INTESTINAL PSEUDO-OBSTRUCTION WITH STEATORRHOEA*

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A hitherto undescribed syndrome is recorded. Its features are gross overactivity of the small intestine with episodes of obstruction, but without any mechanical factor being found, and steatorrhoea. In one patient the inner circular coat showed gross hypertrophy.

In the past few years we have encountered three cases of intractable wasting and diarrhoea in which attacks of intestinal pseudo-obstruction have occurred. Two of them will not be discussed in detail because in one, a woman of 52 years, the jejunum showed the histological appearances of adult coeliac disease as described by Paulley (1954) and Shiner (1956). At laparotomy the peristaltic activity of the small intestine was grossly abnormal, showing violent segmentation movements. It was impossible to alleviate her condition, from which she had suffered for 20 years, and she has recently died elsewhere, some three years after coming under our care. Another patient, a woman, aged 56, who in four years had two attacks of pseudo-obstruction leading to laparotomy and lost 70 lb. from steatorrhoea, was eventually explored for a perforation, and a very small reticulo-sarcoma of the ileum was found, from which she died soon after. It is extremely doubtful whether this sarcoma could have accounted for the long preceding history.

Our third patient, a man aged 36, has been carefully investigated both by us and by Dr. Avery Jones at the Central Middlesex Hospital, and so far we have not been able either to find a known cause for his condition or to alleviate it substantially. The main features of this case, similar in many respects to the two previously mentioned, are (1) a long history of colicky, abdominal pain; (2) fatty diarrhoea, malabsorption, and loss of weight; (3) gross abdominal distention with audible and visible intestinal peristalsis; (4) episodes of pseudo-obstruction; (5) no mechanical cause for obstruction found at laparotomy; (6) marked segmentation movements of the gut visible radiologically and at laparotomy; (7) absence of hydroxyindoleacetic

acid in the urine and absence of gastric hypersecretion (800 ml. per 24 hr.), no anaemia, normal erythrocytic sedimentation rate, and no L.E. cells.

CASE REPORT

CASE 1.—B, a man aged 36, began to have abdominal symptoms of pain and discomfort in 1947 when aged 24. A duodenal ulcer was diagnosed radiologically and he obtained some relief of symptoms by dietary treatment. In 1953, when aged 30, he had an emergency appendectomy but abdominal colic and post-prandial discomfort continued after this. Mild diarrhoea occurred from time to time, and by 1957, the pattern of symptoms was established as noisy abdominal colics at all times of day but worse after meals; three or four loose stools a day; some abdominal distention and progressive loss of weight. A barium meal at this time showed only rapid gastric emptying.

In May, 1958, when in Weston-super-Mare Hospital for the investigation of his symptoms, he developed more severe abdominal pain and tenderness, a rapid pulse, and signs of shock. He was treated conservatively and discharged from hospital, but was readmitted six weeks later with abdominal pain and vomiting. A laparotomy showed that a small abscess cavity had formed in the lower peritoneum from a minute perforation of the small intestine. The terminal ileum appeared to be in spasm but there was no evidence of regional ileitis.

A barium meal in September, 1958, showed great dilution of the barium in the small gut. A blood count and the E.S.R. were normal.

On November 20, 1958, he was admitted to Southmead Hospital, Bristol, where the findings were: Weight 103 lb., a distended abdomen, which showed phases of visible and audible peristalsis accompanied sometimes by pain. The stools were greasy in appearance with a mean daily fat excretion over three days of 17 g. Sigmoidoscopy showed an oedematous mucosa which, on biopsy, appeared normal. A straight radiograph of the abdomen showed distended coils of small bowel (Fig. 1).

Barium studies were fraught with difficulties due to delay in stomach emptying and to great dilution of the

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barium in the small bowel. Segmentation movements were observed and the barium, which flocculated easily, was shuttled back and forth in the loops of intestine.

In December, 1958, he had a seven-day phase of increased abdominal colics and hyperperistalsis.

On December 16, 1958, laparotomy (W.M.C.) was performed and a reddened small intestine covered with flakes of fibrinous exudate was found. Peristalsis was very active and segmentation movements marked. The wall of the small intestine was possibly oedematous.

A biopsy of liver and peritoneal fibrin nodules showed no specific pathology. Biopsy of the jejunal wall showed normal mucosa, submucosa, and muscularis mucosae, but a greatly thickened inner muscular coat with a good deal of vacuolation of the muscle fibres.

After operation, the patient was given a gluten-free diet and this has been continued to date without clear-cut improvement.

From January 20, 1950, he was given "prednisone", 40 mg. per day, with potassium chloride by mouth, but this seemed to make no difference and he was weaned off it after two months.

He had a further attack of pseudo-obstruction on January 31, 1959, but this was successfully treated by intravenous therapy and intestinal suction.

Twenty-four-hour gastric aspiration showed no evidence of hyperchlorhydric hypersecretion (800 ml.

FIG. 1.—Barium in the small intestine.

FIG. 2.—Section of patient's small intestine (left) with serosa at the bottom, showing thickened inner or circular muscle coat, with a section of normal small intestine for comparison (right).

per 24 hours). The urine contained no excess of hydroxy-indoleacetic acid but large quantities of indican.

The patient was subsequently reinvestigated by Dr. Avery Jones at the Central Middlesex Hospital with similar findings and on intraluminal biopsy of the jejunum Dr. Shiner confirmed the normal mucosal appearances. Radiologically it was then impossible to fill the terminal ileum, and possible diagnoses of intraluminal tumour or an achalasia of the ileocaecal valve were considered. However, a further abdominal exploration was not considered justifiable.

Subsequently the patient has been able to resume work. He continues to take a gluten-free diet. He has had in the past six months two attacks of pseudo-intestinal obstruction, the first a minor one treated by a day's gastric aspiration and rest, the second a more prolonged affair when continuous gastric suction and intravenous therapy had to be continued for several days, following a severe mental shock.

The state of the abdomen varies greatly from day to day and hour to hour. Phases of great distension are followed by phases of loud peristaltic activity and occasionally the abdomen is relatively flat. The patient is anxious to keep working and to avoid further operations.

The Morbid Anatomy of the Gut.—The appearances of the small intestine at laparotomy were notable dynamically, segmentation movements being most pronounced. In addition the gut was hyperaemic and the intestinal wall appeared thickened. On histological examination the most striking feature was the thickness of the inner circular muscle coat, which is about 10 times the thickness of the outer or longitudinal coat (Fig. 2). Through the kindness of Dr. Paulley we were able to examine several of his sections of normal small intestine obtained at operation, and these, though they show the difficulty of measuring the thickness of muscle coats due to varying directions of the cut, are quite different because in none is the inner coat more than four times the thickness of the outer coat.

The hypertrophied inner muscle shows marked vacuolation of the fibres and this feature is also present, though less noticeably, in the outer coat. The muscle fibres of the inner coat have indistinct outlines but in both coats the nuclei appear normal. The ganglion cells and nerve fibres of Auerbach's and Meissner's plexuses show no abnormality.

DISCUSSION

This case presents us with many problems. First, have others encountered a similar clinical problem? There is no reference in the literature to a condition specifically like this one, but there are a number of cases described which are similar in many respects. In the literature concerning steatorrhoea, there are cases recorded where obstructive symptoms have developed leading to useless laparotomy. Cooke, Peeney, and Hawkins (1953) quote two patients with adult steatorrhoea who had recurrent intestinal colic, but both were

relieved to a certain extent of their symptoms by potassium repletion. One of them had a negative laparotomy.

Ingelfinger (1943) also describes a patient with steatorrhoea of 15 years' duration who suddenly had episodes of intestinal obstruction. At laparotomy the gut wall was reported as loose and flabby. Two ileostomies were carried out at different times, and later closed after the malabsorption state had been thoroughly treated. Even so the patient still suffered from cramps and noisy gut activity.

Murley (1959) has described the case of a woman with multiple sclerosis and migraine who had symptoms of intermittent intestinal obstruction. There was steatorrhoea, radiological evidence of disordered peristalsis in the duodenum and jejunum with backwash into the stomach, and at operation the jejunum was thickened and the duodenum dilated. Biopsy showed no specific features of any known disease. Dr. Murley has kindly let us see sections of the small intestine from this patient, which do not show the thickening of the inner muscle coat observed in our patient. Oedema of the muscular coat and a degree of cellular infiltration in Dr. Murley's specimen may fall within the limits of the normal.

Ihre (personal communication, 1959) has told us of a patient of his, a 35-year-old man who began to suffer from abdominal pain and meteorism in 1951. Five years later he was worse and had lost 5 kg. in six months. Abdominal distension occurred in bouts every afternoon and was somewhat relieved by passing wind. Two years later, vomiting, diarrhoea, and steatorrhoea were all noted again, as well as the radiological appearance of dilated intestines. A laparotomy revealed no mechanical cause for the obstructive symptoms nor any specific pathology. Only intravenous feeding prevented the attacks of pseudo-obstruction. Jejunostomies were performed in the next two months, but without improvement and he died, necropsy revealing no explanation for the cause of the malady.

Pilkington (personal communication, 1959) has described to us the case of a man with diabetes and polyneuritis who developed marked abdominal distension and steatorrhoea. Laparotomy was carried out but the clinical suspicion of Whipple's disease was not borne out by the histological findings. Abdominal distension and vomiting continued until death ensued six months later. Necropsy revealed no cause for the condition, though the small intestine was found to be distended and thickened in places. We have been privileged to examine this specimen which shows none of the apparent muscular hypertrophy observed in our

TABLE I
MAIN FEATURES OF SOME CASES OF PSEUDO OBSTRUCTION WITH STEATORRHOEA

Patient and Age	Length of History (in years)	Vomiting and Distention During Attacks (Pseudo Obstruction)	Steatorrhoea and Weight Loss	Noisy and Disordered Peristalsis	Gut Histology
Male 36 (Naish and Capper)	10	++	++	++	Inner muscular coat hypertrophied, mucosa normal
Male 36 (Ihre)	7	++	++	++	Not obtained
Male 26 (Pilkington)	1	Continuous distention and vomiting	+++	+	Muscle coats normal, mucosa autolysed
Female 50 (Murley)	2	Vomiting only, no distention	++	++	Muscle coats normal, mucosa normal
Female 46 (Dudley <i>et al.</i>)	16	++	++	+	No laparotomy
Female 73 (Lang Stevenson)		++	++	++	Muscle coats hypertrophied, diverticula present
Female 50 (Naish and Capper)	16	+	+++	++	Resembles adult coeliac disease

case. The mucosa has been damaged by post-mortem digestion and no opinion can be reached on its living state.

Dudley, Sinclair, McLaren, McNair, and Newsam (1958), in reviewing the clinical problem of intestinal pseudo-obstruction or "spastic ileus", record a number of cases where laparotomies were undertaken to relieve a mechanical obstruction which was not found. Their Case 4, a woman aged 46, had bouts of abdominal pain and vomiting for 16 years. Radiographs showed distended loops of intestine and multiple fluid levels and a mouthful of barium remained in the pyloric region for days. In the light of previous experience, conservative treatment was advised.

Lang Stevenson (personal communication, 1959) has drawn our attention to a patient, a woman aged 73, who suffered from repeated vomiting and steatorrhoea and whose jejunum showed disordered and ineffective peristalsis which was visible through the abdominal wall, radiologically and at laparotomy. This patient also had a number of duodenal and jejunal diverticula for which she underwent operation. She gave a six months' history of noisy intestinal rumbling and a shorter history of pain and projectile vomiting of food eaten up to 12 hours previously. She had lost 14 lb. in weight and her haemoglobin was 8.4 g.%. Peristalsis was easily visible through the abdominal wall, and appeared to pass to both right and left. Laparotomy showed dilatation and hypertrophy of the duodenum and the jejunum with diverticula in both areas. In the jejunum, waves of peristalsis were very active and passed in both directions, the

gut writhing violently. Beyond the affected area, peristalsis was orderly and tailwards.

Three duodenal diverticula were removed and the affected area of jejunum (2 ft.) excised, continuity being restored by end-to-end anastomosis. A retrocolic gastro-jejunostomy was also done. After operation the patient did well, lost her symptoms, and regained her former weight.

Microscopical sections of small intestine kindly forwarded to us show striking changes in the muscular coats. The appearances vary a little in sections from different levels but in most the inner muscle coat is considerably thicker than the outer. Where this feature is most marked the nuclei of the muscle fibres of the outer coat are very abnormal, many being greatly increased in size and of bizarre pattern and shape. Vacuolation of muscle fibres is not seen in the inner coat but is present to a slight degree in the outer coat. The mucosa, submucosa, muscularis mucosae, and nerve plexuses appear normal. In the diverticula both muscle coats are thinned out but no other special features are seen.

The features of the cases described are none of them uniform but in all there was evidence of disordered peristalsis of the small intestine, attacks of vomiting and distention, diarrhoea with steatorrhoea, and loss of weight (Table I).

The second major problem posed is the reason for the increased but ineffective peristaltic activity of the small intestine in our case and these other similar cases. Could this be due to an immobile segment of gut at the ileo-caecal valve, comparable with the condition of achalasia of the oesophagus or congenital megacolon? Against this hypothesis

is the absence of any evidence for such a state of affairs in our case at laparotomy, and the absence of any loss of ganglion cells. In other cases the condition appeared to affect the jejunum only. Furthermore increased peristaltic activity is a feature neither of oesophageal achalasia nor of megacolon.

Could the increased peristalsis and segmentation be due to the presence of excess quantities of a chemical substance such as serotonin? None of the end-products of serotonin breakdown have been found in the urine, and the presence of large quantities of urinary indican may be a reflection only of the stagnation of intestinal contents.

Speculation as to possible causes leads us to look for possible clues from the study of other similar but not identical cases. Two of the patients, Murley's and Pilkington's, had diseases of the central nervous system which could conceivably have affected the central connexions of the autonomic nerve fibres supplying the gut. The comment of the surgeons, who were shown the film of our patient's intestinal behaviour at laparotomy at a meeting of the Surgical Section of the Royal Society of Medicine, was that the patient must have been under spinal anaesthesia. This was not so. But could he have

some disease of the spinal cord or its autonomic outgoing connexion which produces the same effects on movement of the gut as spinal anaesthesia? Is perhaps the inhibiting control exercised by the spinal cord and the autonomic nervous system more important in the smooth regulation of gut peristalsis than we are accustomed to recognize? We tend perhaps to regard tailward peristalsis as an innate function of a tube of smooth muscle and we regard the autonomic nervous system merely as the controller of valves and the speed of transit. Anyone who has witnessed the acute abdominal distension and ileus of a patient with an extradural vertebral abscess may doubt this; and the case of the patient we have described must stimulate thought on the mechanisms controlling a process which in most humans is so trouble free.

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