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REVIEW PAPERS

BOERHAAVE`S SYNDROME– OVER 290 YRS OF SURGICAL EXPERIENCES. EPIDEMIOLOGY, PATHOPHYSIOLOGY, DIAGNOSIS

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Boerhaave's syndrome (BS) is a relatively rare condition which involves a spontaneous, full-thickness longitudinal rupture of a previously non-diseased oesophagus. Two years ago 290 years passed after the eminent Dutch physician Dr Herman Boerhaave produced a scientific report of a case of spontaneous rupture of the oesophagus for the first time.

Dr Herman Boerhaave produced an extensive 60-page-long work published in 1724, entitled 'Atrocis, nec descripti prius, morbi historia. Secundum artis leges conscripta', which may be translated as 'A report of a terrifying disease, previously undescribed. Recorded according to the rules of the medical profession'. He described a case of his 51-year-old friend Jan Gerrit van Wassenaer (1). Baron van Wassenaer, Rosenberg coat of arms, was a Great Admiral of the Dutch Fleet during its prime. He also held the function of the Prefect of Rhineland. He was a very gregarious man who enjoyed parties, good drink and gourmet food – a sybarite. On 28 October 1723 during a party he consumed half of baked duck, some bread, two baked larks, white cabbage and spinach lamb stock soup, some smoked and baked veal thymus and pears, grapes and a sweet cake for dessert. He finished his meal with a small amount of beer and Moselle wine. Having come back home he went for a horse-riding trip. He returned after a quarter of an hour due to symptoms of indigestion. Personal physician of the king Dr James de Buy gave baron a few glasses of Carduus Benedictus (blessed thistle root extract) in order to induce vomiting. During not particularly profuse vomiting baron van Wassenaer felt a 'sudden pull' in the upper abdomen and a feeling as if his stomach had displaced. Then strong, relentless pain appeared in the left half of the chest and upper abdomen. Dr H. Boerhaave was called in to help. He examined the patient and gave him milk mixed with flour to drink. Subsequently he performed bloodletting and enema and applied warm compresses. Despite the procedures used the baron's condition sys-

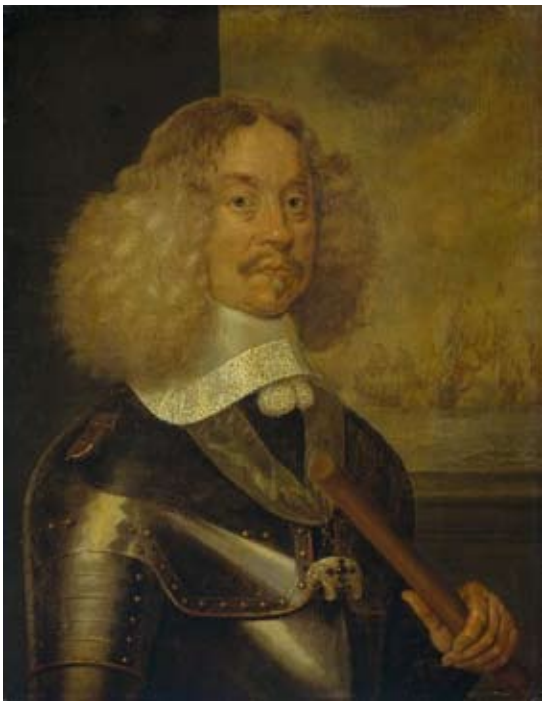

Fig. 1. Jan Gerrit van Wassenaer, Great Admiral

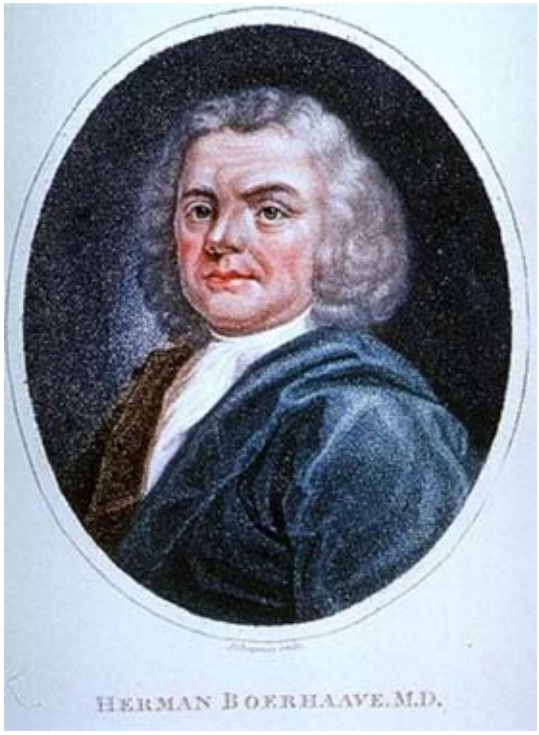

Fig. 2. dr Herman Boerhaave

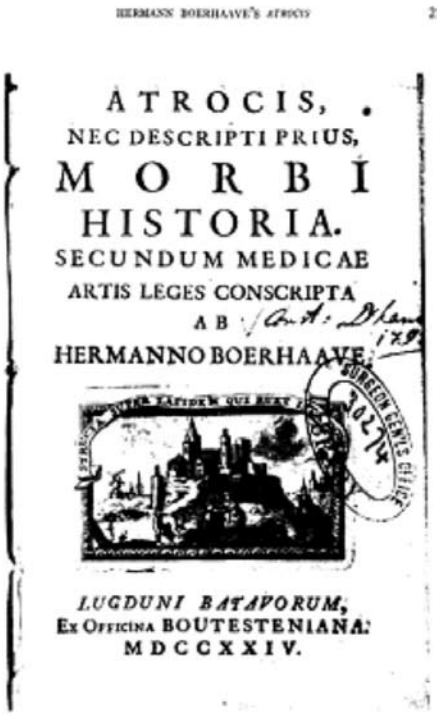

Fig. 3. Title page of the thesis

tematically deteriorated and after 16 hours from the onset of pain the patient died in horrible agony. Due to the ambiguous symptoms and difficulty in determining the cause of death autopsy was performed. A 1.5-inch rupture of the oesophagus at the left posterolateral wall just over the diaphragm, 900 ml of fluid similar to beer in the left pleural cavity and extensive subcutaneous emphysema at the anterior chest wall were found. In addition, the smell of baked duck emerged from the left pleural cavity. Ever since the publication spontaneous full-thickness rupture of the oesophagus has been called Boerhaave's syndrome (BS) after the name of the physician who produced the first scientific report of this condition.

Epidemiology

Reports of spontaneous oesophageal perforation are rare in scientific literature. In over 200 years from the first report of this condition the cases of only 50 patients were presented, the majority of whom died (2, 3). Another review of world literature dating back to 1980 covered only 300 cases of documented Boerhaave's syndrome (4). In 1986 Bladergroen M. et al. collected 127 cases of spontaneous perforation from the literature, 114 of which were diagnosed in live patients, while the remaining ones were autopsy findings (5). The largest literature review on the Boerhaave's syndrome is the work of Brauer et al. (6), who collected 989 cases of 1914–1995 from German, English, French and Italian sources. On the basis of the data the authors calculated mean age (52.4 years), gender distribution (men: 82%, women: 18%) mean size of rupture (3.3 cm) and mortality rates (50% for 1947–1980 and 34% for 1980–1995). One of the last works from 2002–2009 presents slightly over 300 patients (7). The data presented demonstrate that spontaneous perforation of the oesophagus is a very rare phenomenon. An epidemiological study of the whole population of Iceland corroborates this fact. It was found that the incidence of BS in that country was 3.1 cases/1 million citizens/year (8). Another example is the Netherlands, where approximately 10 cases of Boerhaave's syndrome are recorded every year (7). In Poland there are no detailed data; however, one may expect the situation to be similar on the basis of isolated reports of treatment outcomes (9, 10, 11).

Spontaneous oesophageal ruptures account for 10-15% of all oesophageal perforations and occur two to five times more frequently in men

than in women. They are usually found in men aged 40-60 years who abuse alcohol and like to eat large meals. Single cases of BS in patients over 90 years of age as well as neonates can also be found in the literature (5, 12-15).

Perforation of the oesophagus is the most fatal damage to the integrity of the digestive tract. Death is usually the consequence of infection including mediastinitis, pneumonia, pericarditis or pyothorax. Survival largely depends on early diagnosis and the introduction of appropriate surgical treatment. Overall mortality rates are close to 30%. The rates for the different types of perforation are 39% for spontaneous, 19% for iatrogenic and 9% for traumatic perforation (16,17,18). In BS cases every hour of delay of surgical treatment results in a 2% increase in the mortality rate (19). Patients who received surgical treatment up to 24 hours after the damage have 70–75% chances of survival. This rate is lower – 35–50%, if surgical treatment was introduced over 24 hours later and are close to 0% if the delay was larger than 48 hours (12, 19, 20).

Pathophysiology

The term 'spontaneous' is criticised by a number of authors, since the underlying cause of this condition is a sudden rise in internal oesophageal pressure. It is argued that it is caused by disturbed coordination of vomiting which is consciously suppressed. During vomiting rising pressure together with stomach contents is rapidly transferred to the oesophagus where it encounters consciously closed cricopharyngeus muscle. This results in a further increase of pressure in the oesophagus additionally intensified by intra-abdominal pressure. In extreme cases intra-oesophageal pressure may be as high as 290 mmHg (21). It ultimately leads to a longitudinal oesophageal rupture most commonly in the lower 1/3 of the thoracic oesophagus (80% of cases), 3–6 cm above the diaphragm at the left posterior wall, the damage measuring 2–10 cm. Other, much less common sites of rupture include the subdiaphragmatic and upper thoracic areas (5, 7, 22).

There are a number of theories trying to explain the location of a spontaneous perforation most commonly in the lower part of the thoracic oesophagus on the left side. One of them points to the lack of anatomical continuity of the muscular layer in this area due to the presence of muscle fibres in the submucosal connective tissue. Another explanation involves the differences in the structure of the connective tissue between diagonal and circumferential fibres. Another theory explains this fact by vessels and nerves penetrating the oesophageal wall in this area (22,23). A diffe rent theory attributes the location of perforation to the fact that the neighbouring organs adhere to the lower part of the thoracic oesophagus to a lesser extent than elsewhere, which is thought to increase the susceptibility to barotrauma (5,22). Th e actual pathology behind the condition still remains unclear.

The predisposing factors include alcohol abuse, asymptomatic gastroesophageal reflux disease (GERD) and hiatus hernia (5, 22, 23). Appreciating the different causes that might lead to the development of BS may help to establish the correct diagnosis. The examples of rare concurrence of BS and other conditions, complaints and activities are summarised in the tab. 1 (24).

The most important element of pathophysiology of BS that makes it so serious is rapidly deteriorating sepsis as a result of extensive (explosive rupture) infection of the mediastinum, pleural cavity, abdominal cavity and pericardial sac with saliva rich in enzymes and bacteria, gastric juice and bile (25).

Table 1. Concurrence of the Boerhaave's syndrome with other conditions, complaints and activities (24)

Trigger cause:
– bronchial asthma attack
– epileptic seizure
– parturition
– hyperemesis gravidarum
– vomiting:
a) during preparation for gastroscopy
b) during haemodialysis
c) during epidural block
d) during biliary colic
– rhinitis and sneezing
– prolonged cough and hiccup
– urge to defecate
– lifting of a heavy object
– during laughter
– during sleep
– rapid swallowing of a large piece of food
– Heimlich manoeuvre

Diagnosis

An important element of diagnostic investigation is thorough medical interview that indicates the consumption of an excessively large meal, alcohol abuse, violent vomiting or nausea and a rapid onset of very strong pain in the left chest. Vomitus may sometimes include traces of blood, while massive haemorrhage from the upper gastrointestinal tract is extremely rare. Some patients may additionally report cough, dysphonia, dyspnoea, voice change and increased thirst combined with drinking large amounts of cold water mitigating retrosternal pain. Less common symptoms include facial oedema, one-sided proptosis and excessive dilation of jugular veins (22, 24, 26, 27). Sometimes Hamman's sign may be present (in approx. 20% of patients) in a supine position. This involves distinctive crepitation heard over lung fields, synchronous with the heartbeat (28). The most characteristic feature of BS is the presence of three symptoms called Mackler's triad, which includes vomiting, strong chest pain and subcutaneous emphysema in the neck and chest (29). Unfortunately, subcutaneous emphysema, which is the most important symptom facilitating the right diagnosis, is found in only 14–30% of BS cases (16, 19). Anderson's triad may also be helpful, which includes subcutaneous emphysema in the neck and chest, tachypnoea as well as tenderness and tension of muscles in the upper abdomen (19). Reports of atypical or poorly symptomatic course of the disease in patients who present only with isolated symptoms such as chest pain without concomitant vomiting may be found in the literature.

It is assumed that 30–50% of BS cases have an atypical clinical course causing difficulty in documenting and determining the diagnosis (11, 14, 18, 24). The symptoms usually suggest gastric or duodenal ulcer rupture; as a result, in 9% of BS cases the abdominal cavity is unnecessarily opened (30). Differential diagnosis should also include such conditions as myocardial infarction, acute pancreatitis, pulmonary embolism or dissecting aortic aneurysm (7, 16, 19, 30). Diagnostic errors are made in over half of BS cases (30). As a result, sometimes only 5% of patients are correctly diagnosed before 12 hours after the damage and 35% before death (19). Numerous examples of diagnostic errors reported in the literature have been collected by Rokicki et al. (24) (2). In conclusion, the symptoms of the disease are often unstable and ambiguous, which leads to a false or delayed diagnosis, adversely affecting the chances of satisfactory treatment outcomes.

Table 2. Most common diagnostic errors (24)

The following have been diagnosed instead of Boerhaave's syndrome:
– perforated gastric or duodenal ulcer
– myocardial infarction
– pneumonia
– acute pancreatitis
– dissecting aortic aneurysm
– pneumothorax
– pulmonary embolism
– renal colic
– acute appendicitis
– lung abscess
– mesenteric artery embolism
– pericarditis
– splenic haemorrhage
– incarcerated diaphragmatic hernia

The suspicion of BS requires immediate and concurrent diagnostic activities as well as efforts to limit further superinfection of tissues in the area of the damage. The latter goal may be achieved by absolute prohibition of eating and drinking, urging the patient to spit out saliva, administration of wide-spectrum antibiotics and restoring the water-electrolyte balance. A plain chest X-ray film made at an early stage of diagnostic investigation enables one to visualise abnormalities in 81–90% of patients (19, 32, 33). The most common symptom is unilateral pleural effusion on the left side. It corresponds to the fact that the majority of BS perforations occur in the left posterolateral part of the thoracic oesophagus (5, 7, 22, 23). Other radiological signs may include the presence of pneumothorax, pneumothorax with fluid, mediastinal pneumothorax, subcutaneous emphysema, atelectasis or widened mediastinum (32, 33, 34). V-shaped accumulation of air in the mediastinum visible on a PA chest X-ray film (Naclerio V sign) is a distinctive sign of BS (34); unfortunately, it is found in only 10–20% of patients (19, 27, 35).

It is also known that in the first six hours after the perforation a chest radiograph may be normal in 10–33% of patients (16, 27, 34). A further step enabling one to determine the ultimate diagnosis of BS are contrast-enhanced

examinations of the oesophagus (7, 19, 20, 27, 32, 33). An oesophageal radiograph following the administration of an oral water-soluble contrast medium (e.g. 70% Uropolin) may reveal contrast leakage beyond the lumen of the oesophagus and shows the extent of perforation and its location, which facilitates the selection of the right surgical access. Using barium sulphate for contrast examinations of the oesophagus is advised against since it may exacerbate the symptoms of inflammation upon entering the mediastinum. False-negative results of a contrast examination of the oesophagus occur in 10–25% of patients. Such a situation may be caused by too high viscosity and spreading (e.g. of barium sulphate), too rapid passage (e.g. Gastrografin) or blockage of the perforation site by oedema, a clot or food remains (20, 32, 36). If a contrast examination of the oesophagus does not demonstrate contrast leakage beyond its lumen and the clinical index of suspicion of perforation is high, dynamic high-definition computed tomography may be a conclusive examination. Its sensitivity is estimated to be 92–100%. Dynamic CT may reveal additional changes such as effusion into the pleural and peritoneal cavities, accumulation of air around the loose tissues of the oesophagus or a mediastinal abscess. An additional advantage of computed tomography is the possibility to exclude a dissecting aortic aneurysm, perforated gastric or duodenal ulcer and acute pancreatitis, which are most commonly confused with BS. Unfortunately, a disadvantage of CT is the fact that it does not allow for particularly precise locating of the site of damage (20, 32, 33). Oral contrast-enhanced computed tomography is currently the diagnostic method of choice used at the present authors' department.

Oesophageal endoscopy for the ultimate diagnosis of BS attracts the same amount of support as it does opposition. According to the opponents of this solution it should be used exclusively when a wider diagnostic investigation is indispensable or if post-traumatic damage is present (33). They argue that gastroscopy requires insufflation of the oesophagus with air which may enlarge the damage and intensify the migration of air and infection into the mediastinum and pleural cavity (10, 11, 20, 25, 33). According to the supporters of endoscopy it allows not only for a very precise determination of the site of damage, but it also enables one to establish the length of rupture of the mucosa, which is of fundamental importance for surgical repair (16, 18, 19, 37).

Diagnostic investigation of BS should definitely include laboratory tests and microscopic examination of the fluid collected from the pleural cavity. The presence of acidic stomach contents (pH < 6.0), shed salivary gland cells, food remains or high activity of α-amylase (> 512 WU) are clear signs of oesophageal perforation (19, 20). The presence of a blue dye (methylene blue or gentian violet aqueous solution) in the pleural cavity fluid collected by way of paracentesis, earlier administered by mouth (32, 33) has a similar diagnostic utility.

Despite the fact that nearly 300 years have passed from the first report, currently the diagnosis of BS is usually delayed and the proposed forms of treatment are controversial. The opinion that surgical intervention performed as soon as possible as a life-saving procedure remains the standard of treatment is growing in popularity. This clearly indicates that early diagnosis may improve unfavourable prognosis.

REFERENCES

1. *Boerhaave H*. Atrocis nec descripti prius, morbi historia. Secundum medicae artis leges conscripta. Lugduni Batavorum Boutesteniana; 1724.
2. *Olson A, Clagett O:* Spontaneous rupture of the esophagus, report of a case with immediate diagnosis and successful surgical repair. *Postgrad Med* 1947; 2(6): 417-21.
3. *Barrett NR:* Report of case of spontaneous perforation of the esophagus successfully treated by operation. *Br J Surg* 1947; 35: 216-19.
4. *Kish GF, Katske FA:* A case of recurrent Boerhaave's syndrome. *W V Med J* 1980 Feb; 76(2): 27-30.
5. *Bladergroen M, Lowe JP, Postlethwai R:* Diagnosis and recommendet management of oesdophageal perforation and rupture. *Ann Thorac Surg* 1986; 42: 235-39.
6. *Brauer RB, Liebermann-Meffert D, Stein H* et al.: Boerhaave's syndrome: analysis of the literature and report of 18 new cases. *Dis Esophagus* 1997; 10: 64-68.

7. *de Schipper JP, Pul ter Gunne AF, Oostvogel HJM* Spontaneous rupture of the oesophagus: Boerhaave-s's syndrome in 2008. *Dig Surg* 2009; 26: 1-6.
8. *Vidarsdottir H, Blondel S, Alfredsson H* et al.: Oesophageal perforations in Iceland: a whole population study on incidence, aetiology and surgical outcome. *Thorac Cardiovasc Surg* 2010 Dec; 58(8): 476-80.
9. *Minkowski D, Komarowski G, Karoń J:* Zespół Boerhaave'a. *Pol Przegl Chir* 2003; 75 (12): 1221-24.
10. *Kula Z, Kłonowska-Majchrzak K:* Samoistne pęknięcie przełyku- opis przypadku. *Przegl Gastroenterol* 2008; 3(4): 192-95.
11. *Szeliga J, Jackowski M:* Zespół Boerhaave. *Pol Przegl Chir* 2011; 83(8): 523-26.
12. *Plott E, Jones D, Mc Dermott D* et al.: A state-of-the-art. Review of esophageal trauma: where do we stand? *Dis Esophagus* 2007; 20: 279-83.
13. *Rao KS, Malla K, Singh A* et al.: Boerhaave's syndrome unusual presentation in a 4 month old infant, a case report and review of literature. *Ann Pediatr Child Health* 2015; 3(2): 1053-57.
14. *Roy KP, Katz J:* Boerhaave syndrome. *Medscape* 2013 Mar 15.
15. *Brinster CJ, Singhal S, Lee L* et al.: Evolving options in the management of esophageal perforation. *Ann Thorac Surg* 2004; 77(4): 1475-83.
16. *Spapen J, De Regt J, Nieboer K* et al.: Boerhaaves syndrome: still a diagnostic and therapeutic challenge in the 21 century. *Case Reports in Critical Care.* Article ID161286, 4 pages http: // doi org/10. 1155/2013/161286.
17. *Jones WG, Ginsberg RJ:* Esophageal perforation: a continuing challenge. *Ann Thorac Surg* 1992; 53: 534-43.
18. *Cho JS, Kim YD, Kim JW* et al.: Thoracoscopic primary esophageal repair in patients with Boerhaave's syndrome. *Ann Thorac Surg* 2011; 91: 1552-55.
19. *Marinis A, Rizos S:* Boerhaave's syndrome or spontaneous perforation of the oesophagus. *Hellenic J Surg* 2011; 83: 258-62.
20. *Wang Y, Hang R, Zhon X* et al.: Our experience on management of Boerhaave's syndrome with late presentation. *Dis Esophagus* 2009; 22: 62-67.
21. *Maier A, Pinter H, Anneg U* et al.: Boerhaave^s syndrome: a continuing challenge in thoracic surgery. *Hepatogastroenterol* 2001; 48: 1368-71.
22. *Korn O, Zonate JC, Lopez R:* Anatomy of Boerhaave syndrome. *Surgery* 2007; 141: 222-28.
23. *Mc Farlane GA, Munro A:* Oesophageal injury: part 2. The changing face of the management of ruptured oesophagus: Boerhaave's syndrome. *Gullet* 1990; 1: 16-23.
24. *Rokicki M, Rokicki W:* Samoistne pęknięcie przełyku. Zespół Boerhaave. *Pol Merkuriusz Lek* 1996; 1: 348-50.
25. *Vial CM, Whyte RJ:* Boerhaave's syndrome: diagnosis and treatment. *Surg Clin N Am* 2005; 85: 515-24.
26. *Joungon J, Mc Bride T, Delcombe F* et al.: Primary oesophageal repair for Boerhaave's syndrome whatever the free interval between perforation and treatment. *J Cardiothorac Surg* 2004; 25: 475-79.
27. *Khan AZ, Strauss D, Mason RC:* Boerhaaves syndrome: diagnosis and surgical management. *Surgeon* 2007; 5: 39-44.
28. *Hamman LV:* Spontaneous mediastinal emphysema. *Bulletin J Hopkins Hospital:* Baltimore. 1939; 64: 1-21.
29. *Mackler SA:* Spontaneous rupture of the esophagus an experimental and clinical study. *Surg Gynecol Obstet* 1952; 95: 345-56.
30. *Shenfine J, Dreszer SM, Vishvanath Y* et al.: Management of spontaneous rupture of the esophagus. *Br J Surg* 2000; 87: 362-73.
31. *Cho S, Jheon S, Ryu KM* et al.: Primary esophageal repair in Boerhaave's syndrome. *Dis Esophagus* 2008: 21: 660-63.
32. *Janjua KJ:* Boerhaave's syndrome. *Postgrad Med J* 1997; 73: 265-70.
33. *Chirica M, Champault A, Dray Y* et al.: Perforations de l'oesophage. *J Clin Visc* 2010; June: 147(3): 169-81.
34. *Hegenbarth R, Birkenfeld P, Beyer R:* Roentgen findings In spontaneous esophageal perforation (Boerhaave's syndrome). *Aktuelle Radiol* 1994; 4(6): 337-38.
35. *Naclerio E:* The V sign in the diagnosis of spontaneous rupture of the esophagus (an early Roentgen clue). *Am J Surg* 1957; 93: 291-98.
36. *Veno S, Eckardt J:* Boerhaaves syndrome and tension pneumothorax secondary Norovirus forceful emesis. *J Thorac Dis*. 2013; 5(2): e38-e40.
37. *Schweigert M, Beattie R, Solymosi N* et al.: Endoscopic stent insertion versus primary operative management for spontaneous rupture of the esophagus (Boerhaave syndrome): an international study comparing outcome. *Amer Surg* 2013; 79(6): 634-40.

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