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ESPGHAN/ESPEN/ESPR/CSPEN guidelines on pediatric parenteral nutrition: Home parenteral nutrition

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1. Methods

Literature search timeframe: Publications published after the previous guidelines (i.e., from 2004–December 2014), were considered. Some studies published in 2015 or 2016 during the revision process have also been considered. The references cited in the previous guidelines are not repeated here, except for some relevant publications; the previous guidelines are cited instead.

Type of publications: Systematic reviews, randomised controlled trials, case–control or cohort studies

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Table: Recommendations for home parenteral nutrition (PN)

R 12.1	A child who is expected to need PN for more than 3 months can be discharged home as soon as clinically stable for improved quality of life with less complications, e.g. reduced CRBSI and less IFALD, improved psychosocial circumstances, and reduced costs; as long as: <ul style="list-style-type: none"> • There is a safe environment (e.g. running water, reliable electricity) • At least one parent/caregiver has been trained by a specialised nutrition nurse/team, and • appropriate social support is available (LoE 2– and 4, RG 0, strong recommendation for)
R 12.2	Management of home PN by centralised units with expertise in the investigation of intestinal failure rehabilitation and with a multidisciplinary nutrition team to support care at home may minimise complications, improve outcome and allow weaning from PN as soon as possible (LoE 2–, RG 0, strong recommendation for)
R 12.3	Complications can be reduced and quality of life may be improved by: <ul style="list-style-type: none"> • Using existing evidence-based guidelines • Limiting the number of infusions/week if possible • Limiting the hours of PN to the minimum possible aiming for 10–12 h • Incorporating replacement of excessive fluid losses in PN if at all possible • Use of portable pumps • Care as close to home as possible (LoE 3 and 4; RG 0, strong recommendation for)
R 12.4	Paediatric HPN patients must be followed-up by an experienced team on a regular basis with a minimum of about 4 visits per year in older children (LoE 4; RG 0, strong recommendation for)
R 12.5	Monitoring should be considered on an annual/alternate year basis for complications including: <ul style="list-style-type: none"> • Liver disease by US • Bone density, vitamin D and body composition if available • Radionuclear lung perfusion scan for pulmonary emboli if indicated • Chest X-ray to assess appropriate position of central line (LoE 4; RG 0, conditional recommendation for)
R 12.6	PN mixtures that are stable for >7–14 days may be used to minimise the frequency of deliveries required (LoE 4; RG 0, conditional recommendation for)
R 12.7	The use of a single bag may be recommended (LoE 4; RG 0, conditional recommendation for)
R 12.8	The patient should be on a stable regimen before starting home PN (LoE 2– and 4, RG 0, strong recommendation for)
R 12.9	PN solutions should be compounded according to the individual patient's macro- and micronutrients needs (LoE 2–, 4; RG 0, strong recommendation for)
R 12.10	The aim of home PN should be survival into adult life with the best possible growth and psychosocial development, school attendance and participation in other activities, e.g. sport, swimming, family holidays (LoE 4; RG 0, strong recommendation for)
R 12.11	Early referral of long-term PN patients to an expert centre may reduce PN-associated complications (LoE 2–, RG 0, strong recommendation for)
R 12.12	Early referral to a centre for intestinal transplantation can minimise mortality from Home PN related complications whilst on the waiting list (LoE 2–, RG 0, conditional recommendation for)
R 12.13	Home PN expert centres should provide 24-h phone support and support weaning off PN at the earliest opportunity (LoE 4; RG 0, strong recommendation for)

Language: English, Dutch and German

Key words: Infants, children, home parenteral nutrition, quality of life, complications, liver disease, catheter-related sepsis, thrombosis, metabolic bone disease, electrolyte disturbances, hypoglycemia, hyperglycemia, prevention, long-term outcome, costs.

Abbreviations: CRBSI – catheter-related blood stream infections; CVC – central venous catheter; IF – intestinal failure; IFALD – intestinal failure associated liver disease; PN – parenteral nutrition; US – ultrasound examination

2. Introduction

Home parenteral nutrition (Home PN) is the best alternative to prolonged hospitalisation and the best option for improving the quality of life of children dependent on long-term PN [1]. Furthermore, Home PN is associated with lower risks of complications such as catheter related blood stream infections (CRBSI) and overall cost [1,2]. Therefore, Home PN should be considered for any child who is clinically stable and expected to remain dependent on PN for at least three more months [3]. Over the last decade, Home PN has increased rapidly due to improvement in survival with better quality of care of surgical treatment, of neonatal care, of daily catheter care and also of the composition of PN [3]. Reported prevalence varies across studies ranging from 9.6 children per million in the Netherlands to 13.7 children per million in the U.K [3,4]. In this chapter, we will discuss indications, organisational aspects, requirements, follow-up, complications, quality of life and long-term outcome of Home PN.

3. Indications

3.1. Why start a home PN program?

Long-term total or complementary PN is required to preserve nutritional status when oral or enteral nutrition cannot meet

protein-energy needs, especially in diseases that impair digestive function. When a child does not need hospitalisation but depends on long-term PN, home parenteral nutrition (Home PN) is the best alternative to prolonged hospitalisation and is recognized as the best option for improving the quality of life of these children and their families [1]. In addition, compared to PN in the hospital setting, Home PN is associated with lower risks of catheter related blood stream infections (CRBSI) and a decreased risk of intestinal failure associated liver disease (IFALD) [2]. Therefore, Home PN should be considered for any child who is clinically stable and expected to remain dependent on PN for at least three more months [3].

National prevalences are difficult to compare because of the use of different definitions and inclusion criteria. Therefore, it would be helpful to have national or even a European register for Home PN in children using the same definitions and inclusion criteria to improve the quality of data on prevalence, outcome and complications. A Dutch study estimated a point prevalence of 9.6 children per million for instance, but a study from the UK published a higher point prevalence of 13.7 children per million [3,4]. However, in recent years, the incidence of children on Home PN has increased rapidly due to improvement in quality of care, of surgical treatment, of neonatal care, of daily catheter care and also of the composition of PN [3]. Children on treatment with Home PN should be managed by a multidisciplinary team. A European study showed that in both adults and children, the risk of death is increased by the absence of such a specialist team [5].

3.2. Underlying indications

The most common indications for prolonged PN and thus Home PN in children are primary digestive diseases causing intestinal failure (IF). Short bowel syndrome (SBS), mainly acquired during the neonatal period, is the largest group of patients, which

has increased rapidly in frequency, accounting for at least 40% of all cases [1,3–7]. The main other digestive indications are intractable diarrhoea of infancy, such as tufting enteropathy/epithelial cell dysplasia or microvillous inclusion disease and motility disorders, such as chronic intestinal pseudo-obstruction. Other less frequent indications are inflammatory bowel diseases, especially Crohn's disease, and primary nondigestive indications such as immune deficiency, tumours, metabolic diseases and neurological impairment with intestinal hyperaesthesia. The need for Home PN in these diseases is usually shorter than for primary digestive diseases [1].

3.3. Age

There is no minimum age criterion at which Home PN can be safely started, but it will depend on each individual case [1,3–5,7]. Most infants are not sufficiently stable to be discharged home until about 4 months of age (corrected for prematurity), although some have been discharged by 2 months. Data from a cohort of 139 children showed that almost 15% of the children on Home PN were younger than 1 year [3]. It should be considered that starting Home PN prior to 4 months of age may carry a greater risk of mortality [5].

3.4. Condition

Children can be discharged from the hospital to continue PN at home if they are expected to need PN for at least 3 further months, and have a well-inserted central venous catheter. In addition, they have to be in a stable condition regarding their underlying disease and their fluid and electrolyte requirements. Ideally they should be able to tolerate PN infused over just 10–14 h overnight, but in certain cases it may be necessary to send children home on PN for up to 18 h and in extremely unusual circumstances, 24 h.

3.5. Social and family requirements for home PN

Before starting Home PN, the team should decide if the indication is appropriate and ethical. Furthermore, parents/caregivers but also the (older) child have to be able to cope with all the medical, emotional and technical issues related to PN. Practical issues should be discussed such as space for a refrigerator, pumps and the need for home nursing assistance [1]. Preferably, both parents/care givers should be trained simultaneously. A single-parent family is not always a contraindication for Home PN, but social help and home nursing assistance may become more necessary [1]. The professional status of both parents needs also to be discussed, since one parent may have to stop working for a certain time, although parents should be supported to continue working if at all possible.

3.6. Cost savings

An economic evaluation study of pediatric small bowel transplantation in the United Kingdom showed mean costs up to 30 months of £207,000 for those transplanted or on the waiting list, and £159,000 for those stable on home PN [8]. A recent adult study showed that Home PN costs were €13,276 for treatment introduction, followed by €77,652 annually [9]. Home PN is thus very expensive. However, cost-benefit studies have demonstrated that it is about 65% more cost-effective than hospital treatment for children as for adults [10–12]. The longer a patient survives at home on PN, they will have fewer complications with time and the more cost-effective home-treatment becomes.

4. Preparation for PN care at home

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- R 12.1** A child who is expected to need PN for more than 3 months can be discharged home as soon as clinically stable for improved quality of life with less complications, e.g. reduced CRBSI and less IFALD, improved psychosocial circumstances, and reduced costs; as long as:
- There is a safe environment (e.g. running water, reliable electricity),
 - At least one parent/caregiver has been trained by a specialised nutrition nurse/team, and
 - Appropriate social support is available (LOE 2– and 4, RG O, strong recommendation for, strong consensus)
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Once it is established that the child will be discharged home on PN treatment, the medical team needs to discuss the management of PN at home and what is involved with the family. It is best practice for both parents to meet with a consultant specialising in PN and nutrition nurse specialist. If possible a social worker should also discuss any social concerns that the professionals and/or family have. The home needs to be inspected to ensure that it is adequate for caring for a child on PN. There needs to be a reliable electricity supply for the pump and accessible running water to wash hands, preferably without a step between the bedroom and washing area. Accessible toilet facilities are also essential for the older child. If the child is young/shares a room with a young sibling the PN equipment needs to be situated so that the toddler cannot tamper with it.

If at all possible both parents should be trained to administer PN. In a single parent family setting, that parent alone can be trained along with another family member/close friend as well, if available. In practice it is unusual for a non-parent to be willing to be trained when made aware of the responsibility involved, and ascertaining the capability of the caregiver to take care of the child receiving Home PN is part of the evaluation and approval of the home environment as discussed below. If parents live separately and the child spends time in both homes then both parents will need to be trained. They should be trained at the same time in order to ensure that each is aware of the training the other has had, that they cooperate over the care of their child's PN and to use health resources as efficiently as possible. In certain circumstances when no healthy parent is available a trained nurse will need to come into the home to connect and disconnect the PN, but this should be avoided if at all possible since it reduces family flexibility. Furthermore, if several different nurses take turns to do it, it may increase risk of CRBSI.

Funding for the home PN needs to be secured. In many countries this will be obtained from the National Health Care System. A reliable supply of PN and ancillary equipment must be procured. There are three possible methods:

1. Dedicated homecare companies with compounding facilities to manufacture the PN that will also supply the ancillary equipment.
2. Supply of the PN by the hospital pharmacy and ancillary equipment from the hospital or community services.
3. In exceptional circumstances parents have been trained to compound the PN at home with supplies obtained from a hospital [13].

The parents need to be available for a 1–2-week period to undergo a structured training programme to manage PN at home. Flexibility during the 1–2-week period may be needed by the nurse undertaking the training to enable parents to fulfil other commitments (including work/other childcare) during the training period. Training should include learning techniques for connecting and

disconnecting the PN bags from the CVC, understanding how to recognise complications and what to do when they arise. Once training commences all medical investigations, other than the most routine should have been completed. It is difficult for parents to concentrate effectively on learning PN if their child is still undergoing tests/surgical procedures.

In some countries/circumstances PN training can be undertaken by specialist nurses working in the community. These nurses would usually work with a home care company that supplies PN to the home. Parents will be trained to manage the PN after discharge home. On completion of training parents should be assessed and re-trained on any area in which they fail to demonstrate appropriate standard of care.

A planning meeting should be held around the time of discharge to ensure that professionals and family members are all aware of the child's treatment plan and where to manage different complications that may arise. A community nurse/health worker, the specialist hospital consultant and nutrition nurse, the consultant at the hospital closest to the patient's home and the parents should all be present with other available professionals involved in the child's care. A shared care plan should be made between the local hospital, specialist team and community professionals. A system for direct access to the local hospital in an emergency should be set up. Minutes of the meeting should be circulated for future reference.

5. Organisation, monitoring and follow-up

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- R 12.2** Management of home PN by centralised units with expertise in the investigation of intestinal failure rehabilitation and with a multidisciplinary nutrition team to support care at home may minimise complications, improve outcome and allow weaning from PN as soon as possible (LOE 2–; RG 0, strong recommendation for, strong consensus)
- R 12.3** Complications can be reduced and quality of life can be improved by:
- Using existing evidence-based guidelines,
 - Limiting number of infusions/week if possible,
 - Limiting hours of PN to minimum possible aiming for 10–12 h,
 - Incorporate replacement of excessive fluid losses in PN if at all possible,
 - Use of portable pumps, and
 - Care as close to home as possible (LOE 3 and 4; RG 0, strong recommendation for, strong consensus).
- R 12.4** Paediatric HPN patients must be followed-up by an experienced team on a regular basis with a minimum of about 4 visits per year in older children (LOE 4; RG 0, strong recommendation for, strong consensus)
- R 12.5** Monitoring can be considered on an annual/alternate year basis for complications including:
- Liver disease by US
 - Bone density, vitamin D and body composition if available
 - Radionuclear lung perfusion scan for pulmonary emboli if indicated
 - Chest X-ray to assess appropriate position of central line (LOE 4; RG 0, conditional recommendation, strong consensus)
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As mentioned previously, management of children on Home PN should be performed in specialist centres with a multidisciplinary IF rehabilitation hospital PN team in order to reduce the risk of complications and even mortality [5]. Home PN centres should have adequate expertise and resources to ensure a good standard of care by using existing evidence-based guidelines and aiming to wean the child from PN as soon as clinically appropriate [1]. The aim is to try to limit the number of infusions per week and also hours of PN to the minimum, possible aiming for 10–12 h. A portable pump should be used (see chapter “Techniques”). Physicians should be trained and qualified to be responsible for the

appropriate use, prescription and follow-up of patients on Home PN programmes. Nurses responsible for parents' teaching and training should work with the specialist IF team and evaluate their capacity to deal with all medical and technical issues related to the child's treatment. Pharmacists or pharmaceutical companies specialising in PN at home should ensure safety of compounding and storage of the PN mixtures. Replacement of excessive fluid losses should be incorporated in PN if at all possible. Once discharged from hospital, regular out-patient follow-up should be planned to check clinical and biologic parameters (Table 1) with a minimum of 4 visits/year to the specialist centre [14,15]. Investigations should be tailored for each child and will depend on the underlying disease. Visits should be tailored according to each individual situation, initially 1–3 months after discharge, but more frequently if necessary, especially in infants. During visits, oral and/or enteral feeding opportunities should always be reconsidered and discussed with parents or caregivers. A 24 h phone contact with the on-call team at the specialist centre should be provided [1]. A close connection with general practitioners and local non specialised hospital units is warranted. Whilst the specialist team should lead care they should support the child's local medical services in delivering as much care as possible as near to the home as possible. Some children live long distances from the specialist centre and it is not practical or necessary for them to travel all the way to the centre for every problem that arises.

6. Techniques

6.1. Vascular access

A centrally placed cuffed catheter is the most secure venous access for PN at home. A peripherally inserted centrally placed catheter (PICC) can be safely used for many months [16]. A PORT should only be used in exceptional circumstances since it can be difficult to eradicate infection. In exceptional circumstances a-v fistulae have been used successfully [17].

6.2. The infusion cycle

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| R 12.6 | PN mixtures that are stable for >7–14 days may be used to minimise the frequency of deliveries required (LOE 4, RG 0, conditional recommendation for, strong consensus) |
| R 12.7 | The use of a single bag may be recommended. (LOE 4, RG 0, conditional recommendation for, strong consensus) |
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PN at home is normally infused over 10–12 h overnight (up to 16–18 h in some infants) leaving the child free to participate in activities, including school, during the day [18]. The number of nights/week that PN is infused should be minimised at the earliest opportunity. An oral/enteral sodium supplement should be given the day before and after a night off PN in children with high sodium requirements/excessive loss. Some children with excessive stoma/stool fluid losses and those with difficulty maintaining a normal blood glucose level may need a prolonged infusion cycle. In exceptional circumstances children can be sent home on PN over 24 h e.g. a toddler or older child who is unlikely to significantly improve and will benefit from the opportunity to return to the family home and participate in usual activities such as school. The PN infusion rate should be gradually reduced over the final hour prior to disconnection, particularly in the younger child to avoid the risk of hypoglycaemia on stopping [19].

PN for home use may be compounded by a hospital pharmacy, a specialist homecare company pharmacy or even by parents at home [13]. Stability can be obtained for 14–21 days for most

Table 1
Clinical and biologic monitoring in children on PN at home.

Intervals	Clinical assessment	Other investigations
1–3 months	<ul style="list-style-type: none"> • Weight, height • Clinical examination • Dietetic assessment 	<ul style="list-style-type: none"> • Blood count, coagulation • ALT, AST, bilirubin, GGT, alkaline phosphatase, Na, K, Ca, PO₄, Mg, glucose, urea, creatinine • Urinary electrolytes (Na, K, osmol) • Plasma vitamins A, D, E, B12 • Ferritin, iron, zinc, copper, selenium, manganese • When indicated: <ul style="list-style-type: none"> - anti X-a level (in case of low-molecular-weight heparin use), - blood gas analysis, albumin, PTH, thyroid function - duplex ultrasonography (if using an arteriovenous fistula) • Bone densitometry • Body composition (if available) • Liver and biliary tract ultrasonography • Renal ultrasound • Radionuclear lung perfusion scan (when indicated) • Chest X-ray to assess appropriate position of central line
6–12 months		
12 months		

formulations when made by a homecare company enabling delivery to the home on a 2-weekly basis. In smaller children or those with variable stool/stoma losses weekly deliveries may be appropriate when first discharged home. If changes to the PN are needed it is best if these can be made by increasing or decreasing the volume infused in the first instance before changing the formulation. Any excess stoma losses should be replaced by incorporating appropriate fluid and sodium in the PN (see chapter “Fluid and nutrients”). In all cases there should be extra fluid, ‘overage’, in the PN bag in addition to the amount prescribed in case needed.

PN for home use should ideally be provided in a single bag system. Every effort should be made by the pharmacy to obtain stability as a single bag and the clinical team should adjust the prescription if possible to enable an all-in-one bag to be used. In certain circumstances a two-bag system may be needed, but some units are able to avoid two bags – even when achieving 14–21 days stability.

6.3. Pumps, equipment and ancillaries

The PN should be infused via a filter (if available) with a portable pump when at home [20]. The pump should be as quiet and light as possible. It needs to be reliable in order to avoid unnecessary alarming and waking the family at night. A good battery life and rucksack are particularly important when the child is active whilst connected to PN. Ideally, there should be a contract for the pump to be rapidly replaced as if it breaks (a 4-h replacement service may be possible) and serviced on an annual basis. The family should have a second pump at home – essential for children who need a 24-h infusion in order to avoid hypoglycaemia.

Ideally all ancillary equipment should be delivered to the home. Families should not have to collect it from different sources.

7. Fluids and nutrients

PN requirements depend on age, weight, underlying disease, nutritional and current hydration status, and environmental conditions. When PN is not the sole source of protein-calorie intake, intestinal absorptive function should be estimated.

7.1. Fluid and electrolytes

R 12.8	The patient should be on a stable regimen before starting home PN (LOE 2– and 4, RG 0, strong recommendation for, strong consensus)
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The fluid and electrolyte composition of the PN regimen should reflect fluid losses and deficits that may result from medical therapy. Digestive tract losses due to diarrhoea or from stomata should be measured (volume and sodium concentration) and replaced. Adjustments may be required frequently depending on the clinical situation. The patient should be on a stable regimen before starting a home PN programme.

7.2. Vitamins and trace elements

The PN infusion should provide vitamins and trace elements (TE), according to the patient's age, weight and specific needs. Certain conditions may predispose to deficiencies of fat-soluble vitamins by interrupting the entero-hepatic circulation: extensive resection of terminal ileum, bacterial overgrowth, which may lead to deconjugation of bile salts and increased inflammation with further small intestinal injury, and PN-associated cholestasis from prolonged PN [21,22]. Oxygen is the principal agent responsible for degradation of vitamins and originates from PN ingredients, the filling process, air remaining in the bag after filling, and oxygen permeation through the bag wall. Therefore, multilayered bags with reduced gas permeability should be used, and careful oxygen monitoring during the filling process is mandatory [23]. Exposure to light may degrade vitamins (especially vitamin A) and is associated with increased production of peroxides. Light protection should be considered for Home PN that is exposed to strong direct day light (see chapter “Vitamins”). Supplying vitamins in a lipid emulsion can be an additional way to reduce losses due to adsorption onto the tubing materials [24]. Vitamin and trace element (TE) doses in Home PN should be adjusted based on regular monitoring. In long-term PN patients, a restriction of manganese supplementation is needed to avoid accumulation [25,26]. In certain circumstances, e.g. patient weaning from PN, vitamins and minerals may be given orally/enterally.

7.3. Nutrition mixtures for paediatric home PN

R 12.9	PN solutions should be compounded according to the individual patient's macro- and micronutrients needs (LOE 2– and 4, RG 0, strong recommendation for, strong consensus)
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Binary mixtures including glucose, amino acids, electrolytes, trace elements and vitamins (lipids being administered separately on a Y-line) or, ideally, all-in-one mixtures are provided to children on Home PN. Mixtures may be manufactured and delivered to patients with complimentary equipment weekly,

fortnightly or monthly. Vitamins or drugs added to nutrient mixtures might impair their stability whilst, the availability of drugs and vitamins might be reduced when introduced into PN mixtures [27]. Thus, depending on these limiting factors, the “safe” duration of PN bag storage varies from about 14 to up to 21 days. Bags should be stored at 4 °C from the time of their production to their administration to the patient without any discontinuity. The families should receive a dedicated refrigerator for PN bag storage.

Special PN mixtures should be prepared according to individual requirements. The so-called standard PN mixtures compounded by pharmaceutical companies, meant only for adult patients on short-term and/or complementary PN, cannot meet children’s nutritional requirements and are free of vitamins and trace elements. They may be suitable for some adolescents on supplemental PN only. Their use in children at home can lead to metabolic complications with severe electrolyte imbalances. Currently, no pediatric standard formulas are suitable for children on PN at home.

8. Complications

Any complications that can develop in children receiving hospital PN may occur but are less frequent at home. The more common complications of long-term/home PN include infection, disturbances of fluid balance and occasionally renal disease, CVC related thrombosis and abnormalities of growth, bone density and body composition. IFALD is uncommon at home. Initial management of inter-current problems should be started by the parents with support of the community nurse/professional. If urgent medical care is needed the child should be taken to the nearest hospital.

Parents should have a hand held plan for how to manage complications.

The two most common infections are those involving the skin and sub-cutaneous tissue around the catheter exit site and CRBSI. If inflammation develops at the exit site, a swab should be taken and antibiotic treatment commenced. A topical antibiotic can be used initially and, if the infection fails to respond, a systemic antibiotic. Most patients with IF can absorb an oral/enteral antibiotic sufficiently well, but in more severe cases with no oral tolerance intravenous treatment may be necessary.

In order to avoid serious life-threatening complications of septicaemia if the child develops a fever >38.5°C or is unwell with rigors/other symptoms suggestive of infection, s/he should be taken to the nearest hospital with in-patient paediatric facilities. A blood culture should be taken via the CVC and at least two antibiotics to cover both gram-positive and gram-negative infections commenced. Antibiotics should be adjusted according to sensitivities once available. Treatment may be continued at home with antibiotics either given by the parents (after training) or a community nurse, providing the child is stable. Taurolidine and 70% ethanol line locks have been successfully used to reduce the incidence of infection when PN is cycled: see chapter “venous access” for recommendations [28,29].

In children with excessive fluid losses, e.g. those with intestinal dysmotility or pseudo-obstruction, there should be a plan for managing increasing fluid losses at home in the first instance. In most cases the volume of PN infused can be increased, with a plan for hospital admission if losses exceed a certain volume or the child is clinically unwell. By increasing PN volume, fluid and electrolyte replacement will be given. Children will often have reduced appetite when losses increase (if on partial PN) and will also benefit from the increased calories.

Growth on PN can be poor [30]. In order to promote growth protein/nitrogen intake may be increased. When nitrogen intake is increased, the amount of glucose has to be increased as well. If the child’s height gain does not improve s/he may gain excessive fat. If this starts to occur with the weight centile increasing above the height centile, the carbohydrate and protein should be reduced again.

Low bone density is a risk of long-term PN [30]. Monitoring of vitamin D and a DXA scan should be done on an annual basis. Other relevant investigations include parathyroid hormone (PTH), calcium, phosphate and urinary calcium. If vitamin D is low, oral or intra-muscular supplements may be needed in addition to that in the PN.

Catheter-related venous thrombosis is a potential risk and pulmonary emboli may develop [15,31]. Long-term anti-coagulation treatment may be required.

Children are at risk of increased fat mass and low lean body mass [14]. Measurement of body composition using Dual X-ray absorptiometry (DXA) should be considered on a 2–3 yearly basis or annually if previously abnormal.

Hepatobiliary disease is a less of a risk in long-term stable patients at home compared to in hospital. If present on discharge liver function usually improves with time and should be monitored appropriately. There is an increased risk of gallstones. Annual ultrasound examination is recommended and gallstones treated as needed [32].

Episodes of dehydration and possible pre-renal failure when there are excessive fluid losses can predispose to renal disease. Monitoring includes 3-monthly urea and creatinine blood levels and annual renal ultrasound.

9. Quality of life

R 12.10	The aim of home PN should be survival into adult life with the best possible growth and psychosocial development, school attendance and participation in other activities, e.g. sport, swimming, family holidays (LOE 4, RG 0, strong recommendation for, strong consensus)
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The aim of PN at home is to give the best possible quality of life to the child and family. It should be recognised that some children may need to stay on treatment throughout childhood and into adult life. It is important to set up the PN so that the child can have the most normal life possible for weeks, months and years. If set up in the simplest manner possible the PN is incorporated into daily life and any symptoms related to underlying disease are more troublesome than PN itself [18]. It is possible for children on PN at home to have a similar quality to healthy children [33]. Children can attend regular school, participate in non-contact sport and swim and go on family holidays [34]. In many countries there are patient support groups for home PN patients. Support groups can advise on foreign travel, including health insurance amongst other issues. Quality of life and survival on long-term/home PN can be maximised when the child is cared for by a multi-disciplinary team that uses the existing evidence-based guidelines. A portable infusion pump should be used and the specialist centre should facilitate care close to home whenever possible.

In children with other major organ failure, e.g. severe developmental delay without the ability to live an independent life, or those who need palliative care, home PN may prolong suffering rather than improve quality of life. In such cases an ethical review may be needed.

10. Home PN long-term outcome: importance of centralised home PN expert centers and the role of small bowel transplantation

R 12.11	Early referral of long-term PN patients to an expert center may reduce PN-associated complications (LOE 2–, RG 0, strong recommendation for, strong consensus)
R 12.12	Early referral to a centre for intestinal transplantation can minimise mortality from Home PN related complications whilst on the waiting list (LOE 2–, RG 0, conditional recommendation for, strong consensus)
R 12.13	Home PN expert centres should provide 24-h phone support and support weaning off PN at the earliest opportunity (LOE 4, RG 0, strong recommendation for, strong consensus)

Large, long-term pediatric surveys reported a mean Home PN duration of about 2 years with an upper duration longer than 15 years [7,35]. About 40–70% of pediatric patients can be weaned from long-term Home PN depending on their underlying disease and medical condition, the prognosis being better for inflammatory bowel diseases and short bowel syndrome than for other indications [7,36,37]. Patients with motility disorders and structural enterocyte defects have an increased risk for permanent PN dependency [36,38].

Survival rates of long-term Home PN patients vary between 62 and 94% depending on cohort and observation period [7,39]. Mortality during the early years of Home PN is mainly attributable to the underlying disease, whereas in long-term patients PN-related complications predominate [40]. Children on Home PN have better survival rates and greater likelihood of resuming full enteral nutrition than adult patients [7].

However, a subgroup of children receiving Home PN have irreversible intestinal failure and cannot be weaned from PN [38]. In these patients small bowel transplantation might be an alternative to lifelong Home PN, depending on the individual situation (complications of long-term PN, tolerance of the family). Only a small percentage of patients require immediate transplantation for life threatening conditions. When irreversible intestinal failure is diagnosed, patients should be considered to be listed for intestinal transplantation in accordance with the criteria of the American Society of Transplantation [41]. Nevertheless, Home PN can be used for an indefinite period of time without intestinal transplantation, if long-term PN is effective and well tolerated. Some children are now growing up on PN and transitioning to adult care. Careful support is needed to ensure that the young adult engages with their new team and takes on appropriate responsibility for their future health. Since the first isolated small bowel transplantations, major advances resulted from the use of new immunosuppressive treatments [2]. When liver structure and function are impaired by long-term PN, a combined small bowel and liver transplantation should be considered. However, timing of referral and criteria for isolated intestinal or combined transplantation are still a matter of debate [40,41]. Earlier referral may be a contributory factor to improved survival [2]. Home PN expert centers should provide a 24-h phone support and support weaning off PN at earliest opportunity. Early referral of long-term PN patients, especially before irreversible liver failure occurs, can increase their quality of life and survival and reduce the cost of care [2].

Conflict of interest

None declared.

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