

ANAESTHESIA

GENERALIZED SEVERE RECESSIVE DYSTROPHIC EB AND GENERALIZED INTERMEDIATE RECESSIVE DYSTROPHIC EB

This section provides advice of what to observe in case someone affected with EB needs anaesthesia.



Important points in a nutshell

- **Anaesthesia means using an anaesthetic agent for suppressing pain during examinations, treatment and operations.**
- **In principle anaesthesia is possible for those affected by EB. However, certain precautions must be taken for various reasons (including for example, the increased vulnerability of the skin and often the mucous membranes, existing limited ability of opening the mouth, etc.).**
- **Good planning, starting well before the already planned intervention is essential!**
- **What needs and specific practices are used for patients with EB, needs to be communicated to everyone involved (doctors, nurses).**
- **In an emergency, if these precautions require too much time - then lifesaving measures have priority!**
- **Despite all the caution and experience, blisters and sores cannot be completely avoided, especially in severe forms of EB.**

1. General Considerations

There are several types of anaesthesia:

General Anaesthesia:

Also referred to as anaesthesia, induces you into a deep sleep-like state.

Local anaesthetic technique:

Is also called local or regional anaesthesia. When using local anaesthesia, the examination or surgical area is numbed by injecting a local anaesthetic.

The anaesthetist will discuss with you, which type of anaesthesia is most appropriate for you or your child for the planned intervention. Other methods that may possibly come into question may also be discussed and the advantages and disadvantages they have.

Patients affected with EB may require anaesthesia for various reasons: firstly, complications from EB sometimes require surgery (oesophageal stricture, finger adhesions, severely decayed teeth,), and secondly it can also be for **other reasons that have nothing to do with EB, where anaesthesia is necessary** (appendicitis, fractures ...).

There are several reasons why an anaesthetic for patients affected by EB is more challenging, such as:

- The great vulnerability of the skin and/or the mucous membranes depending on the form of EB.
- An increased heat and fluid loss through the skin wounds.
- Depending on the type of EB the patient is affected with, they may have a reduced mouth opening and/or contractures of their large and small joints.

Therefore, there are various procedures that must be carried out a bit differently for those affected with EB, unlike those procedures used for patients with healthy skin and mucous membranes:

- They need to be lying on a soft padding during the operation and careful repositioning without friction must be ensured. To make the repositioning gentle, there should also be the possibility that the patient can move themselves to the OP table, or the patient's repositioning is done together with the padding on which he is lying-in, so that the patient is not even touched.

- Heating blankets and a preheated procedure room prevents heat loss of patients affected with EB.
- Venous cannula, breathing tube and other tubes, ECG electrodes, etc. should not be fixed down with adhesive tape. Instead for example, attachment with the help of micro-adhesive materials, by wrapping with gauze bandages or by sewing is possible.
- Depending on the form of EB it may be necessary that a thinner tracheal tube is used, rather than what is used in people with normal skin.
- Much more often ingenuity and imagination are required!

Pain relief for after the procedure must also be well planned; it's just as important as the pain relief before and during the procedure. It is often necessary to provide pain relief using a painkiller given intravenously, for example when swallowing is still painful.

If adhesive materials were used during the procedure, they can be carefully detached with a special spray that dissolves the adhesive (e.g. Niltac[®], Sensi Care[®], etc.). If that does not work, it is best not to try and remove most of these materials. They will eventually come off in the bath or overtime time on its own accord.

2. Who performs anaesthesia for the person affected with EB?

When someone affected by EB needs anaesthesia, for each procedure they require a good detailed plan of action well in advance of the procedure. Good communication between all participants (patients and their families, doctors, nurses) is needed. For the informed consent discussion with the anaesthesiologist ("anaesthetist") take with you all current medical reports and findings, also a list of the medication and dressing-materials that you use.

Since those affected with severe forms of EB more often need frequent surgical procedures due to the complications of their disease, procedures are usually done in centres where they already have experience with EB patients.

The anaesthesia can also be performed from a doctor who does not have experience with EB. In this case, it is important however that the doctor takes time before the

procedure to familiarise themselves in detail with the specific circumstances and precautions needed while caring for an EB patient (for example in the literature and/or contacting an EB-expert centre).

3. Generalized severe RDEB, generalized intermediate RDEB

In severe and intermediate recessive dystrophic EB anaesthesia is often required, either for dressing changes or due to the complications of RDEB (oesophagus stretching, hand surgery, tumour surgery ...).

The anaesthesia and the whole procedure or the examination must be planned well in advance of the actual procedure, giving careful attention to detail.

Before any procedure that requires anaesthesia an informed consent discussion with both the anaesthetist and the doctor who will perform the procedure (surgeon, dentist), will take place. It is very helpful if you take contact details of your supervising EB specialist and medical reports to this visit.

The procedure must be well planned and thorough, especially if complications of the disease have already occurred:

In most cases the mouth opening is significantly reduced, the tongue is often scarred and deformed and the epiglottis can also have adhesions.

All participating doctors and nurses need to know about the special operation procedures and precautions needed when caring for someone with EB.

If you or your child's mouth opening is reduced, you can even help ease the implementation of anaesthesia by:

In order to improve the **narrowed mouth opening**, there are **various exercises** you can do. Therefore each individual millimetre by which the mouth opening is increased, is a benefit!

A method that works well is to use the so-called mouth gyroscope. This was developed by the Chilean dentist Dr. Susanne Krämer. The mouth gyro is set between the upper and lower incisors and by rotating the gyro the mouth is stretched. The exercises with the mouth gyroscope are best when done in the days before a visit to the dentist or before a procedure with general anaesthesia.