Esophageal Perforation in the Newborn

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Abstract
Esophageal perforations in newborns can occur for a multitude of reasons and are best classified as iatrogenic or spontaneous. Spontaneous perforation is extremely rare and more commonly encountered in full-term infants. Iatrogenic perforations are more often seen in premature, small for gestational age infants and usually occur in the cervical esophagus or hypopharynx. Frequent causes for iatrogenic perforation include pharyngeal suctioning, laryngoscopy, attempted esophageal intubation, and digital manipulation of the neonatal head during breech delivery.

With early diagnosis and close monitoring, even more distally located iatrogenic perforations can often be managed conservatively with good outcomes. Early diagnosis of this condition allows for further surgical treatment options such as closed-chest drainage, primary repair, or stenting. Delayed diagnosis may result in the inability to repair the injury primarily, a significant increase in mortality, and the eventual need for esophageal replacement among survivors.

Keywords
Esophageal perforation · Boerhaave’s syndrome · Cervical esophagus · Hypopharynx · Hydropneumothorax · Pneumomediastinum · Esophagography · Thoracostomy
Introduction

Esophageal perforation can occur for a multitude of reasons in neonates. This condition was first reported in the literature by Eklof and colleagues (Eklof et al. 1969). Over the past 40 years, perforation of the esophagus in extremely premature neonates has become increasingly recognized and reported. Spontaneous perforation of the esophagus (neonatal Boerhaave’s syndrome) is extremely rare, and Fryfogle performed the first successful repair (Fryfogle 1952). Although non-operative management techniques are often successfully employed, neonatal esophageal perforations may be fatal without early diagnosis and treatment, and aggressive surgical therapy is occasionally warranted (Michael et al. 1981; Van der Zee et al. 1966; Gander et al. 2009; Hesketh et al. 2015; Garey et al. 2010). Surgeons must continue to play an early and active role in the individualization of care in these patients.

Classification and Etiology

Esophageal perforations in newborns are best classified as iatrogenic or spontaneous. Spontaneous perforations are extremely rare and are more commonly encountered in full-term infants. The most common site of perforation is the lower third of the esophagus. Although the exact cause in individual cases may remain unclear even after treatment, possible etiologies for spontaneous perforation include increased intra-abdominal pressure at delivery, existing weakness in the esophageal wall (either congenital or from a perinatal ischemic event), and reflux-associated peptic esophagitis (Aaronson et al. 1975).

Iatrogenic perforation of the esophagus is more commonly seen in premature, small for gestational age (SGA) infants and usually occurs in the Pharyngoesophageal junction or cervical esophagus (Rentea et al. 2017; Mileder et al. 2016; Geoghegan et al. 2014, Ducharme et al. 1971). Specific causes in this setting include pharyngeal suctioning, laryngoscopy, attempted esophageal intubation, and digital manipulation of the neonatal head during breech delivery. All of these maneuvers have the potential to cause injury to the delicate hypopharynx and proximal esophagus in premature neonates, and all have been reported in the literature (Astley and Roberts 1970; Su et al. 2009; Lee and Kuhn 1976; Wychulis et al. 1969).

During instrumentation, particularly when the neck is hyperextended, perforation may occur at the level of the cricopharyngeus muscle, as a result of compression of the posterior esophageal wall against the cervical vertebrae. The initial injury by a laryngoscope blade or endotracheal tube may be submucosal, resulting in a pseudodiverticulum. The associated cricopharyngeal spasm predisposes the weakened area to full thickness perforation during subsequent manipulations. Endotracheal intubation, especially in premature, SGA neonates, may further compromise the esophageal introitus. Subsequent oropharyngeal suctioning or insertion of a nasogastric tube may similarly extend the submucosal injury into a full thickness perforation (Girdany et al. 1969).

Iatrogenic injury to the mid-esophagus is usually associated with dilatation of a stricture or a postoperative anastomotic leak following esophageal atresia repair (Sloan and Haight 1956; Eraklis and Gross 1966). An improperly placed chest tube may cause disruption of a fresh esophageal anastomosis or may penetrate a proximal myotomy site (Johnson and Wright 1990). Chest tube-related direct pressure necrosis of an otherwise normal esophagus has been reported in a premature neonate (Cairns et al. 1999). As transesophageal echocardiography has become increasingly used in the evaluation and management of congenital heart disease in neonates, perforations of the proximal and middle esophagus have been reported with this procedure (Miller et al. 2015; Mukerideen-Russell et al. 2001).

Diagnosis and Clinical Manifestations

Newborns with iatrogenic esophageal injury frequently present with hypersalivation, choking or coughing. Many will have overt respiratory distress, either from aspiration of oral secretions or the development of hydropneumothorax. The
examiner will have difficulty passing a nasogastric tube, either as the inciting event or as a result of swelling or a false tract created by prior instrumentation. In patients who are not acutely symptomatic, abnormal position of the nasogastric tube on post-placement chest X-Ray is commonly the first indication of esophageal perforation (Ducharme et al. 1971; Soong 2007).

In neonates, the presence of blood-tinged oral secretions after endotracheal intubation indicates the potential for iatrogenic injury and justifies serial X-ray examinations of the chest. The proper diagnosis often will be missed in the absence of such an examination. The symptoms of perforation may not become evident until the child develops esophageal obstruction and may be difficult to distinguish from esophageal atresia if the event occurred close to the time of birth (Ducharme et al. 1971; Su et al. 2009; Wychulis et al. 1969). Esophageal perforation may be differentiated from atresia of the esophagus by an asymptomatic interval after birth, a history of repeated attempts at intubation or vigorous suctioning, absence of prenatal polyhydramnios, and the position and course of the nasogastric tube on chest X-ray (Ducharme et al. 1971). Some perforations of the esophagus create respiratory distress secondary to the development of hydro or pneumothorax. In these cases, the right pleural space is most commonly affected (Michael et al. 1981; Gander et al. 2009; Su et al. 2009; Wychulis et al. 1969). Thoracentesis or tube thoracostomy will typically yield serosanguinous fluid or the contents of the previous feeding. In the case where persistent cloudy drainage occurs following chest tube placement, chylothorax should be suspected (Kairamkonda 2007).

Anteroposterior and lateral X-rays of the chest and neck are indicated in any suspected case of perforation. Hypopharyngeal and cervical perforations frequently demonstrate extraluminal air in the neck, without pneumomediastinum initially. Mid-esophageal perforations may demonstrate pneumomediastinum, pneumothorax, or hydrothorax. An unusual course of the nasogastric tube (pleural cavity, pericardial cavity (Fig. 1) or right side of the mediastinum (Fig. 2)) supports

Fig. 1 Lateral radiograph showing aberrant course of nasogastric tube, coiled in pericardial sac

Fig. 2 AP radiograph demonstrating aberrant course of nasogastric tube, with tip positioned on right side of mediastinum
the diagnosis and may be confirmatory. Widening of the mediastinum and blurring of the mediastinal margin indicate the development of mediastinitis but are later findings. The absence of these findings does not rule out an injury, but the demonstration of these changes, in the absence of other confirmatory signs, should prompt contrast esophagography. Three types of injury patterns are seen in premature neonates undergoing contrast evaluation of the esophagus: (1) a pharyngeal diverticulum created by a local cervical leak; (2) a mucosal perforation extending posteriorly in parallel to the esophagus; and (3) a free intrapleural perforation where there is obvious leakage of air and esophageal contents into the pleural cavity (Mollit et al. 1981).

In cases where chest X-ray demonstrates the nasogastric tube to be located in the pleural cavity or pericardium, the diagnosis of esophageal perforation can be confirmed. Localization of the site of perforation may be unnecessary in these cases, unless the patient’s clinical condition deteriorates after removal of the nasogastric tube appropriate conservative management. If symptoms suggest esophageal obstruction, esophagography should be performed by administering a small quantity of diatrizoate meglumine (Hypaque), diatrizoate sodium (Renografin), or metrizamide into the proximal esophagus; Gastogarfin and barium should be avoided due to the risk of worsening mediastinitis from extravasation or pneumonitis from aspiration. In cases of pharyngeal-esophageal perforation, cricopharyngeal spasm may be so severe that no contrast material will enter the native esophagus. In these cases, several clues may help to differentiate submucosal perforation with secondary esophageal obstruction from congenital esophageal atresia (Blair et al. 1987).

These include:

- An increased distance between the posterior wall of the trachea and opacified tract on lateral X-ray, as the pouch in congenital esophageal atresia is usually closely associated with the trachea
- A longer, narrower, and more irregular opacified tract then is seen in esophageal atresia, which typically shows a bulbous uniform pouch
- Lack of compression of the posterior trachea by the opacified tract on lateral X-ray, as the proximal pouch in esophageal atresia typically causes tracheal compression

Endoscopic evaluation of the esophagus is typically not indicated at the time of diagnosis, as it has the potential to worsen the injury.

Spontaneous perforation of the neonatal esophagus usually presents with respiratory distress, which presents within minutes to hours after the perforation occurs. These patients are usually quite symptomatic. In contrast to the pattern typically seen in older children and adults, there is a greater predilection for right-sided hydropneumothorax in neonatal Boerhaave’s syndrome (Aaronson et al. 1975). This may be explained by the close adherence of the aorta to the left side of the esophagus in neonates, which provides an additional mediastinal barrier on the left side. If the perforation remains undiagnosed, respiratory distress will worsen with subsequent feedings. Esophagography must be performed in all suspected cases of spontaneous perforation with hydropneumothorax to evaluate the extent and location of the injury.

Management

The overall mortality rate in neonates with esophageal perforation (4%) is significantly less than that in older children and adults (25–50%). (Hesketh et al. 2015; Garey et al. 2010; Engum et al. 1996). The management of this condition should follow a selective approach which depends upon the size of the patient, the location and nature of the injury, the time between injury and initiation of therapy, and the neonate’s response to initial conservative management when this is employed (Garey et al. 2010; Mollit et al. 1981; Johnson et al. 1982; Krasna et al. 1987). Despite the frequency with which conservative therapy is successful in this condition, esophageal perforation can be a rapidly fatal condition that requires
immediate recognition and aggressive management for a successful outcome. As a result, pediatric surgical involvement is warranted early in the evaluation process. Small submucosal perforations of the hypopharynx and esophagus, limited to the mediastinum and without systemic symptoms, may be managed by nonoperative methods. Actual localization of the perforation is not essential in these infants. If the nasogastric tube is noted in the mediastinum or pericardial cavity, the tube can be withdrawn and a new tube placed under fluoroscopic control. A broad spectrum antibiotic should be administered for 7–14 days. IV fluids and hyperalimentation should be given, as oral feedings are withheld. Esophagography should be performed 7–10 days after the injury. If the perforation is completely healed, oral feeding may be resumed. If the perforation has not healed during this interval, conservative treatment for another week will often lead to complete healing.

Routine surgical intervention does not appear to improve the rate of survival in these newborns. Tube thoracostomy should be placed when the chest X-ray indicates pneumothorax (Fig. 3), hydrothorax, or increasing pneumomediastinum. All newborn infants with esophageal perforation must be carefully monitored during treatment (Ramareddy and Alladi 2016), including the use of white blood cell counts or C-reactive protein levels, platelet counts, blood gas analyses, and chest X-ray evaluation. If there is clinical deterioration or respiratory compromise, and closed chest drainage does not handle the leak, direct repair of the perforation is indicated.

Long, linear perforations to the lower end of the esophagus typically require thoracotomy, debridement of the necrotic edges, and primary repair of the defect with pleural flap coverage. A gastrostomy tube may be inserted in these situations to facilitate gastric decompression and limit gastroesophageal reflux during healing. When a secure direct repair of the perforation is not technically feasible because of scarring, inflammation, and tissue friability, diverting cervical esophagostomy with closure of the perforated area and concomitant gastrostomy is indicated. If at all possible, efforts should be made to preserve the native esophagus in an effort to avoid future esophageal replacement.

If there is a delay of more than 48 h in the diagnosis of a spontaneous perforation of the esophagus, unprotected primary repair often cannot be safely accomplished. After adequate debridement, local esophagectomy with closure of the proximal and distal esophagus, proximal esophagostomy, and gastrostomy tube placement should be considered. Critically ill newborns can be successfully managed with chest tube drainage, cervical esophagostomy (with or without ligation of the cardio-esophageal junction), and gastrostomy (Urschel et al. 1974). Broad spectrum antibiotics, IV fluids, and hyperalimentation should be continued until clinical signs of sepsis resolve. When the cardio-esophageal junction is closed, gastrostomy feedings may be attempted after 48 h. In cases where extensive debridement or resection is necessary and adequate native esophagus cannot be preserved, esophageal substitution will ultimately be necessary. This is typically done after an interval of at least 6 months, to allow growth and resolution of mediastinal inflammation.
Perforations that occur following dilation of esophageal anastomotic strictures are usually managed nonoperatively, as long as the leak is contained or can be adequately drained by tube thoracostomy. These perforations may take some time to heal if there is obstruction distal to the site of perforation. In the last decade, endoscopic stenting has been reported as a means of containing leakage and promoting healing in these cases. The use of this technique is currently limited in the neonatal population by the size constraints of commercially available stents (Ahmad et al. 2016; Rico et al. 2007).

Onwuka et al. (Onwuka et al. 2016) recently reported the largest series of neonates treated for esophageal perforations and found that nonoperative management with bowel rest, parental nutrition and antibiotics was successful in 96% of neonates.

**Conclusion and Future Directions**

Perforation of the neonatal esophagus, especially of the iatrogenic variety, is more common than reported in the literature and may be fatal without early diagnosis and management. The incidence of recognized pharyngeal and esophageal perforations is low, however, considering the large number of pharyngeal instrumentations that are performed on premature infants in the modern NICU. Gentle laryngoscopy with proper visualization of the vocal cords during intubation, avoidance of protruding stylets, careful suctioning of the pharynx, and avoidance of forceful nasogastric tube placement are all essential factors in the prevention of these injuries.

It is generally accepted that most iatrogenic perforations of the esophagus in newborns are cervical, occurring as a result of attempted intubation of the trachea or esophagus. With early diagnosis, even more distally located iatrogenic perforations can often be managed conservatively with good outcomes. However, these infants should be monitored closely. If they develop systemic illness, operative intervention may be indicated. Early diagnosis of this condition allows for more treatment options, which include nonoperative therapy, closed-chest drainage, and primary repair. Delayed diagnosis may result in the inability to repair the injury primarily, a significant increase in mortality, and the eventual need for esophageal replacement among survivors. Surgical consultation is warranted in all cases of esophageal perforation to allow timely and selective management, thereby limiting both mortality and long-term morbidity.

**Cross-References**

- Anesthesia and Pain Management
- Congenital Esophageal Stenosis
- Esophageal Atresia
- Gastroesophageal Reflux and Hiatus Hernia
- Specific Risks for the Preterm Infant

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