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How good is quality-of-life for children receiving home parenteral nutrition? – A pilot study

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1 **Abstract**

2 **Background & Aims:** Children on home parenteral nutrition and their parents not
3 only engage with complex nutritional issues but also have to manage difficult social
4 and financial problems with social and clinical support that may not always meet their
5 needs. Baxter's HPN-QOL questionnaire, assesses the QOL of adult patients treated
6 with HPN, and has been developed rigorously using standard guidelines, measuring
7 various dimensions of QOL. Our aim was to use this tool to explore how HPN
8 influences the QOL of paediatric patients.

9 **Methods:** The HPN-QOL questionnaire was modified to suit a paediatric HPN
10 population. Data on demographics, aetiology of intestinal failure and duration of HPN
11 were collected from a departmental database. Quality-of-Life grading of functional
12 and symptom scales, HPN specific items and overall QOL Numerical Rating Scales
13 were determined.

14 **Results:** Fourteen out of 17 families returned the completed questionnaires. QOL
15 was significantly impaired by increased dependency regarding items of daily living
16 such as eating, dressing, washing, and mobility, but was not affected in the domains
17 of school attendance, general fatigue, pain and body image. There were no
18 significant differences in QOL when patients with and without enterostomy were
19 compared. Patients felt well supported by the hospital nutrition team in managing
20 logistics related to HPN.

21 **Conclusions:** QOL in HPN patients was not significantly affected by the medical
22 aspects of care. This descriptive study highlights the need for further integration of

- 1 medical and social care in order to support families of children receiving HPN as
- 2 QOL was impaired in relation to activities of daily living and social functioning.

1 **Keywords:**

2 Home parenteral nutrition; Quality-of-Life; Intestinal Failure; Short Bowel Syndrome;

3 Nutrition team

4

5

6 **Highlights**

7 • Baxter's HPN-QOL tool has been adapted for paediatric patients and used to
8 assess QOL

9 • Overall, children on home parenteral nutrition have a good quality-of-life

10 • Improved social care may be of particular value for these families

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1 **Abbreviations**

2

3 CVC: Central venous catheter

4 HPN: Home parenteral nutrition

5 IF: Intestinal failure

6 PN: Parenteral nutrition

7 QOL: Quality-of-life

8

1 **Introduction**

2 Home Parenteral Nutrition (HPN) allows patients with intestinal failure (IF) to
3 be managed in the community during and after intestinal adaptation to bridge the
4 period of adaptation (1,2). Intestinal failure may be temporary in some children with
5 short bowel syndrome so that weaning from PN can occur over time (3). Providing
6 PN to children is a multidisciplinary team effort requiring the expertise of a skilled
7 nutrition team (3–5). Each team member plays a specific role, providing an overall
8 safe package of care centred around the child, with crucial involvement from other
9 professionals, such as interventional radiologist and surgeons. When the child is to
10 be discharged home on PN, support from community services is essential, as well as
11 that from expert parents, who are taught the complex skills required prior to leaving
12 hospital.

13 Issues commonly associated with HPN, including risk of sepsis and protecting the
14 central venous catheter (CVC), represent a major burden of care and responsibility
15 for parents (6). The presence of stoma or an enteral feeding tube adds further
16 complexity and in older children may negatively affect their body image. The
17 potential for complications such as CVC-related sepsis, liver failure, and repeated
18 hospitalisations are undoubtedly a source of anxiety. Leaking stoma bags or watery
19 diarrhoea, excessive flatulence and abdominal pain are embarrassing, distressing
20 and difficult to manage. These issues can have a major impact on patients' quality-
21 of-life (QOL), school attendance, their sense of independence, sleep pattern and
22 parents' ability to work (7). Some HPN patients may over time experience
23 improvement in symptoms related to their underlying condition, but they still require

1 ongoing nutritional support. PN maintains nutritional status and both prolongs and
2 enhances life, resulting in a better overall survival (8).

3 Defining patients' QOL is a difficult task when considering the subjectivity
4 around quantifying a sense of well-being, but parameters such as emotions, social
5 environment, occupations and physical feeling are clearly relevant. We considered
6 that using a standardized, validated and specific HPN questionnaire that had been
7 widely used was the most appropriate method (9). Baxter et al (10) designed a HPN-
8 QOL questionnaire sent to 100 adults receiving HPN. The aim was to assess the
9 impact of treatment on patients' feelings, their functional capacity and symptoms with
10 which they had to cope. This approach allowed a robust and objective evaluation of
11 the QOL on HPN. As no such instrument has been developed for children, our
12 objectives were to modify the Baxter questionnaire for use with paediatric patients
13 receiving HPN and to use it to explore what was important to child and family in
14 order to improve future care.

15

1 **Materials and Methods**

2 **Participants**

3 We conducted a descriptive, cross-sectional study, over a 3-month period
4 (from May 15th to July 30th, 2016), in patients care for by Leeds Teaching Hospitals
5 NHS Trust. All children who received HPN during the study period were invited to
6 participate. Demographics from the cohort were described (age, duration of HPN,
7 aetiology of IF, presence of stoma, family structure and employment).

8 The study was fully discussed with the family in the outpatient clinic where
9 they were given a copy of the questionnaire; consent to participate was implied by
10 return of a completed questionnaire. The questionnaire was anonymous but there
11 was a possibility that families could be identified by nutrition team members through
12 some of the demographic details provided. For this reason, after extracting and
13 pooling data, the questionnaires were destroyed. The primary intention of the study
14 was to explore through assessment of QOL how effective our service was in
15 supporting families. A formal opinion from a Research Ethics Committee was not
16 requested.

17 **Questionnaire**

18 The original 48 item Baxter scale was reduced to 46 items, comprised of the
19 same functional considerations (general health, physical and emotional function),
20 HPN related items (nutrition team and use of ambulatory pump), items about
21 symptoms, and three related to QOL rating. Each item was associated with
22 qualitative answers, composed of four or five scores, which were ordered from worst
23 (score 1) to the best possible answer (score 4 or 5); except for the three QOL

1 numerical rating scales (Fig 1). Increasing positive scores represented better QOL
2 and negative scores poor quality. Our adaptations were made in order to better fit
3 with children's way of life: items related to attractiveness (n = 1), adults' activities
4 such as work (n = 2), shopping (n = 2), or sexual life (n = 3) were removed, while
5 items related to school (n = 3) and to nocturnal stooling (n - 1) were added. The
6 answers to some questions were combined. For example, "do you feel depressed?"
7 and "do you feel tense" both relate to depression. The higher the scores, the better
8 the QOL.

9 The questionnaires were either given to parents attending HPN clinics or
10 posted with a stamped addressed envelope to families at their home. All
11 questionnaires were filled anonymously, either by the parents and/or by the patient if
12 they were old enough to understand.

13

14 **Statistics**

15 Data was collated on Microsoft Excel sheet and analysed using the median
16 values of the reported answers. Chi-square and Fisher tests have been used to
17 compare patients with and without stoma. A *p-value* < 0.05 was considered
18 significant.

19

20

<p>During the past year (or since your child started HPN) Much worse (1) Worse (2) No change (3) Better (4) Much better (5)</p> <p>1. How has HPN made your child feel? 2. How has HPN affected your ability as a family to go on holiday? 3. How has HPN affected your ability as a family to travel?</p> <p>During the past week Not at all (1) A little (2) Quite a bit (3) Very much (4)</p> <p>4 Has HPN felt a burden to you? 5 Have you had concerns about your child's weight? 6 Has the presence of the central line affected their body image? 8 Have you felt supported by your hospital nutrition team? 9 Have you had access to a portable pump? 9b If so, has it improved your ability to get around? 11 Has your child had trouble taking a long walk or playing? 12 Has your child had trouble taking a short walk outside the house? 13 Has your child needed to stay in bed or a chair during the day? 14 Has your child needed help with eating, dressing, washing or using the toilet? 15 Has your child felt tired? 16 Has your child felt lacking in energy? 17 Has HPN disturbed your child's sleep pattern? 18 Are you worried about your child's current health? 19 Have you worried about the future? 20 Is your child able to socialise? 21 Is your child able to do sports/exercise? 22 Is your child able to attend school? 23 Does HPN affect his/her attendance at school? 24 Does HPN affect his/her learning at school?</p> <p>During the past week Not at all (1) A little (2) Quite a bit (3) Very much (4)</p> <p>23 bis Is your child able to take part in hobbies or leisure activities? 24 bis Is your child able to cope with daily life? 25 Is your child able to feel independent?</p>	<p>26 Does your child feel bloated? 27 Is your child able to eat food? 28 Is your child able to drink fluids? 29 Does your child have pain after eating or drinking? 30 Does your child have nausea/vomiting? 31 Does your child have aches or pains in their muscles or joints? 32 Does your child have other pain? 33 Does your child feel depressed? 34 Does your child feel tense?</p> <p>During the past YEAR (or since your child started HPN) Not at all (1) A little (2) Quite a bit (3) Very much (4)</p> <p>37 Has HPN caused financial worries? 38 Does your child have a stoma (ileostomy/colostomy/gastrostomy)? If your child has a stoma please go to questions 39-40 If your child does not have a stoma please go to questions 41-43</p> <p>During the past week - For patients who have a stoma Not at all (1) A little (2) Quite a bit (3) Very much (4)</p> <p>39 Have you had problems caring for the stoma? 40 Have you had problems with the stoma site?</p> <p>For patients who do not have a stoma Not at all (1) A little (2) Quite a bit (3) Very much (4)</p> <p>41 Does your child have frequent bowel movements? 42 Does your child have difficulty with opening bowels? 43 Does your child have painful bowel movements? 44 Does your child open their bowels in the night after going to bed?</p> <p>Please answer each of the following questions by circling the relevant number: During the past week</p> <p>45 How would you rate your child's quality of life on a scale of 1-10? (1: very bad, 10: very good) 46 How has your child's quality of life been affected by HPN? (-5: very much for the worse, +5: very much for the better) 47 How has your child's quality of life been affected by their underlying illness? (-5: very much for the worse, +5: very much for the better)</p>
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1

2 **Figure 1.** The HPN-QOL from Baxter et al. (10) modified for paediatric patients.

3 Each item was associated with a range of numbers (for example: 1, 2, 3, 4, N/A for
4 non-applicable) which had to be circled by the patient or parent(s).

5

1 **Results:**

2 **Demographics**

3 Fourteen out of 17 families returned the completed questionnaire. Thirteen percent
4 of questions did not have a valid answer (i.e. no selected answer for a particular
5 item). During the study period, the Leeds HPN cohort was two third males, aged
6 between 1 and 17 years-old, and the ratio of patients with and without an
7 enterostomy was 4/3. The median (range) duration of HPN was 32 months (8-182
8 m). The main cause of IF was short bowel syndrome (n=10; mostly complex
9 gastroschisis), followed by motility disorders (n=6), such as long segment
10 Hirschsprung's disease, and one case of microvillous inclusion disease.
11 There was one single parent family; three couples were separated and two others
12 had adopted children (Table 1). In four families parents were unemployed.

	Cohort (n=17)
Number of participants (%)	14 (82%)
Male (%)	11 (79%)
Median age in years [range]	4 [1-17]
Patients with a stoma (%)	10 (59%)
Median duration on HPN in months [range]	32 [8-182]
Aetiology :	
Short Bowel Syndrome	10
Dysmotility	6
Microvillous Inclusion Disease	1
Median distance in km from hospital [range]	19 [5-129]
Number of single parents families (%)	4 (24%)
Number of families having a job (%)	10 (71%)

13

14

Table 1: Demographic data for the HPN cohort (n=17)

15

1 **Items of daily living (Fig 2A and 2B)**

2 Overall, parents reported their children were able to cope with daily living activities
3 and to have a social life. An important need to rest was expressed in about one-fifth
4 of these patients, while taking a short walk was very difficult for only 7%. However,
5 half of these families declared problems associated with simple tasks, such as
6 eating, dressing, washing, or resting. For the majority, learning at school, having
7 hobbies and doing sports were considered to be difficult.

8

9 **Parental anxiety (Fig 2C)**

10 Parents did not describe any financial worries and felt supported by the nutrition
11 team. However, items associated with travel, leisure and worries about the future
12 were highlighted, as well as issues related to stoma care.

13

14 **Numerical scales of QOL (Fig 3)**

15 The median overall QOL score was 8 out of 10 (10 was the best answer and 0 the
16 worst), for 13 answers.

17 When considering how QOL was affected by HPN, the median score was 3, on a
18 scale between -5 (worst collected answer: -1) and +5 (best collected answer: +5), for
19 13 answers. For QOL affected by the underlying disease, the median score was 0,
20 on a scale between -5 (worst value) and +5 (best value), for 13 answers.

21

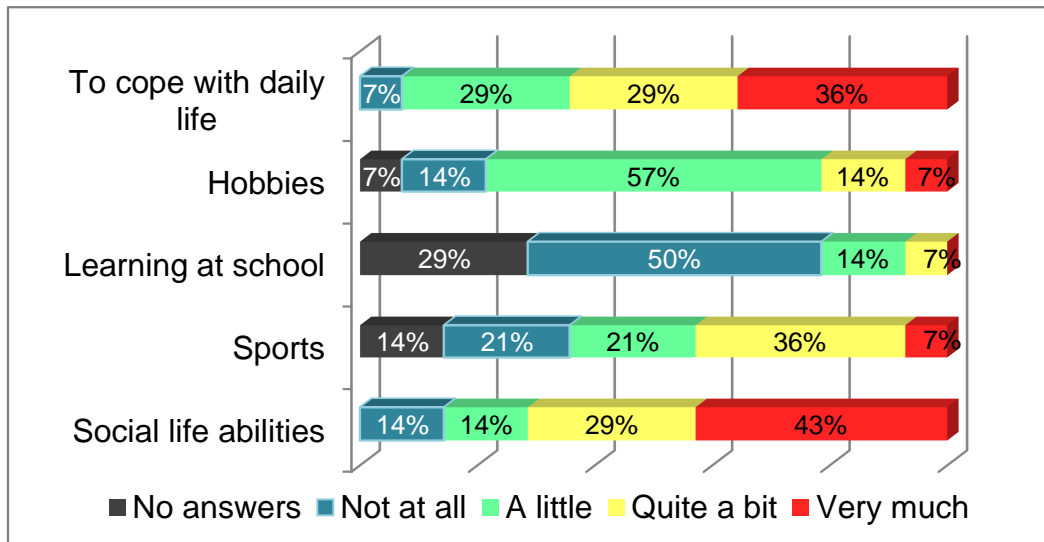


Figure 2 A : Answers about physical and social abilities

On the left side, items regarding children's abilities to do daily life activities. The horizontal bars represent the percentages of different answers to these items. Five different colors correspond to each type of answers : grey means

1

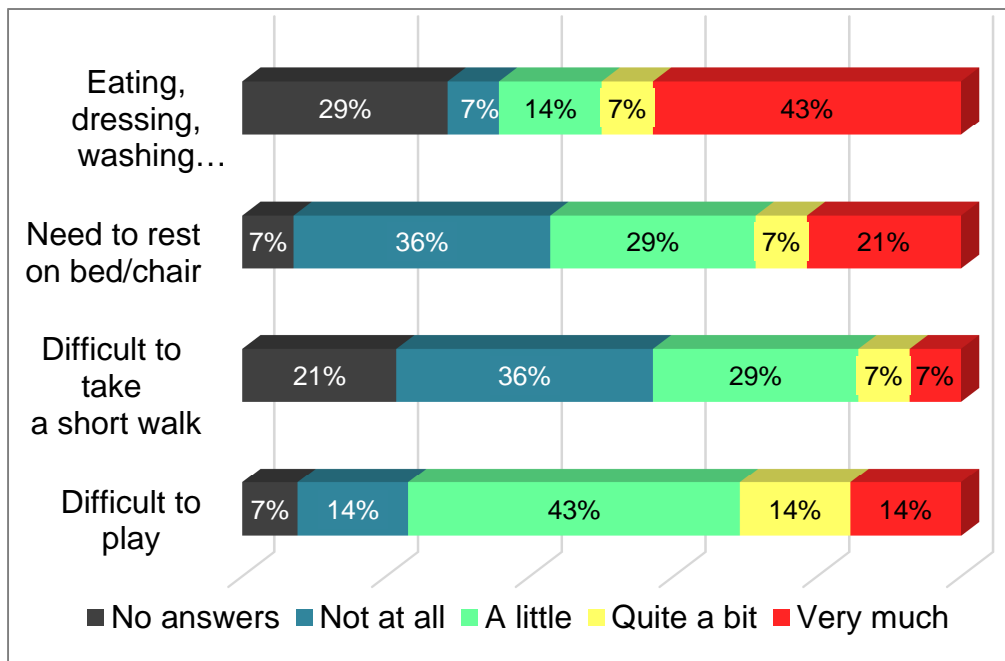


Figure 2B : Answers about dependency

On the left side, items regarding children's dependency with doing simple tasks of daily life.

The horizontal bars represent the percentages of different answers to these

2

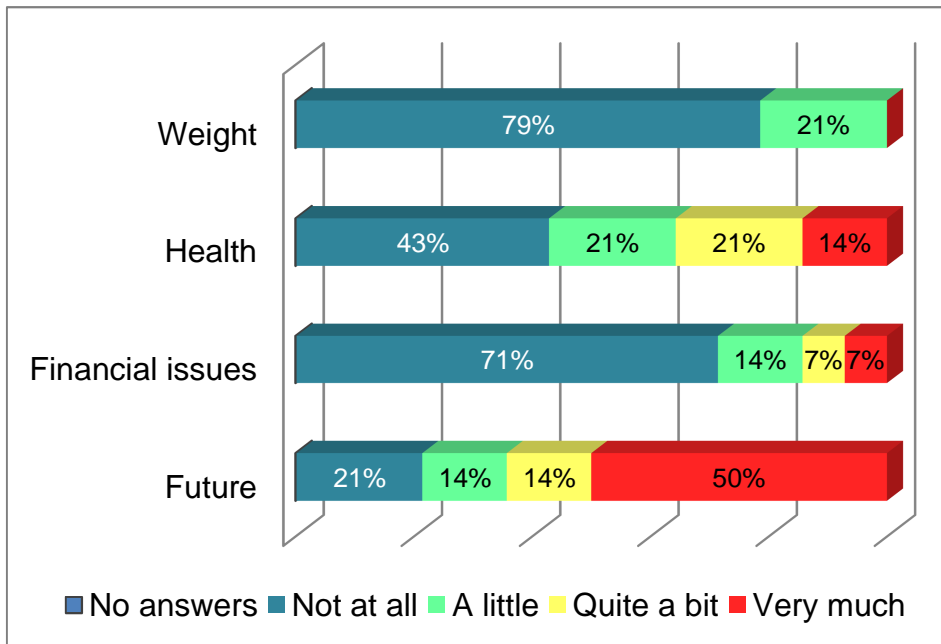


Figure 2C : Answers about parental concerns

On the left side, items regarding parents' concerns about their child's weight, health, their financial difficulties and worries about their child's future.

The horizontal bars represent the percentages of different answers to these items. Five different colors correspond to each type of answers : grey means

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3 **Differences between children with or without stoma**

4 For families completing the questionnaire, no statistical difference was observed

5 between the stoma group (n=8) and the group without stoma (n=6) (Table 2). Four

6 patients were younger than 3 years at the time of the study. School attendances

7 were similar among both groups. Patients without stoma felt better overall and also

8 more able to do sports or exercise, to engage in leisure activities, to socialize and to

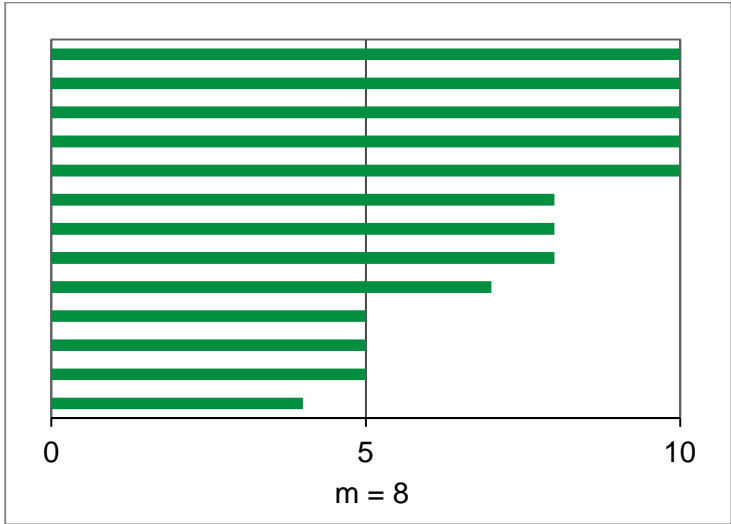
9 go to school. Gastrointestinal symptoms, such as bloating, nausea, vomiting or pain

10 after eating were slightly more frequent among patients without stoma. Interestingly,

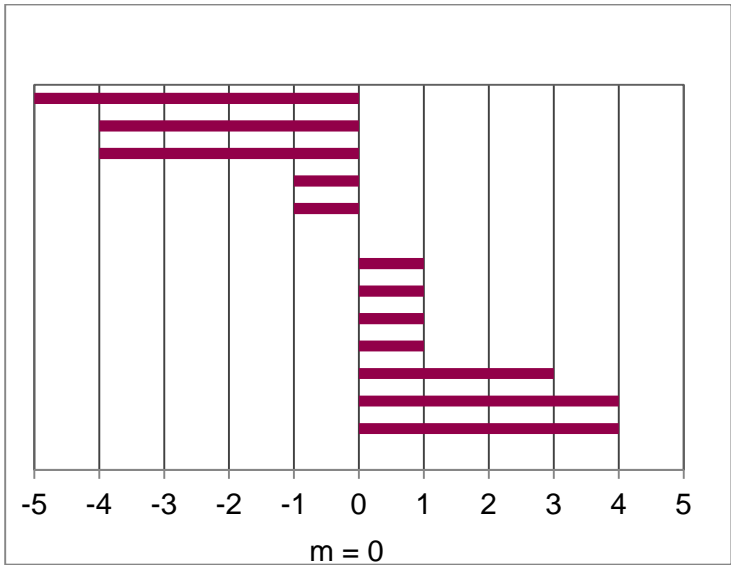
11 there were no statistical differences in QOL items, considering both the overall

12 rating, the QOL affected by HPN rating and the QOL affected by the underlying

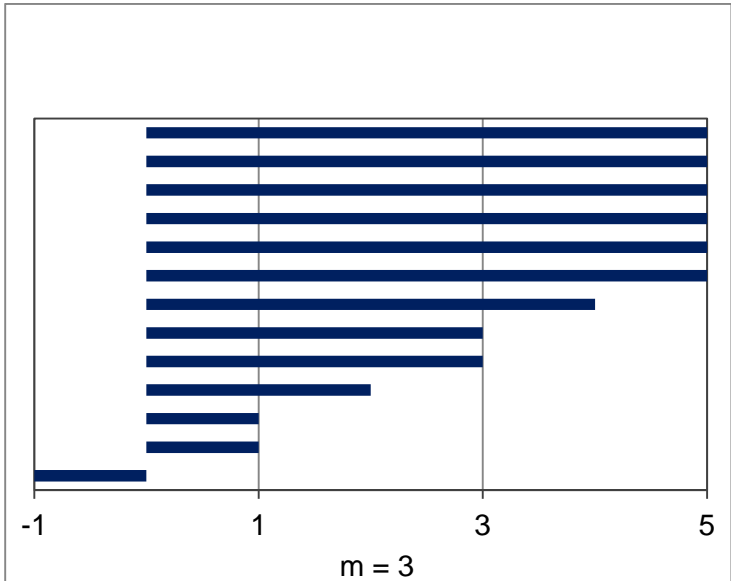
13 disease rating.



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Figure 3. Numerical scales to evaluate the Quality of Life (QOL) of children receiving Home Parenteral Nutrition (HPN): answers from 14 families.

On the upper figure, global score of QOL, between 0 (worse) and 10 (best); on the middle, influence of HPN on QOL, between -5 (much worse) and +5 (much better); on the lower figure, influence of the underlying disease on QOL, between -5 (much worse) and +5 (much better).

m = mean value; each bar corresponds to one answer

Theme	Number of items	Stoma (n=8)	No stoma (n=6)
1. Global feeling: "Much better"	1	3 (38%)	5 (83%)
2. To feel depressed: "Not at all"	2	1 (67%)	3 (63%)
3. Effects on school attendance: "Not at all"	2	5 (64%)	3 (67%)
4. Physical functions: "Very much" ^Δ	4	1 (17%)	2 (41%)
5. To cope with daily life, to feel independent: "Quite a bit"	2	1 (7%)	3 (59%)
6. Fatigue: "Not at all"	2	5 (63%)	3 (42%)
7. Gastrointestinal symptoms [♣] : "Not at all"	3	2 (32%)	2 (45%)
8. Parental anxiety [◇] : "Not at all"	5	4 (48%)	4 (63%)

1

2

Table 2. Adjusted percentages of groups of patients with (n=8) and without stoma (n=6) according to the questions theme.

3

4

Δ: ability to do sports or exercise, to engage in leisure activities, to socialise and to go to school

5

6

♣: bloating, nausea, vomiting or pain after eating

7

◇: burden of undertaking HPN, financial worries and concerns about the child's weight, health and future

8

9

10

1 **Discussion**

2 We have shown in our cohort that QOL of children receiving HPN is globally good
3 (overall score of 8/10). This finding is consistent with the study by Gottrand et al. (7),
4 showing that QOL was similar between HPN-dependent and healthy children,
5 suggesting the effectiveness of coping strategies. In this study, although infants and
6 younger children were more autonomous and needed less attention, adolescents
7 showed lower QOL with regard to social life and concerns for the future. In contrast,
8 in our cohort, there were no significant differences between children with and without
9 stoma.

10 In studies by Engstrom and Emedo (11,12), children receiving HPN were found to be
11 psychologically distressed when assessed by standard questionnaire, and to be
12 aware that their lives were restricted in some respects. . To address negative effects
13 on QOL, Engstrom et al. suggested better contacts with the family through a
14 multidisciplinary team approach and encouraged families to meet together to share
15 their feelings and generate effective mutual support. Similarly, Emedo and co-
16 workers highlighted the importance of support from peers and good counselling as
17 near to home as possible, and extolled the benefits of owning pets. They concluded
18 that the impact of HPN on QOL was far less than the effects of the underlying
19 chronic disease.

20 In a systematic review of the literature of 26 studies including HPN patients aged
21 from 17 to 87 years, treated by HPN from 1 month to 28 years, Huisman de Waal et
22 al. (13) concluded that patients experienced a moderate to good QOL, even though
23 they suffered from depression, fatigue and limited social life. They recommended

1 early interventions in developing coping with these difficulties, starting from when the
2 patient begins to be trained to manage HPN.

3 To determine whether the consequences on QOL are direct results of HPN
4 itself or are associated with the underlying disease is challenging. HPN does allow
5 children to have a relatively normal life outside hospital and for them to mix socially
6 with other children of their age. However, the CVC and the stoma are not only
7 constant reminders of HPN dependence (together with necessary restrictions, for
8 example when swimming or during contact sports), but they are also a constant
9 source of worry in relation to infection, accidental damage or dislodgement. These
10 are the reasons why parents have to be educated very carefully in order to minimize
11 risk of complications. HPN involves high work load and responsibilities and parents
12 know very well how their child's well-being and health might be influenced by their
13 capacity to take care of them. In their study, Friedman-Gruszczyńska et al (14)
14 compared the incidence of catheter infections in trained parents to the hospital
15 pharmacy, and showed that trained parents were safer. That being said, the impact
16 of HPN management on the lives of families caring for their child at home is
17 significant, such that improved social support is likely to be important. In our study,
18 although achieving simple tasks of daily life did not seem to be considered
19 burdensome, parents and their children were likely to encounter difficulties doing
20 them. Almost one-quarter of our families were single-parent ones, for whom social
21 assistance and community nursing support may be essential. This element of care
22 has not always been highlighted in HPN guidelines (15) but needs to be taken into
23 account in clinical practice.

1 After social life, HPN had the next highest impact on freedom of movement,
2 with restricted travel being a common source of frustration. The main limiting factors
3 are stability issues of PN bags, as well as expensive travel insurance. Some airline
4 companies provide medical assistance on request. Charities and hospices are
5 further sources of support for parents, including organisations such as Patients on
6 Intravenous & Nasogastric Nutrition Therapy, Dreams Come True or Destination
7 Florida (16–18) in the UK. Trips abroad require considerable organisation and
8 advanced planning, but can be hugely beneficial experiences for children (19).
9 Parents worries about their child's future were a recurring issue. Among the eight
10 adults with SBS receiving HPN assessed by Carlsson et al. (20), only half of them
11 had a full-time job, perhaps because their situation made them less attractive for
12 employers.

13 Our study suggests that a good QOL can be achieved for HPN patients by
14 developing strong links between the support of a trained hospital nutrition team and
15 the community nursing team. The nutrition team provides ongoing support to families
16 through telephone calls, emails and texts. Training sessions and competency
17 assessment for parents and community nurses include CVC management, aseptic
18 technique, blood sugar testing, starting and stopping intravenous fluids and
19 parenteral nutrition, assessing gastrointestinal fluid losses, management of fever,
20 recognizing an unwell child, as well as safety training for staff in nurseries and
21 schools. Specialist nurses perform periodic home visits (particularly if there are
22 concerns about management), or arrange urgent outpatient or day-case review when
23 necessary. This approach to care is aimed at minimising catheter-related sepsis, the
24 need for readmission to hospital, and the number of routine outpatient visits (once

1 every three months). A weekly multidisciplinary team meeting allows assessment of
2 patients without seeing them, and is informed by feedback on clinical status
3 according to a defined proforma from the community nurse. Furthermore, children
4 can also develop coping strategies when supported by their parents, siblings and
5 their friends. Maintaining good peer relationships during usual childhood activities, as
6 well as taking care of household pets, are undoubtedly sources of happiness in
7 childhood (7), while children's compliance with treatment can be improved with their
8 better understanding of their underlying condition.

9 Our study has several limitations. It was conducted among a small cohort,
10 from one British city. We could have used questionnaires adapted to different age
11 ranges or disease-specific questionnaires. Three families did not complete the
12 questionnaires. We were not able to describe the profile of these non-responders, or
13 determine who filled in the questionnaire (although most of our patients were too
14 young to do this) because of the anonymous design of the study. Comparisons with
15 a control group of patients having the same diseases but without IF and HPN and
16 stratifying the results according to age could bring more precise results; a larger
17 study in the future should incorporate this. The main strength of this study was the
18 use of a detailed questionnaire that has been validated in adults receiving HPN and
19 required only slight modification. The questionnaires were well accepted with a
20 response rate of 82%. Finally, QOL of children treated by HPN is not well described
21 in the literature. It would be useful to repeat this study in a number of other centres
22 working with a collaborative group such as the British Society of Paediatric
23 Gastroenterology. Using the questionnaire in non-English speaking countries would
24 not be justified by this pilot.

1 In the future, follow-up questionnaires to our patients may be helpful in
2 demonstrating improvements after interventions have been put in place, and routine
3 use in HPN clinics will help us to involve young children and families in identifying
4 areas where further support is needed. Reassuringly, our results show that children
5 on HPN can have a good QOL. Key factors in this include ongoing support from the
6 hospital nutrition team following discharge from hospital and close liaison with
7 community services. The burden of care represented by HPN is probably under
8 recognised by social care agencies, and this consideration is most important for
9 those families who already struggle with issues such as being a lone parent or
10 unemployment (21). Greater integration between health and social care services in
11 the future may further assist families who take on such great home care
12 responsibilities.

1 **Acknowledgements**

2 We thank our patients and their families for their contribution.

3

4 **Statement of Authorship**

5 The paper was drafted by Dr Tran and commented on by co-authors. After
6 amendments, the final draft was approved by all authors.

7 **Conflict of Interest Statement and Funding sources**

8 The authors have no conflicts of interest to report and have not received any funding
9 for this study.

10

11

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