Boerhaave's Syndrome

38

Gereon Gäbelein, Christoph Benckert, Uwe Eichfeld, and Sven Jonas

Introduction

Boerhaave's syndrome describes a spontaneous rupture of the esophageal wall due to a sudden increase in intraesophageal pressure combined with negative intrathoracic pressure caused by forceful retching and projectile vomiting. The most common anatomic location of the tear is the left posterolateral wall of the distal third of the esophagus (Korn 2007). Boerhaave's syndrome is associated with a high risk of severe complications and mortality and is fatal in the absence of therapy. The occasionally nonspecific nature of the symptoms may contribute to a delay in diagnosis and a poor outcome.

The syndrome was first documented in the eighteenth century by the physician Herman Boerhaave, after whom it is named (Boerhaave 1724). In contrast to Mallory-Weiss syndrome, in which only the mucosal layer is torn and bleeding complications are predominant, the entire esophageal wall is ruptured in Boerhaave's syndrome. The classical history and primary clinical findings are excruciating retrosternal chest or upper abdominal pain, subcutaneous emphysema with crepitation, and preceding vomiting, also known as Mackler's triad. Subsequently, odynophagia, dyspnea, cyanosis, fever, and shock may develop rapidly. Pain occasionally may radiate to the left shoulder, leading some physicians to confuse an esophageal perforation with a myocardial infarction. Other common misdiagnoses include pancreatitis, lung abscess, pericarditis, and spontaneous pneumothorax. Most patients suffer from alcohol withdrawal, and presentation during an episode of heavy drinking may disguise symptoms and further delay diagnosis (Curci 1976).

G. Gäbelein • U. Eichfeld • S. Jonas (✉)
Department of Visceral, Transplantation, Thoracic and Vascular Surgery, University Hospital of Leipzig, Leipzig, Germany
e-mail: sven.jonas@uniklinik-leipzig.de

C. Benckert
Clinic for General, Visceral and Vascular Surgery, University Clinic Magdeburg, Magdeburg, Germany

If esophageal perforation is suspected, even in the absence of physical findings, contrast radiographic studies of the esophagus and a CT scan should be obtained promptly. The initial plain chest radiograph is almost always abnormal and usually reveals mediastinal or free peritoneal air. With cervical esophageal perforations, plain films of the neck show air in the soft tissues of the prevertebral space. Pleural effusion with or without pneumothorax, a widened mediastinum, and subcutaneous emphysema typically are seen. The CT scan may show esophageal wall edema and thickening; extraesophageal air; periesophageal fluid, frequently but not exclusively gas bubbles; mediastinal widening; and air and fluid in the pleural spaces or retroperitoneum. The diagnosis of esophageal perforation also may be confirmed by a water-soluble contrast esophagram, which reveals the location and extent of extravasation of contrast material. Although barium is superior in demonstrating small perforations, spillage of barium sulfate into the mediastinal and pleural cavities may cause an inflammatory response and subsequent fibrosis; therefore, it is not used as the primary diagnostic tool. Endoscopy has a minor role in diagnosing spontaneous esophageal perforation. Insufflation of air may extend the perforation and introduce air and soilage into the mediastinum.

Conservative measures seem feasible especially, if patients are diagnosed after 48 h and are not septic. In these cases, Boerhaave's syndrome can be controlled medically and by endoscopic or radiologic interventions consisting of the application of antibiotics, insertion of metallic endoprosthesis, and percutaneous drainage of abscesses (de Schipper 2009). Generally, surgery is the treatment of choice. If there are signs of severe sepsis, there is no wide-mouthed cavity draining freely back into the esophagus, or the pleural or abdominal space is contaminated, surgical therapy is mandatory. The earlier surgery is arranged, the more likely conditions will be sufficient to perform primary closure, if possible with local reinforcement, because of the limited extent of contamination and inflammation of the surrounding regions. In the absence of phlegmon or implacable obstruction, primary repair offers excellent results. The surgical approach to

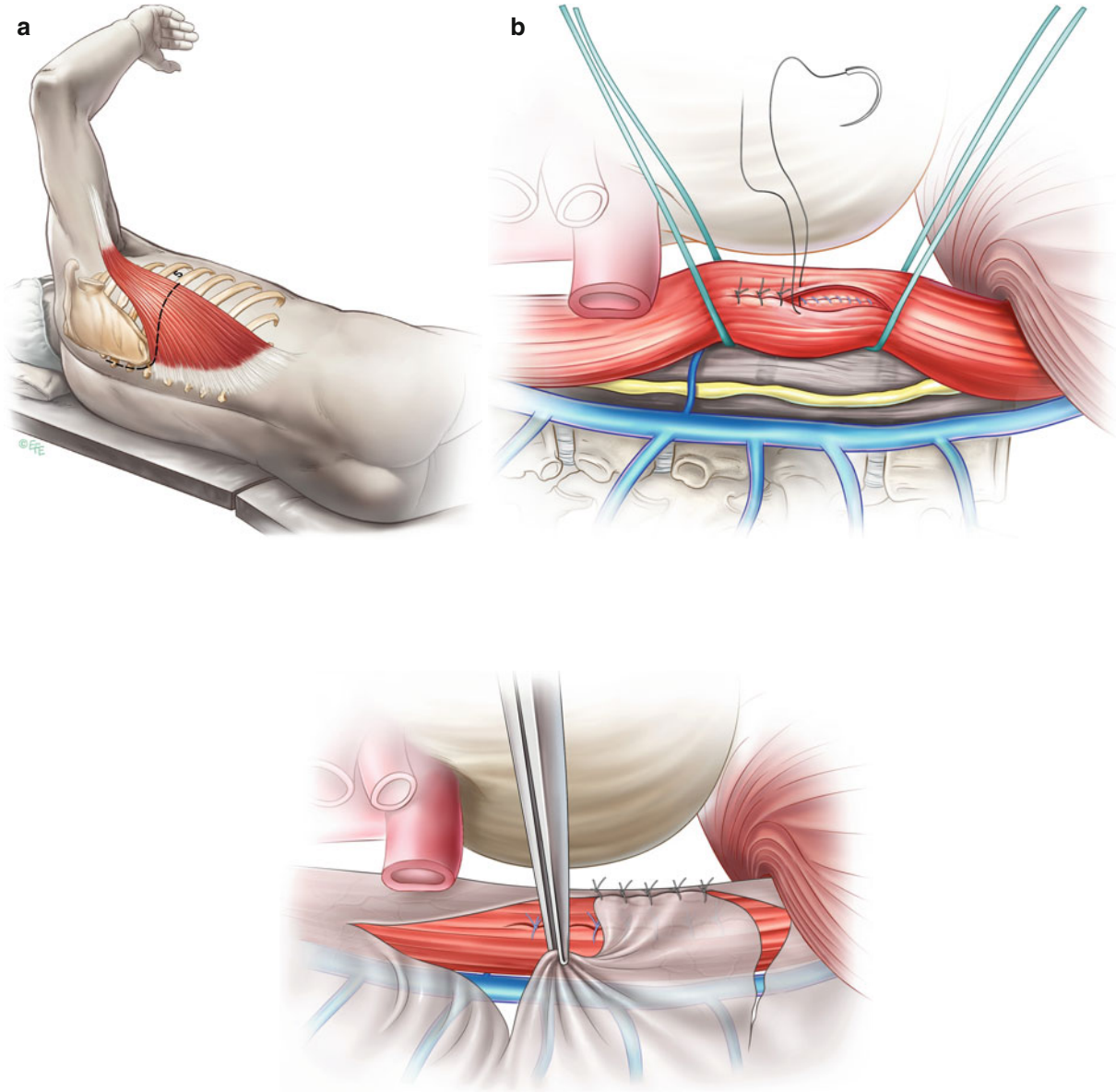
H.C. Dienemann et al. (eds.), *Chest Surgery*, Springer Surgery Atlas Series,
DOI 10.1007/978-3-642-12044-2_38, © Springer-Verlag Berlin Heidelberg 2015

remedy the defect is an individual decision. Resection and reconstruction are the best choices in circumstances in which a significant phlegmon or distal obstruction renders primary repair hazardous or inapplicable. Diversion, preferably with proximal and distal esophageal exclusion, may be necessary for patients too ill to undergo more formidable surgery. Even with early surgical intervention (within 24 h), the risk of death is high.

Figures 38.1 and 38.2

If the extent of contamination and inflammation of the surrounding regions is limited and it is assumed to be localized within the chest, right thoracotomy, necrosectomy, and primary closure of the perforation should be performed, if possible with local reinforcement. A double-lumen endotracheal tube is required for single-lung ventilation. A broad-spectrum antibiotic should be given before skin incision. Because of uncontrollable conditions due to the perforation, no nasogastric tube should be inserted before surgery. The patient is placed in the typical left lateral decubitus position. The table is flexed at the hips, and the right arm is adequately supported. All pressure points are carefully padded. The surgeon stands on the right, at the back of the patient, with the first assistant opposite. Dorsolateral thoracotomy is performed. The incision is carried from the anterior edge of the latissimus dorsi muscle at the level of the mamilla surrounding the lower scapula edge upward in parallel to the paravertebral muscles. The latissimus dorsi muscle is dissected, and the scapula and anterior serratus muscle are lifted off the chest wall. Above the fifth rib, the intercostal muscles and pleura are dissected, and the rib spreader is placed and opened carefully. After exploration of the chest space, mobilization of the esophagus, and assessment of the wound edges, the indication for primary closure must be reevaluated. If the extent of contamination and inflammation of the surrounding regions is limited, a primary closure of the perforation should be performed. At this point, it is possible to move the nasogastric tube within the esophagus, inspecting the esophagus while leading and passing the tube across the perforation. The wound may be reapproximated by a running suture placed only in the mucosa. A second layer of interrupted sutures is placed in the muscular wall of the esophagus. The closure may be reinforced by a pleural flap, which may be used to cover the sutures. The adjacent mediastinal pleura is incised widely to ensure adequate drainage into the pleural space, which in turn is drained in the usual manner. The fascia and skin are closed via standard techniques. If there is additional involvement of the abdominal space, subsequent laparoscopy with exploration, lavage, and selected placement of drains is indicated. (**a**) Approach. (**b**) Closure of the lesion

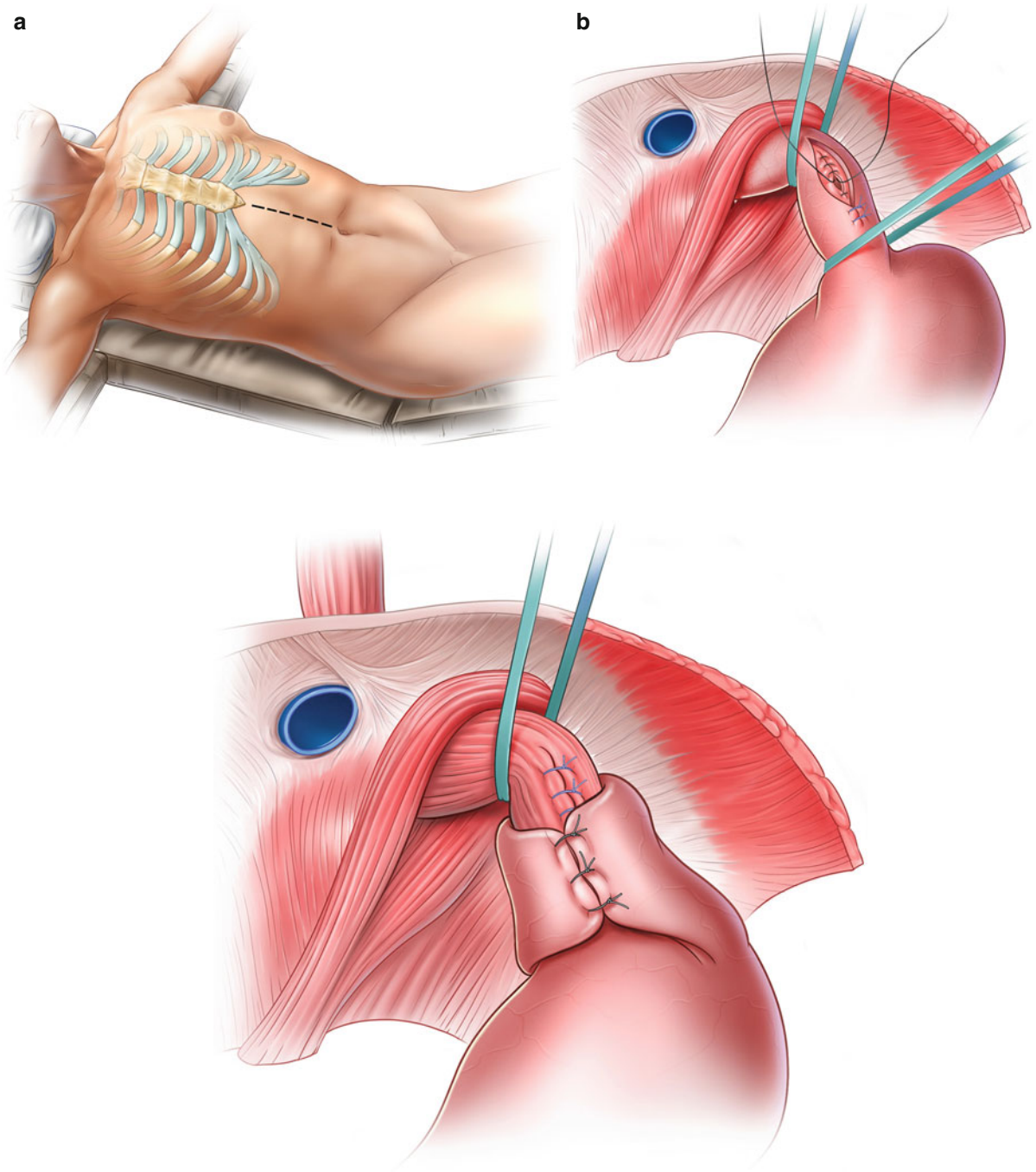
Figures 38.1 and 38.2



Figures 38.3 and 38.4

If the extent of contamination and inflammation of the surrounding regions is limited and assumed to be localized in the abdominal space, median laparotomy, necrosectomy, and primary closure of the perforation with a (hemi)fundoplication to cover the suture should be performed. A broad-spectrum antibiotic should be given before skin incision. Because of uncontrollable conditions due to the perforation, no nasogastric tube should be inserted before surgery. Under general anesthesia, the patient is placed in the typical supine decubitus position. The table is slightly flexed at the hips with the arms extended at the side. All pressure points are carefully padded. The surgeon stands on the right, the first assistant opposite. Median laparotomy is performed. The incision is carried from the xiphoid process down to the navel. The fascia and peritoneum are dissected carefully, and a Rochard retractor is placed. After exploration of the abdominal space and mobilization of the left liver lobe, the gastrohepatic ligament is dissected and divided. Care should be taken to identify and avoid dissection of an aberrant left hepatic artery within the gastrohepatic ligament. Next, the crura of the diaphragm are exposed at the gastroesophageal junction. While assessing the wound edges, the surgeon must reevaluate the indication for primary closure. The space between the esophagus and the crura is opened bluntly. The retroesophageal space is dissected under direct vision. The distal esophagus should be mobilized until the whole perforation is exposed. At this point, it is possible to move the nasogastric tube within the esophagus, inspecting the esophagus while leading and passing the tube across the perforation. If the extent of contamination and inflammation of the surrounding regions is limited, primary closure of the perforation should be performed. The wound may be reapproximated by a running suture placed only in the mucosa. A second layer of interrupted sutures is placed in the muscular wall of the esophagus. The adjacent mediastinum should be opened enough to ensure adequate drainage into the abdominal space, which in turn is drained in the usual manner. (Hemi)fundoplication is used to cover the suture. To achieve a tension-free wrap, it is necessary to mobilize the gastric fundus. The fundus is pulled gently across the back of the gastroesophageal junction. With the same segment of stomach pulled around the esophagus, a 360° or 270° wrap is created and fixed in a length of 2–3 cm and should be located and fixed over the suture of the perforation. Lavage of the whole abdominal space is necessary, and the hiatus should be drained. The fascia and skin are closed using standard techniques. If there is further involvement of the thoracic space, subsequent thoracoscopy with exploration, lavage, and selected placement of drains is indicated. (**a**) Approach. (**b**) Closure of the lesion

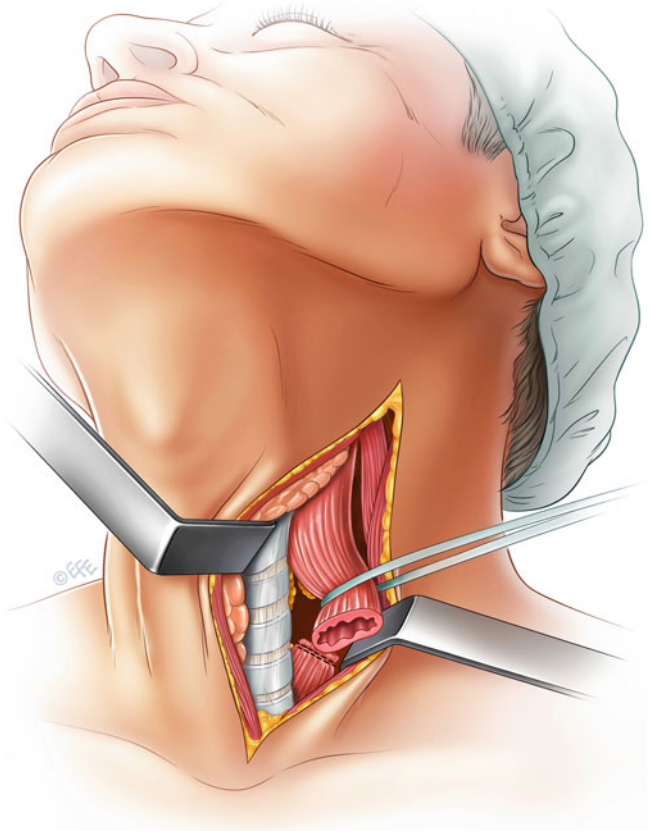
Figures 38.3 and 38.4

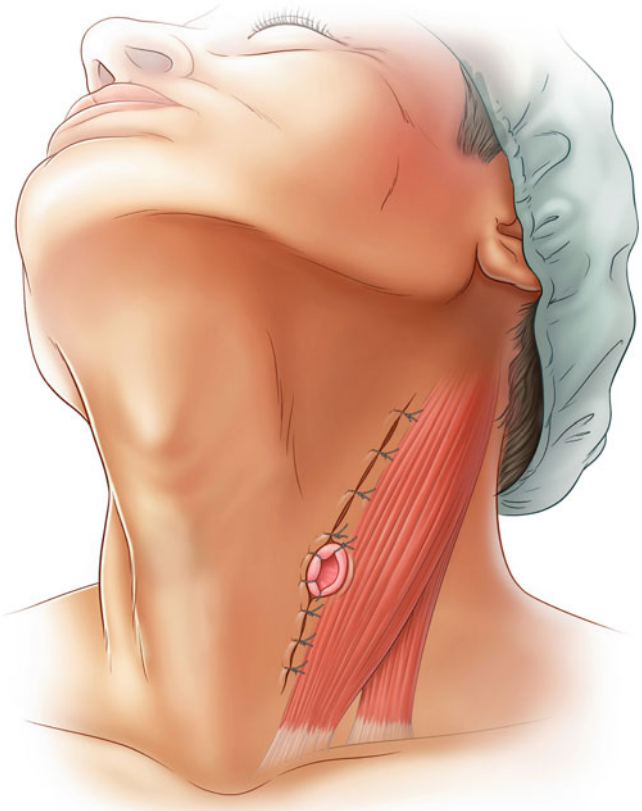


Figures 38.5 and 38.6

If the perforation has been present longer than 24 h, survival decreases. In this setting, primary closure of the perforation may carry a high risk of insufficiency, with the need for further interventions and an additional hazard to the patient. In situations in which the esophagus is destroyed beyond the possibility of primary repair and reestablishment of in situ continuity, the involved segment of esophagus must be removed completely with restoration, at a later date, of pharyngogastric continuity with an interposed segment of bowel or stomach. When the esophagus is being resected, the upper segment should be left as long as possible. Resection of the involved segment of esophagus must be attempted, or at the very least, the area must be drained and the esophagus diverted proximally by collar esophagostomy. In addition, tube gastrostomy and feeding jejunostomy should be performed. This may be the only life-saving strategy. The decision is individualized based on the size of the perforation, the extent of tissue devitalization, and the patient's associated general state and conditions. Rapid control and prevention of ongoing soilage and sepsis must have top priority and are the only hope for survival. Thoracotomy must be attempted to identify the esophageal perforation for débridement of necrotic debris and for resection of the affected site. The upper esophagus must be mobilized as high as possible in the neck. In the case of larger injuries or severely inflamed tissue, wide and extensive drainage is performed after esophagectomy. Drains, as well as chest tubes, are positioned strategically to provide drainage of both the mediastinum and pleural space. For collar esophagostomy, it is helpful to have a nasogastric tube in the esophagus. If one has a choice, it is easier to approach the esophagus from the left side because it tends to lie to the left of the midline; however, approach from the right also is possible. An oblique incision, approximately 5 cm long, is made along the anterior border of the sternocleidomastoid muscle 2–3 cm above the clavicle. The anterior border of the muscle is retracted laterally, and the strap muscles are retracted medially. The carotid sheath is retracted laterally. The thyroid gland, vessels, and recurrent laryngeal nerve are retracted anteriorly and medially by dissecting the prevertebral muscles and fascia toward the midline. The recurrent laryngeal nerve is not exposed purposely, but gentle blunt dissection is necessary to avoid injuring it. The previously placed tube in the esophagus is palpable by a finger inserted in the wound. The esophagus is mobilized enough to dissect its wall with a stapler suture, if this was not already done during thoracic resection of the esophagus. The aboral ending, if not already resected, is closed with an additional suture. The oral ending must be mobilized to be placed in the level of the skin, and the wound is closed with the standard technique upward and downward from the prospective esophagostomy. The esophagus is fixed at the skin with interrupted sutures placed circularly

Figures 38.5 and 38.6





Conclusion

Boerhaave's syndrome results in chemical and infectious mediastinitis, which is lethal unless treated early and effectively. A high index of suspicion is important for early recognition of injury. If a transmural injury with mediastinal or abdominal soilage is identified, definitive surgical management is required. Symptoms of spontaneous rupture often are nonspecific and include acute chest and abdominal pain, odynophagia, dyspnea, and fever. Pleural effusion, pneumothorax, pneumomediastinum, and subcutaneous emphysema are chest radiograph findings. Diagnosis is confirmed by esophagram. Esophagoscopy generally should not be performed for diagnosis.

The approach and extent of surgical intervention must be planned according to the location of the lesion, the time interval, and the region of expected infection. Although there is no conclusive consensus regarding the best surgical strategy, the following points may be considered: Treatment by primary suture should be performed if it is possible and seems secure (Jougon 2004). Surgical treatments include cervical esophagectomy with gastrostomy for high intrathoracic ruptures, primary repair with or without omentoplasty or (hemi)fundoplication for low thoracic ruptures, and transhiatal repair or resection for ruptures at the level of the diaphragm. If necessary subsequently, depending on the primary approach—either abdominal or thoracic—the other cavity may be explored and drained via a minimally invasive approach (de Schipper 2009).

The symptoms, diagnosis, and treatment of esophageal perforations other than those of Boerhaave's syndrome are similar but not fully comparable. All in all, the most common cause of esophageal perforation is iatrogenic. However, it should also be noted that iatrogenic perforations, although still a serious medical condition, are easier to treat and less prone to complications, particularly mediastinitis and sepsis, because they usually do not involve contamination of the mediastinum with gastric contents. Therefore, a nonsurgical approach often may be possible, with fair results (Vallböhmer 2010).

Spontaneous esophageal perforation continues to be a serious disorder with significant morbidity and mortality. Early surgical treatment offers the best chance for a satisfactory outcome. As an exception, we agree to conservative treatment if inflammatory effects are localized and complications can be controlled (Schmidt 2010). However, these conditions are rare in classical Boerhaave's syndrome. Extended perforations with spread of air and fluids to the mediastinum, retroperitoneum, or abdomen should be treated as early as possible surgically before systemic life-threatening symptoms develop.

References

Boerhaave H (1724) Atrocis, nec descripti prius, morbis historia: Secundum medicae artis leges conscripta. Lugduni Batavorum; Ex officine Boutesteniana

Curci JJ, Horman MJ (1976) Boerhaave's syndrome: the importance of early diagnosis and treatment. Ann Surg 183:401–408

de Schipper JP, Pull ter Gunne AF, Oostvogel HJ, van Laarhoven CJ (2009) Spontaneous rupture of the oesophagus: Boerhaave's syndrome in 2008**.** Literature review and treatment algorithm. Dig Surg 26:1–6

Jougon J, Mc Bride T, Delcambre F, Minniti A, Velly JF (2004) Primary esophageal repair for Boerhaave's syndrome whatever the free interval between perforation and treatment. Eur J Cardiothorac Surg 25:475–479

Korn O, Oñate JC, López R (2007) Anatomy of the Boerhaave syndrome. Surgery 141:222–228

Schmidt SC, Strauch S, Rösch T, Veltzke-Schlieker W, Jonas S, Pratschke J, Weidemann H, Neuhaus P, Schumacher G (2010) Management of esophageal perforations. Surg Endosc 24:2809–2813

Vallböhmer D, Hölscher AH, Hölscher M, Bludau M, Gutschow C, Stippel D, Bollschweiler E, Schröder W (2010) Options in the management of esophageal perforation: analysis over a 12-year period. Dis Esophagus 23:185–190