

## EPIDERMOLYSIS BULLOSA SIMPLEX (EBS)

### **WHAT IS IT?**

EB is a group of inherited disorders in which the skin blisters extremely easily. There are 3 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 or junctional).

### **WHAT IS THE CAUSE?**

The problem lies in the genes which 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.

### **HOW DID YOU GET IT?**

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.

## ARE THERE DIFFERENT TYPES OF EBS?

There are 3 main types of EBS -

Weber Cockayne EBS

Köbner EBS

and Dowling Meara EBS

This short information leaflet will describe the care and management of Weber Cockayne and Köbner EBS.

DEBRA produce a separate booklet pertaining to the management of Dowling Meara 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.

### KOBNER

Blistering may be obvious from birth, or develop during the first few weeks of life. Sites of blistering respond 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.

### MANAGEMENT

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 aspirated 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 non-stick dressings that could be tried are Intra Site Conformable (Smith & Nephew), Aquacel (Convatec) and Mepitel or Mepilex (Mölnlycke). These dressings can be held in place with a sock, cotton bandage or tubular

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bandage such as Tubifast. Dressings are continually evolving. What one patient likes another may not. The EB team can advise on up to date appropriate dressings.

## **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 DEBRA EB medical team can offer advice on this to your GP.

## **CONSTIPATION**

Occasionally, a person can get a blister around the anal region (bottom), which can then make it painful when passing a stool. This in turn leads to constipation, as a child is frightened to open their bowels because it hurts. If this occurs, it is important not let this continue. A good diet with plenty of fluids helps. Occasionally medicines to help soften stools or to stimulate the bowel are prescribed.

## **PREVENTION**

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

1. 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 area (e.g. hands and feet) in cool water, especially after a hot day
  - Temperature regulation in house/classrooms

2. Choice of footwear is very important. As stated above natural materials for shoes are good i.e. 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.

Some people find the use of insoles beneficial e.g. Carnation who also make socks containing a silver thread which helps to keep the feet cool. "Silversocks" are available from Carnation Footcare, Cuxson Gerrard & Co Ltd, Freepost BM4571, 125 Broadwell Road, Oldbury, West Midlands B69 4BR. Cushioning insoles are available from the same company.

Walking some days will not be a problem but some days can be difficult. Many people 'save' themselves to do activities that they enjoy.

## WHAT HELP IS AVAILABLE

DEBRA is a charity set up to help people with the condition EB. The contact number is 01344 771961. Their website address is [www.debra.org.uk](http://www.debra.org.uk).

They provide information, practical help, and professional help from nurses, social workers, welfare workers and a means of communicating to families via newsletters and conferences.

Some people with EBS have been successful in claiming D.L.A (Disability Living Allowance), which can help towards extra costs involved in having EBS i.e. expensive footwear, bus fares, taxi fares.

Some children have problems getting teachers and schools to understand their condition. Literature is produced by DEBRA to give to the school and the EB nurses can visit the school if indicated.

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