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Neonatal and pediatric esophageal perforation

Rebecca M. Rentea, MD, Shawn D. St. Peter, MD*

Deparment of Surgery, Children's Mercy Hospital, 2401 Gillham Rd, Kansas City, Missouri 64108

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ABSTRACT

Esophageal perforation (EP) is a rare complication that is often iatrogenic in origin. In contrast with adult patients in whom surgical closure of the defect is preferred, nonoperative treatment has become a common therapeutic approach for EP in neonates and children. Principles of management pediatric EP includes rapid diagnosis, appropriate hemodynamic monitoring and support, antibiotic therapy, total parenteral nutrition, control of extraluminal contamination, and restoration of luminal integrity either through time or operative approaches.

Introduction

Esophageal perforation (EP) is a rare complication of enteric instrumentation in children commonly after endoscopy for stricture dilation in the pediatric or nasogastric tube insertion in the neonatal population.[1,2] In this review, we briefly examine the historic literature, history, anatomy, incidence, etiology, management-operative and nonoperative EP, and outcomes. We additionally discuss in detail some of the specific causes of EP including placement of neonatal iatrogenic EP, esophageal dilation, foreign bodies, Boerhave's syndrome, and transesophageal echocardiography.

History

The first recorded case of EP was described in 1724 by Hermann Boerhaave following an autopsy.[3] The first report of neonatal esophageal perforation by James Fryfogel in 1952 was a spontaneous rupture proximal to an esophageal web, however the literature increasingly suggests that iatrogenic causes are far more common.[4,5] The first reported iatrogenic case of neonatal EP was due to a stiff rubber catheter used for suctioning in 28 day old infant in 1961which was repaired via a right thoracotomy.[6]

Anatomy

The esophagus lacks a serosa and is surrounded by loose stromal connective tissue, allowing infectious and inflammatory response to disseminate easily, making EP a medical emergency.[7,8] Perforation allows bacteria, usually polymicrobial flora, and digestive enzymes to spread into the mediastinum or subphrenic space leading to mediastinitis, empyema, abscesses, sepsis, and multiple organ dysfunction syndrome.[4,9]

In neonates, the injury site is often at the pharyngoesophageal junction, proximal to the cricopharyngeal muscle.[10] This is hypothesized to occur for two reasons (Figure 1). The first is that it is the narrowest point of the esophagus and that instrumentation may cause a reflex muscular constriction and closure of the esophagus with further attempts at intubating the esophagus leading to perforation. The other reason is that on hyperextension of the neck,[11,12] the posterior esophageal wall is compressed against the cervical vertebrae and attempts at intubation may lead to the perforation.[13] On the other hand, in pediatric patients the most common location for perforation is in the thoracic esophagus.[1,8]

A perforation may be characterized as submucosal or free. Submucosal perforation results in a blind pouch or tract that may be indistinguishable from esophageal atresia, requiring a high degree of clinical suspicion to accurately diagnose the injury.[14] Free perforation results in an aberrantly positioned enteric tube, often causing a right pneumothorax due to the close apposition of the aorta to the left pleural cavity.[15] There have been reports however of enteric tubes migrating into other structures including the pericardial sac,[4,16] the right renal pelvis,[17] and even the urinary bladder.[18]

Incidence

EP is a relatively rare diagnosis. The incidence of iatrogenic EP has been evaluated most extensively. The largest series of children with EP identified only 24 patients over a 20-year period at a tertiary pediatric surgical center.[1] A 14-year review of 39,650 upper gastrointestinal endoscopies, in patients 2–90 years of age, reported only 2 esophageal

* Corresponding author.
E-mail address: sspeter@cmh.edu (S.D. St. Peter).

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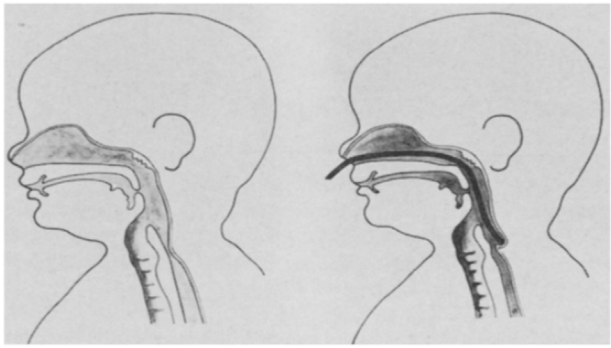

Fig. 1. Iatrogenic esophageal injury usually occurs at the pharyngoesopahgeal junction where the lumen is narrowed by the circopharyngeus muscle.[13]

perforations (0.006%). However, when a therapeutic measure was incorporated, such as stricture dilation, the incidence of perforation increased to 73 in 12,366 patients (0.6%).[19]

Etiology

EP can be difficult to diagnose and manage. The overall mortality associated with EP approaches 20%–30%. Treatment delayed by >24 hours in 50% of patients following perforation resulting in a doubling of mortality.[20,21] The pediatric literature documents 71%–84% of cases of esophageal perforation caused by an iatrogenic source.[1,4,5,15,22] The most common cause of iatrogenic EP in the pediatric population is from stricture dilation, while in the neonatal population is nasogastric tube placement.[23]

The causes of EP are varied and abundant (Table 1) and are outlined in order of descending incidence in Table 2. Etiologic factors of EP may be separated into the following four groups: (1) instrumental/iatrogenic (endoscopy and trauma),[4,5,8,24–27] (2) pressure induced or spontaneous (i.e., Boerhaave syndrome and barotrauma),[28,29] (3) ingestion of foreign body (coins and laundry detergent capsules),[30] and (4) previous esophageal pathology (i.e., diverticula, stricture, and at the junction with a Bochdalek hernia).[9,31,32]

Clinical presentation

The cause, location of the injury, age, as well as the interval between perforation and diagnosis all determine the clinical features of EP. Dysphagia (60%) is the most common presenting symptom followed by dyspnea and fever.[1] Cervical EP is generally less severe as the spread of contamination from the retroesophageal space to the mediastinum is slowed by attachments of the esophagus to the prevertebral fascia.[33] Inflammatory changes may not develop for hours and sepsis may be delayed up to 24 hours.[31] Cervical perforation can present with neck pain, cervical dysphagia, dysphonia, or bloody regurgitation and subcutaneous emphysema.[8]

Thoracic perforations rapidly contaminate the mediastinum which can result in systemic sepsis and shock within hours.[20] Intrathoracic perforations present with chest pain, tachycardia, tachypnea, fever, and leukocytosis.[8] If the integrity of the pleura is maintained, salivary organisms and gastric contents infiltrate the mediastinum and produce mediastinal and later cervical emphysema. Initial chemical mediastinitis is followed by bacterial invasion and severe mediastinal necrosis. Exacerbated by negative intrathoracic pressures, pleural rupture by mediastinal inflammation, or direct perforation result in pleural effusion.

Intra-abdominal esophageal perforations often present with peritonitis or referred shoulder pain due to the irritation of the neighboring hemi-diaphragm. Systemic signs occur much more rapidly, usually within a few hours.[31]

Infants and premature neonates are unique in that they manifest different signs and symptoms.[15] The diagnosis of EP should be considered in any infant presenting with hypersalivation, choking, coughing, or cyanosis after repeated or difficult attempts at endotracheal or enterogastric intubation.[10,15] A contained mediastinal perforation may produce an extrinsically compressing, obstructive mass that may mimic esophageal atresia.[34] In a neonate thought to have esophageal atresia, blood-tinged or coffee ground aspirate from a nasogastric or orogastric tube is more suggestive of EP.[10,21,35] A free perforation into the pleural cavity with consequent pneumothorax or pleural effusion may present with acute respiratory distress and rapid clinical deterioration[34] and possibly formula coming from a thoracostomy tube after placement in these patients.[13] Additionally, EP should be considered in the micropreemie due to minimal symptoms or signs when difficulty with advancement of the enterogastric tube is encountered.

Table 1
Etiology of esophageal perforations. (Modified with permission from Gupta and Kaman.[22])

Endoscopic
Diagnostic endoscopy
Endoscopic biopsy
Endoscopic dilations
Variceal sclerotherapy
Nasogastric tube placement
Endotracheal intubations
Transesophageal echocardiography (TEE)
Missed tracheoesophageal atresia/fistula
Foreign bodies
Operative antireflux procedures
Trauma
Blunt
Penetrating
Non-accidental trauma/child abuse
Pneumatic barotrauma
Spontaneous or Boerhaave's syndrome
Caustic agents
Acid and alkali
Mallory–Weiss tear
Infective causes: tuberculosis, immunodeficiency, candida, herpes, and syphilis
Mediastinal and cervical tumors—rare
Malignancy of esophagus, lung, or other mediastinal structures—rare

Imaging and diagnostic adjunctions

Imaging provides an adjunct to the diagnosis of EP. Early recognition of EP is critical and ensure the best possible outcome. Any child who becomes symptomatic after an endoscopic or esophageal dilation procedure must be evaluated for the presence of EP.[8]

Diagnosis is usually made by plain X-ray and is a useful initial diagnostic modality, although plain films may be normal in 12%–33% of cases.[36] Plain radiography may demonstrate the abnormally located enterogastric tube, pneumothorax, pleural effusion, or pneumodiastinum, providing clues as to the location of the perforation.[31] In instances where the clinical history is unclear, an esophagram may be warranted to rule out other differentials, such as esophageal atresia. Some institutions obtain an esophagram to confirm and localize EP.[37] However, this study does not definitively rule out a leak.[15] One study documented a false negative rate of approximately 10%.[38] A dilute barium contrast may increase the sensitivity of the test.[20] In contradistinction, a single institution study evaluated the utility of follow-up barium study following a negative water-soluble contrast examination and found both to be negative in all 46 patients (age: 10 days to 17 years). This study indicated that a single-contrast water-soluble esophagram alone is 100% sensitive in the diagnosis of esophageal injury or leak, and that follow-up barium esophagram only increases