Achalasia of the Cardia: Reflections upon a Clinical Study of Over 100 Cases

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Adams *et al.* (1961) have stated that the four cardinal changes that typify achalasia are (1) a failure of the cardia to relax, (2) the disappearance of normal oesophageal peristalsis and its replacement by segmental and tertiary contractions, (3) dilatation and hypertrophy of the oesophagus above the cardia, and (4) an increased sensitivity of the oesophagus to cholinergic drugs. They believe that these changes are due to degeneration or absence of the ganglion cells in Meissner's and Auerbach's plexuses. And the cause of these neurological faults is unknown.

There are other conditions in which peristalsis disappears from the oesophagus; but these are generally due to changes in the muscle coats—for example, dermatomyositis—and not to nerve palsies. Achalasia of the cardia has also been compared to Hirschsprung's disease in the rectum, and to Chagas's disease (Brazilian trypanosomiasis) in so far as the latter affects the gullet and the large bowel. These comparisons have not advanced knowledge about achalasia.

In achalasia peristalsis has disappeared by the time the patient consults a doctor: there is no definite prodromal phase of partial paralysis or hypermotility of the gullet. But after peristalsis has gone, uncoordinated muscle movements persist, and these may become accentuated, especially if the gullet has been efficiently drained by operation.

If one is correct in assuming that achalasia of the cardia is due to an absence or a degeneration of the ganglion cells in Meissner's and Auerbach's plexuses it follows that the disease is physiologically incurable. This does not mean that nothing effective can be done to relieve the symptoms. I shall mention a few relevant recent advances in the study of achalasia.

The pharmacological studies of Adams *et al.* (1961) confirmed a number of points that had previously been suspected. They proved that there is a disturbance in the innervation of the whole oesophagus (a point guessed at by Einhorn in 1888) and that ganglion cells are completely absent in the "sac"—that is, the dilated part—and almost completely absent in the "neck"—that is, the narrow segment of the gullet. This also confirmed the work of Lendrum (1937); and it is sufficient explanation for the pathological changes, the signs, the symptoms, and the clinical course of the disease.

Important facts have come to light as a result of the intra-oesophageal pressure recordings and motility studies made by many investigators (notably Code and his associates (Anderson *et al.*, 1958) at the Mayo Clinic). These are that true peristalsis can be differentiated from "uncoordinated" oesophageal movements; that the latter do not pass food onwards through the gullet; that they generally persist after peristalsis has disappeared and may be accentuated because, in the absence of peristalsis, they are more evident radiologically. Uncoordinated

movements may, in the past, have been mistaken for true peristalsis by those who have claimed that treatment of achalasia can often restore the gullet to normal.

Intraoesophageal pressure recordings and motility studies have shown that in achalasia the lower segment of the gullet (the "cardia" or the "neck") does not relax as it should when food or liquid passes through the crico-pharyngeal sphincter. Failure of the "neck" to relax is the cause of the events that characterize achalasia. The fact that the whole oesophagus is aperistaltic is not in itself enough to produce obstruction because it is known (experimentally and clinically) that the gullet can be replaced by a prosthesis, or a graft, and still transmit food, provided that there is no mechanical obstruction; in these artificial circumstances food passes from the pharynx to the stomach as a result of the force imparted to it by the pharyngeal muscles and the effects of gravity. Thus in "achalasia of the cardia" the patient cannot swallow if the pharynx is lower than the stomach, and this is an important radiological test that can be useful in elucidating doubtful cases.

By using cineradiography and the image-intensifier other facts have been amplified. The complexity of the oesophageal movements have been recorded and the fallacy of relying upon a single x-ray film has been underlined. Too often anomalies in size, shape, and contour of the gullet have been diagnosed without evidence that they are persistent, and without regard to the circumstances that may have been responsible for them.

Material

The opinions that follow are based upon clinical and radiological studies of approximately¹ 120 patients who suffered from achalasia of the cardia. Most of these people were under my care and were operated upon by me during the past 30 years, but some of the records of patients operated upon at St. Thomas's Hospital by Mr. Kent Harrison have also been reviewed. About 70 have had serial x-ray examinations over many years, and there are records of the others, made at operation, and a short post-operative follow-up. The cases have been divided into these two groups because I have repeatedly emphasized the fallacy, in all oesophageal work, of opinions on the value of this or that treatment based upon short follow-up—that is, two years or less.

In this series the age and sex incidence was the same as others that have been reported—the disease presented at all ages from birth to old age; but its greatest incidence was between 25 and 60. The youngest patient was 4½ years old and the oldest 84 when they first came for diagnosis (Table I).

¹ In reviewing a series of patients some of whom were seen and treated more than 25 years ago—at a time when the criteria for the diagnosis of achalasia were not definite—the records are often speculative. For this reason the figures quoted in this paper are approximate.

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TABLE I.—*Age and Sex Incidence*

Ages	Men	Women	Total
0-9	0	2	2
10-19	4	3	7
20-29	8	4	12
30-39	12	11	23
40-49	20	9	29
50-59	10	11	21
60-69	6	13	19
70-79	1	4	5
80-89	1	3	4
Total	62	60	122

No patient in this series had neurological lesions elsewhere. All were thought to have true achalasia—as a result of intra-oesophageal motility studies (in doubtful cases), the radiological findings, or the findings at operation. The macroscopic findings at operation—a narrow lower segment (1 to 5 cm. long) of the lower oesophagus, without evidence of peri-oesophagitis or mural inflammation and with a thick circular muscle coat—are typical of the disease.

Age and Sex Incidence.—The proportions of males to females vary. Below the age of 50, males predominate, but after this there are more women than men (Table I).

Onset of Symptoms

Nobody knows how achalasia of the cardia starts nor how long it has been present when the patient first comes for treatment. The disease is unmistakable by the time the patient consults a doctor: it is nearly always fully developed radiologically, and also according to intraoesophageal motility studies.

There is one possible—though as yet unsettled—exception to this statement—namely, that a few patients may present with the condition called “diffuse oesophageal spasm.” This anomaly, which has been described and differentiated from achalasia by Creamer and his colleagues (Creamer and Pierce, 1957; Anderson *et al.*, 1958), is one in which the patient experiences generalized oesophageal pain on swallowing, and radiologically is seen to have useless oesophageal movements (both peristaltic and uncoordinated). The nature and course of “diffuse oesophageal spasm” is unknown, but I have seen two patients who after three and four years changed to achalasia. Thus it has been suggested that achalasia is generally a disease in which the phase of neurological irritation is transient and passes unnoticed, so that the patient presents in the phase of aperistalsis. It may also be a fact that some who have a “corkscrew” oesophagus, affecting the lower third, in which the gullet is actually smaller than normal, are about to become aperistaltic and dilated.

The onset of clinical symptoms is remarkable for its variety; and on looking through these records it soon became obvious that no useful table of “presenting symptoms” could be drawn up, because the disease often starts vaguely and with troubles that seem to be unconnected with the oesophagus. Nevertheless, about two-thirds (or more) of the patients complain of some of the following disabilities: a choking feeling on attempting to swallow, difficulty in swallowing, regurgitation of food and saliva, vomiting, precordial pain, and hunger. But some have no apparent difficulty with swallowing.

Recently the headmaster of a school came under my care: he had had a mass x-ray film taken in which a large “mediastinal tumour” had been found. This was in fact a mega-oesophagus. He did not admit to more than mild occasional attacks of “indigestion.” Another had complained of indigestion for 30 years; and some adults say that they have had intermittent episodes of unaccountable vomiting since they were small children. Achalasia may first be diagnosed after the birth of a baby; and in such cases vomiting, supposedly due to the pregnancy, continues after parturition, and it is then that achalasia is found to be present. Other patients are diagnosed

as being hysterical because they have indigestion of unknown cause; and some give a history that the first symptoms occurred immediately after a fright. Here is a typical story of this type. During the second world war a young man who had never fired an anti-aircraft gun in anger found himself engaged in an air-raid; as soon as the enemy had disappeared he vomited profusely and was taken to hospital; he had a mega-oesophagus at that time. Sometimes a datestone or a piece of apple sticks in the narrow segment of the gullet, and the first intimation is an episode of acute oesophageal obstruction that requires oesophagoscopy for its relief.

Here are some other bizarre episodes that occurred in patients who had not up till then been diagnosed as having achalasia.

An old woman presented with an aerocoele in the left side of the neck: this was the uppermost pouch of a massive oesophagus.

A diplomat dining at the Kremlin had taken somewhat too much vodka. He withdrew and was surprised to vomit several pints of “blood.” Some of this he inhaled and he became ill with bilateral lung consolidations. After the resolution of his “pneumonia” gross achalasia was diagnosed, and was successfully alleviated by Heller’s operation.

A policeman was admitted to hospital with a pyrexia of unknown origin. He had recently returned from Kenya, where all serological tests for tropical diseases had been found to be negative. Achalasia of the cardia was eventually diagnosed by a barium swallow, and his temperature settled with oesophageal washouts and Heller’s operation.

A man who had had slight dysphagia for years came to hospital because he had become so short of breath that he could no longer shout the odds on a racecourse. His dyspnoea was due to long-standing bronchitis and emphysema consequent upon spill-over into the lungs from the contents of a mega-oesophagus.

A child aged 8 was found in a sanatorium allegedly suffering from bilateral pulmonary tuberculosis. This pneumonitis was due to spill-over from an oesophagus distended as a result of achalasia. The diagnosis of tuberculosis had been based upon the finding of bacilli in the sputum: these were in fact atypical mycobacteria that simulated tubercle bacilli (q.v.).

Course of the Disease

There is a lack of information about the course of achalasia, and this is because the moment of onset is unknown, the results of the disease are slow to kill the patient, and there has been no accurate way of assessing the results of treatment or the causes and effects of prolonged oesophageal retention.

The course of the disease is unpredictable (in any individual) as regards length of life, severity of the incapacity, and the complications that may occur. But two things are certain: peristalsis never reverts to normal whatever be done for the patient, and the patient who has achalasia has ultimately a fatal disease. He may not die from achalasia or its complications, because achalasia may take 20 to 50 years to kill, and other things can intervene during this time; but if he lives long enough achalasia will be the cause of death.

Most patients say that in the beginning the symptoms were intermittent. Sometimes there were periods when the disease seemed to have disappeared; periods when swallowing was normal and there were no symptoms. Then, for no apparent reason, dysphagia and regurgitation, pain and hunger, returned. Details of diet played no part in these remissions (though lumps of food tend to stick at any time); but when symptoms have become continuous patients learn that they can never eat certain foods. They also learn that they have an oesophagus, and that food sticks in certain places; they can feel it going through into the stomach, and their observations, checked radiologically, are correct. They become oesophagus-conscious.

In the Guy’s Hospital reports, Adams *et al.*, (1961) described three stages of achalasia. These they said were consecutive and progressive.

Stage I: there is marked painful dysphagia; frequent regurgitation; weight loss; and, radiologically, a small active oesophagus (that looks like a cucumber).

Stage II (the compensated state): the patient feels much better; weight loss and appetite improve; there is scarcely any regurgitation, but the oesophagus has become larger radiologically.

Stage III (the stage of decompensation): the patient has a mega-oesophagus and dies of inanition or from the complications of oesophageal retention, such as carcinoma or ventilatory insufficiency.

This staging suggests that the signs and symptoms of achalasia change as time passes—and this is true. But the sequence of events is not certainly that the oesophagus will be fusiform and irritable at first and grossly dilated and incompetent at the end. A patient who has a cucumber oesophagus (fusiform in shape) may die of the disease without the gullet becoming greatly enlarged, and another who has had a mega-oesophagus for years may be in reasonably "good health."

In the patients I have watched not one oesophagus has changed from cucumber oesophagus to megaesophagus. Each had its characteristic shape *when the patient was first seen*, and each kept that shape until the end. I cannot say that this change from cucumber oesophagus to megaesophagus does not occur, but I have not seen it.

If this observation is correct cucumber oesophagus and mega-oesophagus may be different varieties of achalasia. It is certainly not inevitable that the former precedes, and changes into, the latter in patients who have had dysphagia for a long time.

Approximate Length of History of Symptoms Possibly Attributable to Achalasia (Table II).—It is strange that very few patients who have long-standing achalasia die of starvation. In this series one old woman died in this way: she had a long history of the disease and a megaesophagus; she was admitted in a neglected condition and died before anything could be done to treat the obstruction.

TABLE II.—Length of History of Symptoms

Patients presenting with a Fusiform, or Cucumber, Oesophagus					
Less than 1 year	20	4 years	4	16 years	1
1 year	4	8 "	2	18 "	1
2 years	10	10 "	2	More than 20	
3 "	4	12 "	2	years	3
Patients presenting with a Megaesophagus					
Less than 1 year	4	5 years	4	15 years	1
1 year	4	6 "	6	16 "	1
2 years	6	9 "	1	18 "	1
3 "	1	13 "	4	More than 20 years	6
4 "	1				

Note: The above figures are only approximate, because (1) the history in long-standing cases is nearly always confused—the patient has learned to live with his disease and there are often complicating factors; (2) adequate records and barium studies are not always available; (3) the cases are *not*, in fact, sharply separated into those that have a megaesophagus and those with a fusiform variety; intermediate types occur.

Pathology

Very few specimens of oesophagus taken from patients dead with achalasia are available in medical museums; and the pathological changes that are described are based upon clinical investigations and biopsy specimens. A few points may be mentioned.

Counting ganglion cells in biopsy specimens is not a useful investigation: it gives no indication of how many ganglia are or should be present; and the fact that the oesophagus is dilated means that the ganglia must be more than usually spread out, and thus may appear to be scarcer than normal. And for the same reason, those who believe that the muscle is hypertrophied should recollect that the wall of the oesophagus is often distended to many times its normal size. At operation the circular muscle layer in the narrow lower segment certainly looks to be hypertrophied, but the lumen is narrower here than it should be and the muscle coats are bunched up.

One of the most extraordinary things about achalasia is that the patients do not suffer from serious toxic effects of stagnation of food in the gullet. In a severe case the patient may look

well and have a good complexion, and yet there may be several pints of stale food, mucus, and saliva pent up in the oesophagus. Moreover, most of these patients have little or no inflammation in the mucosa even though the disease has been present for a long time; nor is mediastinal adenitis a feature as it is with benign peptic oesophagitis. On the other hand, pyrexia may be a feature of this disease, and if it is the oesophagus may be inflamed. Such cases respond very well to pre-operative wash-outs.

Tumours occur more commonly than usual in the oesophagus of patients suffering from achalasia. Here is an example of a benign lesion.

A man in his fifties first consulted his doctor in 1938 and was diagnosed radiologically as having typical achalasia of the cardia. He was 6 ft. 5 in. (195 cm.) in height and weighed 17 st. (108 kg.). Treatment by bouginage and amyl nitrite was ineffective and he lost so much weight that a gastrectomy was done and maintained for two years, by which time he weighed about 8 st. (50.8 kg.). During the war he lived on milk alone, and he suffered from many complications, including bronchitis, effusions into his joints, and hypertrophic pulmonary osteoarthropathy. I saw him in 1956—that is, almost 20 years after the onset of symptoms—he was bedridden and a complete invalid. He could swallow nothing without coughing, and the radiographs of his chest showed a lobulated opacity that almost completely filled the right hemithorax. The nature of the opacity was uncertain because the films did not look like a megaesophagus, and the original films showed that the oesophagus had been cucumber in shape. This man died, and at necropsy there was a leiomyoma the size of a big grape-fruit arising in the middle of the oesophagus and two more, the size of small oranges, arising from a point 2½ in. (6.3 cm.) below the cricopharyngeus. The specimen is preserved in the museum of the Royal College of Surgeons. Below the tumours the changes were typical of achalasia, and this oesophagus was still fusiform in shape (cucumber) and not a mega-oesophagus. In retrospect, the original barium swallow in 1938 shows that the largest of the leiomyomata was present at that time and was then about the size of an orange. To-day this man would almost certainly be diagnosed and relieved of his tumours and his symptoms.

Malignant tumours occur in the dilated "sac" of achalasia; they do not arise in the narrow "neck." In the Guy's Hospital series (Adams *et al.*, 1961) carcinoma occurred in 29% of those in whom the cause of death was known; the growths were all in patients who had had achalasia for many years, and they were old when they died. The same was true in the patients who had carcinoma in this series. The growths were large squamous-cell ulcers, situated usually in the middle third of the gullet (but not confined to this position), and they occurred more commonly in megaesophagus than in the cucumber variety. They were inoperable by the time a diagnosis was made. Dysphagia was not a symptom, because the oesophageal lumen was bigger than normal: the patients complained of pain and vomiting of bloody saliva; there was also a characteristic offensive smell and taste in the mouth. The incidence of malignant ulceration superimposed upon achalasia in this group was that seven patients are known to have died of cancer. Others have probably died of carcinoma, and some who are still alive will do so in the future. It would be convenient to be able to state that Heller's operation eliminates the risk that cancer will appear later: this is not true in these patients, several of whom had had "successful" myotomies in the past.

Pulmonary complications, ranging from transient episodes of pneumonitis to pneumonia, abscess, etc., are well known and are due to small amounts of oesophageal contents spilling over from the gullet into the air-passage when the patient lies down. One group of patients are especially interesting. They are the people who have atypical mycobacteria in the obstructed gullet. These organisms are acid- and alcohol-fast and can resemble tubercle bacilli microscopically. In the ordinary course of events these organisms are non-pathogenic; but when coated with certain oils (notably liquid paraffin and butter or milk fats) they can cause lesions in the lungs that simulate pulmonary tuberculosis, radiologically and clinically, and organisms

resembling tubercle bacilli may be found in the sputum. Patients who have achalasia are likely to have small amounts of these oils in the oesophagus, either because they take them in their diets or because the paraffin has been used to lubricate bougies: the oil floats to the top of the column of retained food and liquid in the gullet. Thus the mycobacteria become coated with oil and thence they behave as tubercle bacilli after inhalation into the lungs.

The lesions in the lungs may resemble recent infiltrates of tuberculosis (as in the child already mentioned) or they may behave like chronic phthisis with fibrosis and excavation, as was found in a man aged 60 who had been treated for apical tuberculosis for 15 years and who was ultimately found to have achalasia and atypical mycobacteria. The state of his lung was much improved by Heller's operation.

Thus it is essential to verify the existence of *true* pulmonary tuberculosis in any patient who has achalasia and acid/alcohol-fast bacilli in the sputum. The atypical mycobacteria differ from tubercle bacilli in that they have different cultural behaviours, they do not react to anti-tuberculous drugs, and they are not infectious to other persons.

Apart from atypical mycobacteria there are other saprophytes, such as streptothrixes that exist in food held up in the gullet, and these may also become pathogenic when inhaled into the lungs (*Nocardia asteroides*). Patients who have such pulmonary complications often do not know that they have been caused by achalasia; and even if they do know this, they do not realize that inhalation of small amounts from the oesophagus is going on all the time.

Radiology

Apart from the well-known radiological features of achalasia, two points are important: (1) no patient suffering from achalasia can swallow in the head-down position—this is because there is no peristalsis in the gullet; and (2) the normal gas bubble in the fundus of the stomach is generally absent.

The appearances of achalasia—that is, those of cucumber oesophagus—can sometimes be produced by carcinomata on the lesser curve of the stomach, and by some carcinomata (of the leather-bottle type) that affect the cardia and produce a smooth stricture. Thus it behoves a surgeon who operates for achalasia, and who finds no abnormality in the lower inch of the gullet, to open the abdomen and examine the stomach.

Physiological obstruction resembling achalasia and situated in the lower inch of the oesophagus also occurs after some vagotomies and partial gastrectomies (in which the vagi have been sectioned). This clears up spontaneously within a few weeks and requires no treatment.

The severity of the clinical symptoms bears little or no relation either to changes that the radiologist finds when he does a barium-meal examination or to the length of the clinical history. Sometimes it happens that the radiological findings are atypical of achalasia, but the changes of achalasia at thoracotomy are unmistakable.

Some patients who have a megaesophagus are at first diagnosed as having one of the following:

1. A "tumour" in the lower mediastinum. This is because the oesophagus at the lower end and immediately above the narrow segment doubles back upon itself before passing through the hiatus, and the overlap of shadows produces the appearance of a "tumour" behind the heart.
2. A mediastinal abscess, or a diaphragmatic hernia. These diagnoses are made because in a megaesophagus, of whose existence the patient and the radiologist may be unaware, fluid levels may be seen in the mediastinum. The liquid contents of the oesophagus in a patient who has achalasia do not all gravitate to the lower, obstructed segment; some are held in the redundant curves of the dilated gullet.
3. If no previous diagnosis of achalasia has been made the wall of the dilated oesophagus, impinging upon the right upper lung field,

may simulate an azygos lobe. The distinction is that the linear shadow cast by the wall of the distended (and empty) oesophagus is wider than the hair-like line of an azygos fissure.

There are some general purposes for which radiology is occasionally helpful in these cases: (1) if a patient is being treated by self-bouginage it is important to verify that the bougie actually enters the stomach; (2) to verify the position of a hydrostatic bag before distending it; (3) to demonstrate abnormal movements in the gullet and to synchronize these with episodes of pain of which they may be the cause; and (4) to pick up complications such as leiomyoma and carcinoma of the oesophagus, and to indicate the presence of pulmonary complications.

Radiographs should include the whole length of the gullet, as it is not unusual for complications at the upper end, such as carcinoma, to be missed because the plate used has been too small.

Treatment

Achalasia never resolves spontaneously after peristalsis has disappeared, but the symptoms can generally be dramatically relieved by Heller's operation.

Lavage of the oesophagus is essential in patients who have a considerable retention and pyrexia. It is almost impossible to do this with an ordinary stomach-tube (because the tube gets blocked and siphonage or suction fails), and most surgeons advocate lavage by oesophagoscopy. In this series lavage immediately before operation was never done; it is unnecessary.

There is only one operation that affords the patients relief of symptoms without "undue" risk of reflux (Barrett and Franklin, 1949). The word "undue" is used because some patients get oesophagitis after Heller's operation. This operation achieves its best results in young people who have a short history and an oesophagus shaped like a cucumber. The results in megaesophagus are not so good. Heller's operation should be done through the chest, because only in this way can the surgeon be certain of dividing all the circular muscle-bands over the whole length of the lower narrow segment (the "neck") of the gullet.

At operation three points need consideration. (1) The myotomy must extend on to the stomach below and on to the dilated lower part of the gullet above. (2) *All* the circular muscle fibres must be divided throughout the whole length of the myotomy. Sometimes, in long-standing cases, there are fibrous strands in the layer of the muscularis mucosae. These must be divided, because if left intact they can prejudice the result of the operation. (3) The surgeon must consider the possibility that this operation may produce an incompetent cardia, reflux, and oesophagitis. I believe this series of complications to be rare, provided the peritoneal attachments that ensheath the gullet as it passes through the hiatus are not disturbed. It is unnecessary to reduce the size of the hiatus by stitches, except in the unusual event of a hiatus hernia being present as well as achalasia.

Neither inhalations of amyl nitrite nor bouginage influence the course of the disease, although a few patients believe that they are beneficial. Hydrostatic dilatation differs from bouginage in that the dilating-bag is expanded to a transverse diameter of about 5 cm., and this ruptures the circular muscle fibres in the narrow segment. Anybody who has dilated this segment, and watched the effect upon the wall of the oesophagus when the chest is open, will have misgivings about the safety of the procedure. Nevertheless, there are many records of patients whose symptoms have been relieved by dilatation.

Some criticisms of this method are: (1) the hydrostatic bag cannot always be engaged in the narrow segment—this is particularly true in megaesophagus where the lower part of the gullet is sharply angulated to the left; (2) as the hydrostatic bag is distended it often slips out of the narrow segment into

the distended oesophagus above or, more usually, into the stomach below—there is no way of knowing if this has happened unless the manoeuvre is watched with an image-intensifier; (3) figures from other clinics show that only about two-thirds of the patients (or less) are relieved by a single “successful” dilatation, and the chance of recurrence is higher, and sooner, than after Heller’s operation.

Any open operation that results in incompetence of the cardia and reflux must be accounted a failure, because the symptoms of achalasia will be replaced by those of oesophagitis.

Barrett and Franklin (1949) described the harmful effects of all types of oesophago-gastric bypass procedures done to relieve achalasia and of all types of cardioplasty. Their observations, disputed at the time, are now accepted as correct.

If the exposed mucous membrane be opened inadvertently at operation this does not result in mediastinitis provided the perforation is closed with fine sutures upon atraumatic needles. In this series the mucosa was opened and sutured seven times: there were no harmful results; but one woman aged 83, in whom the mucosa had *not* been opened, died several days after Heller’s operation, from the effects of a subphrenic abscess due to a perforation. Thus in elderly patients, who often have atheroma of the coeliac axis and in consequence have a relatively poor blood supply to the lower oesophagus, there may be a risk of late perforation from ischaemia.

Heller’s operation is more likely to give a good clinical result in patients having a cucumber oesophagus than in those with a voluminous megaesophagus. But as there is no way of knowing, pre-operatively, whether a Heller’s operation will succeed in patients who have a megaesophagus, it was tried as a first expedient. If it failed in a case of megaesophagus it was thought to be unwise to repeat it. Something more radical is then necessary, and in four cases I found that oesophago-jejuno-gastrostomy—after excision of the obstructing lower segment of the gullet—gave a good result. Jejunal grafts heal well, though the dilated lower oesophagus does not appear to be in an ideal condition for anastomosis.

Attempts to alleviate megaesophagus by Heller’s operation combined with fixing the oesophago-gastric junction some inches below the hiatus (thus straightening out the lower redundant loop of the gullet) have failed in my hands; and the operation by which a small fillet of central tendon of the diaphragm, which retains its blood supply and which is sewn into the space created by the myotomy, with a view to keeping the lumen open, has not been done on any of these patients.

Post-operative Findings

A successful Heller’s operation is one in which dysphagia is relieved from the time of operation. These patients state that swallowing feels quite different, that food no longer sticks, and that they can feel it passing through into the stomach. They are grateful for what they consider to be the cure of an asocial disease. They generally put on weight rapidly and engage in activities that they could not contemplate before. For all these reasons Heller’s operation has been claimed to be satisfactory.

It sometimes happens that for some unknown reason the immediate result is not good when expectations seemed to be favourable. There were three such cases in this series and all were operated upon again within three weeks of the first operation. A new myotomy was done and the final results were clinically good.

If after operation the patient’s condition is satisfactory and water can be swallowed without difficulty, there is no need to limit the amount of food or liquid that is taken: a full diet may be safely allowed.

Follow-up

I have heard it said that a patient who has oesophageal obstruction due to achalasia is relatively lucky because the symptoms can be cured so easily. Long acquaintance with some patients who have this disease has suggested to me that the ultimate outlook is not as rosy as many believe; but this is difficult to prove: difficult because the disease kills slowly—perhaps over a span of 20 to 50 years—and no surgeon sees enough cases to enable him to collect significant statistics or to keep in touch with the people who have and those who have not been treated surgically. Moreover, the ultimate assessment of the value of treatment depends upon a series of investigations, done many years after treatment, that could show that the oesophagus had reverted to its normal function. As it is, nobody can tell us whether oesophageal peristalsis ever returns; nor even such essential information as the number of patients who have been relieved of oesophageal obstruction at the expense of having been given an incompetent cardia and reflux oesophagitis. Some surgeons guess that the late incidence of oesophagitis is high, and others proclaim that incompetence of the cardia after Heller’s operation is rare. I do not know.

It is certainly true that the normal oesophagus transmits food from the pharynx to the stomach more efficiently than one that is paralysed. And it is surprising that some patients who have gross oesophageal retention remain in relatively good health for a long time. They may state that they can “eat anything” and are very well, and yet a barium swallow reveals that the oesophageal function has not been greatly changed by treatment.

Those who say that “as long as the patient is satisfied I am satisfied” may be taking too sanguine a view, because there is evidence that prolonged oesophageal obstruction is a pre-cancerous condition, and that spill-over of oesophageal contents into the lungs—as well as crippling complications such as hypertrophic pulmonary osteoarthropathy—occurs especially in patients who have a long history of dysphagia and have become decompensated.

It is probably true that untreated established achalasia of the cardia never undergoes spontaneous cure, although patients often state that during the early phase of their dysphagia the symptoms were intermittent. Heller’s operation does nothing that could remedy the underlying myenteric ganglion paralysis: it merely overcomes (partially or completely) the physiological obstruction at the “neck” of the achalasia, and allows the oesophagus to drain more effectively into the stomach. Thus the oesophagus is never restored to normal function by Heller’s operation; it persists in greater or less degree as an infected sump, and the larger it gets the less the patient experiences dysphagia and the more prone he becomes to other serious complications. In this disease variations in the size of the gullet, as shown by a barium swallow, cannot be taken as a criterion in favour of progress or deterioration; the size depends upon the degree of the paralysis, the position of the patient, the density of the opaque meal, and the normality (or otherwise) of the mediastinal and pleural pressures. After a clinically successful Heller’s operation the oesophagus gets less voluminous; but it also happens that it does not change in size or it continues slowly to get larger.

A post-operative follow-up based on a short post-operative survey, and recording the patient’s own account of his health, is of little interest; and yet it has proved difficult to get more accurate information.

Table III is an attempt to assess the late results of Heller’s operation. It is deficient for many reasons: it has not been possible to trace a consecutive series, the relevant notes and serial radiographs have not always been available, and different radiologists (having various opinions on what to record) have examined the patients.

The cases are here listed in two groups: the first are those who have a fusiform (cucumber-shaped) oesophagus, and the

second a voluminous megaoesophagus. It has already been stated that this distinction is not absolute; but it is a useful approximation. They are classified into three types of result: *group A* means that the patient is well, has no complaints, and the barium studies show little or no oesophageal retention; *group B* are those who feel well and can swallow reasonably but in whom the barium swallow is unsatisfactory; and *group C* are those who have dysphagia, or comparable troubles, and who have complications; it includes patients who have developed carcinoma after Heller's operation and those who have died.

TABLE III.—Results of Follow-up of Some Patients who Had Heller's Operation for Achalasia

Length of History before Operation	Follow-up Time								
	1-2 Years			2-5 Years			5-10+ Years		
	A	B	C	A	B	C	A	B	C
<i>Fusiform (or Cucumber) Type of Gullet</i>									
1-3 years ..	12	3	1	3	1	—	—	2	—
3-6 " ..	4	1	—	1	1	—	—	—	—
6-9 " ..	2	—	—	—	—	—	1	—	—
10+ " ..	2	—	—	—	2	1	—	—	3
<i>Megaoesophagus</i>									
1-3 years ..	6	2	1	—	—	—	1	3	2
3-6 " ..	2	—	—	1	2	—	—	1	—
6-9 " ..	1	1	—	1	1	—	—	1	—
10+ " ..	2	—	—	3	—	1	—	1	2

These figures are too small to allow any conclusions; but two things might be true. The best results are in the fusiform type of oesophagus with a short history, and when the assessment has been made soon after Heller's operation. The worst results are in long-standing cases followed up for a long time.

In the follow-up of these patients certain points have been noted:

1. Although Heller's operation does nothing to restore normal peristalsis, uncoordinated movements of the gullet are often accentuated as the oesophagus drains more efficiently; unless these movements are checked by intraoesophageal pressure recordings they may wrongly be interpreted as peristalsis, and it may then be claimed that the gullet has returned to normal. Uncoordinated movements do nothing to propel the oesophageal contents onwards to the stomach; and they may be painful.

2. After a successful Heller's operation the mucous membrane pouts through the myotomy incision. This can produce the radiological appearances of a hiatus hernia; but this pouting is *above* the "oesophago-gastric sphincter mechanism" and does not produce reflux. Thus before diagnosing a post-operative hiatus hernia reflux must be demonstrated and these two conditions differentiated from each other.

3. Complications of achalasia such as pneumonitis, bronchitis, and emphysema are often dramatically improved by Heller's operation; but they are not cured, because the patients seldom know about the possibility of spill-over and the pathological changes in the lungs are usually fixed by the time an attempt is made to relieve them.

4. Post-operative pain is not uncommon and is of various sorts. It may be due to reflux oesophagitis or uncoordinated oesophageal movements, or it may be due to the incision in the

chest wall. In this series there was one middle-aged man whose dysphagia was completely relieved, but who developed such intractable chest-wall pain that he went from hospital to hospital and had several neurological operations. He has now been completely incapacitated for 20 years.

5. The most serious complication of achalasia is carcinoma. There were seven known cases in this series, but this figure is too low. Carcinoma is prone to develop in patients who have a long history: it occurs in both varieties of achalasia and it followed a clinically successful Heller's operation in two cases in this series.

Summary

The cause and genesis of achalasia of the cardia is unknown. Patients in whom the diagnosis is made practically always have complete aperistalsis when they are first examined: thus the sequence of the antecedent pathological changes is also unknown.

It is arguable that "megaoesophagus" and "fusiform" (cucumber) oesophagus are different varieties of the same disease. It cannot be shown that all patients who have a megaoesophagus have had the disease for a long time, nor that there is a natural progression from fusiform to megaoesophagus. It is not true that all patients who have a megaoesophagus are "decompensated" or that serious complications are imminent for them. It is possible for a fusiform oesophagus to maintain this basic shape for 20 or more years, and carcinoma may develop in this type of gullet as well as in the other.

As Heller's operation does nothing to remedy the underlying neurological defect the gullet does not return to normal, however well the patient may claim to be clinically. The crucial simple test of the presence of peristalsis is whether or not the patient can swallow liquid in the head-down position. As the oesophagus of a patient who has achalasia cannot be restored to physiological normality, it is probable that the late results of this disease are less satisfactory than early post-operative assessments suggest. Some support for this view is given in this paper.

Hardly anybody who has achalasia dies of starvation; they die from the complications of the disease; and because these can take years to become established, intercurrent illness may be the actual cause of death.

Many of the radiological studies on the patients in this series have been done by Dr. J. W. Pierce at St. Thomas's Hospital. To him I give my thanks.

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