Chapter 1
Feeding and Nutrition in Neurodevelopmental Disability: An overview

Peter B Sullivan

Introduction
The feeding and nutritional problems encountered by children with neurological impairment have been overlooked until relatively recently. Much has been written about the diagnosis and management of children with cerebral palsy and marked progress has emerged from medical and technological advances, especially with respect to mobility, communication, education and orthopaedic care. Nevertheless, even as recently as the 1980s detailed accounts of the management of children with neurological impairment neglected to mention their feeding problems or the nutritional consequences of these. It is probable that the feeding problems and growth failure were considered to be an irremediable component of these children’s disorders. The central thesis in this volume is that this view is incorrect and that failure to feed and grow adequately has significant consequences for both the child and their parents and that these consequences are to a degree remediable.

The aim of this book, therefore, is to provide a framework for the multidisciplinary assessment and management of the feeding and nutritional problems in children with neurological impairment.

Oral motor impairment and swallowing dysfunction in children with neurological impairment
Oral motor impairment and swallowing dysfunction are a commonly associated disability in children with neurological impairment. Box 1.1 gives a summary of some of the more common conditions associated with oral motor impairment and swallowing dysfunction.
Box 1.1 Disorders of the central nervous system in children which may be associated with oral motor impairment and swallowing dysfunction

**Acute disorders of the central nervous system**
- hypoxic-ischaemic encephalopathy
- intracranial vascular events
  - infarction, haemorrhage
- infections
  - meningitis
  - encephalitis
  - poliomyelitis
- metabolic encephalopathies
  - aminoacidopathies
- trauma

**Chronic – static – disorders of the central nervous system**
- cerebral palsy
- genetic disorders
- Riley-Day syndrome
- kernicterus
- Arnold–Chiari malformation
- Möbius sequence

**Chronic – progressive – disorders of the central nervous system**
- intracranial malignancies
- degenerative conditions
  - lysosomal storage disease
  - metachromatic leukodystrophy
  - adrenoleukodystrophy
  - Leigh's encephalomyelopathy
  - neuroaxonal dystrophy
  - Rett syndrome
  - Wilson's disease
  - Zellweger's disease
- multiple sclerosis
- amyotrophic lateral sclerosis
- spinal muscular atrophy
- syringobulbia
Chapter 1 Feeding and nutrition in neurodevelopmental disability

Epidemiology – cerebral palsy
Cerebral palsy is the commonest form of neurodevelopmental disability and estimates place its prevalence at 2.4 per 1000 children aged 3–10 years. Recent data suggest that, contrary to initial expectations with improvements in perinatal medicine including the use of fetal monitoring and caesarean section, the prevalence of cerebral palsy has not decreased over the last 20 years. So, although survival in babies of 24–27 completed weeks of gestation has improved, the proportion of survivors with severe disability (25%) has not changed. Many survivors of neonatal intensive care 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. Moreover, against this background of an unchanged prevalence of disability, there is evidence that life-expectancy is increasing in people with cerebral palsy and that this may, in part, be related to improved nutritional care in recent years.

Epidemiology – feeding and nutritional problems
The literature on the incidence and prevalence of feeding difficulties mostly derives from small series reported from specialist hospital centres. More recently, robustly conducted epidemiological studies have provided an accurate measure of the extent to which children with cerebral palsy encounter feeding difficulties and have shown that this occurs in 30–40% of children sampled. The Oxford Feeding Study, for example, in the UK examined 271 children within a defined geographical region who had cerebral palsy and feeding problems and the results (Table 1.1) convey a sense of the range of difficulties faced by these children and their caregivers.

Table 1.1 Feeding and nutritional problems in relationship to the degree of motor deficit in children with cerebral palsy (adapted from Sullivan et al, 2000)

<table>
<thead>
<tr>
<th>Feeding/nutritional problem</th>
<th>Severity of motor impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
</tr>
<tr>
<td>Help with feeding needed</td>
<td>238/268</td>
</tr>
<tr>
<td>Choking with food</td>
<td>142/257</td>
</tr>
<tr>
<td>Feeding reported as stressful or not enjoyable by parent</td>
<td>51/262</td>
</tr>
<tr>
<td>Prolonged (≥3 hours/day) feeding times</td>
<td>71/258</td>
</tr>
<tr>
<td>Parents considered child underweight</td>
<td>93/240</td>
</tr>
<tr>
<td>Child received calorie supplements</td>
<td>23/271</td>
</tr>
<tr>
<td>Gastrostomy feeding</td>
<td>20/265</td>
</tr>
<tr>
<td>Never had feeding nutritional status assessed</td>
<td>169/264</td>
</tr>
<tr>
<td>Frequent vomiting</td>
<td>55/249</td>
</tr>
<tr>
<td>Bowels opened &gt; every 3 days</td>
<td>68/267</td>
</tr>
</tbody>
</table>
The great majority of children with cerebral palsy, for instance, required assistance with feeding and other concerns such as frequent choking and vomiting together with prolonged feeding time contribute to mealtimes which may often be an unpleasant and frightening experience for mothers and children alike.\textsuperscript{9}

The clear correlation between severity of feeding difficulties and the degree of motor impairment was also confirmed in the North American Growth in Cerebral Palsy Project.\textsuperscript{1} This project was a population-based multicentre study that aimed to describe parent-reported feeding dysfunction and its association with health and nutritional status in 230 children with cerebral palsy.\textsuperscript{1} The authors concluded that for children with moderate to severe cerebral palsy, feeding dysfunction is a common problem (occurring in around one-third of their sample) and that it was associated with poor health (more days ill in bed, hospitalization, missed school) and poor nutritional status. The problem is not confined to those with severe oral motor dysfunction alone, even children with only mild feeding dysfunction, requiring chopped or mashed foods, may be at risk for poor nutritional status.

Another feature revealed by the Oxford Feeding Study was that there was an apparent deficit in the degree of input into feeding and nutritional issues in the care of these children. Nearly two-thirds of caregivers of these children reported that they had never had their child’s feeding and nutritional state assessed. Table 1.2 (from the same study) shows that only 17% of children with cerebral palsy had contact with a dietitian in the previous 12 months. It is increasingly recognized now that early involvement of a multidisciplinary team is essential to prevent the adverse outcomes associated with feeding difficulties and poor nutritional status.\textsuperscript{10}

Feeding and nutritional therapy are time consuming and may not be available to some children who are at greatest risk for feeding dysfunction and subsequent malnutrition.

<table>
<thead>
<tr>
<th>Health care professional</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech therapist</td>
<td>74</td>
<td>27</td>
</tr>
<tr>
<td>Occupational therapist</td>
<td>47</td>
<td>17</td>
</tr>
<tr>
<td>Dietitian</td>
<td>47</td>
<td>17</td>
</tr>
<tr>
<td>School nurse</td>
<td>46</td>
<td>17</td>
</tr>
<tr>
<td>Hospital paediatrician</td>
<td>55</td>
<td>20</td>
</tr>
<tr>
<td>Community paediatrician</td>
<td>33</td>
<td>12</td>
</tr>
<tr>
<td>Family doctor</td>
<td>28</td>
<td>10</td>
</tr>
<tr>
<td>School doctor</td>
<td>23</td>
<td>8</td>
</tr>
<tr>
<td>Health visitor</td>
<td>12</td>
<td>4</td>
</tr>
<tr>
<td>Psychologist</td>
<td>8</td>
<td>3</td>
</tr>
</tbody>
</table>
Chapter 1 Feeding and nutrition in neurodevelopmental disability

The lack of support – especially from paediatric dietitians – for caregivers in dealing with feeding problems was also noted in the North American study\(^1\) which found that this may lead to significant stress in the family. This study and others have highlighted the emotional stress and adverse effect on caretaker quality of life posed by the difficulties in feeding a child with oral motor dysfunction.\(^1,11\) High levels of psychological distress have been recorded in mothers who have a child with neurological impairment, and the greater the degree of disability in the child the greater the distress in the mother.\(^12\) A common, and (as will be described in detail later in this book) potentially remediable, cause of this distress is prolonged feeding times related to oral motor dysfunction and inefficient feeding.

**Oral motor dysfunction and its consequences**

The relationship between oral motor dysfunction and growth retardation has been clearly documented.\(^13\)\(^{–15}\) Inefficient and slow feeding limits food intake and mothers spend prolonged periods of time (up to 8 hours a day in some cases) attempting to feed their disabled child.\(^16\) Although prolonged feeding may compensate for feeding inefficiency when the child is small, as body size increases a point is reached at which no further compensation is possible and growth is limited by energy intake. This issue will be dealt with in detail in Chapter 3. It is important to realize, however, that oral motor dysfunction is but one of a constellation of factors – impaired communication, immobility, medication, constipation and so on (see Figure 1.1) that lead to limited food intake in children with moderate to severe cerebral palsy. Nutritional impairment from limited food intake may be further exacerbated by excess losses following vomiting and gastro-oesophageal reflux. Gastrointestinal problems are encountered in around a third of children with cerebral palsy and will be dealt with in detail in a subsequent chapter.

The outcome of limited intake when this remains untreated is undernutrition and the associated short stature, low fat stores and reduced muscle mass have been well described.\(^17\)\(^{–19}\)

Figure 1.2 shows anthropometric data in children with different levels of severity of cerebral palsy; severity was graded as mild (little or no difficulty walking), moderate (difficulty walking but does not need aids or a helper), and severe (needs aids and/or a helper).\(^17\) These data reveal the significant adverse effect of cerebral palsy on linear growth and also the differential growth effect with the lower limb being more profoundly affected than the upper limb. This presumably relates to the importance of mobility and weight bearing in stimulating leg growth as the effect was significantly more marked in the group with severe motor impairment. Nevertheless, it is likely that nutritional status accounts for only 10–15% of the variability in linear growth of children with cerebral palsy.\(^20\) It is probable that genetic and neurohumoral influences exert a greater degree of influence on linear growth than nutritional status. Wasting is, however, predominantly nutrition related and prevalent in children with severe disabilities but in the Oxford Feeding Study was also observed at an individual level in those with mild and moderate mobility defects.
Problems with reference standards

The body composition of the child with severe cerebral palsy differs from that of the average child; a decrease in body cell mass accompanies an expansion of the extracellular fluid volume. The relative immobility of the child with severe cerebral palsy reduces fat free mass (largely muscle but also skeletal mass) as well as energy expenditure. The reduced energy expenditure of children with cerebral palsy is reflected in a lower dietary energy requirement – around 80% of current recommendations for children without neurological impairment. These differences in body composition and energy expenditure, therefore, mean that standard reference data for ideal nutritional input and optimal growth do not apply to children with cerebral palsy. This is a problem for those involved in the nutritional care of children with cerebral palsy because growth assessment requires reliable measures and comparison reference data. Only recently has an attempt been made to produce growth charts derived from observations made on clearly defined samples of children with cerebral palsy and which have been stratified by level of motor functioning and feeding ability.\textsuperscript{19,21} Appendix 1

Figure 1.2 Dotplot of z-scores by disability group. Line indicates median value for each disability group (a) bodyweight; (b) lower leg length
Figure 1.2 Dotplot of z-scores by disability group. Line indicates median value for each disability group (c) upper arm length; (d) head circumference z-score by disability group.
Chapter 1 Feeding and nutrition in neurodevelopmental disability

contains a series of growth charts for children with different levels of motor disability and for those who are also fed by gastrostomy tube. It is important to note, however, that these are descriptive in nature, i.e. they reveal how children with different levels of motor impairment actually grow – but they are not ‘reference standards’.

Nevertheless, the advent of these growth charts is welcome as the poor performance of standard reference charts in identifying malnourished cerebral palsy children has been recognized for a long time. Samson-Fang and Stevenson (2000), for instance, have shown that a triceps skin fold thickness (TSF) performs much more accurately than weight-for-height in identifying those with depleted fat stores and that a TSF below the 10th centile identifies 96% of malnourished children with cerebral palsy.22

Once malnutrition is identified the next problem is to decide how much to feed the child. The central consideration here is the amount of energy that the child requires to grow optimally. There is a wide variation in total energy expenditure (largely attributable to variations in physical activity levels) in immobile children with cerebral palsy. This individual variation, together with the lack of any suitable reference standards, compounds the difficulties in writing an accurate dietetic prescription. Fortunately there are some common sense ‘rules of thumb’ derived from experimental and clinical observations that can guide the clinical management of children with cerebral palsy. For instance, as the energy requirement for growth relative to maintenance is small (about 10 kj/g), satisfactory growth can be used as a sensitive indicator of whether energy needs are being met. It is surprising how little may be required to achieve this. Thus, exclusively gastrostomy-tube fed children with cerebral palsy grow consistently on an energy intake of less than 7 kcal/cm i.e. diets ranging from 500–1100 kcal/day which is 16–50% less than the recommended daily allowance.23 Such extremely low energy intakes often make paediatricians, nurses and dietitians hesitant to accept the adequacy of these diets. The consequences of this particularly in the gastrostomy fed child with cerebral palsy may be overfeeding and the risk of excessive fat storage.24

Treatment of suboptimal nutrition
Undernutrition has significant consequences for the child with cerebral palsy. There is hardly any physiological function which is not compromised by poor nutrition. Some examples of these will be described. Skeletal muscles are weakened further, which for the respiratory system renders the cough weak and predisposes the child (already at risk from aspiration secondary to oral motor dysfunction) to respiratory infection. The risk of infection is further increased as proper functioning of the immune system is compromised by undernutrition. Cardiac output is reduced and circulation time prolonged (Figure 1.3).

When the nutritional problems caused by feeding difficulty are properly addressed the consequences of undernutrition for the child are revealed. One of the first signs reported by parents is that the cold, pale and mottled condition of arms and legs (Figure 1.3) disappears as the limbs become pink and warm.25 Weight gain in the child is also
associated with an improvement in overall health and reduction in hospital attendances and frequency of chest infections have been reported following nutritional restitution.\textsuperscript{26}

Adequate nutrition is also associated with an improvement in levels of alertness and mood and parents report that the child is ‘brighter’. Some published reports have noted significant developmental progress in previously undernourished children with cerebral palsy accompanying improved nutritional status.\textsuperscript{27} This effect of nutrition on neurodevelopmental status is a feature that remains to be fully explored especially when children at risk are identified early and adequate nutritional management instituted before the onset of malnutrition.
Chapter 1 Feeding and nutrition in neurodevelopmental disability

Psychosocial considerations
The central thesis in this volume is that nutritional compromise from failure to feed adequately has significant consequences and is to a degree remediable and it has been approached largely from the standpoint of a biomedical model of health. There is a growing body of qualitative research which points to the need to incorporate a biopsychosocial component to health care especially when interacting with the parents of children with neurological impairment. Medical emphasis on growth and health may overlook parental concerns about oral and tube feeding. Mothers, who may harbour feelings of guilt about their child’s poor growth, can perceive the suggestion that gastrostomy feeding is required as confirmation of failure. Fears about loss of normal eating, dependency on gastrostomy feeds, complications of the procedure and so on can make parents very resistant to the idea of supplemental tube feeding and even if they agree they may opt to use the tube only as a last resort. Thus, a great deal of sensitivity to the fears and feelings of the parents is required when approaching the subject of gastrostomy tube feeding. Furthermore, all members within 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. In the experience of the author there is no better way to assist parents with the decision about whether or not to proceed with gastrostomy in their disabled child when it is clearly indicated than by introducing them to others in a similar situation that have experience of this intervention.

Recurrent pain and irritability
Another issue of great concern to caregivers is the occurrence in their disabled child of persistent irritability often ascribed to chronic or recurrent pain. Given that it is difficult to obtain reliable measures of the degree of pain or a clear indication of the source of pain in a child with a communication disorder, it is perhaps not surprising that this problem has not received much attention in the literature. This issue is covered here because such studies as have been undertaken point to the gastrointestinal tract as a commonly suspected source of recurrent pain. In one study, for instance, more than a third of children with severe cognitive impairment experienced pain that lasted for hours each week and the gastrointestinal tract was the commonest non-accidental source of pain. Results from the North American Growth in Cerebral Palsy project suggest that around 10% of caregivers report that their children experience pain on a regular basis. This study also demonstrated that pain was related to the severity of motor impairment and the presence of a gastrostomy feeding tube and taking medications for gastrointestinal symptoms (e.g. gastro-oesophageal reflux (GOR), motility problems and constipation). Manometric studies have shown that upper gastrointestinal sensory disorders contribute not only to abdominal discomfort in children with cerebral palsy but also to persistent feeding problems in these children. Visceral hyperalgesia may result from a range of sensitizing gastrointestinal disorders including GOR, fundoplication, gastrostomy insertion, chronic constipation coupled with the disordered cross-talk between the cerebral cortex and the enteric nervous system that occurs in children with cerebral palsy. A range of pharmacological approaches to modulate neurotransmission in the enteric nervous system (e.g.
imipramine, amitryptyline, cyproheptadine) or to inhibit central sensitization (e.g. gabapentin) have been shown to have therapeutic value.\textsuperscript{31,32}

References
Chapter 1 Feeding and nutrition in neurodevelopmental disability


