

# **Intestinal failure: recommendations for tertiary management of infants and children**

A Report by the Intestinal Failure Working Group,  
British Society of Paediatric Gastroenterology, Hepatology and Nutrition,  
and the British Association of Paediatric Surgeons

John Puntis (Chair); Gastroenterologist, Leeds

Sue Beath; Hepatologist, Birmingham

Mark Beattie; Gastroenterologist, Southampton

Mark Dalzell; Gastroenterologist, Liverpool

Mark Davenport; Hepatobiliary Surgeon, London

Michelle Gabriel; Administrator, Birmingham

Sue Hill; Gastroenterologist, London

Chris Holden; Nutrition Nurse Specialist, Birmingham

Sarah Macdonald; Dietitian, London

Nigel Meadows; Gastroenterologist, London

Victoria Magnall; Pharmacist, Liverpool

Ian Sugarman; Gastrointestinal surgeon, Leeds

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## **Abbreviations**

BANS – British Artificial Nutrition Survey

BAPEN – British Association for Parenteral and Enteral Nutrition

BAPS – British Association of Paediatric Surgeons

BIFS – British Intestinal Failure Survey

BSPGHAN – British Society of Paediatric Gastroenterology, Hepatology and Nutrition

CRBSI – catheter related blood stream infection

CVC – central venous catheter

HPN – Home Parenteral Nutrition

IF – intestinal failure

MRI – magnetic resonance imaging

NSCAG – National Specialist Commissioning Advisory Group

NST – Nutritional Support Team

PN – Parenteral nutrition (intravenous feeding)

SBS – short bowel syndrome

STEP – serial transverse enteroplasty

## **Introduction**

The National Specialist Commissioning Advisory Group (NSCAG) was established in 1996 by the Department of Health to advise Ministers on the provision of specialised/complex medical services. The current view of the British Society of Paediatric Gastroenterology, Hepatology and Nutrition (BSPGHAN) and the British Association of Paediatric Surgeons (BAPS) is that the management of intestinal failure (IF) should be provided by adequately resourced regional gastroenterology centres (Appendix 1). Although IF services will therefore remain outside the remit of NSCAG, the BSPGHAN has been asked to clarify indications for referral of patients with IF to a regional centre. In response, an IF Working Group was formed and has developed this position statement through meetings and e-mail discussion among members. The composition of the group is shown on page 1 of this document.

The Working Group has considered the expertise, staff, and facilities necessary to provide a regional IF service (Appendix 2). The term 'Intestinal Failure Unit' here refers to a department that is providing regional diagnostic and management services (both medical and surgical) for children with complex gastrointestinal disorders, and in addition to which, is also able to support a home parenteral nutrition (HPN) service (see below). An example of a 'patient journey' (Appendix 3) is presented in order to illustrate to Commissioners the range of personnel and facilities that are necessary to run such a service. Some unpublished data from West Yorkshire is included (Appendix 4) giving information regarding causes and frequency of IF.

## **Intestinal failure - definition and demographics**

IF refers to the situation where gastrointestinal disease precludes adequate digestion and absorption of nutrients to support homeostasis and normal growth. In relation to an anatomically short bowel it is defined as ‘a reduction in the functioning intestinal mass below the amount necessary for adequate absorption to allow for growth’ (1). Survival therefore depends on provision of nutrients directly into the bloodstream (parenteral nutrition – PN), and the avoidance of life threatening complications directly related to the need for this intervention. There are no accurate data regarding numbers of patients with IF. Extrapolating from a recent survey in West Yorkshire (Appendix 4), there may be around 1100 children in the UK annually who receive PN for more than one month. A little over two thirds of these are premature newborn, who have an excellent prognosis. Children needing longer term PN (a few of whom may ultimately need small bowel transplantation, and many of whom are candidates for HPN), are largely comprised of those with short bowel syndrome (SBS; secondary to congenital or acquired gastrointestinal disease), enteropathy, or abnormal gut motility presenting as pseudo-obstruction (2, 3).

In SBS, adaptation of the gastrointestinal tract occurs so that in a high proportion of affected children enteral feeding can be increased slowly until PN is no longer essential to sustain growth. During this process of adaptation (often lasting several years), episodes of life-threatening infection may occur, together with the development of liver disease, venous thrombosis, disordered eating behaviour and a range of important nutritional deficiencies. Such problems are also seen with other causes of IF in which there is less prospect of progressing to full enteral nutrition. When managed in an experienced centre, 90% of children can be expected to survive

(4). However, much time must be invested by specialist staff (with key personnel usually organised as a multidisciplinary Nutritional Support Team - NST) in order to promote growth and development while avoiding life threatening complications.

Long term management includes discharge from hospital and administration of PN at home (HPN) when this is feasible. The British Artificial Nutrition Survey (BANS) registered 81 children receiving HPN from 1996 –1999 (5); these data are incomplete due to inconsistent ascertainment. Some children are unsuitable for HPN as clinical instability dictates the need for hospital care. Others may remain in hospital for an extended period of a time for ‘social’ reasons (including inadequate housing or parents who are unable/unsuitable to take on the responsibility of home care), until PN is no longer required. Discharge planning for HPN together with training of parents, liaison with community services and follow up is an important role for the nutrition nurse specialist, working as part of a multidisciplinary NST.

### **Management of children with intestinal failure**

Within a regional centre appropriate staff (Appendix 2) should be organised as a multidisciplinary NST comprising consultant gastroenterologist, surgeon, nutrition nurse specialist, pharmacist, and dietitian as core members. An NST is required for optimal nutritional support to both enterally and parenterally fed patients (6, 7). Where NSTs have been developed, this has been on an ad hoc basis, without adequate funding and recognition of staff time. We consider the role of an NST as crucial in caring for children with IF and therefore essential to providing an IF service. HPN is a complex process that requires investment in infrastructure. Sufficient numbers of patients are needed to ensure adequate expertise. Care should be shared with referring

hospitals as a part of a managed clinical network (for example the Scottish Home Parenteral Nutrition Managed Clinical Network).

The most difficult patient management problems relate to maintenance of venous access, treatment of catheter related blood stream infection (CRBSI), prevention and management of complications such as liver disease, weaning from PN (balancing enteral and parenteral intakes), teaching carers techniques necessary for HPN, and maintaining oromotor feeding skills. Expert advice should be available for HPN patients on a 24 hour basis, and acute readmission must be available if needed.

There should be designated medical staff (usually surgeons, but sometimes interventional radiologists or anaesthetists) - with a named consultant lead - able to provide a CVC placement service, with expertise in resolving mechanical catheter related problems including occlusion and catheter fracture. Bowel lengthening procedures (e.g. Bianchi operation, or serial transverse enteroplasty – STEP) as well as surgery to improve motility (e.g. tapering or plication) are interventions that are carried out where anatomically appropriate and may have a crucial role in some children who might otherwise remain PN dependent.

There is an extensive literature on the importance of a multi-disciplinary team in patient management and prevention of complications (7, 8, 9, 10, 11, 12, 13). Referral to a regional IF unit can reduce the patients' level of dependency on PN through management strategies well described elsewhere (14,15,16), These include a range of medical (17,18,19,20,21), dietetic (22,23) and surgical treatments (24,25,26,27,28).

### **Appropriate facilities for a regional unit serving a population base of 3 million**

As stated previously there are no accurate data for numbers of children with IF, but an NSCAG sponsored registry (British Intestinal Failure Survey - BIFS) is in progress to address this issue. For a population of three million people we estimate that four beds are required to manage the initial phase of IF and an additional two beds to manage the 10-15 infants and children who remain on PN for six months or more. Access to paediatric hepatology services should be through a managed clinical network; currently there are three supra-regional hepatology centres with which regional IF units would be expected to have close communication.

### **Evidence based practice**

A comprehensive evidence based review of paediatric PN has recently been published (29). The IF unit should adopt an evidence based approach to clinical work, supported by national guidance and protocols adapted for local use. These should include, for example, diagnosis and management of CRBSI, management of a blocked CVC and monitoring of PN. There should be regular meetings between IF units in order to discuss complex cases and audit practice, management strategies, and patient outcomes.

### **Indications for referral to a regional intestinal failure unit**

The majority of infants with congenital anomalies of the gastro-intestinal tract are identified antenatally and delivered in a regional neonatal surgical unit. However,

some infants are delivered outside a regional unit, or the onset of IF occurs later in childhood. Transfer to a comprehensively staffed IF unit may be delayed especially if initial presentation is with protracted diarrhoea. In Table 1 we present recommendations regarding referral criteria intended to simplify the process of transfer and ensure it is timely. It is incumbent on the regional unit to maintain excellent communication with the referring hospital and that where possible, shared care (e.g. out-reach clinics) be undertaken to minimise disruption of family life and assist seamless integration between hospital and community services. The latter is essential for successful HPN.

**Table 1** Criteria for consultation/referral to an IF unit for infants and children

<b>Criteria</b>
<ul style="list-style-type: none"> <li>• Children with massive small bowel resection</li> <li>• All children with congenital enteropathies who are destined for life-long PN dependence</li> <li>• Diagnostic/prognostic uncertainty (e.g. uncharacterised protracted diarrhoea)</li> <li>• All children requiring PN for more than 28 days</li> <li>• Hyperbilirubinemia (greater than 50 micromol/L)</li> <li>• Vascular access problems in patients receiving long-term PN</li> </ul>

### **Criteria for referral for Small Bowel Transplant Assessment**

While acknowledging that it is often difficult to make a perfectly timed referral, it is better to refer too early than too late. Those patients who are at high risk of developing life threatening complications of PN (Table 2) should be referred for assessment to the IF/Small Bowel Transplant unit at Birmingham Children's Hospital. In 2004/2005, 42 patients were referred to Birmingham Children's Hospital for consideration of small bowel transplantation, of whom 22 were listed for transplantation (nine patients received small bowel transplants combined with liver,

one child received an isolated small bowel transplant, and five patients received liver transplant only, because their underlying disease of short bowel syndrome was expected to improve). Due to a combination of late referral and shortage of donated organs, four patients died before surgery could be performed, and three patients were still on the waiting list at the end of 2005. The BSPGHAN consider that thought should be given to the requirement for a second UK small bowel transplant centre (see Appendix 1). The following recommendations are consistent with international criteria (30).

**Table 2** Criteria for consultation/referral for Small Bowel Transplant Assessment

<b>Criteria</b>
<ul style="list-style-type: none"> <li>• Children with massive small bowel resection</li> <li>• Children with severely diseased bowel and unacceptable morbidity</li> <li>• Continuing prognostic/diagnostic uncertainty</li> <li>• Microvillous inclusion disease</li> <li>• Persistent hyperbilirubinemia (&gt;100 micromol/L)</li> <li>• Thrombosis of two out of four upper body central veins</li> <li>• The request of the patient/ family</li> </ul>

The decision to offer transplantation is a complex one and depends on individual factors as well as those outlined above. Small Bowel Transplantation is generally only recommended in PN dependant patients if that patient is considered to have less than a 50% chance of surviving more than 12 months without transplantation and where small bowel transplantation can reasonably be expected to improve quality of life and survival.

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## **Appendix 1**

### **THE FUTURE OF INTESTINAL FAILURE SERVICES IN ENGLAND AND WALES**

Report prepared by RM Beattie and JWL Puntis on behalf of the BSPGHAN,

August, 2004.

#### **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 in its role as supporter of 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 a 13 year period, 152 referrals had 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 were transplanted, although in recent years, this had 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) 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 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 are summarised below; a further discussion meeting was 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 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 $\geq 4$ weeks and $\geq 12$ 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 2: RECOMMENDED STAFFING FOR REGIONAL IF UNIT

Staff Required	Key Responsibilities/Job Role
<b>Paediatric Gastroenterologist</b> (Interest in or some experience of hepatology or local access to hepatology services)	<ul style="list-style-type: none"> <li>◆ Lead multi-disciplinary team - liaise with referring centre</li> <li>◆ Clinical assessment including diagnostic procedures e.g. endoscopy</li> <li>◆ Review of medical, pharmacological, nutritional and surgical strategies in conjunction with referring centres</li> <li>◆ Referral to hepatology service if required</li> </ul>
<b>Paediatric Pathologist</b>	<ul style="list-style-type: none"> <li>◆ To support/confirm diagnosis</li> </ul>
<b>Paediatric Surgeon and anaesthetist</b>	<ul style="list-style-type: none"> <li>◆ Primary management of patients with surgical disorders leading to intestinal failure</li> <li>◆ Lead and deliver central venous catheter insertion service</li> <li>◆ Surgical interventions to be utilized to promote intestinal rehabilitation</li> </ul>
<b>Paediatric Dietitian</b>	<ul style="list-style-type: none"> <li>◆ Dietetic review and manipulation of individual nutrients, depending on patient's absorption</li> <li>◆ Dedicated paediatric dietitian with experience of intestinal failure</li> <li>◆ Review/estimate of energy and hydration needs</li> <li>◆ Optimisation of parenteral/enteral nutrition</li> </ul>
<b>Support of specialised feed unit</b>	<ul style="list-style-type: none"> <li>◆ Accurate formulation of feeds and/or teaching of parents</li> </ul>
<b>Paediatric PN Pharmacist</b>	<ul style="list-style-type: none"> <li>◆ Pharmacist experienced in formulation, compounding and prescribing</li> <li>◆ Liaison with referring pharmacy centre</li> <li>◆ Liaison with home care company for HPN patients</li> </ul>
<b>Nutrition Nurse</b>	<ul style="list-style-type: none"> <li>◆ Liaison with nursing staff from referring hospital, and community support staff</li> <li>◆ Family education regarding disease process and methods to minimise complications related to central venous catheter</li> <li>◆ Training, support and liaison with referral centres regarding protocols and intravenous pumps used in home PN</li> <li>◆ Discussion regarding regional support groups</li> <li>◆ Training carers in home PN</li> </ul>
<b>Laboratory services: Clinical Chemistry, Microbiology, Haematology and Immunology</b>	<ul style="list-style-type: none"> <li>◆ Consultant Microbiologist for advice regarding antibiotics, regimens and infection control</li> <li>◆ Liaison between laboratory staff and NST</li> </ul>
<b>Social Work support</b>	<ul style="list-style-type: none"> <li>◆ Support families with social and financial stress</li> <li>◆ Liaison and review with referring centre</li> </ul>
<b>Psychologist</b>	<ul style="list-style-type: none"> <li>◆ Help to identify and manage psycho-social and chronic problems related to illness.</li> </ul>
<b>Play Therapist</b>	<ul style="list-style-type: none"> <li>◆ To support child and family with pre- and post-procedural work</li> </ul>
<b>Stoma Nurse Specialist</b>	<ul style="list-style-type: none"> <li>◆ Care of stoma problems</li> </ul>
<b>Specialist ward nursing staff with gastroenterology experience</b>	<ul style="list-style-type: none"> <li>◆ To provide expert nursing support for families</li> <li>◆ Liaison with referring centres to ensure continuity of high standard of care</li> <li>◆ Provide essential documentary support regarding monitoring of care and treatment</li> </ul>
<b>Paediatric Radiologist</b>	<ul style="list-style-type: none"> <li>◆ To provide advanced interventional radiological techniques</li> <li>◆ Expert review of radiological investigations supporting diagnostic process</li> </ul>
<b>Physiotherapist (speech and occupational therapists)</b>	<ul style="list-style-type: none"> <li>◆ Assessment of motor development skills, including feeding skills</li> <li>◆ Ongoing support/liaison in the community</li> <li>◆ Development and treatment plans instigated and monitored re ongoing development.</li> <li>◆ Liaison with referring centre.</li> </ul>

## **Appendix 3**

### A PATIENT'S JOURNEY

One of the more common congenital bowel anomalies leading to need for long term PN is gastroschisis (highlighted in the Chief Medical Officer's Report, 2004) (1) complicated by bowel infarction. The following description outlines what should happen to such an infant.

After ante-natal diagnosis (often at the time of the routine fetal anomaly scan), consultation with a paediatric surgeon is arranged so that the prognosis can be discussed and delivery planned in a neonatal unit with paediatric surgical facilities. Following delivery a full assessment is made, and when non-viable bowel is present, surgery is carried out and a temporary stoma formed. Once there is no more need for operative intervention and the surgeons consider that a full post-operative recovery has been made, the patient should be transferred to the care of a paediatric gastroenterologist for ongoing medical management of IF. In some instances, this may mean transfer to a different hospital.

PN is commenced within the first few days, following insertion of a CVC. Small volumes of milk are introduced as tolerated, but if there is less than 30 cm of small bowel remaining (usually in conjunction with loss of ileo-caecal valve and around two thirds of the colon), PN dependency can be anticipated for at least one year, if not permanently. Gastroenterologist, pharmacist and dietitian supervise nutritional intake, while the nutrition nurse specialist develops a training programme for family carers opting for HPN. Ideally, discharge home for HPN should occur from 2-4 months of age in clinically stable patients, and needs to be supervised by a NST. Intolerance of

enteral feed may merit further investigation, including upper and lower gastrointestinal endoscopy with mucosal biopsy.

Biochemical monitoring is performed regularly and abnormalities discussed with a clinical biochemist and pharmacist. Episodes of septicaemia associated with the CVC require antibiotic treatment and expert advice from a microbiologist. Limited tolerance of enteral feeding and episodes of infection are both risk factors for the development of liver disease. Concerns regarding abnormal liver function are discussed with a paediatric hepatologist. Prevention of CVC related infection relies principally on high standards of aseptic technique, maintained by staff training and experience. The CVC must sometimes be removed because of continuing infection or blockage; prompt replacement is undertaken by surgeon, anaesthetist, or interventional radiologist. Venous thrombosis may require detailed radiological investigation (including ultrasound, venography and MRI venography) in order to guide subsequent placement of CVC.

Speech and language, occupational and play therapists all help promote oral feeding skills and general development during prolonged hospitalisation. Psychology and social work colleagues support the family, while the nutrition nurse liaises with community staff preparing for the day when HPN becomes possible. Reversal of the stoma should be planned with the surgeon and carried out before discharge on HPN if appropriate. Once carers have been through a complete training process and demonstrated competency at each step, HPN can be arranged through one of the home care companies (providing housing conditions are adequate). Standards for the supply of nutrients and equipment have been elaborated by the British Association for Parenteral and Enteral Nutrition (BAPEN) (2). Following discharge, close contact is

maintained between nutrition nurse, family and other members of the multidisciplinary nutritional care team. Regular discussion of progress takes place, together with outpatient review.

In the event of fever or acute illness, the child is readmitted urgently to the local paediatric ward for investigation and management of suspected CVC sepsis according to agreed guidelines. Should tolerance of enteral feeding remain limited and monitoring indicate progressive liver disease, referral is made to a supra-regional paediatric hepatology unit for assessment, or to the intestinal failure unit at Birmingham Children's Hospital for consideration of small bowel transplantation.

If the child tolerates a slow build up of enteral feed without developing liver complications, weaning from PN can be anticipated. The dietitian and gastroenterologist manipulate enteral feed intake in order to maximise absorption and hasten weaning from PN. Enteral tube feeding is often required for months or years while oral feeding skills develop. Progress is closely monitored by members of the NST (see Appendix 2). Long term follow up to assess growth and screen for nutritional deficiencies is necessary, even when successful weaning from PN has taken place.

(1) Report of the Chief Medical Officer of Health, 2004.

(2) Current perspectives on paediatric parenteral nutrition. A report by a Working Party of the British Association for Parenteral and Enteral Nutrition, Maidenhead, 2000

(see also diagrammatic representation)

## **Appendix 4**

### **A SURVEY OF CHILDREN WITH INTESTINAL FAILURE IN WEST YORKSHIRE**

Köglmeier J, Martin H, Day C, Puntis JW

#### **Aims**

Lack of data regarding the epidemiology and long term outcome of intestinal failure (IF) in the UK preclude rational planning of tertiary referral and transplant services. Our aim was to identify all cases of IF in a geographical area representative of the UK population: 3.5% of 0-14 year olds live in West Yorkshire (n = 414 137), which also has 4.3% (n = 25,714) of the UK's annual births.

#### **Methods**

Only children from families living within West Yorkshire were included in the study. Hospital pharmacists were contacted by telephone and provided details of 96 children from West Yorkshire who over a two year period (2001/2002) had received parenteral nutrition (PN) for >28 d. 93/96 hospital records were successfully traced, and clinical details obtained including underlying diagnosis, type of surgery, complications, long-term problems and outcome at 1 and 2 years by review of the case notes.

#### **Results**

Patient categories included premature newborn (n = 61), surgical newborn (n = 20), oncology patients (n = 11) and liver disease (n = 1). Underlying diagnoses/indications for PN included gastrointestinal immaturity (n = 37), necrotising enterocolitis (n = 23), congenital bowel anomaly (n = 20); gastrointestinal complications of chemotherapy (n = 11), dysmotility (n = 1), severe post enteritis syndrome following liver transplant (n = 1). At 2 years follow up 85 children were fully enterally fed, had

normal liver function and were well. 6 (7%) died whilst on PN, 3 from gram negative sepsis, and 3 following withdrawal of care. Two patients remained on home PN. Three children were referred to a small bowel transplant centre.

### **Conclusion**

The overall outcome of intestinal failure is good. Almost two thirds of children with intestinal failure are premature newborn infants. If our figures are representative, around 1100 children in the UK require long term PN (>28 d) each year, with approximately 35 being candidates for transplant surgery.

## Appendix 5

### Patient Journey – Gastroschisis with Bowel Infarction

