

The Care of the New Baby with EB

Initial Information

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Care of the new Baby with Epidermolysis Bullosa (EB).

Initial information

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 1:4 risk that both genes will be passed to the baby, and that 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.

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 troublesome 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.

Diagnosis

Diagnosis is made by examination of a small piece of skin. We recommend this be analysed at a specialised centre 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 cot, lay the child on a small soft pad so he 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 must be given prior to dressing changes and as required for general comfort.

Wounds

Must 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.

Mepilex (Molnlycke) provides the optimum environment for wound healing and poses no threat to the surrounding skin on removal of the dressing.

For lightly exuding wounds or for those with EB simplex who react adversely to a thicker dressing; Mepitel (Molnlycke) is a more suitable dressing. The required secondary dressing can be changed daily to remove exudate and inspect the wound. Change the Mepitel dressing every 4 to 7 days.

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.

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.

Tips on care and handling

Ensure all those who are caring for the infant know the correct method of handling.

Clamp umbilical cord with ligature rather than cord-clamp to avoid local blistering.

Avoid plastic identification tags.

NEVER apply adhesive tapes of any kind to the skin.

Use strips of Mepitel/Mepiform (Molnlycke) to secure IV cannulae.

Discourage use of dummies/soothers.

Avoid suppositories/enemas.

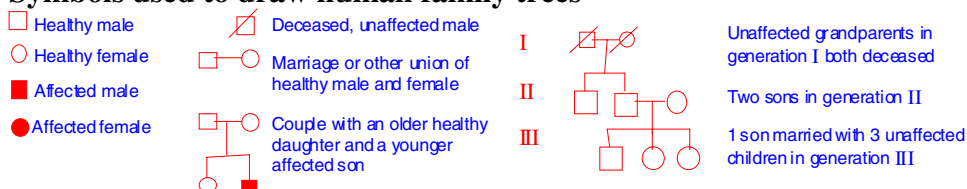
Use commercial plastic kitchen cling film as temporary dressings after bathing.

Select flat-seamed clothes or turn garments inside out to avoid friction.

The Inheritance of EB

You may be concerned about how EB has been passed down your family and how future generations may be affected. As already noted there are two types of inheritance pattern, 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 1 in 2 chance of children having the condition. With dominant inheritance if you haven't got the visible condition you can not pass it on to your children.

Typical Dominant Inheritance – *(Illustration caption)*

Recessive Inheritance

With recessive inheritance there is only a 1 in 4 chance of a child inheriting the condition. But there is a further 1 in 2 chance of offspring carrying the condition and so having the chance of passing the condition on. So there is a 1 in 4 chance of a child being totally free of EB.

Typical Recessive Inheritance – *(Illustration caption)*

Information may be sought from:

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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 specialist medical contact is:

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This booklet was sponsored by Mölnlycke and produced by DEBRA International, registered charity no: 284754

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