

E03/S(HSS)/b

**NHS STANDARD CONTRACT
FOR INTESTINAL PSEUDO-OBSTRUCTIVE DISORDERS SERVICE (CHILDREN)**

PARTICULARS, SCHEDULE 2 – THE SERVICES, A – SERVICE SPECIFICATION

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| Service Specification No. | E03/S(HSS)/b |
| Service | Intestinal pseudo-obstructive disorders service (Children) |
| Commissioner Lead | |
| Provider Lead | |
| Period | 12 months |
| Date of Review | |

1. Population Needs

1.1 National/local context and evidence base

Chronic intestinal pseudo-obstruction syndrome (CIP) is an intestinal motility disorder in which impaired intestinal motor activity causes recurrent symptoms of intestinal obstruction in the absence of mechanical occlusion. CIP comprises a heterogeneous group of uncommon congenital or acquired, localised or generalised, primary or secondary disorders affecting the enteric neuromusculature. The nerves and muscles of the intestine grind, mix and move intestinal contents along the bowel. This normal motility is disturbed in a variety of uncommon congenital and acquired disorders of the gastrointestinal neuromusculature.

These disorders, which can affect 1/75,000 children, typically manifest as chronic or sub-acute obstructive disorders of the gastrointestinal tract which can mimic surgically amenable conditions. It is not unusual therefore for infants to have had 3 - 5 laparotomies/operative procedures before referral to a tertiary centre for definitive diagnostic assessment (Faure C. et al Journal Pediatric Gastroenterology and Nutrition 1997; 24(4): 459). Inappropriate surgical intervention is also common (Heneyke S. et al Archives of Disease Childhood 1999; 81(1): 21-27). The implications of inappropriate operative interventions upon long-term morbidity may also be considerable, potentially hastening progression to permanent intestinal failure.

Evidence base

There is broad international consensus on how these children should be investigated

The broad approach to *initial* investigation has changed little in the last 10 years (Di Lorenzo C. Gastroenterology 1999; 116(4): 980-978) although the diagnostic procedures have been refined considerably with technological advances. It is evident that 3 factors are *essential* to a pseudo-obstructive diagnostic service:

- a multidisciplinary team (MDT) with core members comprising:
 - gastroenterologists
 - general and urological surgeons
 - radiologists
 - histopathologists and,
 - the ready availability of other members with expertise in metabolic, infectious, rheumatological, neuromuscular, neurological and autonomic disorders as well as nutrition support teams
- access to a wide range of manometric methods including:
 - antroduodenal
 - colonic and anorectal manometry
- specialised histopathological services with experience of enteric neuromuscular pathologies (see consensus guideline Knowles CH et al Acta Neuropathol 2009; 118(2): 271-301)

There is consensus that such assessments should take place early and that surgical interventions should be minimised to those for definitive diagnosis and therapy

2. Scope

2.1 Aims and objectives of service

To provide a service for the diagnosis and management of paediatric pseudo-obstructive disorders that will:

- streamline admission for definitive investigations, speeding up diagnosis and as a result reducing morbidity with a target time from referral to admission of less than four weeks
- initiate the first step in definitive management
- facilitate the generation of evidence based guidelines for the management of this diverse group of children by establishing a defined UK cohort of children

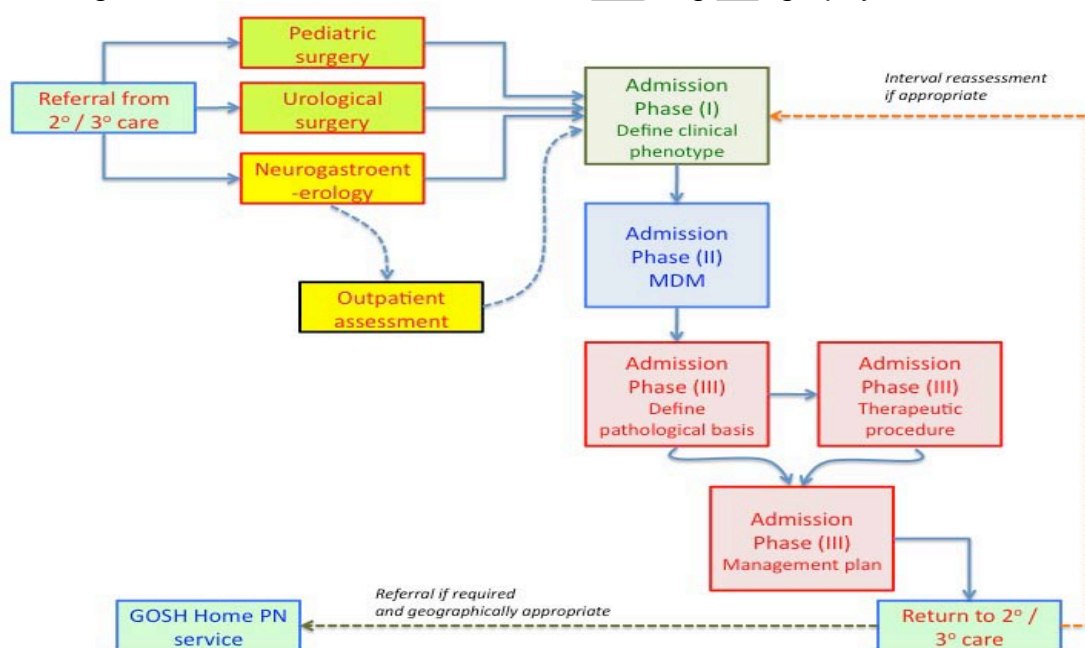
2.2 Service description/care pathway

The service has three elements/phases (figure 1). Patients will enter the service either through direct external referral to the neurogastroenterology department or by internal referral. Referrals typically come from general surgeons or urological surgeons. External referrals may require a fast-track outpatient assessment prior to inpatient assessment. The three essential elements of the service will include:

- definition of clinical phenotype (1-2 weeks)
- Multi-disciplinary meeting
- definition of the pathological basis of CIP in the 70% of patients in whom this is appropriate (1-3 weeks)

To facilitate adherence to presentation and age specific investigative algorithms a minimum armamentarium of gastrointestinal investigative methodologies are necessary:

- manometric methods and expertise in interpretation of foregut and hind gut motility patterns including esophageal high resolution manometry, antroduodenal and ileal manometry, colonic manometry and anorectal manometry with age appropriate manometric catheters and endoscopic and interventional radiographic skills for appropriate catheter placement in infants
- histopathological expertise in the qualitative histopathological assessment of laparoscopic seromuscular biopsies and qualitative and quantitative assessment of intestinal sleeve biopsies using both standard and specialised stains and electron microscopy Clinical Pathology Accreditation - accredited laboratory
- non invasive assessment of gastrointestinal motility including isotopic or scintigraphic gastric emptying of solids and liquids, isotopic determination of regional intestinal transit and surface electrogastrigraphy



Following diagnosis a definitive treatment plan is necessary involving a multidisciplinary discussion between clinicians with expertise in gastrointestinal (GI) motility, paediatric surgeons, dieticians and the nutrition support team. Decisions are made about the need for GI stomas, the need for parenteral nutritional support and the need for disease specific therapies including non-GI surgical interventions.

If there is no local home parenteral nutrition (PN) service then the home PN service could review the patient during their inpatient stay with a view, in due course, to

including the child in this programme when clinically appropriate and if requested by local services. The ultimate decision to refer the child to this service will be between the local health services and the PN team. Provision of on-going PN is beyond the scope of this service

Costs of PN and home PN provision are outside the scope of this service. Outreach clinics may be held occasionally with local providers as required by participants for combined outpatient review and care planning.

2.3 Population covered

NHS England commissions the service for the population of England. NHS England contract includes provision for the service to treat eligible overseas patients under S2 [Under European Union (EU) regulations, patients can be referred for state funded treatment to another European Economic Area (EEA) member state or Switzerland, under the form S2 (for EU member states) or the form E112 (for Iceland, Norway, Liechtenstein and Switzerland)] referral arrangements. Providers are reimbursed for appropriately referred and recorded activity as part of NHS England contract.

Trusts performing procedures on EU-based patients outside of S2 arrangements will need to continue to make the financial arrangements directly with the governments involved, separately from their contract with NHS England.

With regard to S2, the mechanism for recovery of costs has been via the Department for Work and Pensions Overseas Healthcare Team. They are responsible for agreeing reconciliation and recovery of costs with European administrations. These arrangements were implemented in October 2009, though a similar process existed previously. The financial flows are therefore back into the treasury rather than back to trusts.

2.4 Any acceptance and exclusion criteria

The service will consider new referrals of all ages up to their fifth birthday regardless of race, gender or religion. Patients may be admitted and/or managed within the scope of the service specification beyond their fifth birthday providing the referral was received prior to the child's fifth birthday and that the time between referral and admission was in keeping with the usual operation of the service.

Referral criteria, sources and routes

Infants and young children with apparent intestinal obstruction but no anatomical evidence of physical obstruction will commonly have been referred to a paediatric surgeon and undergone a negative laparotomy or will have been investigated by a surgeon and no evidence of a physical cause found. Most cases will therefore come from tertiary paediatric surgery or gastroenterology services. Referral from other routes is extremely unlikely unless the children have been investigated in another country and already have a tentative diagnosis of pseudo-obstruction.

Exclusion criteria

Children with evidence of physical obstruction might be excluded, although children with pseudo-obstruction can also develop physical obstruction secondary to adhesions following previous surgery (they as a group have an increased prevalence of this complication).

Disorders of the extrinsic enteric nerves are excluded as are acute self-limiting forms of pseudo-obstruction.

2.5 Interdependencies with other services

The service will function as a supra-regional referral service. It will be a requirement that referrals arise from secondary or tertiary care and post diagnosis patients will return to the care of these services with continued advice as necessary at the request of these services. It is envisaged that on occasion the service will ask the secondary / tertiary care provider to re-refer for reinvestigation at a specified time-point if there is no unified diagnosis following initial referral and investigation. Such a situation will be uncommon.

3. Applicable Service Standards

3.1 Applicable national standards e.g. NICE, Royal College

The providers of the service must ensure they are fully integrated into their trust's corporate and clinical governance arrangements and must comply fully with clinical negligence scheme for trusts (CNST) and Care Quality Commission (CQC) requirements in terms of quality and governance. The hub centres are responsible for overseeing the governance arrangement of any spoke clinic provided under sub-contractual arrangements.

Each centre will ensure that:

- regular meetings take place with patient representatives
- all practitioners participate in continuous professional development and networking
- patient outcome data is recorded and audited across the service

4. Key Service Outcomes

| <i>Quality Performance Indicator</i> | <i>Threshold</i> | <i>Method of measurement</i> | <i>Report Due</i> |
|---|------------------|------------------------------|-------------------|
| Mortality | | | |
| Unplanned admissions | | | |
| Improving service user & carer experience | | | |
| Clinical outcomes | | | |
| (i) Physiological diagnosis | | | |
| (ii) Histopathological diagnosis (if applicable) | | | |
| (iii) Definitive primary treatment given (irrespective of response) | | | |

5. Location of Provider Premises

Great Ormond Street Hospital for Children NHS Foundation Trust (GOSH)