Idiopathic steatorrhea with intestinal pseudo-obstruction

H. A. McCLELLAND, M. J. LEWIS, AND J. M. NAISH

From Southmead Hospital, Bristol

EDITORIAL SYNOPSIS  This is a case report illustrating a disorder of small intestine motility producing gross muscular hypertrophy and associated with steatorrhoea. The literature is briefly reviewed, and it is suggested that in idiopathic steatorrhoea there is ineffectual peristalsis which leads to stagnation of intestinal contents and hypertrophy of its muscular wall.

Writers on the subject of adult steatorrhoea have commented on the abdominal colic and episodes simulating intestinal obstruction which may occur (Badenoch, 1960; Avery Jones and Gummer, 1960). A disorder of bowel motility is presumed to be the cause. The case presented here is that of a patient in whom severe malabsorption and prolonged attacks of intestinal pseudo-obstruction culminated in death after an illness lasting eight months.

CASE REPORT

A housewife, aged 55, in February 1960 developed severe diarrhoea with increasing abdominal distension. In March she was admitted to Weston-super-Mare Hospital with abdominal pain and vomiting, having passed no flatus rectally for 24 hours. There was abdominal distension and marked peristalsis.

At laparotomy a chylous peritonitis was found, and the upper half of the small intestine was distended, with a considerably hypertrophied wall. A diagnosis of volvulus was made. Following the operation her symptoms improved, but another acute episode of abdominal colic and vomiting three weeks later led to a second operation, with similar findings. Nevertheless, recurrent pain and vomiting continued and in May a plication operation was performed, with no benefit to the patient. Her symptoms persisted, with the passage of six to eight loose bulky stools a day. Investigation revealed steatorrhoea and she was transferred to Southmead Hospital.

At this time the patient said that she had had no diarrhoea before the present illness, but her husband said that she had suffered transient episodes of diarrhoea during the previous four years. There was no family history of bowel disorders.

Examination showed a pigmented, very emaciated woman weighing 43·3 kg. with a persistently low temperature of 96° to 97°F. Her blood pressure was 125/90 mm. Hg. She had marked ankle and sacral oedema. The abdomen was grossly distended. At rest only three to four bowel sounds were heard in seven minutes, but at intervals they reached a normal frequency and coincided with a desire to defaecate. Knee and ankle jerks were absent and there was some sensory loss below the ankles. Her stools were loose, bulky, and very offensive.

The mean daily faecal fat excretion was 18 g., Hb 86%, blood urea 21 mg %, serum albumin 3 g %, and serum globulin 1·9 g %, serum sodium 130 mEq/l., serum potassium 3·0 mEq/l., and serum calcium 7·5 mg %. The alkaline phosphatase was 4 King-Armstrong units, cholesterol 99 mg %. A xylose absorption test showed the excretion of 1·02 g. of a 25-g. test dose in five hours.

Barium cine studies showed a dilated small bowel with many fluid levels and an unusually smooth wall. Transport seemed to be mainly by spillover as peristalsis was very slow and shallow. The barium tended to pool beneath the resting fluid, and remained there for days, although the transit time of a carmine marker through the gastrointestinal tract was only 18 hours. Attempts at jejunal biopsy failed, the Crosby capsule on one occasion lying in the pyloric canal for 36 hours without passing into the duodenum.

From her admission to Southmead Hospital on 14 September 1960 to her death five weeks later, the patient was on a gluten-free diet and was given parenteral vitamins, with no appreciable effect. Intravenous infusions containing high concentrations of potassium did not cause any improvement and broad-spectrum antibiotics were of no value. Cortisone was not given. She had frequent bowel colic, most marked before defaecation, and repeated attacks of intestinal pseudo-obstruction lasting for days and needing suction and intravenous therapy. During one of these episodes the patient died. Formalin was instilled into the stomach immediately after death.

Necropsy was performed 13 hours after death. A small quantity of free fluid was found in the peritoneal cavity. Macroscopically the duodenum and first six feet of the small intestine were greatly dilated and their wall was...
greatly thickened, but thereafter the lumen gradually narrowed and the wall became less thick until in the terminal eight feet of the ileum the appearances were normal, apart from two dilated sections, each about 6 in. long (Figs. 1, 2, and 3). The stomach was dilated but the wall appeared normal. The colon showed no gross abnormality.

Histologically there was no abnormality in the wall of the stomach. In the small intestine the muscle coats were hypertrophied at all levels, the hypertrophy being more marked in the duodenum and jejunum than in the ileum. The individual muscle fibres varied from 10 to 30μ in thickness, the normal, as quoted by Maximow and Bloom (1952), being 6μ. The duodenum had been partially fixed by the formalin, but here and throughout the small intestine post-mortem autolysis obscured the mucosal appearances. The plexi of Auerbach and Meissner were normal. Other organs showed a marked reduction in size from the normal, the heart and liver showing brown atrophy.

DISCUSSION

In this case the most striking features were abdominal colic, episodes resembling intestinal obstruction, and the great muscular hypertrophy of the duodenum and jejunum found at necropsy. At no time was a definite mechanical obstruction of the bowel demonstrated. In retrospect, we must accept that the findings at laparotomy in the early stages of the disease were of pseudo-obstruction mimicking the appearances of a volvulus, especially as later episodes settled spontaneously on conservative therapy. The concept of pseudo-obstruction is necessary in order adequately to discuss this case and other sporadic cases that have been reported in the literature (Dudley, Sinclair, McLaren, McNair, and Newsam, 1958) in which the clinical features of intestinal obstruction have been present but without a demonstrable mechanical cause. It is most unfortunate that in this case no satisfactory histological examination of the mucosa could be made.

In jejunal diverticulosis, in which the histological appearances of the mucosa are normal, hypertrophy of the intestinal muscle, pseudo-obstruction, and steatorrhoea have been reported singly or in combination (Phillips, 1953; Badenoch, Bedford, and Evans, 1955). It has been suggested (Edwards, 1939) that in this disease the primary disorder is irregular peristalsis causing increased tension within segments of relaxed bowel.

Naish, Capper, and Brown (1960) have reported a case in which episodes of disordered hyperperistalsis were associated with steatorrhoea and intestinal pseudo-obstruction. The muscle layers of the jejunum were greatly thickened but the mucosa was normal. Similar cases in the literature were reviewed,
in which the combination of muscle hypertrophy with a normal mucosa indicates that disorders of motility with steatorrhoea need not be associated with mucosal change. In recent years, with the advent of peroral biopsy instruments, attention has been directed to the abnormal mucosal patterns which may be seen in steatorrhoea. Thus, in coeliac disease of children and in many cases of adult steatorrhoea a subtotal villous atrophy ('flat biopsy') is a very common finding (Shiner and Doniach, 1960). The changes of a partial villous atrophy ('abnormal villi') appear to be associated with a group of less well-defined syndromes which may, or may not, respond to a gluten-free diet. Fone, Cooke, Meynell, Brewer, Harris, and Cox (1960) report that of 12 patients in their group II with partial cilioid atrophy who had malaise and diarrhoea, in five colicky abdominal pains were a feature and two of these came to laparotomy. Conversely, Fone et al. found in their group I (subtotal villous atrophy) that abdominal pain and distension were rare except when hypokalaemia was present.

There are very few detailed reports of the intestinal wall in sprue during life or on post-mortem examination. Thinning and atrophy of the wall is the most frequently noted feature (Macgregor, 1960; Adlersberg and Schein, 1947; Himes and Adlersberg, 1958). This accords well with the reduced motility so frequently seen on x-ray examination and with the diminished intraluminal basal pressures and reduced phasic (segmental) activity that have been described by Fink (1959). These findings may well be due, as Schneider, Bishop, Shaw, and Frazer (1960) have shown, to a gluten factor which can inhibit contraction in isolated strips of normal intestine. But though this may be so in the majority of cases of adult steatorrhoea, there appears to be a smaller ill-defined group in which the clinical features and variable mucosal patterns are associated with a peristaltic disorder of a different nature. In these patients, careful examination of the muscle coat would be of considerable interest.

In passing it may be noted that a simple increase in intestinal motility by itself need not necessarily lead to steatorrhoea or muscle hypertrophy. This is seen in 'diabetic diarrhoea' which is usually attributed to a disturbance of the autonomic nervous system. Many such cases with very rapid transit times have been recorded, both with and without steatorrhoea (Malins and French, 1957). Berge, Sprague, and Bennett (1956), in a series of six post-mortem reports on patients with 'diabetic diarrhoea', including one with steatorrhoea, did not find any mucosal or muscle abnormality.

Normal bowel movement is an intricate complex of activity involving muscle tone, segmentation, and peristalsis. A disorder which alters any one of these factors may disturb the normal progress of the peristaltic wave. Thus a local increase in tone or alteration of segmental activity (perhaps initiated by absorption of substances from the bowel lumen) might cause increased but non-propulsive movements. Although basal pressures are the same at all levels of the normal small intestine (Fink) there appears to be a gradient of activity for rhythm, irritability, and susceptibility to drugs (Best and Taylor, 1961) which is maximal in the duodenum and becomes progressively less along the jejunum and ileum. Therefore any product toxic to the affected patient (perhaps derived from food or a bacterial metabolite) may be expected to have its greatest effect on the upper small intestine. This might be related to the finding that the muscle hypertrophy, when present, is greatest in this region.

The subject of adult idiopathic steatorrhoea is under intensive study but more precise clinicopathological descriptions are required, especially from post-mortem reports and from full-thickness bowel biopsies taken at laparotomy. Image intensification has made barium cine records possible but the analysis of the motility patterns, which may vary from day to day in the same patient, calls for new techniques.

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REFERENCES


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