Motility Disorders

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7.1 Introduction

Though a relatively rare disease (with an estimated prevalence of 0.5–1 new cases per 100,000 population a year, and with no clear age predilection), achalasia is the most common motor disorder of the esophagus. Its pathophysiology consists of the loss of esophageal body peristalsis and an impaired lower esophageal sphincter (LES) relaxation, resulting in a residual pressure gradient between the esophagus and the stomach during swallowing, which gives rise to a functional obstruction at the gastro-esophageal junction.

It is widely accepted that the partial or total absence of swallow-induced LES relaxation (the main functional anomaly in achalasia) is caused by a loss of the inhibitory innervation in the myenteric plexus; the exact mechanism behind this loss of inhibitory neurons is far from clear and treatment is still limited to mechanical or surgical disruption of the LES.

7.2 Indications

Dysphagia, for both solids and liquids, is the main symptom of achalasia and the swallowing behavior of achalasia patients (especially those with a dilated esophagus) is unique: they often make food pass through the cardia by drinking large quantities while eating, thus increasing the pressure in the gullet enough to overcome the LES resting pressure. Undigested food regurgitation occurs, especially at night, with corollary symptoms that may include respiratory complications (nocturnal cough and aspiration), chest pain, and weight loss.

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Although the above-mentioned symptoms may seem rather obvious and diagnostic evaluation has certainly improved, there is often still a considerable delay between the onset of symptoms and the diagnosis of achalasia, due not to an atypical clinical presentation of the disease, but to the physician's misinterpretation of its typical signs and symptoms. Patients are often treated for suspected reflux disease, they are sometimes given sedatives and may even be referred to a psychologist.

There is a lively debate regarding the most effective treatment for long-term symptom relief. The relative rarity of achalasia means that most patients are treated according to local preferences and expertise. First-line achalasia treatment has traditionally been pneumatic dilation, reserving surgery for patients requiring repeated dilations or when this treatment fails. Currently, minimally invasive surgery – with its more limited related morbidity – is progressively employed in patients with achalasia.

Non-decompensated achalasia (grades I to III according to the radiological classification), i.e., with a maximum esophageal diameter of about 6 cm and a gullet that is still "straight" is suitable for laparoscopic myotomy. In cases of mega-esophagus (>6 cm in diameter and/or a sigmoid shape), laparoscopic myotomy can still be performed, but the success rate is lower; it may still be an option, nonetheless, before scheduling a patient for esophagectomy.

7.3 Alternative Treatment

Pneumatic dilation is a good alternative to surgery. It aims to disrupt the LES by forcefully dilating it with an air-filled balloon. To date, the most commonly used is the Rigiflex balloon (Microvasive, Boston Scientific, Cedex, France), which is available in three sizes (30, 35, and 40 mm in diameter). Briefly, the balloon is inserted over an endoscopically maneuvered guide wire, positioned across the LES, and inflated under endoscopic visualization. The immediate success rate of this procedure is 55–70 % with a single dilation,

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but this figure can be improved with multiple dilations. The best results can be obtained by using increasing balloon sizes in a stepwise fashion over several months. The long-term results may be less positive, however, with about 60 % of patients still symptom-free after a year, but more than 50 % experiencing recurrent symptoms after 5 years. One of the major risk factors for recurrence is young age (<40 years old), with a 5-year remission rate of only 16 % for patients under 40, as opposed to about 60 % for older patients. Other predictors of failure after pneumatic dilation are male gender, a single dilation with a 30 mm balloon, and a post-treatment LES pressure >10 mmHg.

Esophageal perforation is the most important and potentially life-threatening complication of pneumatic dilation, occurring in up to 6.6 % of the patients. Other potential complications following pneumatic dilation include prolonged pain, gastroesophageal reflux, aspiration pneumonia, gastrointestinal hemorrhage, esophageal mucosal tears without perforation, and intramural esophageal hematoma.

7.4 Preoperative Work-Up

The conventional diagnostic work-up is based on functional studies, such as esophageal manometry and barium swallow. The radiographic features are esophageal dilation and minimal LES opening with a bird's beak appearance, sometimes with an air-fluid level in the gullet and no intragastric air bubble. These radiographic features may be missed by a conventional test, especially in the early stage of the disease. A "timed" barium swallow test has been proposed and is widely used in evaluating patients before and after treatment. A fixed amount of barium (200 ml) is ingested in 2 min, and pictures are taken after fixed intervals (0, 1′, 2′, 5′) to measure the height of the barium column at various times. At manometry, the typical findings are aperistalsis of the esophageal body and incomplete LES relaxation, sometimes with a high intra-esophageal pressure due to the stasis of food and saliva. LES resting tone is often elevated. Swallowing may trigger simultaneous low-amplitude pressure waves with a similar morphology in all channels in the esophageal body, called "common-cavity" waves.

Endoscopy is usually the first test to be performed in a patient with dysphagia. Findings may seem normal in patients with achalasia, especially in the early stages, when the gullet is only mildly dilated. Esophagitis ("stasis" esophagitis) may be identified and should not be confused with reflux esophagitis. Esophageal candidiasis, resistant to the usual treatments, can also be found, and is usually related to the functional obstruction. Malignant tumors can produce an achalasia-like syndrome called "pseudoachalasia" by infiltrating the gastro-esophageal junction and mimicking the clinical and manometric presentation of achalasia; they account for about 5 % of cases of misdiagnosis. In general, patients with pseudoachalasia are older and have a shorter history of dysphagia and weight loss. Endoscopy, with a careful examination of the cardiac and fundic region, is therefore mandatory as part of the diagnosis work-up to avoid this potential pitfall, and if the clinical suspicion is strong, computerized tomography (CT) and endoscopic ultrasound should be considered. These latter tests should be considered, particularly in elderly patients with symptoms of recent onset.

Finally, all of the tests routinely performed before surgery under general anesthesia (blood tests, chest X-rays, EKG) are required. Additional tests may be requested by the anesthesiologist for particular patients, but are rarely necessary.

7.5 Operating Room

The operation is performed under general anesthesia and oro-tracheal intubation. The patient is placed supine on the operating table with legs abducted on flat padded leg boards to minimize the risk of lower extremity neurovascular injury. Alternatively, the patient can be positioned in simple supine position, i.e. with the legs together on the operating table.

The right arm is tucked against the patient's side and the left arm remains on an arm board (Fig. 7.1). The patient should be well secured, as a steep reverse Trendelenburg position is needed to displace the intra-abdominal organs from the subdiaphragmatic area and bring the surgical site closer to the surgeon, who stands between the abducted legs to gain easy access to the upper abdomen. This position demands all the measures usually adopted to prevent deep vein thrombosis (stockings, heparin prophylaxis). Due to the brevity of the operation, a Foley catheter is unnecessary, unless it is requested by the anesthesiologist in the case of frail patients.

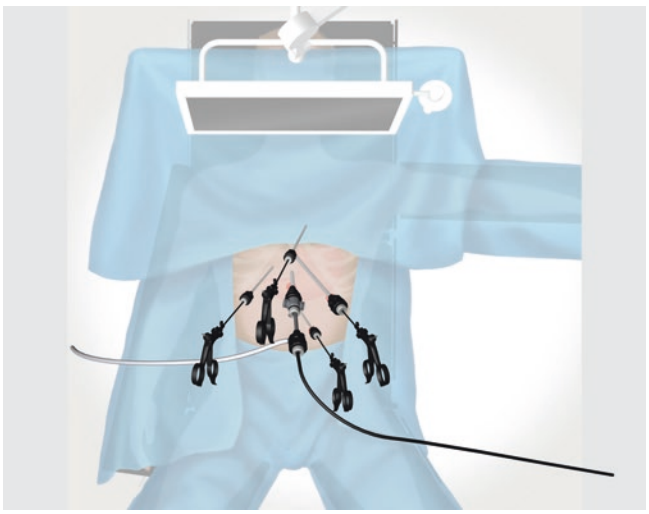

Fig. 7.1 Patient positioning for Heller's myotomy

The poor emptying of the achalasic esophagus makes the gullet retain saliva and ingested food, so there is a high risk of regurgitation and aspiration during the induction of anesthesia in these patients. Patients should be kept on a liquid diet for 48 h before the operation, and a dilated gullet should be mechanically washed and emptied via a naso-esophageal tube the night before the procedure.

The standard laparoscopic instrumentation is required: two 12 mm trocars and three 5 mm trocars are used; a 30° laparoscope offers the best view of the cardiac region; a device for lifting the left liver lobe, atraumatic forceps for pulling down the stomach, and a cautery hook and scissors are the instruments needed for the procedure, plus a couple of standard forceps for tissue handling and a needle holder for suturing. An ultrasonic scalpel or Ligasure are usually not necessary. Small bipolar cautering forceps may be useful to control bleeding from the edges of the myotomy.

During the performance of the myotomy the endoscope may be useful to facilitate the procedure and check for any mucosal lesions. We prefer to position a guidewire across the cardia through the scope before starting the operation: during the myotomy, a 3.0 cm Rigiflex balloon is placed across the cardia and gently inflated and deflated.

7.6 Surgical Technique

7.6.1 Trocar Positioning (Fig. 7.2)

We usually prefer the open technique when performing the pneumoperitoneum: the first 12 mm blunt trocar, used for the laparoscope, is inserted along the midline, halfway between the umbilicus and the xyphoid. A 5 mm trocar is inserted as laterally as possible on the patient's right side, immediately below the costal margin, to lift the left liver lobe. A 5 mm trocar inserted immediately below the xyphoid is used for the operator's left hand, while a 12 mm trocar inserted laterally below the left costal margin provides access for the surgeon's right hand. Finally, a 5 mm trocar, at the same level as the first trocar and on the midclavicular line, is used to pull down the gastric fundus.

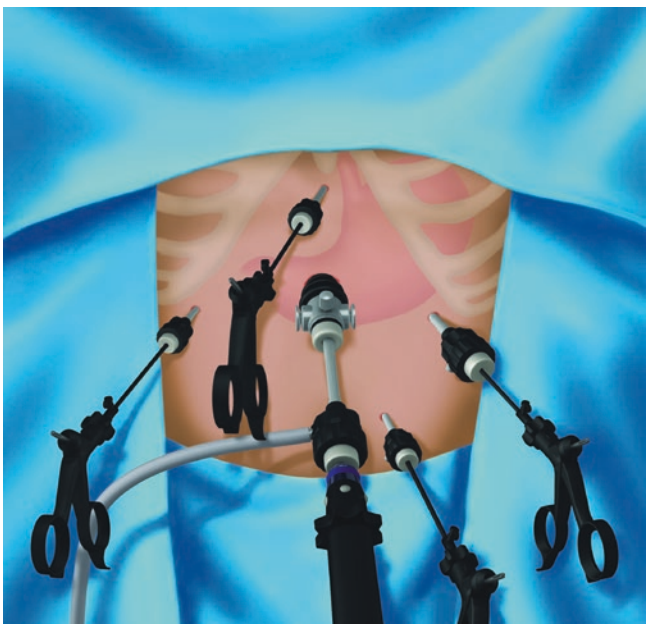

Fig. 7.2 Trocar positioning for Heller's myotomy

7.6.2 Exposure of the Anterior Wall of the Esophagus

An assistant on the surgeon's left-hand side lifts the left liver lobe using an atraumatic retractor, thus exposing the cardia region. It is not necessary to divide the left triangular ligament. An assistant on the surgeon's right-hand side grasps the gastric fundus with atraumatic forceps, maintaining a caudal traction on the esophagogastric junction. The operation begins with a minimal dissection of the anterior part of the esophagus. With the cautery hook, the peritoneum over the esophagogastric junction is divided to expose the anterior wall of the gullet. Any adipose tissue (usually found at this level) is removed, paying attention to the small vessels coming from the gastric wall, which should be coagulated with the bipolar forceps; they usually mark the inferior limit of the myotomy. The left vagus nerve, which becomes anterior at this level by crossing the anterior esophageal wall from left to right, is clearly evident and must not be damaged.

7.6.3 Myotomy

The myotomy is started with the cautery hook on the dilated part of the distal esophagus, above the lower esophageal sphincter that, in cases of long-standing achalasia, is often marked by a whitish, sclerotic area. The myotomy must in any case be started 2 cm above the esophagogastric junction to expose the esophageal submucosal layer with the least risk of perforation. The cautery power is reduced to 15 W to avoid transmitting its coagulating effect to the underlying mucosa. First the longitudinal muscle fibers are hooked, lifted, and coagulated, until the circular ones are exposed; then the latter are hooked, lifted, and divided using the same technique. Then the submucosal layer, which forms a slight bulge between the two margins of the myotomy, is exposed. At this point, using the forceps in the left hand, the margin of the myotomy is delicately lifted, and small scissors are used to bluntly dissect the muscle layer from the submucosal layer. The muscle tissue is then cut and minor bleeding from the edges of the myotomy can be controlled with a careful

use of the cauterized scissors, or simply with the aid of mechanical compression using a small sponge. A myotomy 6–8 cm long is performed, extending it 1.5–2 cm on the gastric side below the oblique muscle fibers that represent the beginning of the gastric muscle, thus exposing the gastric submucosa, which is usually more vascularized than the esophageal mucosa (Fig. 7.3.). Scissors are used in the proximal part and a hook cautery is inserted downwards on the gastric side to lift and divide the circular muscle fibers. Care must be taken during the myotomy to avoid injuring the anterior vagus nerve and prevent any esophageal perforation or spiraling of the myotomy. A feature of our personal technique involves the intraoperative use of a 30 mm Rigiflex balloon. The balloon is placed inside the esophageal cavity at cardia level using an endoscopically-positioned guide wire. During the myotomy, the balloon is gently inflated and deflated with 40–60 cc of air using a syringe: this exposes the circular fibers, which can then be stretched and easily cut or torn apart (Fig. 7.3). The edges of the myotomy are separated and peeled away from the submucosal plane: any minimal bleeding from submucosal vessels is easily controlled by inflating the balloon, thus reducing the need to use the cautery. At completion of the myotomy, a flexible endoscope can be inserted to confirm sufficient dilatation and detect any perforation of the mucosa. Placing the patient in Trendelenburg position during endoscopic inspection and instilling the upper abdomen with saline allows recognition of a small perforation by the presence of air bubbles during endoscopic insufflation.

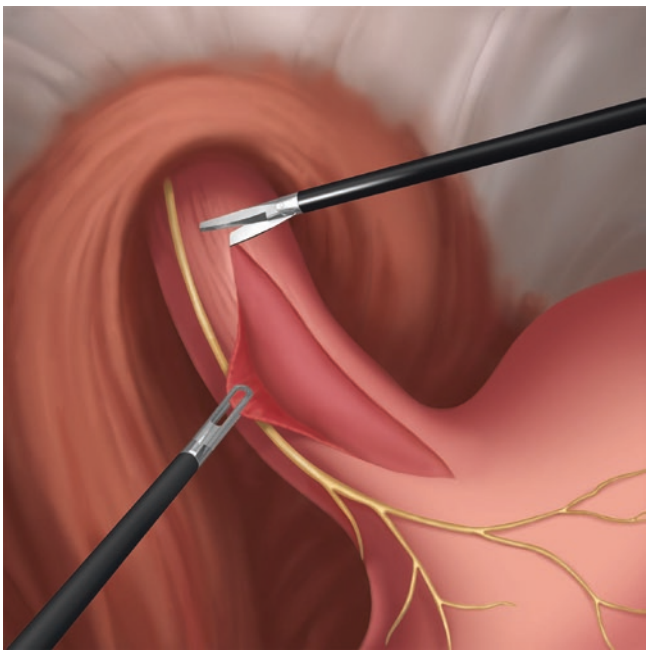

Fig. 7.3 The myotomy is being performed. The anterior vagus nerve must be identified and preserved from damage

7.6.4 Risks of the Myotomy

The main risk in performing esophageal myotomy is a mucosal lesion (perforation). This can be caused directly by an erroneous handling of the hook or scissors, or by the endoscope while checking the myotomy, or even by the Rigiflex balloon if it is inflated too vigorously. Indirect lesions can be caused by an excessive coagulation involving the mucosa, in which case the perforation occurs when the eschar falls out, 48–72 h after the operation. Direct lesions are usually detected during the operation and can be sutured directly with 4–0 interrupted reabsorbable stitches. This is usually done laparoscopically, but conversion to open surgery may be necessary. The suture line is then further protected with the anterior fundoplication. In the case of small indirect lesions, subsequently detected by Gastrografin swallow, a conservative treatment with gastric aspiration, NPO, TPN, and antibiotics usually suffices.

7.6.5 Antireflux Fundoplication

Although some authors perform only the myotomy as described above, a fundoplication is normally added to prevent postoperative gastroesophageal reflux disease, which may be a severe complication in patients with a poor esophageal clearing ability due to the absence of peristalsis. Given this lack of any propulsive peristaltic activity of the esophageal body, a partial fundoplication is usually performed, either anteriorly (Dor) or posteriorly (Toupet). The former has the advantage of protecting the exposed esophageal mucosa and can be performed without completely mobilizing the esophagus, thus preserving the natural antireflux mechanisms.

Using our technique, an anterior partial hemifundoplication according to Dor is added to the myotomy. In general, there is no need to mobilize the gastric fundus by dividing the short gastric vessels. Three stitches are inserted on each side, the proximal one to include the stomach, the edge of the myotomy and the diaphragm (Fig. 7.4).

If a posterior hemifundoplication (Toupet's procedure) is to be performed, the abdominal esophagus must be completely mobilized, and the gastric wrap is passed behind the esophagus: the right and left sides of the wrap are secured to the corresponding edges of the myotomy with three to four interrupted non-reabsorbable stitches.

7.6.6 Postoperative Care

A naso gastric tube is carefully positioned at the end of the operation. A water-soluble contrast esophagogram (Gastrografin) is obtained on the first postoperative day to

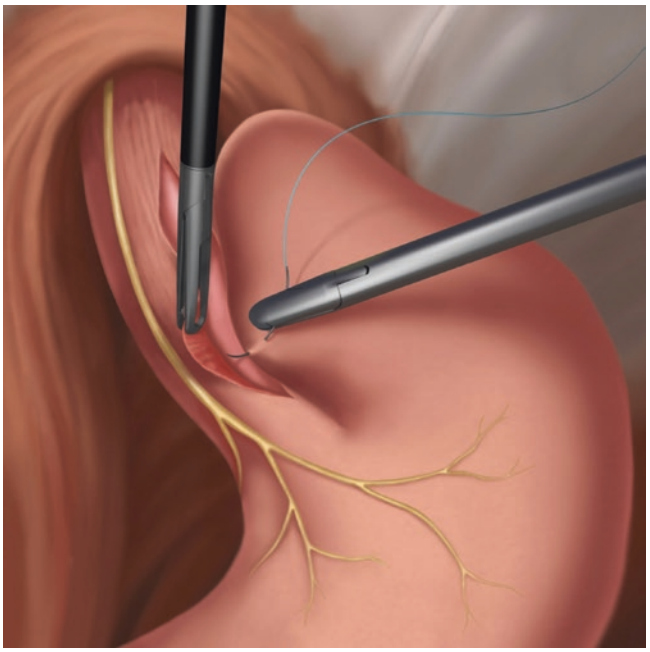

Fig. 7.4 A partial anterior fundoplication using the gastric fundus is added to the myotomy. Usually three stitches per side, securing the wrap to the myotomy edges, are needed, the most proximal ones also include the corresponding diaphragmatic pillar

rule out any mucosal perforation. In case of small perforation the NG tube is kept in position and the patients is maintained in parenteral nutrition for 6–8 days. If a large perforation is detected and the contrast freely diffuses in the abdomen or in the pleura, drainage and immediate suture are reccomended.

If no perforation are observed a liquid diet is started and the patient is discharged after another 24–48 h, once a soft diet has been started and depending on how far away from the hospital they live. A soft diet is recommended for 8–10 days, after which a normal diet is allowed.

Patients usually return to the outpatient clinic 1 month later for a barium swallow. Endoscopy and function tests are performed after 6 months to rule out any postoperative GERD. After that, endoscopy is recommended every 2 years to rule out any cancer growth.

Conclusions

Surgical myotomy is the most effective and durable treatment for achalasia, and since laparoscopy has reduced related morbidity, laparoscopic Heller myotomy has become the first-line treatment for this condition. A partial fundoplication should be performed in conjunction with the myotomy to minimize the risk of postoperative gastroesophageal reflux, a risk that must be avoided as far as possible in an esophagus with poor clearance capabilities. Although it is not clear which fundoplication is best in association with laparoscopic Heller myotomy (a prospective randomized trials is eagerly awaited), we have found the laparoscopic Heller-Dor combination a safe and effective treatment for achalasia.

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