Chronic Idiopathic Intestinal Pseudo-obstruction in Infancy and Its Successful Treatment with Parenteral Feeding*

M. J. GREENALL, F.R.C.S., M. H. GOUGH, F.R.C.S.

Greenall MJ, Gough MH. Chronic idiopathic intestinal pseudo-obstruction in infancy and its successful treatment with parenteral feeding. Dis Colon Rectum 1983;26:53–54.

Intestinal pseudo-obstruction is an illness characterized by obstructive symptoms for which there is no mechanical cause. It is becoming increasingly recognized, and there is now a greater understanding of its classification, pathogenesis, and management. Although usually a disease of adulthood, we describe a case occurring in infancy. The difficulties in diagnosis, and the management of this condition in both the short and long terms, with parenteral nutrition, are discussed. [Key words: Pseudo-obstruction; Infancy]

From John Radcliffe Hospital, Headington, Oxford, England

Report of a Case

The patient was the fourth child of healthy young Americans. Two of the children were alive, and well but one had died in infancy after multiple operations on the gastrointestinal tract for similar symptoms.

After a normal delivery, the patient's progress was satisfactory until the age of three months, when he developed intermittent attacks of vomiting and diarrhea. Manipulation of his feeding regimen produced no improvement, and, because of continued weight loss, he was admitted to the hospital for further assessment.

Examination revealed a child who had failed to thrive and whose weight was below the third centile. The only other abnomalities found on examination were those of a slightly distended abdomen and occasional waves of peristalsis in the left hypochondrium. Bowel sounds were high pitched, but rectal examination results were normal. X-rays of the abdomen revealed a dilated stomach, duodenum and proximal jejunum with fluid levels. This dilatation was confirmed by small-bowel x-ray (Fig. 1).

Initial treatment with nasogastric aspiration and intravenous fluids produced no improvement; therefore, laparotomy was performed to exclude a mechanical cause of obstruction. The findings were of an atonic intestine with dilatation of the duodenum and upper jejunum. No mechanical obstruction was found, but during laparotomy no waves of peristalsis were seen. Full-thickness biopsies were taken of the distended jejunum, terminal ileum, and sigmoid colon. A central venous line was inserted for parenteral feeding. After two weeks of intravenous feeding, the nasogastric aspirate was reduced, bowel sounds returned to normal, and gradually normal feeding was reinstituted.

*Received for publication March 22, 1982.

Address correspondence and reprint requests to Mr. Greenall: John Radcliffe Hospital, Headington, Oxford OX3 9DU, England.

Intestinal biopsies showed normal ganglia and nerve fibers and no abnormality of smooth muscle or acetylcholinesterase activity. Results of all other tests, including those of thyroid function, catecholamine, and cortisol secretion were normal.

After discharge, the patient progressed well and gained weight satisfactorily. However, after a further three months, at six months, and again at one year he was readmitted with similar obstructive episodes. X-rays again revealed upper small-bowel obstruction, and treatment on each occasion was by nasogastric suction and nutritional support with parenteral feeding. On all of these occasions, the obstruction settled after about two weeks of treatment.

In the absence of any obstructing lesion, a diagnosis of chronic idiopathic intestinal pseudo-obstruction (CIIP) was made. The clinical course was typical of the condition, the diagnosis also being supported by the suggestion that the other sibling had died of a similar disorder.

Discussion

The term "intestinal pseudo-obstruction" was originally applied to adults with clinical features of bowel obstruction but in whom no disease was found at laparotomy. Subsequently, three distinct entities have been described. There is an acute and usually transient form affecting the colon of elderly patients who have suffered anoxic episodes. Second, there is a chronic form associated with systemic disease and drug therapy, and third, a chronic idiopathic type.

CIIP usually presents in young adulthood and may affect the small or large intestine. The histologic appearance of the bowel of such patients is very variable since in many cases, the intestine is free from pathologic change, but in others there is muscle hypertrophy, atrophy, or replacement with collagen.[1] The myenteric plexus may be normal, but sometimes the ganglia and nerve bundles are reduced in number and are morphologically abnormal. In one case, there was an absence of the argyrophil plexus and acetyl-

0012-3706/83/0100/0053/$00.90 © American Society of Colon and Rectal Surgeons

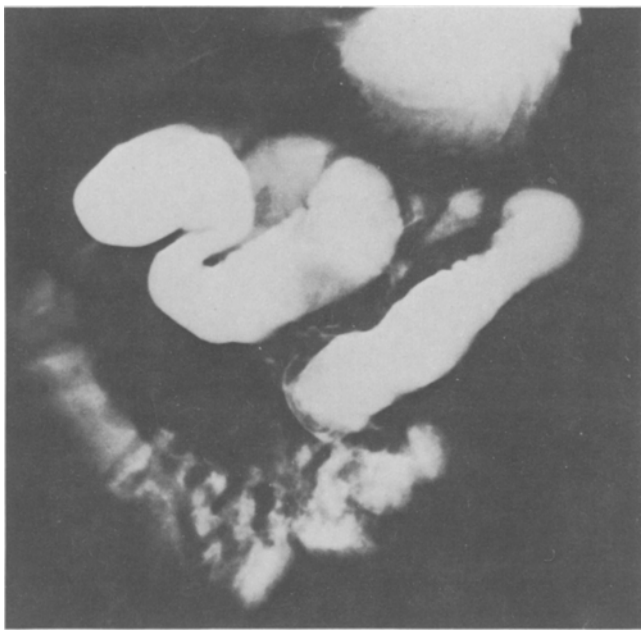

FIG. 1. Small-bowel x-ray showing obstruction of upper jejunum.

cholinesterase activity, in spite of the presence of normal ganglion cells.[2]

CIIP presents with recurrent attacks of intestinal obstruction of varying severity. Unlike organic obstruction, diarrhea with steatorrhea and malabsorption may occur. Remission may last for several years, but eventually chronic subacute obstruction supervenes, leading to progressive malnutrition.[1]

As well as intermittent intestinal obstruction, there may be evidence of abnormal esophageal motility, megaduodenum, urinary retention, ureteric reflux, and defects of pupillary, sweating, and cardiovascular function.[3–5] The widespread nature of the disease and its frequent presence in relatives has led to its description by Schuffler *et al.* as "hereditary hollow visceral myopathy."[3,4]

Infantile intestinal obstruction due to CIIP is uncommon and has a poor prognosis. There were only two survivors out of a total of 11 such cases described by Sieber and Girdany[6] and Kapila *et al.* [7] Other cases presenting in infancy or childhood were described by Puri[2] and Byrne *et al.*[8] "Functional obstruction" due to metabolic disease, septicemia, or electrolyte imbalance must be excluded before CIIP can be diagnosed. Contrast radiology may determine the site of obstruction, but a mechanical cause cannot be excluded unless there is a preexisting family history or other evidence of "hollow visceral myopathy."

If laparotomy is required to establish the diagnosis, full-thickness intestinal biopsies should be taken to exclude Hirschsprung's disease and abnormalities of acetylcholinesterase activity. Bowel decompression with ileostomy or colostomy has not been successful,[7,8] although operations to remove or bypass dilated segments of small intestine have occasionally relieved symptoms.[1]

There is no effective treatment for CIIP. Attempts at medical treatment with steroids, antibiotics, cholinergic or antiadrenergic drugs and gluten-free diets have been disappointing.[1] More recent recommendations, such as the intestinal stimulant cerulein[7] or prostaglandin synthesis inhibitors like indomethacin,[1] are only hypothetical.

Treatment involves the conservative management of obstructive episodes with nasogastric suction and fluid replacement. Nutritional support should be provided with parenteral feeding,[1] this having been the basis of treatment in our case. Early and vigorous conservative management with nasogastric suction and parenteral feeding should be instituted at the first sign of recurrent obstruction. Further surgical exploration should be avoided in spite of the possibility of adhesive obstruction resulting from previous laparotomies.

CIIP is a generalized disease of the gastrointestinal tract which presents as recurrent paralytic or adynamic ileus. Although rare, its recognition, as a cause of obstruction in the neonate or in infancy is important, since the preferential treatment is nonsurgical. This will prevent the augmented effect of even mild adhesions in the presence of an intermittently adynamic bowel.

References

1. Faulk DL, Anuras S, Christensen J. Chronic intestinal pseudoobstruction. Gastroenterology 1978;74:922–31.
2. Puri P, Lake BD, Nixon HH. Adynamic bowel syndrome: report of a case with disturbance of the cholinergic innervation. Gut 1977;18:754–9.
3. Schuffler MD, Lowe MC, Bill AH. Studies of idiopathic intestinal pseudoobstruction. I. Hereditary hollow visceral myopathy: clinical and pathological studies. Gastroenterology 1977;73:327–38.
4. Schuffler MD, Pope CE II. Studies of idiopathic intestinal pseudoobstruction. II. Hereditary hollow visceral myopathy: family studies. Gastroenterology 1977;73:339–44.
5. Bannister R, Hoyes AD. Generalized smooth-muscle disease with defective muscarinic-receptor function. Br Med J 1981;282:1015–8.
6. Sieber WK, Girdany BR. Functional intestinal obstruction in newborn infants with morphologically normal gastrointestinal tracts. Surgery 1963;53:357–61.
7. Kapila L, Haberkorn S, Nixon HH. Chronic adynamic bowel simulating Hirschsprung's disease. J Pediatr Surg 1975;10:885–92.
8. Byrne WJ, Cipel L, Euler AR, Halpin TC, Ament ME. Chronic idiopathic intestinal psuedo-obstruction syndrome in children—clinical characteristics and prognosis. J Pediatr 1977;90:585–9.