PREVENTATIVE NUTRITIONAL CARE GUIDELINE

Constipation management for children and adults with epidermolysis bullosa (EB)

An expert working group consensus

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Disclaimer

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CONTENTS

ABSTRACT

DEBRA International (DI) is undertaking a long-term initiative to develop clinical practice guidelines (CPGs) for epidermolysis bullosa (EB) in order to improve the clinical and social care of people with the condition. This clinical practice guideline (CPG) focusing on the preventative nutritional care management of constipation for children and adults. It describes the development of the CPG recommendations in a rare disease such as EB and highlights key links to previous established guidelines. This guideline appraises the use of adequate fibre and fluid to prevent constipation ***verses*** a low fibre diet. The recommendations follow the Scottish Intercollegiate Guidelines Network (SIGN)[1] methodology and, where strong evidence was lacking, were reached by a working group's consensus expert opinion.

What is already known about this topic?

- There are a small number of publications identifying constipation as a problem in people with all types of EB.
- Enteral feeds containing fibre and supplementary drinks containing fibre have been successfully used to treat people with EB who have constipation. Guidelines are available for management of constipation in the general population.

What does this guideline add?

This article provides an overview of the evidence on the incidence, causes, prevention, and dietary treatment of constipation in people with Epidermolysis Bullosa. It combines what is already known in the literature with expert opinion and current clinical practice. It also provides information on fluid and fibre requirements to act as a working guide for patients and clinicians around the world.

This CPG acknowledges existing guidelines by signposting with this icon though this document. These are referenced to on page 30.

Key words

Epidermolysis Bullosa, Rare disease, Nutrition, Constipation, Fibre, Hydration, Guideline, Recommendations

Table 1. Summary of key recommendations to increase frequency of stools, prevent anal blistering, and improve consistency of stools.

a) Key recommendation	Grade strength of recommendation	Quality of evidence (Rate Average)	Key references
We recommend offering adequate fibre from food sources.	**B ✓**	**2++**	**30, 29, 24*, 14***
We recommend the use of oral or enteral feeds containing fibre when additional nutritional support is required.	**C ✓**	**2+**	**31, 33, 34, 35, 37, 38, 18**
We recommend offering artificial fibre to attain adequate oral fibre intake if dietary intake is inadequate.	**C ✓**	**2+**	**28, 10**
We recommend offering adequate fluid intake.	**C ✓**	**2+**	**27, 34, 4**
We suggest offering polyethylene glycol in combination with fibre and fluids when constipation remains unresolved on a high fibre diet.	**D ✓**	**3+**	**33, 11*, 4 Expert opinion**

	Key	* articles where the sample population did not have epidermolysis bullosa participants
bi)	**Grades**	**Descriptions in accordance to SIGN[1]**
	B	A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overall consistency of results; or extrapolated evidence from studies rated as 1++ or 1+
	C	A body of evidence including studies rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; or extrapolated evidence from studies rated as 2++
	D	Evidence level 3 or 4; or extrapolated evidence from studies rated as 2+
bii)	**Quality rate**	**Descriptions in accordance to SIGN[1]**
	2++	High quality systematic reviews of case control or cohort studies; high quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
	2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
	3	Non-analytic studies, e.g. case reports, case series
	4	Expert opinion
biii)	**✓**	**Recommended best practice based on the clinical experience of the guideline development group[1]**
	Notes:	There was no disagreement on the quality of the appraised articles or the strength of the recommendations. For EB-specific articles used for the recommendations see Annex IV.

C

Recommendation

The EB pain management best practice guideline recommends constipation should be addressed nutritionally, ensuring adequate hydration and fibre in the diet to keep improve stool consistency and reduce medication use[19].

Chronic constipation in which the stool is hard and causes pain and blistering on defaecation is a common feature of EB[2-6] especially in RDEB[21]. Faecal impaction can lead to overflow diarrhoea and soiling. While awareness is high regarding problems of the upper gastro-intestinal tract in EB, (such as oral blistering and oesophageal stricture) affecting dietary intake and growth, the negative impact on appetite from constipation can be underestimated[7]. While many aspects of EB are difficult to manage, constipation can be prevented. This can improve the nutritional intake and improve the quality of life of the person with EB.

Without early diagnosis and treatment, an acute episode of constipation can lead to anal fissure and subsequent chronic constipation in children without EB[8]. Prevention of constipation is not expensive but requires an understanding of the factors involved in causing constipation in order to optimise bowel function. The National Institute for Health and Clinical Excellence (NICE)[8] recommend that once constipation has been diagnosed, the first-line treatment is not diet alone but in combination with prescribed laxatives and behavioural changes relating to bowel habits[8].

Objectives of guideline and purpose

- To provide information regarding current best practice for the prevention of constipation through the use of nutrition for all ages and all types of EB.
- To promote increased oral intake of fibre-containing foods and fluids.
- To encourage regular pain free bowel movement.
- To reduce risk of chronic constipation, faecal impaction, pain and rectal bleeding, anal stricture.
- To be used as a tool for prevention and treatment of constipation when there is no access to a specialist dietitian.
- To encourage collaboration from around the world by the inclusion and addition of high fibre food table (Annex I).

We have not sought to provide guidelines for the diagnosis of constipation or faecal impaction. There are guidelines available for the medical management of constipation[8]. In infants and children it is advisable to rule out other medical causes of constipation in addition to treating it as a current symptom of EB. For example, this may include ruling out coeliac disease or cow's milk protein allergy where constipation may also be a symptomatic feature[9].

Guideline users

This CPG is intended to increase the knowledge of those involved in optimising nutritional status in EB. This includes patients, parents and carers, dietitians and nutritionists, dermatologists, paediatricians, gastroenterologists, nurses, dentists, surgeons, psychologists, speech and language therapists, physiotherapists, occupational therapists, radiologists, and general practitioners. Close collaboration between all health professionals is key in striving for optimal management and quality of life for patients with EB, particularly the severe types.

Target user of guideline

The CPG comprises information relating to EB patients of all ages and diagnosed with any of the four major types of EB: EB simplex, Junctional EB, Dystrophic EB and Kindler EB.

Constipation is defined as reduced frequency of bowel movement with or without hard, dry stools that are difficult or painful to pass[10-11]. It is a symptom, which can present as an acute or chronic condition, not a disease.

In accordance with the world gastroenterology organisation practice guideline12, functional constipation is diagnosed when patients who do not take laxatives report at least two of the following in any 12-week period during the previous 12 months[10-12]:

- Fewer than three bowel movements (BMs) per week for adults[8, 10-12]
- Fewer than four BMs per week for children[10-12]
- Hard stool in more than 25% of BMs[10-12]
- A sense of incomplete evacuation in more than 25% of BMs[10-12]
- Excessive straining in more than 25% of BM[10-12]
- A need for digital manipulation to facilitate evacuation[10-12]

BMs can vary between children and adults. In addition, constipation can be perceived differently by patients; where some may regard constipation as straining (52%), for others it means hard, pellet-like stools (44%), an inability to defaecate when desired (34%), or infrequent defaecation (33%)[12]. Constipation is common in childhood. It is prevalent in 5-30% of the paediatric population, depending on the criteria used for diagnosis[8, 13-14].

During clinical assessments all health professionals in all settings are able to use the Bristol Stool Chart Form scale[15] to identify constipation as well as diarrhoea, Figure 1 (Annex II)[15]. The stool types 1 and 2 are used to identify constipation, (Figure. 1). The “Bristol Stool Form Table” has been adapted and validated in other languages, such as Spanish16 and could be reliably used by doctors, nurses, health professionals, and patients[16].

Figure 1. The Bristol Stool Chart Descriptor Scale[15-16]

Type	Description	Assessment
1	Separate hard lumps like nuts (difficult to pass)	Very constipated
2	Sausage shaped but lumpy	Slightly constipated
3	Like a sausage but with cracks on its surface	Normal
4	Like a sausage or snake, smooth and soft	Normal
5	Soft blobs with clear-cut edges (passed easily)	Lacking fibre
6	Fluffy pieces with ragged edges, a mushy stool	Inflammation
7	Watery, no solid pieces, entirely liquid	Inflammation

The incidence of constipation is common in children and adults in all types of EB as illustrated in Table 2 but is most common in those with recessive dystrophic EB (RDEB).

Table 2. Incidence of constipation in EB

Participants	Incidence of constipation	Rate quality of evidence	Study type and reference
3280	75% in RDEB-HS (n=141) 50% in RDEB-nHS (n=264) 75% in RDEB-inversa (n=17) 20% in JEB-NH (n= 191) 20% in DDEB (n=425) 7% in EBS-WC (n=1092) 25% in EBS-DM (n=114) 12.5% in EBS-K (n=96)	**2++**	Cross-sectional and longitudinal study of both children and adults[21]
22	13.6% in JEB-H (n=22)	**2++**	Longitudinal data on infants diagnosed with JEB-H[22]
101	54% of RDEB (n=19/35) 30% of JEB (n=8/27) 9% of DDEB (n=1/11) 8% of EBS (n=2/24)	**2-**	Cohort study retrospective of both children and adults[6]
108/248	12% DDEB (n=3/25) children 21% DDEB (n=8/39) adults	**3**	Qualitative questionnaire study of both children and adults[17]
Key	RDEB: recessive dystrophic epidermolysis bullosa; RDEB-HS: RDEB Hallopeau-Siemens reclassified as RDEB generalised severe; RDEB-nHS: RDEB non- Hallopeau-Siemens reclassified as RDEB generalised intermediate ; RDEB-I, RDEB Inversa no change in classification: JEB: Junctional EB; JEB-H: JEB Herlitz reclassified as JEB generalised severe; JEB-NH: JEB non-Herlitz reclassified as JEB generalised intermediate; JEB-PA: JEB with pyloric atresia; DDEB: dominant dystrophic EB; EBS: EB simplex; EBS-WC, Weber Cockayne reclassified as EBS localised; EBS-DM: EBS Dowling Meara reclassified as EBS severe generalised; EBS-rec: EBS recessive; EBS-MD: EBS with muscular dystrophy		
Quality rate	Descriptions in accordance to SIGN1		
2++	High quality systematic reviews of case control or cohort studies; high quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal		
2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal		
3	Non-analytic studies, e.g. case reports, case series		

Table 2. Incidence of constipation in EB (Continuation)

Participants	Incidence of constipation	Rate quality of evidence	Study type and reference
223	40.3% in RDEB (N=23) 14.3% in JEB-NH (n= 7) 33.3% in JEB-PA (n=3) 46.3% in DDEB (n=54) 18.9% in EBS-WC (n=37) 37.9% in EBS-DM (n=29) 33.3% in EBS-rec (n=3) 50% in EBS-MD (n=4) 50% in Kindler EB (n=4)	**3**	Cohort study retrospective of children[18]
Key	RDEB: recessive dystrophic epidermolysis bullosa; RDEB-HS: RDEB Hallopeau-Siemens reclassified as RDEB generalised severe; RDEB-nHS: RDEB non- Hallopeau-Siemens reclassified as RDEB generalised intermediate ; RDEB-I, RDEB Inversa no change in classification: JEB: Junctional EB; JEB-H: JEB Herlitz reclassified as JEB generalised severe; JEB-NH: JEB non-Herlitz reclassified as JEB generalised intermediate; JEB-PA: JEB with pyloric atresia; DDEB: dominant dystrophic EB; EBS: EB simplex; EBS-WC, Weber Cockayne reclassified as EBS localised; EBS-DM: EBS Dowling Meara reclassified as EBS severe generalised; EBS-rec: EBS recessive; EBS-MD: EBS with muscular dystrophy		
Quality rate	Descriptions in accordance to SIGN1		
2++	High quality systematic reviews of case control or cohort studies; high quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal		
2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal		
3	Non-analytic studies, e.g. case reports, case series		

Dietary fibre

Dietary fibre or dietary plant non-starch polysaccharides (NSP) and lignin are resistant to hydrolysis by enzymes in the digestive tract, but they are an important component of the diet of all children and adults, benefiting bowel function[23]. Dietary fibre can be present in soluble and insoluble forms within a healthy diet[23]. NSP-rich foods tend to be less energy dense and bulky, which in turn may induce greater satiety, reduce appetite[23-13] and promote peristalsis[14]. There are two mechanisms that can explain the role of fibre in constipation:

- Bacterial decomposition of fibre produces short chain fatty acids, which increases the osmotic load[14]
- Indigestible fibre has the capacity to retain water and soften stools helping colonic motility[14].

The daily requirements for fibre in the United Kingdom (UK) for adults is 30g per day and for children: 15g at age 2-5 years, 20g at age 5-11 years, and 25g at age 11-16 years[23, 24].

- In the United States (US), daily fibre requirements are estimated for children using a formula of age (years) + 5-10 g daily[24, 25]. Adults fibre requirements are shown in Tables a, b, (Annex III).
- Studies have shown the treatment of constipation of non-organic cause with dietary fibre may be of benefit[43], but no clear guidance on type, amount, or duration exists. Therefore, normal fibre intake is advised for children with and without constipation[9].

Artificial fibre

The beneficial effects of fibre on gut health and its moderating effect on constipation[23] and diarrhoea have led to it being included in many manufactured feeds, both those intended for tube or enteral feeding and those intended as oral nutritional supplements (ONS)[6,18]. If enteral feeding is required,an age appropriate fibre feed can be considered from 6-8 months and is highly advisable from 9-12 months[6-7,18,28] and for all adults unless contraindicated.

Fibre intake can also be increased using a pure soluble fibre source such as Optifibre® (Nestle) which comprises partially hydrolysed guar gum[28]. Optifibre® is a virtually tasteless powder and its smooth consistency renders it barely detectable in foods and drinks; a vital factor in promoting acceptance[29]. In this respect, Hyfiber® (Nutrinovo) is a similar product for use by people over three years; it is a ready-to-use liquid containing polydextrose (8.6%) and fructooligosaccharides (FOS) (1.4%), which can be added to food or drinks, or given enterally. These products are easier to use than ones based on other fibre sources which set quickly, for example Fybogel® especially if the child or adult eats slowly, which, can be common in EB. Supplementary pure fibre should be introduced gradually, following manufacturer's instructions, usually over a week, and adequate fluid intake should be ensured to prevent gastric discomfort.

Poor fibre intake

Fibre intake of people with EB has been reported as low. It has been found to be significantly lower in those reporting constipation[29]. This may be due to a number of reasons[13,26,28,31]:

- Weaning diets may be delayed or established more slowly than in infants without EB, which may lead to a longer dependence on breast milk/formula milk with minimal fibre content[7].
- Painful episodes of teething/oral blistering may revert the weaning progress back to soft stages for prolonged periods, reducing the overall fibre content of the diet[7].
- Some patients may only manage a blended or pureed diet if they have oral or oesophageal blistering or stricture. This is likely to have a lower fibre content when compared to the equivalent unblended foods [2,4].
- Dental caries or reduced dentition means intake of foods that are difficult to bite and chew are reduced (e.g. raw fruits and vegetables)[32].
- Oral blistering and ulceration may result in avoidance of foods that are acidic (e.g. citrus fruits and tomatoes) or hard to chew (e.g. wholegrain products, raw fruits and vegetables) [26, 27]. Episodes of blistering make it difficult to maintain a regular pattern of eating and fibre intake.
- Oesophageal blistering or stricture may lead to avoidance of foods that may get stuck (e.g. husks of peas or beans) or foods that are hard, dry, or sharp causing blistering when swallowed (e.g. toast) [4,7].

- People with EB may choose more energy-dense foods and less fibre rich foods for optimal energy intake as the amount of food they consume in total may be small[2, 4, 26, 27].

Pain or discomfort

The need to pass a stool may cause abdominal pain, anal pain, and perianal blistering[6, 31]. This may result in avoidance of defaecation resulting in faecal retention and subsequent constipation[2, 20]. Young children often withhold stools when learning to use the toilet. Painful defaecation or fear of pain will exacerbate this[20]. The presence of perianal streptococcal infection may also contribute to discomfort[8]. Development of an anal fissure may result in avoidance of defaecation resulting in constipation[20].

Poor fluid intake

Fluid balance may be inadequate due to poor oral intake reducing the average number of bowel movements. Loss of exudate through wounds and increased temperature during infection will increase fluid requirements. In infants, a blistered mouth may severely affect milk intake and their ability to drink.[7, 34] Poor mobility and dexterity may reduce independent access to frequent drinks. Some schools and workplaces may not encourage drinking. People with EB may limit drinking to avoid feeling full or needing to go to the toilet during the night.

The side effects of medication

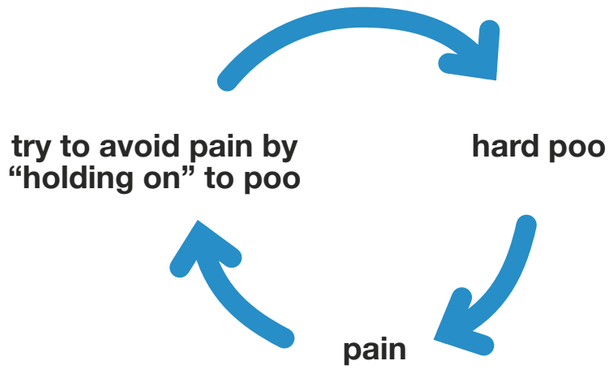
Medications for pain control including opiates and codeine reduce peristalsis[20]. Oral iron preparations for treatment of anaemia and Amitriptyline® used to treat pain can also cause constipation.

Mobility/activity

Mobility stimulates peristalsis but regular activity is often difficult for people with EB. Some children with EB may be slower to start walking. Reduced mobility may reduce appetite and make it more difficult for people to shop for and prepare fibre rich foods.

Other possible causes of constipation

- Episodes of oral blistering, oesophageal stricture, or infections make it difficult to maintain a regular pattern of eating and drinking, resulting in irregular bowel habits.
- In children, changes in routine including potty-toilet training, moving house, starting nursery, or major changes in family life may affect eating and drinking.
- Fears and phobias, including unfamiliar toilet facilities can result in withholding stool.
- Toilet facilities that are lacking in privacy.



B

Strong recommendation

- Optimise fibre using soft fibre rich foods such as oats, pulses, soft fruits and vegetables (Annex I). Liquidised fruits and vegetables in stews, soups and smoothies can be useful if eating is slow and chewing and swallowing difficult, Tables 1, 3 (Annex I).

C

Recommendation

- If a gastrostomy tube is being used for additional feeding then a feed containing fibre or additional fibre supplement (such as Optifibre®) can be added if using a feed that does not contain fibre, Table 1, 3 (Annex III). This is advisable even if constipation is not yet present[33].
- Ensure adequate intake of fluid, based on age and body weight then adjust for additional losses, Tables c, d (Annex III). If a gastrostomy tube is present it can be used to give fluids.

Disclaimer

For the following, Table d (Annex III) does not apply and you should consult a medical team

- In adults who have not reached adult weight.
- Increased fluid requirements will also be required in people with high losses of wound exudate

D

Recommendation

- Fluid used to prepare polyethylene glycol is required to soften the stool and be excreted, Table 1, 3 (Annex III). This should be in addition to the daily fluid requirement.
- Optifibre® can be introduced from 6-9 months of age, or Hyfiber from 3 years. As constipation is so common in EB, it is advised to prophylactically add in a fibre source for all children at 12 months of age[33], Table 1, 3 (Annex III).
- If medication is started in which constipation is a known side effect consider pre-emptive management (such as polyethylene glycol), Table 1, 3 (Annex III).

Table 3a. Evidence for the use of dietary fibre as a specific treatment of constipation

Number of participants	Treatment of constipation and population group	Quality of evidence	Study and global location
Fibre from food sources			
N/A	Dietary fibre, oral nutritional supplements, and tube feeding in EB children	**2++**	UK questionnaire study[30]
15	Dietary fibre, oral nutritional supplements, and tube feeding in EB children`	**2++**	UK prospective study[29]
	Dietary fibre in Childhood of the general population (non-EB)	**2+**	US survey in children[24*]
561/778	Dietary fibre in school children of the general population (non-EB)	**2+**	Japanese prospective questionnaire study[14*]
Use of oral or enteral feeds containing fibre when additional nutritional support is required			
13	Enteral fibre feeds in children with EB	**2+**	UK retrospective evaluation /cross-sectional study using questionnaire[34]
12	Placement of a gastrostomy tube allowed for fibre feeds to RDEB adults	**2+**	UK retrospective review[35]
20	Fifteen out of 20 children using 1-3 ONS drink containing 3.5g fibre per 250ml, increased frequency of defaecation, softer consistency of stools, and a reduction in laxative use	**2+**	UK retrospective evaluation/cross-sectional study using questionnaire in children[31]
10	Placement of a gastrostomy tube allowed for fibre feeds to be given to EB adults	**2-/3**	UK qualitative study[37]
64	Enteral fibre feeds to children with mainly severe RDEB	**3**	UK evaluation of practice[38]
12	Enteral fibre feeds to RDEB adults	**3**	UK adult pilot studies[34]
223	Constipation was common in patients with dystrophic EB (DEB) requiring laxatives and in some cases fibre supplementation	**3**	Cohort of paediatric patients with dystrophic EB (DEB) retrospective case studies[18]

Key	* Articles where the sample population did not have EB; N/A: not applicable; signposts to an **existing** guidelines.
Notes	The appraisers agreed to remove reference 37 as it was considered bad practice
Rate	Descriptions in accordance to SIGN[1]
2++	High quality systematic reviews of case control or cohort studies; high quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
3	Non-analytic studies, e.g. case reports, case series
4	Expert opinion

Table 3b. Evidence for the use of artificial fibre and fluids as a specific treatment of constipation

Number of participants	Treatment of constipation and population group	Quality of evidence	Study and global location
Artificial fibre to attain adequate oral fibre intake if dietary intake is inadequate			
20	Constipation was treated successfully with liquid fibre-containing tube feeding formula in patients with EB	**2+**	UK Retrospective evaluation/Cross-sectional study using questionnaire in children28
	The indigestible matter in dietary fibre increases faecal bulk by promoting faecal water holding capacity and bacterial proliferation	**2+**	Guideline[11*]
Adequate fluid intake			
N/A	Increasing fluids intake was essential in helping with constipation	**2+**	Book chapter[27]
13	Enteral feeding helped meet fluid requirements in children with EB	**2+**	UK Retrospective evaluation /Cross-sectional study using questionnaire[34]
4	Increasing fluids intake was essential in helping with constipation	**3**	US case studies[4]
Offering polyethylene glycol in combination with fibre and fluids			
N/A	Optifibre®	**2+**	Clinical text book reporting current practice [33]
	Polyethylene glycol solution, lactulose, and sorbitol may be useful	**2+**	Guideline[11*]
4	The use of commercial fibre preparations provided constipation relief	**3**	USA case studies[4]
N/A	Use of polyethylene glycol	**4**	Expert opinion

Key	* Articles where the sample population did not have EB; N/A: not applicable; signposts to an **existing** guidelines.
Notes	The appraisers agreed to remove reference 37 as it was considered bad practice
Rate	Descriptions in accordance to SIGN[1]
2++	High quality systematic reviews of case control or cohort studies; high quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
3	Non-analytic studies, e.g. case reports, case series
4	Expert opinion

C

Recommendation

Fibre

Some research has been reported on the use of fibre as a treatment for constipation in EB. 20 children with EB who reported constipation were given oral nutrition supplement (ONS) drinks containing 3.5g fibre per 250ml drink[29], Table 3 (page 22). Subjects drank one to three drinks per day and reported increased frequency of defaecation, softer consistency of stools, and a reduction in laxative use[29]. All 20 children had been described by their parents as having "delay before defaecation" and 15 cried or screamed on defaecation[29]. After use of the fibre drink, three continued to delay and two continued to cry on defaecation. In a further study, 11 out of 13 children chronically constipated before placement of a gastrostomy tube were given fibre feeds and adequate fluid leading to resolution of the constipation[29]. Placement of a gastrostomy tube enables adequate fluid intake, administration of fibre rich feeds, and enhanced compliance with laxatives, all of which will help resolve constipation for many[34].

Fibre requirements should be calculated, Tables a, b (Annex III) and compared to the usual fibre intake and it should be gradually increased. Sudden increase in fibre intake may lead to gastro-intestinal side effects.

The source of fibre can be food (Annex I), oral nutrition supplements containing fibre[29], enteral feed containing fibre or fibre added to food, feed or drink e.g. Optifibre®, Tables 1 (page 10), 3 (page 22).

D

Recommendation

Laxatives

Upon diagnosis of constipation in children and adults, the first-line pharmaceutical treatment should be to introduce stool softeners (polyethylene glycol) before increasing fibre to assist in empting the bowels with minimal trauma. Polyethylene glycol is currently licensed for children over two years but may be appropriate for children with EB over the age of six months under appropriate medical supervision.

- Stool softeners, such as lactulose can be used to treat constipation in younger infants. Side effects may include excess gas[11].
- Osmotic or stimulant laxatives can be used in combination with polyethylene glycols to optimise bowel frequency if needed[11].
- Some people will require maintenance therapy in order to maintain a soft stool which is easy to pass[10]. Some people with EB, prefer a stool that is softer than normal, levels 4-5 on the Bristol Stool Chart to try and reduce peri-anal blistering.
- Fragility of the anal mucosa indicates that rectal washout, suppositories, and enemas could cause trauma so should be routinely avoided and used only under strict medical supervision.
- If faecal impaction is suspected due to symptomatic overflow diarrhoea or is shown on abdominal x-ray, supervised evacuation is indicated using a stool softener, as impaction can lead to nausea, vomiting, abdominal pain, and gastro-oesophageal reflux[33].

C

Recommendation

Meeting fluid requirements

Fluid requirements should be calculated according to age/body weight, Tables c, d (Annex III). Fluid is required to prepare polyethylene glycol. This will soften the stool and should be given in addition to the daily fluid requirement.

- As infants with EB may have higher energy requirements, increased volumes of breast milk or infant formula may be needed to support growth. If additional nutrients are not required, cooled boiled water may be offered in between milk feeds7.
- Professionals can work closely with school teachers and carers to enable children with EB to have regular access to oral fluids where needed.
- Professionals can advise on the timing of fluids to avoid patients feeling full at meal times.

D

Recommendation

Psychological support

Expert opinion

If fear of anticipated pain on passing a motion becomes a problem, the support of a clinical psychologist maybe helpful. This could include offering support to help make the changes that will treat constipation including scheduled toileting and support to establish a regular bowel habit, maintenance and discussion of a bowel diary, information on constipation, and use of encouragement and rewards systems to support change.

Constipation is a known problem in all types of EB but particularly in those with RDEB. By understanding the causes of constipation and both treatment and prevention strategies it is possible to reduce constipation and its painful side effects thus improving quality of life. Treatment is not expensive and can be tailored to suit the individual's food preferences and different types of food consumed around the world. It is hoped this document will impart knowledge to help people with EB prevent and treat constipation.

Future research

An evidence base is required to evaluate and advance current practice in the following areas in EB:

- for the effectiveness of polyethylene glycol
- for the effectiveness of fibre supplements, such as Hyfiber® and Optifibre®
- on drug treatments and strategies to reduce the pain of defaecation when the anus is blistered
- on drug treatments and strategies to optimise healing of the blistering of the anal area

Key limitations with rare disease

Although people living with EB were invited, only a small number could attend the second workshop; this may be due to distance and health issues that can arise in people with EB making attendance at workshops difficult. The draft was circulated by post and email in order to maximise feedback and comments from people living with EB.

Implementations, barriers and evaluation

DI aims to ensure that the EB CPG address the needs of patients internationally. The guidelines were presented at the international DEBRA Congresses. This guideline has supplementary material (annexes) which can be used as tools anywhere in the world by professionals as well as non-professionals. The implementation of these recommendations could be supported by DI to be monitored and evaluated through audits. The completion of a current practice audit, followed by the CPG pre-implementation survey (https://surveyhero.com/c/aabc0100) and post-implementation survey are highly recommended for best practice.

Updating the guideline

This guideline will aim to join a guideline group that has produced guidelines on constipation, and contribute to future updates by adding to previous versions of already consolidated CPG on a section that specifically addresses the issues of this condition in patients with EB. This may improve visibility, dissemination, and therefore adoption and implementation in clinical practice.

CPG development was led by two EB specialist dietitians and supported by a research dietitian. During the CPG development, two multidisciplinary working groups were assembled; the first for the preliminary scoping of the EB nutrition guidelines in 2015 and the second for feedback on the first draft of the guideline in 2016. These groups comprised of EB specialists for both adults and paediatrics (dietitians, nurses, and doctors).Three adults with EB and a parent of a child with EB were able to attend the second multidisciplinary working group and three other adults with EB provided feedback on the draft document. The guideline topics were chosen by the working group based on expert opinion, Figure 2. The final draft was reviewed by panel members and clinical nurse specialists in EB.

Figure 2. EB nutrition guideline topics in order of panel importance and most likely to benefit more EB patients around the world

Ranking	Topics
1	Constipation
2	Anaemia
3	Enteral and parenteral nutrition
4	Bone health
5	Factors affecting intake
6	Nutritional assessment and monitoring
7	Developmental stages
8	Specific nutrients-vitamins, minerals, and trace elements
9	Obesity
10	Infant
11	Breast feeding and weaning

Systematic literature searching, appraisal, and evidence rating

The literature search was conducted by a methodologist at inception on May 7, 2012. It was standardised, limited to English and Spanish languages, and ranged from 1970 to 2012. Extended searches were undertaken between 2012 and 2018. The electronic searching included MEDLINE, EMBASE, CINAHL, and Cochrane Central Register of Controlled Trials. Searches included studies, conference presentations, proceedings, and other unpublished documents meeting the selection criteria were also included. The search criteria were as follows: Epidermolysis Bullosa, constipation, soiling, encopresis, child, children, paediatric, adults, elderly, male, females, trace elements, micronutrients, vitamins, etc. The boolean AND and OR operators were used to combine these terms as appropriate. Citations for the evidence were reviewed by title and abstracts, papers were filtered and included for relevance regardless of study type. In addition, hand searches of journals, grey literature, and other guideline sources, such as NICE, and DEBRA's pain management and oral health care guidelines were carried out. One additional article[43] was added post reviewer recommendation as it has been published during the submission period.

GUIDELINE DEVELOPMENT METHOD

The SIGN[1] methodology was used to appraise and rate the evidence, Table 1bii. Each paper was rated by at least two panel members to compare consensus of rating. A summary of the evidence and the risk of bias in each paper was presented to the panel at a DEBRA member's day. In addition the panel reviewed the first draft of the guideline to ensure it addressed the concerns of adults and children with EB and the parents of children with EB. This meeting was used to agree the recommendations' strengths for a rare disease. The AGREE II tool was consulted to improve the guideline quality where possible.

SIGN grading system

Levels of Evidence			Grades of recommendation	
1	++	High-quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias	A	At least one meta-analysis, systematics review or RCT rated as 1++, and directly applicable to target population; or A body of evidence consisting principally of studies rated as 1+, directly applicable to target population, and demonstrating overall consistency of results
	+	Well-conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias		
	-	Meta-analyses, systematic reviews, or RCTs with a risk of bias		
2	++	High-quality systematic reviews of case controlmor cohort studies	B	A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overalll consistency of results; or Extrapolated evidence from studies rated as 2++
	+	High-quality case-control or cohort studies with a low risk of confounding or bias and a high probability that the relationship is casual		
	-	Case-control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is casual		
3		Non-analytic studies, e.g. case reports, case series	C	A body of evidence including studes rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; or Extrapolated evidence from studies rated as 2++
4		Expert opinions	D	A body of evidence rated level 3 or 4; or Extrapolated evidence from studies rated as 2+
			Good practice points	
			✓	Recommended best practice based on the clinical experience of the guideline development group

Source: SIGN 50 Guideline Developer's Handbook. NHS Scottish Intercollegiate Guidelines, Network 2014

ABBREVIATION

AGREE II	Appraisal of Guidelines for Research and Evaluation II
BMs	Bowel movements
CPG(s)	Clinical practice guideline(s)
EB	Epidermolysis bullosa
NICE	National Institute for Health and Care Excellence
NSP	Non-starch polysaccharides
ONS	Oral nutritional supplementary
PPI	Patient and Public involvement
PEG	Polyethylene glycol
SACN	Scientific Advisory Committee on Nutrition
SIGN	Scottish Intercollegiate Guidelines Network
UK	United Kingdom
US	United States

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Foods	Fibre / 100g	Adult Portion[40] (weight)	Fibre / portion
Almonds, six	12.5	13 g	1.6
Apple, cooked in water	2.4	85 g	2.04
Apple, peeled	1.6	65 g	1.06
Apricots, two	2	80 g	1.6
Apricots, dried, four	7.3	32 g	2.3
Baked beans in tomato sauce	3.7	80 g	2.96
Banana, peeled	1.1	80 g	0.88
Banana-based smoothie	9.2	200 ml	18.4
Barley, boiled	3.8	60 g	0.96
Beef chilli con carne without rice	1.1	155 g	1.71
Beetroot, one	1.7	35 g	0.6
Blackberries	5.3	140 g	7.42
Blueberries	1.5	100 g	1.5
Blueberry muffin	1.6	85 g	1.4
Bread, (slice - brown)	3.5	36 g	1.5
Bread, (slice - 50:50)	4.8	30 g	1.4
Bread, (slice - white)	3	35 g	1
Bread, (slice - wholemeal)	7	38 g	2.45
Bread, (pitta - white)	4.3	75 g	3.2
Bread, (pitta - wholemeal)	4.3	75 g	3.2
Broccoli, boiled	2.3	85 g	2
Brown rice and split peas	1.7	180 g	3.1
Brussel sprouts, boiled, nine	3.1	90 g	2.8
Buckwheat, boiled	2.7	100 g	2.7
Butternut squash. baked	1.4	65 g	0.91
Cabbage	2.5	60 g	1.5
Carrots, boiled	2.8	60 g	1.7
Cashew nuts	4.3	50 g	2.2
Cauliflower, boiled	2.3	60 g	1.4
Cherries, ten	0.9	40 g	0.4
Chia seeds	34.4	10g	3.4
Chick peas, boiled, 2-3 tablespoons	4.3	90 g	3.9
Chives	2.5	1 g	0.03
Courgette	1.2	60 g	0.72
Cranberries, dried	5.3	40 g	2.12
Currants	4.3	25 g	1.1
Dates, dried, one	1.8	15 g	0.3
Digestive biscuit, one	2.2	13 g	0.3
Falafel, three	3.5	85 g	2.98

Key: g = grams; ml = milliliter, * = this is an ingredient of foods, the portion size will depend on how much is used (examples: pasta, tortilla, gravy, etc.)

Foods	Fibre / 100g	Adult Portion[40] (weight)	Fibre / portion
Fig roll, one	4.2	14 g	0.6
Figs, raw, one	2.9	55 g	1.6
Figs, dried, one	9.8	20 g	2
Flaxseed	27.3	10 g	2.7
Flour, (chapatti - brown), one	10.3	42 g	4.33
Flour, (chapatti - white)	4.1	38 g	1.56
Flour - gram	9.1	*	*
Flour - maize (instant corn masa)	3.3	30 g	1
Flour - millett	3.5	*	*
Flour - rice	2.0	*	*
Flour - rye	11.7	*	*
Flour - soya (full fat)	11.2	*	*
Flour - wheat - brown	6.4	*	*
Flour - wheat - white plain	3.1	*	*
Flour - wheat - wholemeal	9.0	*	*
Fruit cake, one slice	1.7	70 g	1.2
Fruit yogurt	0.2	120 g	0.24
Grapes	1.2	100 g	1.2
Hazelnuts	9.7	25 g	2.4
Hobnob biscuit, one	3.5	14 g	0.5
Hummus	2.4	20 g	0.5
Kiwi, one	3	60 g	1.8
Kohlrabi raw	3.6	135 g	4.9
Kumquat raw, one	3.8	8 g	0.3
Lemon, peeled, one slice	2.8	20 g	0.56
Lentil soup	1.2	220 g	2.64
Lentils boiled	1.9	40 g	0.76
Malt loaf, one slice	3.5	35 g	1.2
Mango, one	2.6	150 g	3.9
Meat and vegetable stew	1	230 g	2.3
Melon (yellow)	0.7	200 g	1.4
Mushy peas	3.1	80 g	2.5
Nectarine, one	1.2	90 g	1
Oatcakes, one	9.6	10 g	0.96
Olives in brine drained	2.9	20 g	0.01
Onion, fried	3	50 g	1.5
Orange, one	1.2	160 g	3
Pakora	4.5	100 g	4.5
Paratha, plain	4	140 g	5.6

Key: g = grams; ml = milliliter, * = this is an ingredient of foods, the portion size will depend on how much is used (examples: pasta, tortilla, gravy, etc.)

Foods	Fibre / 100g	Adult Portion[40] (weight)	Fibre / portion
Parsley, dry, one teaspoon	3.3	1 g	0.03
Parsnips	4.7	60 g	2.8
Pasta, cooked (white - wheat)	2.6	230 g	5.98
Pasta, cooked (whole - wheat)	3.9	150 g	5.85
Pea and ham soup	1.4	220 g	3.08
Peaches, tinned/canned	0.8	120 g	0.96
Peanuts, ground	6.2	*	*
Peanut butter	6.6	12 g	0.8
Pear, raw, one	3.1	170 g	5.3
Pear, tinned/canned	1.4	135 g	1.9
Peas, cooked from frozen or fresh, four tablespoons	5.5	80 g	4.4
Pizza - vegetarian, baked from frozen, one slice	2.3	100 g	2.3
Plum	1.6	55 g	0.8
Pomegranate	4	40 g	1.6
Porridge (oat)	7.5	160 g	3.6
Potatoes boiled	1.6	175 g	2.8
Prunes tinned	2.4	135 g	3.2
Prunes, dried, one	5.7	8 g	0.5
Pumpkin seeds, roasted	18.4	25 g	4.6
Quinoa, cooked	2.8	40 g	1.12
Quorn	8.3	100 g	8.5
Quorn chilli without rice	2.4	150 g	3.6
Raisins, one tablespoon	2	30 g	0.6
Raspberries, two tablespoon	2.5	80 g	2
Re-fried beans	5.6	75 g	4.2
Ready Brek cereal	8.5	180 g	3.6
Red kidney beans, cooked, (canned/tinned), 1 tablespoon	6.5	60 g	3.9
Rice Basmati, plain, boiled	1.1	180 g	2
Rice Pilau, plain, cooked	0.3	100 g	0.3
Rice White, plain, boiled	0.1	100 g	0.1
Rice brown, plain, boiled	0.8	100 g	0.8
Risotto, plain, boiled	2.5	180 g	4.9
Samosa - vegetarian, one	1.8	40 g	0.8
Satsumas, one	1.5	70 g	1
Sharon fruit, one	1.6	110 g	1.8
Spinach	2.1	90 g	1.9
Squash, baked	2.8	60 g	1.68
Strawberries	3.8	100 g	3.8

Key: g = grams; ml = milliliter, * = this is an ingredient of foods, the portion size will depend on how much is used (examples: pasta, tortilla, gravy, etc.)

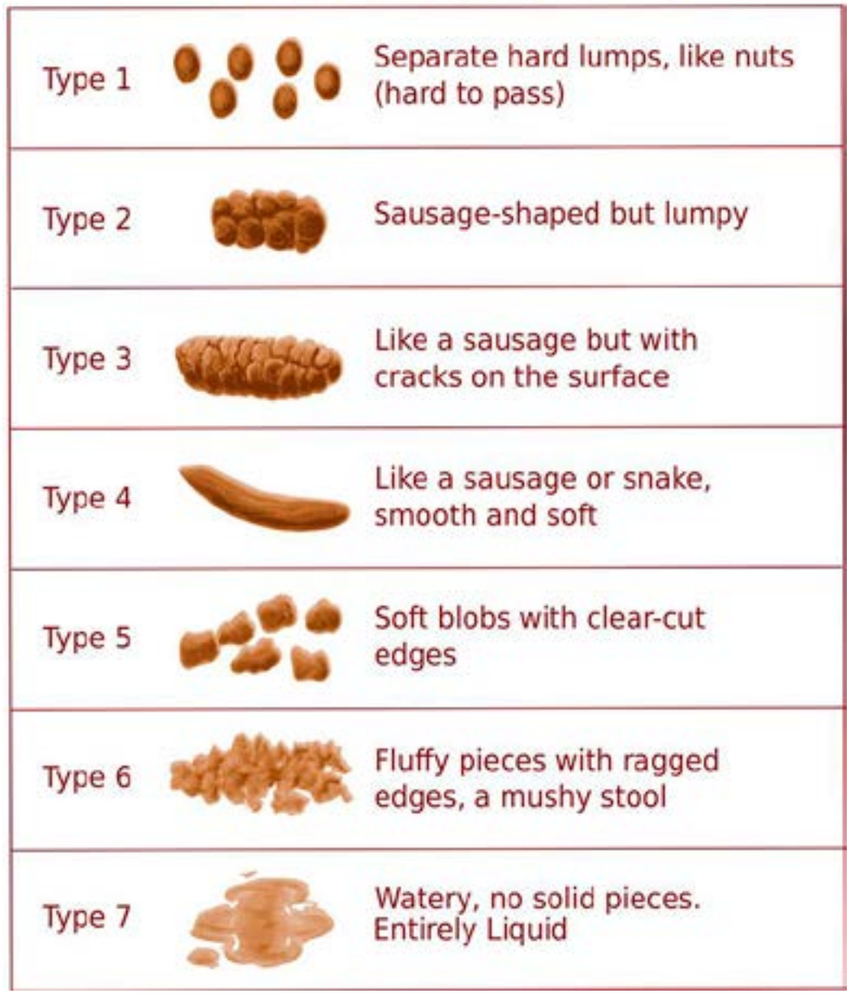
Foods	Fibre / 100g	Adult Portion[40] (weight)	Fibre / portion
Sunflower seeds	6.0	20 g	1.2
Swede, boiled	0.7	60 g	0.42
Sweet potato, cooked	2.4	130 g	3
Sweet-corn, cooked, (canned/tinned)	3.1	75 g	2.3
Tomatoes, raw	1	85 g	0.85
Turnip, boiled	1.9	60 g	1.14
Vegetable curry, 3-4 tablespoons	3.3	200 g	6.6
Walnuts, six	6.7	20 g	1.34
Weetabix, two biscuits	9.7	37.5 g	4
Wrap, (tortilla - maize)	3.3	30 g	1.39
Wrap, (tortilla - wheat - white and corn flour)	2.4	42 g	1.01
Wrap, (tortilla - wheat - white)	2.4	45 g	1.08
Wrap, (tortilla - wheat - wholemeal)	7.3	45 g	3.29
Yam, boiled	1.4	130 g	1.82

Key: g = grams; ml = milliliter, * = this is an ingredient of foods, the portion size will depend on how much is used (examples: pasta, tortilla, gravy, etc.)

How to work out the content of fibre in a portion

1. Weigh the food you are going to eat or assess from weight of package
2. From the table above, take the value of fibre (g) / divide by 100 and multiply by the portion eaten
3. Formula = Fibre (g) / 100g x Portion of food (g)

Bristol Stool Chart



1. How often in the last 14 days were your stools type 1 or type 2?
2. How often in the last 14 days were your stools type 3, type 4, or type 5?
3. How often in the last 14 days were your stools type 6 or type 7?
4. Currently, how often do you pass a bowel action? Please state number of times per: Weeks, e.g. once a week, Days e.g .every other day (2-3 days); Times per day e.g .4-6 times per day

Reference: Heaton, K W & Lewis, S J 1997, 'Stool form scale as a useful guide to intestinal transit time'. Scandinavian Journal of Gastroenterology, vol.32, no.9, pp.920 - 924

A. Fibre requirements in children

US	
> 2 years = age + 5g/day minimum	Range = age + 5-10g/day
UK	
Children 2-5 years Children 5-11 years Children 11-16 years Adolescents 16-18 years	15g/day 20g/day 25g/day 30g/day

B. Fibre requirements in adults

	US	UK[24]
Adults	> 20 years 20-35g/day	30g/day
Key: g: grams; : signposts to a guideline; US: Unites States; UK: United Kingdom		

C. Fluid requirements in children over one month[2, 42]

Weight	Millilitres per kilo of body weight (ml/kg)
Under or equal (£) 10kg	100ml/kg
10 to 20kg	100ml/kg+ 50ml/kg for each kg body weight over 10kg
Greater than > 20kg	100ml/kg + 50ml/kg for each 1kg 10-20kg + 20ml/kg for each 1kg body weight over 20kg

NB: seek specific medical advice in children who may have higher fluid requirements due to high losses of wound exudate.

D. Fluid requirements in adults[22]

Age	
< 65 years old with low intensity exercise (walking, no more than 30mins)	Weight in kg x 35ml
> 65 years old with low intensity exercise (walking, no more than 30mins)	Weight in kg x 30ml
Temperature	
Every degree centigrade once the temperature is over 37°C	+2 - 2.5ml per kg
Physical activity (exercise)[43]	
For intended exercise of more than one hour, it is recommended to weigh before and after (gym, dancing, football etc.)	+1500ml for every 1kg weight loss during exercise
NB seek specific medical advice in persons who do not attain adult weight (WHO growth charts)	

BOX 1

Disclaimer: For the following the above does not apply and you should consult a medical team

- In adults who have not reached adult weight.
- Increased fluid requirements will also be required in people with high losses of wound exudate

Title of Article	Author et al	Journal	Year
Epidermolysis bullosa: gastrointestinal manifestation	Orlando R et al	Annals of Internal Medicine	1974
Epidermolysis Bullosa: radiographic findings in 16 cases	Mauron M et al	American Journal of Radiology	1987
Gastrointestinal Manifestations of Epidermolysis Bullosa in Children	Gryboski J et al	Archives of Dermatology	1988
Gastrointestinal manifestations of Epidermolysis Bullosa: A study of 101 Patients	Ergun G et al	Medicine	1992
Oral and gastrointestinal manifestations of epidermolysis bullosa	Travis S et al	The Lancet	1992
Nutrition in dystrophic epidermolysis bullosa	Allman S et al	Paediatric Dermatology	1992
Constipation in Epidermolysis Bullosa: Successful treatment with liquid fibre-containing formula	Haynes L et al	Paediatric Dermatology	1997
The clinical spectrum of dystrophic epidermolysis bullosa	Horn & Tidman	British Journal of Dermatology	2002
Gastrointestinal manifestations of epidermolysis bullosa	Freeman E et al	Journal of Paediatric Gastroenterology and Nutrition	2006
Gastrointestinal Complications of Inherited Epidermolysis Bullosa: Cumulative Experience of the National Epidermolysis Bullosa Registry	Fine J et al	Journal of Paediatric Gastroenterology and Nutrition	2008
Nutritional support for children with epidermolysis bullosa	Haynes L.	Hintner H & Fine J-D eds. Life with Epidermolysis Bullosa (EB). Springer-Verlag.	2009
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CONTACT INFORMATION

DEBRA International

DEBRA International is the umbrella organisation for a worldwide network of national groups that work on behalf of those affected by the rare genetic skin blistering condition, epidermolysis bullosa (EB). The first DEBRA was created over 40 years ago and is present in over 50 countries around the world.

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EB Without Borders

EB Without Borders is a key initiative of DEBRA International. Its mission is to help patients, families, and doctors in countries where there is no DEBRA structure to support them, and to assist new groups to form and develop.

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EB-CLINET

EB-CLINET is the global clinical network of EB centres and experts.

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