

NUTRITION IN EBS generalized severe

1. Introduction

Food and drink play a major role in the life of all people, whether sick or healthy. And not just because food is essential to life, but also because food is important for the personal well-being and also within the family and society it plays an important role.

For example, our daily routine is marked by meals. For many families eating together is an important time when all members come together. Also holidays are celebrated with "feasts".

Food is therefore ideally not only for the absorption of nutrients, but also for pleasure! If the food is good, varied and composed of rich nutrients, it can also contribute significantly to your health.

A newborn baby and especially one with EB are very tiring and therefore you should also pay attention to yourself and your health by eating well and by indulging in rest and relaxation phases. Only then you will have enough energy to take good care of your child!

Below you will find general information and tips, for specific problems you should contact your dietician or health care provider.



Important points in a nutshell

- **In EBS generalized severe blisters and sores in the mouth and throat may occur.**
- **This lead to difficult food intake.**
- **Prevention through a balanced diet and individual nutrition counseling are recommended.**
- **All treatment measures should be accompanied by an evaluation.**

2. General Nutrition

All babies and children need good nutrition. It is the foundation for growth and a good quality of life. It is not possible to cure EB with a special diet (e.g. by avoiding certain foods or administering certain nutrients in large amounts), yet it can be supported by optimal nutrient absorption for growth, for the immune system and wound healing. The information provided here is general in nature and therefore cannot provide solutions for all situations. If you want to find the best diet for your child, please talk with a nutritionist. They can work with you to develop an individualized plan, which should also be frequently checked to meet the changing needs of your child.

A balanced diet includes in varying proportions:

- Proteins
- Fats
- Carbohydrates
- Fiber
- Vitamins
- Minerals
- Water

This is not as complicated as it sounds, since most foods contain a mixture of the various nutrients. A balanced diet can be achieved by combining many different foods. The more varied the diet, the more likely it contains a good balance of nutrients needed for the growth and repair processes of the body and providing energy. The chemical changes which the food is subjected to in the body are referred to as metabolism. The food requirements are relatively high in childhood and adolescence, while most of the growth process takes place during this period.

Protein

Proteins are particularly important for building a strong healthy tissue during childhood. In EB a high protein intake is needed during your whole life to support wound healing.

The main sources of animal protein are meat, fish, eggs, milk and milk products such as cheese and yogurt. Plant (vegetable) protein is contained in legumes (peas, beans and lentils), nuts and grains. A vegetarian diet is generally not recommended in children up to primary school age. Especially with EB there is a particularly high need for various nutrients that cannot be satisfied by a vegetarian

diet. Whole nuts (especially peanuts) should not be given to children under 5 years of age, since there is a risk of choking!

Fat

Fats are the most concentrated source of energy in the diet.

If your baby has a lot of blisters and sores, its energy demand increases because it needs to invest a lot of energy in the healing process. This is the case with some, but not all babies with EBS, babies with the type EBS generalized severe are especially affected. If your baby is one of those babies that need a lot of energy, you should take advantage of the high energy content of fat and fatty foods and let your baby have an additional portion.

Butter, margarine, whipped cream, oils, lard are obvious sources of fat.

Butter, margarine, whipped cream, oils; lard and gravy are obvious sources of fat. Hidden fats are for example found in whole milk, yogurt (especially full cream or Greek yogurt), many cheeses (including cream cheese), ice cream, meat (especially with visible fat), eggs, fatty fish (salmon, sardines), avocado or almond butter.

Carbohydrates

Carbohydrates are made up of a large group of energy-supplying foods, some of which (cereals, flour, pasta, bread, potatoes, fruit, legumes) include fiber, vitamins and minerals. Other foods in this group are pure energy suppliers, such as cookies, sugar, candy, honey and jam. Cakes and other desserts are valuable because of their energy content, but can also be sources of protein when they contain eggs and dairy products.

With EB all carbohydrates are important in the diet. The sweet carbohydrates should be eaten as often as those carbohydrates (cereals, potatoes) that are less sweet.

Fiber

Dietary fiber is the part of food that passes through the digestive system without being absorbed. Although it has almost no nutritional value, it is still very important for normal digestion and helps to prevent constipation. Fiber is found in cereals, whole wheat bread, whole wheat flour, whole wheat pasta, brown rice, lentils, dried fruits and fruit and vegetables.

If there are blisters in the mouth and esophagus area, chewing and swallowing of high fiber rich foods can be difficult. Citrus fruits (oranges, tangerines, etc.) and tomatoes can also irritate the oral mucosa. A high fiber diet is extensive and filling, but rather low in calories, since less food can be eaten.

Vitamins

Vitamins are nutrients that are essential for growth and health. For most people, if they eat a varied diet, the daily intake of vitamins is sufficient. There are many different vitamins which have different functions:

Vitamin A: Is found in liver, carrots, milk, margarine and oily fish (e.g. mackerel). Dark green, red and yellow vegetables contain retinol, which is converted into vitamin A in the body. Vitamin A is needed for healthy skin and mucous membranes, the eyes and to support the immune system.

Vitamin B group: Is found in dairy products, meat, eggs, bread, cereals and potatoes. The various vitamin B sub-groups have different functions for the optimal use of energy from carbohydrates, for blood formation and skin regeneration and for the support of protein metabolism.

Vitamin C: Is found in fruits, especially in citrus fruits, also in vegetables especially green peppers and potatoes. Vitamin C is important for wound healing and helps the body in the absorption of iron.

Vitamin D: Is needed together with calcium to build strong bones and teeth. It is found in butter, margarine, oily fish, eggs and liver. The richest source is found in Cod liver oil. Vitamin D will only be effective in the body when the skin is exposed to sunlight. For all infants that are breast-fed, vitamin D should be administered in the form of drops (Oleovit) 1x daily. This is especially important for EB children who are barely exposed to sunlight on their skin due to the bandages and long-sleeved clothing. Therefore administration of Oleovit is widely useful beyond the first year of life.

Vitamin and Mineral Supplements

In people with DEB it can be difficult to eat sufficient amounts of food. At the same time, there is a higher vitamin and mineral requirements than people without EB. Hence, vitamin supplements may often be prescribed. Please be aware that an excessive intake of vitamins can be harmful! Please ask about the most appropriate vitamin supplements for your child.

Water

Although water is often not considered a nutrient, it is still essential for life. Adequate hydration also prevents constipation.

3. Nutrition Problems in EBS generalized severe

In EBS generalized severe blisters can occur in the mouth and throat and make eating painful. If your baby has painful sores in its mouth that can affect the breast feeding, you can apply a numbing solution directly onto the painful area such as Herviros[®] with a cotton swab before breast-feeding. Caution: If this solution is distributed throughout the mouth and throat, the "docking" will be disturbed.

If your baby's mouth is too sore to suck or it gets tired quickly you can express the milk or use a breast pump and feed the child with a spoon or a special bottle. In this special bottle called Haberman[®] from Medela (available from pharmacies), the nipple is designed so that no vacuum needs to be established for drinking and therefore the palate of the child is protected. It has a valve and a long neck that facilitates control over the flow of milk, so even babies with a weak sucking reflex get enough milk. The important thing is not too squeeze the bottle to tight or the milk stream is pressed into the back of the throat which can cause choking, whereby the milk can get into the lungs. Another possibility is to enlarge the hole of a commercial nipple with a sterile needle or you can make a small cross-shaped slit with sharp scissors. Then sterilize the nipple before use. Make sure that your child does not cough or choke due to the faster milk flow.

After the first year of life and in adults there are ways to make eating easier due to open wounds in the mouth:

- Offer food that is cooled- this may be perceived as less painful and therefore more likely to be accepted.
- Avoid acid (e.g., tomatoes, citrus fruit), spicy, salty and sharp foods.
- Soft or mushy, pureed meals enable the person concerned to eat quickly and less caution.
- Before eating apply a numbing solution such as Herviros[®] with a cotton swab directly on the painful area.

Of course eating is important yet frequently offering food or when the attention of adults mainly revolves around eating, children may lose the joy of eating which can significantly reduce the appetite.

4. Age Related Nutrition

4.1. In the First Year of Life

Breastfeeding and Breast Milk

Breast milk is perfectly suited for the growth and development of the baby. It contains valuable proteins (immunoglobulins) that are important for the defense against infections, as well as prebiotics (which favor the growth of normal bacterial colonization in the intestine). Breastfeeding may also reduce the risk of the child developing an allergy. Therefore, it is very beneficial for a baby with EB to be breast fed, even if it is only for a short period of time. Here are some tips that have been found useful by mothers with babies who have EB:

- Even if your baby has blisters in their mouth this does not necessarily speak against breastfeeding.
- You can apply lanolin (wool oil) to the cheeks, lips and palate of the baby and the mother's nipple to reduce friction, from searching for the nipple and sucking.
- Careful touching of the baby's lower lip with a finger or nipple encourages the child to open its mouth wide. When the baby does open its mouth, pull the baby toward your chest. Make sure that the baby is well "docked". The baby should have its mouth full of breast and should not be sucking only on the nipple!
- Reposition your baby often and let it suck for as long as it wants.
- Take your time, so that you both do not feel pressured.
- If your breast is very full first press out a little milk so that the child can grasp the breast better and not be choked when suddenly a lot of milk comes out at once.
- Do not be afraid to go to a breastfeeding counselor for open questions or uncertainties.

Bottle Feeding and Baby Formula

You may choose not to breast feed your child, but to provide nutrition with a baby formula. It may be that you give in addition to breast milk, bottle feedings or expressed breast milk to add nutrients, or to give the child fortified infant formula. For whatever reason, you can discuss an optimal nutrition plan for your child with your doctor. If your child develops blisters in the mouth that disturb it when eating, please refer to "Nutrition Problems EBS generalized severe" to find out what you can

do to make it easier to eat and drink. Such blisters occur mainly in EBS generalized severe, and rarely in other affected with EBS.

Weight Gain and Fortified Foods

To judge if a baby is developing well, can best be evaluated in an increase of weight, an increase in length and head diameter. In particular, your child should be weighed regularly to ensure that it is thriving. If possible, you should always use the same scale and weigh your child either always without clothes or bandages/or with similar bandages and clothing. If your baby is only slowly gaining weight, our dietician recommends that you add additional nutrients to expressed breast milk or give your baby formula that has a higher concentration of nutrients than conventional baby formula (e.g. Infantrini®).

Fruit Juices and other Beverages

Babies usually do not need any additional liquid when they are fed with breast milk or infant formula and only require an additional bottle with cooled boiled water when they have diarrhea, a high fever, profuse sweating or during hot weather. Fruit juices and sweetened teas are not necessary, as sweet drinks can reduce the appetite of the child. As a thirst quencher water is ideal. Please be aware that a low fluid intake may cause or aggravate constipation. If you notice that your child needs more fluids, but refuses water and unsweetened tea, you can give the baby well diluted baby juice (at least 1 part water to 1 part juice), available from various baby food suppliers.

Colic can be caused by swallowing air while drinking, especially when the hole in the nipple was enlarged. Make sure that the bottle contents always fills the nipple, and let your baby burp after drinking and also during drinking by tapping the baby gently on its back (do not rub) or rock it on your knees.

Weaning - Transition to Solid Food

Weaning is the one process in which babies learn to distinguish between sucking and biting to gradually start eating solid food. Every baby is an individual and learns something new at its pace. Usually a baby at 5 months is ready to eat baby food, if there is nothing preventing them from doing so. With EB babies it can take much longer to get used to a different flavor and a firmer texture due to a sore oral mucosa or tongue.

During the weaning process: Allow your child time!

It is important that you make the transition from liquid to solid foods slowly; your child will do it, only when it is ready. There is nothing to gain but much to lose if you push the child towards a faster transition. Under no circumstances should you force your child in any way to eat solid foods, by denying it baby milk. This would only lead to increased stress and frustration during feeding times. Check with your doctor or dietician if there are problems.

Babies and children learn primarily by observing their surroundings and therefore are strongly influenced by the behavior of the other members of the family, to eat what the others like and not eat what is disliked. If possible, feed your baby during family meals and allow it to share this important social event. Food should be fun! Your own diet is just as important, especially if you are breastfeeding. A newborn and especially one with EB is very tiring and therefore you should pay attention to yourself and your health by eating well and indulge in resting and relaxation periods. Only then will you have enough power to take good care of your child!

The solid foods may be introduced to babies with EB as well as in all other babies. The solid food is first intended to supplement a milk meal but not replace it. If your baby gets a special energy baby formula, it may have little appetite for solid food. In this case, you do not have to worry as long as you offer a suitable formula so that your child's needs are met. As your child gets older, it will eat more and more solid food and drink less formula. However, milk is an important source of nutrients.

What, When and How to offer and provide Nutrition for your Baby?

In many diet brochures for babies or also suggested by pediatricians you will find a supplementary feeding plan for when certain solid foods should be introduced. This scheme should be more of a directive than a schedule. Let your child take the time it needs! Solid foods can be cooked and prepared by you or purchased in glass jars.

- If you prepare your own baby food, be sure to cook salt-free.
- Puree the food, but do not pass it through a sieve otherwise valuable dietary fiber is lost.
- Use a baby spoon made of plastic or silicone. There are also various extra soft examples available (e.g. Flexy[®] by Dr. Böhm).
- Choose a time of day when your baby has sufficient hunger and you have the necessary patience.

- If your baby is really hungry it may be that it is too impatient to try something new. In this case give him some milk first so that the worst hunger is satisfied.
- Do not be disappointed if your baby refuses the food initially. It has to get used to the unfamiliar taste and texture.
- Increase the solid foods gradually in accordance with the supplementary feeding plan. Please note that even when learning to eat, every baby has a different pace!
- From about the 9th month the baby learns to chew. The children find great pleasure in grabbing the food and holding it in their hands. Allow the baby to touch the food even if there are bandages on its hands and the food ends up not only in its mouth - this phase is important for its development.
- Do not give the baby any rough or raw foods. Foods that are well suited are for example: bananas, soft peaches, apricots, pears, cooked carrots, potatoes and beets.
- Never leave your baby alone while eating - it could choke.

When can Cow's Milk be introduced?

Ideally, all babies should receive breast milk or infant formula in their first year of life. Especially children with EB, are recommended to drink a baby formula (if necessary high-calorie and nutrient-rich) due to their increased nutritional needs, throughout the first year of life and possibly beyond.

Cow's milk and yogurt can be given from the 12 month of life.

4.2. After the First Year of Life:

At around the first birthday the child can already participate in some family meals, provided they have little salt, are not too spicy and have a suitable consistency. If there are blisters in their mouth, citrus fruits (e.g. oranges or tangerines), tomatoes or acidic fruit juices are unpleasant; in which case cold food and drinks are most enjoyable. Foods which may scratch or irritate the mucous membranes of the mouth and throat, such as crackers, toast and bread with hard crust should be avoided.

Small round foods like grapes or cherry tomatoes should be cut into pieces for small children before consumption. It could otherwise be swallowed whole and if it then passes into the trachea it could lead to suffocation. For this reason one should not give candy or peanuts to children under 5 years of age! Fish is a very valuable food however make sure that all the bones are removed.

It is important that the child likes to eat, and if an EB child still prefers pureed or crushed food, this can still be permitted and need not be seen as a lack of progress. Consideration should be taken that the diet is balanced and contains plenty of protein. If your child has a poor appetite, offer him 3 small meals day and additional nutritious snacks. Milk remains an important source of nutrients. Some infants are difficult to separate from the bottle. Nevertheless, they should switch to drinking from a cup as soon as possible as bottle drinking can lead to tooth damage, especially if the bottle is used at night or when the child is drinking frequently from the bottle.

5. Special Issues

5.1. Your Baby will or can not eat

While it is easy to blame everything on the complications of EB, normal occurrences such as teething and febrile infections can affect the intake of food and drink. These events are part of normal development.

Children have a keen sense of their parents moods, so if your child has a day when they eat very little (for whatever reason), try not to show too much concern, as you can make your child insecure. Under no circumstances should you force your child to eat.

5.2. Constipation

Constipation in babies and toddlers is often the result of a low fluid intake, a reduced appetite or an increase in requirements (e.g. in hot weather). Iron supplementation may worsen constipation. However, constipation can develop for no apparent reason. If a baby with EB needs to make an extra effort to empty its bowel, it can cause pain and blisters in the sensitive area of the anus, even with a rather loose stool.

The fear of pain during defecation can lead to a suppression of the urge of a bowel movement and sets a vicious circle: The retained stool gets drier and harder, causing more and more pain with bowel movements. In addition, the appetite is reduced and the general well-being is decreased. Therefore, prevention is most important: make sure that your child drinks a lot, for example, in babies who already eat solid food, 150 ml of fluid per kg body weight in 24 hours. If your child refuses water (boiled and cooled) offer it well diluted fresh fruit juice (e.g. 1 tsp

juice in 100 ml water) or bottled baby juice, diluted at least 1:1. Once you are feeding your baby solids, give it vegetables and fruit porridge daily. Note that bananas, blueberries and rice can sometimes cause constipation. Jars of baby food with apple or pear in contrast, act as a stool softener.

Whole grain products should not be given to a baby! From the 8th month of life Opti Fibre[®] can be slowly added to the diet. This contains soluble, tasteless fiber and is available in the pharmacy. If, despite these measures, there is still constipation, a mild laxative may be helpful. Better yet is to give this regularly and as a preventative measure rather than when a stubborn constipation already exists. Act before the vicious circle of "pain during bowel movements - fear of pain - suppressing the urge for a bowel movement" is already set in motion.

5.3. Sugar and Dental Care

The condition of the teeth really affects the food intake. The better the dental health, the easier the biting and chewing, and the faster and with less caution the person concerned can eat. The development of caries is promoted by several factors among those affected with DEB. For example, the careful brushing is complicated by the vulnerable mucosa and with the resulting tightness from scarring in the mouth. In addition, a limited mobility of the tongue makes the removal of food residues more difficult. On the other hand a high-calorie and sugar-rich diet is often required to cover the increased calorie needs.

Caries or tooth decay occurs when the bacteria that colonize the teeth react with the sugar from the food and then forms acid. This acid attacks the enamel first, then the underlying dentin and then decomposes the tooth. The resulting decay can be painful and the affected teeth must either be filled or pulled. Of great importance for dental health are sufficient breaks between meals and from drinking sugary drinks. The saliva can then neutralize acids and the assaulted tooth substance can build up again.

It is not so much the amount of consumed sugar, but mainly the frequency or duration of sugar consumption that affects the teeth. A sugary soft drink, drunk throughout the day or frequent bites of chocolate are more damaging to the teeth than eating sweets with meals.

A compromise between the sugar as an energy source on the one hand and the dental health on the other hand is therefore necessary. A couple of important points:

- Babies should not receive any sugary drinks in their bottle or training cup. Water (boiled first) is the ideal thirst quencher!
- Pacifiers should be used with caution because they can cause blisters. If they are still used, do not immerse them in honey or syrup.
- Suitable baby beverages are water, baby formula, milk (with the appropriate age) and well diluted fruit juice.
- Soft drinks should be avoided - they contain lots of sugar and acid that can damage the teeth.
- Babies under 1 year old should not be given sweets - they can get used to the taste and reject other nutritious foods!
- If you give the child sweets, it is best that they are eaten with the main meal and not "in between".
- Make sure that your teeth are caries-free. Tooth decay is transferable and is passed from parent to child.
- Go to a dentist as soon as your child's first teeth appear, to get tips for optimal dental care, mouth rinses, fluoride medication etc. . . .
- If you are treated at the dentist take your child with you so that it can get used to the situation by watching.

5.4. Anemia and Iron Deficiency

A chronic anemia leads to fatigue and listlessness. An early sign of this is when the child is in school and has a short attention span or has no drive in the afternoon. Anemia affects the wound healing adversely and also has other subtle effects on health.

Unfortunately iron supplements often leads to constipation (rarely it can lead to diarrhea), especially when taken on an empty stomach. It is very important that undesirable side effects be treated, so that the iron can be taken. Iron can be administered as a capsule or liquid. The absorption in the body is greatly improved when vitamin C is taken at the same time (as a powder or in the form of fruits or vegetables). However, you should avoid taking zinc at the same time or the absorption of both trace elements is reduced. It is best to take them at least 2 hours apart.

6. Special Note

A part of the given information is based on the translation and adaptation of the brochure "Nutrition for EB Children over 1 Year of Age" by Lesley Haynes child nutritionist for EB, London, UK.