

## NUTRITION FOR DYSTROPHIC EB

### 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, 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 is 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

- **Mucosal blisters and sores in the mouth and throat may occur and cause food intake to be difficult.**
- **RDEB may cause esophageal strictures, which at times makes oral feeding impossible.**
- **In RDEB one often suffers from chronic constipation.**
- **Individual nutrition counseling is recommended.**
- **All treatment measures should be accompanied by an evaluation.**

## 2. The Importance of Nutrition in EB

A good diet is important for all of us, whether we have EB or not, and especially during a growth period. Babies, who do not suffer from EB, invest the majority of their diet in growth. Because the skin of EB children is very vulnerable, they need food not only to ensure normal growth, but also:

- to replace the nutrients that are lost through open wounds,
- to provide additional nutrients which are necessary for rapid wound healing,
- to assist the body to fight infections in the area of damaged skin,
- to support functioning bowel movements and prevent constipation.

All babies including those with EB have "bad" days when they are teething or feel uncomfortable for other reasons and therefore consume less food. EB babies may also develop blisters in the mouth and esophagus, and because of the difficulty in swallowing will eat much less. Parents are understandably very worried on these days, but when babies feel this, they often reject the food even more. Therefore, it is important to make the most of the "good" days and try to provide a nutrient-rich diet.

## 3. General Information

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. He 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. Some babies with EB may not be very physically active, yet they need a high caloric intake in order to make effective use of the proteins. If they do not receive enough energy in the form of fats and/or carbohydrates, the valuable protein is used instead for a relatively inefficient or wasted means of energy production. Therefore, EB sufferers with little appetite or difficulty with feeding should take advantage of the high energy content of fats and fatty foods and eat them regularly as part of their meals (see also the section "your baby can not or will not eat").

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,

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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 fruits 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

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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.

## **Minerals**

Mineral substances, as well as vitamins are essential for growth and health. A balanced diet usually contains sufficient quantities. Particularly important minerals in EB are iron, zinc, selenium and calcium.

*Iron:* The main dietary sources of iron are meat (especially liver, kidneys and red meat), bread and cereals (mainly millet and bran flakes) and legumes. Baby formula contains iron and many other baby food products, such as porridge, zwieback and biscuits, baby juices and jars of baby food are also fortified with iron. Only a small portion of iron from the diet is absorbed by the body, the rest is unused and excreted in the feces.

The proportion of iron in animal sources (meat) is much higher than that from vegetable sources. Iron absorption can be doubled when during the same meal vitamin C (e.g. orange juice, green peppers, etc.) is also eaten. When preparing home-cooked meals for babies you should use high-quality oil (such as sunflower seed) and add some orange juice before serving. Iron is needed for blood cell formation and helps prevent anemia. The iron loss can be significant when the skin is fragile and has a number of wounds.

*Zinc:* Is contained in a variety of foods, especially in fish, seafood, meat (especially liver, kidney, heart), but also in legumes, soy, mushrooms, dates, dairy products and bread. Zinc is essential for rapid wound healing and the immune system. It is also part of several complex metabolic processes. Since EB usually consists of a continuous wound healing process, the demand for zinc is high, so it is often supplemented. Zinc deficiency leads not only to growth and immune system disorders but also a lack of appetite.

*Selenium:* Is found in dairy products, nuts, grain products and legumes. It is (in addition to Vitamin E) an important factor in cell protection and it helps the immune system in the production of antibodies.

*Calcium:* Can be found in milk and dairy products like cheese and yogurt. Breast milk and infant formulas contain a lot of calcium, so that most children with EB get enough calcium through milk and milk-based products. Calcium is responsible together with vitamin D, for the formation of healthy bones and teeth. It is also essential for normal muscle and nerve function and blood clotting.

### **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 requirement than for 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.

## **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 its 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.
- If your baby has painful sores in its mouth that can affect breast feeding, you can apply a numbing solution such as Herviros<sup>®</sup> with a cotton swab directly onto the painful area before breast-feeding. Caution: If this solution is distributed throughout the mouth and throat, the "docking" will be disturbed.
- Do not be afraid to go to a breastfeeding counselor for open questions or uncertainties.
- 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<sup>®</sup> 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. Sometimes it may be necessary to increase the nutritional content of the breast milk. There are several options available; your nutritionist can advise you which is best for your child.

### **Bottle Feeding and Baby Formula**

You may choose not to breast feed your child, but to provide nutrition with a baby formula. Babies with EB often have increased nutritional needs, so that breast milk alone may not be sufficient to provide this and to ensure normal growth. For whatever reason, you can discuss an optimal nutrition plan for your child with your dietician. It may be that you give bottle feedings or expressed breast milk to add nutrients in addition to breast milk, or to give the child fortified infant formula.

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<sup>®</sup> with a cotton swab before breast feeding. Caution: If this solution is distributed throughout the mouth and throat, the "docking" will be disturbed.

If many blisters and sores exist in the mouth, sucking can also be facilitated by enlarging the suction opening of the nipple (with a sterile needle or a small crosswise slit with a sharp pair of scissors).

Make sure that your child does not cough or choke because of the faster milk flow. A bottle from Haberman<sup>®</sup> with a special nipple is available in pharmacies (for more information about Haberman<sup>®</sup> nipple see: breastfeeding and breast milk).

### **Weight Gain and Fortified Foods**

To judge if a baby is developing well this 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<sup>®</sup>).

### **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 food 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.

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- 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 food 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.

A light or skim milk is generally not beneficial for children with EB due to their low fat and low calorie content. Yogurt can be given from the 12 month of life. Again, you should avoid low calorie products.

### **At the End of the first Year**

With 11 to 12 months of age the baby 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 its mouth, citrus fruits (e.g. oranges or tangerines), tomatoes

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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. When fish is on the menu make sure that all the bones are removed.

When cooking yourself remove the hard parts from fruit like the skin and core of apples. Provide a good balanced diet with plenty of protein and lots of calories. If your child has a poor appetite offer it nutritious snacks in addition to the 3 meals. Milk remains a valuable source of nutrients. Many babies prefer to drink their milk from a bottle rather than from a glass or training cup.

#### 4.2. After the First Year of Life

At about their first birthday most EB-children may attend family meals, provided the food is not too spicy and the consistency is appropriate. Citrus fruits (e.g. oranges, tangerines), tomato or freshly squeezed juices can irritate the oral mucosa. Foods that can scratch or damage the oral mucosa - such as hard bread crusts, chips or hard raw vegetables (e.g. carrots) 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 a 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. Problems in severe Generalized Recessive Dystrophic EB

The problem is mainly that the wounds heal with scarring and that it also leads to contractures. In this form, hands and feet are fused and are very severely impaired in their function. It is common to have painful mucosal blisters and sores in the mouth and throat which can lead to less food being eaten. If this is the case, you can use a cotton swab to apply a numbing solution such as Herviros<sup>®</sup> directly to the painful area to alleviate their discomfort. A further complication is that the mouth can be difficult to open and the tongue is often rigid. Due to the scar tissue, esophageal strictures also can form and make oral feeding impossible at times. Furthermore, these children also suffer frequently from chronic constipation.

Children with severe dystrophic EB often cannot eat enough to ensure optimal growth and wound healing. This reduces their quality of life significantly and is one of the many concerns and fears of parents. Problems in the mouth and esophagus can lead to a slow, tiring and sometimes painful eating process; such experiences often cause the child to refuse to eat or to try new food. An unpleasant reminder of painful swallowing or choking is enough that long after this experience, the trust is disturbed in swallowing and the fear of eating remains. If the problems with eating and swallowing persist, it may happen that a meal is finished without substantial time interval to the next.

This is unfavorable for the following reasons:

- It is tiring and tedious for the children and their caregivers and the food is not what it should be; a pleasant experience.
- It is very time consuming and leaves so little time for other activities.
- It prevents the development of an appetite.
- It promotes the development of tooth decay.
- It favors the development of constipation.

The pureeing of the food allows the child to eat quickly and with less caution (see also 9.2. Pureed Food). The inclusion of limits for meal times reduces the frustration for all parties involved and also the negative associations with food.

It is important on the one hand to support the child while eating and to let it finish its meals and on the other to give the child enough time to play. Praise not only the large, but also the small progress the child makes. You should never force feed the child; this would cause anxiety and in the long run is counter-productive.

Independent eating of EB sufferers in every case should be supported. There are special spoon holders that can be attached to the hand or forearm, when the fingers are fused and no cutlery can be held.

## 6. 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 juices.

- 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".
- Go regularly to the dentist with your child - preferably every 3 months. If you are treated at the dentist take your child with you so that it can get used to the situation by watching.
- 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. . . . ideal, would be a dentist who has experience with EB patients.

## 7. 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). An EB-baby with pronounced blister formation may have a significantly increased fluid need. 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 their 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. To prevent constipation, it is really important to make sure that your child drinks a lot. 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<sup>®</sup> can be slowly added to the diet. This contains soluble, tasteless fiber and is

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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. Another very effective medication for bowel movements in EB is Movicol<sup>®</sup> (active ingredient: macrogol). This can be prescribed by your doctor.

## 8. Anemia and Iron Deficiency

Children with EB can be anemic for two reasons. One reason is that the skin is very vulnerable and they lose blood through wounds. This can take place internally (mouth, esophagus, intestine and anus) or by external wounds on the skin. The other reason for some children is, that meat (the best source of iron) is difficult or impossible to chew and swallow. 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. In rare cases, iron is also given as a venous infusion.

## 9. Special Issues

### 9.1. Your Baby can not or will 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 illnesses are inevitable during the development of a child and also important for the development of the immune system; nevertheless they are very stressful for parents. Children have a keen sense of their parents moods, so if your child has a

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day when they eat very little (for whatever reason), try not to show too much concern, as you can make your child insecure.

A child, who is used to a nutrition rich diet when it is well, will accept this food even more when it only can eat a little.

Therefore, it is better to use some "tricks" to increase the daily calorie and nutritional intake from food. Prepare as many meals as you can with eggs, meat, fish, milk, and cheese. Take advantage of the calorie content of fat-rich foods such as butter, oil, mayonnaise, whipped cream, sour cream and mascarpone.

**TIPS:**

- Add 2-3 tablespoons of whipped cream or sour cream to mashed potatoes, sauces, soups, yogurt, puddings, creams and applesauce.
- Use plenty of butter, cream cheese, almond or nut butter on bread and cookies.
- Add some grated cheese, mayonnaise, butter, cream cheese or sour cream on top of spaghetti, potatoes, vegetables, scrambled eggs...
- Add 1-2 teaspoons of jam, ripe banana pieces, etc. in cream of wheat, rice pudding, and yogurt.
- Stay away from low fat products, use high-fat products, e.g. high fat yogurts, cream, cheese, etc.
- Prepare savory dips and spreads, such as mashed avocado with mayonnaise, tuna spread mixed with mayonnaise or sour cream.
- Prepare sweet dips, such as jam with sour cream or mascarpone.
- ... and other "tricks" that allow you to meet your child's preferences.

## 9.2. Pureed Food

Some children can only eat very soft, pureed or liquid food. This will soon become boring if it only consists of soup and ice cream. But if suitable meals the family eats are included in the menu, the food will be more varied and just as important, the child feels less "different". Children often want to see the food before it is pureed to know that they get the same as the other family members. Pureed food looks much more attractive when everything is pureed individually and colored foods such as carrots, potatoes or peas contrast with the meat.

This way the child can get to experience the different tastes of food. With a sore oral mucosa, it may be that cool, cold or slightly frozen food is enjoyed more. Some

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dishes can be cooked in advance, in a large amount and frozen in portions to be used when the family meal time is not suitable for pureeing.

Soup, milk, whipped cream or sauces are best suited as a liquid additive for pureeing. If water is used, the nutrient content of puree is diluted and it also tastes bland. If possible, do not pass the food through a sieve; otherwise the fiber content is reduced.

### 9.3. Supplementary Nutrition

If chewing and swallowing are very difficult for your child the tips above can only partially help to meet the increased nutritional requirements. Fortunately, there are a wide range of nutritional supplements.

They are available in different preparations:

- **Complete food supplements:** Are usually produced as a beverage or as a pudding. You can add or replace a meal with it. Since they are packaged "child-friendly" with colorful packaging and a straw, they can be eaten in the school as a snack.
- **Partial supplements:** Are given in addition to the meals. They contain a high concentration of a particular nutrient, such as high-calorie, high protein, high fat or they contain fiber.

Contact a dietician to receive the latest information and to get the food supplement that best suits your individual needs. It is useful to change the products repeatedly, as children usually get tired of the taste.

### 9.4. Dilatation of the Esophagus

Children with recessive dystrophic EB often develop tightness of the esophagus, which leads to difficulty swallowing (dysphagia). Sometimes the intake of liquids is even obstructed. An esophageal dilatation is a surgical procedure done under anesthesia, in which the esophagus is gently stretched so the constriction (stricture) is expanded. In some children a considerable improvement in swallowing can be achieved by this manner. Most often the procedure must be repeated from time to

time. When despite an esophagus dilatation the child still has difficulties to consume enough food, there is the possibility that a nasogastric feeding tube is needed.

### 9.5. Nasal Tubes and PEG (Stomach) Tubes

Sore oral mucosa and painful swallowing have a very negative impact on nutrition and significantly reduce food intake. By administering a part of the necessary food directly into the stomach (thus bypassing the "problem areas" mouth, throat and esophagus), the child can continue to enjoy eating those foods that it tolerates. The nutrition that is "missing" is given through a tube. Nasal tubes (soft, thin flexible silicone tubes are inserted through the nose passed through the esophagus and down into the stomach) are rarely recommended and then only for a short time. The children understandably do not like these as they are clearly visible, as well as the tube is hard to fix to the vulnerable skin are. The so called PEG tube (percutaneous endoscopic gastrostomy) feeding tube is passed directly through the abdominal wall into the stomach during a simple surgery.

One of the advantages is the "button" which is covered by the clothing. The button is the connector on the outer abdominal wall, which is composed of a button with a cover for which the administration of the feeding tube is connected. In addition the PEG tube can be removed at any time when it is no longer necessary. It can be more or less often used: The child eats and drinks as much as it wants or can, and the rest of their nutrition can then be administered via the feeding tube. The tube can also be used to administer bad tasting medications.

#### Nutrition via the PEG Tube:

It is not advisable to try and administer pureed food over the nasal or the PEG tube, because it can get blocked and also it can grow harmful bacteria. Instead, one of the special tube feed products that your doctor or health care professional can prescribe should be used. The dietician will recommend the most suitable product for you, depending on the age and special needs of the child.

There are many different ways of when and how the tube feeding can be given. Each child has individual needs and the daily routine of each family is different - all of which will be taken into account when the dietician creates an individualized plan for your child.

Here are three of the most common options:

- Give a small amount after a meal, e.g. 50 ml, when the child has not eaten enough or has eaten nothing.
- Let the tube feeding drip at night when the child is asleep for example, 300 ml between 8 and 11 pm.
- Let a larger amount drip at night, but this has the disadvantage that the child is not hungry in the morning.

Although we normally do not eat at night, many EB children prefer the administration of nutrition over the PEG tube at this time, so that during the day they have time to eat and drink normally and time is left for other activities.

This nightly diet leads to nighttime needs for urination and defecation, so that the sleep of the child and the parents is often disturbed. This is more likely to occur when more than 500 ml of the tube feeding are given.

The nutrition via the PEG tube can therefore be adapted for each family and for the individual habits and conditions, so that the feedings can be well integrated into the daily routine.

## **10. 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.