

Intestinal Pseudo-obstruction in adults and children
Preliminary Bid for NSCAG Support

A Proposal to BSPGHAN

Background

Chronic Intestinal Pseudo-obstruction encompasses a variety of rare chronic intestinal dysmotilities distinguished from the very common problems of irritable bowel syndrome and functional dyspepsia by characteristic histopathology, intestinal motility on physiological testing and their profound effect on the patient's ability to maintain a normal state of nutrition. They may be primary, or secondary to other systemic illnesses, and may be linked to the more severe disorders of slow colonic transit. Patients suffer from a variety of intensely debilitating symptoms including dysphagia and reflux, high volume vomiting, intestinal distension and bacterial overgrowth, opiate-requiring chronic abdominal pain and severe constipation which is often poorly responsive even to total colectomy, when severe ileostomy dysfunction can be anticipated.

The overall prevalence of chronic intestinal pseudo-obstruction is unknown. A national survey by members of the North American Society of Pediatric Gastroenterology and Nutrition (1) identified a total of 87 cases as fitting the criteria for the syndrome. Five families, three with autosomal dominant inheritance, accounted for 15 cases. The remainder were single cases in families, indicating either autosomal recessive inheritance, spontaneous mutation, or acquired disease. Of the 87 patients, 47 were male; 19 patients were symptomatic at birth and 37 (43%) within the first month of life; 64% were diagnosed by the first year of age and the remainder were diagnosed by 18 years of age. Of those not lost to follow up 31% died, often in the first six months of life.

The prevalence among adults is low also but is unclear. While only a proportion of children with the disease survive into adulthood, patients can present late with primary neuropathies or myopathies, or pseudo-obstruction secondary to other diseases such as diabetes, scleroderma and other autoimmune diseases.

Patients with pseudo-obstruction need to be managed in a specialist facility where they can come in touch with other patients with similar problems and learn that they are not alone. Such a unit needs to be capable of authoritatively investigating oesophageal manometry/pHmetry, anorectal function, intestinal transit, and small intestinal and colonic motility. Histopathological support must be specialist because of the difficulty in making pathological diagnoses in such patients. Surgical care needs to be experienced in order to minimise and optimise surgical intervention - such patients are typically submitted to much fruitless and potentially complicated surgery. While most patients with slow transit constipation do not have intestinal pseudo-obstruction, some do, and it is helpful if the surgical unit involved has an interest in this problem too. Finally a medical clinic familiar with the medical and psycho-social problems of such patients, and capable of coordinating and supervising the overall management, and the nutritional support that

they may need, is mandatory. We must provide a national safe haven for these patients to protect them from over investigation and unnecessary surgery, and to advance their cause if they are treated in ones and twos in a wide variety of units.

Proposal

We at Barts and the London (Powell-Tuck, Sanderson, Meadows, Evans, Williams, Martin, Small, Sharp, Khair, Durman, Vaghjiani et al) wish to form a consortium with the other principal units in the UK in a similar state of development in order to press for NSCAG recognition and funding of adult and paediatric services. At present we envisage this consortium to include, with us, Great Ormond Street (Milla, Pierro, Hill, Smith), St Marks Hospital (Forbes, Silk, Gabe, Kamm) and Hope Hospital, Salford (Shaffer, Carlson et al), but wish to hear from others who wish and are in a position to provide a comprehensive service for these patients as outlined above. *To progress this we seek the formal support of BSPGHAN.*

Reference

1. Vargas JH, Sachs P, Ament ME, et al. Chronic intestinal pseudo-obstruction syndrome in pediatrics. Results of a national survey by members of the North American Society of Pediatric Gastroenterology and Nutrition. 1988;7:323-32.

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Management of Chronic Intestinal Pseudo-obstruction at Barts and the London

At the Royal London Hospital over 45 patients with severe gastro-intestinal dysmotility have been managed in the specialist multiprofessional nutrition/intestinal failure clinic (Powell-Tuck, Durman, Small). Of these 20 have needed home parenteral feeding among whom 13 are current. Patients requiring admission for investigation or treatment are admitted to a specialist combined medical/surgical ward used to the delivery of long-term parenteral nutrition, and to the nursing of the relevant surgical procedures. 50 patients per year are seen by the Academic surgical unit (Williams, Lunniss, Dorudi) for investigation and treatment of severe slow transit constipation of whom a small proportion prove to have pseudo-obstruction. Each year Professor Jo Martin receives requests for opinions on full thickness intestinal biopsies which are needed for the histological diagnosis of pseudo-obstruction.

The Wingate Institute (Evans) is an independently run, purpose-built research unit, affiliated to the School of Medicine and funded in part by a charitable trust, The Whitechapel Society, which has an international reputation for research in the field of neurogastroenterology and motility (Wingate). Areas of interest include the study of neurological mechanisms responsible for functional gastrointestinal disorders. With the Academic Department of Surgery it supports an NHS GI physiology service. The Unit enables measurement of oesophageal function, gastric emptying, intestinal manometry and transit, and anorectal function. It is active in classification of normal motility patterns of the large and small intestine and abnormalities in specific diseases. It is nationally accepted as a key centre for the study of oesophageal, gastric, small and large intestinal and anorectal conditions. It is also closely involved in the evaluation of new techniques in the study of gastrointestinal function with adults and children, and the development of new surgical techniques designed to improve continence and restore quality of life in patients who have undergone anorectal excision. Recent studies have delineated the interaction between gastric electrophysiology, gastrointestinal manometry changes and gastric emptying. The pharmacology of the enteric nervous system (Burleigh) will be shortly added to this group.

Our service is not only well developed in respect of the facilities and expertise available to the adult patient but also by virtue of its intimate links, through the combined academic Department of Adult and Paediatric Gastroenterology with paediatric and adolescent services (Sanderson/Meadows/Croft). We have a close clinical and academic relationship with our paediatrician colleagues and with paediatricians at the Hospital for Sick Children, Great Ormond Street (Milla/Pierro/Hill). We attract patients from all over England and Wales.