RECENT ADVANCES

Nutritional support at home and in the community

J W L Puntis

Abstract

Technical developments in feeding, together with the growth of support structures in the community has lead to a steady increase in the number of children receiving home enteral tube feeding and home parenteral nutrition. In many cases the adverse nutritional consequences of disease can be ameliorated or prevented, and long term parenteral nutrition represents a life saving intervention. Careful follow up of children receiving home nutritional therapy is necessary to establish the ratio of risks to benefits. A considerable burden is sometimes placed on family or other carers who therefore require adequate training and ongoing support. The respective responsibilities of different agencies relating to funding and support tasks require more clear definition.

(*Arch Dis Child* 2001;84:295–298)

Keywords: home enteral tube feeding; home parenteral nutrition; review

Recognition that malnutrition and its adverse clinical consequences are potentially avoidable even in children with a range of severe and chronic disorders has given rise to the widespread use of nutritional support.^{1,2} Technical developments such as flexible fine bore feeding tubes³ and percutaneous endoscopic gastrostomy (PEG),⁴ together with emphasis on treatment out of hospital and in the community have produced a rapid expansion in the number of patients receiving home enteral tube feeding (HETF). For a much smaller group of children with intestinal failure, long term home parenteral nutrition (HPN) has become a life saving intervention.

Home nutritional support

The British Artificial Nutrition Survey (BANS) collates prospective data on home nutritional support in both adults and children from a large number of centres and produces regular reports under the auspices of the British Association of Parenteral and Enteral Nutrition (BAPEN). The August 1999 BANS report showed an annual increase in HETF of around 20%, together with a 5% increase in HPN.⁵ By the end of 1998, 2832 (26%) of the 10 864 BANS registered HETF recipients were children; for HPN the numbers were 64 (18%) and 284 and respectively. This is probably an accurate picture for HPN, but may represent only 90% of HETF patients in the

UK.⁵ Cerebral palsy was the most common diagnosis in children receiving HETF, representing 19% of those less than 15 years of age. Severe dysmotility disorders, protracted diarrhoea, and short bowel account for the majority of patients managed with HPN.

HPN and HETF tend to be long term undertakings, with 80–90% of patients still receiving nutritional support one year after initiation. Data from BANS⁵ show that over this period, among the children with cerebral palsy receiving HETF, 5% had died and only 4.7% returned to oral feeding. The figures were similar for patients with cystic fibrosis, while of those children with “congenital handicap”, 4.4% died and no more than 1.8% returned to oral feeding. A high proportion of children receiving HPN make the transition back to enteral feeding in due course. For example, among 300 patients in France only 30% were considered to be permanently dependent on parenteral nutrition (PN) (and therefore potential candidates for small bowel transplantation), while 23% required PN for less than two years.⁶ The successful re-establishment of full enteral feeding in a high proportion of children may be a reflection of the expertise acquired through the highly centralised French system whereby HPN is limited to only five centres.

BENEFITS OF HETF

It has been estimated that treating a patient on artificial nutrition at home instead of hospital provides a cost saving of 50% for HPN and 70% for HETF.^{7,8} In addition to cost savings, HETF confers a number of benefits including improvements in survival and general well-being.⁷ In children, growth can be used as a proxy for overall nutritional status. Promoting normal growth in some situations is extremely difficult, for example in young infants with chronic liver disease awaiting transplantation, many of whom remain severely malnourished despite HETF.⁹ Several recent studies have investigated the overall effectiveness of HETF in correcting or preventing growth failure in children referred to a nutrition team from a variety of different specialties. In a prospective study by Papadopoulou and colleagues,¹⁰ 44 consecutive patients who underwent HETF for a median period of six months were investigated. While heterogeneous in terms of underlying medical condition, just over half were being treated for malignancy. Patients fell into three similar sized groups with regard to indications for HETF: reversal of stunting, correction of wasting, and maintenance of

Neonatal Unit,
Clarendon Wing, The
General Infirmary at
Leeds, Belmont Grove,
Leeds LS2 9NS, UK
J W L Puntis

Correspondence to:
Dr Puntis
puntisj@ulth.northg.nhs.uk

Accepted 5 December 2000

nutritional status during treatment. Serial anthropometric measurements made during the study failed to show a significant improvement in height standard deviation score, although there was a trend towards improvement that correlated with duration of HETF. Wasting, however, significantly decreased, and growth was maintained in the third group. A similar but retrospective study from Canada in a more diverse group of children also showed that HETF produced significant weight gain but had little effect on height gain, perhaps because of the relatively short duration of follow up.¹¹

Given the large number of patients with neurological handicap receiving HETF, the suggestion by Strauss and colleagues¹² that tube feeding in such children is associated with increased mortality (possibly as a result of pulmonary complications) is of concern. However, the lack of information regarding clinical characteristics of individual patients, type of feeding tube, and surgical procedures undertaken makes it difficult to interpret the findings of this large retrospective survey. This emphasises the need for well designed prospective studies in order to establish the long term benefits and hazards of HETF in this group.

Disease modification and HETF

In many chronic disorders effective nutritional support does not necessarily modify the underlying disease process. This is not true for Crohn's disease, and paediatric gastroenterologists in the UK continue to use exclusion diets as first line management at presentation and for relapses.¹³ Dietary treatment is usually undertaken at home over a six to eight week period, with feeding by tube being reserved for those children who cannot comply with the required intake of liquid feed by mouth. Recent evidence suggests that by continuing supplementary enteral nutrition after disease remission, relapse can be delayed and linear growth improved.¹⁴ The incorporation into feeds of naturally occurring anti-inflammatory cytokines such as transforming growth factor β_2 may represent a further therapeutic advance in the management of Crohn's disease,¹⁵ although formal randomised trials have yet to be carried out. While animal studies have suggested that the addition of glutamine to enteral feeds suppresses mucosal inflammation, a small trial in children with Crohn's disease points to a lack of benefit.¹⁶ In cystic fibrosis, maintaining good nutrition is thought to have a positive effect on respiratory function. Overnight HETF via a PEG is well tolerated, results in significant improvements in nutritional status, and appears to stabilise pulmonary function, both in severely malnourished children and adults with advanced lung disease.¹⁷

GASTROSTOMY FOR HETF

Gastrostomy was once an open procedure carried out during routine neonatal surgery, but is now most commonly a percutaneous endoscopic technique performed to facilitate nutritional support. Experience with regard to the complications of PEG continues to grow. In a

retrospective survey of 130 PEG insertions at Great Ormond Street Hospital, complication rates were assessed with a view to highlighting those children most at risk.¹⁸ There were 51 complications judged to be major, the largest category being the development or exacerbation of gastro-oesophageal reflux disease (GORD). This occurred in 17 children, 10 of whom went on to have antireflux surgery. Nine patients developed significant stomal problems, eight peritonitis, and four gastrocolic fistula. Minor complications including external leak or infection occurred in almost a quarter of patients. The incidence of complications in children who had undergone previous abdominal or gastric surgery was found not to be increased. In a review of the limitations of PEG, Kimber and Beasley¹⁹ estimate that as many as one third of patients require a second PEG related operation including drainage of abscess, debridement of granulation tissue, or repeat endoscopy for flange removal. These reports emphasise that PEG insertion should not be regarded as a minor procedure, and that the potential risks and benefits be fully discussed with parents or care givers. Clearly insertion of a PEG should not be performed as an occasional exercise by someone wishing "to keep their hand in".

Children with neurodisability

Although PEG may provoke or exacerbate GORD (particularly in children with neurodisability), accurate prediction of who should have a fundoplication at the time of PEG is difficult. Boyle²⁰ has proposed that if a trial of supplemental nasogastric feeding produces adequate weight gain without clinical exacerbation of GORD for at least four weeks, PEG insertion does not need to be combined simultaneously with fundoplication. A recent retrospective study of PEG in children with neurological handicap but without clinically gross reflux endorsed this view.²¹ However, just over half of the 26 children in this study developed symptomatic GORD following PEG insertion, 12 then responding to medical treatment and two needing surgery. Prospective studies are required to define when an antireflux procedure should be performed in a gastrostomy fed child with neurological handicap.

For those children with severe GORD who have contraindications to major surgery, transpyloric rather than intragastric feeding can be given. Nasojejunal tubes, or gastrojejunal tubes passed via a gastrostomy are difficult to keep in place and a more reliable approach is to use a modified feeding Roux-en-Y jejunostomy.²² Recent publications have highlighted the problems faced by carers of disabled children receiving HETF,²³ including difficulty over access to education and social services.²⁴ Recommendations urged for the UK²⁴ include a set of national guidelines for local authorities to include the principles that disabled children are entitled to access to education, health, and social services, and that the respective responsibilities of different agencies relating to funding and support tasks should be clearly defined.

Other conditions

The inadvisability of gastrostomy tubes in some patients continues to be reassessed. Gastrostomy has generally been avoided in children with advanced liver disease, varices, or portal gastropathy. This is because of the risk of gastric haemorrhage and the potential problems of access to the abdominal cavity during liver transplantation. However, positive experience of PEG in a small group of children with chronic cholestatic disease has now been reported,²⁵ and it may be that PEG is a safe option in a selected number of children with liver disease such as Alagille syndrome not usually associated with early and severe portal hypertension. In Crohn's disease, although the use of more palatable whole protein in place of elemental liquid diets has reduced the need for tube feeding, anorexia or nausea may still frustrate treatment objectives. Concern over possible enterocutaneous fistula formation meant that nasogastric tubes have been used in preference to gastrostomy. In fact, it seems that PEG is safe and well tolerated in Crohn's disease, in addition to being more acceptable than nasogastric tube to some children.²⁶

Children with end stage renal disease often receive HETF, although in those undergoing peritoneal dialysis (PD) there have been concerns that gastrostomy would increase the risk of peritonitis. In a 15 year retrospective study of children receiving PD,²⁷ Ramage *et al* compared 23 children who also had gastrostomy feeds with a control group of 127 who did not. Gastrostomy was performed by open technique in nine and percutaneously in six. While there was a higher incidence of peritonitis in the children who had gastrostomy tubes compared with controls, this had also been true before insertion of gastrostomy. No significant difference in the incidence of PD catheter exit site infection was seen in the two groups, however, exit site infections were more common in those children who underwent insertion of gastrostomy after having started PD. The authors recommend gastrostomy tube insertion prior to instituting chronic PD.

Home parenteral nutrition

The organisation and benefits of HPN have recently been described by Meadows.²⁸ The long term nature of HPN offers opportunities for bringing to light complications associated with over or under provision of nutrients and then further refining feeding regimens. Life threatening complications of PN remain the principle indication for small bowel transplantation.²⁹

CHOLINE

Choline is an ammonium compound and a nutrient essential for mammalian cells. Low plasma choline concentrations have been described in adult HPN patients and are associated with abnormalities in liver function. In a study comparing HPN children with healthy controls,³⁰ plasma free choline was reduced in HPN patients and showed a steady decline with increasing age. A negative association was observed between plasma free choline and hepatic transaminases. Although some choline

is provided in the emulsifier portion of intravenous lipid emulsion, this appears to be insufficient to meet the needs of patients on long term PN with minimal enteral intake, and may be a factor in the development of liver disease. Guidelines on maintaining an adequate choline intake are now needed.

IRON

Iron requirements during long term PN are uncertain, but an intake of 100 µg/kg/day has been suggested.³¹ This intake, however, has been associated with iron overload and hepatic dysfunction.³² In some children with iron overload, iron status has been shown to improve if parenteral iron is withdrawn.³³ The optimum iron intake during long term PN remains unclear. Iron status should be monitored periodically in children receiving HPN with three monthly measurement of plasma ferritin concentration. Another contributor to PN associated liver disease has been identified as the plant sterols in lipid emulsion.³⁴ In vitro investigations have also pointed to an inhibitory effect of lipid emulsions on cholesterol uptake by hepatocytes.³⁵ Improvement in cholestasis may follow if intake of lipid emulsion can be reduced below 1.5 g/kg/day.³⁴

MANGANESE

Manganese is an essential trace element required for mucopolysaccharide production and the formation of normal bone, cartilage, and connective tissue. Manganese toxicity was described in children on long term PN presenting with liver and nervous system disorders.³⁶ This prompted modification of the trace element preparation by the manufacturers, resulting in a 50-fold reduction in manganese intake. Manganese binds to dopaminergic receptors in the basal ganglia and brain deposition can be seen on MRI scan. Recently it has been shown that in children with subclinical manganese toxicity, MRI scans return to normal over a number of years.³⁷ Whole blood manganese concentrations should be monitored in HPN patients each three months, particularly in the presence of liver disease, when biliary excretion is likely to be impaired.

THROMBOEMBOLISM

A further major concern related to long term PN is the risk of thromboembolic events. Over a 78 month period, 11 of 244 patients reported to the Canadian Childhood Thrombophilia Registry with central venous catheter associated thrombosis were receiving long term PN.³⁸ A paper from the largest UK children's HPN centre identified thrombosis or embolism in 12 of 32 patients, in whom there were four deaths.³⁹ Those children with autoimmune enteropathy appeared to be at increased risk, an observation that may be linked to the development of anticardiolipin and antiphosphatidylcholine antibodies during PN in this group.⁴⁰ There are no generally accepted guidelines for prevention and management of central venous catheter associated thrombosis. How far thromboembolism might be preventable by routine use of anticoagulants, and the

risks of this treatment, remains unclear at the present time. Some units now routinely anticoagulate those patients with underlying autoimmune disease. Ultimately, thrombosis may preclude venous access, although successful stenting of an occluded superior vena cava has recently been reported in one candidate for small bowel transplantation who was then able to continue with HPN.⁴¹

FOLLOW UP

Children who progress from HPN to full enteral feeding may still have abnormalities in growth and nutritional status. Leonberg and colleagues⁴² studied nine children from 2 to 6 years of age who had received PN for a mean of 14.6 months before having re-established full enteral nutrition for at least six months. One child was stunted and five had low total body fat; six had low serum vitamin A concentrations and four abnormal bone mineral density. Psychomotor development was normal in all nine. The authors emphasise the need for long term dietary, growth, and nutritional monitoring in children who have had HPN.

Conclusions

It seems likely that the number of children receiving home nutritional support (particularly HETF) will continue to increase as more aggressive efforts are made to prevent the adverse nutritional consequences of chronic disease. The balance of risks and benefits in some circumstances requires clarification by careful follow up. Training and support is needed for families and carers, including further development of hospital outreach services and cooperation with schools and social services in order to bridge the gap between home and hospital. There is an important role for a multidisciplinary nutritional care team⁴³ in coordinating such activity.

- Booth IW. Enteral nutrition in childhood. *Br J Hosp Med* 1991;**46**:111–13.
- Puntis JWL, Holden C. Home enteral nutrition in paediatric practice. *Br J Hosp Med* 1991;**45**:101–6.
- Sharpe G, Puntis JWL. Nutritional support at home: accessing the gut. *Clin Pediatr* 1997;**7**:213–17.
- Grunow JE, Chait P, Savoie S, et al. Gastrostomy feeding. In: David TJ, ed. *Recent advances in paediatrics*. 12. London: Churchill-Livingstone, 1994.
- British Association of Parenteral and Enteral Nutrition. *Report of the British Artificial Nutrition Survey (BANS), August 1999*. Maidenhead: British Association of Parenteral and Enteral Nutrition, 1999.
- Colomb V, Goulet O, Ricour C. Home enteral and parenteral nutrition in children. *Baillieres Clin Gastroenterol* 1998;**12**:877–94.
- Elia M, ed. *Enteral and parenteral nutrition in the community. A report of a working party of the British Association for Parenteral and Enteral Nutrition*. Maidenhead: British Association of Parenteral and Enteral Nutrition, 1994.
- Puntis JWL. The economics of home parenteral nutrition. *Nutrition* 1998;**14**:809–12.
- van Mourik IDM, Beath SV, Brook GA, et al. Long term nutritional and neurodevelopmental outcome of liver transplantation in infants aged less than 12 months. *J Pediatr Gastroenterol Nutr* 2000;**30**:269–75.
- Papadopoulou A, Holden CE, Paul L, et al. The nutritional response to home enteral nutrition in childhood. *Acta Paediatr* 1995;**84**:528–31.
- Kang A, Zamora SA, Scott RB, Parsons HG. Catch-up growth in children treated with home enteral nutrition. *Pediatrics* 1998;**102**:951–5.
- Strauss D, Kastner T, Ashwal S, White J. Tube-feeding and mortality in children with severe disabilities and mental retardation. *Pediatrics* 1997;**99**:358–62.
- Cosgrove M. Home enteral nutrition in childhood Crohn's disease. *Br J Home Care* 1999;**1**:23–7.
- Wilschanski M, Sherman P, Pencharz P, et al. Supplementary enteral nutrition maintains remission in paediatric Crohn's disease. *Gut* 1996;**38**:543–8.
- Fell JMA, Paintin M, Arnaud-Battandier F, et al. Mucosal healing and a fall in mucosal pro-inflammatory cytokine mRNA induced by a specific oral polymeric diet in paediatric Crohn's disease. *Aliment Pharmacol Ther* 2000;**14**:281–9.
- Akobeng AK, Miller V, Stanton J, et al. Double-blind randomized controlled trial of glutamine-enriched polymeric diet in the treatment of active Crohn's disease. *J Pediatr Gastroenterol Nutr* 2000;**30**:78–84.
- Williams SGJ, Ashworth F, McAlweenie A, et al. Percutaneous endoscopic gastrostomy feeding in patients with cystic fibrosis. *Gut* 1999;**44**:87–90.
- Khattak IU, Kimber C, Kiely EM, Spitz L. Percutaneous endoscopic gastrostomy in paediatric practice: complications and outcome. *J Pediatr Surg* 1998;**33**:67–72.
- Kimber CP, Beasley SW. Limitations of percutaneous endoscopic gastrostomy in facilitating enteral nutrition in children: review of the shortcomings of a new technique. *J Paediatr Child Health* 1999;**35**:427–31.
- Boyle JT. Nutritional management of the developmentally disabled child. *Pediatr Surg Int* 1991;**6**:76–81.
- Puntis JWL, Thwaites R, Abel G, Stringer MD. Children with neurological disorders do not always need fundoplication concomitant with PEG. *Dev Med Child Neurol* 2000;**42**:97–9.
- Gilchrist BF, Luks FI, DeLuca FG, Wesselhoeft CW. A modified feeding Roux-en-Y jejunostomy in the neurologically damaged child. *J Pediatr Surg* 1997;**32**:588–9.
- Spalding K, McKeever P. Mothers' experiences caring for children with disabilities who require a gastrostomy tube. *J Pediatr Nurs* 1998;**13**:234–43.
- Townsend R, Robinson C. What rights for disabled children? Home enteral tube feeding in the community. *Children and Society* 1999;**13**:48–60.
- Duché M, Habès D, Lababidi A, et al. Percutaneous endoscopic gastrostomy for continuous feeding in children with chronic cholestasis. *J Pediatr Gastroenterol Nutr* 1999;**29**:42–5.
- Cosgrove N, Jenkins HR. Experience of percutaneous endoscopic gastrostomy in children with Crohn's disease. *Arch Dis Child* 1997;**76**:141–3.
- Ramage IJ, Harvey E, Geary DF, et al. Complications of gastrostomy feeding in children receiving peritoneal dialysis. *Pediatr Nephrol* 1999;**13**:249–52.
- Meadows N. Home parenteral nutrition in children. *Baillieres Clin Paediatr* 1997;**5**:189–99.
- Reyes J, Abu-elmagd K. Small bowel and liver transplantation. In: Kelly DA, ed. *Diseases of the liver and biliary system*. Oxford: Blackwell Science Ltd, 1999:313–32.
- Misra S, Ahn C, Ament ME, et al. Plasma choline concentrations in children requiring long-term home parenteral nutrition: a case control study. *Journal of Parenteral and Enteral Nutrition* 1999;**23**:305–8.
- Greene HL, Hambidge KM, Schanler R, Tsang RC. Guidelines for the use of vitamins, trace elements, calcium, magnesium and phosphorus in infants and children receiving total parenteral nutrition: report of the subcommittee on Pediatric Parenteral Nutrition Requirements, from the Committee on Clinical Practice Issues of the American Society of Clinical Nutrition. *Am J Clin Nutr* 1988;**48**:1324–42.
- Ben Hariz M, Goulet O, De Potter S, et al. Iron overload in children receiving prolonged parenteral nutrition. *J Pediatr* 1993;**123**:238–41.
- Ben Hariz M, Goulet O, Colomb V, et al. Inappropriate iron intake in children on long-term parenteral nutrition: outcome after iron withdrawal. *Clin Nutr* 1997;**16**:109–12.
- Clayton PT, Bowron A, Mills KA, et al. Phytosterolemia in children with parenteral nutrition-associated cholestatic liver disease. *Gastroenterology* 1993;**105**:1806–13.
- Whitfield PD, Clayton PT, Muller DPR. Effect of intravenous lipid emulsions on hepatic cholesterol metabolism. *J Pediatr Gastroenterol Nutr* 2000;**30**:538–46.
- Fell JME, Reynolds AP, Meadows N, et al. Manganese toxicity in children receiving long-term parenteral nutrition. *Lancet* 1996;**347**:1218–21.
- Kafritsa Y, Fell J, Long S, et al. Long term outcome of brain manganese deposition in patients on home parenteral nutrition. *Arch Dis Child* 1998;**79**:263–5.
- Massicotte MP, Dix D, Monagle P, et al. Central venous catheter related thrombosis in children: analysis of the Canadian registry of venous thromboembolic complications. *J Pediatr* 1998;**133**:770–6.
- Dollery CM, Sullivan ID, Bauraind O, et al. Pulmonary embolism and long term central venous access for parenteral nutrition. *Lancet* 1994;**344**:1043–5.
- Berant M, Shah V, Ben-Barak A, et al. Thromboembolism and parenteral nutrition: association with intravenous fat and antiphospholipid antibodies. *J Pediatr Gastroenterol Nutr* 1995;**20**:453.
- Peters M, Beath SV, Puntis JWL, John P. Superior vena cava thrombosis causing respiratory obstruction successfully resolved by stenting in a small bowel transplantation candidate. *Arch Dis Child* 2000;**83**:163–4.
- Leonberg BL, Chuang E, Eicher P, et al. Long-term growth and development in children after home parenteral nutrition. *J Pediatr* 1998;**132**:461–6.
- Puntis JWL. Establishing a nutrition support team. *Baillieres Clin Paediatr* 1997;**5**:177–88.