Module 19.1

Indications and Epidemiology

Learning Objectives

- Learn about epidemiology and the extent of the use of HPN in adult patients;
- Learn about indications for HPN in adult patients and clinical features (including prognosis) of these patients.

Contents

1. Incidence and prevalence
2. Indications
3. Demographic data of long-term HPN patients
4. Perfusion regimen
5. HPN-related complications
6. Rehabilitation status
7. Prognosis
8. Conclusions

Key Messages

- HPN is worldwide used in industrialized countries;
- For patients with benign diseases, the main indication is short bowel (80%);
- In many European countries as well as in US, cancer has become the main indication for HPN;
- The point prevalence of HPN in US is expected to be 5 to 10 times higher than in Europe (from 2 to 12/10^6 inhabitants);
- HPN-related complications are quite rare.
1. Incidence and prevalence

The use of parenteral nutrition administration started in the early 1960s. Schils et al (1) tried to maintain a patient at home on parenteral nutrition (HPN). Although this patient only survived during a few months, several teams in North America and in Europe initiated a program of HPN during the 1970s. Subsequently, many groups reported their initial experience with HPN, mentioning a low incidence of complications and a good survival rate (2 - 5).

According to the data collected by the North America Registry on HPN patients, the estimated number of HPN patients in the US was approximately 18 000 in 1986 and reached 40 000 in 1992 (6).

A multicentre survey performed in nine European countries in 1997 showed a mean incidence of three newly enrolled patients on HPN per million inhabitants (7) and a mean prevalence of four per million (Fig. 1). The increased use of HPN in Europe is obvious when comparing data obtained in 1986 and in 1993 (7, 8).

However, use of HPN is still ten times higher in the US than in Europe. HPN is routinely used in Japan, Israel and Australia. In the European survey, age distribution of the patients at the onset of HPN was as follow: 28% aged 16-40, 44% aged 41-60, 18% aged 61-70 and 10% over 70 years (7).

Since 1997, data about HPN incidence are only available in a few European countries. In the UK, a national register was started by BANS since 1996 (9). The number of adult HPN registered with BANS has grown progressively since 1996 (Fig. 2).

An increase in the number of centres reporting was seen in 2002, but despite it is felt that HPN is underreported. A total of 103 new adult patients were registered in 2002, making the total number of registered patients receiving HPN at the end of 2002 to 465. It is noteworthy that there are significant national but also regional variations in the reporting HPN.

In Scotland, all patients receiving HPN have been identified with the development of the Managed Clinical network (MCN) and data from 2001 found the point prevalence to be 12 patients per million of the population (10). The figure exceeds the overall UK rate of approximately 8 patients per million of the population. Within the UK, further regional variation has also been identified.

In Spain, data are annually collected throughout a designed questionnaire (11). In 1997, the registration rate of HPN in Spain was about 0.7 patients/10^6 inhabitants/year. In 2000, fourteen hospitals participated and 67 patients - adults and children - were newly enrolled. The registration of patients that was expected to reflect the incidence was 1.9/10^6 inhabitants/year. In 2001, seventeen hospitals participated by enrolling 66 patients (1.65 patients 10^6 inhabitants/year).
In France, a national HPN registry was open in 2001 (12). Between June 2001 and June 2004, 413 adults were included in the registry; the estimated incidence was: 3 newly enrolled patients/1.10^6 inhabitants/year.

2. Indications

Overall, the distribution of underlying diseases requiring HPN is quite similar in Europe, the US and Japan (6, 7).

![Distribution of underlying diseases for HPN patients in Europe (1997; n = 479)](ESPEN-HAN, Clin Nutr 1999, 18, 135)

Cancer has become the largest single indication for HPN throughout the world (40%). Crohn's disease, mesenteric vascular diseases, radiation enteritis and disorders of intestinal motility remain the most frequent benign conditions requiring long-term HPN. HPN is also used in AIDS patients with intractable diarrhoea (Fig. 3). However, the number of AIDS patients receiving HPN recently decreased since the introduction of more efficacious tritherapy. We have to underline that 25% of HPN patients are suffering of "miscellaneous" diseases, including chronic pancreatitis, intestinal mucosa atrophy, anorexia nervosa, cachexia, etc.

However, the distribution of underlying diseases in HPN patients varies among the different European countries (7) (Fig. 4). Crohn's disease accounts for 44% of indications in UK but for only 13% in The Netherlands. On the contrary, cancer represents 60% of indications in The Netherlands and 5% in UK. The survey that was performed in 1993 showed that cancer was the main indication for HPN in Italy (67%) (8).

If we consider the benign diseases, the most common indications are small bowel resection, digestive fistulas and motility disorders. For cancer patients, the main indication is intestinal obstruction which is common in cases of peritoneal carcinomatosis. Intractable diarrhoea associated with severe malnutrition is the major indication in AIDS patients.

3. Demographic data of long-term HPN patients

A survey that was performed by the ESPEN-HAN group included 228 adult patients: 141 females and 87 males, with a median age of 49 years (range 19-92) (13). The underlying diseases were: Crohn's disease (33%), mesenteric vascular diseases (25%), post-surgical (19%), intestinal pseudo-obstruction (8%), radiation enteritis (4%), abdominal trauma (2%) and miscellaneous (8%).
Intestinal anatomy was defined in 222 patients. The remaining small bowel was less than 50 cm in 84 patients, less than 100 cm in 67 patients, less than 200 cm in 44 patients and less than 300 cm in six patients. Twenty-one patients had no small bowel resection (12%).

In patients with short bowel, the intestinal anatomy was a terminal jejunostomy (I) in 41% (Fig. 5), jejunocolic (II) (Fig. 6) in 46% and jejuno-ileo colic (III) in 13%.

The distribution of underlying diseases is typical and similar to other reported HPN series.

As was also expected, 80% of the patients had a short bowel.
The fact that 65% of these patients had less than 1 m of remaining small bowel and that 88% had a type I or II anastomosis confirms previous observations that showed the importance of the length of the residual small bowel and the type of intestinal anastomosis for predicting the HPN-dependency (14).

4. Perfusion regimen

In the majority of the cases (69%), administration of nutritional solutions is performed through a subcutaneous tunnelized catheter and is positioned in the vena cava via the internal jugular vein or a subclavian vein, preferentially on the right side (7).

Based on the reports of the North America Registry on HPN and the European surveys, the use of subcutaneous reservoirs (port-a-cath) is growing (6). This trend is due, on one hand, to its wide use in cancer patients who receive chemotherapy and, on the other hand, to the willingness of some patients who prefer implantable catheters for functional and esthetic reasons, for instance for practicing aquatic sports or for taking a shower.

The number of perfusions that are administered per week may vary along in time in function of intestinal adaptation capacities. The European survey has shown that the percentage of bags/week was as follows: 7 (67%), 6 (9%), 5 (12%), 4 (8%), or less (4%) (7).

Oral feeding is not only allowed but also encouraged in patients without bowel obstruction or need for bowel rest. It has been shown that patients with short bowel are in fact hyperphagic. In the 1997 European survey, 50% of patients had free oral intakes, 27% had limited oral intakes, while 23% ingested nothing (7).

5. HPN-related complications

HPN-related complications are detailed in another chapter. However, the following data were obtained by the ESPEN-HAN group (13).

Within the 12-months period prior to evaluation, the mean number of hospitalisation was 2.7 (0-12), corresponding to a mean period of 23 days (range 0 to 270 days). Reasons for hospitalisation were related either to the underlying diseases in 27% of days admitted to hospital, to HPN complications in 48% or to other medical reasons in 25%. Of the HPN complications catheter related sepsis accounted for 61%, metabolic disorders for 27%, venous access thrombosis for 12%.
One of the main goals of HPN is by definition to avoid prolonged or recurrent hospitalisations. When we consider the 12-month period before the evaluation, the mean time of hospitalisation corresponds to 8% of the year. This seems acceptable for patients with life-threatening intestinal failure. However, we have to accept that a few patients stayed much longer in hospital (up to 270 days).

The mean number of central venous catheters used during the total HPN period was 3 (range 1 to 17), with a mean survival time per catheter of 34 months (range 4-245 months). During the 12-month-period before evaluation, an episode of catheter-related sepsis occurred in 31% of the patients. Central venous thrombosis was reported in 9% and vascular access problems in 13% of the patients.

**6. Rehabilitation status**

When comparing the rehabilitation score before HPN and at the time of evaluation, it appears that the percentage of HPN patients who are capable of coping with a job is about 65% (Fig. 7) (13). Nevertheless, there is a sharp decrease in this percentage in favour of part-time work when on HPN. This can be easily explained by limitations due to the time spent on taking parenteral nutrition.

On the other hand, it clearly appears that the percentage of grade IV (bedridden at home) patients significantly decreased meaning that HPN may improve the status of patients who had a very low rehabilitation score before starting HPN.

**7. Prognosis**

Several studies have shown that survival (prognosis) is linked to the underlying disease (14).

In a European survey performed in 1997, mortality rate after a 6 to 12 month follow-up period was 4% in Crohn's disease, 21% in radiation enteritis, 13% in vascular diseases, 16% in miscellaneous but 74% in cancer and 34% in AIDS (Fig. 8). The North America HPN Registry reported similar results (6). B. Messing et al. performed a study on 217 HPN patients with benign diseases and that have been enrolled in a HPN program between 1980 and 1989 in Belgian - French specialized centres (15).

Seventy-three patients died during the follow-up period. Mortality rate due to HPN was 11%. This work showed a survival probability at 1, 3 and 5 years of 91%, 70% and 62%, respectively.

Multifactorial analysis of prognostic factors showed that independent factors associated with a good survival rate were: an age below 40 years at the start of HPN, initiation of HPN after 1987, that was reflecting the experience of the center - and the absence of chronic intestinal obstruction.
8. Conclusions

- HPN is worldwide used in industrialized countries;
- In many European countries as well as in US, cancer has become the main indication for HPN;
- For patients with benign diseases, the main indications are short bowel and chronic intestinal motility disorders;
- The number of HPN centres increased with a variable degree of expertise;
- The prevalence in US is expected to be 10 times higher than in Europe (from 2 to 12/10^6 inhabitants);
- HPN related complications are quite rare and rehabilitation status is good in the majority of the patients.

References