



eb

CLINICAL PRACTICE GUIDELINES FOR NUTRITION SUPPORT

in Infants and Children with Epidermolysis Bullosa (EB)

including **THINC** (**T**ool to **H**elp **I**dentify
Nutritional **C**ompromise in EB)



Making the difference in paediatric care

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2007

Lesley Haynes RD
Principal Paediatric Dietitian for EB
Great Ormond Street Hospital for Children NHS Trust (GOSH)
Great Ormond Street, London WC1N 3JH, UK
HayneL@gosh.nhs.uk



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Reviewed and endorsed by:-

Vanessa Shaw, Head of Dietetics, GOSH
John Harper, Professor of Paediatric Dermatology, GOSH
Anna Martinez, Consultant Paediatrician, GOSH
Jemima Mellerio, Consultant Dermatologist, GOSH and Guy's & St Thomas' Hospitals



Foreword

Children with severe epidermolysis bullosa (EB) are one of the most nutritionally compromised group of patients that the paediatric dietitian will come across in their practice. The sequelae of these children's fragile epidermis all present a huge challenge: hypermetabolism due to nutrient and heat losses through their damaged skin, heightened by frequent infection; compromised immunity and wound healing; blistering throughout the gastrointestinal tract with resultant dysfunction, strictures and fissures; bone disease, anorexia, poor growth and general malaise.

The provision of nutritional support is becoming an integral part of the management of these children, largely due to the dedicated effort of Lesley Haynes. She has an extensive record of presenting and writing about the impact of nutritional support on children with EB and has gained an international reputation over some 20 years, sharing her knowledge, practice and passion for the optimum care of these children.

These guidelines are not written to replace the expertise of the dietitian or clinician, but rather to complement their existing practice and are based on the published evidence, where available, and the author's experience. They include an easy to use assessment tool, **THINC** (**T**ool to **H**elp **I**dentify **N**utritional **C**ompromise in EB). The complications affecting nutritional status in the different types of EB are well described together with the nutritional interventions needed with each type. The principles of nutritional support, dietary requirements and delivery of feeds and supplements are all presented with a clear statement and rationale and illustrated in algorithm format. The importance of monitoring, using anthropometric and biochemical parameters, and how to interpret these with respect to the underlying nutritional status and dietary intervention is also included. The contribution of the multidisciplinary team is highlighted and is necessary to formulate a comprehensive nutritional treatment plan suitable for the individual child.

With an extensive list of published literature and website resources to support the guidelines, this piece of work is an invaluable aid for those working with children with EB and their families.

Vanessa Shaw
Head of Dietetics
Great Ormond Street Hospital for Children NHS Trust



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Introduction

Relatively little has been published on the subject of nutrition support for children with epidermolysis bullosa (EB). However, provision of optimal nutrition is gaining recognition as an integral part of holistic, multi-disciplinary team (MDT) management and information on best practice is being increasingly sought.

In the current literature, specific guidelines for nutrient intakes are rarely documented, even in papers focusing on nutrition. The complications that compromise nutritional status along with abnormal biochemical, haematological and anthropometric data are comprehensively listed¹⁻⁷. Where nutrient targets are specified, they are based on extrapolation from nationally accepted government recommendations for healthy children and/or “best guesses” from professional experience⁶⁻⁹.

The following guidelines comprise the above data combined with current practice at Great Ormond Street Hospital for Children (GOSH), London, UK. The author is aware that, in their current form, the guidelines are incomplete and not robust. However, they are the best available at present. Constructive criticism, comments and contributions from colleagues working in the field of EB are welcomed. These will be incorporated into future revisions of the document and due credit will be given to contributors.

It is important to emphasise that the guidelines are intended to:-

- complement (not replace) the clinical expertise and judgement of the dietitian (or in the absence of a dietitian, the clinician responsible for the patient’s care)
- be used in conjunction with the referenced papers
- assist the professional to arrive at advice offered to, and agreed with the patient, in conjunction with patient preference and circumstances
- be a starting point for gathering of further evidence to enable best practice

What is EB?

EB is the collective term for a rare group of genetically determined skin blistering disorders. Its hallmark is extreme fragility of the skin and mucous membranes, with recurrent blister formation. Morbidity and mortality differ between, and within, the various types of EB. The cornerstones of management are control of infection, wound management, pain relief, promotion of optimal nutritional status, surgical intervention and provision of the best possible quality of life^{10,11}.



Figure 1 A six year old child with severe RDEB

EB is broadly classified into three main types¹⁰:-

- **Junctional (JEB)** – Herlitz (HJEB, formerly known as lethal), non-Herlitz (NHJEB, formerly known as non-lethal) and Junctional with pyloric atresia (PA)
- **Dystrophic (DEB)** – dominant (DDEB) and recessive (RDEB)
- **Simplex (EBS)** – Dowling-Meara, Weber-Cockayne

In common with clinical manifestations, the extent of nutritional compromise is determined by the type of EB. It tends to be mild in EB simplex, but can be extreme in junctional (JEB) and recessive dystrophic (RDEB) forms¹⁰.



Addressing nutritional support in EB

The 2 main factors responsible for compromised nutrition are:- ^{1,2,7-9}



① Oral, oro-pharyngeal, oesophageal and gastrointestinal complications (ulceration with or without stricture) which limit nutritional intake.

② Hyper-metabolism promoted by external skin lesions with loss of blood and serous fluid, leading to increased protein turnover and heat loss particularly when associated with infection.

Interactions between these and other factors (see **Figure 3**) mean that management is challenging and prioritisation of interventions can be very difficult. A holistic, multi-disciplinary team approach is highly desirable in such a group of conditions that impose enormous long term pressures on affected families^{1-3,6-10}.

Figure 2 Dressings need to be applied to open lesions to minimise infection and promote healing, and to protect vulnerable areas from injury.

Figure 3 Interactions between causes and effects of inadequate nutritional intake in severe EB

(Adapted from Shaw & Lawson 2007)

*Generally confined to RDEB

Dental / gum disease
Oral, pharyngeal and oesophageal blistering
Microstomia*, fixed tongue*
Dysphagia
Oesophageal stricture*
Gastro-oesophageal reflux (GOR)
Painful defaecation +/- constipation
Lower GI tract involvement
Anal fissures
Hand deformity*
PAIN

Reduced:-
• food intake
• mobility & weight-bearing
• sunlight exposure

Growth failure
Nutrient losses via blisters and wounds
Nutritional deficiencies
Compromised wound healing
Compromised immunity
Increased infection rates
Pubertal delay/failure
Osteoporosis/osteopenia

Anorexia
Apathy
MISERY

Aims of nutritional support (modify in cases of HJEB)

- To alleviate under nutrition and the stresses of feeding
- To minimise nutritional deficiencies
- To optimise growth
- To optimise bowel function
- To optimise immune status
- To optimise wound healing
- To promote pubertal development

Prognosis of different types of EB

In patients with severe generalised RDEB, the combination of persistent skin lesions, microstomia, abnormal dentition, oesophageal stricture, lower gastro-intestinal complications and severe hand deformities potentially leads to extreme nutritional deprivation^{1-4,6-12}. Life expectancy is variable and although death in childhood or teenage is not usual, chronic complications such as profound anaemia, septicaemia and dilated cardiomyopathy are commonly implicated. Metastatic squamous cell carcinoma is the most frequent cause of death in adulthood¹⁰.



Figure 4 The mouth of a child with severe RDEB. The degree of opening is limited, the tongue is fixed and there are heavy deposits of plaque and calculus with gingival inflammation, blistering and crusting.



Figure 5 The hand of a child with severe RDEB. The fingers have become encased in a cocoon of skin (pseudosyndactyly).



Figure 6 The hands of a child with Dowling-Meara EB simplex.

Patients with HJEB are unlikely to survive beyond the first or second year of life, and usually die from sepsis, laryngeal involvement and failure to thrive. Survival beyond the first 2 years is occasionally seen¹⁰.



Figure 7 The lower leg of a child with severe RDEB



Figure 8 The arm and trunk of a child with EBS Dowling-Meara

Gastrostomies are often placed in RDEB and NHJEB to forestall or address poor nutritional status^{3,6-10,12,13}. Conversely, in types of EB in which skin lesions gradually become confined mainly to feet and hands (with little or no oral involvement) such as in the Dowling-Meara EB simplex, patients often gain excess weight which exacerbates painful foot blistering and limits mobility⁹⁻¹⁰.



Figure 9 A child with severe RDEB before gastrostomy placement (left) and 6 months later (right).

Painful defaecation, with or without chronic constipation, exerts an extremely detrimental effect on appetite and quality of life. It is common in all types of EB, often bearing no relationship to the overall severity of skin disease. Frequently overlooked, the distress it causes can be significantly underestimated and under treated.^{1-3,6-10,12,13,14,15}

Osteopenia or osteoporosis occurs when reduced mobility leads to increasing wheelchair dependence, and is associated with significant bony pain¹⁶. Promotion of mobility is an important aspect of MDT care.

Gene therapy holds the best hope for this currently incurable set of conditions, and researchers predict that this could be available within the next 5 - 10 years for some EB types. Tables 1 and 2 list the complications of severe EB and the recommended nutritional interventions.

Assessment and regular review

Thorough assessment with regular review of nutritional intake and growth is essential. **THINC**, a **T**ool to **H**elp **I**dentify **N**utritional **C**ompromise is a comprehensive method of assessing the risk of actual or potential nutritional compromise in EB children under (Appendix 1) and over (Appendix 2) 18 months of age. **THINC** comprises a quick and simple scoring system that highlights current and specific nutrition-related problems and suggests appropriate action according to the nutritional compromise score. **Figure 11** suggests points to address when carrying out a comprehensive dietary assessment.



Figure 10 A child with severe RDEB

Figure 11 Suggested proforma for recording factors affecting nutritional intake and other relevant information (adapted from Shaw & Lawson 2007).

NAME:	DOB:	HOSPITAL NUMBER:	Date:
WEIGHT:		HEIGHT:	
kg	centile	cm	centile

CONSISTENCY OF FOOD:

Normal
 Soft
 Puree
 Fluid

REASON(S)

Oral blistering
 Microstomia
 Fixed tongue
 Dental caries
 G-O reflux
 Dysphagia
 Oesophageal stricture
 Excess mucus production
 Regurgitation

FREQUENCY OF DEFAECATION:

x day,
 x week
 Pain
 Bleeding p.r.
 Stool consistency

LAXATIVE(S), PREBIOTIC(S), PROBIOTIC(S) etc Preparation(s), dose, frequency

Stimulant laxative(s)	Stool softener(s)
Fibre	Prebiotic(s)
Probiotic(s)	Other

TYPICAL MEAL PATTERN

Breakfast:

Mid morning snack:

Lunch:

Afternoon snack:

Evening:

Bedtime snack:

Time taken over average meal: mins % eaten of food offered **GASTROSTOMY/other tube**

NUTRIENT-DENSE / ENERGY-DENSE SUPPLEMENT(S) / GASTROSTOMY FEEDS:

Name of preparation(s), dose, frequency

OTHER SUPPLEMENT(S): Name of preparation(s), dose, frequency

Iron	Zinc	Fluoride	Calcium +/- Vitamin D3
Carnitine	Selenium	Other vitamins	Other

APPROX. DAILY INTAKE:

Protein	g (/kg)	Normal DRV* =	g (/kg)
Energy	kcal (/kg)	Normal DRV* =	kcal (/kg)

COMMENTS / ACTION

* DRV = Dietary reference value for age and gender matched healthy child

Table 1 Main complications affecting nutritional status in different EB types

(Adapted from Shaw & Lawson 2007)

EB type	Complications affecting nutritional status
Weber-Cockayne EB simplex (EBS WC)	Lesions usually confined to feet and hands, especially in hot weather, often severely limiting mobility. Frequently painful defaecation with/without constipation.
Dowling-Meara EB simplex (EBS DM)	Generalised blistering tending later to become more confined to hands and feet. Feeding problems often severe in infancy, especially gastro-oesophageal reflux (GOR) but generally resolve before teenage. Often painful defaecation with/without constipation.
Herlitz junctional EB (HJEB)	Recurrent moderate to severe lesions. Dental pain due to abnormal tooth composition. Laryngeal and respiratory complications. Good initial weight gain usually followed by profound failure to thrive; possible protein-losing enteropathy. Opioid analgesia often exacerbates constipation. Massive sepsis and respiratory complications are usual causes of death. Survivors often profoundly anaemic with osteoporosis /osteopenia consequent to immobility and possibly to malabsorption.
Non-Herlitz junctional EB (NHJEB)	Recurrent mild to severe lesions. Dental pain due to abnormal tooth composition. Possible protein-losing enteropathy. Osteoporosis/osteopenia when immobility compromised.
Junctional EB with pyloric atresia (PA)	Mild to severe lesions. PA. Usually fatal in infancy, but there are exceptions.
Dominant dystrophic EB (DDEB)	Usually mild lesions. May have oral and oesophageal involvement. Anal erosions/fissures can cause painful and reluctant defaecation with/without constipation.
Recessive dystrophic EB (RDEB)	When severe, recurrent skin lesions heal poorly with generalised scarring and contractures. Internal contractures cause microstomia, dysphagia and oesophageal strictures. Digits fuse in severe generalized type. Anal erosions/fissures cause painful and reluctant defaecation with/without constipation. Some develop inflammatory bowel disease/colitis. Refractory anaemia. Osteoporosis/osteopenia in less mobile patients.



Table 2 Nutritional interventions associated with particular EB types

(Adapted from Shaw & Lawson 2007)

EB type	Nutritional interventions
Weber-Cockayne EB simplex (EBS WC)	Due to reduced mobility and activity, advice on weight maintenance/reduction may be required. Age-appropriate fibre (and fluid) intakes.
Dowling-Meara EB simplex (EBS DM)	As for RDEB (see below) in early years, but gastrostomy placement rarely necessary. Catch-up in weight often occurs around adolescence and excess weight gain leads to exacerbation of foot lesions and further reduction in activity and mobility. If so, advice on weight maintenance/reduction required. Age-appropriate fibre (and fluid) intakes.
Herlitz junctional EB (HJEB)	As for RDEB (see below) in terms of global supplementation, but with intention of improving quality of life rather than quantity. Intervention has no impact on prognosis. Unlike RDEB, gastrostomy placement not generally appropriate as may result in very poor healing around entry site, skin breakdown and leakage of gastric contents. Specialised formula feeds and exclusion diets have been used experimentally with patients with suspected protein-losing enteropathy.
Non-Herlitz junctional EB (NHJEB)	Global supplementation (as for RDEB) usually required except in mild cases. Specialised formula feeds and exclusion diets have been used experimentally with patients with suspected protein-losing enteropathy.
Dominant dystrophic EB (DDEB)	Intervention generally not indicated other than age-appropriate fibre (and fluid) intakes.
Recessive dystrophic EB (RDEB)	Global supplementation usually required except in mild cases. Oesophageal dilatation +/- gastrostomy feeding often indicated. Specialised formula feeds and exclusion diets have been used experimentally with patients with suspected inflammatory bowel disease/colitis.



What are nutritional requirements in EB?

This is one of the most frequently asked questions in EB management. It is also one of the most difficult to answer due to:-

- the complex, multi-system, inflammatory, infection-prone nature of the disease
- the variability of disease severity of patients even with the same EB sub-type
- the variability over time of individual patient's requirements as a reflection of age, extent of skin lesions, presence of infection, need for catch-up growth etc
- the difficulties associated with estimating desirable weight gain when height is compromised as a result of pain, joint contractures and osteoporosis (see Optimising growth below).
- the difficulties associated with conducting clinical trials in such small patient numbers

Periods of rapid development (infancy, childhood and adolescence) are the most challenging times. While their unaffected peers channel nutrition primarily into growth, those with the more severe EB types face additional demands placed by a wound healing process that strives to function despite inherent defects in the structure of the skin.

Extrapolations and best guesses from work with pressure ulcers and thermal burns form the basis of current practice. However, a rigorous evidence base is lacking in trials involving patients with normal skin^{17,18} As with burns, nutrient requirements in EB probably reflect the severity of lesions^{1,19} However a burn, albeit severe or extensive, is a distinct episode whilst EB is a life long condition and the megadoses of some nutrients recommended for burned patients are very likely to be inappropriate, or even inadvisable, for long term administration¹⁷. In terms of energy requirements, some children become overweight if fed overly-enhanced energy intakes long term.

Energy requirements can be estimated using a calculation based on weight-for-height age and taking into account additional factors, namely degree of blistering, sepsis and requirement for catch-up growth⁶. This method provides a working figure, but the scoring of skin involvement is subjective and the formula is somewhat complex:-

For example:-

Weight (kg) x (kcal/kg for height age) x [1 + (sum of 3 additional factors)]

Additional factors =

- 1. Ratio of blisters to body surface area (BSA):**
20% BSA = 0.19, 40% BSA = 0.5, 100% BSA = 0.95
- 2. Sepsis: mild = 0.2, moderate = 0.4, severe = 0.8**
- 3. Catch-up growth: 0.1 – 0.2**

Example:- 6 year old boy, weight 13kg, height age (25th centile) = 4.7 years
20% BSA blistered, mild sepsis, stunted

Apply formula:- 13 x 90 x [1 + 0.19 + 0.2 + 0.2] = 1860 kcal = 143 kcal/kg



Using a simpler method, based on chronological age and UK Dietary Reference Values²⁰, increases in weight can be achieved by providing 100–150% estimated average requirement (EAR) for energy. Using the above example to estimate energy requirement:-

**100 – 150% of energy requirement for chronological age = 1810 – 2715 kcal/day
= 139 – 209 kcal/kg/day (average 174 kcal/kg).**

Even the lower end of this range is likely to provide a significantly higher intake than that to which the child has been accustomed, so it is advisable to start here and monitor weight gain, increasing kcal/kg until consistent weight gain is achieved.

Protein requirements should be based on 115–200% the UK reference nutrient intake (RNI) for protein, using chronological age. Using the above example:-

22.7 – 39.4g protein/day = 1.7 – 3g/kg/day. Children with extensive / infected lesions will require an intake at the higher end of the range.

Individualised supplementation, oral or via feeding gastrostomy, can undoubtedly improve nutritional status, however, few children report associated improvements in wound healing rates. This is disappointing but predictable, considering the intrinsic flaws in EB skin and many severely affected children endure long term, poorly healing or unhealed lesions that pose a continual infection risk and cause major discomfort and disability.

In spite of the above, it does not follow that nutrition has no impact on the complex events surrounding attempted tissue repair and efforts should always be made to optimise nutritional intake if deficiency or imbalance is suspected. Promotion of wound healing is not the only reason for enhancing nutrition; optimisation of immunity and quality of life are equally important aims. The role of enhanced intakes of single micronutrients e.g. zinc, vitamin C, is unclear even in those with normal skin, and more work is required to identify both the clinical conditions and the doses of individual micronutrients that actively promote or accelerate healing^{17,18}. “Immune enhanced” formulas containing nutrients such as arginine, glutamine and essential fatty acids are marketed for adults as promoting healing, optimising immune status and exerting a beneficial effect on inflammatory conditions²¹. These would be highly advantageous in EB, but as yet, their efficacy has not been tested. Even if this data were available, assessment of wound healing in EB is highly subjective and fraught with confounding factors most significantly infection and anaemia, and clinical trials are required to monitor this more objectively.

Iron and zinc supplements have the potential to interact, possibly leading to reduced absorption of both. However, although some trials have shown that joint supplementation has a smaller effect on biochemical or functional outcomes than does supplementation with either mineral alone, there is no strong evidence to discourage joint supplementation. Whether given separately or together, it is firstly important to establish the level of compliance. Then, if the scheme proves impractical or the physiological response is poor, the regimen can be modified^{8,9}.



Optimising growth in EB

Determination of optimal growth rates for children with severe EB is difficult. Fox et al²² concluded that RDEB children are of significantly lower birth weight than unaffected children, and that the compromise in growth throughout life in RDEB appears to begin in utero. Plotting weight and length or height on nationally approved growth charts is useful in assessment of dietary interventions, but should always be considered in the context of individual disease severity. Pain, fixed flexion contractures around joints and osteoporosis all lead to underestimated or inaccurate height measurements^{1,3,7-9} and more accurate measurement may be possible using a supine stadiometer or in some cases, by segmental measurements^{9,23}. The traditional 'rule of thumb' that disparity between weight and height should not be greater than two major centiles is generally applicable.

Persistent dysphagia and painful defaecation (with or without constipation) are two major reasons for poor weight gain in severe EB^{1-10,12-15} (➤ **Figure 3**). Dilatation of the oesophageal strictures should relieve dysphagia, however, the pharyngeal and oesophageal mucosae are extremely fragile and may rupture. In the past, even in experienced hands, fatalities have occurred when a gastroscope has been used²⁴. Recent evidence shows that, with fluoroscopically guided endoluminal serial balloon dilatations, patients with high oesophageal strictures can gain significant benefit²⁵.

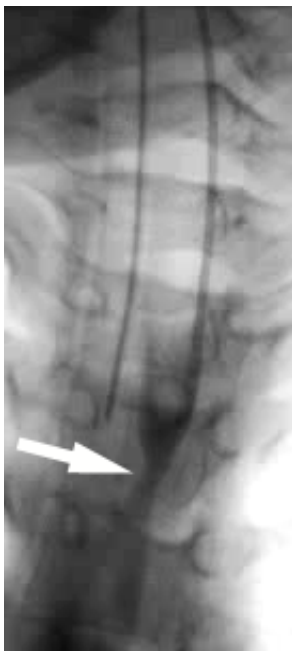


Figure 12

Left A tight stricture (2mm) typically located in the thoracic oesophagus in severe RDEB.

Right The dilated stricture. An 18mm balloon has been inflated



Chronic constipation (often more accurately termed faecal loading) with painful defaecation is one of the most frequent, yet underestimated, complications of all types of EB. Even a moderately bulky stool can tear delicate anal skin causing fissuring and extreme pain with subsequent bowel movements. Fear of defaecation leads to infrequent and incomplete bowel emptying and the gradual accumulation of hard faeces. It should be treated without delay if the vicious cycle of pain, conscious ignoring of the gastrocolonic reflex and secondary anorexia is to be avoided. Management involves adequate fluid and age appropriate fibre intakes and appropriate stool softeners/laxatives^{1-3,6-10,12-15}.

Nutrition support by gastrostomy feeding has definite advantages in EB^{3,6-10,12,13}, but weight gain may be associated with central fat deposition and poor linear growth, causing disproportionate body shape. Reasons for this are probably multi-factorial and inter related and as likely to be due to disturbances in growth hormone production mediated by cytokines and increased cortisol production inherent in chronic inflammatory illness²⁶ as to gastrostomy feeding per se. This seems to be more frequent in more severely affected children (in whom the inflammatory process is great) and whose oral intake is minimal relying almost entirely on gastrostomy feeds to supply their requirements.

The impact of nutrition on mobility in EB

When weight centile deviates upwardly by more than 2 centiles from the height centile, the EB child may be less mobile and more wheelchair dependent. However, maintenance of a balance between mobility, growth and nutritional status is vital, as these 3 aspects are inter-related and inter-dependant. Lack of weight bearing exercise and significant wheelchair dependency compounds the low bone mass often seen in severe EB¹⁶. Increased bone pain and fractures lead to further reliance on a wheelchair. Conversely, children who remain abnormally light may fail to attain puberty and to benefit from its associated protective hormonal effect on bone health. Work is on going to investigate best practice in this area.

Figures 13 & 14 A boy with severe RDEB during a session of physiotherapy



Table 3

Principles of nutrition support in EB

Statement	Rationale
<p>“Severe, recalcitrant nutritional deprivation unparalleled in all of clinical medicine” has described the most complex and serious forms of EB¹.</p>	<p>To be aware that, although every realistic effort should be made to improve nutritional status, beneficial effects may be limited due to the multi-system nature of the disease, particularly when EB type is severe. Goals should be adjusted accordingly.</p>
<p>An understanding of the aetiology and prognosis of different types of EB is highly desirable. It is equally important to appreciate the dynamics of the affected child’s family and their expectations of management and interventions.</p>	<p>To provide appropriate and realistic advice, particularly when life expectancy is short. To modify nutritional interventions, expectations and counsel according to the patient’s individual situation.</p>
<p>Nutrition support should be undertaken as an integral element of a multi-disciplinary team skilled in the holistic management of EB.</p>	<p>To minimise the antagonistic effect of factors beyond the dietitian’s control (e.g. gastro-oesophageal reflux, constipation, poor skin care).</p>
<p>Optimal nutrition support depends on active and regular nutritional assessment and intervention, ideally beginning as soon as possible after birth or as soon as a diagnosis of EB is made. Nutritional evaluations should be undertaken by a registered dietitian. Frequency of evaluation depends on disease severity and age of patient.</p>	<p>To identify likely nutrient deficiency/excess promptly in order to promote optimal immune status and growth, maximize wound healing and last, but not least, to promote optimal quality of life for child and family alike.</p>
<p>Appropriate strategies (compatible with family dynamics) should be recommended in order to address dietary deficit. Realistic time frames should be placed on interventions.</p>	<p>To minimise feelings of guilt and failure by carers and/or non-compliance by imposing impractical or unrealistic recommendations.</p>



Table 4

Details of nutrition support in EB (continues on pages 19 & 20)

➤ Refer to algorithms in Figures 15 and 16

Statement	Rationale
Growth retardation may begin in utero	To modify interventions and expectations accordingly.
Breast feeding is possible, and provided that it permits acceptable weight gain, should be encouraged.	To promote bonding, immunity and desirable gut flora.
Breast milk alone, in all but mild cases, usually fails to satisfy increased requirements, demonstrated by failure to gain weight satisfactorily. If so, measures should be taken to provide a more nutrient-dense intake.	To promote age-appropriate increases in weight, head circumference and length.
Sucking from a conventional teat is often painful and causes trauma to the oral mucosa. Suckling from the breast may induce facial lesions. A specialised teat should be tried.	To provide satisfactory milk flow with even a weak suck, using a specialised teat e.g. Haberman (Athrodax International). To minimise trauma to the gum margin and control feed delivery via the internal valve.
Naso-gastric tube placement is best avoided unless absolutely necessary.	To avoid:- <ul style="list-style-type: none"> nasal, oro-pharyngeal and oesophageal trauma interfering with oral feeding and possibly leading to later food aversion and an increased tendency to develop strictures difficulties in securing (use only non-adhesive materials or silicone tape recommended for fragile skin)
Weight, length/height (and, in infants, head circumference) should be measured and plotted at regular intervals. Supine length or segmental measurements are more practical when pain or contractures prevent accurate height measurement.	To identify promptly periods of faltering growth, and to suggest appropriate interventions
Energy requirements seem to range from 100 – 150% EAR for age, and protein requirements from 115 – 200% RNI. The greater the requirement for catch-up growth and/or area of denuded/infected skin, the greater the energy and protein requirements will be.	To promote optimal immunity, growth and wound healing .
Vitamin and mineral deficiencies are frequent in severe forms of EB and regular nutritional evaluation with laboratory analysis of blood should be undertaken.	To minimise the potential for deficiency and to promote proactive and appropriate supplementation.
Requirements for vitamins are assumed to be increased in severe cases. Provision of 150-200% of the RNI ensures intakes are adequate and within recommended safe limits.	To compensate for losses/poor absorption via the gut, to promote optimal immunity, growth and wound healing. To avoid toxicity from excessively high intakes.

Table 4

Details of nutrition support in EB (continued)

➤ Refer to algorithms in Figures 15 and 16

Statement	Rationale
<p>Requirements for certain nutrients e.g. iron, zinc and selenium and carnitine are based on amounts calculated to address deficiency as demonstrated biochemically and haematologically.</p>	<p>To promote optimal immunity, growth and wound healing. With selenium and carnitine particularly, to reduce the potential for development of dilated cardiomyopathy.</p>
<p>Requirements for all other micronutrients are assumed to be increased. Provision of 150-200% of the RNI ensures intakes are adequate and within recommended safe limits.</p>	<p>To promote optimal immunity, growth and wound healing. To compensate for losses via the gut/poor absorption and to avoid toxicity from excessively high intakes.</p>
<p>Megadoses of vitamins and/or minerals are not routinely recommended.</p>	<p>There is no evidence that megadoses are beneficial; they may even be associated with impairment of the immune response.</p>
<p>Optimal timing of mineral supplements is not agreed. A regimen best suited to the patient and adjusted in the light of laboratory results should be adopted and regularly reviewed.</p>	<p>To optimise compliance and absorption.</p>
<p>Liquid preparations of supplements should be used whenever possible.</p>	<p>To facilitate swallowing in dysphagic patients. To permit administration via feeding tube.</p>
<p>Interpretation of blood test results is complex due to the inflammatory nature of the condition. Nevertheless, it is important to monitor certain parameters regularly.</p>	<p>To assess as well as possible, the impact of intervention, and to modify management accordingly. ➤ Table 5 suggests parameters to be monitored and frequencies.</p>
<p>Blood test results should be interpreted in conjunction with information such as:- extent of lesions, presence of infection, growth rate, gastro-intestinal complications and comparison of nutritional intakes with age appropriate recommended nutrient intakes.</p>	<p>To avoid inappropriate (ineffective) intervention e.g. supplementary zinc when plasma zinc level is spuriously low due to low plasma albumin, supplementary iron when constipation is present without appropriate modification of laxative regimen.</p>
<p>Gastro-oesophageal reflux (GOR) is common in all types of EB and should be managed proactively with medical management using anti-reflux agents and feeds may need to be thickened.</p>	<p>To minimise pain, mucosal erosion by gastric acid (leading to structuring and possible oesophageal foreshortening) and subsequent reluctance to feed. To minimise risk of aspiration of feed/food.</p>

Table 4

Details of nutrition support in EB (continued)

➤ Refer to algorithms in Figures 15 and 16

Statement	Rationale
Oesophageal strictures may be relieved by serial balloon dilatations.	To reduce dysphagia, maximise oral intake and its enjoyment, and to improve social aspects of eating.
Gastrostomy placement should be considered if significant improvement in nutritional intake is not achieved with oral supplementation.	To reduce parental anxieties surrounding feeding, improve nutritional intake and provide a route for unpalatable medications.
Painful defaecation (with or without constipation) is common in all types of EB. It requires proactive medical management with stool softeners/laxatives and feeds with an age-appropriate source of fibre.	To promote a normal bowel pattern and minimise negative associations of feeding with anal pain that leads to reluctance to feed.
Fibre requirements are assumed to be normal. Supplements should be age appropriate.	To promote optimal gut integrity and function.
Prebiotics are assumed to be beneficial.	To promote optimal gut integrity and function.
Calcium and vitamin D intake should be monitored in association with dual X-ray absorptiometry (DEXA scan), biochemical estimation of calcium and vitamin D status and administration of bisphosphonates with a combined calcium and vitamin D preparation.	To minimise the likelihood of low bone mass, abnormal bone mineralisation and fractures.

Table 5

Biochemical and haematological investigations in EB children; suggested investigations and sampling frequencies

(Adapted from Shaw & Lawson 2007)

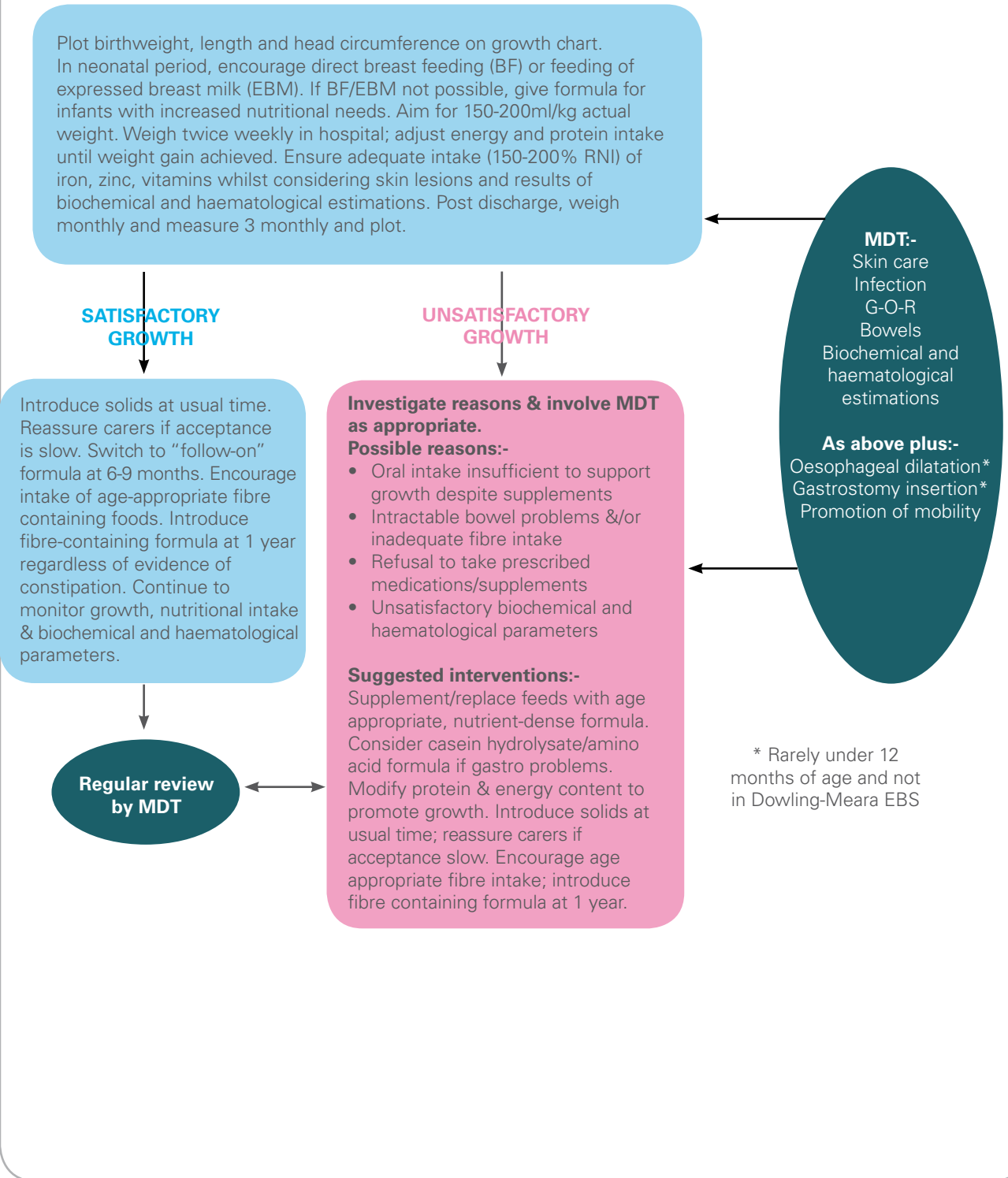
6 – 12 monthly	Yearly	1 – 2 yearly
Urea and electrolytes	Vitamin B1	Vitamin E
Creatinine	Carnitine	
Calcium, phosphate (+/- Vitamin D3)	Vitamin B12	
Total protein, albumin	Folate	
Alkaline phosphatase		
Zinc, selenium		
Serum iron, ferritin, full blood count		
Hypochromic red blood cell		
Transferrin receptors		
Mean corpuscular volume (MCV)		
Reticulocytes, red cell folate		
Erythrocyte sedimentation rate (ESR)		
Free erythrocyte protoporphyrin (FEP)		

N.B. The above is a guide. Sampling frequency depends on individual disease severity and the need to evaluate intervention.

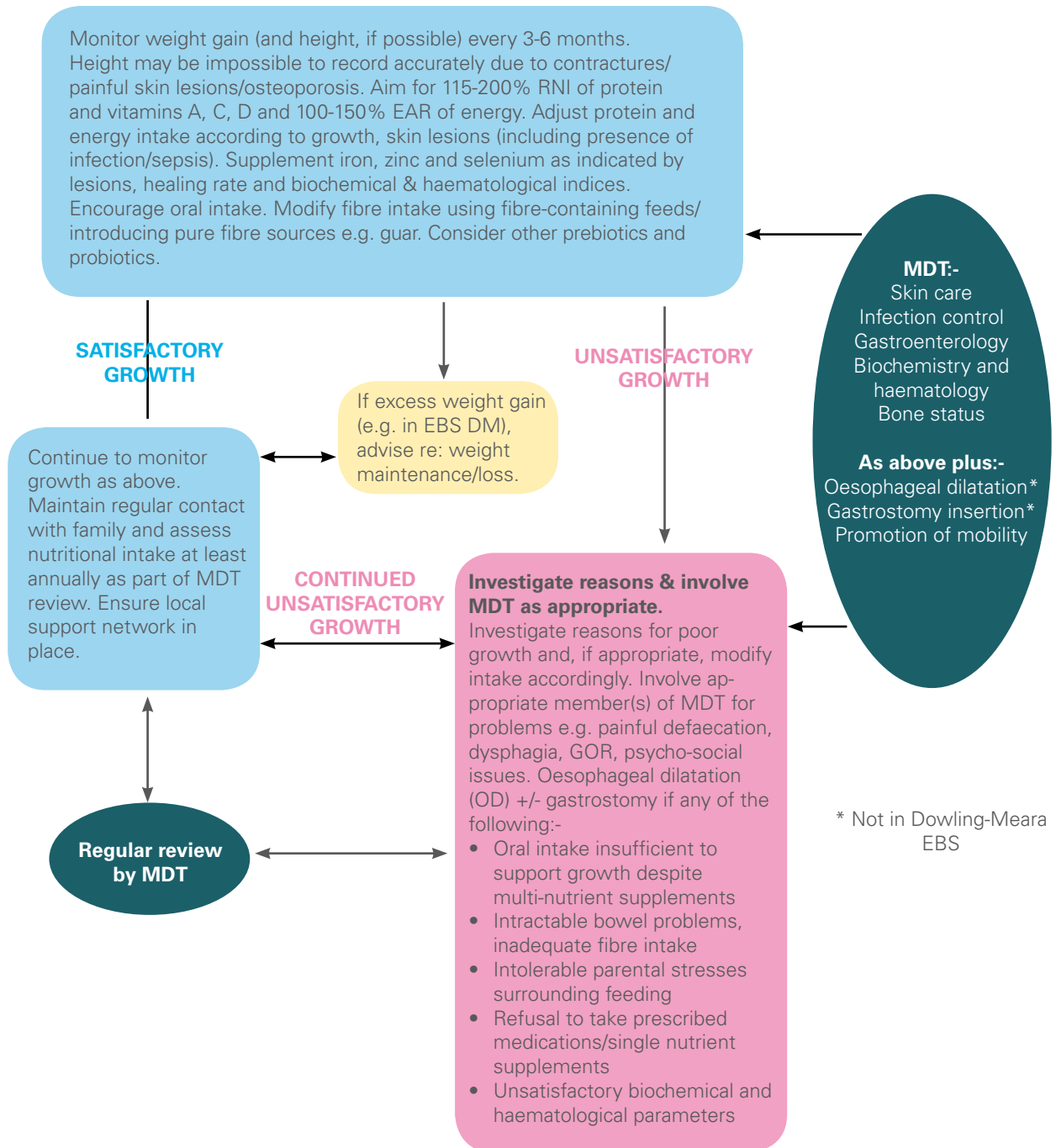


Figure 15

Algorithm for nutrition support in **infants (under 18 months of age)** with RDEB, non-NHJEB and Dowling-Meara EBS



Algorithm for nutrition support in **children** (18 months of age and **over**) with RDEB, non-NHJEB and Dowling-Meara EBS



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Resources

Nutrition information booklets published by DebRA:

Haynes, L. Nutrition in epidermolysis bullosa, for children over 1 year.

Haynes, L. Nutrition for babies with epidermolysis bullosa.

(This information may be suitable for babies with EBS Dowling-Meara and non-Herlitz JEB, but not Herlitz JEB.)

Dystrophic Epidermolysis Bullosa Research Association (DebRA)

DebRA House, 13 Wellington Business Park, Dukes Ride, Crowthorne, Berkshire RG45 6LS.
Tel. 01344 771961, Fax 01344 762661, e-mail debra@debra.org.uk

DebRA is a charity that exists to help people with all types of EB. DebRA also funds research into all aspects of EB care as well as dedicated nursing and social support.

Fora, websites

- www.internationalebforum.org
(international multi-disciplinary professionals' forum) This includes the Nutrition Forum
- www.debra.org.uk (DebRA UK)
- www.debra.org (DebRA of America)
- www.ebanusa.org (US EB Action Network)
- www.ebkids.org (US EB research foundation)
- www.ebinfo.org (Compiled by the US parent of an EB child)



Appendix 1

“**THINC**” for Children with Epidermolysis Bullosa under 18 months of age

THINC about nutrition

This is a ...

Tool to
Help
Identify
Nutritional
Compromise

in epidermolysis bullosa (EB)
for use by professionals

with children under 18 months of age
with recessive dystrophic EB, non-Herlitz
junctional EB and Dowling-Meara EB simplex

by Lesley Haynes RD
Principal Paediatric Dietitian
Great Ormond Street Hospital
for Children NHS Trust
London, UK

Guidelines on the use of **THINC** with children under 18 months of age

The significant nutritional compromise, growth failure and reduced quality of life of severe EB can be improved by measures that minimise the impact of the condition's complications.

THINC's aim is to highlight the child at actual or potential risk of nutritional compromise/ deficiency, so that a proactive treatment plan can be drawn up which minimises the impact of the disease and supports the family in this process.

THINC is intended to be used by a dietitian (or in his/her absence, the health professional supervising the child's medical/surgical care), in association with other appropriate professionals. The tool should aid, not replace, clinical judgment. It should be used alongside Clinical Practice Guidelines (and cited references) which include algorithms of suggested courses of action.

THINC's scoring chart is intentionally brief and the measures are practical to simplify the completion process. Scores for some aspects are unavoidably subjective, however they have been weighted to allow for this. To maximise the accuracy of the final score, and to provide a useful and meaningful result, please take time to read the following notes. They explain the relevance of the particular aspect rated and offer guidance regarding the questions to ask in order to elicit the most authentic picture of the child's current condition and the potential for development of problems that affect nutritional status. The higher the score, the greater is the likelihood of nutritional compromise. The maximum possible total score is 100.

THINC's scoring chart rates 3 main aspects of the child's state :-

- Weight and length
- Gastroenterology
- Dermatology

Weight and length

Weight, in particular, and length are amongst the best gauges of nutritional adequacy.

Weight nude or in a dry nappy and plot on an ethnically appropriate growth chart (where available). If such a chart is unavailable, use a nationally accepted growth chart, e.g. in UK use Four-in-One (Duodecimal) Growth Charts (Birth - 20 years). UK cross sectional reference data : 1990

Expected weight gain means the weight gain achieved by an unaffected child of the same age and sex, according to above chart. If this information is not available, score 5. If catch-up weight gain has taken place, score 0.

Weight loss in EB, is rarely desirable, but occasionally children become overweight and this can be detrimental to later mobility and sometimes to skin care. If desirable weight loss is not achieved, this is a disadvantageous aspect and should score 10.

Length may be difficult to measure, especially when the skin is very fragile and carers are anxious that further damage will occur during measurement. In this case, score 10. If length is unavailable for other reasons, score 5. If catch-up weight gain has happened, score 0.



Feeding and gastro-intestinal aspects

These directly influence nutritional intake and attitude to food. Long term aversion to eating is often a legacy of factors such as poorly controlled gastro-oesophageal reflux (GOR), naso-gastric tube placement, painful defaecation etc.

Oral lesions, reluctance to feed, gastro-oesophageal reflux (GOR) are all strongly associated with compromised nutritional intake, even when GOR is managed by medication.

Painful defaecation/constipation are major causes of poor food intake. They are often not mentioned by parents/children as they assume them to be unavoidable and untreatable. Diarrhoea is frequently actually overflow of liquid faeces in association with faecal loading and is a symptom of poorly managed constipation. Even if the child's defaecation is controlled by prescribed laxatives/softeners), score 10.

Feed Breast milk, for even a short period, may favour gut colonisation with advantageous flora and promote gut integrity in the longer term.

Naso-gastric tube (NGT) placement may be associated with later food aversion. This and gastrostomy placement per se denote deficit in oral intake.

Dermatological Aspects

Body surface area of denuded/ulcerated skin. The greater the area of denuded/ulcerated skin, the more nutrition is diverted from growth into the healing process. Ask the child's carer to shade in affected areas as accurately as possible on the chart overleaf.

Skin infection compromises appetite and food intake and increases nutritional requirements. Ask the child's carer to indicate these areas on the same chart and use the table to calculate approximate body surface area involved.



"THINC" for Children with Epidermolysis Bullosa (under 18 months of age) ²⁹

Name: Hospital No: dob: / / Date: / /

Please read guidance notes before completing	Nutritional Compromise Rating	Nutritional Compromise Score
Weight and length		
Birth weight centile	> 9th = 0 0.4 – 9th = 5 < 0.4th = 10	
Weight gain/loss in past month	~ 100% expected wt gain OR catch-up = 0 Unavailable OR ~ 50% expected wt gain = 5 Static weight /weight loss = 10	
Birth length centile	Unavailable OR > 9th = 0 0.4 – 9th = 5 < 0.4th = 10	
Length increase in past 2 – 4 months	~ 100% expected increase = 0 Unavailable OR ~ 50% expected increase = 5 Too fragile to measure OR No increase = 10	
Feeding and gastroenterological aspects		
Feed in first 4 – 6 months	Fully breast fed = 0 Breast milk only (expressed or direct suckling) for first 1 – 4 weeks = 0 Combined breast milk and formula = 0 No breast milk, fully formula fed = 5	
Current feed	Cows' milk-based term formula = 0 Nutrient-dense formula OR modified formula eg hydrolysed protein formula = 5 Oral (regular teat) = 5	
Mode (past or present)	Oral with Haberman feeder +/- NGT +/- gast = 10 OR combination of above = 10	
Oral lesions &/or reluctance to feed from breast, bottle or spooned solids &/or gastro-oesophageal reflux (GOR) &/or taking GOR medication(s)	No = 0 Occasionally = 5 Usually / always = 10	
Painful defaecation &/or constipation &/or diarrhoea &/or taking laxatives/ stool softeners	No = 0 Occasionally = 5 Usually / always = 10	
Dermatological Aspects		
Body surface area of denuded/ulcerated skin; % (use grids overleaf)	None = 0 1 - 30% lesions = 5 >30%=10	
Skin infection/sepsis: % (use grids overleaf)	None = 0 1 - 30% lesions = 5 >30%=10	
Total Score	Maximum possible = 100	

Suggested action for ranges of Nutritional Compromise Scores (NCS)

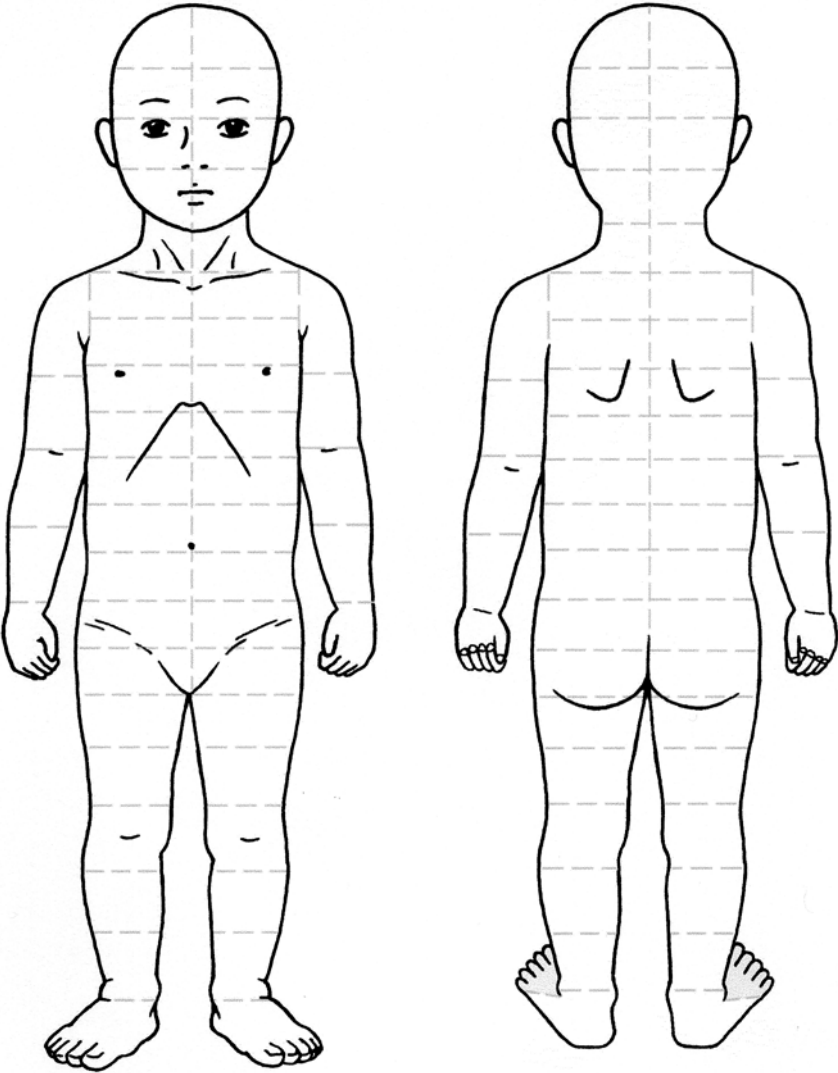
- 0 - 25 Low risk of nutritional problems; review in 3 - 6 months by relevant professionals.
- 26 - 50 Moderate risk, address all scores over 0, involving relevant professionals; review in 1 – 3 months.
- 51 - 75 Significant risk, address all scores over 0, involving relevant professionals. May require admission to hospital for more intensive management; review in 1 month.
- 76 - 100 Very high risk, admit to hospital and address all scores over 0, involving relevant professionals.



Guide to Calculation of Approximate Body Surface Area³⁰ (BSA) of Skin Lesions

Name: Hospital number: Date: / /

- Ask the child’s carer to shade the affected areas, using a different colour for areas of infection.
- Each box shown below represents 1% of total body surface area.
- Count 1% for every box marked if the box is quarter-shaded or more.



Determination of Percentage BSA of Skin Lesions

	% BSA of all lesions	% BSA of infected lesions
Front		
Back		
Total		



Appendix 2

“**THINC**” for Children with Epidermolysis Bullosa
18 months and over

THINC about nutrition

This is a ...

Tool to
Help
Identify
Nutritional
Compromise

in epidermolysis bullosa (EB)
for use by professionals

with children 18 months of age and over
with recessive dystrophic EB, non-Herlitz
junctional EB and Dowling-Meara EB simplex

by Lesley Haynes RD
Principal Paediatric Dietitian
Great Ormond Street Hospital
for Children NHS Trust
London, UK

Guidelines on the use of **THINC** with children

18 months of age and over

The significant nutritional compromise, growth failure and reduced quality of life of severe EB can be improved by measures that minimise the impact of the condition's complications.

THINC's aim is to highlight the child at actual or potential risk of nutritional compromise/ deficiency, so that a proactive treatment plan can be drawn up which minimises the impact of the disease and supports the family in this process.

THINC is intended to be used by a dietitian (or in her/his absence, the health professional supervising the child's medical/surgical care), in association with other appropriate healthcare professionals. The tool should aid, not replace, clinical judgment. It should be used alongside Clinical Practice Guidelines (and cited references) which include algorithms of suggested courses of action.

THINC's scoring chart is intentionally brief and the measures are practical to simplify the completion process. Scores for some aspects are unavoidably subjective, however they have been weighted to allow for this. To maximize the accuracy of the final score, and to provide a useful and meaningful result, please take time to read the following notes. They explain the relevance of the particular aspect rated and offer guidance regarding the questions to ask in order to elicit the most authentic picture of the child's current condition and the potential for development of problems that affect nutritional status. The higher the score, the greater is the likelihood of nutritional compromise. The maximum possible total score is 100.

THINC's scoring chart rates 3 main aspects of the child's state :-

- Weight and height
- Gastroenterology
- Dermatology

Weight and height

Weight (and to some extent height) are amongst the best gauges of long-term nutritional status in severe EB.

Weigh in light indoor clothing and plot on an (where possible) ethnically appropriate chart. If such a chart is unavailable, use a nationally-accepted chart, e.g. in UK use Four-in-One (Duodecimal) Growth Charts (Birth - 20 years). UK cross-sectional reference data : 1990

Expected weight gain = weight gain achieved by an unaffected child of the same age and sex, according to above chart. If this information is not available, score 5.

Height (standing) is difficult or impossible to measure with painful joint contractures and/or skin lesions and osteoporosis. Length may be easier to measure supinely using a mat e.g. Raven Rollametre (Raven Equipment Limited, Unit 4, Ford Farm Industrial Complex, Dunmow, England CM6 1HU). Even this will be impossible in a very severely affected child and a tape measure can be used, although the result will be very approximate. The disparity between height and weight centiles is rated here. If a reasonably accurate height measurement is not possible, score 5.



Gastro-enterology

Gastroenterological factors directly influence nutritional intake and attitude to food.

Tethered tongue causes poor coordination of chewing and swallowing and slows mealtimes significantly. Gauge this by asking the child to protrude the tongue as far as possible, and to move it laterally keeping the jaws aligned.

Severe tethering = cannot protrude tongue beyond lower teeth; cannot move tongue laterally.

Moderate tethering = can protrude tongue beyond lower teeth, but only marginally. Limited lateral tongue movement.

Dysphagia. This can be estimated by asking what texture of food is generally eaten. Although the child who habitually eats a pureed diet may not complain of dysphagia, the long-term nutritional adequacy of such an intake may be very poor and so this should be scored 5 - 10.

Although **microstomia** is a complication of severe RDEB, this has not been included in the scoring due to the absence of a simple and validated measure. Scoring of tongue tethering and dysphagia avoid the need for this.

GOR (gastro-oesophageal reflux). Children and parents may not realize that GOR is present, but recognise, for example, that the child wakes in the night complaining of heartburn type pain that is very likely to be GOR. Even if the child's GOR is controlled by prescribed medication, score 10.

Mucus/phlegm Secretions may be significant and too thick to swallow, requiring regurgitation. This invariably causes nausea and reduces appetite and food intake.

Painful defaecation with or without constipation are major causes of poor food intake. They may not be reported by parents/children because they are assumed to be inevitable and untreatable aspects of EB. Question carefully reports of defaecation several times per day as, paradoxically, this is a cardinal sign of constipation with each defaecation attempt producing a minimal result due to the deterrent effect of abdominal and anal pain.

Diarrhoea may actually be overflow of liquid faeces in association with faecal loading and is often a symptom of poorly managed constipation. Even if the child's defaecation is controlled by prescribed laxatives/softeners/fibre supplements, score 10.

Elimination of dietary components suggests suspected or proven intolerance which is an indicator of increased disease severity.

Dermatology

Body surface area of denuded/ulcerated skin. The greater the area of denuded/ulcerated skin, the more nutrition is diverted from growth. Ask the carer to shade in affected areas as accurately as possible on the chart overleaf.

Skin infection/sepsis compromises food intake, and raises metabolism, increasing nutritional requirements. Ask the carer to indicate these areas on the same chart and use the table to calculate approximate body surface area involved.



Name: Hospital No: dob: / / Date: / /

Please read guidance notes before completing	Nutritional Compromise Rating	Nutritional Compromise Score
Weight and Height		
Current weight kg, centile	> 9th centile = 0 0.4 – 9th centile = 5 < 0.4th centile = 10	
Current height cm, centile Number of centile divisions height centile is greater than weight centile	< 1 = 0 1 – 2 = 5 > 2 = 10	
Approximate weight gain/weight loss in past 6 months	~75-100% expected = 0 ~25-75% expected = 5 >25% OR undesirable weight loss = 10 (Score 0 if child overweight)	
Gastroenterological Aspects		
Tethered tongue*	No = 0 Moderate tongue tethering = 5 Severe tongue tethering = 10	
Dysphagia &/or pureed diet &/or GOR &/or taking anti-reflux medication(s) &/or excess mucus/phlegm	Very rarely/never = 0 Occasionally = 5 Frequently/always = 10	
Painful defaecation &/or constipation &/or rectal bleeding &/or diarrhoea &/or taking laxatives/stool softeners &/or excessive flatus/bloating &/or eliminating dietary component(s) eg cows' milk, wheat	Never = 0 Occasionally = 5 Frequently/always = 10	
Gastrostomy/other feeding tube <i>in situ</i>	No = 0 Yes = 5	
Dermatological Aspects		
Body surface area of denuded /ulcerated skin: % (use grids overleaf)	Nil = 0 1 – 10% = 5 11 – 30% = 10 31 – 50% = 15 > 50% = 20	
Skin infection: % (use grids overleaf)	No = 0 < 25% lesions = 5 26 – 50% lesions = 10 > 50% lesions = 15	
Total Score	Maximum possible = 100	

Suggested action for ranges of Nutritional Compromise Scores (NCS)

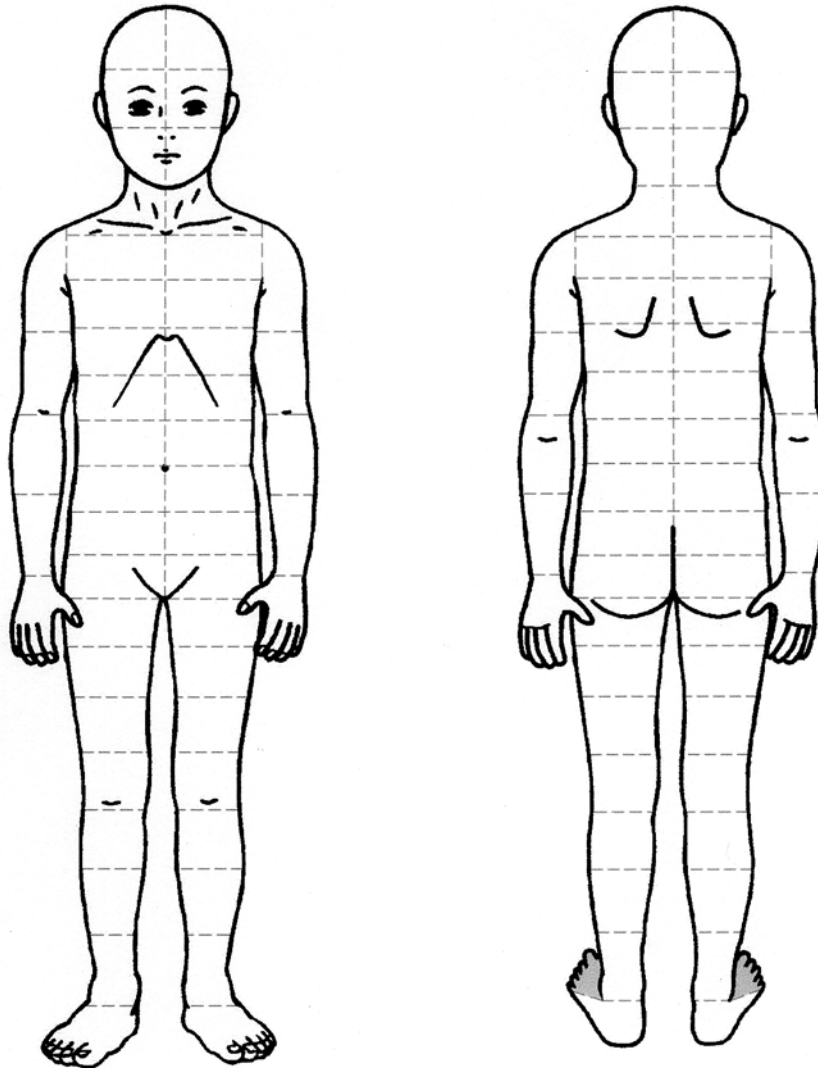
- 0 - 25 Low risk of nutritional compromise; review in 9 - 12 months by relevant professionals.
- 30 - 50 Moderate risk, address all scores over 0 (except that marked *), involving relevant professionals, and review in 4 - 6 months.
- 55 - 75 Significant risk, address all scores over 0 (except that marked *), involving relevant professionals. May require admission to hospital for more intensive management; review in 3 months.
- 80 - 100 Very high risk, seriously consider admission to hospital and address all scores over 0 (except that marked *), involving relevant professionals.



Guide to Calculation of Approximate Body Surface Area (BSA) of Skin Lesions

Name: Hospital number: Date: / /

- Ask the child’s carer to shade the affected areas, using a different colour for areas of infection.
- Each box shown below represents 1% of total body surface area.
- Count 1% for every box marked if the box is quarter shaded or more.



Determination of Percentage BSA of Skin Lesions

	% BSA of all lesions	% BSA of infected lesions
Front		
Back		
Total		



Evaluation and feedback form

The Guidelines are a starting point for the gathering of further evidence to enable best practice and their quality and value will improve only if their users feed back their comments and constructive criticism.

So, please take a few minutes to answer the 3 questions below.

Please communicate this information to me by printing the page off and posting it to me, faxing it to me or copying and pasting it into a Word document and e-mailing to me (see below).

Although inclusion of your contact details is not compulsory, it is requested firstly so that we can communicate if clarification of your comments is required and secondly so that your co-operation can be acknowledged if you so wish. Anonymous feedback will also be very much appreciated.

Please place a ✓ in the box that matches your opinion most closely:-

1. How useful are the Guidelines to you?

(a) Not at all useful (b) Fairly useful (c) Very useful

If you ticked (a) or (b), please describe what additional information would make them more useful

2. How useful is THINC for children under 18 months of age to you?

(a) Not at all useful (b) Fairly useful (c) Very useful

If you ticked (a) or (b), please describe what additional information would make it more useful

3. How useful is THINC for children 18 months of age and over to you?

(a) Not at all useful (b) Fairly useful (c) Very useful

If you ticked (a) or (b), please describe what would additional information would make it more useful

Thank you, now please provide your contact details:

Name Job title

e-mail address@.....

Postal address Phone number:.....

..... Fax number:.....

.....

Please send to: Lesley Haynes, Principal Paediatric Dietitian, Dietetic Department,
Great Ormond Street Hospital for Children NHS Trust, Great Ormond
Street, London WC1N 3JH, UK. e-mail:HayneL@gosh.nhs.uk
Phone: +44-20-7405-9200 ext 5761 Fax: +44-20-7813-8247