Problematic issues of diagnosis and treatment of Boerhaave’s syndrome

Abstract. Boerhaave’s syndrome is a rare disease with an incidence of 3.1/1,000,000 per year, has a high mortality rate due to late diagnosis and difficult treatment. The purpose of the message is to draw the attention of doctors to the variety of clinical manifestations of Boerhaave’s syndrome, which complicates early diagnosis and decision making. Clinical observation of two patients with Boerhaave’s syndrome is presented. One patient was hospitalized 7 hours after the onset of pain in the upper left side, nausea, vomiting. Clinical examination, ultrasound and chest X-ray revealed no pathology. However, after 3 hours, the patient's condition deteriorated sharply, emphysema of the neck and chest appeared. X-ray revealed left-sided pneumomediastinum. Thoracotomy revealed a 3.5-cm long rupture in the lower third of the esophagus on its left lateral wall, which was sustained. The chest cavity and mediastinum are debrided and drained using tubes. A gastrostomy was formed. The postoperative period was complicated by multiple organ failure and sepsis. Patient was in the intensive care unit for 29 days. The esophageal wound healed on the 46th day and the patient was discharged for outpatient observation. Another patient was hospitalized three hours after onset of illness with a left-sided pneumomediastinum and acute abdomen symptoms. Chest tube on the left was placed and a dark brown liquid released under pressure. The peritonitis clinical symptoms prompted surgeons to perform urgent laparotomy during which no pathology of the abdominal organs was detected. Only after computed tomography, a rupture of the esophagus was diagnosed. As a result of delayed surgery, infection complications and sepsis developed, which led to the patient’s death.

Keywords: Boerhaave’s syndrome; rupture of the esophagus; diagnosis; treatment; review

Introduction
Boerhaave’s syndrome (BS) or “banquet esophagus” occurs as a result of a suddenly increased pressure in the esophagus with a closed pharyngeal–esophageal sphincter in combination with negative intrathoracic pressure, which occurs with intense vomiting or straining (lifting loads, excessive coughing, defecation) and is accompanied by a sharp pain syndrome. BS is a rare disease with a high mortality rate due to late diagnosis and difficult treatment. The incidence of esophageal ruptures is 3.1 per 1,000,000 per year but BS is diagnosed only in 15–24 % of cases [1–6].

A wide range of clinical manifestations and comorbidities complicate diagnosis and worsen treatment outcomes. Surgical intervention performed within the first 24 hours leads to the recovery in 75–90 % of patients [1, 7, 8]. Delayed treatment is complicated by sepsis in 78 % of cases [9] with a mortality rate of 40–60 %, which is 100 % if the delay in treatment lasts more than 48 hours [1, 10, 11]. In 17 % of cases, the diagnosis is made only by autopsy [6, 12].

Variety of symptoms and the lack of alertness among physicians regarding the diagnosis of BS, the lack of unified recommendations for surgical procedures require wider coverage and discussion of this pathology in the literature.

The purpose of the message is to draw the attention of physicians to the need for alertness and early diagnosis of BS with the use of modern information technologies and urgent surgical intervention.

Materials and methods
The literature on the diagnosis and treatment of BS is analyzed in PubMed, Scopus, Web of Science, Medscape databases for the 2010–2023 using a combination of terms “Boerhaave’s syndrome”, “diagnosis”, “management”, “review”.

For correspondence: Volodymyr Zhukovsky, PhD, Associate Professor at the Department of Traumatology, Orthopedics and Military Field Surgery, Danylo Halytsky L'viv National Medical University, Pekarska st., 69, L'viv, 79010, Ukraine; e-mail: zukovskiy@ukr.net; fax: +380 (32) 275-76-32, phone: +380 (67) 725-32-39
Full list of authors’ information is available at the end of the article.
Clinical observations of two patients with BS who were treated at Lviv St. Luke’s Hospital of the First Territorial Medical Union in 2023 are presented.

**Clinical observation 1.** Patient K., 49 years old, was urgently hospitalized with complaints of pain in the upper left side, nausea, vomiting, increased blood pressure, headache, general weakness. These complaints appeared after eating 7 hours ago.

Upon admission, the skin is pale pink. Body temperature is 36.6 °C. Auscultation: vesicular respiration on both sides, no wheezing. Respiratory rate is 18 per minute. Heart sounds are clear and rhythmic. Heart rate is 70 per minute, blood pressure is 160/90 mm Hg. The abdomen is soft, not painful. Symptoms of peritoneal irritation are negative, peristalsis is normal, the liver is not enlarged, not painful. Percussion of the right and left abdomen is not painful. Physiological stool is without features. Laboratory tests of blood and urine are within normal limits. Abdominal ultrasound did not reveal free fluid and pathology. Chest X-ray showed no pathological changes.

However, during the diagnostic procedures, the patient’s condition deteriorated rapidly: pain intensified, emphysema appeared on the neck and face. Esophagogastroduodenoscopy was performed during which a linear defect about 3 cm was found in the lower third of the esophagus, on its left lateral wall, which was filled with blood clots, with active bleeding into the esophageal lumen.

X-ray revealed massive emphysema of the neck and chest soft tissues, left-sided pneumothorax with the left lung collapsed, pneumomediastinum, and fluid in the left sinus and mediastinum (Fig. 1).

Thoracotomy was performed urgently with an anterior lateral approach on the left in the 7th intercostal space. During the revision, protrusion and edema of the mediastinal pleura were found in the lower third of the esophagus. The mediastinal pleura was opened, blood clots and hemorrhagic fluid were removed, about 400 ml. A linear rupture of the esophagus in the lower third, 3.5 cm long, was diagnosed. The esophagus is sutured with a two-row suture. Debridement of the pleural cavity was done. Three tube drains were placed: one to the esophageal sutures and two to the left chest (anterior and posterior). The thoracotomy wound is sutured. In order to exclude the passage of the esophagus and for enteral nutrition of the patient, a gastrostomy was formed.

Treatment in the early postoperative period was carried out in the intensive care unit. Two days after the surgery, the patient developed bilateral pneumonia and toxic kidney damage. On the sixth day, a significant increase in the indicators of the systemic inflammatory response and hemodynamic instability developed, blood pressure was 85/50 mm Hg. Signs of respiratory failure were manifested in a decrease in PaO\textsubscript{2}/FiO\textsubscript{2} to 220 mm Hg. Acute kidney injury manifested itself in a decrease in urine output (oligoanuria), an increase in creatinine to 480 μmol/l, and urea to 38 μmol/l. There was about 450 ml of serous-purulent secretions per day through drains from the mediastinum and from the left chest cavity. Thus, mediastinitis and empyema were complicated by multiple organ failure and sepsis.

Bacteriological examination of sputum and secretions from drains revealed mixed microorganisms: *Enterobacter aerogenes* 10\textsuperscript{4}, *Pseudomonas aeruginosa* 10\textsuperscript{4}, *Candida albicans* 10\textsuperscript{4}, *Klebsiella pneumoniae* 10\textsuperscript{4}, which are sensitive to carbapenems. Respiratory support was provided by mechanical ventilation through a tracheostomy for up to 20 days. Correction of the fluid and electrolyte and acid-base balance, transfusion of blood components and nutritional support by the mixed enteral-parenteral route were performed. For analgesedation, dexmedetomidine, non-steroidal anti-inflammatory drugs, and narcotic analgesics were used. In order to treat renal failure, three hemodialysis sessions were performed.

On the 29th day, the patient was transferred to the surgical department. As a result of treatment, the patient’s condition improved. On days 20 and 42, drains were removed from the chest. Fibroesophagogastroscopy on the 46th day after surgery: healing of the esophageal wound was confirmed, and the patient was discharged for outpatient observation.

After 2 months, the patient was hospitalized and closed gastrosomy was successfully performed.

**Clinical observation 2.** Patient B., 35 years old, was admitted to the emergency surgical department at night with complaints of pain in the epigastric region of a cutting nature, which occurred 3 hours ago after repeated vomiting of dark content and general weakness. The day before, he drank alcohol. He denies the injury. The general condition of the patient is serious. The skin and visible mucous membranes are pale. The tongue is moist and clean. Heart sounds are clear and rhythmic. Blood pressure was 120/80 mm Hg, heart rate was 84/min. Electrocardiogram: sinus rhythm, with no signs of ischemia. Auscultation: vesicular breathing is present in all pulmonary fields on the right, without wheezing on the left — there is no breathing. The percussion sound on the right is not changed, on the left it is dulled. The respiratory rate is 20 per minute, SpO\textsubscript{2} is 88 %. The abdomen: pain, swelling, tenderness, vivid symptoms of peritonitis.
X-ray: left — pulmonary pattern is absent, the lung is collapsed, there is fluid in the sinus with a horizontal level, the diaphragm dome is not contoured. On the right, there is the enhancement of the pulmonary pattern, the sinus is free. The shadow of the heart is shifted to the right (Fig. 2).

Abdominal X-ray: no pneumoperitoneum, no Kloiber cups were detected. Abdominal ultrasound: gas under the abdominal wall without free fluid in the abdominal cavity. Laboratory parameters: white blood cells $13.3 \times 10^9/l$, band neutrophils 13 %. Other blood counts are within normal limits. Preliminary diagnosis: peritonitis, spontaneous left-sided pneumohydrothorax.

The left chest tube was placed, air and 150 ml dark brown fluid were released under pressure. Bühlaus drain has been placed. This improved saturation by up to 97 % $\text{SpO}_2$. In connection with the diagnosed free gas in the abdomen, a perforation of the hollow organ was suspected, and a laparotomy was performed but no injury to the abdominal organs was found.

The next day, there was an intense release of dark brown fluid from the pleural drainage, subcutaneous emphysema appeared. 200 ml of 0.9% sodium chloride solution with diamond green dye was injected through a gastric tube. The release of the dye through pleural drainage was obtained. Endoscopy: a longitudinal tear of the esophagus 3 cm long, 2 cm proximal to the Z-line. CT scan: rupture of the lower third of the esophagus, mediastinal emphysema, local mediastinitis, left-sided pneumothorax, atelectasis of the left lung lower lobe (Fig. 3).

Left-sided anterolateral thoracotomy: a linear tear 3 cm long was found on the left esophageal wall, 2 cm above the cardia. The esophagus is sutured, the posterior, anterior mediastinum and the left chest are drained. A jejunostomy is formed for nutrition. Despite intensive care, the disease was complicated by the rapid development of sepsis, which led to the death of the patient on the 18th day after the disease onset.

Discussion

The first report of spontaneous transmural rupture of the esophagus was described by Hermann Boerhaave in 1724 in a publication entitled “A History of a Serious Illness” [13]. Dr. Barrett performed the first successful surgery for this disease in 1946 [14].

Figure 2. X-ray of patient B., 35 years old. Left — pulmonary pattern is absent, the lung is collapsed, there is fluid in the sinus with a horizontal level, the diaphragm dome is not contoured

Figure 3. CT scan: rupture of the lower third of the esophagus, mediastinal emphysema, local mediastinitis, left-sided pneumothorax, atelectasis of the left lower lobe: A — longitudinal section, B — cross section
BS is a barogenic injury of the esophagus caused by a sharp increase in the intraesophageal pressure, which leads to transmural rupture of the esophagus. These ruptures are usually longitudinal, most often occurring in the distal esophagus on the left, about 2–4 cm above the cardia. There is no local protection in this area and the esophageal wall is weak due to the thinness of muscle fibers and the large number of vascular and nerve structures [15, 16]. BS is most common in patients aged 50–70 years who overeat and abuse alcohol, with a male-to-female ratio of 2:1 to 5:1 [15]. Superficial ruptures, which are limited only to the mucous membrane of the stomach and esophagus, are accompanied by the development of Mallory-Weiss syndrome. With transmural injury, gastric contents are thrown into the mediastinum with the development of mediastinitis and empyema [17].

Mackler's triad (vomiting, chest pain, and subcutaneous emphysema) is characteristic of BS with tachycardia, fever and tachypnea. The pain location depends on the site of esophageal perforation and can be in the back, spine, behind the sternum, or epigastrum [18, 19]. As a rule, sudden weakness, dizziness, cold sweat, hypotonia, sometimes loss of consciousness occurs at the moment of esophageal rupture [20–22]. However, these symptoms are not specific to BS and usually occur in only 14 to 50% of all patients [2, 3, 23].

As the researchers note, the diagnosis of BS poses significant difficulties due to the variety of clinical manifestations and the lack of alertness of doctors regarding this disease [21, 24]. Patients are often hospitalized with suspected acute coronary syndrome, pulmonary embolism, diaphragmatic hernia, thoracic aortic aneurysm dissection, acute pancreatitis, pneumothorax and perforated gastroduodenal ulcer [25–27].

According to reports in the literature, a delay in correct diagnosis and the initiation of adequate treatment occurs in more than 50% of cases [28]. The mean time from symptom onset to diagnosis is 27 ± 12 hours [29], which was also in our observation.

In the early stage after damage to the esophagus, 12% of chest X-rays may be perfectly normal [30]. This is why we can explain the absence of pathological changes on the chest X-ray upon admission of our patient. Later, within a day from the moment of rupture, chest X-ray reveals hydropneumothorax on one side and emphysema of the mediastinum [31]. A reliable sign of esophageal rupture is the extravasation of contrast material outside the esophagus [3, 32].

Esophageal radiography with contrast and CT scan is the method of choice for the verification of esophageal perforation with a sensitivity of 90–100% [12, 33]. These methods reveal extravasation of the contrast medium at the site of perforation and mediastinal and chest complications. In addition, it helps to rule out another possible diagnosis that could simulate BS, namely dissecting aortic aneurysm and acute coronary syndrome [12, 17, 24, 34, 35].

Esophageal endoscopy, which has a sensitivity of 100% and a specificity of 92%, can visualize the location and size of the esophageal perforation, but it must be done carefully so as not to increase the size of the esophageal rupture and not cause the development of tension pneumothorax [34, 36]. Therefore, it is relatively safe to perform esophagoscopy after preliminary drainage of the chest. Discharge of air from the chest tube during esophagoscopy objectively indicates a rupture of the esophagus [37].

One should be alert and aware of possible BS, especially with brown fluid discharge from the chest tube, which occurred in one of our patients. Most often, these discharges are purulent, cloudy, or hemorrhagic [4, 17, 38].

The most difficult issue today is the choice of therapeutic approach, which depends on the patient’s condition, the location and length of the rupture, the time period, and concomitant complications [2, 23, 39].

Surgical treatment is currently considered the gold standard for esophageal ruptures and consists of suturing the esophageal perforation through thoracotomy access with debridement and chest and mediastinum tube. The esophagus is temporarily excluded from the digestive system by a gastrostomy, which used for enteral nutrition [1, 26, 28, 40]. It facilitates the diagnosis of esophageal rupture by intraoperative injection of a dye, which can also be used to check the tightness of the esophageal sutures [30]. For patients with perforation localized in the lower third of the esophagus and stable life support, some authors recommend laparoscopic intervention for BS via transhiatal access. This may reduce the risk of postoperative complications [5, 41, 42].

Endoscopic equipment allows clipping esophageal injuries with a system of clamps OTSC [43, 44], endoscopic ligation loops [45, 46], stents [47, 48] and endovacuum aspiration systems [37, 49, 50]. Endoscopic clipping in combination with the chest tube is indicated for early diagnosis, stable hemodynamics, absence of infection, and defects not exceeding 20 mm [2, 43, 45, 51, 52]. Despite minimal invasiveness, endoscopic treatment does not guarantee success, does not allow debridement of the chest, which can pose a potential risk to the patient’s life [39, 49, 53]. Some authors recommend combining video-assisted thoracoscopic surgery and esophagoscopy [37, 42, 54, 55]. Intraoperative esophagoscopy is also useful in determining the exact location and size of the perforation, as mucosal defects are often larger than muscular defects [17, 34].

Attempts to suture a gap that is more than a day old are usually doomed to failure due to the eruption of sutures on swollen and inflamed tissues, which often leads to an increase in the size of the defect [7, 9, 39]. Therefore, the main elements of the surgery are drainage and gastrostomy [40]. Drainage for esophageal ruptures is the most important element of surgery [18]. The first drain is placed near the esophageal suturing site, the second in the posterior mediastinum, and the third in the chest. Active aspiration is advisable. Drains are removed only after cessation of discharge from the mediastinum and chest space.

Esophageal exclusion or resection with gastrostomy are recommended in large esophageal defects and if repair of the esophagus is not possible.

In case of suture failure or the development of an inflammatory process in the chest cavity, the restoration of the esophagus is carried out with a T-shaped tube of a large size [1]. The separation of the esophagus lumen from the mediastinal tissue even for 2–3 days prevents its purulent infection and helps reduce intoxication.

Given the high risk of surgery in patients with BS complicated by multiple organ failure and sepsis, some authors
suggest conservative treatment that includes intensive fluid resuscitation, broad-spectrum antibiotics, proton pump inhibitors, total parenteral nutrition, and chest tube, which can sometimes be positive [4, 37, 40, 52, 56, 57].

In our observations, esophageal rupture was not suspected immediately upon admission due to the underestimation of the medical history by the doctors on duty, the absence of radiological changes and subcutaneous emphysema on the neck, which had not yet developed. Undoubtedly, the initial misdiagnosis was facilitated by the lack of alertness of physicians about this disease. Delayed diagnosis caused the development of severe purulent-septic complications, which led to an adverse outcome in one of our patients. However, it should be emphasized that even timely diagnosis and timely surgical treatment have risks of severe purulent-septic complications associated with failure of esophageal sutures.

Conclusions

Boerhaave’s syndrome in the first hour after onset may not show signs based on which it would be possible to suspect damage to the esophagus.

Each clinical symptom should be analyzed in detail and the diagnostic capabilities of CT should be used urgently.

In patients with complaints of repeated vomiting and chest pain that appeared after excessive food consumption, a possible BS should be suspected and a CT scan with contrast should be performed as a matter of urgency.

Urgent surgical intervention is the optimal method of treating BS the main elements of which are full drainage of the mediastinum and chest space and the formation of a gastrostomy for enteral nutrition.

Comprehensive intensive care and monitoring of complications are the key to a favorable postoperative period.

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Information about authors
Volodymyr Zhukovskiy, PhD, Associate Professor at the Department of Traumatology, Orthopedics and Military Field Surgery, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: zukovskiy@ukr.net; fax: +380 (32) 275-76-32, phone: +380 (67) 725-32-39; https://orcid.org/0000-0002-0594-5316
Ihor Trutyak, MD, DSc, PhD, Professor, Head of the Department of Traumatology, Orthopedics and Military Field Surgery, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: ihortrutukayahoo.com; https://orcid.org/0000-0001-8157-3449
Yaroslav Podhomy, MD, DSc, PhD, Professor, Department of Anesthesiology and Intensive Care of Postgraduate Education, Faculty of Postgraduate Education, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: podhomyjsiukr.net; https://orcid.org/0000-0002-9926-9257
Zhanna Filip, PhD, Assistant at the Department of Anesthesiology and Intensive Care of Postgraduate Education, Faculty of Postgraduate Education, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: zhasya.doc@gmail.com; https://orcid.org/0000-0002-3451-6016
Viktor Kozopas, PhD, Associate Professor at the Department of Disaster Medicine and Military Medicine, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: kozopas@ukr.net; https://orcid.org/0000-0003-3451-6016

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Information about authors
Volodymyr Zhukovskiy, PhD, Associate Professor at the Department of Traumatology, Orthopedics and Military Field Surgery, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: zukovskiy@ukr.net; fax: +380 (32) 275-76-32, phone: +380 (67) 725-32-39; https://orcid.org/0000-0002-0594-5316
Ihor Trutyak, MD, DSc, PhD, Professor, Head of the Department of Traumatology, Orthopedics and Military Field Surgery, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: ihortrutukayahoo.com; https://orcid.org/0000-0001-8157-3449
Yaroslav Podhomy, MD, DSc, PhD, Professor, Department of Anesthesiology and Intensive Care of Postgraduate Education, Faculty of Postgraduate Education, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: podhomyjsiukr.net; https://orcid.org/0000-0002-9926-9257
Zhanna Filip, PhD, Assistant at the Department of Anesthesiology and Intensive Care of Postgraduate Education, Faculty of Postgraduate Education, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: zhasya.doc@gmail.com; https://orcid.org/0000-0002-3451-6016
Viktor Kozopas, PhD, Associate Professor at the Department of Disaster Medicine and Military Medicine, Danylo Halytsky Lviv National Medical University, Lviv, Ukraine; e-mail: kozopas@ukr.net; https://orcid.org/0000-0003-3451-6016

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