Nutrition and Neurodisability

Edited by Peter B. Sullivan, Guro L. Andersen and Morag J. Andrew

A practical guide from Mac Keith Press

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Foreword

Enough and appropriate food and water are essential for human life. This statement is obvious. Nonetheless, lack of food, and starvation are regular occurring consequences of wars and natural disasters. During such crises, children, but also grown-ups, in particular elderly people, may die, or they become more vulnerable to other disorders, such as infectious diseases. In children, somatic growth and psychomotor development will be impaired.

In other settings where food is sufficient, meals are important elements in our social life, ranging from regular daily meals within the family, with friends or colleagues, to the most advanced tasting menus served in the best restaurants. In the short story, *Babette's feast*, the Danish author Karen Blixen, describes how Babette prepares an exquisite meal in a remote fishing village in Northern-Norway, using ingredients imported directly from Paris. Although the locals considered such a meal to be a sinful act, they felt how the meal lifted them both spiritually and physically, how local disagreement and anger evaporated, and love and peace settled on the table.

Persons with disabilities have a range of challenges related to feeding and nutrition. Among the most severe problems is severe oral-motor dysfunction. In early childhood, persons with such dysfunctions are at risk of being truly starved, and undernutrition may impair growth and neurodevelopment. Chewing and swallowing problems, and gastroesophageal reflux may lead to aspiration of food or gastric content into their airways, followed by pneumonia. The other extreme is overfeeding resulting in overweight and adiposity. The latter is often seen in some syndromes, such as Down syndrome and Prader Willi syndrome. However, a too high proportion of body fat may also be the result of misinterpretation of body composition in persons with disabilities where measures such as body mass index applied in the typical developing population may be misleading.

Ten years ago Mac Keith Press published the book *Feeding and nutrition in children with neurodevelopmental disability* edited by Peter Sullivan. That book became a recommended

textbook for many professionals involved in the care for persons with neurodevelopmental disabilities. As Martin Bax stated in his foreword to that book, it was first towards the end of the 20th century that attention was paid to these problems.

This new book edited by Sullivan, Andersen and Andrews, includes the results of the most recent research and practical guidelines regarding assessment of feeding difficulties and body composition as well as the most recent recommendations regarding treatment. The authors of the various chapters are world-leading experts from Europe, Australia and North-America. The authors address all important aspects of this challenging topic. The practical perspective is essential and will certainly be appreciated by inter-professional teams trying to help optimizing the nutritional status of children and adults with disabilities.

In *Babette's feast* there were some original guests, but none with a clear neurodevelopmental disability. Within a family setting, severe feeding difficulties are likely to affect the family's quality of life. The application of the knowledge provided in this new book should provide a basis for enabling persons with disabilities to take part in, and enjoy any meal both in a private and a public setting, and even to experience such meals as presented in *Babette's feast*. Most importantly, the content in the book should contribute to a lighter daily life in families of persons with disabilities, and for the persons themselves.

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Preface

This book is the third produced on this topic by Mac Keith Press over the last three decades. Acknowledgement for this initiative should go to Martin Bax who as Editor of *Developmental Medicine & Child Neurology* realised that the nutritional needs of children with cerebral palsy were not adequately being met by health care professionals. The early texts on cerebral palsy make little or no mention of the feeding difficulties encountered in children with severe neurological impairment and feeding and nutritional assessment was not part of the routine care of these children. In the late 1980's, it was Martin who saw the advantage of teaming up a paediatric gastroenterologist with an interest in nutrition with a paediatric neurologist; this culminated in the first volume *Feeding the Disabled Child* edited by myself and Lewis Rosenbloom and published in 1996. Following Martin's retirement, Lewis, as Chair of the Editorial Board of Mac Keith Press persuaded me that an update in the form of a practical handbook was required. This lead to the publication of *Feeding and Nutrition in Children with Neurodevelopmental Disability* by Mac Keith Press in 2009. Now some ten years later and with increasing research interest and endeavour and with the appearance of guidelines from various learned bodies on the topic of nutritional and feeding problems in children with neurological impairment, Bernard Dan the current Editor in Chief of *Developmental Medicine & Child Neurology* considered it is necessary again to update the handbook.

This volume is designed to be a practical evidenced-based handbook aimed at health professionals who have responsibility for caring for children with the feeding, nutritional and gastrointestinal problems that ensue from neurological impairment and especially cerebral palsy. This book will be of value both for those new to this clinical field and for more experienced practitioners.

An appreciation of the development of the normal anatomy and physiology of the oral-motor apparatus is essential for understanding of the pathophysiology of oral-motor dysfunction that underlies the feeding problems in children with neurological impairment; these aspects are covered in the opening two chapters. Drooling of saliva is an important additional consequence of oral-motor dysfunction and this problem and its

management is covered in a separate chapter. Oral-motor dysfunction is a component of an overall abnormality in gastrointestinal motility resulting in increased gastro-oesophageal reflux, delayed gastric emptying and constipation. Evaluation and management of these gastroenterological problems, which affect the great majority of children with neurological impairment and especially these children with cerebral palsy is covered in detail. Nutritional impairment is frequently a consequence of the feeding problems encountered by children with cerebral palsy and the impact of such undernutrition on growth, metabolism, cognitive and immune function may be overlooked. Recent research has led to a greater understanding of the adverse consequences of undernutrition and these are detailed in this volume.

Successful nutritional management depends upon accurate assessment and so the heart of this handbook comprises a series of chapters on assessment; these cover growth, energy balance, body composition, macro- and micronutrient intake, and dietetic assessment. Because children with neurodisabilities have risk for compromised bone health attributable to a combination of atypical muscle tone combined with lack of weight bearing resulting in reduced bone size and bone density, this new handbook devotes a whole chapter to this topic. All the assessment chapters describe the range of methods and techniques used in nutritional assessment together with an evaluation of the advantages and disadvantages of each technique and a discussion of the validity for their use in children with neurological impairment. The issue of (lack of) appropriate reference standards in children with neurological impairment for growth and nutritional intake is taken into account in the recommendations made.

Following on from detailed feeding, nutritional and gastroenterological assessment comes the development of a management strategy. Children with neurological impairment, and especially those with cerebral palsy, form a rather heterogeneous clinical group. Accordingly, management strategies must be individualised and targeted and the specific needs of each child. Furthermore, successful feeding, nutritional and gastroenterological management will not be the province of one particular professional discipline but will be the outcome of the input and endeavours of a multi-disciplinary team. These basic principles run through all the chapters on assessment and management. Where appropriate clinical case vignettes are used to illustrate points being made in the text and it is hoped that these will prove to be an instructive and valuable addition to the handbook.

Enteral tube feeding has transformed the landscape of nutritional management of children with cerebral palsy in recent years and whilst it circumvents the problems of feeding inefficiency and unsafe swallow associated with oral-motor impairment, the technique is not without its problems. Amongst these are included the complications of the procedure itself, the potential risk of overfeeding and the significant impact – both beneficial and adverse - that gastrostomy or jejunostomy tube feeding has on parents.

These issues are explored in detail in the penultimate chapter. The handbook ends with a resume of all the take home messages from each of the foregoing chapters.

In producing this handbook, the editors have sought contributions from the acknowledged leaders in each topic from around the world. It is our earnest hope that this handbook will prove to be a useful resource for any health professional engaged in the assessment and management of the feeding, nutritional and gastrointestinal problems in children with neurological impairment.

I would like to acknowledge the continuous help and support given to the editors to produce this handbook from Rosie Outred, Lucy White and Sally Wilkinson from Mac Keith Press.

Peter B Sullivan, Oxford
November 2019

CHAPTER 5

Consequences of Nutritional Impairment

Jessie M Hulst

INTRODUCTION

The gastrointestinal problems, oral motor dysfunction, immobility and dentition problems described in the previous chapters contribute to nutritional impairment of children with neurological impairment (NI) (Andrew et al. 2012). The term nutritional impairment used refers to undernutrition, malnutrition or poor nutritional status due to inadequate nutritional intake. Studies have shown that feeding difficulties and nutritional problems are frequent in children with NI (Stallings et al. 1993; Reilly et al. 1996; Fung et al. 2002; Andrew et al. 2012).

There is a strong relation between the grade of disability, the severity of feeding problems, nutritional intake and nutritional status in these children. Nutritional impairment can affect several physical and psychological functions leading to clinical consequences and is associated with worse outcome (Fung et al. 2002; Stevenson et al. 2006b). These consequences include undernutrition, growth failure, micronutrient deficiencies and poor bone health; they will be outlined in the following paragraphs. Figure 5.1 depicts the relationships between factors contributing to feeding difficulties, inadequate nutritional intake and its consequences in children with neurological impairment.

On the other hand there is some evidence that those children with NI with less severe motor impairments may be at increased risk of becoming overweight (Rogozinski et al.

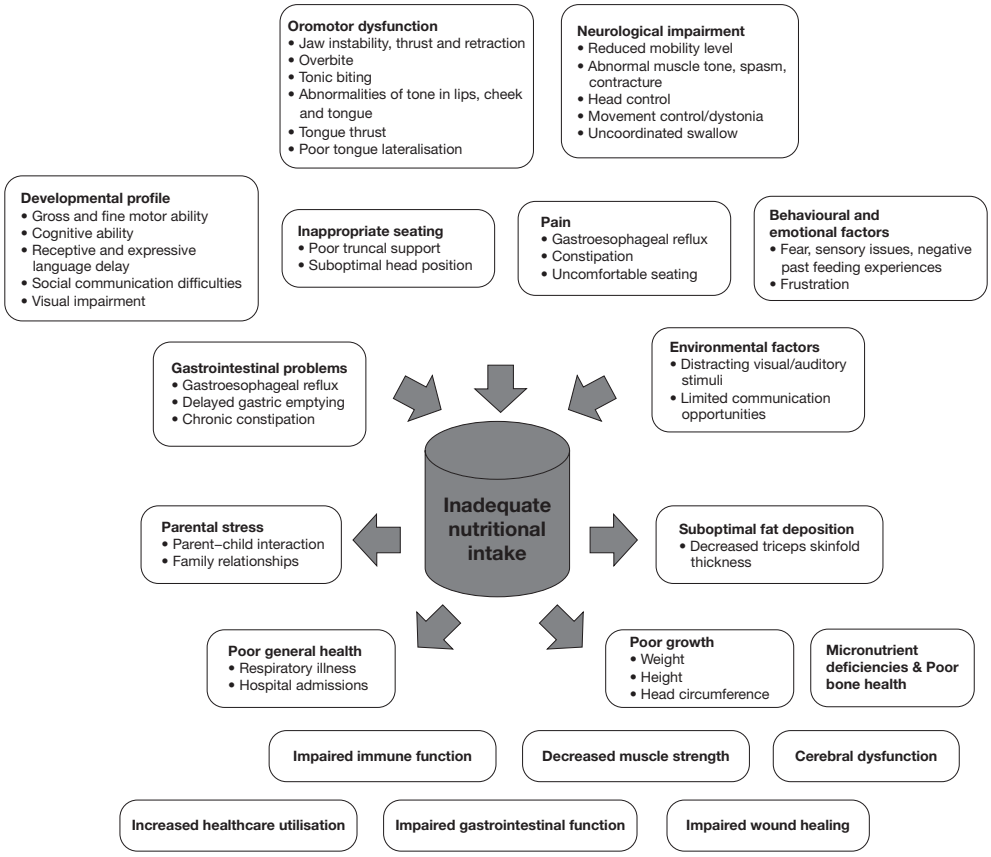

Figure 5.1 Relationships between factors contributing to feeding difficulties, inadequate nutritional intake and its consequences in children with neurological impairment. (Adapted from Andrew et al. 2012 with permission from BMJ Publishing Group Ltd.)

2007; Hurvitz et al. 2008), as are those immobile children receiving gastrostomy feeding. Gastrostomy feeding improves overall weight gain, but has been associated with excess deposition of fat (Sullivan et al. 2006). A study in the USA showed that the prevalence of obesity (based on BMI-for-age > 95th centile) in ambulatory children (GMFCS level I–III) with cerebral palsy (CP) increased from 7.7% in 1994 to 16.5% up to 2004 (Rogozinski et al. 2007). Children with a lesser degree of involvement showed the highest odds of becoming obese. In two other large cohorts of ambulatory children with CP in Korea (Park et al. 2011) and Australia (Pascoe et al. 2016), 11.2% and 7.3% of children were overweight; obesity was identified in 5.8% and 12.1%. In the Australian study, children with GMFCS level III were more likely to be overweight or obese than children with GMFCS level I (Pascoe et al. 2016).

CLINICAL CONSEQUENCES

An overview of the clinical consequences associated with nutritional impairment are shown in Table 5.1.

To examine the associations of impaired nutritional status or impaired intake of nutrients on outcome parameters, it would be necessary to compare two groups with the same condition, one with impaired nutritional status/intake and one without. In clinical practice this can be very difficult to achieve especially in children with severe neurological impairments. That is why well-performed studies are lacking. In reality, some of the consequences may also only be recognised when the nutritional status and/or nutritional intake has improved, for example the child can be more cheerful, more relaxed, less irritable and less often sick (Marchand et al. 2006).

Growth Failure: Low Weight/Impaired Height and Decreased Head Growth

The most obvious sign of poor nutritional intake is poor weight gain or loss of body weight, due to loss of fat and muscle mass. In general, the pattern of change in body composition will depend on the previous storage of fat and muscle and on sex. Depending on which definition is used for nutritional status – BMI/weight-for-age/weight-for-height/skinfold thickness – and depending on the included type of patients in terms of severity and region in the world, various prevalence rates of malnutrition are reported, most often from studies in children with CP. These rates are between 20–90% (Stallings et al. 1993; Stevenson et al. 1994; Marchand et al. 2006, 2006b; Kuperminc & Stevenson 2008; Hariprasad et al. 2017; Minocha et al. 2018).

Sustained nutritional imbalances in children can also result in impaired linear growth (stunting) and decreased head growth. Children with CP tend to be smaller and grow more slowly than typically developing children and the differences in growth increase with advancing age (Stevenson et al. 1994; Samson-Fang & Stevenson 1998). Malnutrition appears to be a major contributing cause but other factors such as endocrine dysfunction are also important (Stallings et al. 1993, 1996; Stevenson et al. 1994; Samson-Fang & Stevenson 1998).

Stunting has been described especially in children with NI and severe motor impairments (Stevenson et al. 2006b; Dabydeen et al. 2008; Hariprasad et al. 2017). It is important to realise that stunting or thinness based on BMI does not have to occur together with undernutrition/wasting as the percentage of body fat assessed by skinfold measurements or dual-energy X-ray absorptiometry can be normal or high (Finbraten et al. 2015).

Table 5.1 Consequences of impaired nutritional status and inadequate nutritional intake in children with neurological impairment (see text for references). (Reprinted from Kecskemethy & Harcke 2014 with permission from IOS Press.)

Consequence
Growth failure – undernutrition
Low weight (wasting)
Impaired linear growth (stunting)
Decreased head growth
Cerebral dysfunction
Reduced potential
Poor concentration
Reduced responsivity
Withdrawal/irritability
Worsening of epilepsy
Depression/apathy
Decreased muscle strength
Decreased respiratory muscle
Increased work of breathing
Weak cough
Impaired cardiac function
Increased circulation time
Poor healing
Micronutrient deficiencies
Low iron: associated with anaemia, fatigue, irritability, cognitive deficits, behavioural abnormalities
Low zinc: associated with anorexia, alopecia, eczema, diarrhoea, reduced growth, skin lesions, stomatitis, impaired wound healing, frequent infections
Low copper: associated with sideroblastic anaemia, reduced growth, osteoporosis, peripheral neuropathy, increased susceptibility to infections
Low folic acid: associated with megaloblastic anaemia, anorexia, behavioural changes, reduced growth
Low vitamin B12: associated with megaloblastic anaemia, muscle weakness, ataxia, spasticity, incontinence
Low vitamin D: associated with osteomalacia/penia, rickets, muscle weakness, decreased immunity, caries, hypocalcaemia, hypophosphataemia
Low carnitine: decreased muscle tone or muscle weakness, fatigue, irritability, poor feeding in infant, hypoglycaemia, cardiomyopathy
Poor bone health (demineralisation/fractures)
Impaired immune function
Increased risk of infections

(continued on next page)

Table 5.1 Consequences of impaired nutritional status and inadequate nutritional intake in children with neurological impairment (see text for references). (Reprinted from Kecskemethy & Harcke 2014 with permission from IOS Press.) (continued)

Impaired wound healing
Poor healing (especially pressure sores)
Impaired gastrointestinal function
Increased healthcare utilisation
Increase in complication rate
Increase in hospital admission and doctor's visits
Increase in length of hospital stay
Decreased level of child and family societal participation
Increased number of missed school days
Increased number of missed family activities
Decreased quality of life/general well-being

The relationship between head growth and poor nutritional intake is shown in a few interventional studies in children with neurological impairments. Additional nasogastric tube feeding improved growth and gross motor functioning compared to a group who did not receive supplements (Campanozzi et al. 2007). Another study looking into the effects of a high-energy and high-protein diet showed increased head circumference and corticospinal tract diameter in infants with perinatal brain injury when compared to infants with normal intake (Dabydeen et al. 2008).

Cerebral Function

Adequate macro- and micronutrient intake are essential for healthy brain development (Lucas et al. 1998). So, in general, learning, behaviour and cognitive functioning can be influenced by nutrition in several ways and at different periods throughout childhood. This has been shown in studies of infants with failure to thrive who were assessed in primary care and hospital clinics at childhood age (Corbett & Drewett 2004). A small-scale study (n = 20, age 5–7 years and 8–10 years) from India investigating the association between stunting and/or wasting (as a result of chronic protein-energy malnutrition) and cognitive development found that malnourished children performed poorly compared with well-nourished children on tests of attention, working memory, learning and memory, and visuospatial ability. Malnourished children, in particular those with stunting, showed a lack of age-related improvement on tests of design fluency, working memory, visual construction, learning and memory (Kar et al. 2008). A large study in children of 8 years of age, The Avon Longitudinal Study of Parents and Children in the UK, found that early growth faltering (defined as < 5th centile for weight gain in the first 8 weeks) was associated with a significantly lower total intelligence quotient by an average of –2.71 points at 8 years of age (Emond et al. 2007).

Studies looking into cerebral function in relation to nutritional status or nutrient intake in children with NI are scarce, but urgently needed (Dan 2016). As adequate macro- and micronutrient intake is needed to support brain development, nutritional inadequacy may limit the brain's capacity for remodelling and repair after injury. This issue is very relevant to children with NI since they suffer neurological injury and they often have inadequate or suboptimal nutrient intake.

A recent double blind randomised controlled trial investigating the effect of early 2-year phosphatidylcholine precursor supplementation (DHA, choline and UMP) in infants with suspected CP showed no statistically significant neurodevelopmental advantage for the treatment group as compared to the control group. There was, however, a clinically meaningful cognitive and language advantage found in the treatment group (Andrew et al. 2015, 2018).

Clinically meaningful changes in the outcome of children with NI such as less irritability, improved responsiveness, better concentration and less apathy are also frequently reported by parents but not adequately studied.

Decreased Muscle Strength

Protein and energy malnutrition can lead to reduced lean body mass which is associated with decreased muscle strength. In healthy individuals, muscle function, as assessed by grip strength, is directly proportional to indices of body muscle mass. It has been suggested that impaired muscle function during low energy intake and malnutrition could be, apart from reduced muscle mass, related to several factors, for example alterations in numbers of muscle fibres, changes in activities of muscle enzymes, defects in calcium channels or sodium-potassium ATPase pumps (Stratton et al. 2003).

Several studies in adults with various diseases have shown associations between loss of muscle mass and outcome parameters such as postoperative complications after gastrointestinal surgery and mortality (Stratton et al. 2003).

In general, malnutrition can also result in loss of cardiac muscle which can result in decreased cardiac output, bradycardia, hypotension and peripheral circulation failure; the last sign is frequently seen in children with NI in clinical practice.

Moreover, malnutrition and protein depletion can adversely affect respiratory muscle structure and function, resulting in reduced muscle mass of the diaphragm and reduced respiratory muscle strength. Impaired respiratory function and muscle strength will weaken the cough and impair airway clearance, which may predispose to and delay recovery form chest infections. Although specific studies in children with NI about the association between nutritional status, muscle strength and adverse events are lacking,

these associations are highly relevant in this population as they frequently experience chest infections (Boel et al. 2018). One study in a small group of children with spastic quadriplegic CP that assessed the health status before, at and 6 months after gastrostomy tube placement showed an improvement in nutritional status over time and in 50% of children a decrease in the number of chest infections requiring antibiotics was seen after 6 months (Vernon-Roberts et al. 2010).

Micronutrient Deficiencies

Poor overall nutritional intake can lead to lower micronutrient intake, which predisposes children with NI to develop micronutrient deficiencies. In general, micronutrients are important for many metabolic pathways in the body and generalised or specific micronutrient deficiencies may have multiple consequences (Table 5.1). These consequences may be difficult to distinguish from the general neurological impairment of NI children as they may affect cognition, behaviour, social interaction and developmental outcomes.

Few studies have evaluated the micronutrient status of children with NI and the implications of various deficiencies on health outcome (Sanchez-Lastres et al. 2003; Hillesund et al. 2007; Tomoum et al. 2010; Kalra et al. 2015; Takeda et al. 2015). These studies show that deficiencies for iron, zinc, copper, vitamin D, carnitine, folic acid and vitamin B12 are common with percentages ranging between 10% and 55%. Factors associated with low levels were found to be vitamin C intake (iron), use of antiepileptic drugs (carnitine, vitamin B12, folic acid, calcium and phosphorus), and reduced exposure to sunlight (vitamin D).

Tube feeding and the use of nutritional supplements were associated with higher concentrations of micronutrients in blood and serum (Hillesund et al. 2007). On the other hand it is also known that deficiencies in micronutrients can still occur when children with NI are exclusively tube fed with a standard formula (Piccoli et al. 2002). Children with NI may require less energy in order to avoid becoming overweight and, as a consequence of a reduced energy intake, their micronutrient intake can be less than daily requirements. Essential fatty acid (FA) deficiency may also be related to suboptimal energy intake as was shown in a study where children with NI were found to have lower levels of docosahexaenoic acid, linoleic acid and total n-6FA in comparison with a healthy reference group (Hals et al. 2000).

The monitoring of micronutrient status in NI children may have a substantial and measurable impact on their nutritional adequacy, hospital costs and future outcomes. In their recent guidelines for the evaluation and treatment of nutritional complications in children with neurological impairment, The European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN) recommends the assessment of micronutrient status (e.g. vitamin D, iron, calcium, phosphorus) as part of nutritional assessment and that micronutrients should be checked annually (Romano et al. 2017).

ESPGHAN also recommends use of the dietary reference intake for micronutrients in typically developing children to estimate the appropriate micronutrient intake for NI children (Romano et al. 2017).

Iron

Iron deficiency is a common problem in children with NI (Sullivan et al. 2002; Papadopoulos et al. 2008) and is related to insufficient intake and inadequate iron absorption. Children with NI on a liquid diet were found to have higher rates of anaemia and iron deficiency compared to children on a normal diet. Prepared liquid diets, although including various types of food, may consist of foods that are a poor source of iron (milk, cheese, cream, yoghurt, rice) or that inhibit iron absorption (vegetables, grains, pulses, cereals) (Papadopoulos et al. 2008).

Since iron deficiency is associated with anaemia, cognitive deficits and behavioural abnormalities, it is important to monitor and supplement when needed. The recommended daily intake of iron is 10mg/day in children (7–10 years), 12mg/day in adolescent males (15–19 years) and 15mg/day in adolescent females (15–19 years). In the treatment of iron deficiency in children with NI it is reasonable to provide iron supplementation as the first diagnostic and therapeutic measure in such patients.

Vitamin D

Decreased 25-hydroxy vitamin D is a major deficiency noted and besides inadequate food intake, inadequate exposure to sunlight and the use of anticonvulsants can be additional risk factors in this group of children. Anticonvulsants can increase the activity of cytochrome P450 mixed function oxidase enzyme, which results in the conversion of vitamin D to an inactive metabolite.

Poor Bone Health

Low bone mineralisation (osteopenia and osteoporosis) is a severe and frequently encountered problem in children with NI and is associated with significant fracture risk. There are multiple factors that can contribute to poor bone health which can all be present in children with NI: vitamin D deficiency, poor intake of calcium and phosphorous, limited ambulation (GMFCS level IV and V), feeding difficulties, low weight-for-age, previous fracture, anticonvulsant use and lower fat mass (Duncan et al. 1999; Henderson et al. 2002; Stevenson et al. 2006a; Mergler et al. 2009; Bianchi et al. 2014). Rickets is infrequently seen in this population because it is a disease of growing children and many children with CP are growing poorly or not at all. This issue is covered in detail in Chapter 8.

Impaired Immune Function and Increased Infection

Protein-energy malnutrition and micronutrient deficiencies can negatively influence the haematopoietic and lymphoid organs and compromise both innate and adaptive immune functions. These changes are associated with impaired ability to prevent, fight and recover from various types of infections. Most of the knowledge about the impact of malnutrition on host defence comes from animal studies and studies in children in developing countries (Ibrahim et al. 2017).

Skin, respiratory and gastrointestinal mucosal barrier integrity can be impaired in children with malnutrition. Children with neurological impairment are prone to respiratory infections (Millman et al. 2016) because of a combination of factors such as decline in lung function, respiratory muscle weakness or atrophy, increased bacterial colonisation of airways and decreased resistance to infection (Boel et al. 2018). Poor nutritional status and decreased nutritional intake may be an important additional risk affecting the different factors.

Micronutrient deficiencies, such as deficiencies of iron, zinc, selenium and vitamins A/C/D, can also have a profound effect on immune function and host defence (Ibrahim et al. 2017). Because children with NI frequently show inadequate micronutrient status, this is highly relevant to this group, and may further contribute to the increased susceptibility to infections and their severity.

Impaired Wound Healing

Nutrition plays an important role in the complex process of wound healing and the development of wounds such as pressure ulcers. Among other factors – for example local factors, presence of chronic disease and age – nutritional status and recent nutritional intake seem to be especially important (Clark et al. 2000). Several studies showed impaired or prolonged wound healing in malnourished patients when compared to patients with normal nutritional status.

Malnutrition goes together with reduced nutrient availability for metabolism, maintenance and repair; losses of fat; physical weakness; decreases in skin resistance; edema; and decreased motility, which are associated with the risk of pressure ulcer development. Specific micronutrient deficiencies, such as vitamin A, vitamin C, vitamin E and zinc, may also play a role in this increased risk. This has been shown by a number of studies, not specifically in individuals with NI. In children with NI several risk factors may be present besides poor nutritional status and nutritional intake, such as less activity (wheelchair bound, bed bound), incontinence (moisture), loss of sensations (reduced pain sensation that would normally cause an immobile individual to change

position), contractures and spasticity (repeated exposure of tissues to pressure through flexion of a joint).

A retrospective study among 79 children with CP who underwent hip surgeries concluded that risk factors for complications were non-ambulant status and on top of this the presence of gastrostomy feeding, indicating the role of nutritional status. Gross motor function correlated well with the risk of complications after osteotomies (Stasikelis et al. 1999).

Another recent study in patients undergoing posterior spine fusion for neuromuscular scoliosis consecutive to CP (n = 66) or muscular dystrophy (n = 30) showed a high rate of early complications especially in the CP group (59%) and especially infectious complications (32%). Infectious complications included 16 wound infections (16.7%) and associations with a lower body weight and poor nutritional status were found (Pesenti et al. 2016).

Impaired Intestinal Function

Adequate nutrition is important for preserving gut structure and function, including digestion and absorption of nutrients and providing the gut barrier. Changes in gastrointestinal structure and function can be seen especially when luminal nutrition is lacking (Stratton et al. 2003). These effects may not be apparent in situations with chronic energy restriction with preserved (minimal) enteral nutrition even though body weight may be dramatically decreased. Few human studies have been performed about the effects of acute and chronic food deprivation on the gastrointestinal tract (absorption, intestinal permeability and transit time), and no studies are available in children with NI. The most evident effect of acute starvation is a reduction of small bowel absorptive surface area due to villous blunting, leading to impaired absorption of monosaccharide and disaccharide that can contribute to diarrhoea (Kvissberg et al. 2016). On the other hand, compensatory mechanisms leading to maximisation of nutrient absorption also come into play. Therefore the clinical significance of changes in gut structure and function in case of malnutrition remain unclear. It is known however that effects of malnutrition on the gut can be more pronounced in the presence of acute illness.

Specific micronutrient deficiencies may also have an effect on intestinal morphology, for example vitamin B2.

It is well known that NI can affect the gastrointestinal system, most notably the oral motor function and motility. The association between nutritional status and gastrointestinal problems in NI children is not well studied. A small uncontrolled study among 21 children with CP and severe intellectual disability looked at the relationship between nutritional status and gastrointestinal problems (gastroesophageal reflux disease [GERD] and/or chronic constipation), and evaluated the role of nutrition on their gross motor

abilities (Campanozzi et al. 2007). Nutritional rehabilitation (increase of daily calories by 20%) of malnourished children took place and children with GERD received additional proton pump inhibitor treatment. While the nutritional status improved, the majority of patients had persistent GERD after 6 months of combined treatment. Improved nutritional status, particularly fat free mass gain, appeared to have a positive impact on motor function in children with CP.

Increased Healthcare Utilisation

In general, malnutrition in children has been shown to be associated with an increased length of hospital stay in a number of studies (Joosten et al. 2010; Hecht et al. 2015). This increase in use of healthcare resources is likely to increase the cost of care of malnourished children. A large retrospective analysis of over 6 million hospitalised children aged < 17 years in the USA found that length of hospital stay among children with a coded diagnosis of malnutrition was significantly longer than those without. In addition, they found that discharge home with care was 3.5 times more common among malnourished patients (10.9% vs 3.1%, $p < 0.001$) (Abdelhadi et al. 2016). In this study hospitalisation costs were US$55 255 for children with a malnutrition diagnosis vs US$17 309 without. The higher requirement of post-discharge home care further suggests higher costs in the community as well.

The impact of poor growth and nutrition on health outcomes in children with moderate to severe CP were investigated in few studies. The children with the poorest overall growth (low arm fat and arm muscle area) had more days of health service utilisation (i.e. doctors' visits, hospitalisations) compared with those children who had the best overall growth (Samson-Fang et al. 2002). This study was the first to document a link between nutritional status, as defined by anthropometry and healthcare utilisation.

In a large multicentre study among 273 children with moderate and severe CP (GMFCS II–IV–V) healthcare use (days in bed, days in hospital and visits to doctor or emergency department) and social participation (days missed of school or of usual activities for child and family) over the preceding 4 weeks were measured by questionnaire. The results showed that children with the best growth had fewest days of healthcare use and fewest days of social participation missed, whereas the children with the worst growth had the most days of healthcare use and most days of participation missed (Stevenson et al. 2006b).

Level of Child and Family Societal Participation

The impact of poor growth and nutrition on levels of participation in children with moderate to severe CP was also incorporated in the study of Samson-Fang and colleagues (Samson-Fang et al. 2002). Low arm fat and arm muscle area were associated with

decreased global health scores, and child and family societal participation. Children with poorest overall growth had lower levels of participation compared with those children who had the best overall growth.

Data in children with CP from The North American Growth in Cerebral Palsy Project showed that low fat stores were associated with lower global health scores, less child and family participation and increased use of health care (Liptak et al. 2001).

Quality of Life

Children with NI have a reduced health-related quality of life (QoL) and the degree is related to the severity of their NI (Samson-Fang et al. 2002; Vargus-Adams 2005). The presence and severity of malnutrition and of feeding problems both have an impact on QoL. Studies directly addressing the relationship between nutritional status, nutritional problems and QoL in children with NI are lacking, but there are some studies looking into QoL after initiating tube feeding with subsequent improvement in nutritional status. A prospective longitudinal study addressing QoL before and after gastrostomy or gastrojejunal tube insertion showed a mean increase in weight for age z-score from −2.8 at baseline to −1.8 at 12 months, with no increase in mean parental-rated QoL and health-related QoL scores in the same period. However, parents felt that the tube had a positive impact on their child's health, particularly with regards to feeding and administration of medications (Mahant et al. 2009).

Indirectly, it has also been shown that the quality of life of caregivers of children with CP can significantly be improved after insertion of a gastrostomy feeding tube in the child in their care, in terms of significant reduction in feeding times, increased ease of drug administration and reduced concern about the child's nutritional status (Sullivan et al. 2004).

On the other hand, being overweight or obese, which is becoming more prevalent in ambulatory children with NI (Pascoe et al. 2016), can also be an important topic in relation to QoL. For children with NI who may already have difficulties with body image and may experience prejudice and discrimination, the combination of being overweight or obese with physical disability can impact QoL to a greater degree than in children who are obese but do not have a physical disability (Rimmer et al. 2011). Increased body mass in conjunction with increasing musculoskeletal impairments may result in progressive loss of function and mobility when compared with peers without a disability (Park et al. 2011).

Mortality

Although high-quality studies specifically addressing children with NI are lacking and data demonstrating that malnutrition has an adverse impact on morbidity and mortality

in paediatrics is limited, it is clear from extrapolation of studies in adults and from studies in children in developing countries that malnutrition is associated with a greater risk (Moy et al. 1990; Stratton et al. 2003). A small recent study about risk factors for mortality in children with CP in Indonesia reported on nutritional status but could not relate it to increased mortality risk (Prastiya et al. 2018). A 1-year follow-up study in 81 children and adolescents with CP in Chile did not find an increased mortality in children at high nutritional risk, but both nutritional risk and mortality were found to be significantly higher in gastrostomy-fed children (RR 2,98 CI 95%: 1.32–6.75 combining both variables) (Figueroa et al. 2017). A possible reason for this is that gastrostomy tube feeding is in itself an indicator of severity of impairment and therefore of an increased risk of mortality.

SUMMARY

Children with NI frequently have nutritional impairment due to inadequate nutritional intake and this has enormous impact on overall health and QoL. There is a strong relation between the grade of general motor function and cognitive ability, the severity of feeding problems, nutritional intake and nutritional status in these children. Nutritional assessment and support should be an integral part of the care of children with NI aiming at early identification of children at risk of nutrition-related comorbidities. To ensure success of interventions, a multidisciplinary team should perform close monitoring of nutritional status.

Case Study

An 18-year-old boy with severe intellectual and motoric disability because of a chromosomal disorder, known with tetraplegic spasticity and GMCFS level IV, was admitted to undergo several orthopaedic surgical corrections in one session. He had been on the waiting list for more than 18 months and was followed by the rehabilitation physician. His preoperative anaesthetic screening took place 2 days before the planned surgery and a weight of 38kg and height of 160cm were obtained. His nutritional status parameters were automatically calculated in the digital patient system and were: WFA –5.1SD, WFH –2.2SD and HFA –3.9SD based on national reference for healthy children, indicating undernutrition. He was used to eating orally and parents told that he enjoyed that.

All surgical interventions went well, but took a total of 11 hours. Postoperatively he developed several problems including respiratory failure because of aspiration pneumonia necessitating non-invasive ventilation at the paediatric intensive care unit and an extended length of hospital stay. In addition he developed a pressure sore at the left knee and severe nutritional problems necessitating nasojejunal tube feeding. After one month, he was discharged home with a weight of 33.7kg (WFA –6.08SD, BMI –7.1SD based on healthy population reference) with additional measures including sputum drainage system and tube feeding.

Already after one day he was re-admitted to the paediatric ward because of respiratory problems with aspiration of enteral formula. He was very tired, not able to sit for more than

15 minutes and showed signs of skin pressure at his spine. As he was not able to tolerate enteral nutrition, he was started on parenteral nutrition to try to let him gain some weight and recover. Because of increasing respiratory problems with incidents with low saturation, because of inability to clear sputum and swallowing difficulties, he was transported to paediatric intensive care again for non-invasive ventilatory support. After some clinical improvement, a barium contrast swallowing study was performed which showed an unsafe swallowing with aspiration. After careful discussion with parents and the treatment team, it was decided to give him a jejunostomy in order to try to feed him enterally as placement of nasal gastrojejunal tubes were not successful. In the days after the jejunostomy placement he developed a pneumonia again and became respiratory insufficient. After some days without improvement, it was decided to continue with palliative comfort care (no intubation) because of the total deterioration of his condition. Six weeks after the initial admission for orthopaedic surgery he sadly passed away.

- *This clinical case gives an example of how a poor nutritional status can play a role in the development of complications after surgery. A multidisciplinary assessment (surgeon, paediatrician, dietician and speech therapist/occupational therapist) in the preoperative period could have given insight about nutritional status and its contributing factors and may have led to a (nutritional) intervention in order to try to improve nutritional status before major surgery. Although it is difficult to actually tell and considering other relevant aspects in a neurologically impaired child, this may have led to a better outcome.*

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CHAPTER 10

Enteral Tube Feeding: Practical and Ethical Considerations

Peter B Sullivan

INTRODUCTION

Progress in neonatal intensive care techniques have resulted in improved survival of extremely preterm and low-birthweight neonates. Of those that survive from extreme prematurity, 25% will have permanent psychomotor problems including feeding and nutritional problems (Costeloe et al. 2012). Many of these children will grow up with a disability so profound that they are never likely to become independently mobile, to communicate effectively with others or to feed themselves.

At least a third of children with moderate to severe cerebral palsy (CP) will have feeding difficulties. Malnutrition should not be considered normal in children with CP. Early, persistent and severe feeding difficulties are a marker for subsequent poor growth and developmental outcomes. Growth patterns in children with CP are associated with their overall health and social participation. Growth restriction increases progressively with age and thus mandates early nutritional intervention. In children with severe CP such nutritional intervention is increasingly being administered by gastrostomy feeding tube but controversy surrounds the evidence base for this approach. Moreover, parental decisions about gastrostomy feeding are complex and difficult and must be taken into account in making therapeutic recommendations. This chapter discusses the available research evidence and psychosocial issues around gastrostomy feeding in children with severe CP. It seeks to provide a basis for rational clinical decision making based upon the integration of the best available research evidence with clinical experience and patient values.

CONSEQUENCES AND MANAGEMENT OF UNDERNUTRITION

The main consequence of undernutrition is growth failure. Chapter 5 deals with this issue in detail. Children with moderate or severe CP have poor growth compared with typical children and this correlates with markers of health and social participation; well-nourished children with CP have better health and more social participation than similar undernourished children (Liptak et al. 2001; Stevenson et al. 2006).

Enteral Tube Feeding

The principles of nutritional assessment and management are described in the preceding chapters. Enteral tube feeding is frequently indicated in children with CP with significant oropharyngeal incoordination who are unable to meet their nutritional requirements orally.

Nasogastric tube feeding is mostly used for short-term enteral tube feeding – often prior to insertion of gastrostomy tube. There are several limitations for long-term nasogastric tube use, including nasal discomfort, irritation or penetration of the larynx, oesophageal erosion, recurrent pulmonary aspiration, and blockage or displacement of the tube. Weight gain in children followed for over a year is better in those with a gastrostomy compared with those with a nasogastric tube (Samson-Fang et al. 2003). Furthermore, survival rates among children with severe neurological disabilities fed by gastrostomy tube are significantly better than those fed by nasogastric tube.

Indications for Gastrostomy Tube Feeding

The range of indications for insertion of a gastrostomy feeding tube in paediatrics is extensive. The commonest indication for gastrostomy insertion is to overcome oral motor impairment and feeding difficulties in children with neurological impairment (predominantly CP). Insertion of a gastrostomy feeding tube is an increasingly common intervention in neurologically impaired children who have a clinically unsafe swallow; are unable to maintain a satisfactory nutritional state by oral feeding alone; have an inordinately long (> 3h/day) oral feeding time; and are dependent on nasogastric tube feeding (Sullivan et al. 2005). Nevertheless, children who have a percutaneous endoscopic gastrostomy tube inserted for the treatment of aspiration are twice as likely to be subsequently admitted to hospital than those fed orally; this is largely related to postoperative complication of the procedure (see below) (McSweeney et al. 2016).

Methods of Insertion

For much of the twentieth century the Stamm gastrostomy, which requires surgical laparotomy, was the most commonly accepted insertion technique. The Percutaneous

Endoscopic Gastrostomy (PEG) technique introduced in the 1980s has the advantage that it is minimally invasive, can be performed by a gastroenterologist, is relatively inexpensive and, if the patient's condition precludes use of a general anaesthetic, it can be performed under sedation. PEG tube insertion can be performed as a day case, preferably under general anaesthesia, and takes less than ten minutes. Nevertheless, children are often admitted to hospital for a period postoperatively for monitoring and initiation of feeds.

Shortly following the introduction of PEG, a percutaneous image-guided alternative to surgical and endoscopic gastrostomy placement, the percutaneous gastrojejunostomy (GJ) was introduced. In contrast to the open and PEG approaches, the radiological technique obviates a laparotomy incision or gastroscope, respectively, and is therefore considered the least invasive gastrostomy insertion technique (Ho 1983). Furthermore, image-guided gastrojejunal tubes may be a useful alternative to fundoplication and gastrostomy for neurologically impaired children with gastroesophageal reflux (Wales et al. 2002). Nevertheless, GJ necessitates a continuous feeding regimen, requires additional procedures if the tube becomes dislodged or has to be repositioned, and in rare circumstances can cause a life-threatening intussusception (Livingston et al. 2015). Despite being less invasive, this relatively blind approach has been associated with unique complications including placement of the catheter through a lobe of the liver and fistulation into the small bowel. This technique is not often used in children as the evidence base is so poor although antegrade percutaneous fluoroscopically guided gastrostomy is growing in popularity (Nah et al. 2010).

In 1990, laparoscopic assisted gastrostomy (LAG) placement was introduced, combining the minimally invasive advantages of PEG with the safety of the open procedure allowing for tube placement under direct visualisation. PEG is associated with an increased risk of major complications when compared to the laparoscopic approach. Advantages in operative time appear outweighed by the increased safety profile of laparoscopic gastrostomy insertion (Baker et al. 2015). Hansen (2017) also found that postoperative complications (mostly minor and which occurred in over half of the children in their study) were more common following the pull-through technique than with the laparoscopic approach (Hansen et al. 2017). Nevertheless, a less clear-cut situation emerged from a systematic review and meta-analysis which failed to find sufficient evidence on which to base a firm recommendation on the choice between the PEG and LAG techniques (Suksamanapun et al. 2017).

Skin level 'button' gastrostomy tubes provide an easy and comfortable approach to enteral nutrition. Development of a single-stage percutaneous technique for gastrostomy button insertion rather than the two-stage technique is popular in some centres.

An antireflux procedure (i.e. fundoplication) to decrease risk of aspiration due to gastric reflux may be done simultaneously with surgical gastrostomy although such an approach

should not be routine (Langer et al. 1988; Wheatley et al. 1991; Puntis et al. 2000; Kakade et al. 2015; Aumar et al. 2018). Current guidelines and reviews do not provide evidence to support superiority between either fundoplication/gastrostomy (FG) or GJ feeding in children with neurological impairment (NI) who have gastroesophageal reflux disease unresponsive to medical management; the risk of early and late major complications appears to be higher with FG whereas the frequency and burden of tube changes appear to be greater with GJ (Livingston et al. 2015).The gastrostomy tube in a child with a fundoplication can be changed without sedation at home or in clinic, while changing a gastrojejunal tube requires conscious sedation and a procedure in hospital.

Benefits of Gastrostomy Tube Feeding

In children with NI, gastrostomy placement has been shown to significantly increase weight gain and to be associated with a reduction in all of the following: feeding time, drooling, feed-related choking episodes, vomiting and frequency of chest infections (Rempel et al. 1988; Sullivan et al. 2000, 2005; Samson-Fang et al. 2003; Marchand & Motil 2006; Romano et al. 2017). Malnourished children with severe CP show significant increases in body fat with gastrostomy tube feeding (Sullivan et al. 2006; Vernon-Roberts et al. 2010). Such children have a rapid response to nutritional support through gastrostomy with catch-up growth regardless of age, even though there is a more pronounced state of malnutrition as age increases. Furthermore, death rates are distinctly higher in the subgroup of children with the most pronounced state of malnutrition and multiple secondary chronic conditions before gastrostomy.

Anecdotal reports in different studies have suggested that early developmental progress, pubertal development and emotional temperament improved following gastrostomy feeding but this needs more detailed research.

Family stress is significantly reduced and quality of life of parents increases after gastrostomy insertion to assist feeding (Mahant et al. 2009). Parents spend less time on child care once tube feedings are initiated and find feeding less difficult. This leads to evidence of caregiver satisfaction with gastrostomy tube feeding in the majority of studies (Nelson et al. 2015).

Complications of Gastrostomy Tube Feeding

It is difficult to make meaningful statements about risks and complications from the published data because types and rates of complications are not reported in a standard way and some children experience multiple complications. Moreover, reports on complications related to PEG are hampered by the lack of consensus regarding the definition of complications and because complications can occur several months after placement. There is, therefore, a need for agreement on the definition of complications

related to gastrostomy insertion (Hansen et al. 2017). Insertion of a gastrostomy feeding tube carries with it a relatively low risk of complications. Published literature suggests a procedure-related mortality of 1%, a major complication rate ranging from 6% to 12% and a minor complication rate of at least 50% (Hansen et al. 2017).

Reported major complications of gastrostomy insertion include adverse anaesthetic events, oesophageal laceration, pneumoperitoneum, peritonitis, colonic perforation and cologastric fistula formation. Many of these complications are now avoided or reduced in likelihood by refinements to the technique of insertion.

Later complications include stoma leakage, cellulitis, granulation tissue formation around the gastrostomy site and displacement. Gastrostomy site infection is the commonest problem occurring in up to 20% of cases but is easily and successfully treatable. More serious later complications such as bowel obstructions, gastrointestinal bleeds, ulceration and peritonitis are rare. Other later gastrointestinal complications include constipation, diarrhoea, cramping and vomiting. Although in the majority of cases PEG placement does not induce symptomatic GOR, it may worsen GOR and necessitate the use of antireflux medication or surgery.

Recent evidence suggests that half of children develop delayed gastric emptying after laparoscopic gastrostomy; this increases the risk of postoperative leakage and feeding intolerance and may be associated with GOR (Franken et al. 2017). This study, which was the first prospective study of delayed gastric emptying before and after gastrostomy tube placement, did not however demonstrate a worsening of GOR following gastrostomy tube insertion.

Death rates following gastrostomy range from 14% (after 1 year) to 26% (after 5 years). Most workers concur that these death rates are indicative of the severe morbidity (usually related to chronic secondary conditions including oesophagitis and lung disease from repeated pneumonias) in the children before gastrostomy.

What to Feed with Enteral Tubes

A consideration of what type of feed to use with enteral tubes is beyond the scope of this chapter and has been dealt with in detail in Chapter 9. Suffice to say that against a background of published evidence to support the efficacy of standard polymeric tube feeding for children with NI (Dipasquale et al. 2018), the current topic of debate concerns the use of home blended diets versus proprietary enteral feeds (Breaks et al. 2018). At the present time, authorities recommend caution when use of home blended diets is considered (European Society for Paediatric Gastroenterology, Hepatology and Nutrition; British Dietic Association) as concerns continue over the nutritional adequacy and safety of such diets (Coad et al. 2017; Romano et al. 2017).

Risk of Overfeeding

Immobile children with spastic four limb CP who are exclusively fed by gastrostomy tube grow consistently on an energy intake of less than 7kcal/cm, that is, diets ranging from 500–1100 kcal/day. Remarkably, this intake is 16–50% less than the recommended daily allowance. These extremely low energy intakes often make doctors, nurses and dietitians hesitant to accept the adequacy of such diets. The consequences of this may be overfeeding with the attendant risk of excessive fat storage. Use of high energy proprietary enteral feeds in children with CP fed by gastrostomy tube exacerbates the risk of overfeeding and has a potentially adverse effect on body composition (Sullivan et al. 2006). Conversely, low energy but nutritionally complete enteral feeds can produce weight gain without excess fat deposition (Vernon-Roberts et al. 2010).

Timing of Gastrostomy Insertion

Given what we now know about the benefits of gastrostomy tube feeding, there is a remarkable variation in its application across Europe with usage in over two thirds of children with CP (Gross Motor Function Classification System levels IV–V) in western Sweden but only 12% in such children in Portugal (Dahlseng et al. 2012a). This variation in usage was accompanied by variation in degree of growth retardation with those in Sweden being the least and those in Portugal being the worst affected. There was similar variation found in age at gastrostomy tube placement ranging from 16 months of age in Sweden to 70 months of age in northern England.

As increasing numbers of children with CP are undergoing gastrostomy insertion and are now surviving beyond childhood into adulthood, the question arises as to whether optimum long-term benefit could be achieved by gastrostomy placement at an early stage within this patient group. The evidence from several studies is that attainment of minimum growth standards occurs more frequently in children treated early before malnutrition and morbidity become established (Martinez-Costa et al. 2011; Sharma et al. 2012). Early nutritional supplementation by gastrostomy results in improved linear growth and weight-for-age z-score in children with severe CP if commenced early in life (less than 18 months) (Marchand & Motil 2006). Conversely, morbidity and mortality are most prevalent in those who receive gastrostomy later in life, that is, children treated 8 years or more after central nervous system insult.

Exhaustion with feeding as a neonate and persisting feeding difficulties at 6 months are associated with more severe neurodevelopmental and growth impairments in school age children with CP (Hawdon et al. 2000; Strand et al. 2016; Jadcherla et al. 2017). These observations may help identify those children with CP who may benefit from early assessment and interventions including gastrostomy insertion.

Recent evidence from Norway is that, although the prevalence of gastrostomy tube use there is increasing, only two thirds of children with significant feeding problems are tube fed, which possibly suggests that too few children have this treatment. Dahlseng and colleagues found that children with poor motor and speech function were older at placement of the gastrostomy tube and that the age of placement of a gastrostomy tube had an inverse effect on growth status. Some children have their gastrostomy tube inserted too late to achieve optimal growth and health (Dahlseng et al. 2012b).

Later insertion of gastrostomy may relate to parents' concerns of complications such as reflux-related respiratory illness in the most severely affected children and abhorrence of the idea of gastrostomy feeding (see below). Nevertheless, as noted above, most studies show that that the majority of parents would have agreed to earlier gastrostomy feeding of their children had they acknowledged its benefits.

Parents' Perceptions

Application of a strict biomedical model with emphasis on growth and symptoms is likely to neglect parental concerns about gastrostomy tube feeding. Craig approached this issue from a feminist poststructuralist perspective: she notes 'women are faced with enormous responsibility for making decisions about a child's nutritional management but may feel almost powerless in the face of uncertainty as to whether or not their child will benefit from a gastrostomy tube, and if so, at what cost?' (Craig & Scambler 2006). Such factors can significantly influence quality of life outcomes.

The severity of the disease as well as the presence and severity of malnutrition both have a negative impact on quality of life (QoL). Gastrostomy feeding is effective in reversing malnutrition in children with NI and has positive effects on the QoL of the patients and the caregivers. QoL is often the most important outcome of treatment for chronic conditions such as NI (Bjornson & McLaughlin 2001). Children with NI have reduced health-related QoL and the degree to which it is reduced is related to the severity of their NI (Samson-Fang et al. 2002; Vargus-Adams 2005). Almost 50% of children with NI are able to self-report their perceptions of their health-related QoL. Healthcare professionals and parents should, therefore, rely on a proxy report only when the children are not capable of self-report, or to ascertain potential differences in perceptions between children and their parents (Varni et al. 2005). Nevertheless, for severely disabled children parent-proxy reported QoL are the only available data. Caring for a child with NI affects a parent's physical well-being, social well-being, freedom, independence, family well-being and financial stability (Davis et al. 2012). Carers of children with NI have poor QoL, worse mental health and higher burnout levels than controls (Basaran et al. 2013). Parents often feel unsupported by the services they access (Craig et al. 2003). Children with the most severe motor disability who have feeding tubes are an especially frail group, that is, having the poorest health, the worst well-being and using the most

health-related resources (Liptak et al. 2001). Previous qualitative studies have found issues of social isolation, difficulty in obtaining care and high caregiving demands among parents of children with NI who are fed through a gastrostomy tube (Thorne et al. 1997; Craig & Scambler 2006).

Gastrostomy tube placement is often delayed because of negative caregiver perceptions. This delay may occur despite multiple hospital admissions for respiratory infections due to aspiration. Paradoxically, these parents are often less tentative about allowing other more invasive procedures such as orthopaedic surgery.

The decision making process for parents when a gastrostomy is first proposed is characterised by conflict (Mahant et al. 2018, 2011). Multiple negative perceptions may coexist in varying degrees. Despite being a struggle, mothers may view feeding by mouth as an enjoyable activity and an important social process. Mothers may feel guilty about their child's poor growth and may perceive the recommendation that gastrostomy feeding is required as confirmation of failure and a disruption of maternal nurturing and bonding. For some mothers gastrostomy feeding represents a loss of normality and may be seen as a confirmation of the permanence of the disability (Thorne et al. 1997; Petersen et al. 2006; Craig et al. 2006). In addition, the loss of oral feeding may be seen as a denial of a basic or essential human pleasure. Fears about loss of normal eating, dependency on gastrostomy feeds, complications of the procedure and the like can make parents very resistant to the idea of gastrostomy tube feeding and, even if they agree, they may choose to use the gastrostomy only as a last resort.

Nonetheless, the majority of caregivers recognise improvement in the children following placement and show high levels of satisfaction (Brotherton et al. 2007; Åvitsland et al. 2012; Nelson et al. 2015). Importantly, the majority admit that they would have accepted an earlier placement of the gastrostomy tube had they anticipated the overall outcome. Both management of the affected child and family relationships are usually accepted as having improved considerably when the feeding difficulties are ameliorated by gastrostomy insertion.

Using a validated instrument for measurement of QoL, Sullivan et al. (2004) found a significant improvement in the QoL of carers 6 and 12 months after insertion of a gastrostomy feeding tube in children with CP (Sullivan et al. 2004), A clear need for additional support for parents of children with a PEG has been identified that goes beyond simply meeting clinical need. Ongoing medical and psychosocial support is needed after initiation of non-oral feeding and is best provided through the collaborative efforts of the family and a team of professionals (Adams et al. 1999).

Interaction with healthcare professionals in relation to the proposition to introduce gastrostomy tube feeding is, however, often characterised by poor communication, lack

of information, paternalism and a lack of caregiver participation in decision making (Brotherton et al. 2007). It is clear from the foregoing that a great deal of sensitivity to the fears and feelings of the parents is required when approaching the subject of gastrostomy tube feeding. Parents need detailed information about gastrostomy feeding and support during decision making without pressure from family and healthcare professionals. Understanding these perceptions will help healthcare workers to develop effective, family-centred, patient appropriate intervention and adherence strategies for gastrostomy fed children with CP. All members of the multidisciplinary team should be well informed about the indications for and advantages and disadvantages of tube feeding so that a consistent message is conveyed to parents.

ETHICAL ISSUES

In an update of an earlier systematic review (Sleigh et al. 2004) which found no evidence of sufficiently high quality to merit inclusion, Sanchez (2016) remarked on the unfortunate fact that no randomised control trials comparing the outcomes of gastrostomy and oral feeding for children with CP were conducted in the 9 years between the publication of two Cochrane Reviews (Gantasala et al. 2013; Sanchez et al. 2016). The practical challenges inherent in conducting randomised control trials in this field are considerable. Sleigh and colleagues point out the potential 'complexity and costs of organizing what would necessarily be a large multi-center RCT with power sufficient to detect moderate-sized trial effects' (Sleigh et al. 2004). Furthermore, there are significant ethical challenges in randomising children to treatment groups when this intervention is already widely provided. This issue is poignant given that providing or withholding the intervention may have a significant impact on participants' growth, health and neurodevelopment and that the intervention may have significant side effects, as outlined above (Samson-Fang et al. 2003; Sullivan et al. 2005; Craig et al. 2006).

PEG feeding for reversing malnutrition in children with NI is a therapeutic intervention and as such is governed by standard ethical rules. The decision on initiation of the treatment is based on the likely net balance between advantages and disadvantages in order to promote the best interest of the individual patient. As is the case in other treatments, informed and educated consent of the parents is an important ethical principle. The parents need to be given detailed information on the benefits, risks and alternatives of the treatment and also, enough time to consider the information in order to make a conscious decision. Healthcare workers need to develop effective, family-centred, patient appropriate adherence strategies for gastrostomy fed children with NI. Furthermore, education and training on gastrostomy feeding, both in hospital and in the community, helps the carers of patients to cope during the transition from oral to gastrostomy feeding while continuing social support is essential in order to improve QoL of carers.

PEG feeding is associated with several complications and is costly. Gastrostomy feeding has been shown to reverse malnutrition (Sullivan et al. 2005) and to reduce the number of feed-related choking episodes, vomiting and chest infections. However, the decision making process for parents is often difficult because of negative caregiver perceptions (Petersen et al. 2006) and the gastrostomy tube placement is often delayed (Mahant et al. 2011). Nonetheless, the majority of caregivers recognise improvement in the children following placement and the majority admit that they would have accepted an earlier placement of the gastrostomy tube if they had anticipated the overall outcome (Petersen et al. 2006; Martinez-Costa et al. 2011).

SUMMARY

Malnutrition should not be considered normal in children with CP. The Nutrition Committee of the Canadian Pediatric Society declared that 'it is unacceptable not to treat undernutrition associated with neurodevelopmental disability' (Canadian Pediatric Society 1994). Nutritional intervention should be provided by a multidisciplinary team of professionals to ensure adequate growth, improve quality of life and optimise functional status. Early intervention, ongoing support and continuing follow-up are necessary to safeguard adequate growth and nutrition. One of the most important decisions to be taken in managing a child with severe CP and feeding difficulties is whether and when to insert a gastrostomy feeding tube.

Samson-Fang et al. critically appraised the effects of gastrostomy feeding in children with CP and concluded that the evidence, although limited, was generally in favour of the intervention (Samson-Fang et al. 2003). However, the review was based on only 10 published studies, largely comprising case series with relatively small samples and without a control (i.e. strength of evidence level IV). A systematic review failed to identify any trials that met the review criteria and concluded that there was continued uncertainty about the effects of gastrostomy (Sleigh & Brocklehurst 2004). Most studies have concentrated on severely affected children and are frequently flawed by a lack of valid and repeatable methods for assessing linear growth and body composition in this population.

Existing studies have focused primarily on the impairment domain measuring surgical and anthropometric outcomes. Evidence from the most comprehensive case series reported improvements on all weight- and growth-related outcomes (weight, head growth, linear growth, arm circumference and skinfold thickness). In fact, all studies to date report significant weight gain after gastrostomy with the implicit comparison being no weight gain without intervention (Ferluga et al. 2014). Some children remain underweight after intervention but given a lack of appropriate reference standards for the CP population, these results should be interpreted cautiously. Few studies have encompassed considerations about the broader health, psychosocial and economic aspects of gastrostomy feeding in children with CP.

Although the hierarchy of research evidence underpinning the use of gastrostomy in children with neurodevelopmental problems may not be strong (Ferluga et al. 2014; Sanchez et al. 2016) it is important to appreciate that one of the central tenets of 'evidenced-based medicine' is that evidence alone is never sufficient to make a clinical decision. A sound clinical decision is based upon the integration of the best available research evidence with clinical experience and patient values. This will involve a trade-off between benefits and risks, inconvenience and costs and the concerns, preferences and expectations of the patient/carer. When these elements are assimilated healthcare professionals and parents of children with CP can form a 'therapeutic alliance' to optimise growth, health and quality of life.

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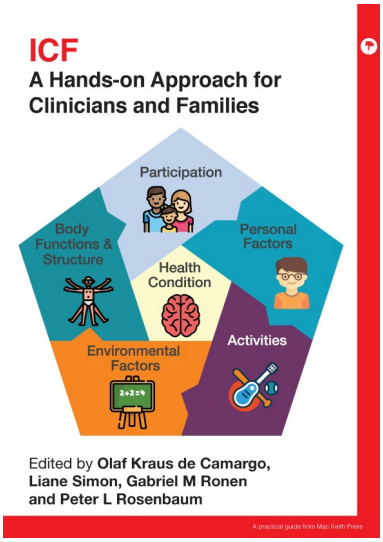

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