Achalasia

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J. Duncan Phillips

Achalasia is an uncommon disorder of esophageal dysmotility, with an annual incidence of only 1 in 100,000 individuals. Although usually thought of as being simply a failure of normal relaxation of the lower esophageal sphincter (LES) during swallowing, it is actually part of a more diffuse disease of esophageal function. Because treatment for diminished or absent esophageal peristalsis is so poorly developed at this time, clinicians tend to focus on the LES, which is the only part of esophageal anatomy that lends itself to intervention.

Normal swallowing is a complex process that involves antegrade propulsion of ingested liquids and solids by coordinated peristaltic contractions of the esophageal body and a transient relaxation of the LES from its normal resting pressure. In patients with achalasia, this normal relaxation fails to occur. Although the precise etiology of achalasia is uncertain, the primary cause is likely an abnormality of the esophageal myenteric plexus. Patients with achalasia often have a reduced number of ganglion cells and the ganglion cells are surrounded by an inflammatory infiltrate. Unlike Hirschsprung disease, which is thought to be due to failure of normal migration of ganglion cells during fetal life, the etiology of achalasia might involve an autoimmune mechanism, with progressive partial destruction of the ganglion cells and the inhibitory neurons that normally mediate LES relaxation.

Achalasia in children has been associated with trisomy 21, triple-A syndrome (achalasia, alacrima, ACTH insensitivity), and familial dysautonomia, but most are sporadic. Although the disease has rarely been described in toddlers, the typical pediatric patient is a teenager between the ages of 13 and 17.

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Diagnosis

Patients typically present with a history of progressively worsening dysphagia that begins with solid foods and then progresses to soft foods and eventually even liquids. Most describe food getting stuck in the cervical region and point to the base of the neck. They usually discover that they can propel that food into the stomach by swallowing liquids frequently throughout their meals. Most also experience intermittent regurgitation of undigested food that occurs immediately or up to a few hours after meals. They and their parents frequently describe this as "vomiting" but careful questioning usually reveals that the vomitus is nonbilious and composed of only chewed-up food.

Substernal chest pain is another very common symptom. This pain is typically described as "heartburn" and often incorrectly attributed to gastroesophageal reflux disease, leading to empiric treatment with acid blocking medications or promotility drugs. These agents are typically of no benefit in patients with achalasia. Most children with achalasia will experience weight loss that ranges from mild to severe (10–20 % of body weight). The weight loss is gradual and often subtle.

The initial diagnostic test in the child with dysphagia is usually a contrast esophagram. It is important that the study be performed in both the supine and upright positions, to properly assess esophageal emptying. It is also helpful to take video recordings with emphasis on the pattern of esophageal contractions. Classic findings include a dilated, dysmotile esophagus and a "bird's beak" deformity at the gastroesophageal junction (Fig. 35.1). Early in the course of the disease, the radiographic findings can be subtle, while, in cases of long-standing disease, the progressively redundant esophagus can adopt a sigmoid shape. Careful observation by the radiologist will usually reveal the absence of relaxation of the GEJ.

Probably because GERD is so much more common, most children with achalasia are initially thought to have lower

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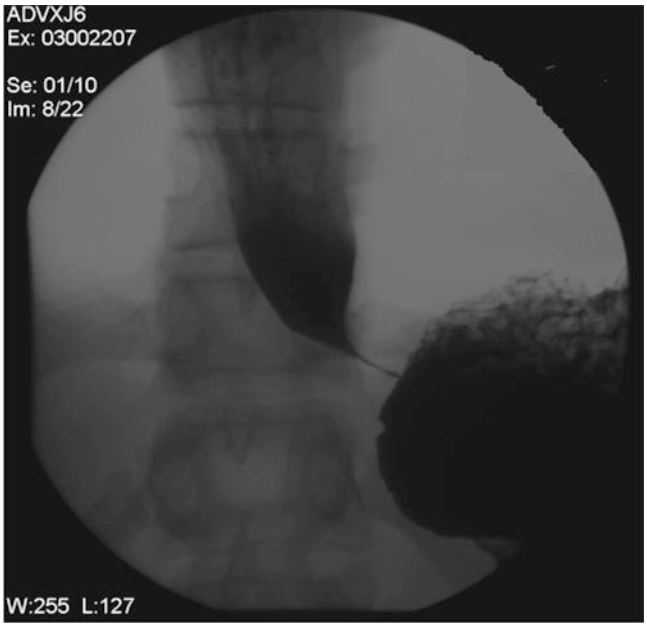

Fig. 35.1 Esophagram demonstrating the "bird's beak" deformity at the gastroesophageal junction, indicating long-standing esophageal achalasia. This 16-year-old girl had dysphagia for over 1 year. Resting lower esophageal sphincter pressure was 42 mmHg

esophageal spasm or stricture due to GERD. Following the esophagram, most physicians will usually then recommend a flexible esophagogastroduodenoscopy. Endoscopy may demonstrate pooling of retained fluid within the esophagus and esophageal inflammation related to stasis. The endoscopist might encounter mild resistance but can usually pass the endoscope into the stomach, essentially ruling out a fibrotic stricture.

The standard test for esophageal achalasia remains esophageal manometry. Pressure recordings show absent or diminished peristalsis in the upper esophagus, elevated LES pressures, and minimal or absent LES relaxation. It is important to note that these pressure differences are typically not as impressive as they might be in adults, affected children having resting LES pressures only about two-thirds that of adults.

Preoperative Preparation

Current treatment strategies for achalasia are directed specifically at the LES. Balloon dilatation of the LES under general anesthesia is the most common nonoperative technique utilized in children. Symptomatic relief is unfortunately almost always transient and thus repeated dilatations are required. Dilatation also carries with it the risks of general anesthesia and approximately a 3 % risk of esophageal perforation. It is useful mostly as a diagnostic tool to help children and their parents understand the potential benefits of Heller myotomy.

An alternative nonsurgical treatment for esophageal achalasia is endoscopic intra-sphincteric injection of botulinum toxin, which lowers LES pressure by inhibiting acetylcholine release from nerve endings. This effect is also transient, requiring repeated injections for long-term success. Several investigators have found that a repeated botulinum toxin injection induces scarring within the wall of the distal esophagus. As a result, the risk of esophageal perforation during subsequent Heller myotomy is increased. For this reason, many pediatric gastroenterologists do not advocate botulinum toxin injections for their patients and most experienced pediatric laparoscopic surgeons are recommending against it.

The occasional patient with achalasia will present with severe weight loss and malnutrition. These children benefit from preoperative supplemental feeds delivered by nasogastric tube. Positive nitrogen balance is associated with decreased perioperative morbidity but can take several weeks to achieve.

Treatment

Esophageal myotomy, originally including longitudinal incisions on both anterior and posterior sides of the LES, was described by Ernest Heller in 1913. Since essentially all authors now utilize a single anterior myotomy, a more accurate term for the operation as it is performed today is the modified Heller myotomy. Nevertheless, most authors consider "the Heller" the procedure of choice for the surgical treatment of children with achalasia. Excellent results have been reported with long-term follow-up of children treated with this procedure.

Controversy regarding specific technical aspects of the Heller myotomy include (1) whether it is better to approach the LES through the chest or the abdomen, (2) to what degree balloon dilatation or botulism toxin injection increases the complication rate of the operation, (3) whether one should also perform an antireflux operation, and (4) whether minimally invasive techniques are as good as the traditional open approach.

Heller originally performed the myotomy through a left thoracotomy, as he felt this allowed better visualization of the esophagus and that the myotomy could more easily be extended superiorly to the level of the inferior pulmonary veins. In fact, until the late 1980s, this was the standard approach at most centers. More recently, however, most have found that the abdominal approach allows for an adequate myotomy, is associated with less perioperative pain and morbidity, and makes fundoplication easier to perform.

Nonoperative treatments such as balloon dilatation and injection of botulinum toxin offer at best a transient improvement in symptoms and are associated with a small risk of significant complications. In addition, extensive scarring within the wall of the esophagus can make subsequent myotomy difficult or even dangerous. Therefore, most pediatric surgeons experienced with the laparoscopic technique recommend against both dilatations and injections.

After disruption of the LES by myotomy, many patients develop GERD, which may or may not be clinically apparent. Most surgeons therefore advocate a concomitant antireflux operation when performing the Heller procedure. This is mainly due to the concern that long-standing GERD places the patient at risk for Barrett's esophagus and esophageal carcinoma. No single technique is clearly favored and various authors have advocated the Nissen, Dor, Toupet, and Thal procedures.

Both laparoscopic and thoracoscopic Heller myotomy have been described. The thoracoscopic approach was initially more popular, but it soon became clear that up to 60 % of patients have significant gastroesophageal reflux. Since most surgeons felt that concomitant fundoplication was exceedingly difficult to do thoracoscopically, there was interest in developing a practical laparoscopic approach that would accommodate both operations.

Surgical Technique

The patient is positioned in the supine position with the surgeon standing to the patient's right side and the assistant on the left. Older children may have the legs extended on stirrups with the knees flexed 20–30° so that the surgeon can stand between the legs. We prefer to place a Foley catheter to evacuate the bladder and a nasogastric tube to decompress the stomach. A single dose of intravenous antibiotic is given prior to incision.

Five trocars are standard, with initial access at the umbilicus. We use a 5-mm trocar and a 5-mm 30° telescope but initial access with a larger trocar is preferred by some. We then place four more trocars under laparoscopic view. We have found it best to insert these trocars fairly high, along the costal margins, for optimal advantage.

A trocar along the right anterior axillary line allows for introduction of a liver retractor. We use a "snake" retractor to elevate the left lateral segment, exposing the anterior gastric wall and GE junction. A telescope holder can be attached to the operating room table to hold this liver retractor, thus freeing up the surgeon and assistant to each use both hands during the remainder of the procedure.

The three additional trocars are usually inserted in the right midclavicular line for insertion of a grasper, in the left anterior axillary line for the assistant's grasper, and in the left midclavicular line. The assistant can move the laparoscope to the left midclavicular trocar and control the camera with the left hand. This allows the surgeon, standing at the patient's right side, to operate with the left-hand instrument inserted via the right midclavicular trocar and the right hand to control instruments via the umbilical trocar.

The dissection is typically started by incising the gastrohepatic ligament to expose the right crus of the diaphragm. We then incise the peritoneum overlying the intra-abdominal esophagus, just superior to the phreno-esophageal fat pad. One can use the hook cautery or Harmonic Scalpel for most of this dissection. As during a Nissen fundoplication, we typically also incise the peritoneum along the anterior surface of the right crus. The anterior and poster vagus nerves are usually visible on the surface of the esophagus and should be carefully protected. If one has chosen to perform an anterior fundoplication, the posterior esophageal attachments may be left intact.

The myotomy is typically performed with the monopolar hook cautery at the 10 or 11 o'clock position, just to the right of the anterior vagus nerve (Fig. 35.2). We usually start this just cephalad to the phreno-esophageal fat pad, which is sometimes quite thick, and extend this cephalad for about 5 or 6 cm. The surgeon typically uses the left-hand instrument to grasp the right edge of the myotomy and the assistant typically uses a gentle grasper to control the left edge of the myotomy so that the muscle edges can be gently separated. We find that 5-mm Hunter bowel graspers work quite well for this step. To avoid thermal injury to the underlying esophageal submucosa, it is important to not set the current too high and avoid arcing of the current. The myotomy incision must be carried through the outer longitudinal muscle and also through the inner circular muscle, exposing the

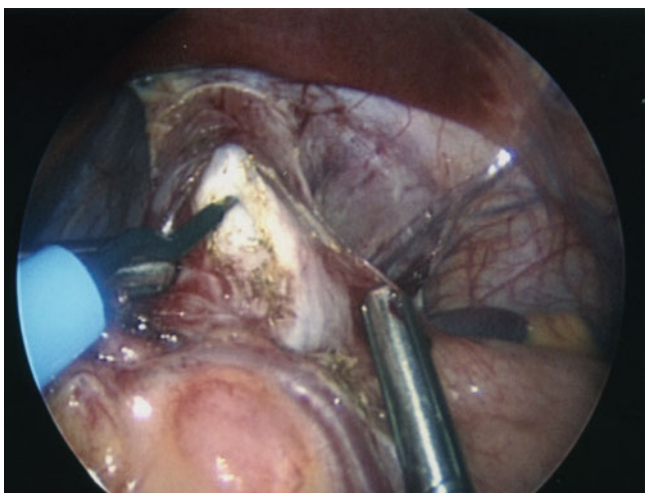

Fig. 35.2 Laparoscopic Heller myotomy. Peritoneum overlying the intra-abdominal esophagus has been divided. The hook cautery is used to begin the myotomy on the anterior surface of the esophagus

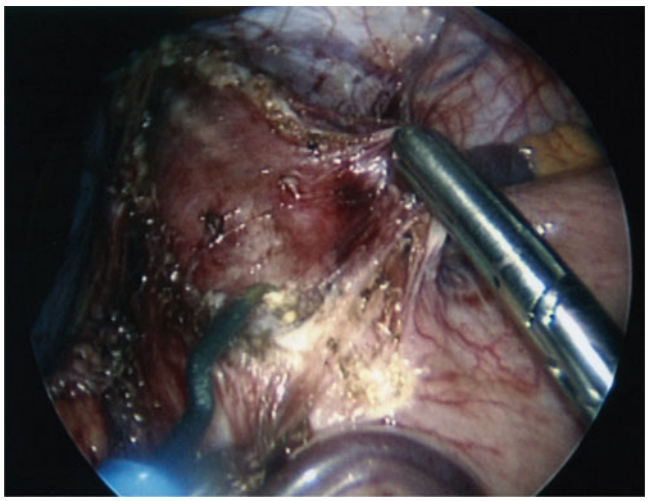

Fig. 35.3 Laparoscopic Heller myotomy. The myotomy of the distal esophagus has been completed and is being extended onto the stomach for 1–2 cm

submucosal vascular plexus. Gentle downward traction on the cut muscle edges allows the surgeon to extend the myotomy well up into the lower mediastinum. The myotomy is then extended 1–1.5 cm onto the anterior gastric wall (Fig. 35.3). In anticipation of the fundoplication, we typically divide the short gastric vessels at this point, using either cautery or the Harmonic Scalpel.

Due to the risk of causing significant dysphagia, most surgeons prefer to avoid creation of a 360° Nissen fundoplication in patients with disorders of esophageal motility. To create an anterior 180° fundoplication (Dor), two rows of permanent sutures are used. We prefer to use 2-0 braided nylon or a similar multifilament coated suture. To help the needles pass through the 5-mm trocars, they need to be straightened slightly into a canoe or ski shape.

The first row of sutures secures the gastric fundus to the left edge of the myotomy. Three or four interrupted sutures are typically needed (Fig. 35.4). The stomach is then "folded over" the myotomy site (from patient's left to right) and a second row of interrupted sutures secures the fundus to the right edge of the myotomy. The uppermost stitch on each side usually includes the crus. We typically place one or two additional sutures between the fundus and the diaphragm anteriorly.

Many authors have described the intraoperative use of a flexible esophagoscopy to aid in myotomy assessment and suture placement. As an alternative, we pass a 6 Fr Fogarty catheter by mouth, inflate the balloon with air or water, and pull it retrograde from stomach to esophagus under laparoscopic visualization. This allows assessment of myotomy completeness. Some authors inject dilute methylene blue dye to exclude occult inadvertent esophageal perforation.

If one has chosen to perform a posterior 180° (Toupet) fundoplication, one must make sure that the posterior esophageal attachments at the esophageal hiatus have been completely divided during the initial esophageal dissection. Following completion of the myotomy, the surgeon reaches posterior to the esophagus with a left-hand instrument to grasp the mobilized fundus and pulls it from the patient's left to right toward the caudate lobe of the liver. If the short gastric vessels have been properly divided, there should be no significant tension on the fundus and it does not snap back toward the patient's left.

Three rows of interrupted sutures are typically placed to complete the Toupet: an interrupted row securing the fundus to the right edge of the myotomy, a second row securing the fundus to the right crus of the diaphragm, and a final row securing the stomach to the left edge of the myotomy. In the Montupet modification of the Toupet, a crural stitch is placed posterior to the esophagus, to approximate the crura in the midline.

To perform the Heller myotomy using an open approach, most surgeons prefer an upper midline incision, beginning at or just to the side of the xiphoid process and extending to just above the umbilicus. A Thompson or Buckwalter retractor is extremely helpful for improved exposure of the GEJ. The left lateral segment of the liver can be detached from the undersurface of the left hemidiaphragm using electrocautery and retracted to the patient's right with a smooth blade of the retractor. The remainder of the intra-abdominal portion of the procedure is similar to that described above.

A thoracoscopic approach may useful in certain circumstances. For left-sided thoracoscopic procedures, single lung ventilation can often be achieved via right mainstem intubation or by the use of a bronchial blocker. In older children, a double-lumen endotracheal tube can be used. An alternative is the use of valved trocars and gentle CO_2 insufflation with pressures of 5–8 mmHg.

Typically, a four-trocar technique is utilized: in the sixth intercostal space in the midaxillary line, in the fourth intercostal space approximately 2 cm posterior to the posterior axillary line, in the fifth intercostal space in the anterior axillary line, and in the eighth intercostal space in the posterior axillary line. With the lung retracted anteriorly and superiorly, the inferior pulmonary ligament may be divided as high as the inferior pulmonary vein. The mediastinal pleura is incised to expose the distal thoracic esophagus. This can be facilitated by the placement of a flexible esophagoscope.

The myotomy is then performed in a fashion similar to that described for the laparoscopic procedure, using 5-mm hook cautery. The myotomy is typically extended just beyond the GEJ. A chest tube is usually placed through one of the port sites.

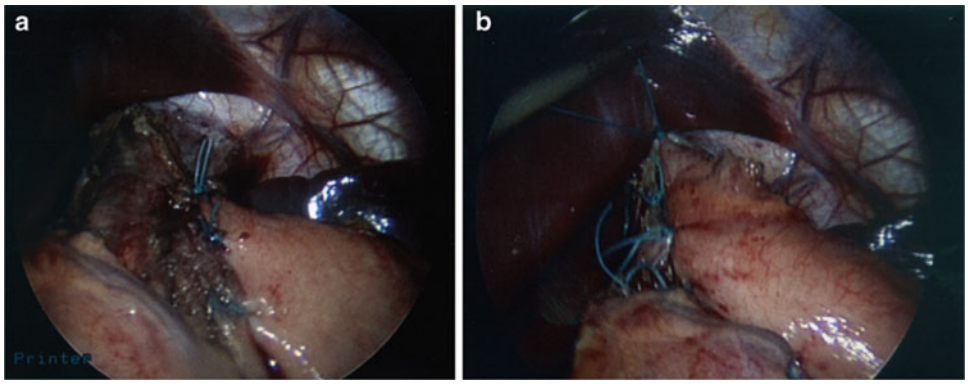

Fig. 35.4 Laparoscopic Heller myotomy with Dor fundoplication. (**a**) The first row of sutures (*arrows*) secures the fundus to the left edge of the myotomy. (**b**) The second row of sutures (*arrows*) helps to "fold" the fundus over the anterior wall of the esophagus at the level of the myotomy to secure the fundus to the right edge of the myotomy

Postoperative Care

Most surgeons at our institution obtain an esophagram on the first postoperative day prior to beginning oral feedings to verify proper esophageal emptying and to rule out the presence of a leak. It should be noted that the esophagram will not reveal any significant change in esophageal dilatation and often shows a rather long area of apparent esophageal narrowing at the myotomy site. This is often quite confusing or even alarming to those unfamiliar with the typical early postoperative radiographic appearance.

Following minimally invasive surgery, diet can usually be resumed immediately following the esophagram. Intake of soft foods is preferable to hard foods, such as crusted breads. Some authors recommend a "no-chunk" diet for 2–4 weeks after surgery. Most surgeons continue empiric acid blocking medications for a period of approximately 6 weeks, though there is little evidence that this is necessary. Most patients can be discharged home within 1–2 days of the minimally invasive operation.

Since achalasia is just one part of a more diffuse esophageal disorder, affected patients should be followed indefinitely by an experienced gastroenterologist. The association between long-standing achalasia and esophageal carcinoma is well-established. Surgical treatment of achalasia during childhood would be expected to reduce this risk but long-term follow-up data are lacking.

Editor's Comment

Achalasia is rare in children and the clinical presentation, though often insidious, is distinctive: dysphagia, chest pain, and regurgitation of undigested food. Nevertheless, patients frequently present for surgical consultation only after many months of misery, failed interventions, and ineffective medical therapy. This is unfortunate, not only because the patient suffers needlessly, but because there is a safe and effective operation available that becomes more difficult and more dangerous when the patient presents late after having been dilated or injected. Primary physicians and gastroenterologists should be encouraged to refer these patients to an experienced surgeon early in the course of the disease rather than as a last resort.

Laparoscopic Heller myotomy is an advanced minimally invasive technique but is generally safe in the hands of an experienced laparoscopist. The operation can be done safely with the patient in the supine position and the surgeon standing to the right, which avoids the inherent delay and added risk of placing the patient in stirrups. Some have touted the use of intraoperative adjuncts like manometry or endoscopy to assess the adequacy of the myotomy, but these are unproven and probably unnecessary. To protect the exposed submucosa and to avoid unnecessary dissection posterior to the esophagus, most recommend an anterior fundoplication, but this is certainly open to debate. A postoperative esophagram is admittedly overkill but avoiding even a single rare case of unrecognized perforation seems like adequate justification for this generally harmless exercise.

The robotic-assisted approach seems very well suited to Heller myotomy—visualization and the ability to manipulate instruments and suture are all superior to the laparoscopic approach. It seems likely that this will become the standard surgical approach in years to come.

The key to the success of the Heller myotomy, like most complex operations, is proper management of expectations. Patients naturally expect instantaneous relief of their symptoms and the ability to eat anything and everything immediately after the operation. In most cases, this is unrealistic and patients should be counseled to expect that some degree of dysphagia will persist for some time after the operation. This is because the disease affects the motility of the entire esophagus, the chronically dilated esophagus is ineffective at peristalsis, and there is a partial functional obstruction at the LES after myotomy. Their esophagus will empty principally by gravity and pressure from the advancing bolus of swallowed food or liquid. These symptoms resolve gradually over the course of several weeks or months, but in the meantime patients should be encouraged to avoid food with large chunks (meat, bread crust), to chew their food well, and to drink fluid frequently during meals. Some patients can

develop intermittent painful episodes of esophageal spasm that usually eventually cease. This has been treated with variable results using calcium channel blockers and medications that counter smooth muscle spasm.

Intraoperative perforations should be primarily repaired and then "patched" with the fundoplication, in which most patients should be able to tolerate oral intake after a brief (48–72 h) period of observation and a negative esophagram. The rare patient with an esophageal perforation noted on postoperative esophagram is treated initially with NPO, antibiotics, and careful observation. The leak usually seals spontaneously in 5–7 days but occasionally will require percutaneous drainage or reoperation for local or systemic sepsis. Perforation should be an extraordinarily rare event.

Suggested Reading

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