

THE FUTURE OF INTESTINAL FAILURE SERVICES
IN ENGLAND AND WALES

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on behalf of the BSPGHAN

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Executive summary

- following a meeting with the National Service Commissioning Group (NSCAG) in January 2004, the BSPGHAN membership was asked to comment on the desirability and role of supra-regional intestinal failure centres; feedback is summarised below
- there was much positive support for optimising management of intestinal failure, but with the proviso that services remain primarily regional rather than supra-regional (i.e. without the remit of NSCAG)
- there was support for an NSCAG funded intestinal transplant assessment centre, and acknowledgement of the role of such a centre in providing ongoing management advice in patients not listed for transplantation
- there should be further exploration of the possible need for a second intestinal transplantation centre
- there was recognition of the need for integration of gastroenterology and hepatology services within supra-regional liver transplantation units for effective joint management of patients developing liver disease while requiring long term parenteral nutrition

Introduction and Background

In January 2004, representatives from the BSPGHAN and BAPS (British Association of Paediatric Surgeons) were invited to meet with members of the National Service Commissioning Advisory Group (NSCAG) at the Department of Health to discuss the future configuration of services for children in England and Wales with intestinal failure. Although Birmingham has remained the single provider for small bowel transplantation in the UK (a service funded by NSCAG under its specific remit to support national or specialised services for rare diseases) discussions focused on whether Birmingham should be seen as having a wider remit, and included the possibility of further NSCAG funding for one or two additional 'supra-regional intestinal failure units'.

Some background data relating to Birmingham activity were presented. Over the past 13 years, 152 referrals have been assessed by the intestinal transplantation unit. Transplantation was judged not to be indicated in 70 patients, and 28 others were thought unsuitable; 54 were recommended for transplantation. In all, 32 patients have been transplanted, although in recent years, this has been isolated liver transplantation rather than combined liver and small bowel. Developments in management of complex intestinal failure including isolated liver transplantation and non-transplant surgery (bowel tapering or lengthening) therefore raise the question as to whether Birmingham should more appropriately be designated as a supra-regional centre for managing complex intestinal failure rather than specifically as a small bowel transplantation unit. If so, was there a case for having several such supra-regional

intestinal failure units (even if small bowel transplantation was confined to only one site), and what patients would it be appropriate to refer?

The meeting concluded with an agreement to canvass opinion among the membership of both the BSPGHAN and BAPS. A document summarising the issues (Appendix) was circulated electronically to all BSPGHAN members on three different occasions and comment invited. Replies were obtained from 12 specialist centres providing tertiary gastroenterology services; some of these were individual rather than institutional responses. Three paediatricians with an interest in gastroenterology working within district general hospitals also replied. The views expressed by the membership are summarised below. A further discussion meeting has been arranged with NSCAG in September 2004.

Is intestinal failure (when separated from bowel transplantation) a specialised and vulnerable service that is appropriate for NSCAG designation?

There was broad consensus that intestinal failure (and in particular home parenteral nutrition programmes) require additional funding and currently often represent an example of innovative service development without adequate resources. In arguing that regional services need supporting and developing, the role of NSCAG is, de facto, negated. A minority view expressed by five regional centres was that intestinal failure should come under the remit of NSCAG, with between 1 and 3 identified supra-regional units. Logically, these should be sited in currently designated supra-regional liver transplant centres since the main issues at stake were assessment of parenteral nutrition associated liver disease, and the possibility of isolated liver transplantation. Two tertiary referral units felt that consideration should be given to a

second intestinal transplant unit. However, the majority of respondents were not in favour of supra-regional intestinal failure services, other than for assessment of those patients who might require small bowel transplantation. There was acknowledgement of the important role performed by Birmingham in this respect, the accumulated experience in this centre and the combined availability of both gastroenterology and hepatology expertise.

Common problems of long term parenteral nutrition (e.g. catheter related sepsis) and training needs dictate that management needs to be 'local', although based in a centre with multidisciplinary nutritional care team, gastroenterologist, surgeon, etc. working in close liaison with hepatologists. In other words, management should be supervised in tertiary level gastroenterology units rather than district general hospitals. Potential disadvantages of NSCAG designation for intestinal failure include over centralisation, de-skilling of regional units, and an unnecessary burden of travelling for families and patients. Such concerns lead some to conclude that 'complex intestinal failure' comprised only those cases requiring small bowel transplantation. Of course, provision of small bowel transplantation ideally starts with an assessment of high risk patients before major complications have occurred. Not all of these will merit (or be suitable for) transplantation; follow up is required for determining outcomes, and other interventions (isolated liver transplantation, non-transplant surgery) may be appropriate. These aspects of an intestinal transplantation unit deserve recognition by NSCAG.

What kind of patients should be referred (i.e. what constitutes ‘complex’ intestinal failure)?

The suggested definitions of complex intestinal failure (see ‘Appendix’) were broadly accepted. With the exception of bowel transplantation and life-threatening difficulty maintaining venous access issues, complex patients could be managed in appropriately staffed and funded regional centres.

How many supra-regional intestinal failure units would be ideal?

Among the minority of respondents supporting the concept of NSCAG funded supra-regional intestinal failure centres, 1-3 units based on current supra-regional hepatology services were proposed.

Information was requested from BSPGHAN members regarding the number of children PN dependent for ≥ 4 weeks and ≥ 12 weeks, in addition to numbers of home PN patients managed during 2003.

Almost no information in response to this request was received. This probably reflects a lack of readily available data, emphasising the problems we have with defining the scope of the problem of intestinal failure, and represents a significant future challenge for the Society.

What facilities/expertise should be available at such a centre?

All intestinal failure should be managed by an expert multi-disciplinary team working in close collaboration with a hepatology unit. Shared care protocols for home parenteral nutrition need to be developed in conjunction with referring district general hospitals. Supra-regional services would principally provide for small bowel

transplantation, and offer advice on non-transplant management of those patients referred for assessment.

Future challenges for the BSPGHAN

Comments received reflect a consensus that the Society should be striving to implement and maintain the highest possible standard of care for children with intestinal failure. Rising numbers of children both with extreme prematurity/necrotising enterocolitis and complex gastroschisis means that numbers of patients are likely to increase. The Society should be working towards national standards for clinical care, shared care protocols, collaborative research, and a supporting network for definitive diagnosis. An intestinal failure registry and home PN register need to be coordinated in conjunction with BAPS, and BAPEN/BANS (British Association for Parenteral and Enteral Nutrition/British Artificial Nutrition Survey) in order to define the level of need. Funding of regional gastroenterology services in a way that reflects work performed is also an area of priority. The Society should consider setting up a permanent intestinal failure committee, which would liaise with the similar ESPGHAN body.

APPENDIX

Introduction

Intestinal failure services for children are currently managed regionally.

Improvements in paediatric intensive care, medical and surgical facilities in the past twenty years has resulted in much greater numbers of children receiving parenteral nutrition for prolonged periods in hospital and at home. Birmingham is currently the single provider for small bowel transplantation in the UK with greater than 50% long-term survival. The service is funded by the National Specialist Commissioning Advisory Group (NSCAG) who fund national services or specialised services for rare diseases. The current service agreement with NSCAG covers the cost of assessment for potential transplant candidates, the transplant episode and follow up.

The current contract does not include the management of those children with complex intestinal failure in whom small bowel transplantation may not be necessary. In recent years such management has included isolated liver transplantation rather than combined small bowel and liver transplantation in children with short bowel syndrome.

This has led to consideration of setting up supra-regional units for the management of children with complex intestinal failure as such patients are small in number and require concentration of expertise.

NSCAG has asked the BSPGHAN to comment on the need for supra-regional centres for the assessment and management of patients with complex intestinal failure. We have not been asked to comment on whether there should be more than one unit performing small bowel transplantation.

Potentially there may be increased funding for children with intestinal failure, and possibly the development of additional supra-regional centres.

The size of the problem

We do not have accurate knowledge of the numbers of patients requiring long-term parenteral nutrition (PN) nationally, and 'complex intestinal failure' is even more difficult to delineate.

If intestinal failure is defined as the need for PN for greater than four weeks there are clearly many patients already being managed in regional centres by gastroenterologists and/or surgeons, often through the agency of multidisciplinary nutritional care teams. If the definition is arbitrarily extended to 6 (or even 12) weeks of PN, the numbers will be less.

For the management of children with intestinal failure complicated by liver disease, regional paediatric units will need either to have hepatology services on site or close links with supra-regional liver units.

Consultation, and report back to NSCAG, September 2004

We are now in the period of a consultative process, involving both members of the BSPGHAN and the British Association of Paediatric Surgeons (BAPS), with feedback to NSCAG scheduled for September 2004. We are therefore requesting your view as a member of the BSPGHAN on this important issue and would be grateful for any comments in relation to how services for intestinal failure should develop.

Specific questions to be addressed include:

1. Is intestinal failure (when separated from bowel transplantation) a specialised and vulnerable service that is appropriate for NSCAG designation?
2. What kind of patients should be referred (i.e. what constitutes 'complex' intestinal failure)?

Possible examples of patients that might be referred include:

- Patients with intestinal failure in whom diagnostic uncertainty persists (e.g. protracted diarrhoea, motility disorders)
- Short gut patients not making reasonable progress with enteral feeding (e.g. tolerating <50% of their nutritional requirement enterally after 6 months of PN)
- Patients with progressive liver dysfunction (rising conjugated bilirubin after three months on PN; not tolerating an increase in enteral feeding)
- Patients in whom venous access is difficult and there is a real possibility that PN cannot be maintained in the near future

- Patients with short gut in whom surgery aimed at bowel lengthening or improving motility (gut tapering) might be considered as a means of establishing full enteral nutrition
 - Patients with recurrent episodes of life threatening catheter related blood stream infection
3. How many supra-regional intestinal failure units would be ideal (1, 2, 3?; it is likely that location would be the subject of a formal bidding process)

Our initial view is that a strong case can be made for more than one intestinal failure centre, but that further development should not be at the cost of weakening current regional services.

The role of the supra-regional intestinal failure unit as part of a managed clinical network could be

- To assess and advise on the management of the small number of complex cases
- Collaborate in the development of management protocols
- Aid in diagnostic difficulty
- Evaluate new treatments
- Support regionally based services
- Facilitate research

The majority of children with intestinal failure would remain within region as now.

In addition, the supra regional unit may also facilitate the standardisation of home parenteral nutrition (HPN) services, although we would not anticipate the need for all HPN patients to be referred to a supra-regional intestinal failure centre. One could therefore envisage most children managed within region with a register of children on home parenteral nutrition. There would be a facility for children from centres with only small numbers of children on HPN to be seen for review in the larger unit if appropriate.

In addition to general views we would also be interested in a number of specific responses including number of children who were PN dependent for 4 weeks or more (and 12 or more) in your centre, and how many home PN patients you managed, during 2003 if these data are available.

What constitutes Complex Intestinal Failure (time frame, clinical condition)?

Whether you feel such patients should be managed in conjunction with a supra-regional centre.

What facilities/expertise should be available at such a centre?

We are grateful for your input so as to ensure that views expressed represent the society's view on this important topic

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On behalf of the BSPGHAN

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