

NUTRITION FOR BABIES WITH EB Lesley Haynes RD, Peincipal Paediatric Dietitian, Great Ormond Street Hospital, London, UK

All babies and children need to be well nourished to thrive and to enjoy the best possible quality of life. This is especially important for those with the more severe EB types, (Recessive Dystrophic EB, Dowling-Meara EB Simplex and non-Herlitz Junctional EB) because some of their nutrition is diverted away from growth and into wound healing and fighting skin infections. Although it is not possible to cure EB with “special” diets (eliminating particular foods or containing megadoses of certain nutrients), by promoting an optimal nutritional intake growth, immunity and healing will have the best chance of succeeding. Sadly, the nature of Herlitz Junctional EB (HJEB) is such that nutritional modifications cannot exert any long term benefit. However some dietary manipulations may positively influence the HJEB baby’s quality of life, and individual advice should be sought about this.

This booklet describes the nutrients that make up a normal balanced diet for all of us regardless of our age and whether we have EB or not. This has been done in order to offer comprehensive information, but please note that some foods mentioned are not appropriate for babies and young children. This is followed by practical information specific to feeding EB babies up to the age of about 18 months.

A booklet like this can give only general advice and cannot provide answers to every situation. To aim for the best nutrition for your child, the advice of a paediatric (children’s) dietitian should be sought. An individualised plan can then be drawn up and reviewed regularly to ensure that it remains realistic and feasible. EB affects girls and boys in equal proportions, but for ease of reading, the baby is referred to as “he” throughout.

Why is nutrition important in EB?

Babies who do not have EB channel most of their nutrition into growth. However, because EB babies’ skin is fragile and easily damaged, they may need individually-tailored diets not only to promote growth, but also:-

- to compensate for nutrients lost through open wounds or lesions
- to supplement the nutrients necessary for optimal healing
- to help maintain an effective immune system to counteract infection
- to promote normal gut function and avoid constipation
- last, but by no means least, to feel as well as possible and enjoy a good quality of life.

All babies have “off days”, e.g. due to teething or minor illness, when food intake is reduced and EB babies are no different. However, this happens more often for EB babies if they have also to contend with blisters in the mouth and throat which make feeding uncomfortable and may reduce food intake significantly. Parents naturally become very anxious at these times, but when babies sense this, they can become more reluctant to feed. However, by capitalising on the good days when your child is happy to eat, you will feel better able to cope with days when his intake is poor.

What is a nutritious diet?

A nutritious diet contains a balance of:-Proteins Carbohydrates Fats Vitamins Minerals Fibre Water This is not as complicated as it sounds because most foods are a mix of several nutrients, and a nutritious diet can be achieved with many different combinations of foods. The more varied the diet, the more likely it is to contain a good balance of nutrients. Very simply, most nutrients are used to satisfy the body's need for growth and repair (normal "wear and tear"), and to supply energy (calories). Complex chemical processes cause food to be broken down into small units and then reabsorbed in forms that enable us to live (this is called metabolism). Nutritional requirements are relatively high during infancy, childhood and adolescence when growth rate is fastest. In EB, blistering and loss of body fluids require constant repair work and the metabolic rate (the speed at which metabolism takes place) can be higher than normal, especially when infection is present.

Proteins

Proteins are important to build strong, healthy body tissues and to promote wound healing. Milk (and other dairy products such as cheese and yoghurt), meat, fish and eggs comprise the main dietary sources of animal protein. Vegetable protein is found in foods such as pulses (peas, beans and lentils or dhal), nuts and cereals. A vegetarian diet needs careful planning to ensure its adequacy for any child. Nuts (except for smooth nut butters) should not be given to children under 5 years in case they inhale them or choke on them.

Fats

Fats are an important source of calories and although some EB children may not be very physically active, they need enough calories to metabolise protein efficiently. If they do not consume enough calories from fats and carbohydrates, protein is used as a wasteful calorie source instead of being used to build body tissues. The high calorie content of fats and fatty foods is useful for children with small appetites (see "Can't eat, won't eat"). Butter, margarine, cream, oil, lard, suet and dripping are obvious sources of fat. Less obvious sources are full cream milk, full fat yoghurt, most types of full fat cheese (including fromage frais), ice cream, meat (especially when there is visible fat), eggs, oily fish (e.g. sardines, pilchards and salmon), avocados, nut butters and chocolate.

Carbohydrates

Carbohydrates are also an important source of calories and, in some cases, of fibre, vitamins and minerals. The carbohydrates that are useful mainly as a source of calories include biscuits, sugar, sweets, glucose, honey, jam and syrup. Carbohydrates that contain a wider range of nutrients include cereals, breakfast cereals, flours, pasta, bread, potatoes, fruits and pulses. Puddings and cakes are valuable mainly for their energy content, but can also be good protein sources if they are made with eggs and milk products. All carbohydrates are important in the EB diet. The sweet ones (biscuits, sugar etc) should be included with, but not instead of, the less sweet ones (cereals, potato etc). (See "Sugar and tooth care").

Fibre

Fibre (roughage) is the part of food which is unabsorbed as it passes through the gut. Far from being of no value, fibre is very important for keeping the gut healthy, the bowels working normally and encouraging the beneficial bacteria there to flourish (which in turn helps the immune system to work effectively). Fibre is found in wholegrain breakfast cereals, eg. Weetabix®, porridge, bran flakes & muesli, wholemeal bread, wholemeal flour, wholemeal pasta, brown rice, pulses, dried fruit and fresh fruits and vegetables. Children with mouth and throat blisters can find high fibre foods difficult to chew and swallow. Citrus fruits (oranges, tangerines etc) and tomatoes may irritate the mouth if it is blistered or sore. A high fibre diet is bulky and filling and consequently can be low in calories, as less total food is eaten. (See “Constipation”).

Vitamins

Vitamins are essential for growth and health. If a sufficiently varied diet is eaten every day, vitamin intake is usually satisfactory, however if food intake is limited supplements may be recommended (see “Vitamin and mineral supplements”). There are several vitamins and each has a specific function in the body. For example:-

Vitamin A is found mainly in liver, milk, oily fish such as mackerel, fortified margarine and yoghurt. Dark green, red and yellow vegetables contain a substance called carotene which can be converted by the body to vitamin A. Vitamin A is needed to maintain the health of skin, eyes and mucous membranes and to help strengthen the immune system.

The B group of vitamins is found in dairy foods, meat, eggs, bread, cereal products and potatoes. Different B vitamins have specific functions eg. helping to metabolise energy from carbohydrates, maintaining healthy blood and skin and promoting protein metabolism.

Vitamin C is found mainly in fruit, especially citrus fruit and some vegetables, eg. green vegetables and potatoes. Vitamin C is important in activities such as wound healing and iron absorption.

Vitamin D is found in butter, margarine, oily fish, evaporated and dried milk, eggs and liver, with fish liver oils being the best source. Sunlight on the skin produces vitamin D in the body. With calcium, Vitamin D helps to build strong bones and teeth.

Vitamin E has several important functions such as protection of cell membranes and promotion of immunity. It is found in vegetable oils, nuts, green leafy vegetables and wheatgerm.

Minerals

Minerals, like vitamins, are essential for health and growth and a well balanced diet usually provides adequate amounts for normal requirements. Iron, zinc, selenium and calcium are particularly important in EB.

Iron The main sources of iron in the diet are meat (especially liver, kidney and corned beef), bread and fortified breakfast cereals. Iron is needed for healthy blood and prevention of anaemia. Iron losses can be significant when skin is fragile and frequently damaged.

Zinc is found in a variety of foods, particularly protein foods e.g. meat, shellfish and dairy products as well as bread and cereal products such as

wheatgerm. It plays essential roles in many complex metabolic processes including wound healing.

Selenium has a vital role (together with Vitamin E) in protecting body cells from damage; it also helps the immune system to produce antibodies. It is found in Brazil nuts, dairy foods and some seeds and grains.

Calcium, with vitamin D, builds healthy bones and teeth. It is also needed for normal muscle and nerve functioning and blood clotting. Calcium is found in milk and milk products such as cheese and yoghurt. Breast and formula milks are good sources of calcium and EB babies rarely need supplements.

Water, although often not considered as a nutrient, is essential to keep us well hydrated. An adequate fluid intake is also important to avoid constipation. (See "Constipation").

Vitamin and mineral supplements

If food intake is restricted by factors such as sore mouth or painful swallowing, or if requirements are increased, supplements can be prescribed. However, because excessive intakes can be harmful, it is always advisable to ask for dietetic advice before introducing any form of supplementation.

Breast feeding

Human milk is perfectly designed to promote babies' growth and development. It contains precious antibodies which contribute to the baby's defence against infection as well as prebiotics (substances which encourage the growth of beneficial gut bacteria that are involved in many vital processes); it may also reduce the chance of development of allergy. So, it is very advantageous for a baby with EB to have breast milk, even if only for a short period.

Here are some tips that mums of EB babies have found helpful :-

- Even if your baby has mouth blisters, this does not necessarily rule out breast feeding. Blisters often burst during suckling, but if not, follow the EB Nurse Specialist's advice
- To minimise friction when he nuzzles against your breast, apply white soft paraffin or Vaseline® to your nipples and to his cheeks, lips and gums
- Put him to the breast often and let him suckle as long as he wishes
- Allow plenty of time so that neither of you feels rushed
- If your breasts are very full, express a little milk first so that he does not choke when the milk comes down
- Make sure that he is properly latched onto the nipple and not askew.

If your baby's mouth is too sore to suckle, or if he tires easily, you could express milk and feed it from a dropper or spoon, or a specially-manufactured bottle and teat such as the Haberman Feeder® (Athrodax Healthcare International). This teat is specially shaped and is less likely to damage the gums than a regular teat. Its internal valve and long shaft allows control over the flow of milk, helping a baby with a weak suck to receive a good flow. However, it is important not to squeeze the shaft so strongly that milk hits the back of his throat, stimulating the gag reflex and possibly causing him to breathe feed into his lungs. Alternatively, the hole in a conventional teat can be enlarged using a needle or by making a small cross cut with a pair of sharp scissors. Sterilise the teat before use.

Take care that the faster flow of milk does not cause coughing or choking. Sometimes breast milk needs to be supplemented to increase its nutrient content (this is called fortifying) and there are various ways in which this can be done. You can discuss with the dietitian the best option for you and your baby.

Formula feeding

You may prefer to give infant formula rather than breast milk to your baby. On the other hand, perhaps your baby's requirements cannot be fully supplied by breast milk and he needs to have formula milk in addition to, or in place of, breast milk. Whatever the reason, you can discuss with the dietitian the best feeding plan for you and your baby. This may involve adding specific nutrients to your expressed breast milk (EBM), or giving some infant formula in addition to your breast milk, or giving only formula feeds. (See "Weight gain and fortified feeds").

Weight gain and fortified feeds

The best gauges of any baby's progress and development are increased weight, length and head circumference and your baby's weight in particular should be checked regularly to ensure that he is thriving. This may mean weighing on alternate days in hospital and weekly to start with once he goes home. Ideally, this should be done on the same scales each time and in similar dressings and/or clothes.

If your baby's weight gain is a little slow, the dietitian can suggest adding extra nutrients to your EBM and/or to give him a commercially-available feed specially formulated to contain a higher concentration of nutrients than regular babymilks. Examples of these include Infatrini® (Nutricia) and SMA High Energy® (SMA Nutrition); both are available on prescription from your general practitioner (GP). Alternatively, you might be advised to make up his feed in a concentration greater than the usual dilution of one scoop of baby milk powder to 30 ml (1 fluid oz) of water. This will give a feed richer in all nutrients but in the same volume of fluid as before. This is called fortifying the feed.

It is safe, provided that it is done under medical or dietetic supervision and reviewed regularly.

An example of such a feed would be:-

4 scoops of babymilk such as SMA Gold® (SMA Nutrition) or Premium® (Cow & Gate) plus 90ml (3 fluid ounces) of cooled, boiled water the usual dilution is 3 scoops plus 90ml water.

If your baby needs more calories, these can be provided in the form of a specially manufactured supplement of carbohydrate and/or fat mixed with the babymilk. The most commonly used carbohydrate supplements are called glucose polymers and brand names include: Caloreen® (Nestle), Polycal® (Nutricia), Maxijul® (SHS Nutricia) and Polycose® (Abbott).

An example of a feed incorporating a glucose polymer would be:-

4 scoops of babymilk plus 1 scoop glucose polymer (using babymilk scoop) plus 90ml cooled, boiled water. Calogen® (SHS Nutricia) is the brand name of a fat supplement in the form of an oil and water emulsion. A combination of glucose polymer and fat is available in two forms, Duocal® (a powder) and Liquid Duocal® (SHS Nutricia). All these calorie supplements can be obtained

on prescription from your GP. Always ask your dietitian for advice regarding their suitability for your baby and directions for their use.

Fruit juice and other drinks

Babies do not usually need any fluids other than breast milk/formula milk to nourish them and boiled water to quench any extra thirst. Fruit juices and herbal infusions are not an essential part of any baby's diet and sweet-tasting fluids may suppress the appetite for feeds. Babies often cry because of thirst as well as hunger, so it is a good idea to offer your baby cooled, boiled water in between feeds, especially in hot weather. If he is thirsty, water is the perfect drink. Too little fluid can cause and aggravate constipation. If you feel that your baby needs more fluid, but he refuses water, offer ready-to-feed baby juice diluted with an equal amount of water or 1 teaspoon of fresh fruit juice plus 100ml cooled boiled water. (See also section on "Constipation".)

Weaning

Weaning is the process during which babies learn to graduate from sucking to biting, chewing and swallowing progressively more solid foods. In the UK, the recommended age for introducing weaning is around 6 months. Some babies need, and can safely be given solids earlier than this, but not before 17 weeks. If you do decide to give solids before six months, foods containing gluten, eggs, cheese, dairy products, fish and shellfish should be avoided. Delaying until then will help to minimise the risk of developing adverse reactions to foods and allergies. Every baby is an individual, and arrives at the weaning stage in his own time. Signs that your baby is ready to start taking solids include:-

- Holding his head up, maintaining a steady, upright position when supported
- Being curious about what you are eating and making chewing motions
- Picking up food and putting it in his own mouth
- Having cut one or two teeth
- Being hungry even though you have tried increasing milk feeds for a few days.

Starting off

At first, solids should supplement milk feeds, not replace them. If your baby is having a more concentrated feed, it is very likely that he will not be interested in solids as early as other babies. So try not to be disappointed if friends' babies are progressing faster than your baby - he will get there. A sore mouth or tongue, or fear of choking on the unfamiliar consistency or flavour are all common reasons for EB babies to be reluctant to progress onto solids. Fear seems more common if there is a history of vomiting or reflux, or if these are current problems. Reflux is the return of acidic stomach contents from the stomach back up the gullet or oesophagus and it is a very unpleasant experience. It is quite common in all babies, but in EB babies it is especially important to treat it promptly because of the burning effect of stomach acid on the delicate lining of the oesophagus. Your specialist can prescribe anti-reflux medication and you may also be advised by the dietitian to thicken feeds. Thicker feeds are less likely to reflux back up the oesophagus. The important thing is not to rush your baby into taking solids; he will get there in his own time. There is nothing to be gained and much to be lost by trying to make him

go faster than he is willing to go. Never try to “starve” him into taking solids by withholding some of his milk feeds. This will lead to weight loss and only add to the frustrations and stresses surrounding feeding. Contact your dietitian and she will be able to suggest strategies appropriate for your individual situation. As your baby gets older, he will eat larger amounts of solids and drink less milk. However, milk will continue to provide a valuable source of nutrients and he should continue to drink about 500 ml per day after he has been weaned.

Take it slowly and allow your baby to set the pace

There is a suggested plan for introducing solids at the back of this booklet. It is meant as a guide rather than a strict timetable, so let your baby set his own pace.

- Choose a time when your baby is most hungry and you have time to relax
- Always give solids from a spoon; don't add them to bottles
- Use a spoon especially designed for baby feeding e.g. the flexible, plastic Baby Basics® spoon made by Heinz
- Offer solids before he is too hungry for the next feed. Give some milk feed first to satisfy him if he is crying
- Don't be disappointed if he refuses new foods at first. It takes time to get used to new flavours and textures
- Other weaning foods can be home cooked or commercially prepared
- Do not cook with or add salt to home cooked food
- If you do need to puree food in a food mill (mouli) or liquidiser, if possible, do not sieve it as this reduces the fibre content
- Between 6 and 9 months, babies gradually learn to chew and foods need to be thicker. Fruit and vegetables should be mashed and meat minced
- Babies rapidly lose confidence if they are frightened by having to control different consistencies at the same time and may even choke, so offer foods of a uniform consistency rather than foods with soft lumps in a liquid
- At 6-7 months, introduce a teacup or cup for some drinks
- At 6-7 months, encourage finger foods (see below), even if your baby's hands are bandaged. He needs to touch food and feed himself and be allowed to make a mess! Avoid sharp or rough foods that scratch the mouth and gums
- Don't give honey until your baby is a year old. It may contain bacteria that produce toxins that can cause serious illness (infant botulism) in younger babies.

Finger Foods

Babies and young children with EB are often very reluctant to try new foods. Holding food and feeding themselves gives them more control and is an excellent way to promote their confidence. It will inevitably be a messy business, but it is well worth it, even if more food is played with than eaten. Here are some ideas:-

Bite and dissolve foods

- trifle sponge, meringue
- soft wafer biscuits (eg. those with a cream filling)
- rice paper *
- ice cream wafers *
- Pom Bear® snacks, Wotsits® **, Skips® **, Quavers® **, prawn crackers **

Bite and chew foods - cut into strips or small pieces if necessary

- ripe peeled pears, peaches, nectarines, mango, banana
- cooked, peeled eating apple
- soft ready-to-eat stoneless prunes, e.g. d'Agen (check no fragments of stone remain), soft, ready-to-eat, stoneless apricots, dried mango, dried pineapplefruit
- "sticks" e.g. Hum Zingers Fruit Stix®, marshmallows
- jelly - made from a packet with half the usual water to get a stiffer consistency
- ripe peeled avocado
- cooked potato, soft chips – no sharp points
- cooked carrot, parsnip, swede
- soft bread, white or wholemeal, not Granary
- soft boneless fish, chicken
- tinned frankfurter sausages eg. Plumrose® **, "fingers" of ham **
- processed cheese eg. Dairylea® triangles **
- plain soft cooked pasta, pitta bread
- pancakes, omelette

* Use with care as they tend to stick to lips, but are useful as they encourage licking.

** Very salty; don't give under 9 months. Give no more than once daily under 1 year.

Towards the end of the first year

By 9-12 months, many family meals can be suitable for the EB baby, provided that they are of a suitable consistency and not too spicy or salty. Citrus fruits, e.g. oranges or tangerines, tomatoes or fresh fruit juices may cause discomfort if the mouth is blistered. Foods which are likely to cut or scratch and blister the mouth or throat should be avoided e.g. hard crusts and sharp chips, toast, crackers, raw carrot and celery. Fish is very nutritious, but always check that all bones have been removed. Take care with hard pieces of fruit e.g. apple, the skin on fresh sausages (tinned sausages e.g. Plumrose® have very soft skin), and small, round foods such as grapes and cherry tomatoes that can be swallowed whole and cause choking - it is safer to cut these in half.

Encourage a good, balanced diet with the emphasis on protein and energy. If the appetite is small, offer 3 small meals a day with nutritious snacks in between. Milk continues to be an important source of nutrients, so try to give not less than 500ml daily of the recommended milk formula. Some toddlers are reluctant to move from a bottle to a cup or teacher beaker. Although this may take time, it is worth persevering because prolonged bottle feeding can cause tooth decay, especially if bottles are drunk during the night when saliva production is low.

When can cows' milk be introduced?

Although it is recommended that all babies should receive either breast milk or an infant formula (or a combination of these) until one year of age, many are switched to cows' milk at about 6 months. However, EB babies who have higher nutritional requirements should continue to take the recommended formula or feed, fortified if necessary (see section on "Weight gain and fortified feeds"), for at least the first year and possibly longer. A "follow-on" milk, designed to bridge the gap between infant formula and cows' milk may be suitable after 6 months. Skimmed and semi-skimmed milks are generally unsuitable because they are low in fat and therefore low in energy. Yoghurt and fromage frais made from pasteurised cows' milk can be given from about 9 months. Choose the full fat, sugar-containing varieties of these products as the energy content is higher than the low calorie types.

Can't eat, won't eat

Normal events such as teething, minor illness and the "Terrible Two's" (which often starts before the age of 2) inevitably interfere with feeding. Although all these are part of normal development, they can be very stressful times. Children rapidly pick up parental worries about feeding, so if your child is having an "off day", try not to transmit your anxiety and never force-feed. When a child's appetite is small, it helps to know how to boost the nutrient content of his meals without increasing their bulk. The child who is used to having his food boosted when he is well is more likely to accept this when he is unwell. So, include these boosts on a regular basis, rather than waiting until an episode of poor eating. A well-nourished child is better able to handle short periods of poor intake and you will be less anxious, knowing that he has "reserves" on which to draw. Boost foods in any of these ways:-

- Add 2-3 teaspoons single/whipping cream or evaporated milk to mashed potato, sauce, yoghurts, custard, mousse, jelly. (Cream or evaporated milk can be frozen in ice cube trays for later use)
 - Add a butter, margarine, olive oil or mayonnaise to hot vegetables
 - Spread butter, margarine, cream cheese or smooth nut butter liberally on bread, soft toast and plain sweet and savoury biscuits
 - Add grated cheese/cream cheese/mayonnaise to mashed potato, baked beans, spaghetti and scrambled eggs
 - Serve white or cheese sauce with fish and vegetables
 - Avocado pear is naturally high in calories; offer it mashed or as a finger food
 - Add 1-2 teaspoons of jam, honey or lemon curd to custard and rice pudding
 - Add 1-2 teaspoons of sugar to breakfast cereals, yoghurts, stewed fruits
 - Make up packet jelly with some of the water replaced by evaporated milk
 - Top bread or biscuits with jam, honey, lemon cheese or chocolate spread.
- Mix any of these spreads into milk puddings or yoghurt
- If your child prefers pureed food, use soup, milk, white sauce or full fat coconut milk rather than water to obtain the right consistency. Water dilutes the food making it taste bland and reducing the nutrient content. Avoid sieving pureed food as this reduces its fibre content
 - When the mouth is sore, cool foods may be better accepted.

If weight gain falters at this time, your dietitian may suggest introducing a different milk that is more concentrated in nutrients such as one of the Nutrini® (Nutricia) or Paediasure® (Abbott) ranges. These are prescribable by your GP.

Constipation

Constipation is often the result of too little fluid, due to a reduced intake of feeds and/or increased requirements in hot weather. An EB baby with extensive blistering may have higher fluid requirements than normal.

Constipation can be caused or aggravated by iron supplements; it may also occur for no apparent reason. How often the bowels are opened is less important than the degree of discomfort felt. As long as motions are soft and painlessly passed, it is not essential that the bowels are opened daily.

For an EB baby, passing even a moderately bulky motion may cause pain and blistering of the delicate skin around the anus. Fear of pain on passing further motions can quickly lead to withholding motions. A vicious cycle is rapidly set up as he becomes more constipated and appetite is reduced. Regular bowel movements also depend on a regular intake of food, so a poor appetite and irregular feeds can lead to harder, drier motions. Try to ensure a generous fluid intake i.e. at least 150ml per kg (2-3oz per lb) per 24 hours for young babies who are not receiving fluid from foods. If he refuses plain cooled, boiled water, offer well diluted fresh fruit juice (i.e. 1 teaspoon juice with 100 ml water) or give ready-to feed baby juice diluted with an equal volume of water.

The importance of treating constipation cannot be overstated. If extra fluid does not help, a gentle laxative will be required and you should ask for specialist advice about this. It is very important to give this regularly as a preventative measure and not to wait until he is very constipated.

Once your baby has started taking solids, try to include fruit and vegetable puree daily. From about 9 months, offer wholegrain cereals such as Weetabix®, and from 10-12 months, include baked beans (and sweetcorn if he is able to chew this).

Prunes are very useful and often well-accepted when they are pureed and mixed with yoghurt, fromage frais, custard etc. The fibre in these foods, combined with an adequate fluid (about 100ml per kg, 1-2oz per lb) will help to keep the motions soft and they will be more comfortably passed. (See "Fibre".) Your dietitian can advise on suitable fibre sources; unprocessed bran is not appropriate.

Sugar and tooth care

In some types of EB, the tooth structure is affected making decay virtually inevitable. In others, tooth structure is normal but decay is common because:-

- blistering and scarring limit the tongue's ability to clean food debris from the gum margin
- bacteria in the plaque accumulated around the teeth react with the sugars in food to produce acid. The acid softens and dissolves the tooth enamel, exposing the sensitive nerves
- tooth brushing is often difficult in a fragile or tight mouth.

Decay causes pain and reduces food intake. Extraction or filling of decayed teeth are unpleasant procedures and if several are needed, a general

anaesthetic may be necessary. Extensive extraction causes permanent scarring in some types of EB, leading to tightness of the mouth and increased problems achieving good oral hygiene routines. So, it is important to try to preserve the teeth as much as possible.

Because children with the more severe types of EB need calorie intakes that cannot always be achieved without the consumption of sugar and frequent meals, it is important to strike a realistic balance. The number of times sugar containing items are consumed is as important as the overall amount taken. So if sugary foods are restricted to mealtimes, they are much less damaging to teeth than if they are eaten alone. A chocolate biscuit eaten straight after a savoury snack or main meal is less harmful than when it is eaten between meals, or worse, when it is nibbled slowly over several hours. A sugary drink should be drunk quickly, with or straight after a meal and followed by some sips of plain water to rinse the teeth.

Prevention is better than cure and compromise is possible. If sugar is used sensibly, its benefits as a high energy food can be exploited, whilst at the same time minimising the likelihood of tooth decay. Here are some important points:-

- Babies should not normally have sugar, cordial or fruit squash added to bottles

and dinky feeders should never contain anything except boiled water

- Dummies should never be dipped in honey, jam or sugar

- Lemonade, coca cola, cordial squash etc. should be avoided because of their sugar and acid content. Give instead water or the recommended milk formula

- For snacks, try cheese; sandwiches filled with smooth nut butter, Marmite®, cheese spread or meat/fish paste; or savoury biscuits which melt in the mouth

- Avoid giving sweets and high-sugar foods e.g. chocolate biscuits to babies under 1 year as this encourages a preference for these to the exclusion of more nutritious foods. If they are given, restrict them to mealtimes

- Finish meals with a savoury rather than a sweet food

- Even young children with only one or two teeth should see a dentist, preferably

one familiar with EB, who can give advice on cleaning techniques, mouthwashes, fluoride supplements etc.