

Nutrition in Epidermolysis Bullosa for children over 1 year of age

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DEBRA is the national charity that supports individuals and families affected by Epidermolysis Bullosa (EB) – a painful genetic skin blistering condition.

A charity registered in England and Wales (1084958) and Scotland (SC039654)

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About this booklet

All children need to be well nourished to thrive and to enjoy the best possible quality of life and this is especially important for children with the more severe EB types. Although there can be wide variation in severity between and within EB types, it tends to be children with the more severe forms of recessive dystrophic EB (RDEB), Dowling-Meara EB simplex (DM EBS) and junctional EB (JEB) who experience nutrition-related problems. This is because their nutritional intake is limited by factors associated with their EB and at the same time, some of their nutrition is diverted away from growth into wound healing. It is not possible to cure EB with "special" diets (eliminating particular foods or containing megadoses of certain nutrients). However, by promoting optimal nutrition, 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 young HJEB child'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 very young children with or without EB. This is followed by practical information specific to feeding EB children from about 1 year of age onwards.

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.

For information on nutrition for babies with EB, please refer to:

Nutrition for Babies with Epidermolysis Bullosa by Lesley Haynes RD

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Why is nutrition important in EB?

Children who do not have EB channel most of their nutrition into growth. Because EB children's 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 children have "off days", e.g. due to minor illness, when food intake is reduced, and EB children are no different. However, this happens more often for EB children if they have also to contend with blisters in the mouth and throat which make eating uncomfortable and may reduce food intake significantly.

Parents naturally become very anxious at these times. 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.

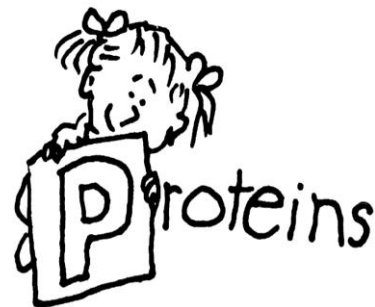
What is a nutritious diet?

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 the most concentrated sources of calories in the diet 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. Butter, margarine, cream, oil, lard, suet and dripping are obvious sources of fat. Less obvious sources are full cream milk, full fat yoghurt, many cheeses (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. On the other hand, some EB children can become overweight, often because of reduced mobility and/or reliance on a wheelchair. They may need to limit their fat intake to avoid further reduction in mobility.

Carbohydrates

Carbohydrates are an important source of calories and, in some cases, of fibre, vitamins and minerals. Carbohydrates that are useful mainly as a source of calories include biscuits, sugar, sweets, glucose, honey, jam and syrup. Carbohydrates containing a wider range of nutrients include cereals, breakfast cereals, flours, pasta, bread, potatoes, fruits and pulses. Puddings and cakes are valuable mainly for their calorie content, but can also be good protein sources if they are made with eggs and milk products. All carbohydrates are important; the sweet ones (biscuits, sugar etc) should be included with, but not instead of, the less sweet ones (cereals, potato etc). However, if your child has a tendency to become overweight, he may need to restrict his intake of sugary foods.

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 and bowels).

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, blackcurrant drinks eg. Ribena® 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 eg. protection of cell membranes and 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 is mainly found in 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 to a lesser extent in dairy products, bread and cereal products such as wheatgerm. It plays essential roles in many complex metabolic processes including wound healing.

Selenium has a vital role, with Vitamin E, in protecting against cell damage and promoting immunity. It is found in Brazil nuts, dairy foods, some seeds and grains.

Calcium, with vitamin D, is essential for healthy bones and teeth, also for muscle and nerve functioning and blood clotting. Calcium is found in milk and milk products such as cheese and yoghurt.

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, excessive intakes can be harmful, so always ask for dietetic advice before introducing any form of supplementation.* Blood tests can help to identify a need for supplementation and, depending on the type of EB, are carried out as part of routine medical care.

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.

After the first year

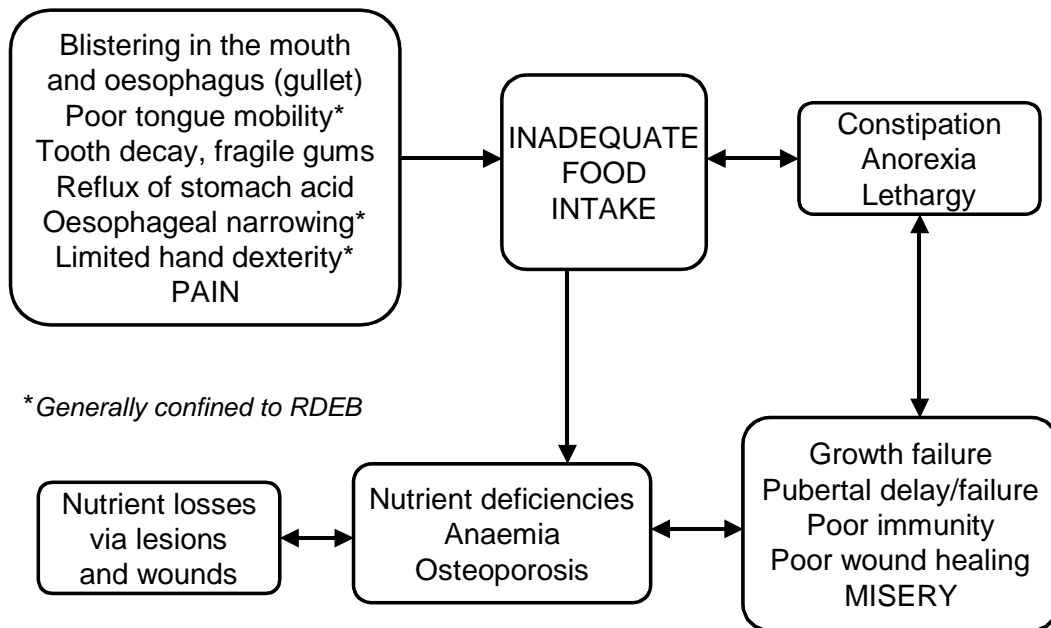
By the age of about 1 year, many EB children can eat the same meals as other family members provided that foods are of a suitable consistency and not too spicy or salty. Citrus fruits, e.g. oranges or tangerines, tomatoes or fresh fruit juices may cause discomfort if the mouth is blistered. Foods which are likely to cut or scratch and blister the mouth or throat should be avoided e.g. hard crusts and sharp chips, toast, crackers, hard raw vegetables such as carrot and celery. Take care with foods that can be somewhat tough such as the skin on fresh sausages (tinned sausages e.g. Plumrose® have very soft skin), some brands of baked beans (mash these slightly to break up the 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. Fish is very nutritious, but always check that all bones have been removed.

Promotion of confidence in eating is very important, and if EB children prefer to continue with pureed or well mashed foods, this should be encouraged rather than being viewed as lack of progress.



The emphasis should be on a good, balanced diet with plenty of protein. If the appetite is small, offer 3 small meals a day with nutritious snacks in between (there is a suggested meal plan on page 22). 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.

Nutritional problems in children with severe EB - causes and effects



Children with severe EB are often unable to eat enough to satisfy hunger and to promote growth and healing. This significantly reduces their everyday quality of life and adds greatly to their parents' stresses and anxieties. The diagram below shows how various factors can interact and how easily vicious cycles become established, despite parents' very best efforts.

Oral and oesophageal problems lead to slow, tiring and sometimes painful eating and such negative experiences often lead to reluctance to eat or to try new foods. The memory of even just one unpleasant episode of painful swallowing or choking can cause fear of eating and lack of confidence long after the event. Gastro-oesophageal reflux (GOR, the return of acidic stomach contents back up the oesophagus) is common in EB, it can be very painful and invariably causes some degree of aversion to eating.

It is very important to treat GOR promptly, not only because of its negative impact on eating, but also because of the corrosive effect of stomach acid on the delicate lining of the oesophagus causing an increased likelihood of narrowing (stricture). In some cases, GOR produces no obvious symptoms, but still causes internal damage, so it is very important to follow your child's specialist's advice about medication dosage.

When eating and swallowing problems are persistent, one mealtime commonly drifts seamlessly into the next and this is undesirable for several reasons:-

- it is wearisome and demoralising for both the child and his carer, detracting from the pleasure that should be a part of mealtimes
- it is very time-consuming, leaving less time for other activities
- it inhibits the development of an appetite

- it increases the likelihood of tooth decay
- it increases the likelihood of constipation

Pureeing foods allows the child to eat more quickly and with greater confidence. Setting time limits on meals minimises the frustrations felt by all concerned and reduces reinforcement of negative associations with food. It is important to strike a realistic balance between encouraging and helping the child to finish meals and still allowing time for play. Praise small achievements and don't be tempted to force feed; this increases fear and is counter-productive in the long run.

Pureed foods



Some children can manage only very soft, pureed or liquid foods. This rapidly becomes boring if it relies on soup and ice cream, but the inclusion of suitable family dishes makes for much more variety and, equally importantly, makes the child feel less different from others. Children usually like to see food before it is pureed so that they know that they are having the same as everyone else.

Pureed meals look more attractive if each food is pureed separately and if colourful ones such as carrots, baked beans and peas contrast with meat on the plate. This also allows the child to savour the different tastes of each food on his plate. He may find that cool, cold or lightly frozen foods soothe a sore mouth or throat.

Some dishes can be frozen in individual portions for use when the family meal is not suitable for pureeing. Use soup, full fat milk, white sauce or full fat coconut milk to puree foods rather than water. Water dilutes the nutrient content of the dish and makes it taste bland. Avoid sieving food as this reduces its valuable fibre content.

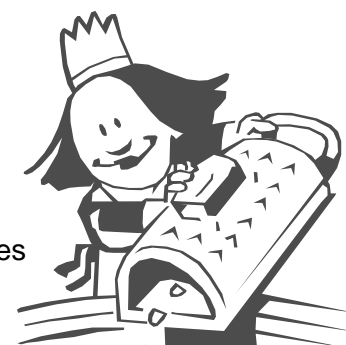
Boosting the nutrient content of meals

Although it is easy to blame everything on the complications of EB, normal events such as illness also interfere with eating and drinking. These are an unavoidable part of every child's development, but they are very stressful times for parents. Children rapidly pick up on parental worries around food, so if your child is having an off day, for whatever reason, try not to transmit your anxiety as this can increase his fears.

When the appetite is small, it helps to boost the nutrient content of meals without increasing their bulk. The child who is accustomed to having food boosted when he is well is more likely to accept this when he is less able to eat. So it is a good idea to include boosts on a regular basis to minimise the likelihood of their being rejected at a time when they are most needed. Make the most of protein foods such as eggs, meat, fish, milk, cheese and dishes containing these and exploit the calorie content of high fat foods (butter, margarine, oil, mayonnaise, cream, full fat coconut milk, evaporated milk) and sweet foods (sugar, jam, honey, sweet spreads and syrup). Try home-made milk shakes (page 23) and encourage your child to help you make them.

Here are some ideas:

- Add grated cheese/cream cheese/mayonnaise to mashed potato, baked beans, spaghetti, omelette/scrambled eggs
- Serve white or cheese sauce with added cream with fish and vegetables
- Add butter, margarine, olive oil or mayonnaise to hot vegetables
- 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 easy access)
- Spread butter, margarine, cream cheese or smooth nut butter liberally on bread, soft toast and plain sweet and savoury biscuits
- Top bread or biscuits with jam, honey, lemon cheese or chocolate spread. Mix any of these spreads into milk puddings or yoghurt
- Make savoury dips e.g:
 - Avocado mashed with mayonnaise
 - Hummus/taramsalata mixed with mayonnaise/crème fraîche
 - Smooth liver pate mixed with mayonnaise/crème fraîche
- Make sweet dips e.g:-
 - Mango mixed with fromage frais
 - Banana mashed with brown sugar and crème fraîche
 - Banana mashed with lemon cheese
- Add a raw egg and/or evaporated milk to home-made milk pudding e.g rice/tapioca before baking
- 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 using a small tin of evaporated milk to replace an equal volume of water
- Add 4 tablespoons skimmed milk powder per 500ml full fat milk



Commercial nutritional supplements

When chewing and swallowing are particularly difficult for your child, the ideas above may only partly satisfy his increased nutritional requirements. Fortunately, a very wide variety of commercial nutritional supplements is available and most can be obtained on prescription from your general practitioner (GP). The supplements come in various presentations:

Multi-nutrient, sweet drinks and soups, some containing fibre

The sweet drinks are available in a wide variety of flavours and are presented in very “child friendly” packaging, often in a tetra-pack with a straw. As they resemble regular supermarket milk shakes, children are often happy to take them to school to supplement or replace a meal or break-time snack. The soups can make a welcome savoury change for children who tire of sweet tastes

Multi-nutrient (dried) pureed meals, some containing fibre

These are available in several flavours and need to be reconstituted with milk or water.

Multi-nutrient desserts

These resemble regular smooth, blancmange-style desserts

Pure energy supplements (carbohydrate or fat or a combination)

These are available in powder and liquid, neutral and flavoured. They can be added to foods or feeds; the liquid can be taken alone as a medicine

Energy and protein supplements (carbohydrate, fat and protein)

These are available as powder and liquid, neutral and flavoured. They can be added to foods or feeds; the liquid can be taken alone as a medicine.

Pure fibre (powdered) supplements

These can be added to foods or feeds.

The number of available products is constantly changing and published information quickly becomes out of date, so individual product details have not been included here. Please contact your dietitian for the most current information and advice on the best supplements for your child’s needs. Children tire very quickly of supplements and it is important to “ring the changes” frequently - another good reason to keep in regular contact with your dietitian so that she can tell you about new additions to the available range.

Iron-deficiency anaemia and iron supplements

EB children can become anaemic (as indicated by a blood test) for two main reasons. Firstly, when their skin is very fragile, they regularly lose blood from wounds and this can happen internally in the mouth, oesophagus, gut and anus as well as externally from skin lesions. Secondly, some children find meat (the best dietary source of iron) difficult or impossible to chew and swallow. Chronic anaemia causes tiredness and an early symptom is poor attention span in school or lethargy in the afternoons. Anaemia also negatively affects wound healing and exerts other subtle adverse effects on overall health.



Unfortunately, iron supplementation often causes or worsens constipation (less commonly it causes diarrhoea), especially if it is taken on an empty stomach. It is extremely important that such unwanted consequences are dealt with so that the iron can continue to be taken, rather than stopping it. Supplementary iron is available in tablet and liquid form on prescription from a GP and it is absorbed best if the total daily dose is split into at least 2 doses (this also minimises adverse side-effects) and taken at the same time as a rich source of Vitamin C. To maximise absorption, iron and zinc supplements should be taken at different times of the day.

Zinc deficiency and zinc supplements



Meat is one of the best dietary sources of zinc. However, many EB children find meat difficult to chew and swallow, so their intakes may be poor. Zinc supplementation should not be started solely on the basis of a low level of zinc in the blood as this may be a reflection of additional dietary shortfalls which your dietitian can discuss with you and make appropriate recommendations. Zinc supplements are usually prescribed in liquid or effervescent tablet form for EB children. Lozenges that are designed to be sucked can also be bought over the counter from chemists and health food stores. Unfortunately, zinc often causes nausea and occasionally vomiting, and this invariably leads to poor compliance. To minimize unwanted side-effects, zinc should not be taken on an empty stomach. To maximise absorption, zinc and iron supplements should be taken at different times of the day.

Selenium deficiency and selenium supplements

Selenium is needed in only very small amounts and deficiency usually takes years rather than months to develop. Selenium deficiency usually occurs in association with other nutrient shortages, so the whole diet should be assessed and all deficits addressed, rather than prescribing supplementary selenium in isolation. Selenium cannot be prescribed by a GP; it must be prescribed by a hospital doctor and dispensed from a hospital pharmacy. It is available only as small tablets which should be crushed for those with swallowing difficulties.



Oesophageal dilatation (OD)

Children with RDEB often develop narrowing of the oesophagus which limits their ability to swallow food, and sometimes even liquids. Oesophageal dilatation (OD) is a surgical procedure during which the oesophagus is gently stretched, under anaesthetic, to relieve the narrowing (or stricture). For some children, this procedure significantly eases swallowing difficulties and it generally needs to be repeated from time to time. Children who are unable to maintain an adequate nutritional intake despite OD, may be recommended to have gastrostomy tube placed.

Feeding tubes e.g. naso-gastric (NG) and gastrostomy tubes

A sore mouth and painful swallowing are huge disincentives to eating and significantly reduce food intake. By supplying some of the child's nutritional requirements straight into the stomach (by-passing the main trouble spots of mouth, throat and oesophagus), he can continue to enjoy the foods he is able to take by mouth and be "topped-up" by the tube feeds. NG tubes (soft, narrow tubes passed via the nose, down the oesophagus and into the stomach) are recommended rarely and then only for short-term use. Children understandably object to the attention that they attract and they are difficult to secure to fragile skin.

Gastrostomy tubes (often called "buttons"; small devices surgically placed in the stomach wall) are concealed under clothing. The operation to place them is reversible, and they can be used as much or as little as necessary, depending on the child's oral intake. The child eats and drinks as much as he is comfortably able to, with the remainder of his nutrition given through the button. The button can also be used for giving unpalatable medicines and for pain relief and sometimes buttons are placed for this purpose alone.

Tube feeds

Passing pureed food through a naso-gastric or gastrostomy tube is not recommended because of the likelihood of blockage and growth of harmful bacteria. Instead, one of the many commercially-manufactured tube feeds, which are available on prescription from your GP, should be used. The dietitian can advise on the best choice of feed, depending on your child's age and requirements.

Although, physiologically, it is natural to fast during the night, many EB children prefer to have their tube feed at this time, leaving the daytime for meals and other activities. Overnight feeding sometimes leads to the need to pass urine or open the bowels during the night, involving a trip to the toilet or a soiled nappy and broken sleep for the child and other family members. These consequences are more likely when the feed volume totals more than about 500ml.

Tube feeds can be given during the day and if the required volume is not over about 200ml per feed, this can easily be fitted into the daily routine, at home and in school. There are many different permutations of feed timing and delivery. The important thing is that each child has his individual situation and requirements assessed by the dietitian and that an appropriate plan is drawn up and regularly reviewed.



The role of sugar

Children with the more severe types of EB need calorie intakes that often cannot be achieved without the consumption of sugar and frequent meals, so it is important to strike a realistic balance. Dentists advise that the frequency of sugar consumption is as important as the overall amount taken. So if sugary foods are restricted to mealtimes, they are less damaging to teeth than if they are eaten alone.

A chocolate biscuit eaten with a meal is less harmful than when it is eaten between meals, or worse, nibbled slowly over several hours. A

sugary drink should be drunk quickly, with or straight after a meal and followed by sips of water to rinse the teeth. Compromise is possible and used sensibly, sugar's benefits can be exploited, whilst minimising the likelihood of tooth decay.

Here are some important points:

- If young children are still using feeding bottles, they should never contain anything except water
- Encourage the progression from bottle to cup
- Even low sugar varieties of lemonade, coca cola, cordial squash etc should be restricted to mealtimes as they contain acid which weakens tooth enamel
- Instead of sweet snacks, offer cheese, sandwiches filled with smooth nut butter, Marmite[®], cheese spread or meat/fish paste; or savoury items which melt in the mouth such as Tuc[®], Wotsits[®], Skips[®], Quavers[®], prawn crackers
- End meals with a savoury rather than a sweet food
- Even young children with few teeth should be seen by a dentist, ideally one familiar with EB, who can give advice on cleaning techniques, mouthwashes, fluoride supplements .etc

Tooth decay

In some types of EB, tooth structure means that decay is virtually inevitable. In others, structure is normal but teeth decay because:

- food debris accumulates around the gum margin because blistering and scarring limit the tongue's ability to clear it away
- 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



Tooth decay causes pain and reduces food intake. Extraction or filling of decayed teeth is difficult to carry out in EB and involves additional pain. If several extractions are needed, a general anaesthetic may be necessary. Extractions cause permanent scarring in some types of EB, leading to increased tightness of the mouth. In turn, this causes further problems achieving both good oral hygiene routines and a satisfactory nutritional intake.

So, it is extremely important to try to preserve the teeth as much as possible. Ask your dental specialist for advice about fluoride supplements and suitable mouthwashes and toothbrushes.

Constipation and bowels

Regardless of the severity of their skin problems, many EB children experience extremely painful bowel motions and become increasingly reluctant to open their bowels. This can be one of the least well-recognised and distressing aspects of EB.

Constipation makes EB children feel listless and apathetic and it can have a devastating effect on appetite and quality of life. Some children's motions are hard and they are constipated according to the medical definition. Others feel pain and avoid opening their bowels even when their motions are soft. In this booklet, both situations are called "constipation" as the management is similar for each.



The negative effect that chronic constipation exerts on eating is underestimated. The anal skin is very delicate and even a "normal" motion has the potential to tear it, after which pain is felt every time the child opens his bowels. He learns quickly to oppose or ignore the urge to open his bowels (sometimes for several days). He may be unable to resist passing a small motion, but retains the rest which becomes progressively drier and harder. A bowel that is loaded with hard "rock-like" motions makes the child feel bloated and very uncomfortable. This reduces his appetite and so he eats less. Less food means less bulk in the bowel to stimulate a motion and so on as the vicious cycle develops (see page 9).

The child may feel abdominal or anal pain as soon as he smells food or is asked to sit at the table for a meal. Refusal to sit or eat is often mistaken for naughtiness and manipulative behaviour. Even the anticipation of eating can be enough to cause abdominal pains, as the muscles in the bowel try to move the motions along to make room for the next meal.

The main ways of managing constipation are:

- Increasing fibre (and fluid) intake and/or
- Taking laxatives/softeners

Both these approaches influence the consistency and bulk of the motions and affect the speed at which they pass through the gut. Increases in dietary fibre and fluid can often very successfully help constipation and for those whose EB is relatively mild, and who can eat the appropriate foods while still maintaining satisfactory growth, this should be the treatment of choice.

Suggestions for increasing fibre intake



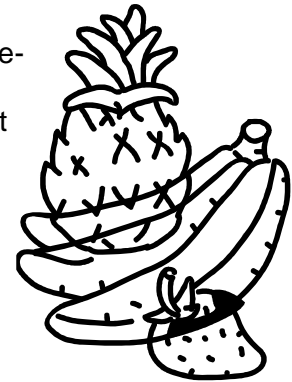
Include cereal fibre by using wholegrain breakfast cereals such as Weetabix[®], Shreddies[®], Bran Flakes[®], Ready Brek[®], porridge. Try wholemeal and pumpernickel (German rye) bread, wholegrain biscuits e.g. Digestive, flapjacks and cereal bars, brown rice and wholemeal pasta. Try using 50% wholemeal and 50% white flour in home baking.

Eat fruit, especially raw. Leave the skin on apples, pears, apricots, peaches, grapes etc. Include prunes, figs, dried apricots, raisins, sultanas. Many fresh fruits can be made into “smoothies”.

Some words of caution about fibre-containing foods

Children with mild EB should be able to chew and swallow regular fibre-containing foods and these can be regarded as part of regular healthy eating pattern. However, for children with severe EB, it is generally not appropriate to advise these because:

- Foods such as muesli, wholemeal bread, fresh fruit and salads are difficult or impossible to swallow.
- Fibre-containing foods are comparatively low in energy. Severely affected EB children need enough calories to grow properly and heal.
- The bulky nature of such a diet makes it very filling and may reduce the appetite for more valuable foods.
- Unprocessed bran is not suitable for children.

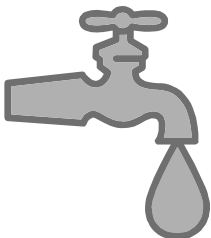


Other sources of fibre

Children who can't (or won't) eat conventional forms of fibre may benefit from:

- A pure fibre source such as Resource Benefiber[®] (Novartis). This is virtually tasteless, mixes with many liquids and foods and is generally well-accepted
- A fibre-containing, multi-nutrient supplement. Several companies produce these in different flavours.

Ask your dietitian for advice regarding their suitability for your child. If the item is available on prescription, the dietitian can write to your GP requesting this.



Encourage your child to drink plenty of fluids preferably water. As a general rule of thumb, children should have 6-8 drinks a day. The size of the drink depends on the size of the child. A toddler would take about 150ml per drink; an older child could drink 200-300ml.

Laxatives and softeners

These can help greatly to counteract the difficulties that many EB children experience in opening their bowels, however they should be introduced with care when a child is already significantly constipated. If the bowel contains “rocks”, a stimulant laxative (e.g. senna [Senokot]) may only increase abdominal pain as the bowel muscles squeeze on the hard motions.

A softening laxative (e.g. lactulose) will have a limited effect on the “rocks”, softening some of them but leaving the rest behind. Very loose motions often cause “accidents” when the child sneezes or laughs and motions are unintentionally leaked. Such outcomes are sure routes to non-compliance, so it is very important to ask your child’s specialist for advice on the best regime.



A suggested meal plan for a child with mild to moderately severe EB

<i>Breakfast</i>	<p>Cereal (ideally containing fibre, eg. Weetabix[®], or porridge) plus milk*</p> <p>Bread or toast (preferably wholemeal) with butter/margarine, honey, jam, marmalade, peanut butter</p> <p>Milk*</p>
<i>Mid-morning</i>	Milk*, water, diluted fresh fruit juice or fruit squash plus fresh fruit, biscuit or cake
<i>Lunch</i>	<p>Meat with gravy or fish and sauce</p> <p>Potato, rice or pasta</p> <p>Two vegetables</p> <p>or a dish such as lasagna, shepherd’s pie, macaroni cheese</p> <p>Fresh or tinned fruit, ice cream, custard</p> <p>Milk*, water, diluted fresh fruit juice or fruit squash</p>
<i>Evening</i>	<p>Egg, ham, cheese, baked beans, sardines, liver sausage</p> <p>Jacket potato or wholemeal bread and butter/margarine</p> <p>Yoghurt or fromage frais or milk pudding e.g. custard, rice</p> <p>Milk*, water, diluted fresh fruit juice or fruit squash</p>
<i>Bedtime</i>	Milk* or cereal and milk*



* Aim for 500ml full fat milk per day unless excessive weight gain is a problem in which case semi-skimmed milk should be used. Skimmed milk is unsuitable for small children unless on the advice of a dietitian.

Recipes for home-made milk shakes

(These are best served chilled)

Ice cream milk shake	
200 ml full cream milk 2 tablespoons skimmed milk powder. Milk shake flavouring eg. Crusha or Nesquik 1-2 scoops ice cream	Whisk the first three ingredients together until well- combined and the milk powder has dissolved. Float ice cream on top.
Yoghurt shake	
150 ml yoghurt, preferably full fat eg. Greek yoghurt 3 teaspoons runny honey 1 small, ripe banana	Liquidise all the ingredients together until smooth.
Peach or apricot shake	
100 ml evaporated milk 100 g tinned peaches or apricots	Liquidise all the ingredients together until smooth.
Prune and orange shake	
75 ml prune juice 75 ml orange juice 2 teaspoons brown sugar	Whisk all the ingredients together until the sugar has dissolved.
Banana shake	
150 ml full cream milk 1 dessertspoon double cream 2 tablespoons skimmed milk powder 1 small, ripe banana 3 teaspoons brown sugar or maple syrup	Liquidise all the ingredients together until smooth.

