Spontaneous Rupture of the Oesophagus: Boerhaave's Syndrome in 2008

Literature Review and Treatment Algorithm

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Key Words

Boerhaave's syndrome · Rupture of oesophagus · Oesophagus

Abstract

Objective: Boerhaave's syndrome is a spontaneous rupture of the oesophagus with a lack of diagnostic and treatment consistency in the literature. Therefore, we reviewed all published literature in order to design a treatment algorithm based on the literature. **Study Design:** A systematic literature review written in the English language since 1975. **Results:** We reviewed all known literature. Treatment of the Boerhaave syndrome was divided into three categories: conservative, endoscopic and surgical approach. The survival rate of all treatments was 75, 100 and 81%, respectively. **Conclusion:** Boerhaave's syndrome should be treated endoscopically when diagnosed within 48 h and when there are no signs of sepsis. However, when a patient is diagnosed within 48 h and has a septic profile, thoracotomy with hemifundoplication and pleural/mediastinal drainage should be performed; and in case of intra-abdominal leakage, a laparotomy for local repair should be performed. When a patient is diagnosed after 48 h, conservative treatment should be followed and only when a patient gets a septic profile is surgical treatment indicated.

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Introduction

Boerhaave's syndrome is a rare condition with a high mortality rate. Between 2003 and 2005, 10 cases with a spontaneous rupture of the oesophagus were reported annually in The Netherlands [1]. Spontaneous oesophageal perforation was first described by the Dutch physician Hermann Boerhaave in 1724 [2]. Classically, presentation of pain, dyspnoea and shock is followed by forceful vomiting.

Boerhaave's syndrome is a form of barogenic rupture caused by a rapid rise in intraluminal pressure in the distal oesophagus [3–5]. Spontaneous rupture of the oesophagus or Boerhaave's syndrome is the most sinister cause of oesophageal perforation with estimated mortality rates of 20–40%. The rupture is usually (in 90% of cases) in the lower third of the oesophagus and in the left lateral position (fig. 1). This is believed to be due to an anatomic weakness at that point. The average tear is 2.2 cm long and 3–6 cm above the diaphragm and rarely causes massive haemorrhage [6]. Boerhaave's syndrome is a rare disease. No consensus exists on the most suitable treatment.

Diagnostic errors are prevalent; the most common misdiagnosis is perforated ulcer, followed by myocardial infarction, pulmonary embolism, dissecting aneurysm

Fig. 1. Dye X-ray of distal oesophagus with leakage due to rupture.

Fig. 2. CT scan in the pulmonary window showing free air and leakage around the distal oesophagus.

Fig. 3. Dye X-ray showing no leakage or stricture after 5 months.

and pancreatitis [7, 8]. Diagnosis can be made earlier and more accurate with additional radiological examinations such as CT scan [9] (fig. 2, 3). Multiple treatment modalities are described, ranging from conservative measures to extensive surgery. However, no consensus exists about preferred treatment for a spontaneous rupture of the oesophagus.

A systematic literature review is performed to detect the best possible treatment strategies. Based on available literature, a treatment algorithm is formed, describing timing and place of conservative, endoscopic and operative treatment.

Methods

A systematic literature search was performed in PubMed searching for English literature available since 1975. The search was performed on mesh terms 'spontaneous rupture', 'oesophagus' and 'treatment'. Since the absence of any level 1 or 2 studies, we included all multi-patient level 3, 4 and 5 articles containing the word 'Boerhaave' in the title. All related articles were examined. All articles concerning diagnosis and treatment of Boerhaave's syndrome were included. Articles concerning treatment of Boerhaave's syndrome and non-Boerhaave's syndrome oesophagus perforations were included. In these studies, patient data about true Boerhaave's syndrome were analysed. The main outcome criterion of all studies was survival.

Results

In the available literature on treatment of a rupture of the oesophagus, it should be considered that these ruptures are not necessarily the ruptures occurring within the Boerhaave syndrome. Considering both treatment and outcome, one of the main differences with non-spontaneous ruptures is that the diagnosis Boerhaave's syndrome is often made later than in patients with an iatrogenic rupture of the oesophagus or a rupture in malignancies of the oesophagus. During our analysis we only included those patients who sustained a non-iatrogenic rupture of the oesophagus.

Table 1. Review of treatment modalities and outcome

Reference (first author)	Oesophageal ruptures, n	True Boerhaave n	Treatment of Boerhaave patients	Survival of true Boerhaave patients, y/Y
<i>Conservative</i>				
Ivey [13]	3	3	Hyperalimentation and antibiotics	3/3
Cameron [15]	8	2	Hyperalimentation and antibiotics	2/2
Michel [18]	72, 19 conservative	11	Antibiotics, nasogastric suction	7/11
Subtotal				12/16
<i>Endoscopic</i>				
Fischer [19]	15	5	Endoscopic stent and pleural drainage (2/5 additional operation)	Not reported
Siersema [20]	11	5	Endoscopic stent and pleural drainage (2/11 additional operation)	5/5
Chung [21]	3	3	Endoscopic stent and pleural drainage (n = 1)	3/3
Subtotal				8/8
<i>Operative</i>				
Huetting [23]	11	3	Primary repair, omentoplasty, hemifundoplication and/or gastrostomy or cervical oesophagostomy	Not reported
Lawrence [24]	21	21	2 patients conservatively, 17 primary repair <72 h, 2 thoracic debridement	18/21
Gupta [25]	59	6	Transhiatal esophagectomy, intercostal chest tube drainage	2/6
Nesbitt [26]	115	22	Closure or diversion with drainage	18/22
Kiernan [27]	48	13	Primary closure, resection and reconstruction, diversion and/or exclusion	12/13
Brauer [28]	18	18	Resection after 24 h, primary suture and fundoplication <24 h	17/18
Subtotal				67/80

Treatment of Boerhaave's syndrome can be divided in three categories: conservative, endoscopic and surgical approach.

Conservative Treatment

The available literature on the conservative approach group is small (table 1). In general, conservative treatment consists of antibiotic treatment and/or percutaneous drainage of abscesses [10–12].

In the literature, Ivey et al. [13] described 3 cases of conservatively treated patients with Boerhaave's syndrome: they stated that conservative management of spontaneous oesophageal perforation is feasible when (1) the perforation is already 5 days old, (2) there are no signs of severe sepsis, (3) oesophageal barium study shows a wide-mouthed cavity draining freely back into the oesophagus, and (4) the pleural space is not contaminated. However, they also stated that when the diagnosis is made promptly, surgical therapy remains the treatment of choice, and patients managed conservatively who show signs of sepsis should be operated on without hesitation.

Other semi-conservative therapies are described as follows: Santos and Frater [14] passed the nasogastric tube down to the level of perforation and used it as a suction-rinsing drain with physiologic saline or antibiotic solutions. Cameron and Kieffer [15] are against the use of a nasogastric probe, claiming that it merely increases the gastro-oesophageal reflux, a most untoward effect as far as healing is concerned. Lyons and Seremetis [16] created a gastrostoma or a nutrient jejunostoma under the heading of non-operative treatment. Junginger et al. [17] found 54% (7 of 13) mortality in the non-operative treatment group and advocated operative treatment. Considering the outcome, Michel and Grillo [18] reported on non-operative treatment in 11 Boerhaave syndromes out of 72 oesophageal perforations with 64% survival.

Endoscopic Treatment

Spontaneous rupture of the oesophagus can also be treated endoscopically. Fischer et al. [19] performed a non-randomized observational study with spontaneous and iatrogenic oesophageal perforations treated with self-expandable metal stents. All non-iatrogenic patients

Fig. 4. Treatment algorithm for Boerhaave's syndrome.

were initially treated with stent and additional chest tube drainage of pleural empyema. Two out of 5 Boerhaave patients were operated on because of insufficient drainage with chest tube drainage of the pleural empyema.

Siersema et al. [20] reported 5 Boerhaave patients out of 11 oesophagus perforations treated with metallic endoprosthesis. Patients were treated with endoprosthesis and one or both pleural cavities drained with chest tubes. One Boerhaave patient deteriorated and required thoracotomy and esophagectomy due to multiple abscesses. All stents were removed after about 7 weeks. Mortality was 1/11.

Chung et al. [21] also presented 3 cases treated with metallic endoprosthesis. In 2 of the 3 subjects, endoscopic treatment was chosen because of the high risk of surgery. All 3 patients survived. Johnsson et al. [22] reported about a metallic endoprosthesis that seals oesophageal leakage from a different aetiology. However, of the 22 patients with oesophageal perforations treated, only 1 was diagnosed with Boerhaave's syndrome. This 76-year-old patient died after 25 days, although no leakage or infectious complication could be found.

Operative Treatment

The last category in treating Boerhaave's syndrome is the surgical approach. Different case reports and case-control studies are available in the literature. In general, surgical treatment consists of strategies for early (<24 h) or late (>72 h) diagnosis of Boerhaave's syndrome.

Hueting et al. [23] retrospectively reviewed 3 Boerhaave patients out of 11 oesophageal perforations. All 3

of these patients in this series survived. In case of rupture to the pleural cavity, thoracotomy with hemifundoplication was performed. Otherwise laparotomy with local suturing and drainage was performed.

Lawrence et al. [24] presented a retrospective series of 21 patients treated over a 10-year period. The surgical approach was through primary oesophageal repair with a single layer of interrupted absorbable sutures combined with mediastinal debridement and drainage gastrostomy. The majority was treated after 24 h. 18 out of 21 patients were operated with repair due to systemic sepsis or non-localized collections. Two patients were thoracoscopically drained due to delayed presentation more than 2 weeks after rupture. The mortality rate in this series was 3/21 patients.

Gupta and Kaman [25] reported 6 Boerhaave patients with 1 survivor. Two of these 6 patients underwent transhiatal resection. Nesbitt and Sawyers [26] reported a significant difference in survival when patients were treated within the first 24 h. 22 patients out of a series of 115 cases had a true Boerhaave syndrome. The overall mortality rate among these patients was 4/22. All patients treated within 24 h survived. For patients treated after 24 h, the mortality rate was 2/7, and for treatment started after 48 h it was 2/5. If treatment was possible within 48 h, primary closure was performed. After 48 h, diversion of exclusion procedures was performed.

Kiernan et al. [27] reported in an article in 2006 their experience with 13 Boerhaave cases out of 48 patients with a rupture of the oesophagus. They recommend aggressive, definitive surgery for thoracic oesophageal per-

forations, regardless of time of diagnosis. In the absence of phlegmon or implacable obstruction, primary repair offers excellent results with the shortest length of stay. Resection and reconstruction are the best choices in circumstances where significant phlegmon or distal obstruction render primary repair hazardous or inapplicable. Diversion, preferably with proximal and distal oesophageal exclusion, may be necessary for patients too ill to undergo more formidable surgery.

Brauer et al. [28] retrospectively reported 18 cases of Boerhaave's syndrome. 11 patients were treated by oesophagectomy, indicated by a rupture of >3 cm and time interval of >24 h. 7 patients were treated by primary suture and fundoplication, on the base of a tear <3 cm and presentation <24 h. Overall mortality was 1/18.

Conclusion

This literature study showed that data are scarce and that the level of evidence on these data is limited to retrospective level 3 case series, and expert opinions. The available data are biased and show favourable results in conservative, endoscopic and operative treatment. Discrimination of the preferred treatment and appointing decisional preoperative factors is not possible. Also a positive publication bias is experienced as merely tertiary expert centres present their (positive) results. However, a treatment rationale and algorithm based on clinical experience and best available evidence is offered here and for discussion.

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Conservative measures seem feasible especially if patients are diagnosed after 48 h and patients are not septic, but conversion to operative treatment remains necessary in quite a number of patients. Conservative treatment is merely reported in case reports. Endoscopic treatment is often advocated. The literature is impressive, but not clear about the success rates in true Boerhaave syndromes. Operative treatment still seems to be the best treatment for cases with an early diagnosis. Reported mortality rates vary from 0%, if treatment is started within 24 h, to 29%, if treatment is started after 48 h. No consensus exists about the best surgical strategy. Dependent on the localization of the perforation, cervical esophagectomy with gastrostoma for high intrathoracic ruptures or primary repair with or without omentoplasty or (hemi)fundoplication for low thoracic ruptures or ruptures at the level of the diaphragm are used as surgical treatments.

Based on this non-systematic review, a level 4 evidence treatment algorithm is proposed as presented in figure 4. The validity of such an algorithm can be best evaluated through a multicentre, national, web-based database. This may solve the difficulties of many hospitals in dealing with single incident cases throughout the country without a thorough knowledge and the lack of a solid treatment algorithm for such hospitals. Since patient characteristics such as age, ASA classification and history of previous diseases are not evaluated in the treatment algorithm, these should be considered separately from the algorithm. A working conference and launching of such a database is being prepared.

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