

EPIDERMOLYSIS BULLOSA

INFORMATION PRACTICAL ADVICE



EPIDERMOLYSIS
BULLOSA
GET INFORMED
& ORGANISED

Different forms of epidermolysis bullosa (EB) can affect around 1: 20,000 new born babies and radically alter the way the child is looked after. The cutaneous fragility caused by the illness means that every action has to be carefully considered in order to limit the formation of blisters and the resulting pain.

Support associations for families affected by EB exist throughout the world and help to resolve the concerns that this disease can raise for new parents: "How can we pick him up without hurting him?" "Can we cuddle him", "How can we comfort him?" The baby's frequent hospitalisation during the first few weeks should be used as an opportunity for finding out as much information as possible and planning for the future. With the help of the hospital team, parents will learn simple activities and methods to enable them to care for their baby safely. The objective of this brochure, compiled by URGO MEDICAL, is to help them during this "learning period".

URGO MEDICAL is committed to supporting families affected by EB.

We hope that the practical advice offered here will help you to care for your baby.

URGO MEDICAL

FINDING OUT MORE ABOUT EPIDERMOLYSIS BULLOSA

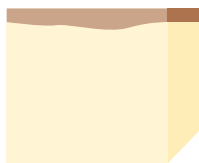
Epidermolysis bullosa (EB) is a group of dermatological diseases of genetic origin. From a very young age the skin is extremely fragile and the slightest friction can cause the skin to blister and shear off. The blisters are typical of EB. There are numerous types of EB that are closely linked to whichever genetic anomaly is responsible. Some forms may disappear progressively, whilst others will develop over time and become an integral part of the lives of the people affected.

EB, an orphan disease

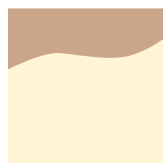
The term “epidermolysis” means detachment of the epidermis. The total number of sufferers – children and adults – is not precisely known but is estimated to be around 500,000 worldwide for all types of EB, 10,000 of which are in France.

The skin lesions are generally present at birth sometimes affecting the mucous membranes. Skin fragility is caused by a problem with the protein production that should provide cohesion between the dermis and epidermis: these two layers of the skin are no longer held together which allows blisters, similar to those caused by burns, to form following the slightest trauma. There are around twenty different forms of the disease belonging to three main groups: EB simplex, junctional EB and dystrophic EB.

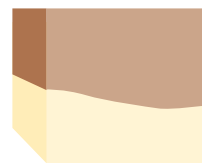
EB is categorised into 3 main types according to how badly the skin is affected



**Dolesto commolum
zrriliquat prat.**



**Vulla
consequatem**



**Veliqua
tummolore**

Cases of EB simplex are quite common. Often the blisters form on the hands and feet. They don't leave scars because the cutaneous separation is located within the epidermis, ie, on the superficial layer of the skin. The disease usually appears during the early years of life and reduces at adolescence. Different forms of EB simplex exist, varying in localisation and the amount of pain suffered.

Junctional and dystrophic EB are less frequent and of variable severity. Junctional EB frequently affects nails, teeth and hair. Certain junctional forms improve over time. Dystrophic EB is diagnosed by the presence of blisters in the dermis. These leave scars can contract and be disabling. Nevertheless certain forms will also improve over time.

The baby's condition at birth doesn't predict how the disease will progress: only a precise genetic diagnosis of the type of EB concerned can allow for a medium or long term prognosis.

Daily care

The **medical care** of EB children aims to both prevent and to treat their skin lesions, manage pain and avoid complications. Whatever the form of EB, precautions should be taken to avoid mechanical stress, particularly friction, on the child's skin. You will learn to handle your baby with minimum risk, to select suitable clothing, to protect his surroundings and to treat his wounds. In the common forms, renewing dressings can take time and must be carried out according to a strict protocol. Pain should be alleviated and preventative measures taken prior to the care routine.

Today, research is advancing and new cellular therapy techniques look promising. Researchers, health professionals, families and sufferers are hoping that one day it will be possible to treat and conquer this disease.

To find out more...



- EB INFO WORLD - www.ebinfoworld.com/abouteb_fr.htm
This site gives clear and precise explanations particularly in the question and answer section. For the French version, click on "FQR".
- The MAGEC Booklet – Can be downloaded from the site www.ebae.org
This booklet was written by a specialist group at the Necker Hospital. It discusses the problems posed by EB from birth to adolescence.

SEEKING SUPPORT AND FINDING HELP

New born babies suffering from EB need careful monitoring every day because of their vulnerability. They often stay in hospital for several weeks following diagnosis, to allow the parents enough time to learn how to look after them and to prepare for returning home. It is also a good idea to use this period for contacting the various authorities that can provide financial and practical aid.

Following the birth...

The new baby suffering from EB will usually stay in hospital for several weeks: this allows doctors to make a precise diagnosis of disease type, to monitor the child while there is a risk of complications and to plan for his/her future care.

The diagnosis of EB type is made by taking samples of skin and blood from the child and his parents. The analyses are carried out by specialist centres.

Daily monitoring in a hospital setting ensures early detection of any infection or absence of weight gain. The wounds are caused by a significant loss of protein in the skin which can cause nutritional deficiencies and disturb the baby's growth.

Good nutritional intake is essential in the diet. Breast feeding is possible, although often supplemented by bottles. The teat must be very soft and moistened so that it doesn't hurt the baby and the bottle should be given at lukewarm temperature. If oral feeding is impossible or insufficient, a small tube inserted through the nose can be used to feed the baby on a temporary basis until he/she begins to gain weight at a satisfactory rate.

Nursing care is carried out following administration of an analgesic. It takes place after the baby has been fed (to limit distress and crying), in a bathroom that is well-heated, peaceful and subtly lit. The dressings are changed after the bath according to a strict protocol (see page 10), every day if possible, otherwise every second day.

Psychological support can help you to better understand this disease which can initially be perceived as intolerable: you are afraid of injuring your baby, you want to cuddle and pamper him but you are scared of hurting him, you worry that you won't be able to cope with the everyday challenges... The hospital team is there to support you, to prepare the return home and to help you to organise a family and social life that is as full as possible.

**The hospital train parents in the daily care routine and monitoring.
Team members help to organise the baby's return home
and subsequent follow up.**

**Support can be arranged with the hospital psychologist:
the social worker helps families with their administrative procedures.**

Contact a patient association



The main association is DEBRA. They will provide you with documentation and are there to support you and answer your questions.

- WEBSITE:
www.debra.org

ORGANISING DAILY LIFE

During his/her early years, you need to protect your child within his surroundings whilst encouraging his development and progress. Little by little he has to learn how to live alongside other people with his severe skin fragility; initially this will be at home with family and then later at school. The objective during early childhood is in fact the same as that for a child who is not ill; to become independent.

How should you pick up your baby?

You may be reluctant to do so for fear of causing blisters. However, a newborn baby needs physical contact and cuddles to help him to develop. You will soon learn how to handle him gently: having removed any jewellery that could hurt the baby, roll him slightly onto his side away from you, then place one hand under his head and the other under his bottom and let him roll into your arms as you lift him up from his bed without letting him slide across the sheet. If your friends want to pick him up too, pop the baby onto a pillow and then they can carry him in complete safety.

When he is bigger and holds out his arms for you to pick him up, place one hand under his bottom and the other behind the nape of his neck. Don't grab him under the arms because this is a fragile area that heals with difficulty.

Even if you have taken all the necessary precautions, blisters may appear but this is still a better option than no cuddles at all! You should also explain to your friends and family how to look after your baby; you must be able to leave him without any worries.

How should you feed your baby?

For EB simplex cases, food does not pose any particular problems. For the type of the disease that affects mucous membranes, feeding takes longer and meals are generally split into small quantities taken often. Anti-regurgitation treatment may be prescribed to protect the digestive membranes from any acid reflux. The baby's diet should allow for the possibility of constipation that could arise because of blistering in the anal region. Every "little tip" is useful to avoid this complication: increasing the amount of fluid, water rich in minerals, particularly magnesium, prune juice or laxatives. A varied diet can be given in the normal way.

How should you dress your baby?

For the first few months soft babygrows are ideal for your baby. Afterwards, put his underclothes on inside out so that the stitching doesn't irritate him. Chose clothes that are sufficiently loose and remove any labels that could easily cause blisters. For difficult cases, specially designed clothing and shoes are available. Check out the websites or contact the associations directly.

Making his environment safe

Children with EB often take longer to learn to walk because they are afraid of hurting themselves. You can use elbow and knee protectors when your child begins to crawl and move around on all fours. If you use a playpen, place a soft blanket or sheep skin at the bottom and protect the sides.

Toys shouldn't have any corners or sharp edges; select quiet games so the baby doesn't get over-excited and hurt himself by waving his arms about. Mobiles and musical toys are useful for calming the baby while you are changing him; he will also enjoy bath books that are soft and easy to grab.

For car trips, you can use a standard baby seat by inserting soft material under the seatbelts to avoid any risk of rubbing. Do the same for the high chair and baby bouncer.

To find out more...



- Discussion forms can be found on the association websites. They will allow you to compare experiences with the parents of other children suffering from the disease.
- You will also find links to specialist products such as clothing and shoes.

LEARNING TO TAKE CARE OF EB WOUNDS

After the neo-natal hospitalisation period, wound care will be carried out at home, in close collaboration with your doctor and the hospital team. Every effort should be made to ensure that the baby is relaxed and comfortable. Pain should be kept to a minimum by using analgesics and dressings that don't adhere to the wounds. We strongly advise you to keep a care routine record book.

Treatment preparation: an essential step

The care routine should be carried out in an atmosphere that is as reassuring as possible, in a warm, softly lit bathroom. Relaxing music and colourful mobiles can have a soothing effect. The baby should have been fed; he can be pacified with a dummy or a little sweetened water. An analgesic, administered 30 minutes before starting any treatment may be necessary. Everything should be ready before you undress your baby: take out all the necessary equipment and place a soft towel along the bottom of the bath. Run a bath at 37° adding some antiseptic soap solution. Make sure that there are enough towels, dressings and bandages. A sterile gown and gloves should be worn to limit the risk of wound infection.

Bath time and the care routine

Two people are needed for the care routine: the baby could be restless so you need to be able to attract his attention, to speak to him or to play with him. It is much better for everyone concerned if the care routine can be stressfree.

Undress your baby and remove any dressings that come away easily. The others will come away in the bath water. Take your time, play and talk to your baby to reassure him and to watch his reactions during his bath, which should last for 10 to 15 minutes (see boxed section).

Each wound should be inspected for signs of infection or healing problems which require medical advice (delayed healing or on the other hand, excessive skin growth). The whole body should be carefully examined to detect and count new blisters. Scabs should be delicately removed or softened with vaseline.

The next step is to lance and clean the blisters to prevent them from spreading and prolonging the healing process.

Antibiotic creams should not be used unless there is obvious super-infection; their prolonged usage can cause bacteria to become resistant and also encourage allergic reactions. Moisturising cream can be used on healthy skin.

Dressings: avoiding pain and adherence

The wounds should be covered with non-adhesive dressings such as Urgotul® contact layer both to avoid pain and to encourage healing: In some areas, even with this type of dressing, it may be useful to apply a layer of vaseline. The dressing should be covered with sterile compresses held in place by a very soft bandage such as Nylexfix® and adhesive tape taking care that it doesn't stick directly onto the skin and that the bandage isn't too tight. For wounds on the hands or feet, fingers and toes should be separated from each other to allow healing to take place: dressings should be inserted between the digits and held in place with a small band around the wrists and ankles.

A treatment record book should be kept to note any important monitoring issues for your baby: appearance of the dressings (colour, odour), amount of pain observed before, during and after the bath to adjust analgesic treatment if necessary, the baby's behaviour, the number of blisters lanced...

After bath time, rest time!

Your baby will be tired after his bath; get him dressed, give him a cuddle, congratulate him for being brave and put him to bed. Try to avoid any further treatment or handling during the hours that follow, to allow him to rest.

To find out more about caring for EB go to:

- PATIENT ASSOCIATION WEBSITES: where you will find details of other parents' experiences and advice
www.debra-belgium.org
www.ebae.org
- SPECIALIST SITE FOR URGO LABORATOIRES: lots of information, including care techniques and useful products
www.eb-urgo.fr



CARING FOR EPIDERMOLYSIS BULLOUS

Prepare the treatment record book

Wash your hands and prepare the equipment you will require

- Sterile gloves and gowns
- Antiseptic soap for the bath
- Several flannels
- Bath towel
- Sterile waterproof draw sheet
- Clothes
- Antiseptic solution
- Instrument box with sterile scissors
- Sterile needles
- Sterile and non sterile compresses
- Non adherent dressings (such as Urgotul®)
- Stretchy bandages (such as Nylexfix®)
- Adhesive tape to hold the bandages in place (such as Urgoderm®)

If necessary: tubular netting (such as Surgifix®), emollient cream, antibiotic cream, ophthalmic solution...

Prepare the bath

- Temperature 37°, with some antiseptic soap solution

Undress your baby and inspect the dressings

- Identify any dressings that are too tight
- Note any abnormal colouration or odour
- Try to ascertain whether the baby is itching

If possible, gently remove the dressings

The bath: 10 to 15 minutes

- Remove the remaining dressings
- Dab the face and any other areas not immersed in the water with a wet flannel
- Finish by washing his hair
- Rinse with clear water, if necessary covering the shower head to reduce the pressure of the stream
- Dry using compresses (non sterile); most importantly do not rub
- Wrap the baby in a bath towel and place him on a sterile, waterproof draw sheet.

Remove the scabs or apply vaseline

Clean the blister contents

- Prick the small blisters with the point of a sterile needle
- Absorb the liquid with sterile compresses
- Nick the tops of the largest blisters with the sterile scissors but leave them in place. Then disinfect with antiseptic solution
- Note the number of pierced blisters

Dressing the wounds

- Apply a contact dressing such as Urgotul® to the wounds, taking care to separate fingers and toes
- Cover with sterile compresses and keep them in place using narrow bandages or tubular netting
- If your baby can sit up, you can begin with his bottom. Then you can sit him up and continue doing the dressings in the following order: feet, knees, thighs, thorax, abdomen, shoulders, arms, elbows, forearms, hands.

URGO products suitable for the care of Epidermolysis Bullosa



Dressings and contact layers reimbursed on prescription



URGOTUL

This fine mesh dressing was developed by URGO research and uses Lipido-Colloid Technology (TLC): the dressing does not adhere to the wound and can be removed with minimal pain. Its great flexibility makes it easy to use for hands and feet. New Urgotul Flex is the answer when even more flexibility and elasticity are required.

URGOTUL DUO

The TLC contact layer is backed with a thin, very flexible foam pad. The dressing does not adhere to the wound. It isn't always necessary to apply a secondary dressing with compresses.

Other products available from the pharmacy



Dry Urgo compresses



Nylexfix bandages

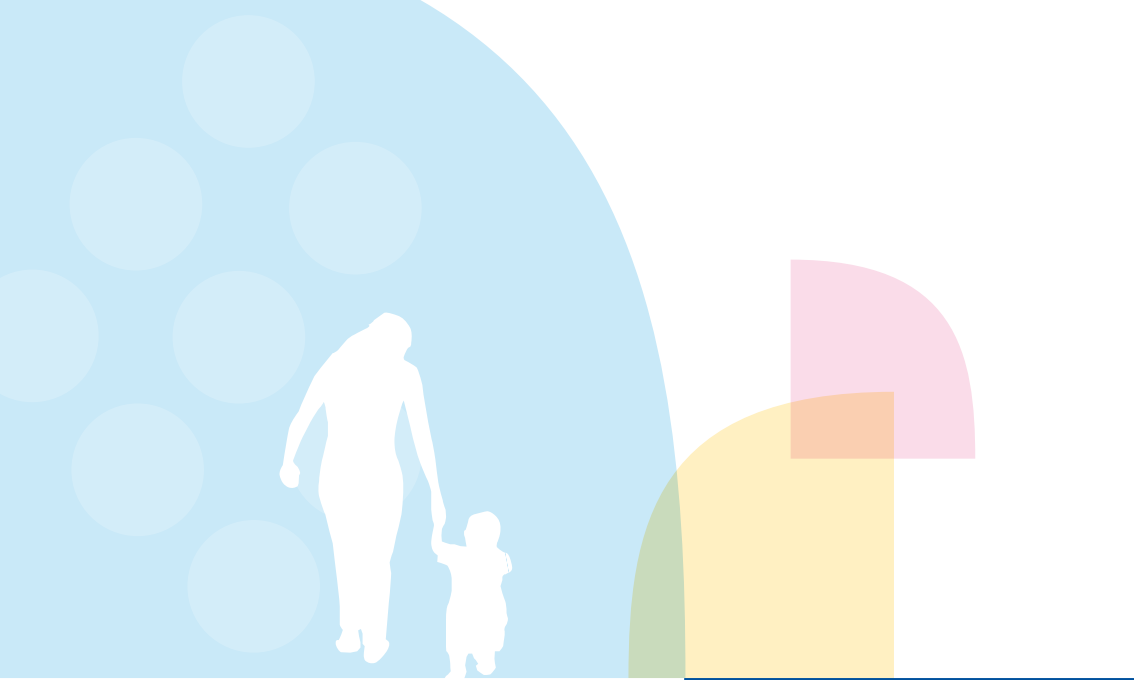


Surgifix tubular netting



Urgoderm adhesive tape

For more information, please visit our website www.urgo.fr



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