

SKIN CANCER IN OTHER FORMS OF EB **(not for EBS generalized severe, DEB, JEB --> separate sections)**

1. Introduction

In some forms of EB, there is the possibility of developing some form of skin cancer as a serious complication. The reasons for this are still not entirely clear, but the effects are often dramatic for those affected. The earlier the signs of cancer are identified, the greater the chance is for a cure! This also applies to the dreaded skin cancer often seen with EB. Special care and attention is necessary, therefore we will make you familiar with the warning signs that you need to pay attention to, in this chapter. We will also give a brief overview of the treatment options. We would like to inform you to pay attention only to the chapter that pertains to the form of EB that you or your family lives with. Only these chapters are actually relevant to you.



Important points in a nutshell

- **Risk for skin cancer**
 - **is very high in dystrophic EB**
 - **is high in junctional EB**
 - **is slightly increased in EBS generalized severe**
 - **by all others no higher than for the healthy skin population.**
- **Early detection is vital!**
- **Prevention: Regular examinations of the entire skin surface every 6 months.**
- **Warning signs: Wounds**
 - **when the healing process is to long**
 - **the wound is somehow different**
 - **is especially painful.**
- **Treatment options: all should be surgically removed.**

2. General Information Section

Cancer - this word alone makes most people shudder. Why is this by some people – yet still not with all people! Why some forms of EB develop skin cancer, has not yet been fully explained. To date very little is known about the precise cellular and molecular mechanisms that ultimately lead to tumor formation. One can imagine that in EB the repair mechanisms of the skin cells are particularly stressed during your lifetime. It now seems to be that these repair mechanisms can be restricted in their function with increasing age. Thus it may happen that sometimes skin cancer cells arise. An essential characteristic of a cancer cell is its unchecked growth. If such a cell multiplies, the body cannot prevent this. After a few weeks many skin cancer cells are already present, so that the area affected is visible. **The sooner this area is noticed, the better the chances are for a successful treatment!** It does not help to simply ignore the danger. It is much better to consciously deal with this threat, to respond quickly in the event of suspected skin cancer you can save precious time.

We need to distinguish the different types of skin cancer in EB. In most cases a form called squamous type develops in the cells of the upper skin layers. This type of skin cancer is the most feared. It is a malignant tumor which starts in one location and changes into a destructive and rapidly growing tumor in the surrounding tissue and after some time also frequently metastasizes, thus forms secondary tumors.

This form of cancer most often occurs in people with severe generalized dystrophic EB. It can also occur in other dystrophic forms of EB and occasionally with junctional EB. With increasing age the risk for skin cancer also increases, so checkups in which the entire skin is examined is more important. Unfortunately in some cases, skin cancer was found in adolescents, therefore semi-annual examinations should be the rule. Once when skin cancer has been detected, the examination must be performed more frequently, about every two or three months.

In rare cases, it may also result in the formation of other types of skin cancer. A distinction must be made especially between basal cell carcinoma, which is called "white skin cancer" and a melanoma, "black skin cancer ". At this point in time there is no known direct relationship with EB. There is also no indication that the incidence of these cancers is elevated in people with EB compared to people with

healthy skin. The only exception is EBS generalized severe, where the risk of white skin cancer (basal cell carcinoma) seems to be slightly higher.

The possibility that you developed a form of skin cancer over the course of your life is the same for all people, so of course in people with EB. The incidence is increasing throughout the world and everyone should be familiar with the early signs of the "black skin cancer" (melanoma) and the "white skin cancer" (basal cell carcinoma).

Briefly summarized: When a dark pigmented mole's appearance changes, is asymmetric or has more than one shade of brown or starts to bleed, caution is advised. The same applies to reddish, rough skin areas on the head and facial area, or poorly healing areas where scabs repeatedly form. Sometimes even small nodes form on the skin that slowly grows in size. If you notice any of these early signs, you should undergo a medical examination.

The same precautionary measures that are valid for all people also apply to people with EB: Excessive sun exposure, especially sunburns should be avoided, apply a good UV protection from sunlight and perform regular mole checks.

At this point the so-called "EB nevi" should be mentioned. These are EB typical pigmentary changes that can be best compared in appearance with moles. They often occur in the area of blisters, can be quite large and are variable in shape and size. Sometimes they completely resolve and disappear. These EB nevi should always be examined during skin checks. No case has yet been described in which a pigmented nevus has developed into a malignancy, but since these pigmentary changes and the early stages of melanomas are similar, they should also be examined. So play it safe, that there is no malignant skin cancer hiding among the pigment changes.

For all forms of skin cancer: regular skin examinations cannot prevent the occurrence, but with an early recognition, therapy is often successful in many cases.