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BAPEN Symposium 1 on 'Nutritional support in children and adolescents'

Managing children and adolescents on parenteral nutrition: challenges for the nutritional support team

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Managing infants, children and adolescents, ranging from premature infants to 18-year-old adolescents, on parenteral nutrition (PN) is a challenge. The ability of children to withstand starvation is limited and, unlike adults, children require nutrition for growth. PN in children is often required secondary to a congenital bowel problem rather than because of an acquired condition. Conditions requiring PN include motility disorders, congenital disorders of the intestinal epithelium and short-bowel syndrome (SBS). Intestinal failure may be temporary and children with SBS may be weaned from PN. However, other children require permanent PN. There are no comprehensive guidelines for the nutritional requirements of children and adolescents requiring PN. Practice in individual centres is based on clinical experience rather than clinical trials. Requirements are assessed on an individual basis according to age, nutritional status and clinical condition. These requirements need regular review to ensure that they remain appropriate for the changing age and weight of the child. Assessments of intakes use different methods, e.g. reference tables and predictive equations. Complications of PN include infection, accidental damage to, or removal of, the line and cholestatic liver disease. Home parenteral nutrition (HPN) is associated with fewer line infections and allows continuation of nutritional support in a more normal environment, encouraging normal development and participation in family activities. However, having a child at home on HPN is associated with physical and psychological stresses. A feeling of depression, loneliness and social isolation is common amongst children and their families. Home-care services are essential to supporting children at home and should be tailored to, and sensitive to, the individual needs of each family.

Parenteral nutrition: Intestinal failure: Children and adolescents

The need for parenteral nutrition results from the loss of normal intestinal absorptive function, and in childhood both acute and chronic intestinal failure can occur. In developing countries gastrointestinal infection is the commonest cause of acute intestinal failure, but in developed countries intestinal failure describing more chronic disturbances of intestinal function is more familiar.

Chronic intestinal failure may result from inherited conditions diagnosed in the early neonatal period, including motility disorders, such as long-segment Hirschprung's disease and chronic intestinal pseudo-obstruction, and congenital diseases of the intestinal epithelium, such as microvillous inclusion disease and tufting enteropathy.

Many children with motility disorders are permanently PN dependent and cannot be managed even with the most specialised enteral feeds. Long-segment Hirschprung's disease raises the same problems as short-bowel syndrome (SBS). However, the colon will never be fully functional and the remaining bowel, even if it is ganglionic, may be dysmotile. If the length of ganglionic bowel is <500 mm,

Abbreviations: HPN, home parenteral nutrition; PN, parenteral nutrition; SBS, short-bowel syndrome.
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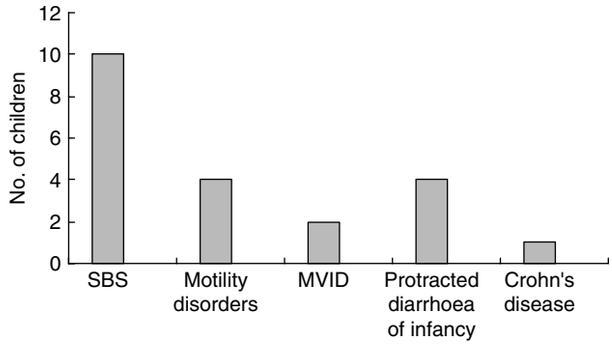

Fig. 1. Home parenteral nutrition by diagnostic group in Birmingham Children's Hospital during 2005. SBS, short bowel syndrome; MVID, microvillous inclusion disease.

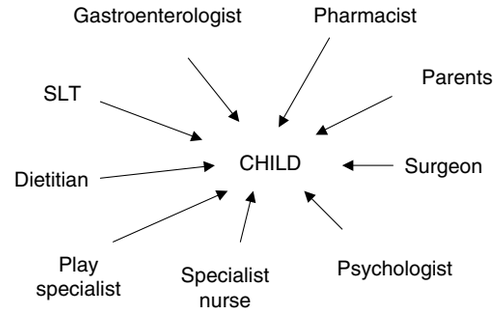

Fig. 2. The members of the paediatric nutrition team. SLT, speech and language therapist.

permanent PN is almost inevitable (Goulet *et al*. 2004). Some children with chronic intestinal pseudo-obstruction also require permanent PN, particularly those who have intractable vomiting. However, some children may survive on enteral feeds but require intermittent PN. Commonly, PN may be needed through their teenage years to maximise potential for linear growth and pubertal development.

Children with microvillous inclusion disease and tufting enteropathy will always require PN. In some children there is some intestinal function and enteral nutrition may be successful as an adjunct to PN. Extensively- or partially-hydrolysed enteral feeds may be helpful to maximise nutrient absorption.

SBS is also commonly seen in the neonatal period, but is an acquired condition resulting from extensive resection of the bowel. In the Birmingham Children's Hospital centre children with SBS are the largest diagnostic group of children on home PN (HPN; Fig. 1). Although children with SBS may require PN for long periods, unlike other chronic causes of intestinal failure in children there is potential for progression to full enteral nutrition. Managing this transition from PN to enteral nutrition can be a challenge, as progression can be both prolonged and unpredictable. The development of PN is the most important factor in the improved survival of children with SBS. PN assures adequate balanced nutrition to maintain hydration and nutritional status and to give time for intestinal adaptation to occur. Although PN provides essential fluid and energy, prolonged exclusive PN can lead to complications and the importance of giving 'PN' rather than 'total PN' has been learnt. Enteral feeds may be nutritionally inconsequential, but they are nevertheless very important because even minimal enteral feeds are an essential factor in preventing intestinal failure-related liver disease (Puntis, 2002). It is also known that intraluminal nutrients are the single most important factor in promoting intestinal adaptation in SBS and will promote pancreatic secretions and bile and may also help to prevent bacterial translocation (Goulet *et al*. 2004).

Managing infants, children and adolescents on PN is a challenge. First, it is important to remember that children are not just miniature adults. The patient age range is very wide. The nutrition team are responsible for providing nutritional support for children ranging from premature infants to 18-year-old adolescents. Compared with adults the ability of children to withstand starvation is limited; an adult will survive for 90 d whereas a premature infant will only withstand starvation for 4 d (Puntis, 2002). Perhaps the most important difference between adults and children is that children require nutrition to allow essential brain growth and linear growth.

The nutrition team

It is well established that the provision of PN to children requires the skills of a multidisciplinary nutrition team (Puntis & Booth, 1990; Sexton *et al*. 2000). An understanding of the individual roles within teams is essential. The team members will vary in individual centres, but core members should include an experienced paediatrician, pharmacist, dietitian and specialist nurse (Agostini *et al*. 2005). In a paediatric setting the expertise of a speech and language therapist, clinical psychologist and play specialist can also be valuable. If a child is to be discharged home on PN the additional support of community services is paramount, and parents who are taught the complex skills required for the safe administration of HPN also become important team members (Fig. 2).

Assessing requirements

There are no comprehensive guidelines for the nutritional requirements of children and adolescents requiring PN. The European Society for Paediatric Gastroenterology, Hepatology and Nutrition has produced guidelines (Koletzko *et al*. 2005). These broad guidelines reflect practice in individual centres across Europe, and are largely based on clinical experience rather than clinical trials. Requirements need to be assessed on an individual basis according to age, nutritional status and clinical condition, and need regular review to ensure that they remain appropriate for the changing age and weight of the child. Assessment of requirements can be based on reference tables or predictive equations but should also include anthropometric assessment of nutritional status.

There is no ideal method of assessing the energy requirements for a child on PN. The Department of Health (1991) and Food and Agriculture Organization/World Health Organization/United Nations University (1985,

2004) provide guidelines for intakes based on both energy per kg body weight and total daily energy intake. However, these recommendations refer to groups and should not be applied too closely to individuals, but serve as a useful reference point. Requirements vary with weight, age and gender, and reference tables adopt 'standard age-groups' with a calculated average or 50% centile weight for both male and female children. There is an assumption made when using these recommendations that a child fits into these standard groups, and it is therefore essential that a careful assessment of an individual child is made to reduce the risk of providing either an inadequate or excessive nutritional intake. An important role of a dietitian is to use their assessment skills and knowledge of nutritional requirements to ensure that every child receives optimum PN and that PN is regular reviewed in relation to a child's clinical condition, weight gain and enteral intake.

Predictive equations to calculate the BMR of individuals have been validated and can be used as an aid to assessing energy requirements (Schofield, 1985). Estimated energy requirement can then be calculated from the BMR with the addition of activity and stress factors, which can be particularly useful for children >3 years of age.

A global assessment of nutritional status is important when commencing PN. Serial data can help to set nutritional goals for weight and energy intake and are important for monitoring progress. Weight and weight history based on current measurements and retrospective data are commonly available, but additional measurements of length or height, mid-upper-arm circumference and skinfold thickness are of value.

Home parenteral nutrition

HPN is increasingly used for children with irreversible intestinal failure. Children who are expected to achieve the transition from PN to full enteral nutrition will also benefit from HPN if nutritional support is required for prolonged periods. HPN allows a child to achieve normal growth and development and a quality of life as close as possible to that of other children of the same age (Holden *et al*. 2000). Importantly, although HPN is associated with reduced incidence of complications (Bisset *et al*. 1992), it is a complex treatment and carries risks that are life threatening. Consequently, careful discussion with families and carers is needed to assess suitability for HPN. Successful HPN depends on commitment from the nutrition team and the family, but most crucially a well-organised training programme, careful discharge planning and ongoing support.

Complications of parenteral nutrition

PN has given life to children with chronic intestinal failure, but paradoxically is associated with many potentially fatal complications. These complications include infection, accidental damage to, or dislodgement of, the catheter and intestinal failure-related liver disease.

Infection

Catheter sepsis is a serious life-threatening complication of PN. Infection is more common in children than adults, with reported incidences of 2–29% (Holden *et al*. 1996). Care of the central venous catheter is the most important factor in preventing infections. Training of nursing staff and parents in aseptic techniques for accessing the line according to established protocols is an essential part of the care of a child on PN (Sizer, 1996). Intervention by specialist nutrition nurses has been shown to markedly decrease central-venous-catheter sepsis rates (Puntis *et al*. 1991). Many infections are associated with Gram-positive organisms, commonly staphepidermis (Puntis *et al*. 1991). Unlike adults, young children commonly attempt to touch and pick at their catheter and site, and practical measures to conceal and secure the catheter under clothing are often needed. Lower sepsis rates are achieved in children on HPN (Holden, 1991; Bisset *et al*. 1992), and where possible HPN should be considered if the requirement for nutritional support is likely to be prolonged.

Contamination of central venous catheters with faeces is another complication seen in the paediatric field. Measures to minimise contamination should be taken in young children whilst they are in nappies and also in children with stomas.

Catheter damage or dislodgement

As young children are both inquisitive and active, accidental damage to, or dislodgement of, the catheter is common. Children have been known to pull, bite and cut their catheter, and it is essential to educate nursing staff and parents in techniques to minimise risks. The catheter needs to be carefully looped and taped securely under a vest and other clothing and out of reach from resourceful toddlers. Catheter fatigue can also arise from persistent twisting of the line, a complication that can also be minimised by careful securing of the catheter.

Intestinal failure-related liver disease

The cause of liver disease in children on PN is multifactorial. It is more common in children than adults and key risk factors are prematurity (Merritt, 1986), sepsis (Sondheimer *et al*. 1998), failure to establish enteral nutrition (Vanderhoof & Matya, 1999; Andorsky *et al*. 2001) and excessive administration of specific nutrients (Koletzko *et al*. 2005).

Prevention of catheter sepsis, aggressive use of enteral nutrition and cycling of PN are useful strategies to minimise cholestasis. Protocols have been suggested for minimising liver disease in SBS (Meehan & Georgeson, 1997), but if irreversible liver damage occurs, consideration for liver transplantation or combined liver and small bowel transplant is required (Beath *et al*. 1995; Goulet *et al*. 2004).

Enteral feeding

It is well recognised that prolonged exclusive PN can lead to complications, and the importance of giving 'PN' rather

than 'total PN' has been learnt. There is generally no consensus of opinion relating to the optimal feed for managing chronic intestinal failure, and in some cases it will depend on the diagnosis. Every child will have a unique anatomy and physiology, and feeding regimens need to be tailored to the individual child. Continuous enteral feeds are often better tolerated than bolus feeds because of improved absorption associated with continuous saturation of brush-border enzymes and carrier proteins.

Weaning from PN can be successful for some children. As enteral feeds are increased PN can be reduced, but regular assessment of nutritional intake and the contributions of enteral nutrition and PN are needed.

Another important aspect is encouragement to maintain oral feeding skills. Young children who miss out on early experiences of taste and texture are more likely to develop feeding problems. Despite dependence on PN as well as nasogastric or gastrostomy feeds children should be offered bottles and solid food. This practice will help to develop their oro-motor skills whilst experiencing a range of tastes. Older children should also be encouraged to continue to try different foods within their limits of tolerance, as it will help to protect against liver disease and allow them to enjoy the important social aspect of mealtimes.

Psycho-social issues

Whilst families can be trained to safely administer PN at home, many psycho-social problems can arise (Holden *et al*. 2000). The impact on the lives of families caring for their child at home should not be underestimated. Parents and carers are aware that the survival of their child is dependent on their skills to manage PN at home, and many of them find that this responsibility is a great burden (Bisset *et al*. 1992; Carlsson *et al*. 1997). Studies have shown three emergent themes related to physical, psychological and burden of care issues at home (Sexton *et al*. 2005). Ethnicity, family structure, coping strategies, educational ability, housing and geographical location will vary between families, and a good understanding of family dynamics is crucial. Specialist home-care companies are used to deliver disposable equipment and PN to the child's home. Furthermore, there has been an increase in the provision by these companies of nursing support to facilitate the discharge home and to provide ongoing support for families. The requirements of families will differ and home-care services should be tailored to, and sensitive to, the individual needs of each family (Fig. 3).

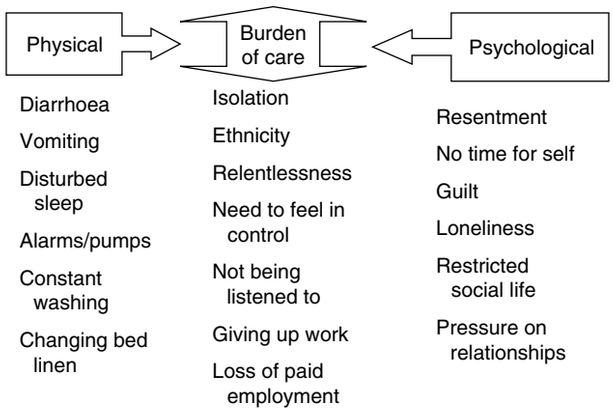

Fig. 3. Model of the key themes identified among families caring for their child and managing their parenteral nutrition at home.

Transition

The Society for Adolescent Medicine USA has defined transition as 'the purposeful, planned movement of adolescents and young adults with chronic medical and physical conditions from child-centred to adult-centred health care systems' (Royal College of Paediatrics and Child Health, 2003). The survival of children on PN is increasing and therefore transition is becoming more common. The lack of evidence has resulted in current practice in transitional care being based largely on expert opinion, although both the Royal College of Nursing (2004) and the Royal College of Paediatrics and Child Health (2003) have provided guidelines for transitional care. Important factors have been identified to help with both the successful transition and the recognition of potential barriers for enterally-fed adolescents moving to adult services (Rollins, 2005). These factors can be applied to PN and highlight the need for flexibility to ensure the needs of individual children are met.

Conclusion

PN in a paediatric patient was first described in the 1940s (Helfrick & Abelson, 1944), and in the 60 years since that report PN has been used extensively. The availability of PN has improved survival in infants following extensive resection of the gastrointestinal tract, in children with protracted diarrhoea and in premature infants.

Challenges to the nutritional care team are diverse and multifactorial. The team must coordinate and supervise the provision of PN to children with wide ranging ages and diagnoses. This process involves maintaining normal growth and development, minimising complications and personalising regimens to meet the requirements of individual children and their families.

Children with chronic intestinal failure requiring prolonged PN can be discharged to home care and have the potential to continue treatment out of hospital and maintain a good quality of life.

Paediatric PN has changed the outlook of children with intestinal failure and has to be one of the major medical developments of the twentieth century. Further clinical audit and research is required at regional, national and international level to enable nutrition teams to provide a high standard of evidence-based care to individual children and their families.

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