

Achalasia:

Results of Operation in 84 Patients

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ACHALASIA of the esophagus is an uncommon neuromuscular disorder originally described by Willis in 1672.¹⁹ Ernst Heller performed the first successful cardiomyotomy for this disease on April 14, 1913.⁴ Zaaier modified Heller's operation in 1923,²¹ using only an anterior myotomy instead of anterior and posterior incisions as done by Heller. The Zaaier modification of the Heller procedure is most often performed today despite its acceptance only recently.⁴

Notwithstanding the general success of this operation there are two complications that detract from good results. One is recurrent obstruction of the esophagus due to achalasia and the other is peptic esophagitis.¹⁰ It is difficult to determine the incidence of these complications.

This paper reports experience in the surgical treatment of 84 cases of achalasia at The New York Hospital-Cornell University Medical Center where the disease has been encountered in more than 150 patients since 1932. Excluding patients treated by

dilatation 84 have undergone one or more surgical procedures for achalasia or its complications.

An aggressive surgical approach to the disease was initiated at our institution in 1944. Between 1944 and 1952, 18 patients underwent various cardioplastic procedures or esophagogastrostomy. Initial results were promising¹³ but the substitution of peptic esophagitis for obstruction due to achalasia made these technics unacceptable. We include in this report late follow-up results in these 18 patients.

In 1951 the Zaaier modification²¹ of the Heller cardiomyotomy was adopted as the procedure of choice for most patients with achalasia.⁵ Our experience with this procedure now extends over 18 years and 59 patients have undergone cardiomyotomy alone or with additional procedures designed to lower gastric acid secretion or to facilitate gastric emptying or both.

Esophagitis with cicatricial stricture may complicate the course of patients operated upon for achalasia.

This complication may require a more radical surgical approach. Esophagogastric

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TABLE 1. *Associated Conditions*

Condition	Number of Patients
Hiatal hernia	4
c̄ esophagitis	2
c̄ Schatzski's ring	1
Esophagitis	10
c̄ stricture	3
Esophageal diverticulum	4
Midesophageal	2
Epiphrenic	1
Multiple	1
Duodenal ulcer	5
Leukoplakia	4
Duodenal and jejunal diverticula	1

resection was initially employed in seven patients with this complication. Late follow-up results in these patients are also included.

Clinical Material

Adult Patients

Among 81 adult patients there were 49 women and 32 men. The average age at the time of operation was 48.4 years.

The average duration of symptoms prior to operation was nine years. The most commonly encountered symptoms were: (1) dysphagia—81 patients, (2) regurgitant vomiting—76 patients and (3) substernal distress—36 patients. A history of upper gastrointestinal bleeding was obtained from 17 patients. In five of these massive bleeding was induced by dilatation. Weight loss prior to operation averaged 22.4 pounds, however 15 patients did not lose weight.

A history of single or repeated episodes of aspiration pneumonia was obtained in 14 patients. An additional eight patients complained of chronic cough.

Four patients underwent emergency operations for esophageal perforation second-

ary to pneumatic dilatation. Associated upper gastrointestinal tract lesions were found in 25 patients and are listed in Table 1.

Pediatric Patients

There were two boys, each aged seven years, and one girl aged 20 months. Each of the boys experienced dysphagia, regurgitant vomiting and retarded growth. Both had had at least one episode of aspiration pneumonia. One boy had had a previous Heller procedure. The girl had regurgitant vomiting and failed to thrive.

Diagnosis and Degree of Esophageal Dilatation

The diagnosis was established by a combination of esophagoscopy and barium roentgenograms in the earlier cases. In patients encountered during later years cine-esophagrams and motility studies were frequently used.

Using the estimate of the endoscopist and the preoperative esophagram the degree of esophageal dilatation was graded as follows: grade 1—mild dilatation, 11 patients; grade 2—moderate dilatation, 27 patients; grade 3—marked dilatation, 43 patients.

Operative Therapy

The 84 patients were divided into four groups on the basis of the operation employed (Table 2).

TABLE 2. *Types of Operations*

Group	Operative Procedure	No. of Patients
I	Cardioplasty	18
II	Cardiomyotomy	44
III	Cardiomyotomy plus*	15
IV	Esophagogastrectomy	7

* Plus indicates additional procedures designed to diminish secretion of hydrochloric acid from the stomach and/or to facilitate gastric emptying.

Group I. Eighteen patients underwent various cardioplastic procedures or esophagogastrostomy (no portion of stomach being resected).

Group II. The Zaaier-Heller procedure was employed in 44 patients.

Group III. Fifteen patients underwent cardiomyotomy in addition to other operative procedures at the time of the myotomy in attempts to diminish gastric acid secretion or to facilitate gastric emptying or both. Seven underwent cardiomyotomy and pyloroplasty, five were treated with cardiomyotomy, vagectomy and pyloroplasty, two underwent myotomy and partial gastric resection and one underwent cardiomyotomy, gastroenterostomy and vagectomy.

Group IV. These seven patients had severe peptic esophagitis in addition to achalasia and were treated by esophagogastric resection. Two had had previous cardiomyotomies elsewhere. Esophagogastric resection was employed in all seven patients. Gastrointestinal continuity was restored by Roux-en-Y esophagojejunostomy in three, jejunal interposition in two and esophagogastrostomy in two.

Follow-up Evaluation

Eight of the 84 patients were lost to follow-up 1 month to 6 years postoperatively. The remaining 76 are currently being followed or have died during the follow-up period of known causes.

Any patient who maintained a stable weight did not require dilatation and had no further episodes of aspiration is classified as a good result. Periodic recurrence of mild symptoms at infrequent intervals was common and does not, in our opinion, constitute a bad result. We agree with Bradham and Sealy⁴ that "in evaluating the results of the modified Heller operation, it may be realized that motility is still abnormal; therefore some symptoms per-

TABLE 3. *Immediate Postoperative Complications of Surgery—All Groups*

Complication	No. of Patients
Empyema thoracis	1
Left phrenic nerve paralysis	2
Pneumonia	1
Empyema with pleuro-cutaneous fistula	1
Empyema c̄ esophagopleuro-cutaneous fistula	1
Massive upper gastrointestinal tract hemorrhage	2
Herniation stomach into chest c̄ necrosis of stomach and esophagus	1
Retained stay suture requiring operation	1
Esophagitis with stricture	1
Gastric outlet obstruction	1

sist." Patients who lost weight, had peptic esophagitis, recurrent aspiration pneumonia or severe recurrences of functional esophageal obstruction were classified as bad results.

Results of Therapy

Immediate Results

There were no postoperative deaths. Complications encountered among 12 patients are listed in Table 3. The complications were evenly distributed among the procedures used but individual groups were small making valid comparisons impossible. Two instances of empyema occurred secondary to entry into the esophagus during cardiomyotomy. These complications as well as phrenic nerve paralysis occurred early in our experience and there have been no similar complications in the past 10 years.

Gastric outlet obstruction occurred secondary to inadvertent sacrifice of the vagi in a patient whose mediastinum was encased in the granulomata of sarcoidosis. The problem was readily remedied by gastroenterostomy.

TABLE 4. *Poor Surgical Results*

Group	No. of Patients	Late Complications	Patients Requiring Reoperation
1	12/18	Peptic esophagitis	8
2	8/44	Recurrent symptoms one patient Peptic esophagitis seven patients	5
3	2/15	Peptic esophagitis	1
4	3/7	Stricture	0

Massive upper gastrointestinal bleeding was encountered in two patients. One required reoperation at which time a bleeding esophageal vessel was ligated. Gastric herniation occurred through a disrupted diaphragmatic suture line and was treated successfully by excision of the necrotic area of stomach.

Late Results

Group I. Four patients were asymptomatic for periods ranging from 2 to 6 years and then were lost to follow-up. One patient died in an auto accident 6 years postoperatively at which time she had no symptoms. One patient followed for 17 years remains entirely well.

The remaining 12 patients have been followed for periods up to 20 years (six patients) or until death. Eight of these patients have required at least one additional operative procedure because of severe peptic esophagitis. The remaining four all have esophagitis with strictures or ulcers. Two of the 12 are dead. One died 5 years after a second operation for peptic esophagitis of carcinoma of the esophagus. The other patient bled to death 6 years postoperatively from an esophageal ulcer.

Group II. Among 44 patients who underwent cardiomyotomy alone three were lost to follow-up within one year. Two died of cerebrovascular accidents 2 months and 12

years postoperatively. Both were free of symptoms at the time of death. The remaining 39 patients have been followed from 4 months to 17 years.

Results were good in 31 patients of whom five have been followed 10 or more years, 16 more than 5 years and the remainder 6 months to 4 years. Three with good results were in the pediatric group.

Eight patients had poor surgical results. Recurrent functional obstruction occurred in one. This patient underwent a second Heller procedure only to develop severe peptic esophagitis requiring three additional operative procedures.

Severe peptic esophagitis following cardiomyotomy accounted for poor results in seven of the eight patients. Two had peptic esophagitis with strictures as well as achalasia preoperatively. Myotomy aggravated their symptoms. Closure of an esophageal perforation was apparently related to a poor result in two patients. In one the perforation occurred preoperatively during pneumatic dilatation and in the other it occurred during operation and was associated with a postoperative empyema. In each case a stricture was noted on postoperative upper gastrointestinal roentgenograms.

Group III. There were 15 patients in this group. One was lost to follow-up after 3 months. A second died of leukemia 7 months postoperatively. At autopsy he had severe peptic esophagitis with leukemic infiltration of the esophagus. A third patient died 4 years postoperatively of metastatic adenocarcinoma of undetermined origin. This patient had no recurrent symptoms.

Ten patients have been followed one to 11 years. Severe peptic esophagitis occurred in one who has since required 12 hospital admissions because of esophageal obstruction. This patient had moderate esophagitis with stricture formation prior to cardiomyotomy and presented with

TABLE 5.

Authors	Year	No. of Patients	Poor Results	Recurrent Obstruction	Peptic Esophagitis
Hawthorne, Frobeses, Nemir ¹⁴	1956	35	7	2	5
Acheson, Hadley ¹	1958	33	7	4	3
Douglas, Nicholson ⁷	1959	28	8	6	2
Ferguson, Burford ¹¹	1960	44	8	4	4
Payne, Ellis, Olsen ¹⁷	1960	96	7	3	4
Browse, Carter ⁶	1961	15	4	3	1
LeRoux, Wright ¹⁶	1961	26	4	4	0
Barlow ²	1961	59	7	1	6
Dubourg, Fontan, Gourdon ⁹	1962	48	12	0	12
Root, Wangenstein ¹⁸	1962	19	6	2	4
Drake ⁸	1962	17	1	0	1
Jekler, Lhotka, Borek ¹⁵	1964	21	3	0	3
Sawyers, Foster ²¹	1967	22	5	0	5
Rees, Barnes, Thorbjarnarson	1969	59	10	1	9

Number in parentheses following the authors names refers to the reference number in the bibliography.

complete esophageal obstruction. The remaining nine patients had good results.

Two patients have been followed for less than one year and are both in good health.

Group IV. All 7 patients in this group had severe peptic esophagitis associated with achalasia preoperatively.

One patient died 16 years postoperatively of esophageal carcinoma after 15 years of good health. Two patients have anastomotic strictures that require periodic dilatation. One has chronic low grade bleeding from alkaline esophagitis but is markedly improved over his preoperative status. Three are well and have had no more difficulty. In Table 4 are listed the poor results in each group and the causes.

Discussion

It is evident that operative procedures which destroy the esophageal sphincter mechanism substitute reflux esophagitis for obstruction due to achalasia. This is apparent in 12 of our 18 patients who underwent cardioplasty or esophagostomy.

Because the surgical treatment of achalasia is mechanical therapy for a neuromuscular disorder the patient is never in

a true sense "cured." The Zaaier-Heller procedure consistently gives good results in 64-87% of patients as reported in many series.^{1, 2, 3, 6, 7, 8, 9, 11, 15, 16, 17} There are however, two pitfalls with the procedure.¹⁰ An insufficient myotomy may result in recurrent dysphagia. An incision sufficient to release the mechanical obstruction may result in reflux and subsequent peptic esophagitis because of destruction of the esophageal sphincter mechanism.

Results of insufficient myotomy usually become apparent almost immediately. One patient in group two was not at all improved after operation. A second myotomy was attempted 3 years later with resultant reflux and esophagitis. Late recurrences of dysphagia have been ascribed to bridging of the muscle gap with scar tissue.¹⁰

The reported incidence of peptic esophagitis following cardiomyotomy varies from four to 25% in collected series (Table 5). Barlow reported six instances of peptic esophagitis with stricture among 59 patients who underwent cardiomyotomy and an additional 15 patients who had reflux.² Hawthorne, Frobes and Nemir¹⁴ reported that 11 of 21 patients had reflux following cardiomyotomy when barium x-ray studies

were obtained with the patient in the Trendelenburg position and executing a Valsalva maneuver. Hiatal hernia with or without peptic esophagitis may occur after a Zaaier-Heller procedure¹² and some authors advise reconstruction of the hiatus at the time of operation to prevent this complication.^{11, 15}

We agree that when a hiatal hernia is present it is desirable to repair this at the time of cardiomyotomy. A greatly dilated hiatus associated with megaesophagus should always be repaired and in particular the phrenoesophageal ligament must be reconstructed. When a long intra-abdominal segment of esophagus is present the ligament may be divided, shortened and resutured to the esophageal wall above its normal position of attachment. The cardia of the stomach should be sutured to the diaphragm to elevate and accentuate the angle of His. Whenever possible a temporary gastrostomy should be done to avoid an indwelling nasogastric tube that might increase reflux in the immediate postoperative period.

The esophageal hiatus should not be narrowed when the esophagus is particularly large. In achalasia the esophagus empties only by gravity. A "tight fit" at the hiatus may result in markedly delayed emptying.

The occurrence of esophagitis in patients in Groups II and III of our series was the most significant factor in poor surgical results. We were unable to relate this complication with the approach used for cardiomyotomy (transthoracic versus transabdominal) or the length of the cardiomyotomy incision.¹⁰ Nor were we able to correlate the occurrence of peptic esophagitis with other upper gastrointestinal lesions as has been suggested by Sawyer and Foster²⁰ as a result for late failure following the modified Heller procedure.

In our series the single most significant

factor in predicting a poor surgical result was the presence of peptic esophagitis preoperatively. Among seven patients treated by cardiomyotomy who had esophagitis (proved by biopsy) preoperatively, three had poor results. The amount of preoperative dilatation of the esophagus did not appear to be a factor in predicting a poor surgical result.

Since peptic esophagitis is the greatest obstacle to good long-term results in patients who undergo cardiomyotomy the addition of vagotomy and drainage procedure or vagotomy and partial gastric resection should be considered. While cardiomyotomy alone carries an almost negligible mortality,⁴ the addition of other operative manipulations at the time of myotomy might add to morbidity and perhaps mortality. Our generally good initial results in 15 patients (Group III) suggest that better late results may be obtained by adding vagotomy with a drainage procedure or partial gastric resection to cardiomyotomy. These seem to be indicated especially in patients who have esophagitis associated with achalasia preoperatively.

Summary

The late occurrence of peptic esophagitis was the most significant factor causing poor late results among 84 patients who underwent operative treatment for achalasia.

Cardioplasty alone resulted in peptic esophagitis in more than half of patients in whom this procedure was employed—and it should not be used.

Esophagogastric resection may be necessary in patients who have achalasia with an associated cicatricial stricture.

A modification of Heller's cardiomyotomy continues to give the best results. These results may apparently be improved by addition of vagotomy and a drainage procedure.

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