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Non-surgical management of Boerhaave's syndrome: a case series study and review of the literature

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ABSTRACT

Background and study aims Boerhaave's syndrome (BS) is a life-threatening condition with morbidity and mortality rates as high as 50 % in some reports. Until recently, surgical intervention has been the mainstay of management plans. With advances in therapeutic endoscopy, however, there has been increasing interest in non-surgical options including endoscopic esophageal stenting.

Patients and methods We reviewed the medical records of all patients diagnosed with BS and managed with endoscopic interventions between November 2011 and November 2016. The following variables were collected: patient demographics, clinical presentations, locations of esophageal perforation, primary interventions, complications, and outcomes.

Results Six patients were found to be diagnosed with BS during the study period. The median age at presentation was 55. There were 4 males and 2 females. The most common site of perforation was in the distal esophagus. The most common presenting symptom was chest pain (67 %) following an episode of vomiting or retching. Four patients (66.7 %) developed septic shock. Endoscopic treatment with a fully covered esophageal stent was the primary intervention in all patients (100 %). Interventional radiology was consulted in all cases for fluid drainage and chest tube placements. Clinical resolution of the BS was achieved in all patients (100 %) without any subsequent surgical interventions. There were no deaths within the study group, and the average follow-up duration was 2 years.

Conclusion Endoscopic treatment seems to be an effective management strategy in patients with BS. We also noted satisfactory results in patients presenting with sepsis, presumably due to urgent, interventional radiology-guided fluid drainage.

Introduction

Boerhaave's syndrome (BS), or spontaneous esophageal perforation due to high intraesophageal pressure, is a life-threatening condition with high morbidity and mortality. Delayed diagnosis is associated with mortality as high as 30 % to 50 % [1]. Until recently, surgical intervention has been the mainstay of treatment. Surgery in acute presentations, though, carries a high burden of complications. With advances in therapeutic endoscopy, there has been an evolving interest in the use of techniques such as esophageal stent placement and/or endoscopic suturing in combination with interventional radiology (IR)-guided techniques.

Patients and methods

We reviewed the medical records of all patients who underwent endoscopic interventions for esophageal perforations between November 2011 and November 2016. We collected the data on patients who received BS as their diagnosis upon presentation.

▶ **Table 1** Patient details.

Case	Age/sex	Comorbidity	Presentation	Diagnosis	Sepsis	Organ failure
1	42 M	Cirrhosis, Barrett's esophagus, GERD, asthma	3 days	40 hours	Septic shock	Respiratory/renal failure
2	57 F	Barrett's esophagus, Nissen fundoplication, asthma	2 days	6 days	Septic shock	Respiratory failure
3	67 M	Diabetes mellitus, Barrett's esophagus	1 day	< 24 hours	sepsis	Respiratory failure
4	55 M	Eosinophilic esophagitis, Barrett's esophagus, Nissen fundoplication	3 hours	1 hour	Sepsis	None
5	54 M	Alcohol abuse, GERD, rheumatoid arthritis	2 days	1 day	Septic shock	Respiratory/renal Failure
6	24 F	Diabetes mellitus, gastroparesis	3 days	1 day	Septic shock	Respiratory/renal failure

▶ **Table 2** Patient intervention and outcome.

Case	Perforation size	Intervention	Stent duration (days)	Hospital stay (days)	Stricture	Patient status (after 2 years)
1	6 cm	23 mm × 12.5 cm WallFlex fully-covered Stent	156	14	No	Alive
2	1 cm	23 mm × 12.5 cm WallFlex fully-covered Stent	6	21	No	Alive
3	1 cm	23 mm × 12.5 cm WallFlex fully-covered Stent	Lost Follow up	17	No	Alive
4	4 cm	23 mm × 12.5 cm WallFlex fully-covered Stent	35	14	No	Alive
5	1.5 cm	23 mm × 15.5 cm WallFlex fully-covered Stent	60	33	Yes	Alive
6	2 cm	18 mm × 12.3 cm WallFlex fully-covered Stent	76	46	No	Alive

The following variables were collected: patient demographics, clinical presentations, time to diagnosis and endoscopic intervention from presentation, locations of esophageal perforation, primary interventions, any other subsequent interventions or supportive measures, complications, and outcomes.

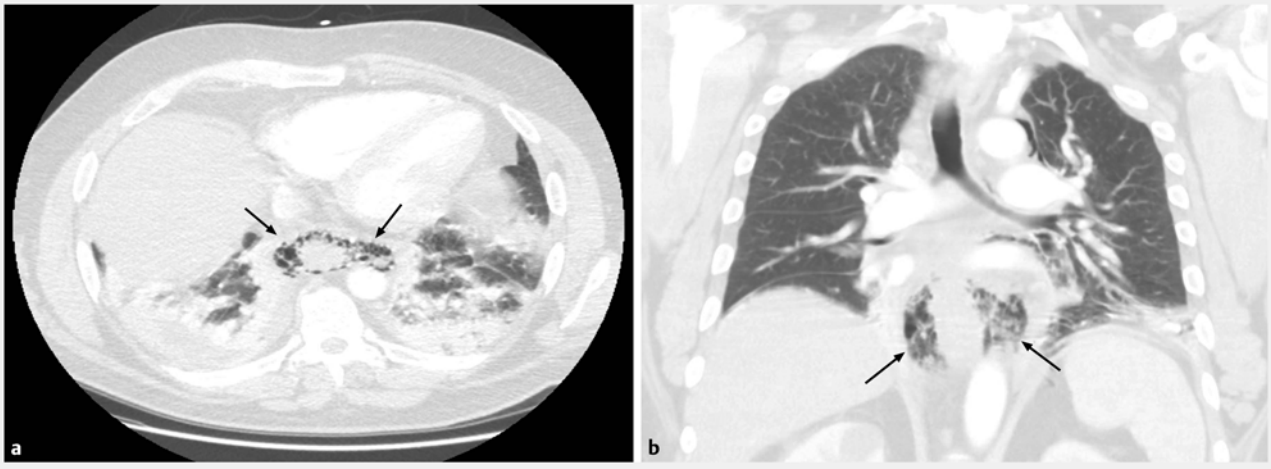
Results

We identified a total of 6 patients (▶ **Table 1**) who received non-surgical management of BS during the 5 years' time period. The median age at presentation was 55. There were 4 males and 2 females. The most common site of perforation was in the distal esophagus, just proximal (within 3 cm) to the gastroesophageal junction. The most common presenting symptom was chest pain (67 %) following an episode of vomiting or retching. Three patients (50 %) had underlying Barrett's esophagus. Four patients (66.7 %) developed septic shock requiring inotropic support, while 5 patients (83.3 %) developed respiratory failure requiring mechanical ventilation support. These patients mostly presented 2 to 3 days after the development of symptoms. Endoscopic treatment with a fully covered esophageal stent was the primary intervention used for all of these patients (100 %). IR was consulted for all patients. IR-guided interventions were aimed at drainage of the mediastinal/pleural fluid collection. Three patients required re-stenting (50 %). Two of them was due to stent migration and 1 due to continuous leakage at the perforation site. Endostiches and clips were used on these cases to prevent subsequent migration. Four patients required subsequent feeding tube placement, either jejunostomy or gastrostomy. Clinical resolution of the BS was achieved in all patients (100 %) without any subsequent surgical interventions (▶ **Table 2**). All the patients were discharged home following a median hospital stay of 19 days. There were no deaths within the study group. The average follow-up duration was 2 years.

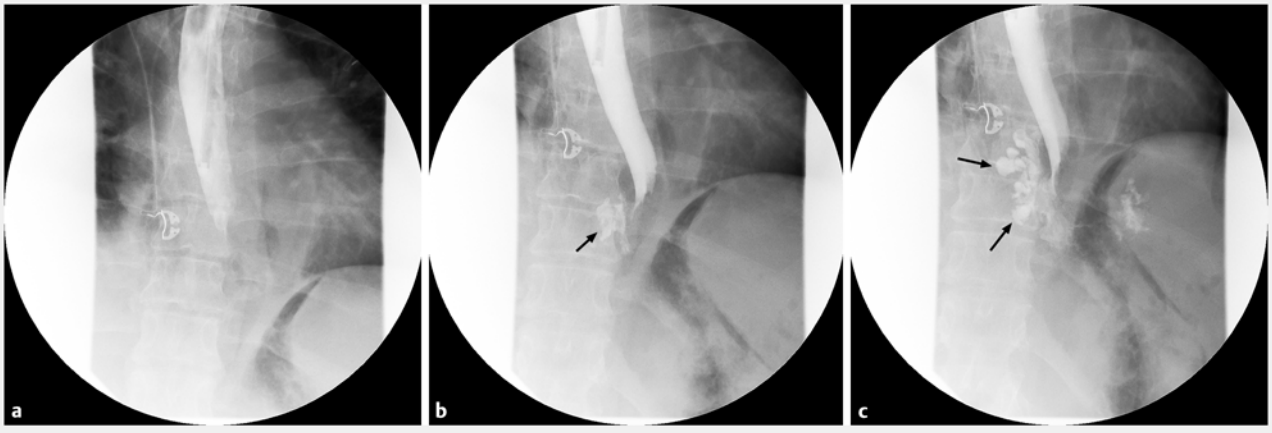
Discussion

After its initial description in 1724, BS has come to be considered the most lethal gastrointestinal perforation, with a mortality rate of close to 100 % without treatment. BS may be difficult to diagnose up front, with the possibility of an erroneous diagnosis at presentation in almost half of reported cases. In 90 % of the cases, the rupture is in the lower third of the esophagus and in the left lateral position. This is believed to be due to anatomic weakness.

Presenting symptoms for BS include vomiting (84 %), thoracic pain (79 %), dyspnea (53 %), epigastric pain (47 %), or dysphagia (21 %). Mackler's triad (thoracic pain, vomiting, and emphysema) is highly suggestive of BS, but this was found only in one-third of the cases. A physical examination may reveal subcutaneous emphysema and signs related to the development of hydropneumothorax. A radiographic examination may reveal pneumomediastinum (▶ **Fig.1**), pleural effusion, hydropneu-



▶ **Fig. 1** CT scan showing pneumomediastinum (Arrows) in the lower part of esophagus at the site of perforation in both Axial (**a**) and Coronal (**b**) view.



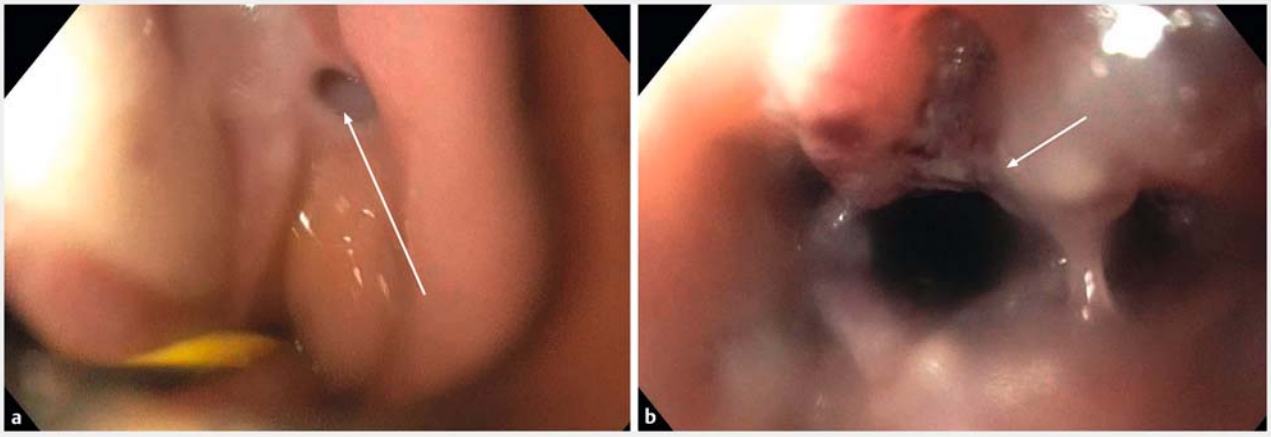
▶ **Fig. 2** Esophagogram (**a**) showing leakage of water soluble contrast material (arrows) into the right side of the chest and/or mediastinum (**b, c**).

mothorax, subcutaneous emphysema, and subdiaphragmatic air. However, it can be normal in around 12 % of patients.

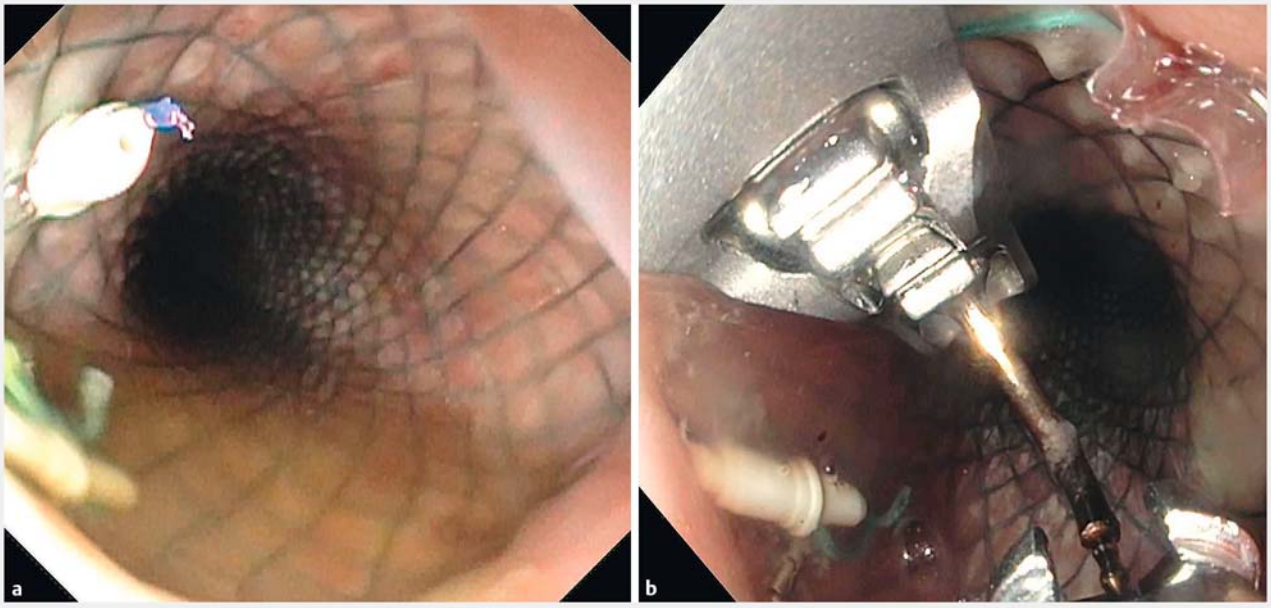
The gold standard for the diagnosis of BS is an esophagogram with a water-soluble contrast (▶ **Fig. 2**), which has a false-negative rate of up to 10 %. However, computer tomography with an oral contrast is able to illustrate a perforation and the surrounding inflammatory process (e. g., mediastinitis). Most authors recommend upper endoscopy to confirm BS (▶ **Fig. 3**) with a sensitivity and specificity of 100 % and 83 %, respectively [2]. There is, though, concern regarding increasing the size of the existing defect.

The treatment paradigm for BS seems to be evolving with less invasive modalities being introduced. Data on these newer and less invasive treatments are limited, as BS is a relatively uncommon condition. Moreover, it mostly occurs as a life-threatening emergency, which requires emergent actions to avoid fatal outcomes. Therefore, randomized prospective trials are difficult to implement. Further, most of the available literature on the treatment of esophageal ruptures is not specifically about BS and instead includes mostly cases of esophageal perforation or leaks. Interestingly, a BS diagnosis is usually delayed compared to iatrogenic rupture diagnosis because of the vague clinical picture, and this has detrimental effects on survival.

The available therapeutic options for BS mainly include conservative approaches with supportive and expectant management plans, as well as endoscopic or surgical interventions. Most of the previously published studies have established the time period between the onset of symptoms and diagnosis (less than 24 hours or greater than 24 hours) as the major factor influencing morbidity, mortality, and the overall outcome [3]. That is evident in our study group, as the patients who presented late had more complications like septic shock and empyema that affected the clinical success of treatment and the length of their hospital stay (▶ **Table 1**).



▶ **Fig. 3** Endoscopic images showing esophageal tear in patients with BS (**a, b**).



▶ **Fig. 4** Endoscopic images showing implemented esophageal stents with various techniques to prevent stent migration. In the first image (**a**) the stent was fixed using a clip. In the second image (**b**) the stent was fixed using Endostitch.

In general, the conservative treatment plan consists of strict oral intake; the initiation of feeding enterostomy or total parenteral nutrition; an intravenous, broad-spectrum antibiotic; and intravenous administration of proton pump inhibitors as well as fluids. The mortality rate with these measures in selected cases was reported to be between 20 % and 22 % [4].

Surgical intervention retains a dominant role in management, and this includes esophageal resection or chest drainage with or without esophageal repair. The primary repair of an esophageal perforation remains the gold standard of therapy, with a 94.7 % survival rate, provided the treatment is performed within 24 hours in the absence of esophageal diseases [4].

The use of self-expanding metal stents as a palliative measure for esophageal malignancies is well established and effective [5]. There is growing interest in the use of self-expanding covered metal stents in benign esophageal perforation, including BS. The endoscopic treatment aims mainly to prevent continued septic contamination and to guide the re-epithelialization of the esophageal mucosa with the esophageal stent placement (▶ **Fig. 4**) and/or endoscopic suturing. Closure of the mucosal defect also allows for early oral feeding. In addition, it results in a great reduction in morbidity and mortality associated with the surgical repair of esophageal perforations [6]. A stent placement was first reported for spontaneous

▶ **Table 3** All cases published on endoscopic treatment of Boerhaave's syndrome.

Author	# of cases	Age	Type of stent	Injury to stenting	Stent placement duration	Follow-up period	Complications and outcome
Dumonceau, et al. 1996 [11]	1	63	Ultraflex	8 weeks	40 weeks	10 months	Recurrent stricture and fistula
Eubanks, et al. 1999 [12]	1	61	Ultraflex	2 weeks	8 weeks	8 months	Asymptomatic
Yuasa, et al. 1999 [13]	1	56	Ultraflex	1 weeks	1 months	6 months	Dysphagia due stricture
Davies, et al. 1999 [14]	1	85	WallFlex	2 weeks	4 weeks	8 weeks	Asymptomatic
Chung, et al. 2001 [15]	3	55 – 58	Song/Niti-S	4 days to 4 weeks	8 – 42 weeks	6 – 32 months	Asymptomatic
Fischer, et al. 2006 [16]	5	44 – 77	Ultraflex	12 – 120 hours	3 – 4 weeks	2 months	1 died, others asymptomatic
Ghassemi, et al. 2007 [17]	1	91	Polyflex	10 days	10 weeks	1 month	Asymptomatic
Freeman, et al. 2009 [18]	19	26 – 69	Polyflex	6 – 78 hours	14 – 32 days	3 months	Leaks, stent migration, no death
Koivukangas, et al. 2012 [19]	14	47 – 81	Hanarostent/ Microtec	8 – 280 hours	13 – 59 days	6 months	2 died, others asymptomatic
Darrien, et al. 2013 [20]	5	30 – 75	Polyflex	24 – 72 hours	6 weeks	109 days	1 died, 2 had strictures
Ota, et al. 2014 [21]	1	56	CHOOSTENT	–	31 days	–	Asymptomatic
Van Weyenberg, et al. 2014 [22]	1	51	Hanaro	–	6 weeks	6 months	Asymptomatic
González-Haba, et al. 2016 [23]	6	63	Fully covered stent	–	8 weeks	–	Asymptomatic

esophageal rupture in 1995 [7], when a plastic-covered, self-expanding stent was used and the patient was discharged 15 days later. Unfortunately, the patient died of massive hematemesis as a result of esophageal necrosis about 8 weeks following the procedure.

The main drawbacks of stent placement are migration of the stent; adverse effects of the endoprosthesis on normal esophageal tissues, including pressure-induced ischemia, ulceration, and perforation; development of new reactive stenosis at the ends of the endoprosthesis; bleeding or injury upon removal; and unsuccessful retrieval of the device at a later date. When stents are placed at the esophagogastric junction, reflux esophagitis is an additional concern. In our case series, we addressed some of these issues by using endostitch to prevent stent migration and using a stent with low radial outward pressure to minimize normal tissue injury. In addition, all of our patients underwent percutaneous drainage of pleural effusion, empyema, and mediastinal abscess on a priority basis.

In terms of type of stents used, Boeckel et al. [8] found no difference between self-expanding metal stents (SEMS) or plastic stents and a clinical success rate of 85 % for different causes of esophageal perforation. Partially covered SEMS exhibit a favorable balance of a low migration rate and easy removability. Over-the-scope endoclips have been successfully used in the management of BS, provided the size of the perforation is less than 10 mm.

Even though the earlier the intervention, the better clinical success and overall prognosis, 4 patients in our series who presented late and developed septic shock underwent endoscopic treatment with good outcomes. Still, they required prolonged hospitalization. Darrien et al. [9] published similar results in a case series of 5 patients who were all septic and managed with minimally invasive endoscopic therapies. Four patients survived, while the fifth died because of co-comorbidities. In fact, many other studies published on BS endoscopic treatment in general showed satisfactory outcome (▶ **Table 3**).

The point of importance to note here is that this challenges current belief that endoscopic therapy should be reserved for patients who present early and in whom there is no associated sepsis, as Dickinson et al. [10] conclude in their recently published study. They compared endoscopic therapy to surgery for BS in 2 groups of patients. Although all of the patients achieved clinical resolution, the mortality rate (2/8) was higher, as well as the readmission rate (3/6), in the endoscopic therapy group. The inherent problem with this study and most BS studies is the sample size, as it is usually small. Also, we don't have much information about patient comorbidities and clinical presentations, and this information would help in determining a patient's prognosis, which could potentially affect the study results.

In conclusion, endoscopic treatment seems to be an effective management strategy for patients with BS. We also note

the satisfactory results in patients presenting with sepsis with mediastinal or pleural abscesses, presumably due to urgent IR-guided drainage and aggressive initial intensive care unit management.

Competing interests

None

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