

Esophageal Perforation, Rupture and Tears

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Author: Corey M Long, MD, Resident, Department of Emergency Medicine, Bellevue Hospital Center, New York University Medical Center

Coauthor(s): Ugo Anthony Ezenkwele, MD, MPH, Assistant Professor of Emergency Medicine, Department of Emergency Medicine, New York University School of Medicine/Bellevue Hospital Center

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Introduction

Background

In 1724, Dr Hermann Boerhaave described the first, and likely most well known, case of esophageal perforation. Baron von Wassenaer, the Grand Admiral of Holland, followed a large meal with his customary bout of emetic-induced vomiting. However, on this occasion, the Admiral experienced a sudden and severe pain in his upper abdomen after violent but minimally productive retching. Dead less than 24 hours later, his autopsy revealed a transverse tear of his distal esophagus and gastric contents in the pleural spaces. Spontaneous esophageal rupture is a rare and dangerous entity, which today is commonly known as [Boerhaave syndrome](#).

Today, most instances of esophageal perforation are iatrogenic, but this remains a potentially devastating condition. Rapid diagnosis and therapy provide the best chance for survival; however, delay in diagnosis is common, resulting in substantial morbidity and mortality. This article discusses the causes, investigation, and initial therapy for this potentially lethal gastrointestinal condition.



Nonenhanced CT scan through the mid esophagus in a patient with esophageal perforation after upper GI endoscopy shows a false tract emanating from the esophagus (arrow).

Pathophysiology

The esophagus is more vulnerable than the rest of the alimentary tract due to the lack of a serosal layer, which provides stability through elastin and collagen fibers. Perforation may be due to several mechanisms, including direct piercing, shearing along the longitudinal axis, bursting from radial forces, and thinning from necrosis of the esophageal wall.

Iatrogenic injury through esophageal instrumentation is the leading cause of perforation by either piercing or shearing and may be due to any number of procedures, especially endoscopy and dilatation of strictures. Such tears often occur near the pharyngoesophageal junction where the wall is weakest. Because the esophagus is surrounded by loose stromal connective tissue, the infectious and inflammatory response can disseminate easily to nearby vital organs, thereby making the esophageal perforation a medical emergency and increasing the likelihood of serious sequelae. Underlying esophageal disease (tumor, stricture) predisposes toward perforation with instrumentation, which often occurs distal to the affected area. Perforation during surgery most often occurs in the abdominal esophagus.

Spontaneous esophageal rupture (Boerhaave syndrome) occurs secondary to a sudden increase in intraluminal pressures, usually due to violent vomiting or retching, and often follows heavy food and alcohol intake. In more than 90% of cases, perforation occurs in the lower third of the esophagus; most frequently, the tear is in the left posterolateral region (90%) and may extend superiorly. The predilection for left-side perforation is due to the lack of adjacent supporting structures, thinning of the musculature in the lower esophagus, and anterior angulation of the esophagus at the left diaphragmatic crus. Fifty percent of ruptures occur in patients with [gastroesophageal reflux disease](#), suggesting that ease of pressure transfer from the abdominal to thoracic esophagus may facilitate rupture.

Shearing forces due to rapid increases in intragastric pressure against a closed pylorus result in a Mallory-Weiss tear (MWT). These longitudinal mucosal lacerations occur most commonly at the gastroesophageal junction or gastric cardia, especially if a hiatal hernia is present, and often present with hematemesis. Ultimately, these tears can perforate if the pressure increases are unrelieved. Further discussion of MWTs is reserved for another section.

The cervical esophagus is the most common site of perforation by several other mechanisms as well, particularly in the region of the pyriform sinus. [Trauma](#), almost uniformly penetrating, shows an affinity for the upper esophagus, while toxic ingestions and foreign bodies can directly damage the cervical esophagus or become lodged and cause insidious erosion of the muscle wall.

Frequency

United States

Boerhaave syndrome is rare, accounting for 15% of all cases of esophageal perforation.

Iatrogenic causes account for 50-75% of esophageal perforations. The actual incidence depends on the procedure; rigid endoscopy carries a perforation rate 0.1-0.4%, while that of flexible endoscopy varies from 0.01-0.06%. Rates increase quickly when interventions are undertaken, such as pneumatic balloon dilatation in achalasia (2-6%) or any procedure involving strictures secondary to radiation or tumor (10%). Furthermore, the rate of perforation is increased in the presence of a large hiatal hernia or esophageal diverticula.

Penetrating trauma to the neck (2-9%), thinning of esophageal membrane secondary to variceal sclerotherapy (1-3%), and foreign body or toxic ingestions (5-15%) make up the bulk of the remaining causes.

Mortality/Morbidity

Even with prompt therapy and advances in surgical technique, the mortality rate can be very high, varying from 5-75%; higher rates correlate with delays in both presentation and diagnosis.

- Mortality also varies by etiology and location of the perforation. The highest rates are attributed to Boerhaave syndrome (up to 72%), partly because of the difficulty in making the diagnosis, followed by iatrogenic (19%) and traumatic perforations (7%). Cervical perforations portend a lower mortality compared with abdominal and thoracic perforations due to containment of potential contamination by tight fascial attachments and mechanisms, which may make injury more obvious.
- The morbidity and mortality in esophageal perforation is most often due to an overwhelming inflammatory response to gastric contents in the mediastinum, pleural spaces, and adjoining tissues, as well as swift spread of infection to paraesophageal structures. Negative intrathoracic pressure may draw gastric material out of the esophagus, exacerbating injury.
- Morbidity may be due to pneumonia, mediastinitis, empyema, polymicrobial sepsis, and multiorgan failure.

Race

No information on racial predilection is available.

Sex

Boerhaave syndrome is generally associated with vomiting and customarily occurs after drinking and eating binges. It is more commonly observed in males than in females. Iatrogenic perforation shows no predilection.

Age

- Boerhaave syndrome is most common among patients aged 40-60 years old, but isolated case reports in children have been described.
- Iatrogenic perforations are associated with preexisting pathology and so are more common in fifth and later decades of life.

Clinical

History

- The classic presentation of spontaneous esophageal rupture is severe vomiting or retching followed by acute, severe chest or epigastric pain.
- Boerhaave syndrome has also been reported with abdominal or chest pain following straining, childbirth, weight lifting, fits of coughing or laughing, hiccuping, blunt trauma, seizures, and forceful swallowing.
- The presence of fever; pain in the neck, upper back, chest, or abdomen; dysphagia; odynophagia; dysphonia; or dyspnea following esophageal instrumentation should raise suspicion for perforation.

- Patients with thoracic or abdominal perforations may present with any of the above symptoms, as well as low back pain, shoulder pain referred from diaphragmatic irritation, increased discomfort lying flat, or true acute abdomen.
- The ingestion of a caustic toxin or foreign body preceding any of the above symptoms may indicate perforation.
- A history of preexisting upper gastrointestinal pathology (gastroesophageal reflux disease, hiatal hernia, carcinoma, strictures, radiation therapy, Barrett esophagus, varices, achalasia, infection) raises a patient's risk of perforation.
- Hematemesis, while occasionally present, is normally not a predominant symptom.

Physical

- Although the physical examination is often nonspecific, certain findings can be helpful.
- Subcutaneous emphysema is palpable in the neck or chest in up to 60% of perforations but requires at least an hour to develop after the initial injury.
- Tachycardia and tachypnea are common initial physical examination findings, but fever may not be present for hours to days.
- The Mackler triad, consisting of vomiting, chest pain, and subcutaneous emphysema, is classically associated with spontaneous esophageal rupture, though it is only fully present in about 50% of cases.
- Auscultation of the chest can be of particular value.
 - The Hamman sign is a raspy, crunching sound heard over the precordium with each heartbeat caused by mediastinal emphysema, often present with thoracic or abdominal perforations.
 - Breath sounds may be reduced on the side of the perforation due to a contamination of the pleural space, often on the left.
- In cases of delayed presentation, patients may be critically ill and present with significant hypotension.

Causes

- Iatrogenic etiologies predominate the causes of esophageal perforation, accounting for up to 85% of cases.
 - Instrumentation modalities commonly include endoscopy, sclerotherapy, variceal ligation, pneumatic dilation, bougienage, and laser treatment.
 - Placement of endotracheal, nasogastric, and Blakemore tubes represent less common iatrogenic causes.
- As detailed above, Boerhaave syndrome consistently accounts for about 15% of all perforations, normally secondary to vomiting after heavy food and alcohol intake, but possible by any action that abruptly increases intra-abdominal pressure against a closed superior esophageal sphincter.
- Swallowed foreign bodies may directly injure the esophagus by penetrating the tissue or becoming lodged at a point of esophageal narrowing, leading to pressure

necrosis and wall weakness; pills and coins are common culprits. Ingestion of caustic chemicals may lead to direct wall inflammation and damage.

- Trauma represents an important cause of perforation, estimated at up to 10% of cases. Penetrating trauma is much far more prevalent than blunt, often in the form of knife or gun wounds, and is associated with significant injury to important adjacent cervical structures. Blunt trauma may affect any portion of the esophagus¹, and the diagnosis is often delayed secondary to other injuries.
- Intraoperative esophageal perforation is a recognized complication of surgery, especially cardiothoracic or fundoplication, accounting for around 2% of all perforations.