

## **EPIDERMOLYSIS BULLOSA SIMPLEX LOCALIZED, KINDLER SYNDROME AND ITCHING**

### **1. Introduction**

Years ago, around 330, the physician Samuel Hafenreffer said that itching is "an unpleasant sensation that provokes scratching". There is not a better definition even today. The medical term for itching is "pruritus", prurire the Latin term meaning "burning". Itching is a particularly agonizing challenge for many people with EB. The itching is very uncomfortable, it has a spontaneous response - the scratching. It can have serious consequences in EB, because new blisters and sores usually form immediately in the places you have scratched. It is necessary to at least relieve itching and it is often possible.



#### **Important points in a nutshell**

- **Itching is a common problem for many people with EB.**
- **There are different causes of itching.**
- **There is no "silver bullet".**
- **You can often provide relief through a combination of general measures, good skin care and medication.**

## 2. General Information

Although itching is a very unpleasant skin sensation and occurs in many skin diseases, we still know relatively little about the mechanisms that lead to its origin. In the skin, there are so-called "receptors" (perception points) for different sensations such as pain, cold, heat or pressure. For a long time it was believed that the same perception points that are responsible for the sensation of pain were also responsible for itching, involving to varieties with the same sensory perception points for pain and itching. A big difference however, is that the perception point that can cause itching is in contrast to the one for pain only in the skin. Meanwhile, there are recent findings that suggest that perhaps there are separate neural pathways for the itching, but not all transmitter substances and their interactions are known. The phenomenon of the induction of itching is actually not really clarified.

Itching is usually "answered" with scratching or rubbing. This leads to inflammatory reactions of the skin, causing a number of neurotransmitters to be released from the cells which further strengthens the existing need to itch. We have all had this experience, often after an annoying insect bite: first a stimulus, you scratch and scratch, it briefly gave you relief but if you stop, the itch is worse than before. Then you scratch again.

What ever the perception points may be, in each case a stimulus to the perception point is always needed. This can be mechanical or chemical in nature. Inflammation of the skin is often the cause of the itch. They are the messengers to the inflammatory cells, especially histamine which produces the itch by stimulating the perception points for the itching. The stimulus is forwarded via nerve fibers to the spinal cord and the cerebrum and perceived as itching.

## 3. Epidermolysis Bullosa and Itching

Itching is unfortunately a very common problem for people with EB. The frequency and intensity that occurs with itching differs significantly in the individual forms of EB. Yet there are also large differences within the same form of EB between each individual concerned. While for some only occasionally itching creates a problem, in others they suffer very pronounced itching that is constant, intense and agonizing.

Itching is unfortunately a very common problem for people with EB, but fortunately in the localized form of EBS and in the Kindler Syndrome it is rare.

Since itching normally leads to scratching and scratching EB-skin promotes the formation of blisters, it is of particular importance in EB to interrupt the vicious cycle of itching and scratching.

The main causes of itching in EB are probably dry skin, healing crusty wounds, overheating, inflammation and persistent skin damage caused by the constant wound healing.

Some sufferers report that an itch mainly occurs when the healing of the blisters begin. Maybe the itch in this case is limited by two factors: one is a situation that is similar to a light (not caused by bacteria) inflammation. Inflammation accompanies the healing process. The other factor often comes from dryness of the wound area, when the wound edges pull together. Both the dryness and sensitivity to inflammation could be the cause of itching during the healing process.

When there is prolonged itching in those effected by EB, you have to think the cause maybe coming from something completely different. In EB not only the blistering and the wound healing process can trigger itching, other secondary problems of EB can be the cause or intensify the itching. Anemia due to iron deficiency and/or malnutrition in a person severely underweight are common in severe forms of EB and can induce or exacerbate itching. You need to be aware that a number of other diseases may be associated with itching. EB does not exclude the possibility of other diseases causing the itching. The following can also influence or enhance the itching mechanism under certain circumstances in people affected with EB:

- All types of skin inflammation
- Medication and food
- Environmental allergens (eg. flowers and grass pollen)
- Immunogenic or allergenic substances (antibodies, lymphocytes)
- Substances that are released or introduced by insects and parasites
- Contact materials (allergens such as colorants, perfumes, fragrances)
- Persistent skin damage (dehydration, sunburn)
- Metabolites from internal diseases
- Psychological stress (enhances)

If one of the above are a possibility for the cause of itching, then it must be treated accordingly. There is a great advantage of having a family doctor or pediatrician who knows you and your family well. It is not always easy to really find and treat the underlying cause of the itching. In addition, itching is usually caused by a combination of factors in EB. And sometimes it is quite difficult to treat a cause without worsening other issues.

## 4. Treatment Possibilities

Severe itching impairs the quality of life significantly. Some sufferers feel itching is even more distressing than pain. Therefore everything needs to be done to try all treatment options available. No one can promise you that itching is so treatable that it will completely disappear, as the circumstances involved in itching are too complex. But it should be possible to reduce the itching, at least to a tolerable level.

Depending on the suspected cause of itching different approaches are recommended, these can be nursing care measures, medications or behavioral therapeutic actions.

In addition to fighting known causes (if they were found) there are a number of things you can do that can alleviate the symptom of itching, even without knowing the exact cause. In the following section we'll give you a few tips that you can use to help relieve itching for you or your loved one. Try them out and see which ones you can use or seems useful in your individual situation.

### 4.1 General Preventative Measures against Itching

- Drinking enough (to prevent dehydration)!
- Do not overheat rooms, install air conditioning (apartment, car)
- Avoid use of desiccants such as gels, alcoholic solutions for rubbing and powder
- Sparing use of soaps
- Tepid water is preferable for bathing and showering, you should not take too long and it should not be too hot. At the end (if tolerable!) try a cool shower, as this closes the pores of the skin and does not allow too much moisture to evaporate

- 
- Damp cool compresses of different types can also relieve itching for example a black tea compress: black tea should be left to sit 15 minutes and then cooled, it contains tannins which may also have a pure cooling effect
  - Light clothing made from allergy free materials such as cotton and silk (for example Dermasilk®)
  - Bed linens made from cooling materials (eg. Microair®)
  - Fingernails should always be cut short so scratch injuries are reduced
  - In the night cotton gloves can be worn to at least limit the possibility of scratching
  - Relaxation techniques can also be very helpful to learn, you should look for opportunities to do this locally (autogenic training, progressive muscle relaxation according to Jacobsen, yoga ...)

Itching worsens especially in the evening with children, yet can also be a problem for adults. In the German language there is a suitable expression: they are really "exhilarated", which generally speaking is due to the diverse experiences of children and adults everyday. With those affected by EB it is no different, but it can have serious consequences. Therefore it can be helpful for both children and adults to develop a fixed bed time ritual which has a calming effect and is not causing additional excitement. An example would be a cup of tea or cocoa, quiet music or a good book before going to bed. Television, computer games and arguments should be avoided for at least 2 hours before bed time. This is not always easy to do, yet one should try his best!

#### 4.2 Local Therapy and Skin Care

There are a variety of products available that lead to relief of itching from various causes. Regular skin care, also for skin that is not effected by blistering, is important, in any case to prevent dry skin. There is also bath products, lotions, creams and ointments that are especially effective against itching. Many of them contain zinc or urea and are well suited for EB. Unfortunately what products can specifically help your situation can not be predicted. It is best if you try out together with a doctor or a nurse miscellaneous products to find the most suitable for yourself. Always ask whether a product that is recommended to you may also be applied in the vicinity of wounds or on open wounds. Some products (for example

with the active ingredient capsaicin) are in fact not for open wounds. If you need assistance in finding the correct product or you want to ask whether a particular product might be suitable for you, then you are always welcome to ask for help from our outpatient clinic in the EB House Austria.

### 4.3 Pharmacologic Measures

If all general measures have been exhausted and you have also tried enough products that have not provided relief, then taking itch-reducing medication may be necessary. If this is the case so-called antihistamines, which come in tablets, syrup, drops, ointments or as injection can be prescribed. You may be familiar with antihistamines as they are frequently used for the treatment of all types of allergies. Some of them make you tired and are therefore more suitable for the evening dose. However, there are now some which hardly make you tired and are suitable use during the day. Please seek advice from your physician/health care professional as to which of these medications is best for you.

Sometimes a brief treatment with a Glucocortikosteroid (simply referred to as "cortisone") containing preparations for intolerable itching, might be necessary as a cream or in tablet form. Short term (about 2 weeks) is recommended and also useful in EB, yet long-term use should be avoided. Such treatment must be used under medical supervision.

Furthermore, there are also other medications for severe itching in EB that have occasionally been used with good results. Whether these could also be useful in your situation needs to be discussed with an experienced EB-specialist.

In summary it must be said that there is no simple "magic formula" for the treatment of itching associated with EB. But there are a number of things you can do that can help to reduce the itching at least to a tolerable level. Do not give up too quickly, with a little support and a lot of patience you often can achieve a lot!