

# Megaduodenum due to hollow visceral myopathy successfully managed by duodenoplasty and feeding jejunostomy

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## Abstract

**A 29 year old man with a history of childhood polymyositis developed insulin dependent diabetes and was found coincidentally to have chronic intestinal pseudo-obstruction due to visceral myopathy. Multiple full thickness biopsy specimens showed severe disease in the duodenum and the proximal jejunum only, with less involvement distally. Total parenteral nutrition has been avoided for more than a year by enteral feeding through a fine bore jejunostomy catheter positioned with its tip in the distal jejunum.**

Chronic intestinal pseudo-obstruction is a rare condition<sup>1</sup> which may be secondary to systemic disease – for example, scleroderma – or due to idiopathic degeneration of visceral nerves<sup>2</sup> or muscle.<sup>3</sup> Clinically, the final common pathway is motor failure of the intestine leading to abdominal distension, vomiting, and, less commonly, pain. In each case diagnosis of the specific cause requires full thickness small bowel biopsy. Laparotomy may also be needed to exclude other causes of intestinal obstruction such as bands or tumours. Treatment for chronic intestinal pseudo-obstruction remains problematic, although some success has been claimed for prokinetic agents.<sup>4</sup> Many patients undergo intestinal bypass but this often fails to relieve symptoms as it may be impossible at operation to differentiate between normal and abnormal bowel. Multiple laparotomies should be avoided because subsequent adhesions may add to the patient's symptoms and cause diagnostic problems. In the past many patients died of inanition but recently the radical solution of removal of all of the small bowel followed by total parenteral nutrition has been advocated in end stage chronic intestinal pseudo-obstruction.<sup>5</sup>

The case reported here illustrates the successful use of a feeding jejunostomy together with a temporary draining gastrostomy in the management of a patient with insulin dependent diabetes and chronic intestinal pseudo-obstruction due to hollow visceral myopathy.

## Case report

The patient presented to another hospital at age 13 years with a six month history of muscular weakness. Polymyositis was diagnosed on the basis of a creatine kinase activity of 1090 mU/l (reference range <100 mU/l), an electro-myogram showing a mixed neuropathic and myopathic picture, and histology of skeletal muscle biopsy (discussed below). He improved

clinically and biochemically on prednisolone which was discontinued after 12 months.

He remained well until April 1988 when, aged 29, he presented with typical symptoms and signs of insulin dependent diabetes mellitus including weight loss (15 kg in total), thirst, polyuria, and fatigue. He also described epigastric discomfort and occasional vomiting during the previous three months.

On examination, the striking findings were emaciation (weight 42.5 kg, body mass index 12.8 kg/m<sup>2</sup>), a lack of subcutaneous adipose tissue, and mild contractures in the arms consistent with previous polymyositis (Fig 1). There were no diabetic complications. He had ketonuria, a blood glucose concentration of 24.0 mmol/l, a glycosylated haemoglobin (HbA<sub>1c</sub>) of

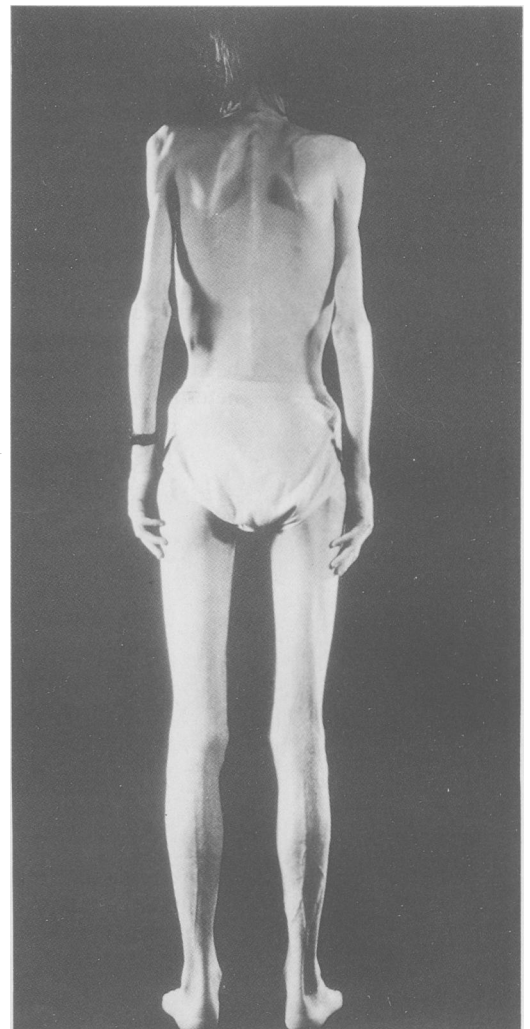


Figure 1: Patient with chronic intestinal pseudo-obstruction due to visceral myopathy.

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14.7% (reference range 4–8%), and pancreatic islet cell antibodies in a titre of 1 in 32. Urea and electrolyte concentrations and thyroid function tests were normal.

He was started on Monotard insulin, 8 units twice daily, but over the next three months, his weight fell to 40 kg despite good diabetic control (HbA<sub>1c</sub> 10.2%) and his symptoms of epigastric fullness and vomiting worsened. A plain abdominal x ray film showed a full stomach six hours after his last meal. When admitted for investigation there were no new findings. Full blood count, prothrombin ratio, serum hydroxycobalamin, and red cell folate concentrations were normal and the erythrocyte sedimentation rate was 1 mm in the first hour. Serum sodium, potassium, calcium, magnesium, bicarbonate, urea, and creatinine concentrations, creatinine kinase activity, and liver function tests were also normal except for a marginally low serum total protein concentration of 58 g/l (reference range 63–78 g/l). Normal inspiratory/expiratory variation in the heart rate excluded autonomic neuropathy. Serum immunoglobulin, C3, C4, C3 degradation products, and C reactive protein concentrations were normal, autoantibodies (other than islet cell) were negative, antiextractable DNA antibody was not detected and x rays of the hands were normal. In view of his weight loss and poor general condition total parenteral nutrition was instituted.

A barium meal (Fig 2) showed the oesophagus and stomach to outline normally. There was a megaduodenum with apparent obstruction at the junction of the second and third parts and, even after three hours, hardly any barium was seen in the small bowel, which appeared dilated. Gastroscopy showed mild reflux oesophagitis; there was

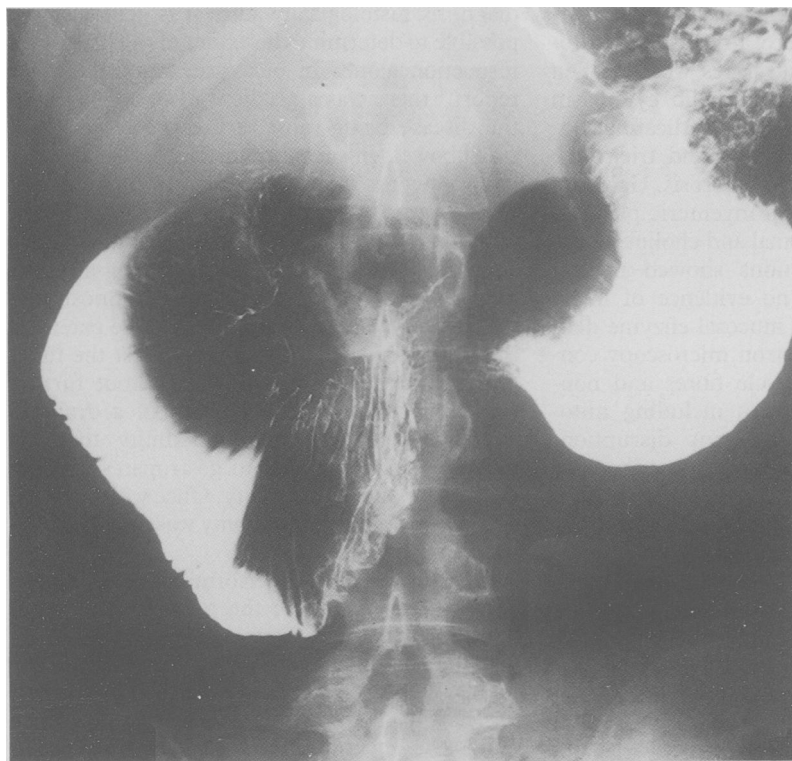


Figure 2: Barium meal in chronic intestinal pseudo-obstruction due to visceral myopathy showing dilated stomach and megaduodenum which failed to propel barium into the jejunum even after three hours.

a residue of food in an otherwise normal stomach in which peristalsis was observed; the pylorus was normal but the first and second parts of the duodenum were dilated. Histology of gastric and duodenal mucosal biopsy specimens was normal.

Upper gastrointestinal motility studies were performed, the provisional diagnosis now being chronic intestinal pseudo-obstruction. Oesophageal manometry was normal but since the probe could not be passed beyond the first part of the duodenum, small bowel motility could not be recorded. The gastric recording, however, did show intact migrating motor complexes. A barium enema was normal.

After four weeks of total parenteral nutrition his nutritional state had improved enough for a diagnostic laparotomy. The stomach and duodenum were dilated but otherwise appeared normal, as did the small bowel and other abdominal organs. A full thickness biopsy specimen was taken from the jejunum two feet below the duodenojejunal flexure and the rectus abdominis muscle was also biopsied. Histology of the jejunal biopsy specimen was consistent with visceral myopathy (see below). On the basis of this definitive diagnosis a management plan was devised. A further laparotomy was performed in which the excess duodenum was excised with duodenoplasty to render the duodenum narrower. A drainage gastrostomy tube was inserted and a fine bore feeding jejunostomy catheter sited in the mid-jejunum. Full thickness biopsy specimens were taken of the stomach, mid-jejunum, and upper and lower ileum to assess the extent of the disease (see below).

One week after operation the gastrostomy tube was removed in view of the low volume of drainage. At the same time jejunal tube feeding began and was so well tolerated that total parenteral nutrition could be discontinued. The patient was taught to administer his own enteral feeding and was discharged, taking 2000 ml of Osmolite (Abbott Laboratories, Queensborough, Kent) providing 8.36 MJ/day. The enteral feed is infused continuously at 200 ml/hour overnight and is discontinued during the day which allows him freedom for his work and daily activities. This unusual pattern of energy intake necessitates a tailored insulin regimen. Currently he injects Monotard 5 units in the morning, and a mixture of 15 units of Monotard and 15 of Actrapid before starting the nighttime enteral feed. Diabetic control is good with no hypoglycaemic attacks and an HbA<sub>1c</sub> of 10.6%. He weighs 47 kg (body mass index 14.8 kg/m<sup>2</sup>) and can tolerate small amounts of food by mouth but still vomits two or three times a week. Hydroxycobalamin (1000 µg) intramuscularly is given every three months. He has remained clinically and biochemically stable on this regimen for 13 months.

## Histology

### SKELETAL MUSCLE

The biopsy specimens of deltoid and vastus lateralis taken at age 13 showed typical features of polymyositis with foci of muscle degeneration and necrosis, an interstitial lymphocytic infil-



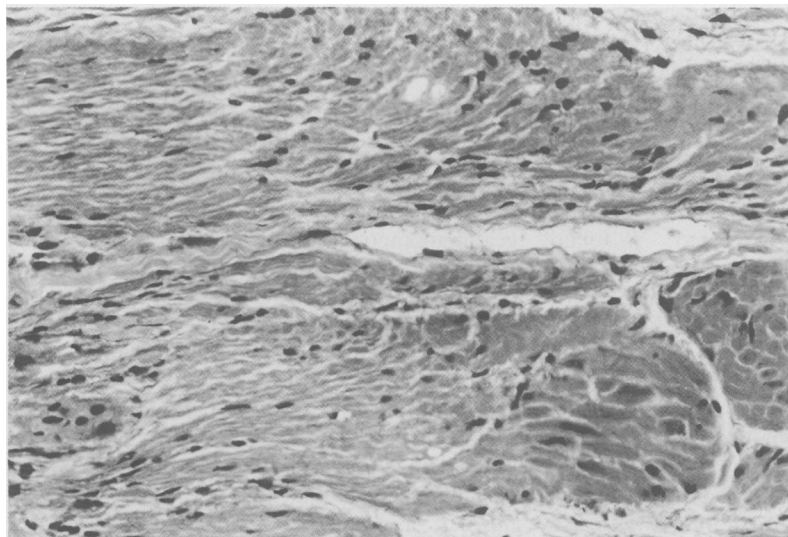


Figure 3: Muscularis propria of jejunum (inner circular layer). This area shows the main abnormalities of the muscle ranging from thin, atrophic fibres towards the outer aspect (left of field) to large eosinophilic fibres with hyperchromatic nuclei towards the submucosal aspect (right of field). Small vacuoles are also seen at the centre. (Haematoxylin and eosin, cryostat section, original magnification  $\times 310$ ).

trate, and a lymphocytic vasculitis. A few fibres showed central core and 'moth eaten' change but there were no other features of mitochondrial dysfunction. Recent biopsy specimens of rectus abdominis show no histological abnormality.

#### STOMACH AND SMALL BOWEL

The initial jejunal biopsy specimen showed normal mucosa, muscularis muscosae and submucosa with no evidence of inflammation or villous architectural distortion. The muscularis propria, however, was severely abnormal, with changes predominantly in the inner circular layer (Fig 3). The muscle layer was thinner than normal and the remaining fibres arranged haphazardly, with focal severe atrophy and cytoplasmic vacuolation. Some fibres were swollen with abundant deeply eosinophilic cytoplasm and large hyperchromatic nuclei, indicating continuing degeneration. Reticulin and trichrome staining confirmed interstitial fibrosis. Ganglion cells of the submucosal and myenteric plexuses were morphologically normal and cholinesterase staining of cryostat sections showed normal innervation. There was no evidence of intraneuronal storage disease, mucosal enzyme deficiency, or vasculitis. Electron microscopy confirmed vacuolation of muscle fibres and non-specific degenerative features including mitochondrial swelling and filament disruption. Changes were similar to those described by Mitros *et al.*<sup>6</sup>

Examination of biopsy specimens obtained at the second laparotomy showed that the duodenum was most severely involved with abnormalities of the muscularis propria as described above, diffuse atrophy of the inner layer (reduced to approximately one third normal thickness), and segmental loss of the outer layer with replacement by fibrous tissue. The jejunum showed milder muscle damage. The ileum showed occasional vacuolated muscle fibres but no overt atrophy. In the stomach there was mild interstitial fibrosis of the muscularis propria.

#### Discussion

The diagnosis and management of visceral myopathy in our patient was complicated by his coincident insulin dependent diabetes mellitus, which may cause gastrointestinal symptoms.<sup>7</sup> Temporary gastric stasis sometimes occurs in diabetic ketoacidosis due to a functional autonomic neuropathy, although this was never a serious possibility since our patient was not acidotic. Gastroparesis usually occurs in long-standing diabetes in conjunction with other severe complications, although it can occasionally be an isolated finding; it was considered here but excluded by the absence of other diabetic complications, normal autonomic function tests, and normal gastric motility. A relation between chronic intestinal pseudo-obstruction and the previous polymyositis in our patient seems unlikely because of the 16 year gap, although there are poorly documented reports of such an association.<sup>8</sup>

The management of a case of chronic intestinal pseudo-obstruction has three stages: (i) making a definitive diagnosis, (ii) determining the extent of the disease, and (iii) planning a strategy to relieve symptoms and supply adequate nutrition. Against this must be weighed the need to minimise the number of operations, particularly as repeated surgery carries the risk of inducing intestinal adhesions, symptoms of which may mimic those of chronic intestinal pseudo-obstruction.

Characteristic motor patterns can be recorded during manometry in chronic intestinal pseudo-obstruction<sup>9</sup> but only if the probe can be introduced into the abnormal gut which may be impossible when intestinal motility is poor. An initial laparotomy is usually necessary to exclude other lesions and to take full thickness biopsy specimens of the small bowel to establish the diagnosis histologically since it is generally not possible to determine the extent of the disease on inspection alone. In our case, and in another report,<sup>1</sup> there was a gradient of pathology with the disease being most severe in the proximal small bowel. In such circumstances duodenal or jejunal bypass or excision may be followed by several years of a good quality of life provided that the immobile bowel is excluded.<sup>1</sup> We adopted a different approach by taking multiple biopsies and inserting a feeding jejunostomy, thereby allowing us to both assess the extent of the disease histologically and also test the function of the distal small bowel without further surgery. Simultaneous insertion of a drainage gastrostomy covered the possibility that the gastric residue was due to gastroparesis rather than to duodenal distmotility. Once we knew that drainage from the gastrostomy was minimal, this was removed.

Duodenoplasty was performed to reduce duodenal stasis and hence the possibility of small bowel bacterial overgrowth, although we recognise that the effect of such a procedure will be limited by progression of the disease which tends to lead to further dilatation. The longterm outlook is uncertain but we hope to have postponed the need for total parenteral nutrition with its attendant hazards and expense. A major advantage of a trial feeding jejunostomy is that it

does not close any of the other surgical options. Later, if the disease progresses the tube can be resited more distally and proximal bowel excised. Subtotal enterectomy and total parenteral nutrition remain a likely eventual outcome but at present the quality of life is reasonable and nutritional state preserved.

Surgery was expertly performed by Mr J Bourke. We thank Dr S P Allison for his advice on nutritional management.

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