Achalasia: Willis or Heller?

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Minimal-access surgery permits many older, tested procedures to be performed in new and better ways that make the operations less painful and more accurate and that reduce hospital stays and disability. These features make it reasonable to reassess the relative indications for surgery and nonsurgical therapy in achalasia of the esophagus. Achalasia comes from a Greek word that means “failure to relax.” Cardiospasm and achalasia refer to the same condition (1, 2). This report addresses esophageal achalasia—its history, diagnosis, pathophysiology, and treatment options. We report our initial experience in treating this disorder surgically using laparoscopic-modified Heller myotomy combined with partial gastric fundoplication.

RESULTS

Between December 1994 and May 1998, we did laparoscopic-modified Heller myotomies on 5 patients. There were 3 men and 2 women, ranging in age from 26 to 87 years (mean, 52 years). All patients had grade 2 or 3 achalasia of the esophagus and had barium esophagram, esophagoscopy, and esophageal manometry before surgery. We did 4 Dor anterior partial fundoplications and 1 Toupet posterior partial fundoplication. We had no intraoperative perforations, and no patient required conversion to an open procedure. When esophagrams with water-soluble contrast medium were taken on the morning following surgery, before patients started a diet, no leaks were shown. All patients tolerated a clear liquid diet that day and advanced to a mechanical soft diet before hospital discharge. All patients went home on postoperative day 2 except an 87-year-old man who had chronic obstructive pulmonary disease and was on steroids. He had a previous Heller myotomy through the chest and had been treated with both hydrostatic dilation and clostridium botulinum injections. All patients were able to return to work within 2 weeks. There was no mortality or major morbidity.

Surgical outcomes can be graded on a scale of 1 to 5, with grade 1 being an outcome worse than before surgery; 2, unimproved; 3, a fair outcome; 4, a good outcome; and 5, an excellent result. A good outcome is distinct improvement of the patient's complaints with few symptoms. An excellent outcome describes a patient with essentially normal swallowing and no regurgitation or heartburn. Using these criteria, 1 patient had a good result and 4 had excellent results (mean, 4.8) (Table). No patient had dysphagia postoperatively. All our patients were pleased with the results and their decision to have the operation.
HISTORY

Sir Thomas Willis, an Englishman, described cardiospasm and treated a patient by dilation using a sponge attached to a whalebone in 1672 (1, 3). Ernest Heller, a German, did the first successful esophagomyotomy 241 years later on April 14, 1913 (1, 4, 5). In 1937, F. C. Lendrum proposed that failure of the lower esophageal sphincter to relax causes functional esophageal obstruction, and the name changed from cardiospasm to achalasia (2, 3). Dor reported his anterior partial fundoplication in 1962 (6), and Andr? Toupet reported his posterior partial fundoplication in 1963 (7). Shimi and colleagues in the United Kingdom did the first Heller myotomy laparoscopically in 1991, 77 years after Heller's operation (8–10).

PATHOPHYSIOLOGY

Achalasia can be categorized by the diameter and length of the esophagus. An esophagus with a diameter <4 cm is grade 1, grade 2 is 4 to 6 cm, grade 3 is >6 cm, and grade 4 is a sigmoid esophagus (11).

Achalasia is a primary motility disorder of the esophagus, occurring in approximately 1 per 100,000 population per year (2, 12, 13). In the normal esophagus, peristaltic waves follow each swallow. In achalasia a neuromuscular defect produces a marked decrease or absence of esophageal body peristalsis (1, 9, 13–16). Occasionally peristalsis returns after successful treatment (17, 18). These patients may have low-level peristaltic activity that becomes apparent after relief of the functional obstruction. Microscopically, there is degeneration of ganglion cells in Auerbach's myenteric plexus (2). This condition is in some ways analogous to megacolon. Normally there is a lower esophageal high-pressure zone, or lower esophageal sphincter. In achalasia the lower esophageal sphincter is hypertonic, producing resting pressures above normal and relaxing incompletely after swallowing (13, 15). This produces a functional obstruction, resulting in dilation and elongation of the body of the esophagus with rapid narrowing at the cardia. In vigorous achalasia, simultaneous nonpropagated high-amplitude pressure waves occur throughout the esophageal body. Some suggest that achalasia and diffuse esophageal spasm may be different phases of the same

<table>
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<tr>
<th>Patient</th>
<th>Age</th>
<th>LOS*</th>
<th>Postoperative dysphagia</th>
<th>Postoperative problems</th>
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<td>2</td>
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<td>Occasional heartburn</td>
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</tr>
<tr>
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<td>2</td>
<td>0</td>
<td>None</td>
<td>5</td>
</tr>
<tr>
<td>6</td>
<td>97</td>
<td>4</td>
<td>1x</td>
<td>Food hung briefly on one occasion</td>
<td>4</td>
</tr>
</tbody>
</table>

*LOS indicates length of hospital stay (days).
motility disorder (12, 19, 20). The etiology of achalasia is unknown (9).

**DIAGNOSIS**

The clinical history of dysphagia, retrosternal pain, regurgitation of stagnant food, and weight loss should lead one to suspect achalasia. Patients with achalasia may have pulmonary symptoms such as cough and hoarseness caused by aspiration (2, 21). They suffer the complications of aspiration and have an increased incidence of cancer of the esophagus (2, 22). Malignant obstruction, gastroesophageal reflux disease stricture, diffuse esophageal spasm, and nutcracker esophagus can mimic achalasia (9, 20).

Characteristic chest x-ray findings in achalasia are a widened mediastinum, an air fluid level in the esophagus, an absent gastric air bubble, and aspiration pneumonia (2, 20). Barium esophagram reveals a dilated esophagus with a “bird's beak” narrowing at the cardia (20). Cine esophagram demonstrates aperistalsis. Elongation and dilation of the esophagus produce tortuosity, leading to a sigmoid esophagus in extreme cases (20).

Esophagoscopy finds retained food in a dilated esophagus that often has significant esophagitis. The lower esophageal sphincter is tight, but the esophagoscope passes with gentle pressure (20). All patients being evaluated for surgery should have esophagoscopy to rule out an organic obstruction due to carcinoma or other coexisting diseases (11, 20).

Esophageal manometry is an essential study in the diagnosis of achalasia. It shows absence of esophageal body peristalsis and a high-pressure lower esophageal sphincter that fails to relax normally with swallowing (1, 20).

**TREATMENT**

The treatment of achalasia is palliative since the underlying motility disorder cannot be corrected (9, 23). Relief of dysphagia and the obstructing high-pressure lower esophageal sphincter with prevention of gastroesophageal reflux constitutes the goal of therapy (11, 13, 24). Pharmacological therapy (25), clostridium botulinum toxin injection (16), forceful hydrostatic dilation (14, 24, 26), and surgical treatment (1, 2, 5, 27–30) are currently available treatments.

The medications predominantly used are the organic nitrates (isosorbide mononitrate) and the calcium channel antagonists (nifedipine) (25, 30). They are largely ineffective and have a transient effect and a high failure rate. Their primary value is in patients with mild symptoms or those who are poor candidates for definitive therapy (20).

The injection of botulinum toxin into the lower esophageal sphincter is a relatively new procedure (16). It has a high failure rate and a temporary benefit, requires repeated injections, and may produce scarring that can make subsequent surgery more difficult and hazardous (5).

Pneumatic dilation is the most effective nonsurgical therapy for achalasia (5). The technique
consists of placing a balloon in the lower esophageal sphincter and rapidly inflating it to a
diameter of 3 to 5 cm to disrupt the lower circular muscle fibers (24, 26). It requires only
overnight observation, and the initial treatment is less expensive than surgery (23, 24). One
pneumatic dilation treatment produces good results 40% to 78% of the time (2, 24, 27, 30,
31). Retreatment of the failures becomes progressively less efficacious (2). Complications
of pneumatic dilation include perforation, bleeding, and gastroesophageal reflux (32). The
reported incidence of perforation of the esophagus ranges from 3% to 15%. Perforation can
lead to mediastinitis, empyema, or even death (24, 27, 32). Perforations or repeated dilations
can cause esophageal scarring that may interfere with later surgery (30).

Sauer et al reported long-term follow-up of 66 patients treated by primary dilation (32).
Two of 66 patients had pulmonary aspiration, and 12% had perforation. Two patients died, a
3% mortality rate. At follow-up (average, 4 years), only 50% of the patients were
swallowing well, 30% had symptoms of gastroesophageal reflux disease, and 20% had
recurrent dysphagia. Abid et al reported 88% good to excellent results with pneumatic
dilation (33). However, only 77% of their patients achieved a good result with initial
dilation, and 7% of their patients had perforation.

Esophagomyotomy has become the primary surgical treatment for achalasia, although
patients with a severely decompensated sigmoid esophagus or carcinoma may require
esophagectomy (22, 30, 34). Esophagomyotomy has fewer complications and a higher
success rate than pneumatic dilation (24, 27–31, 35). Csendes et al demonstrated good
results in 65% of patients treated with pneumatic dilation and in 95% of the surgical group
in a prospective randomized study comparing forceful dilation and esophagomyotomy (24).
Studies from the Universities of Iowa, Wisconsin, and Illinois, as well as the Mayo Clinic,
report 30% to 85% better results from myotomy than from forceful dilation (29, 31, 35, 36).
A collected review by Ferguson found a 71% rate of improvement with pneumatic dilation
in 1049 patients and an 89% rate in 1199 surgical patients (20).

Bonavina et al observed 193 surgical patients for 1 to 12 years. They found good or
excellent results in 94% (11). Parrilla Paricio et al observed 48 surgical patients who had
received myotomy plus a Toupet partial fundoplication for 3 to 12 years (mean, 5.4 years)
(15). They found 92% good to excellent results. A collected review of 5002 surgical patients
by Andreollo and Earlam revealed 89% good or excellent results and a mortality of 0.2%
(1).

Several controversies surround esophagomyotomy (8, 20). Is open or videoendoscopic
better? Is the abdominal or thoracic approach preferable? How long should the myotomy
be? How far proximal should the esophageal portion of the myotomy go? Should the
myotomy extend onto the stomach, and, if so, how far? Is an antireflux procedure
necessary? If it is, which procedure is best?

Ellis et al have championed a transthoracic limited esophagomyotomy, extending no more
than 1 cm onto the stomach. This avoids a dissection of the hiatus and preserves the oblique
sling muscles of the proximal stomach that form the “collar of Helvitus.” They do not do an
antireflux procedure, contending that it is meddlesome and hazardous. They argue that their
incidence of postoperative dysphagia and gastroesophageal reflux is acceptable (37). We believe it is difficult to carry the myotomy far enough onto the stomach without going too far. An inadequate myotomy fails to relieve the dysphagia, and one that is too long causes gastroesophageal reflux (8, 20).

The videoendoscopic approach produces less pain and yields a shorter hospital stay, shorter disability, lower cost, and a better cosmetic result than the open approach (8, 38). Because the operative field is magnified, it is less disruptive and more accurate (8). It produces results at least as good as those from open procedures (8, 9, 11, 38).

The laparoscopic approach is easier technically than thorascopy, since the instruments are in line with the long axis of the esophagus instead of being perpendicular to it. Laparoscopy provides easier access to the distal esophagus and proximal stomach—the most difficult and important part of the procedure (5, 8, 9). The mediastinal esophagus is relatively short, and transhiatal laparoscopy provides excellent access to the majority of it (8). Laparoscopy produces less pain and morbidity and causes a shorter hospital stay (5, 8, 9).

The indications for surgery are proven symptomatic achalasia and the ability of the patient to tolerate general anesthesia (9).

The same hiatal dissection used in a laparoscopic Nissen fundoplication provides excellent exposure of the gastroesophageal junction and the lower two thirds of the thoracic esophagus. The magnification of the operative field allows good visualization of the circular muscle fibers. We make a myotomy of 8 to 14 cm, carry the incision 2 to 4 cm onto the stomach, and dissect the esophageal muscular layer away from the mucosa. A long myotomy ensures division of all the lower esophageal high-pressure zone circular muscle fibers and may prevent chest pain from nonpropagated simultaneous contractions or diffuse esophageal spasm (8, 11, 20).

An adequate myotomy produces gastroesophageal reflux, so we add a partial fundoplication. Both the Toupet and Dor procedures have their advocates. Since the Dor partial fundoplication is an anterior wrap and covers a portion of the exposed mucosa, it should help protect against leakage from an unrecognized perforation or devascularization of the mucosa. However, it may carry a higher incidence of dysphagia and gastroesophageal reflux than the Toupet (5, 9). Following myotomy, we use esophagoscopy to confirm adequacy of the myotomy and air insufflation with the esophagus submerged in saline to ensure that there is no unrecognized perforation. If a perforation occurs, laparoscopic suturing can close it safely (5, 9, 11, 39). Since the aperistaltic esophagus has negligible propulsive activity, a loose partial fundoplication is desirable. We dissect the muscular layer away from the mucosa for 180° and suture the cut edges of the myotomy to the fundoplication. This produces traction that keeps the edges apart, lessening the likelihood that the edges will heal together and cause a fibrous stricture producing a recurrence of the high-pressure lower esophageal sphincter.

The most common causes of a poor result following esophagomyotomy are incorrect diagnosis, inadequate myotomy, fibrosis and narrowing of the myotomy, gastroesophageal
reflux, and the development of esophageal carcinoma (1, 11, 30). The giant megaesophagus may continue to empty poorly after myotomy and require esophagectomy. This is especially true in patients who have had a prior myotomy or who have developed a gastroesophageal reflux disease stricture (20, 21, 30, 34).

CONCLUSION

Should we use the whalebone or the laparoscope to treat achalasia? Laparoscopic Heller myotomy with a partial fundoplication is a safe and effective treatment for patients with achalasia. We consider it the treatment of choice for achalasia because it is more effective and durable than any other form of therapy, and it leads to minimal pain, a short hospital stay, and low morbidity and mortality. Early evaluation of achalasia patients and prompt surgical intervention can help prevent progression of the disease and make better surgical results more likely.

References


