Unclassified oesophageal motor disorders simulating achalasia

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Achalasia of the oesophagus is characterized by the absence of peristalsis, failure of the inferior oesophageal sphincter to relax after swallowing (Harris, 1966), and a positive mecholyl test (Kramer and Ingelfinger, 1951). This combination of features is regarded as specific for this disease. Nevertheless, in the literature a number of cases have been designated achalasia despite the presence of some peristalsis or inferior sphincter relaxation (Vantrappen, van Goïdsehoven, Verbeke, van den Berghe, and Vandenbroucke, 1963) or even a negative mecholyl test (Hightower, Olsen, and Moersch, 1954). Therefore, absolute diagnostic criteria for the disease seem to be incompletely defined.

We have studied six patients with severe oesophageal motor abnormalities referred to us with the clinical diagnosis of achalasia. The oesophageal motility findings in these patients were, however, not characteristic for achalasia; moreover, they were unlike any other recognized oesophageal motor disturbance.

Because these patients clinically simulate achalasia but demonstrate some preservation of normal oesophageal motor function, it is possible that they represent variations of true achalasia.

PATIENTS

Five of the patients were males, one was a female. Ages ranged from 23 to 76 years. Standard barium oesophagrams in these patients showed the features of achalasia: oesophageal dilatation, barium retention, and distal oesophageal narrowing in all (Fig. 1). These radiographs and the presence of dysphagia had led to the clinical diagnosis of achalasia in each patient.

CASE 1

N.D., a 38-year-old construction worker, initially noted difficulty in swallowing in 1962. One year later he experienced severe anterior chest pain associated with a ‘catching sensation in the middle of the chest’. He was conservatively treated for ‘obstruction of the oesophagus’. The return of substernal pain and progressive

FIG. 1. Oesophagrams of the six patients demonstrating marked oesophageal dilatation, barium retention, and distal oesophageal narrowing.
dysphagia led to a loss of 30 lb in weight in January 1964 and subsequent admission to hospital. Oesophagrams showed a 'markedly dilated oesophagus with barium retention for 15 minutes', and a cineoesophagogram supported a diagnosis of achalasia. Oesophagoscopy showed a dilated oesophagus with mild distal oesophagitis but the instrument 'passed with ease into the stomach'. After dilatation of the distal oesophagus with a pneumatic bag, the patient became asymptomatic, gained 15 lb, and the oesophageal diameter decreased. The symptoms returned three months later. An oesophagram performed in 1965 again revealed marked dilatation.

CASE 2 H.H., a 69-year-old retired security guard, related a history of dysphagia, intermittent pyrosis, postprandial vomiting, and 'gurgling in the chest' upon recumbency, dating back to 1925. Oesophageal bouginage had been ineffective on several occasions during the following two decades. Dysphagia was frequently aggra- vated by excitement and relieved by a Valsalva manoeuvre. There was no history of weight loss or pneumonia. In 1961 the patient's oesophageal disease was clinically diagnosed as achalasia. Oesophagoscopy at that time disclosed a 'markedly dilated oesophagus containing a half-gallon of retained food' and cineoesophagrams were 'consistent with achalasia'.

CASE 3 S.S., a 69-year-old retired foundry worker, complained of vomiting without nausea immediately following meals for 18 months. After emesis he could successfully resume eating. Dysphagia was experienced at intervals during this time. He had lost 10 lb in weight. Neither pyrosis nor chest pain was present. Cineoesophagrams revealed a dilated oesophagus and a marked delay in emptying the barium contents. A clinical diagnosis of achalasia was made.

CASE 4 M.P., a 61-year-old housekeeper, experienced haematemesis in 1958. Radiographs suggested the presence of a hiatus hernia which prompted surgical division of the left phrenic nerve. Occasional substernal burning and vomiting were experienced afterwards, and, early in 1965 dysphagia was first noted. Oesophagrams showed moderate dilatation of the oesophagus, narrowing of the distal segment, and a sliding hiatal hernia. At thoracotomy the hernia was repaired but an oesophageal stricture could not be found. Follow-up radiographs again disclosed 'widening of the proximal oesophagus and distal oesophageal narrowing'. The hernia had recurred. Oesophagoscopy and mucosal biopsy performed at this time were considered normal. Recurrence of dysphagia with radiological evidence of delayed oesophageal emptying and continued distal oesophageal narrowing prompted a repeat operation in October 1965. 'Angulation of the distal oesophagus' was described but narrowing or stricture was not present. A subdiaphragmatic vagotomy and hiatal hernia repair were performed. Dysphagia recurred within three months. A cinefluoroscopic study of the oesophagus again showed distal narrowing, dilatation of the body of the oesophagus, and marked barium retention. The clinical diagnosis of achalasia was made. Oesophagomyotomy was performed in March 1966. Again, distal oesophageal stenosis or stricture was not present. Full-thickness excisional biopsy of the distal oesophageal wall showed normal mucosal and muscular elements, but complete absence of ganglion cells in serial microscopic sections.

CASE 5 L.J., a 64-year-old Negro labourer, experienced the onset of singultus, frequent regurgitation of food, and dysphagia in May 1953. Surgical interruption of the left phrenic nerve performed the following year failed to relieve these symptoms. A bleeding gastric ulcer led to subtotal gastric resection without vagotomy in 1957. Continued 'hiccups and vomiting' resulted in further x-ray examination of the oesophagus that year and 'no evidence of cardiospasm' was reported. A history of past alcoholism was obtained in April 1960. Because of continued symptoms, the gastrojejunostomy was revised to a gastroduodenostomy in December 1960. A vagotomy was not performed. Symptoms recurred, however, and continued for the next four years. Oesophagrams in 1964 were considered 'normal'.

An episode of acute pancreatitis resulted in the patient's admission to hospital in 1965; he continued to complain of dysphagia. Oesophagrams at that time disclosed a 'dilated oesophagus with barium retention throughout—compatible with achalasia'. Cinefluorographic examination of the oesophagus showed atony, dilatation, and barium retention.

CASE 6 E.F., a 23-year-old schoolteacher, noticed dysphagia 15 months before admission to hospital in 1967. The dysphagia was located at the suprasternal notch and was produced by solids and occasionally by liquids, although the patient felt his distress was sometimes helped with water ingestion. Occasional relief was obtained by induced vomiting. Two months before admission, the patient experienced retrosternal discomfort in the morning before breakfast. At night, while supine, he frequently regurgitated food into his throat and mouth. Symptoms were aggravated by nervousness, rapid eating, or ingestion of alcoholic beverages. He had lost 5 lb in weight during the last six weeks. Oesophagrams showed a markedly dilated oesophagus with distal narrowing.

OESOPHAGEAL MOTILITY STUDIES

Manometric studies of oesophageal motility were performed utilizing a triple-lumen, water-filled, polyvinyl tube. The three distal recording tips of 1.8 mm internal diameter opened at 4 cm intervals. Pressure changes occurring at the distal tips were transmitted to Sanborn pressure transducers and recorded on a Sanborn multi-channel direct-writing instrument. Oesophageal distension was produced by air injected into a rubber balloon of 20 ml capacity attached to a separate polyvinyl tube. Radiopaque markers adjacent to the balloon and recording tips permitted fluoroscopic localization within the oesophagus. Swallowing and respiration were registered on a fourth channel from pneumographic belts placed around the patient's neck and chest, respectively.

The pressure recording tube was introduced through
the nose into the stomach. With the patient supine, it was then withdrawn in 0.5 to 1.0 cm increments. The patient was asked to swallow at intervals. Most swallows were dry; occasionally, water was administered. The balloon, introduced through the mouth, was positioned proximal to all recording tips, but distal to the aortic arch, and the oesophageal motor response to brief distension was recorded (balloon volume: 5 to 20 ml; duration of distension: 3 seconds).

All pressures were recorded with atmospheric pressure as baseline; resting pressures were measured in the end-expiratory phase. The manometric characteristics of oesophageal motor function were evaluated according to previously described criteria (Ingelfinger, 1958).

The resting pressure in the distal half of the oesophagus was recorded before and after subcutaneous administration of acetyl-beta-methylcholine chloride (Mecholyl). In addition, the response to Mecholyl was recorded by balloon kymography either simultaneously or during a separate study. A condom balloon of 30 ml capacity was positioned in the distal half of the oesophagus and allowed to fill with air isobarically. Oesophageal contraction could freely empty the balloon of air; the volume of air in the balloon was recorded on a kymograph. Mecholyl was administered first in a dose of 5 mg. If the baseline pressure increased no more than 10 mm Hg within the next 10 minutes, the result was considered negative (Nagler and Spiro, 1961), and a second dose of 10 mg of Mecholyl was administered. During balloon kymography, the second dose was given if the balloon had not been completely emptied of air by a prolonged contraction. The batch of Mecholyl used was known to produce a positive oesophageal response in patients with classical achalasia and produced sweating, flushing, tachycardia, and profuse salivation in all patients studied.

and H.H. Patients M.P. (surgical vagotomy) and L.J. demonstrated no increase in gastric acid production following insulin hypoglycaemia.

Distal oesophageal stenosis or stricture was not found in any case. Oesophageal mucosal biopsy in patients N.D. and M.P. showed mild inflammation. Serial sections of a full-thickness oesophageal biopsy from patient M.P. failed to reveal any evidence of ganglion cells.

**OESOPHAGEAL MOTILITY STUDIES** Four of the patients demonstrated primary peristalsis in the oesophagus at some time during the motility study (Fig. 2). The incidence was markedly reduced in the distal two-thirds in all, however, and in only two of the patients were the progressive contraction waves noted to pass through the distal oesophagus. Accordingly, the incidence of nonpropulsive contraction or of no motor response to swallowing was high. Nonpropulsive, repetitive high-pressure waves of the type seen in diffuse spasm were seen in only one patient (S.S.), but these occurred inconsistently and only after a liquid bolus. Two patients (N.D., E.F.) elicited a contraction of some type after every swallow. In one case (N.D.) peristalsis was registered only in the proximal and middle thirds of the oesophagus, and in the other (E.F.) no peristalsis was recorded. An inferior oesophageal high-pressure zone was present in all cases (Fig. 3). The resting pressure of the inferior oesophageal sphincter was

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**RESULTS**

**CLINICAL AND LABORATORY INVESTIGATIONS** There was no history of exposure to toxins or drugs known to affect the oesophagus. None of the patients had travelled or lived in areas endemic for Chagas’ disease.

No evidence of collagen disease was found in the history or on physical examination, nor was Raynaud’s phenomenon present. Skin biopsy of the dorsum of the hand was normal in the five cases examined.

Peripheral neuropathy, nerve paralysis, or muscle disease were not present. SGOT and SGPT levels were normal; the electrocardiograms disclosed no cardiac arrhythmias. Two patients (H.H. and S.S.) had remote histories of moderate ethanol intake, and one patient (L.J.) had been treated for acute and chronic alcoholism in the past and during the period of observation. Patient H.H. exhibited a diabetic oral glucose tolerance test curve.

Hollander tests, performed in four of the patients, showed normal secretory responses in patients N.D.

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![Figure 2](http://gut.bmj.com/)

**FIG. 2.** The oesophageal motor response following deglutition recorded in the proximal (P), middle (M), and distal (D) third of the oesophagus of each patient is represented by a vertical bar. The percentage of swallows eliciting primary peristalsis (black), non-propulsive contractions (stippled) and no motor response (open), are shown. The number of recorded swallows is listed beneath each bar. Normally, 90% or more of swallows result in a primary peristaltic wave (Nagler and Spiro, 1961).
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FIG. 3. Inferior oesophageal sphincter. (a) The resting pressure (mm Hg) of the inferior oesophageal sphincter was normal in all but one case (E.F.) where consistently high inferior oesophageal sphincter pressure was demonstrated (35 mm Hg). The shaded area represents normal range (Nagler and Spiro, 1961; Pert et al, 1959). (b) The incidence of relaxation of the inferior oesophageal sphincter following deglutition was decreased in all patients. No inferior oesophageal sphincter relaxation was demonstrated in patient M.P. The normal incidence of relaxation is from 90 to 100% (Siegel and Hendrix, 1961).

FIG. 4. Oesophageal motility tracing of patient E.F. showing elevation of resting inferior oesophageal sphincter pressure with normal relaxation after deglutition (DS). Distances are measured from the teeth.

FIG. 5. Abnormal delay (five seconds) in relaxation of the inferior oesophageal sphincter of patient N.D. following deglutition.

FIG. 6. Balloon distension of the oesophagus. (a) Secondary peristalsis following balloon distension of the oesophagus was not demonstrated in four of the patients, and the incidence was less than normal (Siegel and Hendrix, 1961; Fleshler et al, 1959) in the other two (H.H., S.S.). (b) Relaxation of the inferior oesophageal sphincter following balloon distension was present in five patients. A normal incidence (Ingelfinger, 1958; Fleshler et al, 1959), was found in only one patient (E.F.)
normal in five patients (4 to 20 mm Hg pressure above that of the stomach) but was elevated in one patient (E.F.) to 35 mm Hg. In all but one (M.P.) the resting pressure of this zone decreased appropriately following deglutition, although the frequency of this response was decreased. An example of raised pressure in the inferior oesophageal sphincter with normal relaxation after swallowing in patient E.F. is shown in Figure 4.

One patient (N.D.) exhibited marked delay in relaxation of the inferior oesophageal sphincter following deglutition. In some instances relaxation was delayed as long as five seconds after swallowing, compared with one or two seconds in normals (Ingelfinger, 1958) (Fig. 5). This finding is similar to that reported by Beck, Hernandez, and Solymar (1966) in a patient who otherwise demonstrated the manometric features of achalasia.

The superior oesophageal sphincter demonstrated normal function in all patients.

Balloon distension elicited secondary peristalsis in two patients and relaxation of the inferior oesophageal sphincter in five (Fig. 6). Non-propulsive contractions occurred after approximately one third of balloon distensions in five patients and was a consistent finding in one patient (E.F.).

The Mecholyl test was positive in three patients, H.H., S.S., and E.F. In each instance the positive response to Mecholyl was recorded as a sustained increase in oesophageal resting pressure of over 35 mm Hg and as complete expulsion of air from the balloon (Fig. 7).

Repeat motility studies were carried out in four patients. The results obtained from separate studies in patient H.H. were strikingly different. During the first study no peristalsis, no inferior oesophageal sphincter, and a negative Mecholyl test were observed. Non-propulsive contractions occurred following one third of the swallows or balloon distensions. The second study performed four months later, although abnormal, demonstrated some primary and secondary peristalsis and an inferior oesophageal sphincter which sometimes responded to deglutition or balloon distension. The Mecholyl test on this occasion was unequivocally positive. No change in symptoms or medication had occurred in that interval. Repeat motility studies in three other patients demonstrated no significant changes from the initial study.

**DISCUSSION**

Classification of the oesophageal motor disturbances in these patients is difficult since they are unlike those typically seen in other recognized oesophageal diseases.

It is clear that none of our patients fulfil all the criteria for the diagnosis of achalasia. Prior to motility studies each of the patients had the presumptive clinical diagnosis of achalasia based on symptoms of dysphagia, the radiographic demonstration of oesophageal dilatation and atony, and the absence of stricture. During motility study, however, oesophageal peristalsis or relaxation of the inferior oesophageal sphincter was demonstrated in all patients. Response to Mecholyl stimulation was negative in three patients, but positive in three others. All patients with a positive response to Mecholyl exhibited relaxation of the inferior oesophageal sphincter and two showed peristalsis.

An achalasia-like syndrome has been reported in Chagas' disease (Castro and Grossi, 1963). None of our patients had visited areas of the world endemic for *Trypanosoma cruzi*.

Oesophageal scleroderma may be manifested by aperistalsis of the distal two-thirds of the oesophagus, loss of function of the inferior oesophageal sphincter, and failure of response to Mecholyl (Code, Creamer, and Schlegel, 1958). While oesophageal involvement may produce the earliest manifestation of scleroderma, in almost all cases with demonstrable motor abnormality a history of Raynaud's disease is present (Atkinson and Summerling, 1966). None of our patients had clinical features suggestive of any collagen disorder, nor was Raynaud's phenomenon
present. Skin biopsy was normal in the five patients examined. All had negative tests for rheumatoid and antinuclear factors and serological tests for syphilis were negative.

None of our patients exhibited neurological or neuromuscular abnormalities. One patient (L.J.) was a chronic alcoholic. The effect of alcohol alone on oesophageal motor function is unknown, although Fischer, Ellison, Thayer, Spiro, and Glaser (1965) have reported two patients with alcoholism with peripheral neuropathy who had diminished to absent oesophageal peristalsis. We have observed the same phenomenon in 10 subjects with chronic alcoholism and peripheral neuropathy (Hogan, Zboralske, and Winship, 1967). The patient in the present study, however, had no evidence of peripheral neuropathy. His oesophageal motor function was characterized by complete aperistalsis and diminution in frequency of relaxation of the inferior oesophageal sphincter.

The role of vagal denervation in the pathogenesis of human oesophageal disease remains to be established. Vagal nerve degeneration has been reported in electron microscopic studies in patients with achalasia (Cassella, Brown, Sayre, and Ellis, 1964), and diffuse oesophageal spasm (Cassella, Ellis, and Brown, 1965), and distal vagotomy in man may temporarily produce dysphagia, atony of the distal oesophagus, and lack of relaxation of the inferior oesophageal sphincter (Anderson, Schlegel, and Olsen, 1966). One of our patients (M.P.) had an infra-diaphragmatic vagotomy several years after the initial complaint of dysphagia. The entire oesophagus was dilated, however, and microscopic examination of the serial sections of a full-thickness surgical biopsy of the distal oesophagus disclosed no ganglion cells. A Hollander test was negative in this patient and in patient L.D. Two other patients, N.D. and H.H., each had a normal gastric secretory response to insulin. This test has been reported to produce a negative result in eight of 32 patients with achalasia (Woolam, Maher, and Ellis, 1967).

The oesophageal motor abnormality associated with aging simulates diffuse oesophageal spasm (Soergel, Zboralske, and Amberg, 1964). Three of our patients were in the seventh decade but had motility disturbances mimicking achalasia, not diffuse spasm. Ages at onset of oesophageal symptoms in our group makes the diagnosis of presbyoesophagus a reasonable possibility in only one (S.S.). This patient exhibited a positive Mecholyl test, however, which was not observed in the study of oesophageal abnormalities due to aging.

The manometric recording in diffuse oesophageal spasm is characterized by non-propulsive, repetitive contractions of high amplitude occurring either spontaneously or after deglutition. The inferior oesophageal sphincter mechanism is usually unaffected (Fleshler, 1957). Patient S.S. exhibited occasional manometric features of diffuse oesophageal spasm although his only symptom referable to the oesophagus was dysphagia. Despite the occurrence of repetitive, non-propulsive contractions on oesophageal pressure tracings, he showed only marked oesophageal dilatation and atony on a cineoesophagram.

A positive oesophageal response to Mecholyl remains the confirming diagnostic test in true achalasia (Kramer and Ingelfinger, 1951). A tetanic muscular contraction simultaneously involving the distal half to two-thirds of the oesophagus occurs following the subcutaneous administration of Mecholyl to patients with this disease. This response has been interpreted as indicating loss of cholinergic innervation of the oesophagus (Kramer and Ingelfinger, 1949). A positive Mecholyl test, however, has been reported in patients with diffuse oesophageal spasm (Kramer, Fleshler, McNally, and Harris, 1967), Chagas' disease (Castro and Grossi, 1963), in one patient with oesophageal carcinoma (Ingelfinger, 1964), and in a patient with marked delay in inferior oesophageal sphincter relaxation and aperistalsis (Beck et al, 1966). Slight increases in oesophageal resting pressure may even occur in normal young subjects. Nagler and Spiro (1961) demonstrated increases in baseline pressure of from 2 to 10 mm Hg following Mecholyl administration in three of 10 such subjects. On the other hand, a negative Mecholyl test has been reported in patients with achalasia (van Goidenhoven, Vantrappen, Verbeke, and Vandenbroucke, 1963), and an insignificant response to Mecholyl stimulation has been noted in four patients with achalasia and a 'markedly dilated' oesophagus (Hightower et al, 1954).

We are making no attempt to classify these six patients into one category of oesophageal disease. Rather, we wish to emphasize that a variety of severe oesophageal motor disorders simulating achalasia may occur and may be difficult clinically to differentiate from true achalasia. These patients may, however, all represent examples of an oesophageal disease the full expression of which is true achalasia.

**SUMMARY**

Dysphagia, oesophageal dilatation, and profound oesophageal motor dysfunction led to the clinical diagnosis of achalasia in six patients. This diagnosis could not be substantiated, but appropriate studies also excluded other oesophageal diseases, such as organic stricture, collagen disease, generalized neuromuscular disorders, diffuse spasm, and presbyoesophagus. Oesophageal motility studies demon-
strated primary peristalsis and/or inferior sphincter relaxation following deglutition in all patients. While these phenomena were not persistently absent (achalasia), the frequency of their occurrence was much less than normal.

Intraluminal balloon distension evoked secondary peristalsis in two patients and spincteric relaxation in five. A positive Mecholyl test was elicited in three patients. One patient, with a negative Mecholyl test, had no demonstrable ganglion cells on full-thickness distal oesophageal biopsy.

Since these patients clinically resemble those with achalasia, but exhibit preservation of some features of normal oesophageal motor function, it is possible that some or all of them represent incomplete forms of true achalasia.

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REFERENCES


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