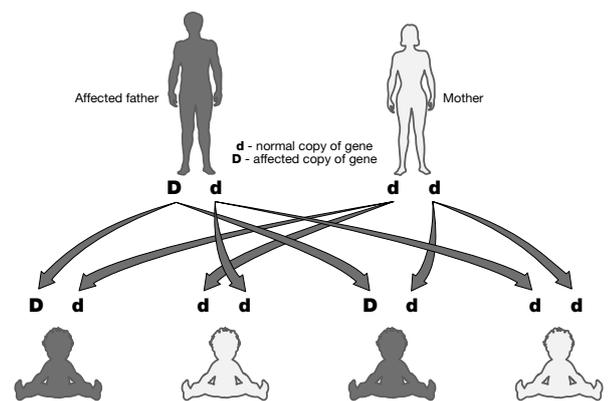
This leaflet explains about the localised and generalised forms of epidermolysis bullosa simplex and how they can be treated. It also contains suggestions for making everyday life more comfortable and contact details for a support organisation. The localised form is also known as Weber Cockayne epidermolysis bullosa simplex and the generalised form Köbner epidermolysis bullosa simplex.

What is epidermolysis bullosa simplex (EBS)?

EB is a group of inherited disorders in which the skin blisters extremely easily. There are four main types of EB. Each is a quite distinct disorder. If you have EB simplex then you cannot develop one of the other forms of EB (dystrophic, junctional or Kindler syndrome).

What caused it?

The problem lies in the genes that hold the instructions necessary for production of certain proteins in the top layer of skin. These instructions have a minor fault, rather like a typing error, with the result that the proteins are incorrectly formed, and unable to fulfil their role as scaffolding for the topmost layer of skin. The result is that the top layer of skin does not 'stick' securely to the layer beneath it, and where the two layers separate a blister develops.



EBS is almost exclusively a dominantly inherited condition seen equally in males and females. This means anyone who has EBS can pass the condition onto his or her children. Each time a pregnancy occurs there is a 1 in 2 chance that the child will inherit EBS if one parent is affected.

However, EBS can sometimes be seen as a 'new mutation' when there is no family history.



Is there a cure?

Not yet, but research continues. There is still a long way to go, but an effective treatment to prevent the blistering may ultimately be possible.

What are the different types of EBS?

Localised EBS (Weber Cockayne)

This is the most common type of EBS. Blistering is localised to the hands and feet. Blisters may not become evident until the child begins to walk.

Generalised EB (Köbner)

Blistering may be obvious from birth, or develop during the first few weeks of life. Occasionally babies are born with raw open wounds which need to be covered with special dressings. Sites of blistering correspond to areas where friction is caused by clothing and frequently appear around the edges of the nappy. Blisters are often seen inside the mouth but do not generally cause a problem during feeding.

EBS Dowling Meara

A separate booklet explaining the management of this type of EB is available.

How is EBS managed?

Management involves identifying new blisters, lancing them with a sterile needle or snipping with a sharp pair of scissors, to release the fluid and prevent spread. If compressing the blister to expel the fluid is very painful, then the fluid can be drawn off using a fine needle attached to a syringe.

Many people find dressings unnecessary, preferring to dust the area with cornflour to help dry up the blisters and reduce friction. However, some people find non-stick dressings helpful.

Pain relief

Blisters can be very painful and limit mobility. Some people find it helpful to take simple painkillers such as paracetamol and ibuprofen when their feet are especially sore. Longer-term pain management may be necessary – sometimes only in the summer months when blistering is at its most troublesome. The EB medical team can offer advice on this to your family doctor (GP).

Constipation

This is a very common problem in all types of EB. Blisters around the anal region (bottom) can make it painful to open the bowels. Constipation then develops because the child is frightened to poo in case it hurts. A good diet, high in fibre with plenty of fluids will help. Sometimes medicines are prescribed to help soften the faeces (poo) or stimulate the bowel.



Prevention of blisters

There are no measures that will totally stop all blisters forming, however the following actions can be carried out to minimise the risk:

- Heat, especially when associated with high humidity can exacerbate blister formation, so measures taken to keep affected areas cool will often reduce the rate of blistering.
 - Wear natural materials
 - Soak the affected area such as the hands and feet in cool water, especially after a hot day
 - Temperature regulation in house/classrooms
- Choice of footwear is very important
 - As stated above, natural materials for shoes are good, such as leather.
 - Check the insides of new shoes for excessive seams, as these could cause blisters.
 - Children should get their feet measured to ensure good shoe fit.
 - It helps to have several pairs of shoes of different styles and to change one pair for another regularly to alter sites of friction. It is helpful to have soft, well ventilated shoes. Shoes in line with the uniform policy of schools are often unsuitable and permission needs to be granted to wear more suitable footwear such as black trainers. A wide fit prevents contact of the shoe with the top and sides of the feet.
 - Some people find special socks beneficial in keeping the feet cool. Many of these products can be supplied on prescription from your GP.
 - Walking some days will not be a problem but other days can be difficult. Many people 'save' their feet to do activities that they enjoy. However, it is important to encourage your child to pace themselves during activities to avoid excess blistering as this will make the following days more difficult.



Suitable dressings and recommended products

The EB Team produce a list of suitable dressings and recommended products, which is updated frequently or as new products become available.

What happens next?

Children with EB simplex will need to attend regular outpatient appointments at their EB specialist centre and then move on to adult services at around 16 years of age.

Further information and support

EB team at Great Ormond Street Hospital (GOSH) – 020 7829 7808
or eb.nurses@gosh.nhs.uk.

DEBRA is the national charity that supports people living and working with Epidermolysis Bullosa (EB) – a genetic condition which causes the skin to blister and shear at the slightest friction or even spontaneously.

DEBRA provides information, practical help and professional advice to anybody living or working with EB, including individuals, families, carers and healthcare professionals, and funds research into the condition. To find out more about how DEBRA can support you, please visit www.debra.org.uk.

Notes



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Dressings and products for babies and children with localised and generalised EB Simplex

This dressing and product list has been compiled by the Epidermolysis Bullosa team at Great Ormond Street Hospital (GOSH). While specific dressings and products are included in this document,

alternatives may be available from other suppliers. If you have any questions, please call the EB team at GOSH on 020 7829 7808 or email eb.nurses@gosh.nhs.uk

| Type of product | Brand | Manufacturer | Indication (used for) | Contraindications and comments |
|----------------------------|------------------------|----------------------|---|--|
| Cornflour | Various commercial | Various commercial | <ul style="list-style-type: none"> Use to reduce friction on feet or to dry up lanced blisters | <ul style="list-style-type: none"> Avoid organic brands as these can be abrasive |
| Lipido-colloid | Urgotul® | Urgo Medical | <ul style="list-style-type: none"> Primary wound contact layer | <ul style="list-style-type: none"> Needs a secondary dressing such as Mepilex® Transfer over the top |
| Soft silicone mesh | Mepitel® One | Mölnlycke Healthcare | <ul style="list-style-type: none"> To cover minor wounds or blisters | <ul style="list-style-type: none"> Can be used alone over blisters without a secondary dressing Use sticky side down |
| Soft silicone foam | Mepilex® Transfer | Mölnlycke Healthcare | <ul style="list-style-type: none"> Blisters sites and areas of skin loss | <ul style="list-style-type: none"> A thin foam for fingers and toes, underarms and groins |
| Polymeric membrane | PolyMem® | Aspen Medical | <ul style="list-style-type: none"> Wounds present at birth | <ul style="list-style-type: none"> Put directly onto wound – no need for primary dressing Change when staining noted on the dressing |
| Bordered dressings | Mepilex® Border Lite | Mölnlycke Healthcare | <ul style="list-style-type: none"> Blister sites or minor wounds or for protection | <ul style="list-style-type: none"> A thin plaster equivalent May need a Silicone Medical Adhesive Remover to take off |
| Hydrogel impregnated gauze | IntraSite® Conformable | Smith & Nephew | <ul style="list-style-type: none"> Wounds and blisters in the nappy area – use over barrier cream. Wrap around toes to heal blisters and protect from rubbing | |



| Type of product | Brand | Manufacturer | Indication (used for) | Contraindications and comments |
|----------------------------|-------------------------------|-------------------|---|---|
| Sheet hydrogel dressing | ActiForm® Cool | Activa | ■ For blister sites | <ul style="list-style-type: none"> ■ Cooling and soothing. ■ Keep blue backing on prevent it from drying out |
| Hydrocellulose dressing | Suprasorb® X | Activa | ■ For blister sites | <ul style="list-style-type: none"> ■ Cooling and soothing. ■ Wrap loosely around fingers and toes as it may tighten if dries out. ■ Change before it gets dry and hard |
| Bandage | ActiWrap® | Activa | ■ To hold on dressings | <ul style="list-style-type: none"> ■ Sticks to itself - do not stretch as you apply or may become too tight |
| Barrier cream | Proshield® Plus | H&R Healthcare | ■ To protect skin | <ul style="list-style-type: none"> ■ Use under nappy or on areas which need protection from friction |
| Cream for minor infections | Flaminal® | Crawford Medical | ■ To manage minor infection | <ul style="list-style-type: none"> ■ Apply in a thick layer ■ Flaminal® Forte for wet wounds ■ Flaminal® Hydro for scabs and drier wounds |
| Moisturiser | QV range | Crawford Medical | ■ For dry skin | <ul style="list-style-type: none"> ■ QV wash, bath oil, cream and intensive ointment |
| Antiseptic wash lotion | Octenisan® | Schülke and Mayr | ■ Open wounds at risk of infection | <ul style="list-style-type: none"> ■ Use in bath or apply to skin and wash off in bath or shower |
| Footwear | Shoes and boots | Geox® | ■ Allow sweat to evaporate and cool the feet | |
| | Climacool® | Adidas | ■ Allow sweat to evaporate and cool the feet | |
| Socks | Silversocks | Carnation® | ■ Contain silver thread which helps to keep the feet cool | |
| Clothing | Silk socks and other garments | Dreamskin® Health | ■ Prevent friction | |
| | | DermaSilk® | | |