Feeding problems in merosin deficient congenital muscular dystrophy

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Abstract

Feeding difficulties were assessed in 14 children (age range 2–14 years) with merosin deficient congenital muscular dystrophy, a disease characterised by severe muscle weakness and inability to achieve independent ambulation. Twelve of the 14 children were below the 3rd centile for weight. On questioning, all parents thought their child had difficulty chewing, 12 families modified the diet, and 13 children took at least 30 minutes to complete a meal. On examination the mouth architecture was abnormal in 13 children. On videofluoroscopy only the youngest child (2 years old), had a normal study. The others all had an abnormal oral phase (breakdown and manipulation of food and transfer to oropharynx). Nine had an abnormal pharyngeal phase, with a delayed swallow reflex. Three of these also showed pooling of food in the larynx and three showed frank aspiration. These six cases all had a history of recurrent chest infections. Six of eight children who had pH monitoring also had gastro-oesophageal reflux. As a result of the study five children had a gastrostomy, which stopped the chest infections and improved weight gain. This study shows that children with merosin deficient congenital muscular dystrophy have difficulties at all stages of feeding that progress with age. Appropriate intervention can improve weight gain and reduce chest infections. The severity of the problem has not been previously appreciated in this disease, and the study shows the importance of considering the nutritional status in any child with a primary muscle disorder.

(*Arch Dis Child* 1999;**80**:542–547)

Keywords: congenital muscular dystrophy; feeding difficulties; aspiration; videofluoroscopy

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Accepted 9 February 1999

In recent years there has been increasing awareness of the feeding difficulties experienced by children with neurodevelopmental disability.[1 2] The majority of studies have been in children with cerebral palsy and many have been found to have major problems with eating and swallowing.[3–6]

Swallowing is a highly complex process[7 8] involving a total of 31 pairs of striated muscles, which are potentially affected by neurological and primary muscle diseases. During the oral phase food is prepared into a bolus by the action of the tongue, jaw, teeth, palate, and lips. The tongue then propels the bolus backwards until it reaches the posterior pharyngeal wall, triggering the swallow reflex. During the pharyngeal phase the bolus is propelled by the pharyngeal muscles to the oesophageal sphincter. The epiglottis closes and the laryngeal strap muscles pull the larynx upwards and forwards, protecting the airway from penetration with food. Respiration also ceases as the bolus moves towards the upper oesophageal sphincter. During the oesophageal phase, this sphincter relaxes allowing the bolus to move down into the stomach by peristalsis.

Feeding difficulties can be distressing for the individual and their family.[9] Prolonged mealtimes and concern about poor weight gain with continual emphasis on diet lead to psychological problems, especially as the child gets older. Lengthy mealtimes at school encroach on playtimes and affect peer relationships. Malnutrition not only leads to poor growth and delay in puberty but also affects the child's general health, including cognitive function.[10–12] Pharyngeal and oesophageal dysfunction can lead to aspiration and gastro-oesophageal reflux which may result in chronic pulmonary disease.

We were aware that some children in our muscle unit were failing to thrive. In the past this has been partially attributed to decreased muscle bulk. Light weight was thought to be an advantage in helping the child achieve mobility and in helping carers lift and transfer. In addition, many of the children had a history of frequent chest infections, which had been thought to be a result of poor respiratory muscle function.

Only when questioned directly about feeding did many parents and children express concerns. Our impression was that this was being overlooked in our clinical practice. We therefore studied 14 children, all with the muscle disease merosin deficient congenital muscular dystrophy, to assess feeding in more detail.

"Classic" congenital muscular dystrophy is an autosomal recessively inherited disease, presenting at birth or within the first six months of life, with hypotonia, muscle weakness, contractures, and motor developmental delay.[13] It has long been known that the clinical phenotype in this disease can vary substantially[13] and until recently there was no explanation for this finding. In 1994, a proportion of children with congenital muscular dystrophy was found to have a deficiency of an extracellular matrix protein called merosin (laminin α2 chain) on immunocytochemistry analysis of the muscle.[14] This extracellular matrix protein links to dystrophin inside the muscle cell through a transmembrane glycoprotein complex.[15] This discovery divided "classic" congenital muscular dystrophy into

two groups—merosin positive and merosin deficient. Although the disease is not progressive, children with merosin deficient congenital muscular dystrophy are severely affected and the creatine kinase is greatly increased, usually above 1000 iu/l.[16] They present at birth or by six months of age with hypotonia and delayed motor development. The muscle weakness is severe and maximal motor function achieved is usually standing with support.[16] This group was chosen for the study as they were followed every six months in the neuromuscular clinic and appeared to have poor weight gain.

Method

Clinical details of 14 children with merosin deficient congenital muscular dystrophy were examined. This represents our total population of affected children who are followed regularly in the neuromuscular clinic, with the exception of two children whose parents declined to take part in the study. Ages ranged from 2–14 years. The children have been followed every six months for the last six years by the same two clinicians (JP, FM). At each clinic visit weight has been recorded. Since commencing this study they have also been assessed on at least three separate occasions by the same speech and language therapist and dietician.

FEEDING HISTORY

A speech and language therapist obtained a detailed feeding history from all the families by questionnaire and then by direct questioning. They were asked about concerns over their child's weight, chewing and swallowing problems, dietary adaptations, and mealtime length. Symptoms of choking, coughing, vomiting, and recurrent chest infections were recorded. Finally, the family were asked about whether they found mealtimes a stressful or enjoyable experience.

EXAMINATION

The speech and language therapist performed an assessment of oral anatomy including lip, tongue, jaw, and palate structures and assessed function. Eight cases were directly observed eating a meal and the mealtime length recorded.

VIDEOFLUOROSCOPY

All 14 children had an abnormal feeding history examination and therefore proceeded to a videofluoroscopy. This is a dynamic radiological study of contrast medium passing from mouth to upper oesophagus that allows oral manipulation and swallowing efficiency with food of various consistencies, from liquids to solids, to be viewed.[17 18] The oral phase gives information about chewing and the ability to manipulate food into a bolus and transfer it to the bolus oropharynx to initiate swallowing. The pharyngeal phase shows the efficiency of bolus transfer through the pharynx and allows any penetration of the laryngeal inlet to be seen. All videofluoroscopies were analysed by the same speech and language therapist (AB).

Abnormal oral motor dysfunction included deficient buccal closure, causing drooling and abnormal tongue and jaw movements leading to difficulties with bolus formation and transfer to the oropharynx. Abnormalities in the pharyngeal phase included delayed onset of swallowing, nasopharyngeal penetration, reduced pharyngeal peristalsis, and failure in pharyngeal emptying. Residual food in the pharynx after the swallow increases the risk of aspiration. It also shows if the larynx is protected during the swallow. It will show if contrast penetrates the laryngeal inlet and in the child frankly aspirating it will show contrast passing below the vocal cords into the trachea.[19]

OESOPHAGEAL pH STUDIES

These were performed in 11 children who gave a history of chest infections, choking, vomiting, or abdominal pain. Three of the 11, however, did not tolerate the procedure (cases 2, 7, and 9) and therefore no results are available. Cases 6, 8, and 14 gave no history of respiratory symptoms, vomiting or recurrent abdominal pain and therefore did not have pH monitoring performed.

Results

When last assessed in clinic, before diet supplementation or gastrostomy, 12 of the children were below the 3rd centile for weight (table 1). One of them was born below the 3rd centile, eight were below the 3rd centile by three years of age, and three were below by six years. Seven of the 12 children had a growth chart similar to that of case 10 shown in fig 1, which shows the weight continuing to fall further away from the 3rd centile with age. In

Table 1 Data for 14 children with feeding difficultes, showing birth weight centile, present weight and centile, and feeding history

Case	*Birth weight centile*	*Age (years)*	*Weight (kg)*	*Weight centile*	*Modified food*	*Poor chewing*	*Meal time (mins)*	*Stress at meals*	*Choking*	*Chest infections*
1	< 3rd	2	8.65	< 3rd	No	Yes	60	No	Yes	Yes
2	10–50	3.5	10.94	< 3rd	Yes	Yes	60	Yes	Yes	Yes
3	90	3.5	11.52	< 3rd	Yes	Yes	60	No	Yes	Yes
4	3rd	4	11.4	< 3rd	Yes	Yes	30	No	No	Yes
5	3rd	4.5	11.25	< 3rd	Yes	Yes	30	No	Yes	Yes
6	3–10	7.5	7.1	< 3rd	Yes	Yes	60	Yes	No	No
7	3–10	7.5	16.96	< 3rd	Yes	Yes	60	No	Yes	Yes
8	3–10	8	15.84	< 3rd	Yes	Yes	60	Yes	No	No
9	10–50	8	27.4	50	Yes	Yes	30	Yes	Yes	Yes
10	10–50	10	14.68	< 3rd	Yes	Yes	60	Yes	No	Yes
11	10–50	11.5	22.95	< 3rd	Yes	Yes	60	Yes	Yes	Yes
12	90–97	12	15.72	< 3rd	Yes	Yes	15	No	Yes	Yes
13	10–50	13.5	24.45	< 3rd	Yes	Yes	60	No	Yes	Yes
14	3–10	14	47	50	No	Yes	45	No	No	No

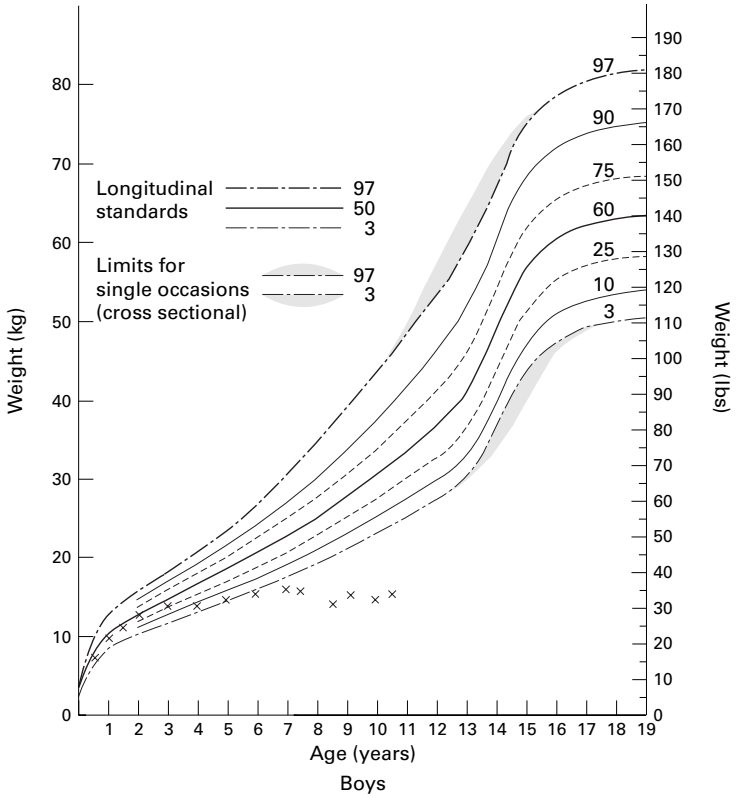

Figure 1 Weight chart of a boy (case 10) with merosin deficient congenital muscular dystrophy. Note the progressive failure to thrive.

the other five children, once the weight had fallen below the 3rd centile it continued to run below but parallel to this centile with increasing age.

FEEDING HISTORY AND CLINICAL EXAMINATION

All the children were assessed by a speech and language therapist. When directly asked, all parents reported that their child had difficulties chewing food and 12 families reported that they had to modify the food by mashing or liquidising it (table 1). All the children took at least 30 minutes for mealtimes except case 18, who only took 15 minutes to complete her meal. However, she had put no weight on from the age of 5 to 12 years.

A history of choking was obtained in nine children and all of them had more than three chest infections a year requiring antibiotic treatment. Chest infections throughout the group were common, with 11 children giving a positive history. Results of chest radiography were not available as the children in the study were appropriately seen at their local hospital during these acute respiratory episodes. Interestingly, despite the numerous feeding problems obtained on history and concerns about weight, only seven families perceived mealtimes as stressful.

On examination the three youngest children were either normal or had mild oral abnormalities (table 2). All the other children, including those with a normal weight, were very abnormal on examination, with poor lip closure, an anterior open bite, high arch palate, and reduced tongue movements. Eight children were observed feeding and the time taken to complete a meal corresponded to that reported by the parents.

VIDEOFLUOROSCOPY

Fourteen children underwent videofluoroscopy (table 3). Only the youngest child, aged 2 years, had a normal swallow. The other children had abnormal swallows, including two children with a normal weight (cases 9 and 14). All had an abnormal oral phase, showing impairment of chewing and bolus formation. They all also demonstrated difficulty propelling the food bolus to the oropharynx to initiate the swallow reflex.

The pharyngeal phase was abnormal in nine children. Once food reached the oropharynx the swallowing was delayed and the swallow function poor with numerous inefficient swallow attempts. This resulted in food residue coating the posterior tongue and pharyngeal wall, increasing the risk of late aspiration. The pharynx appeared to have become desensitised to food residue and no compensatory coughing or swallowing was seen to try to clear the food.

In six children, all with a history of chest infections, food was actually penetrating the laryngeal inlet. In three of them, contrast pooled in the larynx and coated the vocal cords, greatly increasing the risk of aspiration. In the other three, the swallow was definitely unsafe, with aspiration of food below the level of the cords into the trachea. In all three cases no cough reflex was triggered, indicating silent aspiration. These six children also showed reflux on pH monitoring, indicating the need to consider a Nissen fundoplication if planning gastrostomy insertion.

pH STUDIES

In the eight children who tolerated the procedure, oesophageal reflux was demonstrated in six.

CLINICAL COURSE AFTER INTERVENTION

In the children failing to thrive but with safe swallows—that is, no evidence of laryngeal pooling or aspiration on videofluoroscopy (cases 1, 2, 6, 7, 8, and 12)—diet supplementation to increase the energy concentration of ingested food was tried over at least a six month period. Initially they were given advice on increasing the energy density of the diet by increasing the fat content—that is, advising on

Table 2 Oral examination results

Case	*Weight centile*	*Lip closure*	*Bite*	*Palate*	*Tongue movement*	*FVC %*
1	< 3rd	Adequate	Open	Normal	Good	Not able
2	< 3rd	Poor	Open	Normal	Good	Not able
3	< 3rd	Adequate	Closed	Normal	Good	Not able
4	< 3rd	Poor	Open	High	Poor	35
5	< 3rd	Poor	Open	High	Poor	Not able
6	< 3rd	Poor	Open	High	Poor	39
7	< 3rd	Poor	Open	High	Poor	55
8	< 3rd	Poor	Open	High	Poor	16
9	50	Poor	Open	High	Poor	18
10	< 3rd	Poor	Open	High	Poor	11
11	< 3rd	Poor	Open	High	Poor	Not able
12	< 3rd	Poor	Open	High	Poor	20
13	< 3rd	Poor	Open	High	Poor	12
14	50	Poor	Open	High	Poor	59

FVC %, forced vital capacity % predicted for age, height, sex; Not able, children unable to perform test.

Table 3 Videofluoroscopy results

Case	*Oral phase*	*Pharyngeal phase*	*Laryngeal penetration*	*pH studies*	*Management*
1	Normal	Normal	None	Reflux	Feeding advice
2	Abnormal	Abnormal	None	Not tolerated	Feeding advice
3	Abnormal	Abnormal	None	Reflux	Gastrostomy
4	Abnormal	Abnormal	Pooling	Reflux	Gastrostomy
5	Abnormal	Abnormal	Aspiration	Normal*	Gastrostomy
6	Abnormal	Normal	None	Not done	Feeding advice
7	Abnormal	Normal	None	Not tolerated	Feeding advice
8	Abnormal	Abnormal	None	Not done	Feeding advice
9	Abnormal	Abnormal	Pooling	Not tolerated	Feeding advice†
10	Abnormal	Abnormal	Pooling	Reflux	Feeding advice†
11	Abnormal	Abnormal	Aspiration	Reflux	Feeding advice†
12	Abnormal	Normal	None	Normal	Gastrostomy
13	Abnormal	Abnormal	Aspiration	Reflux	Gastrostomy
14	Abnormal	Normal	None	Not done	Feeding advice

Not tolerated, pH study unsuccessful.
*pH study normal before gastrostomy; †gastrostomy discussed.

high fat alternatives to "healthy" low fat foods. They were also started on 1 kcal/ml nutritionally complete liquid supplements (volume depending on age), with powdered carbohydrate polymer added to drinks and occasionally sprinkled on solids. If no response in weight was seen after three months the energy density of the liquid supplements was increased—that is, 1.5 and 2 kcal/ml. One of the major difficulties with diet supplementation was compliance of the child who grew tired of each supplement after a few months. Advice was also given verbally and in writing about seating and positioning, and adaptive skills to assist feeding. Advice included taking small mouthfuls, limiting mealtimes to avoid tiredness, minimising distractions during feeding to allow the child to concentrate on eating, and making sure the mouth was empty after a meal. Advice was also given on always sitting upright during and for at least half an hour after feeding. No child, however, showed an increase in rate of weight gain and all remained below the 3rd centile.

As a result of the study we offered gastrostomy to seven children, either because of videofluoroscopic evidence of an unsafe swallow associated with recurrent chest infection (six children), or because of severe failure to thrive (one child), as described below.

Unsafe swallow on videofluoroscopy associated with recurrent chest infection

Cases 4, 5, and 13 had a gastrostomy because of a history of recurrent chest infections and evidence of an unsafe swallow on videofluoroscopy. All three cases also had a weight below the 3rd centile. The gastrostomy was performed under general anaesthesia in a hospital with postoperative paediatric intensive care facilities. The three cases highlighted the difficulty of whether also to perform a fundoplication at the same time. Case 4 had a gastrostomy two years ago. Videofluoroscopy showed that he was at risk of aspiration. Although his pH study showed mild reflux, it was felt that it was not sufficient to warrant a fundoplication, and since gastrostomy he has had no problems with vomiting. Cases 5 and 13 had gastrostomies 18 months and 12 months ago, respectively. Case 13 also had a fundoplication as the pH study showed reflux. Case 5 had a gastrostomy only as his pH study was negative. After three months, however, he developed severe oesophageal reflux, causing vomiting with weight loss, and therefore had a subsequent fundoplication. Since gastrostomy the chest infections have stopped in all three children and none has required hospital admission. The weight in all three cases has increased to the 10th centile.

Gastrostomy was also discussed with cases 9 and 10, who showed pooling of food in the larynx, and case 11, who was aspirating on videofluoroscopy. At present the families have elected not to proceed with gastrostomy. They were given feeding advice to minimise the risk of aspiration. As well as the usual information describe above, advice included thickening of food, avoiding foods with mixed consistencies, and encouraging the child to swallow or cough if food stuck in the throat rather than drinking fluid. Seating advice was strongly emphasised. Cases 10 and 11 were also given calorie supplementation.

Severe failure to thrive

Case 12 is the first case who has had a gastrostomy purely for poor weight gain (15–15.72 kg from 5–12 years). She had no history of chest infections and videofluoroscopy and pH studies were normal. We found it more difficult to recommend surgical intervention in the asymptomatic children with poor weight gain alone, as we had no evidence that gastrostomy would be effective in this group of children and the procedure was not without risk. Since insertion she has put on 2.5 kg in two months.

One other child (case 3) had a fundoplication for severe gastro-oesophageal reflux not responding to medical treatment at which time a gastrostomy was also performed. Her weight was also below the 3rd centile. She has not had a chest infection since the procedure and her weight has increased up to the 10th centile in six months.

Discussion

Our study has shown that feeding problems are frequent in merosin deficient congenital muscular dystrophy. Problems included difficulty chewing the food, abnormal swallowing of the bolus with or without aspiration, and gastro-oesophageal reflux. As a consequence of these difficulties the children's failure to thrive was notable and they had recurrent chest infections.

There are very few studies on feeding in children with a primary neuromuscular disease. Energy expenditure is much lower in patients with a disability who are not ambulant than in those who are.[20] Little is known, however, about the nutritional needs of patients with severe motor disability. In addition, the nutritional status of a child with a muscle disease is difficult to measure because musculoskeletal deformities make total length measurements impossible.[21] Estimates of body fatness by measurement of skinfold thickness in prepubertal children is associated with large errors at the individual level.[22] Problems also arise in measuring weight, as this includes muscle atrophy which may be caused by underlying disease rather than poor nutrition.

In this study 12 of 14 children with merosin deficient congenital muscular dystrophy were below the 3rd centile for weight. In seven of these children the rate of weight gain continued to be very poor, with the weight slipping further away from normal with age.

ORAL PHASE

This stage of feeding is dependent on both the structure and function of the oral cavity and both were frequently affected in our series of patients. Feeding difficulties in children are unique because their swallowing apparatus is growing, developing, and changing. Studies in children with cerebral palsy have found a high prevalence of oral motor dysfunction.[3 6] In our series the three youngest children had relatively normal oral examinations. After the age of 4 years, however, all children had developed abnormal mouth architecture. The anterior open bite allows spillage of food and greatly reduces the surface area for chewing. One previous study has also shown a high prevalence of open bite and delayed dentition in primary muscle disease.[23] Why malocclusion has developed in our series of children is not known. They are mouth breathers and this has been shown to cause abnormal mandibular growth and an open bite.[24] It is also possible that the facial muscle weakness influences jaw growth in childhood, resulting in malocclusion. As well as the structural abnormalities, dysfunctional, weak tongue movements makes manipulation of food difficult and poor lip closure impedes initiation of swallowing. Oral muscle weakness and the development of an abnormal mouth architecture in early childhood, combined with weaning and introduction of more solid foods that require chewing, may explain the increasingly poor weight gain with age seen in many of our patients.

All parents said their children had difficulty chewing and most had to modify the diet. Although some of the children had been followed for several years, this information was not volunteered until we specifically asked about diet and feeding. Supplements were tried in children with poor weight and safe swallows but without success. Their eating ability is probably impaired to a degree that they are unable to meet their energy needs even with energy supplements.

PHARYNGEAL PHASE

As well as difficulties during the oral stage of feeding with food manipulation and bolus formation, nine of the 14 children had notable pharyngeal impairment of food handling. Videofluoroscopy showed ineffective swallow reflex, poor pharyngeal peristalsis with prolonged transit time, and pharyngeal residue after completion of the swallow. Three of these children also showed penetration of contrast into the laryngeal inlet and three others were frankly aspirating with contrast entering the trachea. During the videofluoroscopy study no choking or coughing was observed in these six children, indicating silent aspiration. This finding highlights the difficulty of deciding whether a child is aspirating from the history alone.

OESOPHAGEAL PHASE

Gastro-oesophageal reflux was found in six of the eight children who had pH studies. One of the cases required a fundoplication with gastrostomy for persistent reflux not responding to medical treatment.

There are now several reports on the use of gastrostomies in children with a neurodisability.[25–28] It can be used to help weight gain when the child is failing to thrive and when the oral route is unsafe for all nutritional needs. As with case 5, many studies found that a high proportion of children fed via a gastrostomy tube develop oesophageal reflux.[27–29] A negative pH study result before gastrostomy does not guarantee that there will be no reflux once gastrostomy feeding is introduced. Some investigators therefore advocate performing a fundoplication at the same time as gastrostomy to prevent this complication.[30] Performing a fundoplication, however, makes the procedure more invasive.[31] This is an important consideration in children with merosin deficient congenital muscular dystrophy because of their severe weakness affecting respiratory function.

Two children in the study were not failing to thrive, despite clinically having the same changes in mouth architecture and function on examination. In fact, in early childhood case 14 had a weight below the 3rd centile but it then increased to the 50th centile when he was started on sodium valproate for epilepsy. We have no explanation for case 9 who is one of the most severely affected children, in terms of muscle function. He is the only one of the group totally unable to feed himself and his videofluoroscopy was very abnormal and yet his weight was on the 50th centile.

Our results show that children with merosin deficient congenital muscular dystrophy have notable feeding problems which were not apparent unless specifically inquired about and investigated. The vast majority of the children were failing to thrive and their frequent chest infections had previously been attributed to poor respiratory muscle function. We have now shown that these children have difficulties at all stages of the feeding process. The abnormal mouth architecture makes chewing and initiation of swallowing difficult and very time consuming. Videofluoroscopy showed that the pharyngeal phase was poor with inefficient swallowing. In six of the 14 children, all with a history of chest infections, contrast was also seen entering the laryngeal inlet. Oesophageal reflux was also common. As a result of this study three children with unsafe swallows have had a gastrostomy and in all cases it has stopped the chest infections. One child has had a fundoplication with gastrostomy for gastro-oesophageal reflux which has also stopped the chest infections and vomiting. A further child has had gastrostomy for poor weight alone which has been effective.

Although our results relate to one congenital muscle disease, congenital muscular dystrophy, they probably apply to the other congenital myopathies and even muscle disorders of later onset. Recognition and management of feeding

problems is important, resulting in improved quality of life and probably affecting prognosis.

This work was supported by the Muscular Dystrophy Group of Great Britain and Northern Ireland.

1 Jones PM. Feeding disorders in children with multiple handicaps. *Dev Med Child Neurol* 1989;**31**:404–6.
2 Nutrition Committee, Canadian Paediatric Society. Undernutrition in children with a neurodevelopmental disability. *Can Med Assoc J* 1994;**151**:753–9.
3 Gisel EG, Patrick J. Identification of children with cerebral palsy unable to maintain a normal nutritional state. *Lancet* 1988;i:283–6.
4 Reilly S, Skuse D. Characteristics and management of feeding problems of young children with cerebral palsy. *Dev Med Child Neurol* 1992;**34**:379–88.
5 Stallings VA, Charney EB, Davies JC, Cronk CE. Nutrition-related growth failure of children with quadriplegic cerebral palsy. *Dev Med Child Neurol* 1993;**35**:126–38.
6 Reilly S, Skuse D, Poblete X. Prevalence of feeding problems and oral motor dysfunction in children with cerebral palsy: a community survey. *J Paediatr* 1996;**129**:877–80.
7 Dodds WJ, Stewart ET, Logemann JA. Physiology and radiology of the normal oral and pharyngeal phases of swallowing. *AJR Am J Roentgenol* 1990;**154**:953–63.
8 Stevenson RD, Alliare JH. The development of normal feeding and swallowing. *Pediatr Clin North Am* 1991;**38**: 1439–53.
9 Douglas JE, Bryon M. Interview data on severe behavioural eating difficulties in young children. *Arch Dis Child* 1996;**75**:304–8.
10 Grantham-McGregor SM, Powell CA, Walker SP, Himes JH. Nutritional supplementation, psychosocial stimulation and mental development of stunted children: the Jamaican study. *Lancet* 1991;**338**:1–5.
11 Dobbing J. Early nutrition and later achievement. *Proc Nutr Soc* 1990;**49**:103–18.
12 Rosso P, Winick M. Relation of nutrition to physical and mental development. *Pediatr Ann* 1973;**2**:33–43.
13 Dubowitz V. *Muscle disorders of childhood*, 2nd ed. Philadelphia: WB Saunders, 1995.
14 Tome FMS, Evangelista T, Leclerc A, *et al*. Congenital muscular dystrophy with merosin deficiency. *C R Acad Sci Paris, Life Sciences* 1994;**317**:351–7.
15 Ervasti JM, Campbell KP. Membrane organization of the dystrophin glycoprotein complex. *Cell* 1991;**66**:1121–31.
16 Philpot J, Sewry C, Pennock J, Dubowitz V. Clinical phenotype in congenital muscular dystrophy: correlation with expression of merosin in skeletal muscle. *Neuromuscul Disord* 1995;**5**:301–5.
17 Jones B, Kramer SS, Donner MW. Dynamic imaging of the pharynx. *Gastrointestinal Radiology* 1985;**10**:213–24.
18 Dodds WJ, Logemann JA, Stewart ET. Radiologic assessment of abnormal oral and pharyngeal phases of swallowing. *AJR Am J Roentgenol* 1990;**154**:965–74.
19 Kramer SS. Radiologic examination of the swallowing impaired child. *Dysphagia* 1989;**3**:117–25.
20 Bandini LG, Schoeller DA, Fukagawa NK, *et al*. Body composition and energy expenditure in adolescents with cerebral palsy or myelodysplasia. *Pediatr Res* 1991;**29**:70–7.
21 Spender QW, Cronk CE, Charney EB, Stallings V. Assessment of linear growth in children with cerebral palsy: use of alternative measures to height and length. *Dev Med Child Neurol* 1989;**31**:206–14.
22 Reilly JJ, Wilson J, Durnin JVGA. Determination of body composition from skinfold thickness: a validation study. *Arch Dis Child* 1995;**73**:305–10.
23 Ghafari J, Clark RE, Shofer FS, Berman PH. Dental and occlusal characteristics of children with neuromuscular disease. *Am J Orthod Dentofacial Orthop* 1988;**93**:126–32.
24 Leighton BC. Aetiology of malocclusion of the teeth. *Arch Dis Child* 1991;**66**:1011–12.
25 Fried MD, Pencharz PB. Energy and nutrient intakes of children with spastic quadriplegia. *J Pediatr* 1991;**119**:947–9.
26 Wesley JR, Coran AG, Sarahan TM, *et al*. The need for evaluation of gastro-oesophageal reflux in brain-damaged children referred for feeding gastrostomy. *J Pediatr Surg* 1981;**16**:866–70.
27 Mago H, Chen CL, Wesson DE, *et al*. Incisionless gastrostomy for nutritional support. *J Pediatr Gastroenterol Nutr* 1986;**5**:66–9.
28 Hull MA, Rawlings J, Murray FE, *et al*. Audit of outcome of long-term enteral nutrition by percutaneous endoscopic gastrostomy. *Lancet* 1993;**341**:869–72.
29 Mollitt DC, Golladay S, Siebert JJ. Symptomatic gastro-oesophageal reflux following gastrostomy in neurologically impaired patients. *Pediatrics* 1985;**75**:1124–6.
30 Dedinsky GK, Vane DW, Black CT, *et al*. Complications and reoperation after Nissen fundoplication in childhood. *Am J Surg* 1987;**153**:177–83.
31 Fung KP, Seagram G, Pasiekka J, *et al*. Investigation and outcome of 121 infants and children requiring Nissen fundoplication for the management of gastro-oesophageal reflux. *Clin Invest Med* 1990;**13**:237–46.