

Top Tips for Managing Children Receiving Long-term Parenteral Nutrition

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The number of children requiring long-term parenteral nutrition (PN) from birth has increased over the past two decades and their outlook has dramatically improved. The principles of long-term PN administration to children are similar to those in adult practice, with the important difference of the energy required for growth. This necessitates a high nutrient intake per kg body weight, particularly during early infancy and later during puberty. This can be achieved by an obsessive attention to detail, permitting normal growth during childhood and minimising the development of intestinal failure associated liver disease (IFALD). The current cohort of children dependent on PN from birth are expected to enjoy a good quality of life, grow normally, and transition to adult PN programmes during their teenage years.

Key steps

1. Identify neonates with a long-term PN requirement (i.e. short bowel) as early as possible and involve a long-term PN multi-disciplinary team (MDT)
2. Maintaining central venous access and good catheter health is a priority from birth, using neonatal percutaneous lines and a dedicated infant line insertion team
3. PN should be cycled (from 24 hours continuous infusion to about 12 hours) as soon as neonates can maintain blood glucose levels without a continuous glucose infusion
4. Neonates should have trophic feeds (e.g. 10 ml/kg) started as early as possible to promote enterohepatic communication
5. Surgery is planned carefully, aiming to achieve gastrointestinal continuity and the closure of a high stoma if present
6. Use a parenteral lipid preparation containing fish oil for the long-term care of children
7. Keep pace with growth, monitoring energy and micronutrient requirements according to age and growth rates
8. Maximise and optimise gut digestive function.

Explanations

- A.** Short bowel syndrome accounts for approximately 50% of children requiring long-term PN, and most cases present in the neonatal period. Identifying cases as early as possible, and involving Paediatric Home PN teams early, is important for families. Controlling the early development and progression of IFALD is vital; early institution of fish oil containing lipid preparations and ursodeoxycholic acid if biliary stasis occurs, reducing infusion time on PN as soon as neonates can sustain blood glucose levels during limited fasting, and using enteral feeds as soon as tolerated, all promote the development of intestinal progression.
- B.** Peripherally inserted neonatal long lines may be used for extended periods to administer PN in neonatal life. It may be tempting for a surgical team operating on a baby in the first few days to consider a surgical approach for inserting a central venous catheter, but long-term maintenance of central venous access is of paramount importance. When infants are beyond the neonatal period, long-term central venous catheters should

be inserted percutaneously under ultrasound guidance by a dedicated paediatric line insertion team.

- C.** The first surgery in these children is typically an emergency procedure in the first few days of life. Further surgery needs to be carefully planned with the MDT, concentrating upon achieving gut continuity and stoma closure as soon as possible. The benefits of bowel lengthening procedures and other non-transplant surgery have not been rigorously established in early childhood. Infants and neonates show a greater ability for bowel adaptation and subsequent intestinal progression than has often been appreciated.
- D.** Prompt and appropriate treatment of catheter-related blood stream infection is a cornerstone of long-term PN management in children as it is in adults. Families should have a written plan to take with them when presenting to their local hospital, so that rapid diagnosis and treatment of sepsis can be made, appropriate treatment commenced, and unnecessary antibiotic use avoided.
- E.** Parents or carers will be administering the home PN. They need to be adequately trained, and a 'back-up plan' in place, which usually entails training a second parent or carer who can administer PN either for respite care or in an emergency.
- F.** Keep pace with nutritional requirements to achieve normal growth. Lipid is usually required every day in infancy unless there is significant enteral nutritional intake and absorption. Monitoring height/length in addition to weight, and head circumference especially during the first year of life is essential. Most units employ an additional anthropometric measure in children and infants beyond the neonatal period, typically mid upper arm circumference (which has the lowest intra and inter observer error of any measure in childhood) for routine monitoring. Expertise in micronutrient requirements for long-term childhood PN continues to develop. These require experienced monitoring; recent examples of clinical relevance include the effect of amino acid preparations on copper bioavailability. Awareness of pubertal staging and monitoring of endocrine changes are important to ensure children's nutrition allows them to proceed through puberty at their normal age and achieve their full growth potential.

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- G.** There are two UK centres able to undertake small intestinal transplantation in children. These centres should be made aware of all children requiring life-long PN, as detailed assessment of liver disease including biopsy and wedge pressures is likely to be required. Hepatic fibrosis and portal hypertension frequently occur without biochemical evidence of IFALD. Varices often occur in atypical sites remote from the oesophagus/fundus.
- H.** Maintaining optimum gut digestive function may entail management of gut microbiota. Small intestinal bacterial overgrowth is difficult to diagnose but may be associated with episodes of D-lactic acidosis in children. Standard approaches to long-term management of children who experience severe episodes includes use of non-absorbable antibiotics, as in adult practice, but there is developing experience of the use of prebiotics and probiotics as an alternative means of managing gut flora without recourse to long term antibiotic therapy.
- I.** Dedicated and experienced multi-disciplinary teams improve outcomes. Children should be managed by centres large enough to sustain such a team and facilitate their transition to adult care. Progression to adult services should commence as soon as appropriate, with the understanding that this process will ideally take many years. Young adults who have required PN since birth will become increasingly common over the next decade; adult PN services will develop expertise in managing congenital disorders that have previously not been encountered beyond childhood.

Suggested reading

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- Neelis E, et al. Bone health of children with intestinal failure measured by dual energy X-ray absorptiometry and digital X-ray radiogrammetry. *Clin Nutr.* 2018; 37(2): 687-694.
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