



PREVENTATIVE NUTRITIONAL CARE GUIDELINE

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

An expert working group consensus



Funding

This document has been developed by DEBRA International and funded by DEBRA UK. The views or interests of DEBRA UK and DEBRA International have not influenced the final recommendations.

How to cite this document

Hubbard L and Jones R. Preventative nutritional care guideline: Constipation management for children and adults with epidermolysis bullosa. DEBRA International, 2019.

Conflict of Interest

The authors declared no potential conflicts of interest (CoI) with respect to the publication of this guideline. One working member declared a potential CoI with respect to the publication of this guideline as they were associate with DEBRA International (DI) as the guideline Coordinator. One referee declared they were an employee of DI. No one involved in the guideline has any connection to manufacturers.

Disclaimer

The information contained in this CPG does not indicate an exclusive course of action or serve as a standard of medical care. Variations, taking individual circumstances into account, may be appropriate. The authors, DEBRA UK, and DEBRA International accept no responsibility for any inaccuracies, information perceived as misleading, or the success of any recommendations, advice, or suggestions detailed in this CPG. The information provided on the following pages is subject to change without notice. For the most up-to-date information on available CPGs and contact information, please visit: www.debra-international.org

Copyright

© Copyright DEBRA International 2019. All rights reserved. This booklet may be copied and redistributed in any medium or format provided appropriate credit is given. This document or any portion thereof may not be remixed, transformed, or built upon without express written permission from DEBRA International.

Acknowledgements

We thank the working experts for giving their time and sharing their knowledge. We would like to thank Lesley Haynes, former principal dietitian at Great Ormond Street Hospital for her research contribution in addressing this problem.

Guideline development team

Lynne D Hubbard, Principal dietitian in adult EB, Department of Nutrition and Dietetics, St Thomas' Hospital, Westminster Bridge Road, London SE1 7EH, UK (Lead)

Corresponding Author
Lynne.Hubbard@gstt.nhs.uk

Rosie Jones, Specialist dietitian in paediatric EB, Department of Nutrition and Dietetics, Birmingham Children's Hospital, Steelhouse Lane, Birmingham, B4 6NH, UK (Co-lead)

People living with EB and public representatives

Working group

G. Jaega, person living with EB, UK
T. Guild, parent of person living with EB, UK

Review group

S. O'Sullivan, person living with EB, UK
C. Kapourani, person living with EB, UK
J. Hinchcliffe, person living with EB, UK
L. Prujean, EB Without Borders International Coordinator, DEBRA International, Austria

DEBRA International Production

Kattya M. Mayre-Chilton PhD RD, DEBRA International CPG Coordinator, Austria; Specialist Adult Research Dietitian, UK
Olivia Mullins - DEBRA International Business Manager, Austria
Michael Cardoza - Graphic Designer, UK

Experts

Lesley Haynes, Retired Principal dietitian in paediatric, EB Great Ormond Street Hospital, UK

Kattya Mayre-Chilton PhD RD, Research dietitian, St Thomas' Hospital, UK and DEBRA International CPG Coordinator, Austria

Natalie Yerlett RD, Specialist dietitian in paediatric EB Great Ormond Street Hospital, UK

Petra Kučan, Paediatric dietitian in EB, Croatia

Julio Villanueva Maffei, Professor Faculty of Dentistry, University of Chile and San Borja-Arriarán Clinic Hospital. Coordinator of the Cochrane Associate Group of the Faculty, (Methodologist) Chile

Elizabeth Pillay, Research nurse; Retired Clinical nurse Specialist (Adult Lead), St Thomas' Hospital, UK

Jane Clapham, EB Clinical Nurse Specialist (Adult lead), St Thomas' Hospital, UK

Pauline Graham-King, Retired EB Clinical nurse specialist (Adults), St Thomas' Hospital, UK

Dawn James, EB Clinical Nurse Specialist (Paediatrics), Birmingham Children's Hospital, UK

Carol Knowles, EB Clinical Nurse Specialist (Adults), Birmingham Children's Hospital, UK

Jacqui Finnigan, EB Clinical Nurse Specialist, North Island, New Zealand

Sharon Cassidy, EB Clinical Nurse Specialist, South Island, New Zealand

Kristina Soon, Psychologist paediatric EB specialist, Great Ormond street Hospital, UK



ABSTRACT..... 07

SUMMARY OF KEY RECOMMENDATIONS..... 08

INTRODUCTION..... 09

WHAT IS CONSTIPATION?..... 11

INCIDENCE OF CONSTIPATION IN EB..... 12

WHAT IS THE ROLE OF FIBRE?..... 15

THE CAUSES OF CONSTIPATION IN EB..... 17

PREVENTION OF CONSTIPATION..... 18

EVIDENCE TABLES..... 20

SUPPORTIVE TREATMENT..... 23

CONCLUSIONS & RESEARCH..... 26

GUIDELINE DEVELOPMENT METHOD..... 27

ABBREVIATIONS..... 29

REFERENCES..... 30

ANNEX I..... 33

ANNEX II..... 37

ANNEX III..... 38

ANNEX IV..... 39

CONTACT INFORMATION..... 40

NATIONAL DEBRA GROUPS..... 42

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.

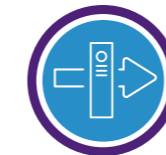
Key words

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 SIGN¹	
	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 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	
	4	Expert opinion	
biii)	✓	Recommended best practice based on the clinical experience of the guideline development group¹	
	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.	

Chronic constipation in which the stool is hard and causes pain and blistering on defaecation is a common feature of EB²⁻⁶ especially in RDEB²¹. 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 underestimated⁷. 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 EB⁸. 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 habits⁸.

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 constipation⁸. 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 feature⁹.

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 use¹⁹.



Guideline users

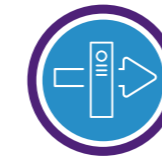
This CPG is intended to increase the knowledge of those involved in optimising nutritional status in EB. This includes patients, parents and carers, dietitians and nutritionists, dermatologists, paediatricians, gastroenterologists, nurses, dentists, surgeons, psychologists, speech and language therapists, 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 pass¹⁰⁻¹¹. 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 months¹⁰⁻¹²:

- ▶ Fewer than three bowel movements (BMs) per week for adults^{8, 10-12}
- ▶ Fewer than four BMs per week for children¹⁰⁻¹²
- ▶ Hard stool in more than 25% of BMs¹⁰⁻¹²
- ▶ A sense of incomplete evacuation in more than 25% of BMs¹⁰⁻¹²
- ▶ Excessive straining in more than 25% of BM¹⁰⁻¹²
- ▶ A need for digital manipulation to facilitate evacuation¹⁰⁻¹²

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 scale¹⁵ 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 patients¹⁶.

Figure 1. The Bristol Stool Chart Descriptor Scale¹⁵⁻¹⁶

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 function²³. Dietary fibre can be present in soluble and insoluble forms within a healthy diet²³. NSP-rich foods tend to be less energy dense and bulky, which in turn may induce greater satiety, reduce appetite²³⁻¹³ and promote peristalsis¹⁴. 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 load¹⁴
- ▶ Indigestible fibre has the capacity to retain water and soften stools helping colonic motility¹⁴.



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 benefit⁴³, but no clear guidance on type, amount, or duration exists. Therefore, normal fibre intake is advised for children with and without constipation⁹.

Artificial fibre

The beneficial effects of fibre on gut health and its moderating effect on constipation²³ 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 gum²⁸. Optifibre® is a virtually tasteless powder and its smooth consistency renders it barely detectable in foods and drinks; a vital factor in promoting acceptance²⁹. 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 constipation²⁹. 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 content⁷.
- ▶ 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 diet⁷.
- ▶ 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 this²⁰. The presence of perianal streptococcal infection may also contribute to discomfort⁸. Development of an anal fissure may result in avoidance of defaecation resulting in constipation²⁰.

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



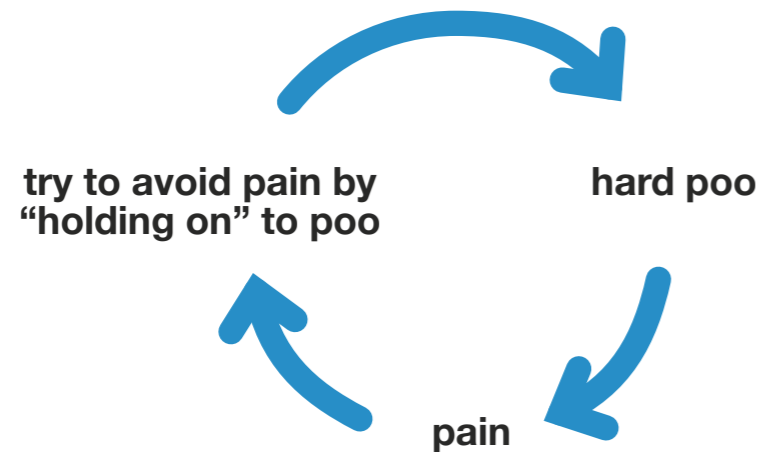
Medications for pain control including opiates and codeine reduce peristalsis²⁰. Oral iron preparations for treatment of anaemia and Amitriptyline[®] used to treat pain can also cause constipation.

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.



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).

B

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 present³³.
- ▶ 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 age³³, 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
 561/778	Dietary fibre in Childhood of the general population (non-EB)	2+	US survey in children ^{24*}
	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 children ²⁸
	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		

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 drink²⁹, 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 use²⁹. All 20 children had been described by their parents as having “delay before defaecation” and 15 cried or screamed on defaecation²⁹. 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 constipation²⁹. 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 many³⁴.

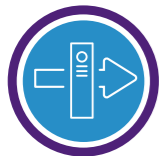
Fibre requirements should be calculated, Tables a, b (Annex III) and compared to the usual fibre intake and it should be gradually increased. Sudden increase in fibre intake may lead to gastro-intestinal side effects.

The source of fibre can be food (Annex I), oral nutrition supplements containing fibre²⁹, 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 emptying 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 gas¹¹.
- ▶ Osmotic or stimulant laxatives can be used in combination with polyethylene glycols to optimise bowel frequency if needed¹¹.
- ▶ Some people will require maintenance therapy in order to maintain a soft stool which is easy to pass¹⁰. 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 reflux³³.

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 may be 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 article⁴³ was added post reviewer recommendation as it has been published during the submission period.

The SIGN¹ 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, systematic 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 control or cohort studies	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 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 studies 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	<ul style="list-style-type: none"> ✓ 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

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



1. The Scottish Intercollegiate Guidelines Network (SIGN). 50 Guideline Developer's Handbook, NHS Revised Edition January 2008
2. Orlando RC, Bozymski EM, Briggaman RA, BREAM CA. Epidermolysis bullosa: gastrointestinal manifestations. *Ann Intern Med.* 1974;81:203-6
3. Mauron MA, Parker LA, Hartley WS, Renner JB, Mauron PM. Epidermolysis Bullosa: radiographic findings in 16 cases. *AJR.* 1987; 149: 925-7
4. Gryboski JD, Touloukian R, Campanella RA. Gastrointestinal manifestations of epidermolysis bullosa in children. *Arch dermatol.* 1988; 124: 746-52
5. Travis SPL, Turnbull AJ, Thompson RPH, Schofield OM, Chan O, O'Connor AF, et al. Oral and gastrointestinal manifestations of epidermolysis bullosa. *The Lancet.* 1992; 340: 1505-6
6. Ergun G, Lin A, Dannenberg A, Carter DM. Gastrointestinal manifestations of Epidermolysis Bullosa: A study of 101 Patients. *Medicine.* 1992; 71: 121-7
7. Haynes L. Nutrition for children with epidermolysis bullosa. *Dermatol Clin.* 2010; 28: 289-301.
8. National Institute for health and clinical Excellence (NICE). Constipation in children and young people: diagnosis and management. *Clinical guideline.* 2010; 99.
9. Tabbers MM, DiLorenzo C, Berger MY, Faure C, Langendam MW, Nurko S, et al. Evaluation and Treatment of Functional Constipation in Infants and Children: Evidence-Based Recommendations From ESPGHAN and NASPGHAN. *JPGN.* 2014;58: 258-74
10. Drossman D A. The Functional Gastrointestinal Disorders and the Rome III Process. *Gastroenterology* 2006;130:1377-90
11. Longstreth GF, Thompson WG, Chey WD, Houghton LA, Mearin F, Spiller RC. Functional Bowel Disorders. *Gastroenterology.* 2006;130:1480-91
12. World gastroenterology organisation practice guideline- constipation. 2016. <http://www.worldgastroenterology.org/guidelines/global-guidelines/constipation>. Accessed 22 April 2016.
13. Kranz S, Brauchla M, Slavin JL, Miller KB. What do we know about dietary fiber intake in children and health? The effects of fiber intake on constipation, obesity, and diabetes in children. *Adv Nutr.* 2012; 3: 47-53
14. Ip KS, Lee WT, Chan JS, Young BWY. A community-based study of the prevalence of constipation in young children and the role of dietary fibre. *Hong Kong Med J.* 2005;11: 431-6
15. Heaton K W, Lewis S J. Stool form scale as a useful guide to intestinal transit time. *Scand J Gastroenterol.* 1997; 32: 920-4.
16. Parés D, Comas M, Dorcaratto D, Araujo MI, Vial M, Bohle B, et al. Adaptation and validation of the Bristol scale stool form translated into the Spanish language among health professionals and patients. *Rev Esp Enferm Dig.* 2009; 101: 312-6
17. Horn HM, Tidman MJ. Quality of life in epidermolysis bullosa. *Clin Exp Dermatol.* 2002; 27: 707-10
18. Freeman E, Martinez AE, Mellerio J, Sebire NJ, Smith V, Lindley KJ, et al. Gastrointestinal manifestations of epidermolysis bullosa. *J Ped Gastroenterol Nutr.* 2008; 158:1308-14
19. Kim KY, Namgung R, Lee SM, Kim SC, Eun HS, Park MS, et al. Nutritional Outcomes in Children with Epidermolysis Bullosa: The Experiences of Two Centers in Korea. *Yonsei Med J.* 2014; 55: 264-9
20. Goldschneider KR, Good J, Harrop E, Lioffi C, Lynch-Jordan A, Martinez AE, et al. Pain care for patients with epidermolysis bullosa: best care practice guidelines. *BMC Med.* 2014, 12:178
21. Fine JD, Johnson L B, Weiner M, Suchindran C. Gastrointestinal Complications of Inherited Epidermolysis Bullosa: Cumulative Experience of the National Epidermolysis Bullosa Registry. *J Pediatr Gastroenterol Nutr.* 2008; 46:147-58

22. Yuen WY, Duipmans JC, Molenbuur B, Herpertz I, Mandema JM, Jonkman MF. Long-term follow-up of patients with Herlitz type junctional epidermolysis: *British Journal of Dermatology.* 2012; 167; 374-82
23. Department of health; RHSS. 41 Dietary reference values for foods energy and nutrients for the United Kingdom. In London: TSO 13th impression; 2004; p. 61-71
24. Public Health England. The Scientific Advisory Committee on Nutrition (SACN) recommendations on carbohydrates, including sugars and fibre: 2015. ISBN 9780117082847
25. Williams CL, Bollella M, Wynder EL. A New Recommendation for Dietary Fiber in Childhood. *Paediatr.* 1995; 96: 985
26. Braegger C, Decsi T, Dias JA, Hartman C, Kolacek B, Koletzko B, et al. Practical approach to Paediatric Enteral Nutrition: A comment by the ESPGHAN Committee on Nutrition. *JPGN.* 2010; 51: 110-22.
27. Haynes L. Epidermolysis bullosa. In: *Clinical Paediatric Dietetics* (Shaw V, Lawson M eds), 3rd edn. Oxford: Blackwell Science 2007; p. 482-96.
28. Haynes L. Nutritional support for children with epidermolysis bullosa In: Hintner H & Fine J-D eds. *Life with Epidermolysis Bullosa (EB)*. Springer-Verlag. 2009
29. Haynes L, Atherton D, Clayden G. Constipation in Epidermolysis Bullosa: Successful treatment with liquid fibre-containing formula. *Pediatr Dermatol.* 1997; 14: 393-6
30. Allman S, Haynes L, Mackinnon P, MacKinnon P, Atherton DJ. Nutrition in dystrophic epidermolysis bullosa. *Pediatr Dermatol* 1992; 9: 231-8.
31. Haynes L. Nutritional support for children with epidermolysis bullosa. *J Hum Nutr Diet.* 1998; 11:163-173.
32. Kramer SM, Serrano MC, Zillmann G, Gálvez P, Araya I, Yanine N, et al. Oral Health Care for Patients with Epidermolysis Bullosa – Best Clinical Practice Guidelines. *Int J Paediatr Dent.* 2012; 22 Suppl. 1: 1-35
33. Shaw V. *Clinical Paediatric Dietetics* 4th edition. Wiley-Blackwell; 2014. ISBN: 978-0-470-65998-4
34. Haynes L, Atherton D, Ade-Ajayi N, Wheeler R, Kiely EM. Gastrostomy and growth in dystrophic epidermolysis bullosa. *Br J Dermatol.* 1996; 134: 872-9.
35. Hubbard L D. Long-term outcomes in adults with recessive dystrophic epidermolysis bullosa fed by a gastrostomy tube in situ. *Int J Dermatol.* 2016; 55: 181-6.
36. Zidorio AP, Dutra ES, Leão DO, Costa IM. Nutritional aspects of children and adolescents with epidermolysis bullosa: literature review. *An Bras Dermatol.* 2015; 90: 217-23
37. Hubbard L, Haynes L, Sklar M, Martinez AE, Mellerio JE. The challenges of meeting nutritional requirements in children and adults with epidermolysis bullosa: proceedings of a multidisciplinary team study day. *Clin Exp Dermatol.* 2011; 36: 579-584
38. Hubbard L D, Mayre-Chilton K M. Quality of Life Among Adults With Epidermolysis Bullosa Living With a Gastrostomy Tube Since Childhood. *Qual Health Res.* 2015; 25:310-9.
39. McCance & Widdowson's. *The Composition of Foods* 5th edition. 1992 Published by the Royal Society of Chemistry
40. Food Standards Agency (FSA). *Food portion sizes* 3rd edition. 1988 Published by London: TSO
41. Paediatric Formulary Committee. *BNF for Children* (online) London: BMJ Group, Pharmaceutical Press, and RCPCH Publications. <http://www.medicinescomplete.com>. Accessed 11 Nov 2017
42. Griffin J. *Food for Sports, Eat well, Perform better.* 2008 Published by The Crowood Press Ltd.
43. Axelrod CH, Saps M. The Role of Fiber in the Treatment of Functional Gastrointestinal Disorders in Children. *Nutrients.* 2018 Nov 3;10(11).



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

Type 1		Separate hard lumps, like nuts (hard to pass)
Type 2		Sausage-shaped but lumpy
Type 3		Like a sausage but with cracks on the surface
Type 4		Like a sausage or snake, smooth and soft
Type 5		Soft blobs with clear-cut edges
Type 6		Fluffy pieces with ragged edges, a mushy stool
Type 7		Watery, no solid pieces. Entirely Liquid


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 **Rome Foundation approved DEBRA International Copyright and License Date 21/06/2018**

A. Fibre requirements in children

US	
> 2 years = age + 5g/day minimum	Range = age + 5-10g/day
UK	
Children 2-5 years	15g/day
Children 5-11 years	20g/day
Children 11-16 years	25g/day
Adolescents 16-18 years	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 adults²²

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)



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

BOX 1

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
Nutrition for children with epidermolysis bullosa	Haynes L.	Dermatologic Clinics	2010
The challenges of meeting nutritional requirements in children and adults with epidermolysis bullosa: proceedings of a multidisciplinary team study day	Hubbard L et al	Clinical and Experimental Dermatology	2011
Long-term follow-up of patients with Herlitz type junctional epidermolysis bullosa	Yuen W et al	British Journal of Dermatology	2012
Quality of Life Among Adults With Epidermolysis Bullosa Living With a Gastrostomy Tube Since Childhood	Hubbard & Mayre- Chilton	Qualitative health research	2014
Pain care for patients with epidermolysis bullosa: best care practice guidelines	Goldschneider K et al	BioMed Central Medicine	2014
Nutritional Outcomes in Children with Epidermolysis Bullosa: The Experiences of Two Centers in Korea	Kim Y et al	Yonsei Medical Journal	2014
Nutritional aspects of children and adolescents with epidermolysis bullosa: literature review	Zidorio C et al	Anais Brasileiros de Dermatologia	2015
Long-term outcomes in adults with recessive dystrophic epidermolysis bullosa fed by a gastrostomy tube in situ	Hubbard, L.D.	International Journal of Dermatology	2016

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.

www.debra-international.org
office@debra-international.org

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.

ebwb@debra-international.org

EB-CLINET

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

www.eb-clinet.org
office@eb-clinet.org

**This is how life feels
to people with EB.**



-  ▶ **Argentina**
www.debraargentina.org
info@debraargentina.org
+54 (011) 3965 4298
-  ▶ **Australia**
www.debra.org.au
admin@debra.org.au
+61 (0) 427 937 003
-  ▶ **Austria**
www.debra-austria.org
office@debra-austria.org
+43 1 876 40 30
-  ▶ **Belgium**
www.debra-belgium.org
info@debra-belgium.org
+32 (0) 4 267 54 86
-  ▶ **Bosnia & Herzegovina**
www.debra.ba
djeca.leptiri@hotmail.com
+387 33 328 284
-  ▶ **Brazil**
www.debrabrasil.com.br
web@debrabrasil.com.br
+55 (047) 3237 6243
-  ▶ **Bulgaria**
www.debrabg.net
contact@debrabg.net
+359 (0) 882 919 167
-  ▶ **Canada**
www.debracanada.org
debra@debracanada.org
+1 800 313 3012
-  ▶ **Chile**
www.debrachile.cl
info@debrachile.cl
+56 2 22 28 67 25
-  ▶ **China**
www.debra.org.cn
debra_china@163.com
+86 139 1850 3042
-  ▶ **Colombia**
www.debracolombia.org
info@debracolombia.org
+57 1 62 365 09
-  ▶ **Costa Rica**
www.debracr.org
info@debracr.org
+506 2244 3318
-  ▶ **Croatia**
www.debra.hr
info@debra-croatia.com
+385 1 4555 200
-  ▶ **Cuba**
debra_cuba@yahoo.com
-  ▶ **Czech Republic**
www.debra-cz.org
info@debra-cz.org
+420 532 234 318
-  ▶ **Finland**
www.allergia.fi/suomen-eb-yhdistys-ry
suomenebyhdistys.debrafinland@gmail.com
+358 9 473351
-  ▶ **France**
www.debra.fr
contact@debra.fr
+33 (0)7 78 95 83 44
-  ▶ **Germany**
www.ieb-debra.de
ieb@ieb-debra.de
+49 (0) 6461 9260887
-  ▶ **Hungary**
www.debra.hu
posta@debra.hu
+36 1 266 0465
-  ▶ **Ireland**
www.debraireland.org
info@debraireland.org
+353 1 412 6924
-  ▶ **Italy**
www.debraitalia.com
info@debraitalia.com
+39 02 3984 3633
-  ▶ **Italy (Südtirol - Alto Adige)**
www.debra.it
info@debra.it
+39 335 10 30 23 5

-  ▶ **Japan**
www.debra-japan.com
info@debra.sakura.ne.jp
+81 (0) 11 726 5170
-  ▶ **Malaysia**
www.debramalaysia.blogspot.com
debramalaysia@gmail.com
+60 12 391 3328
-  ▶ **Mexico**
www.debra.org.mx
debramexico@gmail.com
+81 8008 0352
-  ▶ **Netherlands**
www.debra.nl
voorzitter@debra.nl
+31 030 6569635
-  ▶ **New Zealand**
www.debra.org.nz
debra@debra.org.nz
+64 04 389 7316
-  ▶ **Norway**
www.debra.no
-  ▶ **Poland**
www.debra-kd.pl
sekretariat@debra-kd.pl
+48 501 239 031
-  ▶ **Romania**
www.minidebra.ro
contact@minidebra.ro
+40 0763 141 326
-  ▶ **Russia**
www.deti-bela.ru
info@deti-bela.ru
+7 (495) 410 48 88
-  ▶ **Serbia**
www.debra.org.rs
office@debra.org.rs
+381 (0)64 308 6620
-  ▶ **Singapore**
www.debrasingapore.com
debrasingapore@gmail.com
+65 9237 5823
-  ▶ **Slovakia**
www.debra-slovakia.org
debra.slovakia@gmail.com
+421 940 566 667
-  ▶ **Slovenia**
www.debra-slovenia.si
info@debra-slovenia.si
+386 31 348 303
-  ▶ **Spain**
www.debra.es
info@debra.es
+34 952 816 434
-  ▶ **Sweden**
www.ebforeningen.se
malin.ch.net@gmail.com
+46 (0)703 146 048
-  ▶ **Switzerland**
www.schmetterlingskinder.ch
debra@schmetterlingskinder.ch
+41 62 534 16 90
-  ▶ **Taiwan**
www.eb.org.tw
taiwaneb2010@gmail.com
+886 03 5734493
+886 0966 521836
-  ▶ **Ukraine**
www.debra-ukraine.org
info@debra-ukraine.org
svitdebra@gmail.com
+380 50 331 94 97
-  ▶ **United Kingdom (UK)**
www.debra.org.uk
debra@debra.org.uk
+44 (0)1344 771 961
-  ▶ **United States of America (USA)**
www.debra.org
staff@debra.org
+1 (212) 868 1573
+1 (855) CURE-4-EB

Get involved with DEBRA International

THE WORLD'S LEADING EB PATIENT ADVOCACY AND SUPPORT NETWORK

DONATION ACCOUNT IBAN AT65 6000 0005 1004 5254 BIC BAWAATWW

DEBRA INTERNATIONAL IS REGISTERED AS A CHARITY IN AUSTRIA (ZVR 932762489)



www.debra-international.org



office@debra-international.org



[@DEBRAInternational](https://www.facebook.com/DEBRAInternational)



[@InterDEBRA](https://twitter.com/InterDEBRA)

