

NUTRITION IN JEB 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!



Important points in a nutshell

- **Mucosal blisters and sores in the mouth and throat may occur and cause food intake to be difficult.**
- **Even if your baby has blisters in the mouth it does not necessarily mean they can not be breastfeed.**
- **Individual nutrition counseling can be helpful.**

2. Nutrition for a Baby with JEB generalized severe

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 their 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[®] 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[®] 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 in addition to breast milk, bottle feedings or expressed breast milk to add nutrients, 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[®] 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[®] with a special nipple is available in pharmacies (for more information about Haberman[®] nipple see: breastfeeding and breast milk).

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.

Supplementary feeding and weaning

Weaning is the process in which babies learn to distinguish between sucking and biting to learn how to eat solid food. Every baby is an individual and has its own time to learn something new. A baby with EB can have a sore oral mucosa or tongue causing it to take much longer than a healthy baby to get used to a different flavor and a firmer texture. Your baby will make clear to you, if it is interested in solid food. If your baby with JEB generalized severe type however, should make no attempt to want to eat solid food or rejects this, this is not unusual and you can continue using the usual formula milk. Just be sure that the baby gets all the essential nutrients and sufficient calories it needs through the formula. Let your dietitian advice you accordingly.

3. Constipation: Prevention and Therapy

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