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Original article

The continued rise of paediatric home parenteral nutrition use: Implications for service and the improvement of longitudinal data collection

Andrew R. Barclay [a, *], Paul Henderson [a], Henry Gowen [b], John Puntis [c], BIFS collaborators

[a] Department of Paediatric Gastroenterology, The Royal Hospital for Sick Children, Glasgow, UK
[b] Children's Liver Diseases, Research Unit, Birmingham Children's Hospital, UK
[c] The Department of Paediatric Gastroenterology, The General Infirmary at Leeds, UK

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SUMMARY

Background & aims: Robust data from the United Kingdom (UK) regarding the current epidemiology of patients with types II and III intestinal failure (IF; ≥28 day parenteral nutrition; home parenteral nutrition) are limited. We aimed to analyse trends in type II and III IF in children in the UK using historical and novel data.

Methods: A point survey of the 32 nutrition support teams that register patients with the British Intestinal Failure Survey was carried out in November 2012. Basic demographics for patients on home parenteral nutrition and receiving parenteral nutrition for ≥28 days were collected. Data were anonymised, collated by the registry coordinator and compared to previous surveys by the British Paediatric Surveillance Unit in 1993 and data from 2010.

Results: All 32 participating centres responded giving complete UK ascertainment. There were 195 type III patients, representing a four-fold increase since 1993. The proportion of patients with short bowel syndrome had almost doubled from 1993 (27% vs. 50% $p = 0.001$). The ratio of type II to type III IF patients varied considerably between centres.

Conclusion: These data suggest that type III IF point prevalence has risen in the short term, coincident with individual centres' reporting improved survival in IF. Refinement in the methodology for prospective data collection is needed to gather more accurate incidence, period prevalence and outcome data for UK type II and type III IF.

1. Background and objectives

Parenteral nutrition (PN) is the treatment of choice for children with established intestinal failure (IF). Type II IF can be defined as needing greater than 50% of calories parenterally for ≥28 days post term corrected gestational age [1–4] and can be separated into three main categories by pathogenesis. These are: (1) short bowel syndrome (SBS) [5]; (2) neuromuscular disorders of the gastrointestinal tract including long segment aganglionosis (Hirschsprung's disease) and congenital intestinal pseudo-obstruction syndromes (CIPOS) [6]; and (3) congenital enterocyte disorders [7,8]. Ideally, such a complex and resource-intensive treatment such as long term PN should be provided from an appropriately staffed specialist centre with a multidisciplinary nutrition support team (NST), the latter having been shown to be cost effective and to reduce complication rates [9–11]. Home care (HPN) is advisable with long term PN dependency as it reduces sepsis risk, consequent IF-associated liver disease, and improves quality of life [12]. Patients requiring PN for greater than 3 months, those receiving HPN or being prepared for HPN are defined as type III IF [1,12]. The demand for long term PN has risen in the last two decades due mainly to increased numbers of preterm infants surviving after surgical intervention for necrotising enterocolitis [13] and a rise in the incidence of congenital gastroschisis [14].

The British Intestinal Failure Survey (BIFS) was set up in 2005 by the British Society of Paediatric Gastroenterology Hepatology and Nutrition (BSPGHAN) and the British Association of Paediatric

* Corresponding author. Department of Paediatric Gastroenterology, Royal Hospital for Sick Children, Yorkhill, Dalnair Street, Glasgow G3 8SJ, UK. Tel.: +44 0141 201 0313; fax: +44 0141 2010 0303.
E-mail address: andrew.barclay@ggc.scot.nhs.uk (A.R. Barclay).

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Surgeons (BAPS) to examine trends in paediatric type II and type III IF (http://bspghan.org.uk/working_groups/nutrition.shtml). The two arms of BIFS comprised a longitudinal registry of type II IF and an annual point prevalence survey of type III IF. A previous BIFS point prevalence survey demonstrated a significant rise in UK type III IF in comparison to a 1993 British Paediatric Surveillance Unit survey (4.4–13.7/million population) [15,16]. Recent studies have reported improved short term IF outcomes due to specialist multidisciplinary care from NST [17,18], novel medical therapies [19] and surgical intervention [20,21]. This suggests an ongoing rise in patients surviving type II IF and thus requiring HPN. However the 2010 BIFS survey did not examine the change in relative proportions of underlying diagnoses for type III IF patients. In addition, the previous surveys did not record the prevalence of in-patients with type II IF (who may go on to require HPN) known to individual NST. We aimed to perform a further census of paediatric IF units in the UK to examine the underlying trends in prevalence and demographics in type II and type III IF.

2. Methods

An Excel spreadsheet (Microsoft Corporation, Redmond, WA) was sent to the local investigators for BIFS at the 32 NHS Hospital Trusts in Scotland, England, Wales and Northern Ireland with a specific multidisciplinary NST working in the framework of a paediatric gastroenterology service, and who are already cooperating with the BIFS. Data were requested for children (<16 yrs) with type II IF (in-patient ≥28 days PN) and type III IF (HPN or imminent discharge for HPN) on the 26th November 2012, including: patient initials; year of birth, diagnostic category and first half of home postcode. The information from the postcode was used to identify patients by their place of residence, rather than hospital providing PN, in order to obtain a more accurate regional prevalence and exclude the possibility of double-counting. Replies from the participating Trust were anonymised by the BIFS administrator. Statistics were accessed from the relevant office for England, Scotland, Wales and Northern Ireland to provide population estimates for children less than 16 years at the time of the study [22–25]. Data were compared with the 1993 BPSU survey [16] and the 2010 BIFS survey [15]. Comparisons of changes within centres between 2010 and 2012 were calculated using a paired *t*-test; calculations of prevalence over time were calculated using Poisson regression with correction for over dispersion. Statistics were performed in GraphPad Prism v4.03 (GraphPad Software, CA, USA) and R v2.14.1 (R Foundation for Statistical Computing, Vienna, Austria). Ethical committee review for the survey was not required.

3. Results

All 32 centres responded to the survey, corresponding to services caring for all of the UK population under 16 years (11.8 million). A total of 95 type II IF and195 type III IF patients (171 HPN; 24 being prepared for HPN) were reported. Type III IF patient demographics and comparison with other surveys are displayed in Table 1. Calculated point prevalence of type III IF has risen significantly from 4.4/million in 1993 to 16.6/million at risk population in 2012 ($p < 0.001$). The rise from 13.9/million in 2010 to 16.6/million at risk in 2012 was not statistically significant at the population level ($p = 0.38$), but represented a 19% increase in clinical workload. Proportions of patients by diagnostic category changed significantly from 1993 to 2012 with those with SBS rising from 27% to 50% ($p = 0.001$) and those without a diagnostic category falling from 37% to 4% ($p < 0.001$).

Data for type II and type III IF caseload per individual centre is shown in Fig. 1. As a whole, patients with type II IF, that NST were aware of, represented only 31% of the total IF case-mix. However, for larger centres (≥2 type III IF cases) the proportion of patients with type II IF varied from 0% of total caseload (centre 16) to 75% of total caseload (centre 15).

4. Discussion

The point prevalence of paediatric HPN has risen four-fold in the last two decades and these data suggest that the observations of the 2010 survey, this trend continues. The findings agree with other work suggesting that specialist services for paediatric type II and type III IF continue to grow [3,4,15,26–28]. The close working collaboration between the BSPGHAN and the BAPS, the centres contributing to the BIFS, and the centralisation of HPN provision contracts, means we are confident of complete ascertainment for the UK and of our findings that the point prevalence of type III IF continues to be much greater than two decades ago. Our intentions repeat of our survey in 2014, will give us formal epoch data that may clarify whether an increase in point prevalence has occurred within this decade. The increase in the proportion of SBS patients requiring HPN is most likely to be genuine and multi-factorial including: the increased survival of extremely preterm infants who are at risk of NEC after the first week of life [13,29]; increasing incidence of gastroschisis [14]; greater therapeutic morale with a more aggressive surgical approach to patients with extensive NEC or gastroschisis [30] so that ultra-short segment SBS is not seen as a barrier to survival [31]; improved prevention and treatment of IF-associated liver disease reducing mortality from this complication [19,32]. Whilst we have complete ascertainment for this point prevalence study, the limitations of the use of point prevalence data have been discussed in previous regional survey and been shown to potentially underestimate total service needs by 50% in comparison to period prevalence data [28]. Therefore improvements in prospective longitudinal survey of Type II IF incidence and prevalence are warranted.

The marked difference in relative ratios of type II to type III IF between individual centres is of note (e.g. Centre 16 where type II IF represented 0% of NST caseload vs. Centre 15 where type II IF represented 75% of NST caseload). This highlights the marked variability in service design and potential under-reporting of type II IF across the UK. Criteria for referral to specialist NST vary from unit to unit, from being triggered by a patient receiving 28 days of PN, to only when a patient has been identified as requiring HPN. Historically the time of ≥28 day PN for defining IF was based on the observation that the vast majority of patients were PN dependant

Table 1
Comparison of patient numbers, point prevalence and demographics of patients with type III IF in three surveys BPSU 1993, BIFS 2010 and BIFS 2012.

Survey (year)	Type III IF	Calculated point prevalence/million population at risk (<16 yr)	Short bowel syndrome (%)	Enterocyte disorder (%)	Neuromuscular (%)	Other (%)
BPSU(1993)	66	4.4	18 (27)	12 (18)	12 (18)	24 (37)
BIFS (2010)	164	13.7	–	–	–	–
BIFS (2012)	195	16.6	99 (50)	44 (23)	44 (23)	8 (4)

IF, intestinal failure.