

TEETH

Severe Generalized Recessive Dystrophic EB

1. Introduction

Dental problems can occur with different frequencies and different degrees of severity in many forms of EB. In some (not many!) forms of EB tooth damage is hard to avoid, since genetic enamel defects exist. But even with forms of EB where there are no enamel defects, normal dental condition often leads to tooth decay, because oral hygiene can be difficult due to blister formation or difficulty in opening the mouth.

With [appropriate care](#), a [dental healthy diet](#), [regular dental check-ups](#) and [professional oral hygiene](#), damage to the baby teeth as well as the permanent teeth due to caries in many cases is preventable. It is worthwhile in any case - despite all the other worries and problems caused by EB - to pay regular attention to your teeth and their care. When parents and children are well informed and from the beginning a good working relationship exists between the family, the supervising dentist and EB-specialists, the condition of the teeth can generally be positively influenced.



Important points in a nutshell

- **Dental care starting with the first tooth is important in EB to avoid many subsequent problems.**
- **It is based on regular teeth cleaning at home, at best twice daily with a soft toothbrush, with a small brush head and age-appropriate fluoride toothpaste.**
- **Find a good dentist as early possible, because professional cleaning and rapid intervention when problems arise is necessary.**

2. General Information

In epidermolysis bullosa dental and oral hygiene may be difficult due to blistering of the mouth. The best possible dental and oral hygiene is necessary, despite the risk of blistering, to avoid developing dental caries. A healthy and functional set of teeth is very important for eating and nutrition.

People with [Severe Generalized Recessive Dystrophic Epidermolysis Bullosa](#) have several [problems](#) with their teeth ([tooth enamel](#), [caries](#)) and/or their [oral mucosa](#). [Excessive tooth decay](#) with risk of premature tooth loss affects all severe forms of EB. [Blisters and erosions on the lips, tongue and oral mucosa](#) complicate a consistent oral hygiene and food intake. Frequently it comes to [changes of the gums](#) caused by [chronic inflammation](#) of the gums (gingivitis) and [mechanical damage](#) by the necessary dental care

Also in people [with Recessive Dystrophic Epidermolysis Bullosa](#) tooth decay even with normal tooth condition can develop due to difficult oral hygiene from blister formation or a limited ability to open the mouth. Younger patients often suffer a rapidly progressing destruction of the teeth. As a result, this creates additional problems in food intake. However, the prognosis of the disease depends largely on an adequate diet and thus on the state of the teeth.

[Nutrition](#) in itself is a big problem; often the person concerned cannot eat - and/or - swallow the calories they need, so they will often consume a lot of high calorie foods, often food containing a high amount of sugar. This is understandable, yet it does not make the dental problems any simpler.

Once a cavity occurs you subsequently develop pain. When this is the first dental visit that has taken place, it is not uncommon to be associated with a poor experience. It may be that made fillings are needed or even teeth are pulled. This is sometimes only possible under general anesthesia, which is often not easy in EB.

In [Recessive Dystrophic Epidermolysis Bullosa opening the mouth](#) (to brush your teeth, at the dentist) also may often be [very restricted](#), so basically brushing is already [difficult](#).

Frequently [blisters form](#) on the [oral mucosa](#).

In recessive forms of EB chronic blistering and erosions can lead to scarring of the oral mucosa. Resulting in adhesions at the tongue base with restriction of tongue movement and speech, loss of tongue papillae and taste buds though too the running mucosal folds. Likewise, milia ("milia, grits") occur with preferential localization of the hard palate. Problems with swallowing and eating, as well as biting and chewing can be exacerbated by this sequence of complications.

3. Options for Prevention of Dental Caries

With appropriate dental [care](#) at home, [a healthy dental diet](#), [regular dental check-ups](#) and [professional oral hygiene](#), the destruction of the milk teeth as well as the permanent teeth due to caries in many cases is preventable.

Special Measures:

Consistent daily teeth brushing/oral care

Regular professional cleaning by a dentist/dental hygienist

Fluoride prophylaxis

The classical [pillars of prevention](#) include "[diet](#)", "[oral hygiene](#)", "[Fluoride](#)" and "[fissure sealing](#)". Fluoride tablets now play a secondary role. More important is the topical [fluoride treatment](#). This includes child fluoridated toothpastes and fluoridation with coatings or gels applied by the dentist. Mouthwashes are only useful if the children have learned to spit the mouthwash back out properly. The nature and frequency of the respective fluoride treatment should be discussed with your dentist. Professional oral hygiene in a dental office, in conjunction with the examination of all teeth should be done at least 4x annually, in severe forms of EB more frequently.

When special requirements are needed, oral hygiene can be supplemented by special mouthwashes such as chlorhexidine. There are chlorhexidine rinses as well as gels; the mode of administration should always be discussed with a dentist. This treatment usually takes place as a short-term treatment for about one to two weeks, which may be repeated periodically as needed. Prolonged use will lead to discoloration of the teeth and an irritating taste left in the mouth. The repeated use of chlorhexidine about every 2-3 months for a period of 1 week has been proven to be effective in children with EB. A more recent approach is the use of pastes made from a calcium and phosphate base. This has to be used in addition to fluoride toothpaste and is well accepted by most children because these pastes do not

irritate the mucous membrane and have a pleasant taste. These pastes (e.g. GC tooth mousse) are only available from dentists or from the Internet.

In addition to the above-mentioned fundamentals of prevention, namely "diet", "oral hygiene" and "fluoridation" is dental sealing of molar fissures. Dental sealing is the only measure which lies solely in the hands of the dentist. For this treatment a plastic coating is applied to the surfaces of the molars (back teeth used for chewing) this can prevent the development of dental caries. The teeth need to be kept completely dry for a short time, which can be difficult when you have a hard time opening your mouth.

4. Age Related Information

4.1 Infants

For the "primary prevention" of the so-called "[early childhood caries](#)" (formerly called bottle tooth decay) you should pay attention to the spread and [prevention](#) of dental caries. This usually starts with a rapidly progressive destruction of the upper incisors. While it used to be caused by instant baby teas that were offered in the baby bottle, today it is rather caused by supposedly healthier fruit juices. Pure fruit juice also contains fructose which corresponds to approximately a 10% sugar solution. In combination with a very low pH in juices, continuous bottle drinking leads to massive early destruction of the milk teeth. Studies have shown that children with early childhood dental caries that have been treated are still at a higher risk of developing dental carries later in life.

Infants are often found to have a greater acceptance of a toothbrush during the so-called "oral phase" because the mouth is the contact element to the outside world. This is just as frequently followed by an absolute "Toothbrush refusal" phase.

Even with "tooth brushing opposition" consistency is important, it often helps to integrate teeth brushing into a ritual. The objectives in this age group are preventing the transmission of germs and making oral hygiene a "[daily habit](#)".

The bottle should be [weaned as soon as possible](#), this is usually around the first birthday and the child should then become accustomed to drinking water from a cup.

In the so-called "window of infectivity" (window for infection), between the age of 6-30 month - is the phase where the "milk teeth", primary dentition break through the gums. During this period of time, the child can be infected with caries from contact with the saliva of caregivers, usually their mother. The top priority at this time is to keep the milk teeth healthy. Therefore, an early examination by a dentist is important. In this way, the visit is experienced in a relaxed atmosphere and time can also be used by the dentist for educating the parents.

4.2 Toddlers

Starting between the ages of 2 - 3 years, most children want to try to brush their own teeth. This is important so that they can learn a certain technique and not lose the desire to brush their own teeth. Kindergarten aged children are particularly susceptible to praise and reward, they often live in a fantasy world that can be supported by stories or songs in oral hygiene. Make brushing teeth fun.

The best way is to always clean with the same technique; we recommend the "CEI" system that is - chewing surfaces 1st, then Exterior surfaces followed by Interior surfaces. Tooth brushing should be started as soon as the first tooth breaks through the gums. No later than the 1st birthday a tiny bit of fluoridated toothpaste should be used, at least 1 time a day, preferably in the evening. It is particularly important that the parents help with brushing and check for spots that are missed. When the permanent teeth breakthrough the gums, you should change the toothpaste from a child's to a junior type of paste. Tooth paste containing Menthol can cause discomfort for children with EB. In this case a tooth paste without menthol, yet containing fluoride is a good alternative.

Particularly **dangerous are frequent snacks!**

Even seemingly healthy things such as bananas can cause caries due to their stickiness. If you cannot reduce eating snacks due to general medical reasons and teeth cleaning after every meal is not feasible, you can make do with a little compromise: for example a dental chewing gum, which stimulates the flow of saliva and promotes self-cleaning or at least rinsing your mouth with water between meals and snacks.

4.3 Children

It is not until [about 10 years of age that the fine motor skills are advanced enough that brushing your own teeth is possible](#). Electric toothbrushes are from about kindergarten age just as suitable for use as manual toothbrushes. Many children find electric toothbrushes simply "exciting" and are therefore more motivated to brush their teeth. Electric tooth brushes are especially helpful with disease-related limitation of fine motor skills in generalized EBS. It is also easier for the parents to use an electric tooth brush when helping the child get spots they have missed while brushing.

However, an effective plaque removal by the child himself is not possible until about the age of 10. Until then, the parents really need to check for spots missed. Good teeth brushing technique and [help from the parents is far more important](#) than the frequently asked question about the best toothbrush or toothpaste to use. True to the saying "you cannot teach old dog new tricks" means that behaviors that are practiced in infancy are very resistant to future changes. It does not matter whether these behaviors are positive or negative. In other words, at this age the foundations should be laid for a good dental health-conscious life.

4.4 Adolescents

The closer the children get to puberty, the less parents can influence choices in diet and oral hygiene. With pocket money they can buy sweets and oral hygiene is no longer regularly monitored by the parents. Negative motivation along the lines of "if you eat too many sweets and do not brush your teeth, you're going to get bad teeth" is usually unlikely to be successful. It is usually better to use [positive motivation](#), for example, "beautiful teeth look so attractive".

The aim of these measures is to motivate children from the beginning, so that [diet, professional prophylaxis and oral hygiene](#), at home are included in a [daily routine](#) so that as adults the risk of caries can be held correspondingly low.

Another topic at this age can be [braces](#). In such cases, a precautionary approach is recommended. Especially important is a good working relationship with the orthodontist.

4.5 Adults

In adulthood, the regular daily dental care, professional prophylaxis and dental appointments should be **routine**. Problems that might arise in recessive dystrophic Epidermolysis bullosa should be discussed with a dentist.

The **restoration** of already damaged teeth by removing corroded altered tooth substance and the **reconstruction** of the tooth structure and function of teeth, with fillings and "protective crowns" is sometimes necessary. The **extraction** of badly damaged teeth to eliminate inflammation is needed to prevent bacterial infections and abscesses. **Root canal treatments** are possible, yet treatment with implants in recessive dystrophic EB is rarely considered.

5. Special Problems and Therapy

At this point we would like to mention a few products to help make your search a little easier, with products that have proven effective for EB. However, we would like to point out that there are a large number of similar products that have the same or a similar effect. Which products you use, should be discussed with your dentist.

For rinsing and treatment of **open areas on the inner cheek, sage tea or chamomile teas have** been found to be useful. An oral ointment (Kamistad A gel, available in Germany), which also contains a topical anesthetic in addition to chamomile extract, is another option. Other topical anesthetic creams, also used for pressure areas from false teeth, are for example Solcoseryl ointment or Dynexan gel. While the open areas do not heal faster, it does take some pain away, for example before eating. For rinsing we can recommend Bepanthen mouth rinse.

EB children:

Non-alcoholic mouthwashes containing chlorhexidine ("CHX", has an antibacterial) and fluoride (caries protection) for the daily care:

Gum Paro ex 0.06% CHX + 950ppm sodium fluoride
Curasept rinse solution 0.05% CHX + 0,05% fluoride bottle 200 ml
Mirafluor Liquid CHX 0.06% CHX + 250ppm amine fluoride
Elmex caries protection 0.025% sodium fluoride rinse bottle 400 ml
Meridol med 0.2% CHX

What form the toothbrush should have should be discussed together, at this time for children with EB. Depending on the severity of the disease softer bristles or a small brush head may need to be used. Ultrasonic powered toothbrushes with a very small brush head have proven useful. Depending on the severity of the disease, the condition of the teeth and of course the ability to open the mouth one must improvise with children affected by EB. Also aids such as interdental brushes, Q-tips or gauze swabs can be used, the dentist or dental assistant should find the aids that are most suitable for the child.

Local anesthesia:

Whether a dental treatment can be performed under local anesthesia or not depends amongst other things on the cooperation of the patient, the possibility of adequate mouth opening and the extent of blister formation and the need for treatment.

General anesthesia:

In some cases it may be useful or even necessary to perform dental treatment under general anesthesia. Indications in favor of such an approach are for example, too small of a mouth opening, severe pain and blistering, or the need for extensive treatment.