Achalasia

Javier Lopez-Gutierrez and B. Mario Cervantes

Introduction

Achalasia is the result of a progressive degeneration process of the ganglion cells of the myenteric plexus, located in the esophageal wall. The disorder motility that characterizes achalasia appears to result primarily from the loss of inhibitory neurons within the wall of the esophagus itself. This loss of the inhibitory innervation in the LOS causes the basal sphincter pressure to rise and renders the sphincter muscle incapable of normal relaxation. The loss of inhibitory neurons from the smooth muscle portion of the esophageal body results in aperistalais [1]. The manifestations of the disease depend on the degree and location of ganglion cell loss [2]. Loss of peristalsis in the distal esophagus and LOS failure to relax with swallowing, both impair esophageal emptying. Most of the signs and symptoms of achalasia are due to the defect in LES relaxation. Esophagogastric junction (OGJ) outflow obstruction. The risk of developing esophageal cancer increases up to 3.3% after a mean symptom duration of 13 years [3].

J. Lopez-Gutierrez (✉)
Minimally Invasive Surgery and Gastrointestinal Endoscopy CMN 20 de Noviembre, ISSSTE CDMX, Mexico City, Mexico

B. M. Cervantes
Minimally Invasive Surgery and Robotic Surgery CMN 20 de Noviembre, ISSSTE CDMX, Mexico city, Mexico

Clinical Features and Manifestations

- Usually has an insidious onset of mild symptoms, with gradual progression through the years.
- Mean duration of symptoms before proper diagnosis is 4.7 years [4].
- The most frequent manifestations are:
  - Dysphagia for solids 91%.
  - Dysphagia for liquids 85%.
  - Regurgitation of undigested food or saliva in up to 91%.
  - Aspiration of retained material in the esophagus.
  - Vomit induction.
  - Difficulty in belching in 85%.
  - Substernal chest pain.
  - Heartburn in 40–60%.
  - Hiccups.
  - In order to overcome the distal obstruction:
- Patients slow down when they eat.
- Adopt specific maneuvers (neck lifting and throwing the shoulders back) in order to enhance esophageal emptying.
  - Mild weight loss.
  - Significant weight loss may suggest malignancy (psuedoachalasia).

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Diagnostic Evaluation

- Clinical History.
- Chest X-Ray.
- Barium Swallow Study.
- OGD:
  - Dilated esophagus.
  - Food debris.
  - Mucosal ulcerations (esophagitis).
  - Mild resistance on passing the endoscope through the union.
- Esophageal manometry. Typical manometric findings are:
  - Aperistalsis in the distal two-thirds of the esophagus.
  - Incomplete LOS relaxation.
  - Elevated resting LOS pressure. The loss of inhibitory neurons may cause resting LOS pressures to rise above 45 mmHg.
  - High-resolution manometry. Achalasia is diagnosed by an elevated median integrated relaxation pressure (IRP), which indicates impaired OGJ relaxation, and absence of normal peristalsis.
- Endoscopic Ultrasound.

Findings Include

- Bird beak sign or rat-tail sign
- Esophageal dilatation.
- Pooling or stasis of barium in the esophagus when the esophagus has become atonic or noncontractile (a late feature in the disease).
- Failure of normal peristalsis to clear the esophagus of barium when the patient is in the recumbent position, with no primary waves identified.
- When the barium column is high enough (with the patient standing), the hydrostatic pressure can overcome the lower esophageal sphincter pressure, allowing passage of esophageal content [5].
- Endoscopic evaluation to exclude malignancy at the esophageal-gastric junction that can mimic achalasia. It may reveal a dilated esophagus with residual material. LOS appearance may range from normal to thickened muscular ring with a rosette configuration on retroflexed view. The contracted LOS may appear with an increase in the passage of the endoscope through the esophagogastric junction. However, endoscope can usually be traversed easily with a gentle pressure of the endoscope. The esophageal mucosa usually appears normal [6]. Some nonspecific changes may be seen. Stasis may predispose to esophageal candidiasis, which may be seen as adherent whitish plaques.

Differential Diagnosis

Achalasia may be misdiagnosed as gastroesophageal reflux disease, especially in patient with chest pain of a burning quality of heartburn. The differential diagnosis includes other motility disorders and pseudo achalasia due to malignancy or Chagas disease.

Treatment

1. **Pharmacological therapy.**
2. **Endoscopic Therapy.**
   (a) **Pneumatic Dilation** [7].
   (b) **Peroral endoscopic myotomy (POEM)** [8].
   (c) **Botulin Toxin (BT) injection** [9].
3. **Surgical Myotomy for Achalasia**.

Dr. Heller described in 1913 a surgical myotomy with a fundoplication as the optimal surgical treatment of achalasia [10]. The effectiveness of symptom control ranges from 90 to 97% of patients [11]. The muscle fibers of the lower esophageal sphincter are incised without disrupting the mucosal lining of the esophagus. The primary goal is to relieve the functional obstruction of the LOS while preventing reflux. Original Heller's technique was modified to anterior myotomy only, and nowadays, is the most common operative procedure to treat achalasia [12]. The esophagus can be approached through the abdomen or thorax.

Patient Selection Criteria The key component for selecting the appropriate patient for surgical management is to differentiate achalasia from other motility disorders, pseudoachalasia, malignancy, and mechanical obstruction.

POEM Vs Heller's Myotomy For patients not willing to have surgical treatment, or have relative surgical contraindications, POEM may be an option. It is an incisionless surgery, using flexible endoscopes. Submucosal tunneling is made, and the dysfunctional circular muscle of the LOS is divided leaving the longitudinal muscle layer intact, which differs from surgical myotomy, where both layers are incised. POEM has an additional margin of safety. However, the incidence of pneumoperitoneum or pneumothorax remains high (up to 40%). More long-term studies are needed in order to appreciate the real advantages and disadvantages compared with Heller's Myotomy, as well as to evaluate the long-term results [14].

Contraindications Patients who prefer to avoid surgery have undergone multiple prior abdominal surgeries or would be unable to tolerate the pneumoperitoneum required for the laparoscopic procedure.

Surgical Technique

Patient Position Patient can be placed in supine, split leg position for optimal ergonomics. Surgeon stands between the legs. The patient is positioned in a steep reverse Trendelenburg position, which allows the stomach and other organs to fall away from the esophageal hiatus.

Abdominal Access and Port Placement We can establish pneumoperitoneum by open Hasson technique, Veress needle, or optical trocar entry. After establishing the pneumoperitoneum, we insert the first port, preferably with an optical trocar. Then, four more ports (two for the surgeon, one for the scope, and the rest for the assistant) are placed under direct laparoscopic vision. Liver retraction should always be considered and can be achieved by one of many different devices available for that purpose (Fig. 1).

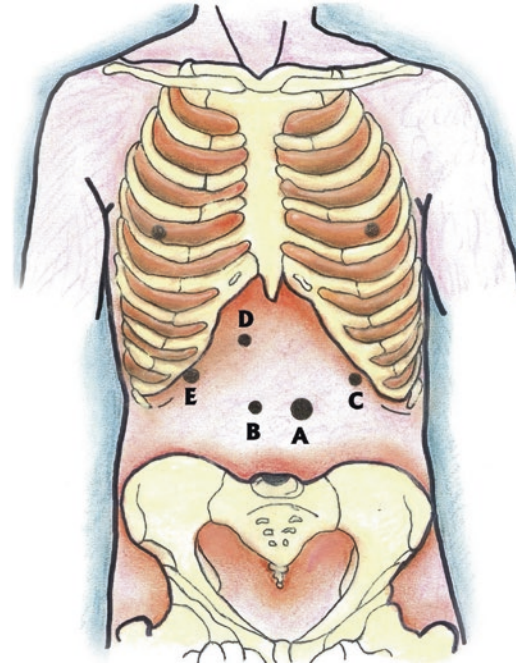

Fig. 1 Port placement

Mobilization of the Gastroesophageal Junction and Proximal Stomach

1. Incise the gastrohepatic ligament (Pars flaccida) in an avascular plane.
2. Preserve the nerve of Latarjet and avoid injury to an accessory or replaced hepatic artery.
3. Divide the anterior phrenoesophageal ligament and the peritoneum overlying the anterior abdominal esophagus.
4. Preserve the anterior vagus nerve, which lies immediately posterior to the right anterior phrenoesophageal ligament.
5. If a posterior partial or a total fundoplication is performed, a posterior esophageal window is created, then divide the left phrenogastric ligaments by dividing the short gastric arteries, starting at the inferior pole of the spleen to the exposed left crus of the diaphragm. In creating this window, the posterior vagus nerve is identi-

fied and protected. If an anterior fundoplication (Dor) technique is used, a posterior esophageal window is unnecessary unless a hiatal hernia and/or a relatively short esophagus is encountered and there is a need for further mobilization to allow more intra-abdominal length in order to construct a proper fundoplication.

Mobilization of the Mediastinal Oesophagus The distal portion of the mediastinal esophagus is mobilized to achieve sufficient length to perform a myotomy incision that divides the entire length of the LOS and permits a tension-free fundoplication.

Myotomy When performing the myotomy, it is essential to have adequate visualization and exposure in order to prevent mucosa injuries.

1. The cardioesophageal fat pad and the anterior vagus must be cleared from the esophagus and the OGJ.
2. Once cleared, a myotomy is performed on the esophagus and the stomach. This is done using a grasper in the left hand and Maryland forceps in the right hand. The muscles are gently split layer by layer till the submucosa is clearly seen this would help avoid injury to the mucosa.
3. It is useful to have a stable platform and lighting. A lighted bougie may be used, or even better, an endoscope, in order to illuminate, stretch the muscle fibers by insufflation, and, therefore, facilitate their division.
4. The anterior surface of the esophagus is completely exposed, and slight tension is created by retracting caudally with a Babcock retractor.
5. The incision may be started on the stomach or the esophagus.
6. The myotomy is performed by individual dividing the esophageal and gastric muscle fibers. Longitudinal muscle fibers are divided first, which exposes the underlying circular muscles (Fig. 2).
7. Division of the circular layer reveals a bulging mucosa plane that should appear smooth and white. The esophageal portion of the myotomy should be approximately 6 cm in length.
8. The most critical and challenging factor is to create a 3 cm myotomy caudal to the OGJ, where the tissue plane becomes less readily identifiable. A careful layer-by-layer dissection helps prevent injury.
9. The total length of the myotomy should be 9 cm.
10. It is highly advisable to perform an endoscopic inspection of the mucosa, before the next steps, in order to identify and repair any mucosal perforations.
11. Perform a hiatal closure and when possible, it is advisable to perform a fundoplication procedure, partial or total. Please refer to the fundoplication chapter for further details (Fig. 3).
12. During fundoplication take care to do the following: Place an inner row of interrupted sutures to secure the medial aspect of the fundus to the left side of the myotomy. A second row of interrupted sutures is placed to fix the leading edge to the right side of the myotomy.

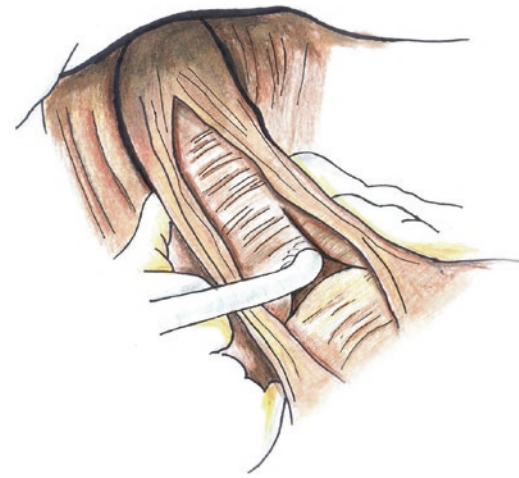

Fig. 2 Esophageal myotomy

Intraoperative Technical Risks

- Esophageal or gastric perforation—It ranges from 10 to 16%. The mucosal perforations, when adverted, should be repaired with 4–0 or 5–0 absorbable monofilament suture. The Dor fundoplication will buttress the repair.
- Division of vagus nerve—It is rare. If an injury to the anterior or posterior vagus nerve occurs, it is not repaired.
- Splenic injury—Ranges from 1 to 5%.
- Pneumothorax.

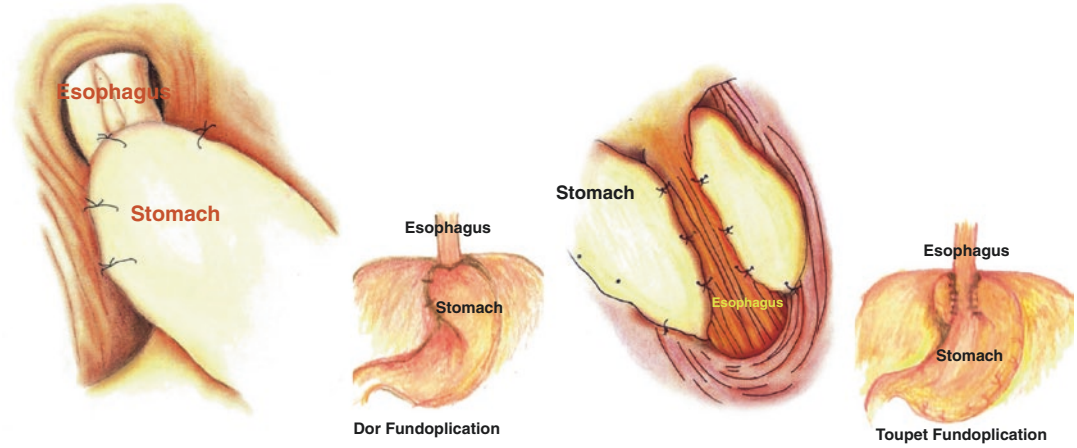

Fig. 3 Partial fundoplication

Postoperative Complications

The morbidity rate ranges between 1 and 10%. The mortality rate is <0.1% in the first 30 days after the procedure [15, 16].

Perforation It is the most common early postoperative complication and occurs in 1–7% of patients [17]. Late perforations usually result from either direct unrecognized mucosal injury or inadvertent thermal injury. Perforations may result in peritonitis or mediastinitis, or both, and may be life-threatening [16]. When a perforation is suspected, a water-soluble contrast radiograph should be obtained. Thoracic and abdominal CT scan with oral water-soluble contrast may show liquid extravasation and collections on abdomen and/or thorax. OGD is controversial. Once the perforation is confirmed, early reexploration is advisable with lavage and drainage placement. Primary repair may have acceptable results if performed in the first 24 hours after the perforation was produced [18].

Recurrent Dysphagia Is usually a late complication of a Heller myotomy + fundoplication. It presents in 3–10% of patients [19, 20]. The most common cause is incomplete myotomy. Is more common in patients that underwent thoracic approach [21]. Other reasons for dysphagia include herniated fundoplication, perihiatal scarring, peptic stricture, and tumors.

GORD If the patient underwent only a myotomy, the rate of GORD is higher [13]. If this happened despite a fundoplication, they should be treated medically.

Other Complications Bleeding is a rare complication and is reported in approximately 3% of patients.

Postoperative Care

- Analgesics.
- Antiemetics. Very important in order to avoid nausea and retching that may increase pressure on the myotomy, increasing the risk of complications.
- Clear liquids may be started the night of the day of the procedure, or when bowel function returns.
- If the patient does not present dysphagia after liquids, diet may be advanced to a soft diet the following day.
- If the patient develops early symptoms suggestive of perforation, a contrast X-ray should be considered. Symptoms include chest pain, epigastric pain, fever, tachycardia, emphysema (subcutaneous or mediastinal), and leukocytosis.

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