

## THE NEW BORN WITH EPIDERMOLYSIS BULLOSA (EB)

### 1. Introduction

A newborn that comes into this world with EB or suspicion of EB, like any other baby, needs especially shelter, security, food, closeness and a stimulating environment. Due to the vulnerability of their skin, the baby also needs professional medical and nursing care. The first days with a new baby is always exciting and characterized by uncertainty, which applies even more to a child born with a disease like EB. This section serves to simplify this first experience, with all its ups and downs and help to make it easier for you. We hope and wish that we can help you quickly to become familiar with this new and unexpected situation. With a little help and support you will soon be able to take good care of this newborn baby with his special needs.



### Important points in a nutshell

- **Safety, security and love are the most important things for a child with EB.**
- **Your baby may and should be touched one way or another and formation of a blister ("cuddly blister") can be taken into account.**
- **If wearing gloves for a dressing change is needed, they should be rubbed with a high-fat concentrated cream or Vaseline.**
- **It is better not to lift the baby under the arms first, but bring a soft blanket into position and then carefully lift it.**
- **When dressing the baby, make sure the clothing has no pressure points.**
- **Breastfeeding is often possible with EB.**
- **Not all babies with EB have pain, through careful observation it is possible to distinguish between pain and other causes of discomfort.**
- **If there is pain, pain treatment should be recommended by a physician/health care professional.**
- **Preparations are needed for the home during hospitalization.**
- **Find a pediatrician or family doctor as soon as possible that you trust in your neighborhood.**

## 2. General Information

If you have started to read this, then you probably already have a very stressful time with ups and downs now behind you. A time when you probably had a totally surprising change in your life situation: when your newborn child (or perhaps even the child of a relative or friend) is born with the suspicion of having Epidermolysis bullosa. We want to help you adapt to this new situation in your life.

If you do not know for sure whether it really is EB, if maybe it can be another disorder, we encourage you to await the results of the medical tests. EB is a disease with many forms and facets, it is extremely difficult to record and classify all the existing information about EB, and you may read a lot of information that is not relevant for the EB form from which your child may suffer. But since the diagnosis or suspicion of EB appears obvious, we want to help you survive during this first period of uncertainty in this following chapter. If it is later determined that it actually is EB, that's ok as in the beginning of life almost all applicable precautions are the same, for all forms of EB - and if it is not EB, you can do no harm.

Of course, the information presented here can never and should never replace a discussion with experienced EB-therapists. You should show a bit of courage to obtain all the information you need to gain confidence in dealing with this newborn child as soon as possible. Information and education about the disease as well as good and clear instructions for the next steps to take are in our view necessary, for the basis on which you can accompany your newborn and later your growing child, as affectionate but also safe parents. Trust the fact that like many other parents and families you will create a loving environment to accompany this child with their special needs through life.

There is a suspicion that EB exists when immediately after birth or within the first few hours and days blisters and/or wounds are observed on the skin of a baby. It may be that only a very few small blisters are seen, but can also involve large body surfaces covered in sores and wounds. Sometimes, the skin on a body part appears to be absent entirely, particularly on the foot. This of course is for parents, midwives and the medical staff at first a great fright, and it takes most everyone completely by surprise. In the normal preliminary examinations during pregnancy, these skin problems cannot be observed, they appear therefore unexpectedly.

At this time a lot of medical tests and examinations were probably very quickly conducted on your child to find out what your child is missing. If you are now

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reading our EB-manual, we assume that all other diseases that may appear similar were excluded by your doctors. He has told you your child has a suspected Epidermolysis bullosa. A rare disease of which you probably did not even know it existed. Within these pages we want to try and give you help to better understand what's going on with your child and how to best care for your child. What EB actually is and how it is caused, you can see in a separate chapter „[General Information about EB](#)“ described.

When a newborn baby comes into the world and it is suspected that they may have EB, this usually triggers a series of reactions. The parents especially have mood swings during the first days of the baby's life. From the time they are informed that "something with their newborn is wrong", until the final diagnosis the parents understandably experience uncertainty and fears, which are very difficult for them to bear. Ultimately it does not even matter what the medical diagnosis is.

It may take a while until they can tell you exactly which form of EB; you now have to live with. No final diagnosis can be made just by the appearance of your child's skin (many blisters or a few blisters, small or large wounds ...) one can definitely not draw conclusions on what the final diagnosis is. Babies with extensive wound surfaces after birth can have milder forms of EB, and some initially mild looking type is ultimately a very serious one. Even care givers experienced with EB can be fooled. So please wait for the final outcome of the investigation, even if it is very difficult and puts your patience to the test. Sometimes the diagnostic tests progress very quickly, sometimes it can take longer for many different reasons. In some cases it can even take months before we know the exact form. On the one hand, there are only a few laboratories worldwide that can perform these diagnostic tests and on the other hand it can be very difficult to find the exact underlying genetic change causing the disease.

There are over 30 different forms of EB, all of which we have been described in our manual. However, we strongly advise you to read this information only if you know exactly what form of EB has now been proven to exist in your child. The general information about EB, of course you can read at any time.

No matter which form of EB it is now, it always will be the same: It's your child, and your child needs, you and all the love and care you can give. No one can predict how each individual case, how living with EB, will exactly look for this new little person. This will only show over time. Despite, or perhaps because of this you have to be aware that a little person has been born into the world. In addition to medical

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skin care and meeting basic needs (like food, warmth, etc.) and safety, do not forget that also security and love need to be given.

While doctors are busy determining the exact diagnosis for your child, you should initially focus toward welcoming your baby into your life - even if it has surprised you with this unexpected challenge. Even if EB is suspected: you can and should touch your baby, even if their skin is very vulnerable. This is true even if your baby is in an incubator, however it is only used in medically well justified cases (e.g. after a premature birth) because the heat can cause increased blistering.

It is best to touch your child using bare hands and avoiding gloves. If this is not possible due to medical reasons (which your doctor should decide!) then you can wear the type of gloves used in the hospital. If you do need to wear gloves, you should apply a high-fat cream or petroleum jelly over the gloves directly after putting them on. Especially in the first days of life, the skin in some newborns with EB are so sensitive that there is a risk that parts of their skin will end up sticking to the gloves. After a few days you will know if this is the case with your child. In most cases you will find that you can cuddle and touch your child easily albeit cautiously - without causing it any harm. And if it gets one or the other blister due to an insecure touch, then do not blame yourself. Try to find out how it happened and learn from it. Do not make yourself crazy over it: these blisters arise very often without apparent reason; they just belong to this disease. It will never be possible to avoid all the blisters before they develop. Particularly sensitive areas are therefore protected by dressings. Body contact and cuddling a baby with EB is just as important as with any other child, and far more important than avoiding development of one or more blisters. One or the other "cuddle bubble" harms your child less than the anxious avoidance of body contact with your baby.

You should learn as soon as it is possible how to care for your baby by yourself, but also how to give it security and physical closeness without hurting it.

If it is possible, then seek contact with an experienced therapist and caregiver. Often this contact is already mediated by the hospital where the child was born.

For the care of blisters and sores that occur in EB, there are some special needs that you have to consider. Therefore we have covered this topic in a separate chapter [„Wound Management“](#).

## **Suggestions for Dealing with Babies who have Vulnerable Skin:**

There are a few possible ways and different behaviors with which you can protect your child's sensitive skin and we want to give you some suggestions:

Babies who have EB should initially not be lifted from under the arms, as this can very easily form large blisters under the armpits. There are babies in which the latter is quite possible despite EB, which depends heavily on the subtype. From the beginning of life you usually can not exactly say which form the baby has, yet you should still be cautious at first anyway. With time you will find out what is working well with your baby and what is not!

For example, you can place your baby on a small soft padding such as a sheepskin, a thick blanket or pillow. In this way, the baby together with the underlying padding, can be picked up to avoid further injury.

If you want to lift the baby without padding, you flip it first carefully from their back to side position, then put a hand gently under their buttocks and the other hand under the head or neck, the baby can roll back slowly and then you can gently lift the baby up.

Caution is also advised with the use of pacifiers: It may lead to blister formation at the edges of the pacifier, as well as the lips and mouth area. Despite EB some babies have few or no problems with it.

With clothing for your child you need to consider a few things. Places where a piece of clothing has buttons, zippers or thick seams can hurt your child's skin. In order to protect their skin before this happens, you can use one piece "soft bodies" inside out, this way there are no hard seams near the baby's skin. For more suggestions see the chapter „[Clothing](#)".

## **Nutrition:**

Contrary to all fears, breastfeeding a baby with EB is still quite often possible and should at least be tried. Breast milk is still the most appropriate food for a baby, and also breastfeeding encourages the positive bond between mother and child. Breastfeeding is therefore highly desirable in children with EB. This applies to all forms of EB, because even in severe forms of EB with mucosal involvement, breastfeeding is the gentlest way of eating. A good lactation consultant can provide

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a lot of support - if she/he has patience, is informed a bit about EB and your baby is well observed.

But if for any reason breastfeeding is not yet possible, you should not be stressed: of course you can provide your baby with good nutrition using infant formula and the mother-child bond can be promoted in other ways.

### **Pain:**

If you have to deal with EB, then you will also often encounter a particularly serious issue: the issue of pain. It is a fact that there are a number of causes that can lead to pain in EB. From the beginning you were told that you do not always have pain and especially not occurring continuously. Experience has shown that especially the dressing change is a time that is connected to with pain for the child. For this reason, it is important to develop an appropriate procedure for your child for changing the dressing. You can and you should get help as soon as possible from people who have experience with EB!

During the first phase of life, experience has shown that it is not so easy to distinguish if a newborn child with EB is experiencing pain now or is not. This is as difficult for the medical staff as it is for the parents. A baby with EB cries and/or screams and does this for many different reasons, just like babies with healthy skin. In addition, a child's behavior is of course influenced by its particular personality. Some children complain more loudly than others, some are rather quiet from the beginning, some are impatient and fidgety, others seem to be content with themselves. In this case close observation of the baby and good communication between the attending physicians and family are particularly important.

Much of what is interpreted as pain in children with EB, in fact has very different causes. You will learn very quickly how to find out if your baby actually has pain or whether his cries are simply "baby related". If it is hungry or wet, or if it just wants your attention. If this baby is your first child, it might take a little longer than if you already have other children, but you will learn just like so many other parents - and certainly faster than you think!

However, if you find that a baby is actually suffering from pain that can only be treated by drug therapy, then you must act quickly with the administration of a sufficient amount of painkillers. There are several possibilities. Medical monitoring of

pain management is essential and a good and open communication between parents and physicians is essential.

### **Preparations for the First Time at Home:**

As soon as it is medically justifiable and once you get the confidence as a parent, the discharge home can be slowly scheduled. You should prepare for this as much as possible while you are still in the hospital. Besides all the preparations you have probably already taken for your baby, you may need to add a few more things. A soft pad for the crib is useful because babies spend a lot of time sleeping. Sheepskins of good quality are a good example, or even a soft blanket or flat cushion. Perhaps you have already found out in the hospital, on which pad your baby feels most comfortable. Then use this or something similar even at home.

You may have already received as gifts, purchased or been lent various items of clothing. You need to check them all out, and sort out those that are soft, do not have seams that are too thick and the possibility of any pressure points. Some garments can be worn turned inside out, others will have to be sorted out and not worn.

Since you always need the dressings for your baby's dressing changes, you need a place where this dressing change can be carried out. Caring for a child with EB's dressings can often take a very long time; therefore it must be ensured that the caregivers (parents, relatives, home health nurse) find a good suitable workplace for this. Consider where it is best for you to do it at home, or where you may be able to set up a new dressing table. Often this will be the place where the diapers are changed. Check to see if you and another person can also stand there for longer periods without straining your back by bending improperly. Especially in the beginning, you will probably need help with dressing changes; therefore, a second person should also have room to stand in a proper position next to the changing area.

You should feel the changing pad and test whether it is suitable for your child, otherwise you may have to purchase a new one. In most cases it is usually enough to use the existing changing pad, with an extra soft blanket or towel and on top of this a soft cloth diaper. Next to the dressing changing table you will also need a space for dressing materials.

Please, you must avoid in any case your child's crib as the place for dressing changes. We really strongly advise you to use the crib, because the bed should be a safe place for the baby and then later for the toddler. The crib is a place to which it can retreat to, where it feels comfortable and safe, away from unpleasant or painful procedures such as the dressing changes.

Look as soon as possible for a pediatrician or family doctor that is willing to accompany you and your baby, to whom you can turn to, and to whom you have confidence in. Because this disease is so rare, it may be that he or she has not had much or no experience with a child who has EB. But this is not so important, what counts is the willingness to stand together with you in this new situation and to be available for you as a first point of contact for questions and issues and to help you with other problems not caused by EB. A person that works with your experienced EB-specialists, to search for the best treatments for your child. This is the basis for successful medical care of your child.

Soon it would also be useful to think about who can help the parents with the care of the child at home. These could - for example - be relatives and/or friends. For the first time at home it is often helpful when a home health care nurse is organized, so she can help with dressing changes. Unfortunately, this is not always possible or available since it is not offered everywhere. But where it is possible, you should absolutely use it!

We also advise you to accept social counseling, which involves getting informed about the financial aid available to you in your situation and where you need to apply for this. That sounds very pragmatic and it probably is. But your child deserves the best possible care and you should not have to deal with financial problems, so you may and should be well informed about your rights!

We are fully aware that this first information is only a start and that you certainly have many more questions regarding your child and his future. This is according to the eventually diagnosed subtype which indeed is very different in each form, therefore we cannot go into detail with all the questions you have on your mind. We therefore encourage you to seek experienced contacts that are determined and ready to help you to answer the many questions posed during the beginning of a life with EB. They are especially good and will help you get through life with EB despite all the challenges!