

**A Guide for Purchasers of Paediatric Gastroenterology,
Hepatology and Nutrition Services**

**British Society for Paediatric Gastroenterology, Hepatology
and Nutrition**

CONTENTS

BACKGROUND	
The scope of paediatric gastroenterology & nutrition	3
Other documents	3
Parents support groups	4
THE PROVISION OF SPECIALISED SERVICES	
Optimising clinical outcomes	4
Specialised Centres	6
Supra-regional services in paediatric hepatology	7
Managed Clinical Network for Paediatric Gastroenterology	8
REFERRAL GUIDELINES FOR SPECIFIC FORMS OF GASTROINTESTINAL DISEASE	
Complex motility disorders (incl. gastro-oesophageal reflux)	8
Recurrent abdominal pain	9
Small intestinal disease	9
Intestinal failure	9
Post-gastroenteritis syndrome	10
Coeliac disease and other enteropathies	10
Inflammatory bowel disease	11
Acute gastrointestinal bleeding	12
Pancreatic disease	12
Neonatal gastroenterology	12
GUIDELINES FOR SPECIFIC FORMS OF LIVER DISEASE	
Neonatal liver disease	13
Chronic liver disease	13
Acute liver failure	14
Liver transplantation	14
Metabolic liver disease	14
NUTRITION SERVICES	
Nutritional Care Team	15
Home enteral and parenteral nutrition	15
Home enteral nutrition	15
Home parenteral nutrition	16
REFERENCES	17
APPENDIX ONE	
Home parenteral nutrition	20
APPENDIX TWO	
Specialised Services Definition Set	23

BACKGROUND

This document has been produced in response to a request from purchasers to provide guidelines by professionals for the provision of specialised services. It describes the ideal configuration of services, which should be available to children in all parts of the country.

Paediatric gastroenterology is a clinical speciality comprising the investigation and management of disorders of the gastrointestinal tract (the oesophagus, stomach, pancreas, small intestine and colon) in infants and children.

It also encompasses two related specialities: first, paediatric hepatology (liver diseases) and second, clinical nutrition; the latter because diseases of the gastrointestinal tract are potent causes of nutritional disorders, often requiring specialised nutritional care.

Paediatric gastroenterology has important interfaces with a number of related medical disciplines:

- general and neonatal paediatrics
- paediatric and adult surgery
- oncology
- inborn errors of metabolism
- immunology and infectious diseases
- gastroenterology and hepatology in adults

Other documents

Purchasers may find it helpful to make reference to the following documents:

- Children's National Service Framework
<http://www.doh.gov.uk/nsf/children.htm>
- National Specialized Services Definition Set
<http://www.doh.gov.uk/specialisedservicesdefinitions/23children.pdf>
- Directory of Acute Paediatric Services 1999
http://www.rcpch.ac.uk/research/Acute_Directory.html
- DGH GI, Hepatology and Nutrition Services: *Final Report – review April 2004*
<http://bspghan.org.uk/document/DGH%20SERVICES%20BSPGHAN.PDF>

Parent support groups

Advice has been taken from the following support groups when preparing this report:

- Crohn's in Childhood Research Appeal (CICRA)
<http://www.cicra.org/>
- National Association for Colitis and Crohn's Disease (NACC)
<http://www.nacc.org.uk/>
- Children's Liver Disease Foundation (CLDF)
<http://www.childliverdisease.org/>
- Patients on Intravenous and Nasogastric Nutrition Therapy (PINNT)
<http://www.pinnt.com/>
- Coeliac society
<http://www.coeliac.co.uk/>
- Cystic fibrosis trust
<http://www.cftrust.org.uk/site/>

THE PROVISION OF SPECIALISED SERVICES

Optimising clinical outcomes

The best clinical outcomes are achieved when the number of patients being treated in a unit is sufficient for a high level of medical and nursing expertise to be maintained. This has been clearly demonstrated, for example, for paediatric oncology and cystic fibrosis (1,2). The proposed model of 'hub' hospitals providing specialised services to a number of 'spoke' hospitals was first suggested as in the provision of adult cancer care (Calman-Hine Report, DoH April 1995). It is now increasingly accepted in paediatrics (3) and was suggested by the Bristol Royal Infirmary Enquiry to allow greater integration of healthcare services for children (4).

The NHS Management Executive, in its guidelines on contracting for specialist services, emphasises that “Sensible contracting needs to take into account the optimum population size not only for the stability of contracted referrals but also to give sufficient “critical mass” for clinical effectiveness”.

Within the United Kingdom, the management of children with paediatric gastrointestinal disorders remains patchy without organised clinical networks and pathways for care that are patient focussed. Fundamental to such models are the paediatric gastroenterologist, general paediatrician and general practitioner supported by other medical and allied professionals.

Significant inroads have been made over recent years to this with specialist centres, outreach clinics and within district hospitals the recognition of the fundamental role of the paediatrician with an interest in paediatric gastroenterology. It is essential that the latter has access to the facilities and support services available in the tertiary centre and is part of a unified service for children which is appropriate for the area they serve. It does however remain the case that many paediatricians are unfamiliar with the presentation and management of liver disorders in childhood for example. This may lead to delay in diagnosis, and in starting treatment, which in turn may allow progression of disease to an irreversible stage (cirrhosis), or development of acute liver failure.

Other examples from the literature include

- jejunal biopsy failure and inappropriate use of gluten-free diets in children suspected of having coeliac disease (5).
- patchy quality of care of children with inflammatory bowel disease (6,7).
- improved outcomes in children receiving parenteral nutrition when cared for in a specialist centre with a multidisciplinary nutritional care team (8-10).
- Adverse outcomes are associated with delay or failure, to refer children with liver disease:
 - increased morbidity and necessity for urgent liver transplantation in biliary atresia (11-13).
 - irreversible brain damage following delay in initiating treatment in children with inborn errors of metabolism
 - increased morbidity and mortality from delayed referral of infants and children with a choledochal cyst (14).
- failure to recognise or refer children with acute liver failure may lead to irreversible brain damage and death, or insufficient time to acquire a donor for liver transplantation (15).

Specialised Centres

Specialist services for paediatric gastroenterology, hepatology and nutritional care must be provided in specialist units.

The requirements for a specialist unit are:

- a population and referral base of sufficient size (approx 2 million) to justify the appointment of at least 3 gastroenterologists to provide on-call cover. Exceptions should be made for remote areas
- 3 trained paediatric gastroenterologists who fulfil the RCPCH/BSPGHAN higher professional training criteria and who have JCHMT accreditation with sub-specialist recognition on CCST.
- An endoscopy service, provided in a child-friendly setting, with at least 75 procedures per consultant endoscopist per annum
- Provision of anaesthetic support for paediatric endoscopy must be available. This does not preclude the use of sedation in appropriately selected cases.
- Specialist nursing staff, with particular training in nutritional care and paediatric intensive care / recovery (for management of children having procedures under sedation / GA)
- Paediatric gastroenterology clinical nurse specialist
- Easy access to the full range of specialist services for children on the same site, including specialist paediatric / adult surgery and intensive care. Similarly, specialist centres undertaking complex neonatal and paediatric gastrointestinal surgery should have access to a full range of gastroenterological and nutritional support services.
- a full range of diagnostic services, including:

Radiologists with specific training in paediatrics and expertise in paediatric diagnostic procedures

A full range of diagnostic and therapeutic endoscopy

Histopathologist with expertise in paediatric gastroenterological pathology

oesophageal pH monitoring

pancreatic function testing*

small intestinal disaccharidase assays*

Breath testing
Motility studies
Liver Biopsy and ERCP*

**some centres only*

- paediatric dietetic service, staffed by dieticians who spend the majority of their time working with children and available to input to the wards on a daily basis and to work alongside the clinicians supporting gastroenterology clinics and gastroenterology and nutrition ward rounds
- paediatric pharmacist
- psychological support from within child psychology / psychiatric services
- parent's accommodation and support services catering for the special needs of children including social workers, play therapists and teachers
- a multi-disciplinary nutritional care team which will include a paediatric speech and language therapist for assessment of children with feeding difficulties
- close links with adult gastroenterologists and an established mechanism for handing on adolescent patients to gastroenterological services for adults
- a commitment to undertake outreach clinics and shared care with general paediatrics

Supra regional services in paediatric hepatology

There are supra-regional units at King's College Hospital, London and The Children's Hospital, Birmingham and St James's University Hospital, Leeds. Refer to guidelines for services provided by paediatric hepatology (NSCAG funded April 1999, Children's NSF document at <http://www.doh.gov.uk/nsf/children.htm>)

Managed Clinical Network for paediatric gastroenterology

Each specialist centre should aim to provide the above services to referral centres. Centralised expertise should be easily accessible .

The following would be essential for this system to function:

1. Access to expert opinion by telephone 24 hours/day.
2. Ability for rapid outpatient referral to defined 'urgent / emergency referral clinic'
3. Capacity to accept in-patient transfers at short notice.
4. Capacity to admit children directly for specialist investigations without prior clinical assessment
5. Regular joint outreach clinics at referring hospitals to assess new patients and review shared care patients as necessary
6. Designated paediatrician at shared care hospital with adequate support services to provide shared care and act as referring consultant to specialist centre
7. Rapid communication of out-patient and in-patient management plans to designated referring consultant
8. Regular meetings teams within managed clinical network to review guidelines / communication / training needs

REFERRAL GUIDELINES FOR SPECIFIC FORMS OF GASTROINTESTINAL DISEASE

Complex motility disorders (including the severe end of constipation and gastro-oesophageal reflux)

Constipation is a very common disorder in childhood, with only a few affected children having an indication for referral to a specialist centre:

- severe, intractable constipation
- major psychological disturbance
- a suspicion of Hirschsprung's disease or intestinal pseudo-obstruction
 - delayed passage of meconium
 - onset in the newborn period, or first few months of life
 - minimal soiling
 - marked abdominal distension
 - episodes of severe diarrhoea (enterocolitis)
 - failure to thrive
 - persistent vomiting

The interpretation of rectal biopsies performed for suspected Hirschsprung's disease requires specialised histopathological techniques and should not normally be performed outside specialist centres. Although a large proportion of children with constipation fall within the remit of primary and secondary care, children with severe and treatment resistant constipation require referral to a specialist centre.

GOR is a common cause of regurgitation and vomiting in the first 12 months of life. It usually resolves spontaneously and can often be managed, without specialised investigation, by simple measures such as feed thickening and appropriate positioning.

Indications for referral to a specialised centre are:

- i) diagnostic doubt
- ii) failure to resolve on simple treatments
- iii) blood in vomit, or iron deficiency anaemia (suggesting oesophagitis)
- iv) dysphagia
- v) persistent pulmonary symptoms (? aspiration) / recurrent apnoea
- vi) failure to thrive and/or food refusal

Recurrent abdominal pain

Ten per cent of school-age children experience recurrent abdominal pain. Few are associated with organic pathology. Some are related to abdominal migraine (paroxysmal pain, pallor and a family history of migraine) or disorders of gastrointestinal motility. Indications for referral are:

- i) diagnostic uncertainty
- ii) night-time pain
- iii) close family history of peptic ulceration
- iv) evidence of gastrointestinal bleeding
- v) pain in association with weight loss, growth failure, anorexia, mouth ulcers, diarrhoea or jaundice
- vi) poor school attendance

Small intestinal disease

Intestinal failure

Severe small intestinal disease results in an inability to maintain normal nutritional status without parenteral nutrition (intestinal failure). At present about 200-250 children in the UK require parenteral nutrition for longer than 6 weeks each year.

Intestinal failure usually results from major congenital malformations of the gut, massive small intestinal resection, inherited abnormalities of the intestinal mucosa or a severe disturbance of gut motility (intestinal pseudo-obstruction). Long-term survival with good quality of life is possible with parenteral nutrition (intravenous feeding), often at home. In a proportion of affected children, complications of parenteral nutrition arise, particularly severe liver disease. These complications are less common in children cared for by a multi-disciplinary nutritional care team, including skilled paediatric dietetic input. Close links with paediatric surgeons experienced in the management of this complex group of patients is crucial.

Small bowel transplantation (16) combined with liver transplantation (17) when necessary, is now available in one centre (Birmingham) for patients with intestinal failure complicated by end-stage liver disease. The need for small bowel (with or without liver) transplantation is calculated to be 10 children per annum (extrapolation from West Midlands data). Constraints include late referral, donor availability and uncertain long-term outcome.

Post gastroenteritis syndrome / protracted diarrhoea

Following acute infectious diarrhoea, a few infants and young children develop chronic diarrhoea and malnutrition. The exact frequency is unknown. When brief and self-limiting, this will usually be managed by a general paediatrician, using a soya or casein-hydrolysate formula. If the diarrhoea is resistant to the use of commercially available infant formulas, expert dietetic advice from an experienced paediatric dietitian, and occasionally parenteral nutrition, are central to successful management.

Referral to a tertiary referral centre is therefore necessary for all patients whose diarrhoea continues for more than three weeks and who are continuing to lose weight.

Coeliac disease and other enteropathies

Guidelines for the diagnosis of coeliac disease have been published by the European Society of Paediatric Gastroenterology and Nutrition (ESPGAN) (18). Despite the availability of screening tests, small intestinal biopsy remains essential to the diagnosis. Diagnosis is difficult, even in those major paediatric centres, which do not have specialised paediatric gastroenterology services: biopsy failures and the inappropriate use of gluten-free diets remain common (5).

Increasingly in tertiary referral centres, biopsies are being obtained endoscopically. This method is quicker, more reliable, avoids irradiation, there

is no possibility of biopsy capsule failure, and multiple biopsies can be obtained. In order to maintain endoscopic skills, there should be an endoscopic activity of at least 75 procedures (oesophago-gastroduodenoscopy or colonoscopy) per annum per consultant.

An experienced histopathologist is also needed to interpret the biopsy. Not all children with an abnormal intestinal biopsy will have coeliac disease. Other dietary protein intolerances, auto-immune enteropathy and microvillus inclusion disease, for example, may all produce abnormalities which can be confused with coeliac disease, and lead to inappropriate treatment. A paediatric dietitian is therefore required to institute and supervise a gluten-free diet for these young patients.

When all these services are unavailable in a district general hospital efficient biopsy, histopathology, paediatric dietetics, either singly or in concert, referral to a tertiary referral centre is advisable.

Inflammatory bowel disease (IBD)

In the UK, nearly all IBD is caused by Crohn's disease or ulcerative colitis. Crohn's disease in children has an incidence in children under age 16 of about 3 per 100,000 population in the United Kingdom (6). Delay in diagnosis of this disorder in childhood is recognized as a major problem (7, 19). There are many reports suggesting an increasing prevalence of Crohn's disease in Europe (20,21).

Approximately 1/3 of childhood IBD is ulcerative colitis. Misdiagnosis is also a problem. The main error had been to commence treatment without a tissue diagnosis of chronic inflammatory bowel disease. Expert surgical services are essential as many children come to surgery (22). Timing is particularly crucial as delay in surgery beyond puberty may lead to permanent stunting of growth (23,24).

Initial investigations should include upper endoscopy, colonoscopy with ileoscopy. This may be performed under general anaesthesia to minimise stress for the child and ensure a complete study. Referral to a centre where this is possible is preferable to an incomplete investigation under sub-optimal conditions, as such investigations often lack diagnostic certainty and need to be repeated.

All children with suspected IBD should therefore be referred to a specialist unit. The advent of new therapies and changing treatment algorithms make centralised care even more important for children with IBD. In order to be able to provide a high quality of care in every region, close collaboration with easy access to the diagnostic services at the 'hub' is mandatory.

Experience of the newer agents is likely to be limited to larger centres, with many being available only in the setting of multi-centre trials.

Acute gastrointestinal bleeding

Acute gastrointestinal bleeding in childhood liver disease may be secondary to a variety of disorders, including oesophageal, gastric or rectal varices, portal gastropathy or peptic ulcer disease. Centres receiving children for management of GI bleeding should have facilities for the necessary medical support. Urgent endoscopy within 24 hours of admission is often necessary. The ability to perform sclerotherapy and/or surgical techniques such as shunt surgery, gastric and oesophageal transection, etc should be available on site. Units unable to provide these facilities should have arrangements for immediate transfer to a specialist centre e.g. children with variceal bleeding should be stabilised and referred to a supra-regional liver unit for further assessment and treatment.

Pancreatic disease

As pancreatitis and pancreatic exocrine insufficiency (other than in cystic fibrosis) are both very rare, referral to specialised centres is essential. Expertise in this area is likely to be limited to only a few centres nationwide. These centres should be able to provide full pancreatic diagnostic and therapeutic endoscopic services, mostly in conjunction with an adult gastroenterological service.

Neonatal gastroenterology

Neonatal surgery. The nutritional management of neonates requiring surgery is crucial to optimise recovery. In addition to maximising nutritional status, gastrointestinal complications may occur that require placement of feeding tubes and/or neonatal endoscopy. The involvement of paediatric gastroenterologists in the peri-operative management also provides continuity of care for children with conditions such as NEC and gastroschisis. These children may require long-term sub-specialist support and benefit from early and close liaison between paediatric surgeons, neonatologists and gastroenterologists.

Neonatal medicine. Neonates frequently present with gastrointestinal problems. Access to a paediatric gastroenterologist is ideal for the diagnosis and management of disorders such as gastro-oesophageal reflux and allergic/congenital enteropathies. Neonatal upper endoscopy may be required for the diagnosis of ulceration or an enteropathy, while the placement of percutaneous feeding tubes may be useful in some larger infants.

GUIDELINES FOR SPECIFIC FORMS OF LIVER DISEASE

Neonatal liver disease

The accurate diagnosis of children presenting with neonatal liver disease (jaundice lasting for more than 2 weeks within eight weeks of birth) is essential. Extra-hepatic biliary atresia for example, often presents in this way, and early diagnosis has a major impact on outcome (25). Units providing this service must have:

- diagnostic facilities to differentiate between extra hepatic-biliary atresia, neonatal hepatitis, inborn errors of metabolism. These facilities would include radiology, nuclear medicine, microbiology, diagnostic biochemistry, specialised histopathology.
- a surgeon skilled in performing the Kasai portoenterostomy with good results (achieving bile drainage in more than 50% cases). Published evidence suggests that results are better in centres treating more than 5 cases a year and with facilities for comprehensive long term care of the patient.
- a dietetic department able to provide nutritional management of infants with liver disease.
- multidisciplinary team to provide support in education and counselling, nutrition and metabolic diets, etc.
- such neonates should be seen as an emergency and the above facilities should be available within one week of referral to exclude biliary atresia.

Referral to a specialist paediatric supra-regional liver unit is recommended:

- for biliary atresia. In England and Wales this is a Dept of Health requirement (DoH 199/0268 30.4.99)
- for patients with liver failure.

Chronic liver disease

Children with chronic liver disease will require accurate and prompt diagnosis and early evaluation for transplantation. Examples of treatable disorders which may progress without early diagnosis and appropriate treatment are autoimmune hepatitis and Wilson's disease.

These children will require assessment at a centre which is in a position to provide:

- biochemical, immunological, microbiological or histological diagnosis.
- the necessary diagnostic procedures (endoscopy, liver biopsy, etc)
- the necessary nutritional, educational and counselling support.

Such children should be seen within four weeks of referral and the above facilities should be available on site to enable a diagnosis to be made within two weeks.

Children with decompensated liver disease should be referred to a specialist centre for management.

Acute liver failure

All children with acute liver failure should be referred to a supra regional/specialist centre capable of providing liver transplantation. These units should be able to establish the diagnosis and manage the acute liver failure in an appropriate setting (outlined above). Facilities for transplantation should be available on site.

Liver transplantation

Patients should only be assessed for paediatric liver transplantation in a centre which has all the facilities of a supra regional/specialised centre and a skilled multidisciplinary team. Outcomes from the three UK centres are closely audited.

Metabolic liver disease

Inborn errors of metabolism may affect many organs and systems. In some, liver involvement is the most significant problem, whilst in others there may be multi-system disease. The quality of the available laboratory expertise is of paramount importance, and centres offering a metabolic service must demonstrate, in addition to the above hepatological competencies,

- (a) a metabolic laboratory, with a senior paediatric biochemist, participation in a quality control system, and a sufficiently large catchment area to maintain experience
- (b) skilled paediatric dietitians
- (c) related paediatric specialties including neurology, nephrology and genetics.

NUTRITION SERVICES

Children with diseases of the gastrointestinal tract and liver frequently become malnourished, and an essential part of their medical care involves nutritional assessment and support. Nutrition is therefore an integral part of specialist paediatric gastroenterology and hepatology services.

In addition to children with gastrointestinal and hepatic disease who require nutritional support, many children, particularly in disadvantaged parts of the UK, have unrecognised, but primary nutritional problems (e.g. failure to thrive, iron deficiency).

Nutritional care team

Nutrition services should be provided by a nutritional care team, comprising a senior clinician (consultant paediatrician trained in clinical nutrition), nutrition nurse specialist, paediatric dietitian and pharmacist, who together with surgical and biochemical, back-up, co-ordinate the delivery of enteral and parenteral nutrition support (27-30).

Home enteral and parenteral nutrition

Increasing numbers of children on long term nutritional support are being cared for in the community. A recent British Paediatric Surveillance Unit study has suggested that this aspect of nutritional support is under-utilised and it is anticipated that the needs for home nutritional care will increase. This therapy requires intensive supervision and support and should not be undertaken in units without a nutritional care team. It has been further suggested by the British Association of Parenteral and Enteral Nutrition that because of the small numbers involved, each region should have one unit responsible for home parenteral nutrition, which will provide the lead in management of the parenteral nutrition and be responsible for the execution of the discharge plan.

Home Enteral Nutrition

This form of treatment consists of the delivery of liquid feeds into the stomach (through a nasogastric tube or a gastrostomy) or into the intestine (via a jejunostomy). In many cases the feed will be delivered continuously by a feeding pump, often overnight. The treatment may be used for a variety of conditions and may be entirely supportive, as in the management of children with malignant disorders, severe neurological disease or as primary therapy, as

in cystic fibrosis and Crohn's disease. Such children may require long-term feeding via gastrostomy or jejunostomy.

The treatment requires a shared care approach between general practitioners who prescribe the feeds, community paediatric services that provide the disposables, and the hospital and specialist paediatric services that provide the lead management.

Home Parenteral Nutrition

Parenteral nutrition is the administration of an individual's nutrition directly into the bloodstream. It is the only method of providing nutrition to children with intestinal failure and allows them to survive, grow and develop.

Parenteral nutrition is administered through a central venous catheter into the right atrium. The nutrient solutions are infused over 12 to 16 hours through the evening and night. Strict aseptic technique is essential whenever the catheter is handled.

Home parenteral nutrition has evolved to enable families who might otherwise spend long periods of time in hospital with associated detrimental effects on child development and family life, to return to a normal environment. This form of treatment requires highly motivated caregivers, who can administer parenteral nutrition safely while being constantly vigilant for the potentially serious complications that can occur. Despite the constraints associated with parenteral nutrition and in some cases the underlying disease, a good quality of life is possible for the child and family. However, this is only achievable with appropriate management of the disease process, psychosocial support and a well organised system for the provision of nutrient solutions and ancillary products.

As home parenteral nutrition is a difficult and expensive treatment, specific criteria and guidelines for its provision are provided (Appendix).

Treatment of children requiring home parenteral nutrition is complex and requires considerable expertise as well as financial and personnel resources. As a result, a degree of rationalisation to concentrate this service into designated regional centres is preferable.

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APPENDIX ONE

HOME PARENTERAL NUTRITION

Clinical Services

In order to provide good quality care for the child, clinical care should be shared with the family practitioner, local hospital and the specialist centre in the following manner:

1. The family practitioner should continue to provide appropriate primary health care for the child in partnership with the referring hospital and specialist centre.
2. The referring hospital and specialist centre should share paediatric care, with the referring hospital available if necessary for emergency treatment.
3. The specialist centre will provide the lead in the management of the parenteral nutrition and be responsible for the execution of the discharge plan.

Discharge planning

Pre-discharge

1. Liaison with local health care professionals to identify funding arrangements.
2. Home visit by nutrition nurse specialist to assess suitability of home for parenteral nutrition
- ~~5.3.~~ Training for local health care professionals, schools and nurses as required.
- ~~6.4.~~ Formulation of home nutrient regimen.
- ~~7.5.~~ Co-ordination of delivery of nutrients and ancillary products to family home. Identification of equipment requirements. Liaison with designated home care provider
- ~~11.6.~~ Training programme and preparation of child and family for home parenteral nutrition. Teaching programme for 2 caregivers will be tailored to individual needs. Prior to discharge caregiver will usually be required to be resident in hospital, carrying out independent care. Written guidelines will be provided.
- ~~12.7.~~ Professionals meeting to inform GP, community and paediatrician community nurses.

Post-discharge

1. Provision of 24 hour on-call service available to local health care professionals and family.
2. The patient should have an outpatient consultation in the specialist home parenteral nutrition clinic at least 3 monthly. At this clinic, nutrition nurse specialist, pharmacist, dietitian and social worker will be available. There will be the opportunity to socialise with other families with children on home parenteral nutrition. More frequent appointments may be necessary depending on the child's medical progress.

~~5.3.~~ Nutritional monitoring

~~6.4.~~ A progress report should be sent to the patient's GP within 7 working days of the clinic visit.

Supply of intravenous nutrients and equipment

The family should know how the deliveries are to be made, when and by whom. Written information about the system should be given to the family.

There should be clear lines of communication between the prescriber, the supplier and the patient. The supplier contact person should understand the needs of home parenteral nutrition patients and how the items being supplied are used. The family should know the name of the contact person at the supplier and be informed when staff changes occur.

The supplier should provide the nutrient solutions and equipment as specified by the nutrition team. Substitutions should not be made without reference to a member of the team. The supplier should respond within an agreed period to the formulation of nutrient regimes and changes in formulation made by the prescriber.

Deliveries should arrive on the designated day. Anyone visiting the home should be smart in appearance, friendly and have knowledge of the products being delivered. Identification badges should be worn.

Nutrient solutions should be placed in the patient's fridge and all packing removed by the deliverer. Equipment should be put in a safe place and care taken to avoid damage to sterile products.

The supplier should maintain safe stock levels. Out of stock items should be delivered as quickly as possible within five working days.

The supplier should avoid additional deliveries other than those detailed in the patients delivery calendar. Ideally fluids should be delivered for two weeks supply with additional bags for safety stock. To avoid the family needing to store large quantities of ancillary goods equipment should not be delivered more often than three to four weekly.

There should be a safe method of ensuring that deliveries can be made if no-one is at home. The service should be flexible enough to enable the patient to take holidays and travel away from home. There should be arrangements for the disposal of clinical waste. The family should be notified of any changes in the service. The service should be open to evaluation from the family and the nutrition team.

APPENDIX TWO

SPECIALISED SERVICES DEFINITION SET

<http://www.doh.gov.uk/specialisedservicesdefinitions/23children.pdf>

Gastroenterology including Hepatology (NSCAG) and Nutritional Support

Paediatric gastroenterology encompasses not only disorders of the bowel but also hepatology and complex nutritional support. Whilst a proportion of paediatric gastroenterology and nutrition belongs within primary and secondary care there are elements which by virtue of their rarity in general paediatric practice, the severity of the condition (e.g. severe constipation or gastro-oesophageal reflux), the specific difficulties encountered in very young children or the complexity of their management (e.g. intestinal failure) means that they must be considered by tertiary or specialist services. In addition there is an increasing array of specialised diagnostic tools that cannot be appropriately provided outside the specialist arena. When a child is referred to a paediatric gastroenterologist, the activity is regarded as specialised.

There is a national service for paediatric hepatology funded by the National Specialist Commissioning Advisory Group (NSCAG) and provided in 3 national units. This service covers liver failure, paediatric liver transplantation, surgery for biliary atresia and paediatric intestinal transplantation. There are established shared care arrangements and referral pathways between the three national units and specialist gastroenterology units. A list of conditions that should be referred to the national service developed by NSCAG and the British Society for Paediatric Gastroenterology, Hepatology and Nutrition, is available at

<http://www.doh.gov.uk/specialisedservicesdefinitions/23children.pdf> as an appendix of the main document.

The aspects of paediatric gastroenterology that should be regarded as specialised are:

Gastroenterology

- Intestinal failure
- Inflammatory bowel disease
- Gastrointestinal bleeding
- Complex motility disorders including the severe end of constipation and gastro-oesophageal reflux
- Protracted diarrhoea (greater than 3 weeks duration and weight loss)
- Congenital transport disorders
- Multiple food intolerances
- Coeliac disease (unless pathology and paediatric dietetic service available locally)
- Pancreatitis
- Pancreatic exocrine insufficiency other than cystic fibrosis

Hepatology

- Neonatal hepatitis syndromes
- Childhood chronic liver disease
- Surgical liver disease
- Jaundice not due to uncomplicated hepatitis A infection

Nutrition

- Home parenteral nutrition
- Nutrition for complex disorders whether supportive or primary treatment
- Gastrostomy and jejunostomy tube feeding

Specialised investigations

- Upper and lower GI endoscopy
- Liver biopsy
- Intestinal biopsy
- Rectal biopsy
- Oesophageal pH monitoring
- Breath hydrogen tests
- Pancreatic function tests
- Motility studies and manometry