Nutritional support at home and in the community

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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.

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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.

Technical developments such as flexible fine bore feeding tubes and percutaneous endoscopic gastrostomy (PEG), 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. 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.

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 BANS 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. 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. In addition to cost savings, HETF confers a number of benefits including improvements in survival and general wellbeing. 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. 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, 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...
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.\textsuperscript{13}

Given the large number of patients with neurological handicap receiving HETF, the suggestion by Strauss and colleagues\textsuperscript{17} 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.\textsuperscript{15} 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.\textsuperscript{14} The incorporation into feeds of naturally occurring anti-inflammatory cytokines such as transforming growth factor β\textsubscript{2} may represent a further therapeutic advance in the management of Crohn’s disease,\textsuperscript{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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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 gastrocutaneous fistulae. 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 Beasley\textsuperscript{19} 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. Boyle\textsuperscript{20} has proposed that if a trial of supplementary 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.\textsuperscript{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.\textsuperscript{22} Recent publications have highlighted the problems faced by carers of disabled children receiving HETF,\textsuperscript{23} including difficulty over access to education and social services.\textsuperscript{24} Recommendations urged for the UK\textsuperscript{24} 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, 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.

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, 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. 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.

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, 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. This intake, however, has been associated with iron overload and hepatic dysfunction. In some children with iron overload, iron status has been shown to improve if parenteral iron is withdrawn. 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. In vitro investigations have also pointed to an inhibitory effect of lipid emulsions on cholesterol uptake by hepatocytes. Improvement in cholestasis may follow if intake of lipid emulsion can be reduced below 1.5 g/kg/day.

MANGANISE
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. 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. 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. 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. 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. 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 colleagues 7 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 team in coordinating such activity.


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