



## Care of the new baby with EB

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# Care of the new baby with Epidermolysis Bullosa (EB).

## What is EB?

Epidermolysis Bullosa (EB) is a large group of genetically determined skin conditions. The common factor within the group is the tendency for the skin and mucous membranes to blister and break down in response to minimal friction and trauma.

There are two ways in which EB can be inherited:

1. Dominant inheritance in which one parent generally has symptoms themselves, and there is a 50% chance in each pregnancy that their children will be affected.

Dominant inheritance can also occur as a new mutation, parents are unaffected, and the symptoms occur for the first time in their child.

2. Recessive inheritance in which both parents carry a hidden or silent gene for EB, in every pregnancy there is a 25% risk that both genes will be passed to the baby, and that the baby will be affected.



## Types of EB?

There are 3 major types of EB, these are simplex, junctional and dystrophic.

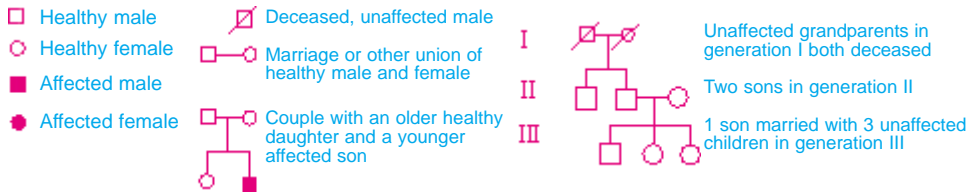
Within each group there are many different sub types and so each type of EB has a wide range of symptoms, varying from very mild to very severe.

It is not possible for one type to change to another within the family.

## The inheritance of EB

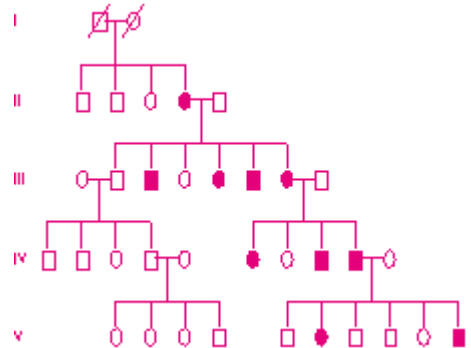
You may be concerned about how EB has been passed down in your family and how future generations may be affected. As already noted, there are two types of inheritance patterns dominant and recessive. They have different inheritance patterns and these are outlined below.

### Symbols used to draw human family trees

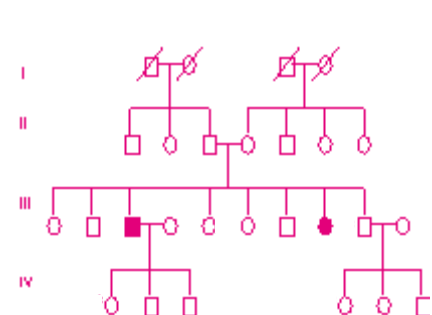


### Dominant Inheritance

The basic thing to remember about dominant inheritance is that it is visible and that there is a 50% chance of children having the condition. With dominant inheritance if you haven't got the visible condition you cannot pass it onto your children.



### Recessive Inheritance



With recessive inheritance there is only a 25% chance of a child inheriting the condition. But there is a further 50% chance of offspring carrying the condition and so having the chance of passing the condition on. So there is a 25% chance of a child being totally free of EB.

## Diagnosis

Diagnosis is made by examination of a small piece of skin. We recommend this be analyzed at a specialized center where diagnosis of EB is routinely made.

Blood samples for DNA analysis are required from the infant and parents for identification of specific mutations.

## Handling

Nurse the baby in an incubator only if medically necessary for reasons such as prematurity. Wherever possible nurse the baby in a bed, lay the child on a small soft pad so he/she can be lifted on this rather than risking further damage.

Where it is necessary to lift using your hands, roll the infant onto his side, place your hands behind the head and under the buttocks, allow the baby to roll back on your hands, and lift.

Never lift from underneath the arms. Remember, friction and shearing forces will cause blisters and skin loss, direct pressure is safe.



## Blisters

These must be lanced with a sterile needle as they are not self-limiting and will extend if left unchecked. Where the roof remains on the blister there is no need for a dressing

## Analgesia

Appropriate analgesia may be given prior to dressing changes and as required for general comfort.

## Wounds

It is recommended that wounds be dressed with a non-adherent dressing. Choice of dressing is limited as many dressings described as non-adherent behave differently on the skin of those with EB.

For lightly exuding wounds or for those with EB simplex who react adversely to a thicker dressing; Mepitel (Mölnlycke Health Care) is a transparent micro-adherent contact layer dressing and therefore may be more suitable. Mepitel dressings may be changed every 4 to 7 days. The required secondary cover dressing can be changed daily to remove exudate and inspect the wound. Depending on the condition of the wound, the surrounding skin and the presence of infection, the physician may modify the frequency of dressing changes.



Please consult the physician about the use of medicated and non-medicated ointments. Apply ordered ointment on top of Mepitel once it is placed upon the wound. Ointment can be reapplied upon Mepitel during secondary cover dressing changes.

Mepilex (Mölnlycke Health Care) is an absorbent foam dressing that is non-adherent to moist wounds and reduces the chance of trauma to the surrounding skin upon removal. It may be left in place for up to 7 days. It is usually indicated for low to moderately draining wounds. Depending on the condition of the wound, the surrounding skin, and the presence of infection, the physician may modify the frequency of dressing changes.

## Feeding

Wherever possible, oral feeding should be encouraged. If the mouth is very sore use of a special feeder recommended for a baby with cleft lip or palate reduces the need for strong sucking and allows a good delivery of milk. Babies with extensive skin loss may need additional calories to avoid competition of nutrients between healing and growth.

## Clothing

Naked babies with EB tend to cause damage to their skin by kicking their legs together and rubbing their arms across their chests. For this reason we recommend dressing the infant in a soft all-in-one baby suit.

## Tips on care and handling

- Ensure all those who are caring for the infant know the correct method of handling.
- NEVER apply adhesive tapes of any kind to the skin. May be helpful to use strips of Mepitel/Mepiform (Mölnlycke) to secure IV cannulae.



- Avoid plastic identification tags.
- Discourage use of pacifiers.
- Avoid suppositories/enemas.
- Select flat-seamed clothes or turn garments inside out to avoid friction.

## EB Simplex



This is generally a dominantly inherited condition, although there are a few cases of recessively inherited EB simplex, but these are very rare.

There are 2 main types of EB simplex:

1. Weber Cockayne which generally affects hands and feet only and is at its most trouble-some in the summer months.

2. Dowling Meara, which causes more wide spread blistering particularly in young children. Infants with Dowling Meara may be very sick in the first few weeks of life, but the majority will survive and the extensive blistering will gradually resolve. Blistering and thickened skin on the palms and soles can cause problems in the longer-term.

## Junctional EB

This is recessively inherited. Junctional EB can be a very mild condition, causing little disability and long term problems. However, in its most severe form junctional EB can lead to death in early infancy or childhood.

## Dystrophic EB

Dystrophic EB can be either dominantly or recessively inherited. In common with most genetic disorders, the dominant form is often a milder disease. However, recessive dystrophic EB varies in severity from very minor symptoms to severe skin loss at birth, and increasing disability as a result of contractural scarring.

**Information may be sought from:**

DEBRA International.  
(Dystrophic Epidermolysis Bullosa Research Association)  
13 Wellington Business Park  
Dukes Ride, Crowthorne  
Berks. RG45 6LS  
United Kingdom.  
Tel: +44 (0)1344 771 961  
Fax: +44 (0)1344 762661  
E-post: [debra.uk@btinternet.com](mailto:debra.uk@btinternet.com)  
[www.debra-international.org.uk](http://www.debra-international.org.uk)

DEBRA employ specialist nurses who can offer advice by telephone and practical help in the UK by visiting the infant and demonstrating care and handling techniques.

**Your local DEBRA contact is:**

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New York  
NY 10006  
Tel 212 513 4090

**Your local Mölnlycke contact is:**

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110 Baldwin Tower  
Eddystone, PA 19022  
USA  
Tel 610 471 0160

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2010 Winston Park Drive  
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Oakville, Ontario, L6H 5R7  
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