



Care manual for **Epidermolysis bullosa**

Author:

Luciana Mendes - Stomatherapist Nurse / EB Specialist

Content reviewed by:

Carla Duque, Stomatherapist Nurse/ Technical Advisor, Epidermolysis Bullosa Mölnlycke

Prof. Dr. Roseanne Montargil Rocha Pos, Doc in Health Sciences Stomatotherapist TiSobest, CNPQ Researcher Coordinator of the UESC Stomatherapy Nucleus, NUET Coordinator of the UESC, LAET Stomatic Academic League

Livia Xavier, Stomatherapist Nurse at Hospital HC, BA

Maria Efigenia de Queiroz Nutritionist, Federal University of Bahia, UFBA, Nutritionist, Neonatal Screening Reference Service, Doctor of Medicine and Health – UFBA

The information in this manual is based on our guidelines, care guidelines and scientific evidence available in the latest literature. The aim of these instructions is to make self-care and information more easily accessible for people with EB, enabling necessary care in daily life.

Note: It is important to always be evaluated by a doctor.



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Introduction

This manual is designed for carers, family members and healthcare professionals to learn about epidermolysis bullosa (EB). Its aim is to provide up-to-date guidelines and daily care information and practices about EB.

This manual was created to guide, clarify and provide specialised support for those who need it.

Epidermolysis bullosa has a major impact on the lives of patients and family members, and represents a great emotional and physical challenge.

Most patients and their carers need extra support for EB care in the home environment. This manual considers the educational process, post-discharge treatment or consultation, and the promotion of self-care.

Epidermolysis bullosa

Epidermolysis bullosa (EB) is a **non-contagious disease that is part of a group of genetically transmitted skin disorders**. It is characterised by the formation of blisters after minor trauma or spontaneously.

It is classified into four types:
EB simplex, junctional EB, dystrophic EB, and Kindler syndrome.

All types of EB are characterised by fragile skin and the formation of blisters, mainly on the hands and feet.

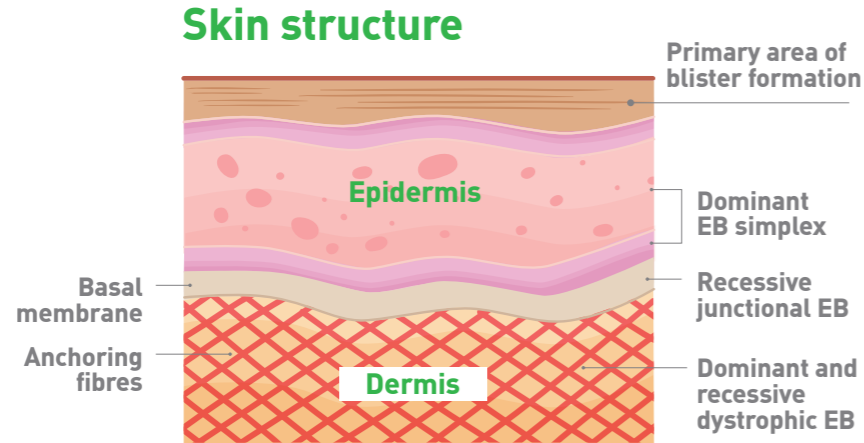


The presence of multiple wounds, which vary in duration, makes EB care complicated.

There is no cure for EB yet, but numerous studies and research projects are being conducted.

Diagnostic confirmation can be made by skin biopsy, immunofluorescence testing, electronic microscopy, or genetic sequencing.

Many EB complications can be reduced or even avoided through adequate care.



How will I be able to take care of my child with EB?

You will be able to take care of your child normally, as if they did not have EB. You only need to change the way in which you give some types of care.

Children with EB have fragile and easily injured skin. Therefore, it is important to handle the skin with care.

When you hold your baby, it is important to place one hand behind the head and the other hand on the buttocks. Avoid picking the child up by the armpits. This pressure could cause injuries and blisters. In some cases, you may need to use a pillow or foam to pick up your child.



Blister care

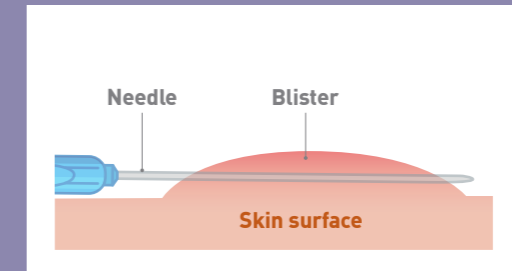
Blisters are a major challenge in EB care because it is impossible to prevent them from appearing.

Blisters

Blisters are caused by friction, trauma, or even spontaneously from heat. It is recommended that blisters be popped in order to avoid their spreading. Once blisters are popped, use a non-adhesive dressing (preferably silicone).



- 1 Take out all materials you will use, placing them in a clean area. Before and after the procedure, wash your hands thoroughly
- 2 Ensure the correct protection is in place before the person lies or sits down (back and buttocks), protecting them from trauma and ensuring their comfort
- 3 Wash the spot with saline solution. Avoid using products that could cause skin trauma. Clean the area gently
- 4 Pop the blisters with a sterile disposable needle. Do not pull the skin away from the blister, ensuring that you keep natural protection in place
- 5 To prevent fingers and toes from 'getting glued together' (sticking to each other), dress each finger and toe separately, both on the hands and on the feet. After that, protect them by wrapping each finger and toe of the hands/feet



Nutrition

Breastfeeding



Mother's milk is an important nutrient for building the baby's immunity.

Always encourage breastfeeding.

Before starting to breastfeed, check the baby's mouth and the mother's nipples. If necessary, moisten the areas using petroleum jelly. Be aware of the areas that might cause friction.

If the baby has problems sucking, pump the milk and give it to the baby in a feeding bottle with flow control, a plastic cup, a silicone spoon, or a feeding bottle with silicone nipple (soft). Avoid using tubes!



Food consumption



Wounds on the mouth and throat cause discomfort and may cause children with EB to reject food, but it is important to offer options.

The child should be monitored regularly by a nutritionist for a nutritional protocol and to measure growth and development.

It is important to encourage the child to participate in family meals.

- Increase calories and use nutritional supplements
- Do not offer hard or crunchy foods
- Offer pureed foods
- Ingest liquids in large quantities
- Avoid excess salt and spicy seasoning
- Offer more meals in smaller quantities



Bath time

Bathing is important and should preferably take place daily. Making it pleasant is a challenge. Find a calm place with good lighting and, if possible, turn on music or cartoons that distract the child.

If prescribed by a doctor, give the child an analgesic before a bath to alleviate pain.

Before the child takes a bath, make sure to disinfect the bathtub. Place everything you will need in a clean space: **soft towel, neutral soap, and appropriate emollient.**

Always pay attention to water temperature. If you are using dressings, take them off during the bath, avoiding trauma.



Newborns



Newborns require special attention. Be careful, protect the bathtub (e.g., with foam). Wash the baby one body part at a time, pouring water from a cup (pay attention to the water temperature).

Never scrub the child, make gentle movements to avoid the formation of new blisters and injuries.

During the bath, watch for any areas of suspected infection.

Once finished, use a soft and clean towel to dry.

Oral health

From an early age, dental care is needed. It is important to brush the teeth daily. Blisters may appear on the inside of the mouth. Use a soft brush, cotton swab or cotton pad. The toothpaste should be suitable for the child's age.

Consult a specialised dentist for instructions and monitoring once the first teeth come out.

If necessary, use a mouth sanitiser without alcohol to avoid discomfort or pain.



Skin hydration

It is important to keep the skin moist with appropriate solutions (talk to the dermatologist).

Dry skin leads to itchiness, so it is important to keep it hydrated. If the discomfort persists, consult a doctor for the right prescription.



Clothing

Wearing loose, soft clothing with few or no stitches is essential. It is important to use appropriate clothing to prevent blisters. Tips: use clothing without labels, without rough seams, without zippers - make sure the elastic does not squeeze the skin.



Always remember when selecting clothes that they should be easy to put on and take off. Take special care with areas of the head, ears, and armpits when putting on or taking off clothes.

Clothes may be worn inside out to help avoid the friction to the skin caused by the clothing's inner seams.

Disposable nappies should be the right size for the baby so that they are not tight on the legs (groin area). On the sides of the nappies, to protect the skin, use of a silicone cover is recommended.

For bedding, choose soft cotton material; it is more comfortable.

Footwear

Wearing shoes is a challenge for some people with EB.

It is important to wear appropriate shoes, preferably soft, without seams, and wide to accommodate the bandages and dressings that are necessary in some cases.



Soft socks without seams work well. When choosing shoes, consider the ease of putting them on and taking them off. Some feet get many injuries, requiring extra padding (e.g., foam).

It is very important to think about comfort, so that the child can move well, walk, play, and exercise.

Dressings

Perform a careful evaluation of the skin and wound(s) every day. People with EB and their carers need to be trained to manage (and care for) the injuries.

Routines and choices in the care of injuries must balance efficacy of product, patient's choice, and quality of life.

Non-adhesive and absorbent dressings are fundamental in the treatment of injuries. When they get dirty, dressings should be changed, preferably daily.

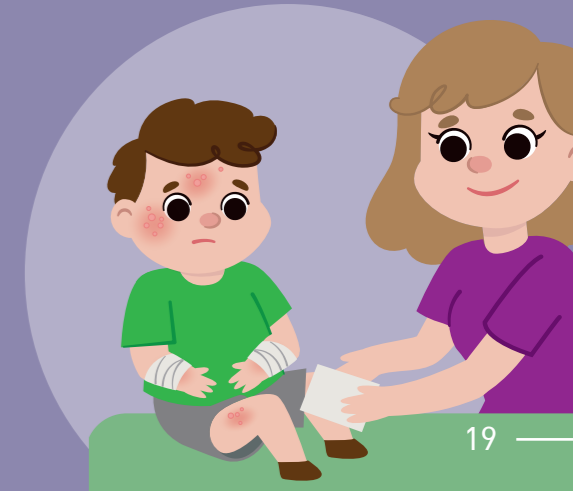
Apart from preventing trauma and minimising pain, dressings provide treatment efficacy. Silicone foam dressings are the most complete type. When combined with silver, they have antimicrobial properties (to help fight infection).

Avoid using adhesive tape. When necessary, apply over mesh or bandages, but never directly onto the skin.

Consult a doctor or specialist nurse to instruct you and prescribe the right dressing.

Applying dressings

- 1 Before and after applying dressings, wash your hands well
- 2 Get out the material you will use, placing it in a clean area
- 3 Inspect the skin and examine any injuries, skin colour or discolouration, secretions, odours, and note whether pus or new blisters have appeared
- 4 Take your time when doing dressing changes, always ensuring comfort
- 5 Clean all the injuries with gentle movements. Do not cause friction
- 6 Choose the dressing. If necessary, use the cutting guide. Using appropriate scissors, cut the dressing to ensure better comfort for the selected area
- 7 Gently place the dressing on the spot, using meshed fabric or bandages for attaching firmly if necessary
- 8 A specialist healthcare professional should provide instructions and prescriptions for the treatment of infected injuries, ensuring use of the right dressings for infection control



Signs to potential infection

It is important to pay attention to signs of infection. Watch out for an increase in or change in colour of secretions, as well as if they have a bad odour. Fever is also an indication of infection.

Consult a specialist if you observe any abnormalities. If an antibiotic is prescribed, use it as instructed by the medical professional.

In case of questions, **consult a doctor.**



Vaccinations

Consultation with a doctor with whom the child's EB diagnosis has been discussed should guide decisions on when, how and which vaccinations should be administered.

When receiving an injectable vaccine it is important to explain to the health professional:

- That the child has EB, a genetic, non-contagious disease that makes the skin extremely fragile
- The cotton pad should be applied gently so that blisters are not formed and the skin is not hurt
- The vaccine should be applied in the area where the skin is intact, without wounds. It could be over a scar
- After application, adhesive tape should not be applied to the skin



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