Hypopharyngeal Perforation Mimicking Esophageal Atresia: A Case Report of an Extremely Low Birth Weight Infant Emphasizing the Need for Preoperative Endoscopy

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We report a case of an extremely low birth weight premature infant born at 27 weeks of gestation, transferred to our tertiary pediatric referral center for surgical repair of an esophageal atresia. Endoscopic evaluation before the start of surgery revealed a hypopharyngeal perforation, resulting in the false impression of esophageal atresia. If no tracheoesophageal fistula is found during tracheoscopy, esophagoscopy should be done before surgical intervention as the inability to pass a nasogastric tube into the stomach is not sufficiently reliable for a correct diagnosis of esophageal atresia. (A&A Practice. 2021;15:e01414.)

GLOSSARY

CARE = Case Report; **Fio_2** = inspiratory fraction of oxygen

The diagnosis of esophageal atresia should be confirmed before attempts at repair because of the risks of surgery, especially in infants with extremely low birth weight.[1] Unfortunately, false and missed diagnoses have been described repeatedly over the past decades.[2] Despite multiple reports of conditions which may be misinterpreted as esophageal atresia, controversy remains whether bronchoscopic examination is mandatory before surgical interventions.[3–5] Tracheobronchcoscopy is still not performed in all cases of presumed esophageal atresia before surgical interventions, even though such endoscopy is safe and can be expeditiously performed. In addition, practitioners of pediatric anesthesiology and surgery should be aware that all preoperative history and evaluations, including radiographs, need to be critically evaluated for evidence that might contradict the diagnosis of esophageal atresia, such as what was seen in this case. Tracheoscopy and esophagoscopy are safe, simple, and quick procedures for trained personnel. This article adheres to the Case Report (CARE) guideline. Parents provided a written informed consent.

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Accepted for publication January 14, 2021.

Funding: None.

The authors declare no conflicts of interest.

Supplemental digital content is available for this article. Direct URL citations are provided in the HTML and PDF versions of this article on the journal's website (www.cases-anesthesia-analgesia.org).

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DOI: 10.1213/XAA.0000000000001414

CASE DESCRIPTION

We report the case of an extremely low birth weight premature female infant, born at 27 weeks of gestational age. Pregnancy followed in vitro fertilization in a 34-year-old secundigravida. The course of the pregnancy was complicated by premature rupture of membranes at week 19. Preterm labor started by the second half of gestational week 26. Following tocolysis and 2 cycles of corticosteroids to accelerate antenatal lung maturation, the infant was born by an uneventful cesarean delivery with a birth weight of 980 g. Neonatal adaptation was poor (Apgar scores: 5/5/7). The newborn did not stabilize with initial bag-mask ventilation and demonstrated persistent bradycardia and an undetectable peripheral oxygen saturation. Tracheal intubation was performed 6 minutes after birth and was successful with the second attempt (uncuffed, 2.5 mm inner diameter). After administration of surfactant 10 minutes after birth, the heart rate accelerated above 100 beats per minute for the first time and pulse oximetry became measurable at levels of 80% with ventilation with an inspiratory fraction of oxygen (Fio_2) of 1.0. As the respiratory functions improved further, the Fio_2 was subsequently reduced to 0.3.

Advancement of a 6 charrière nasogastric tube was not possible. A chest radiograph showed that the tip of the nasogastric tube was stuck at the level of the upper esophagus. A second attempt was equally unsuccessful. After instillation of a radiopaque contrast agent via the nasogastric tube, a second chest radiograph demonstrated a "markedly dilated proximal esophagus with stoppage of the contrast agent at the level of the tracheal bifurcation" (Figure 1). Additionally, a "small amount of contrast agent in the gastric fundus and little air filling of the stomach and proximal intestine" were reported. The diagnosis of an esophageal atresia with distal tracheoesophageal fistula (type Vogt IIIb) was made. When the respiratory function deteriorated again on the second day of life, the infant was transferred to our tertiary pediatric referral center for surgical correction of the esophageal atresia.

After induction of general anesthesia with sevoflurane, sufentanil, and neuromuscular blocking agent in the intubated neonate, a straight rigid endoscope was inserted into the oral cavity. A lesion immediately above the upper esophageal sphincter was noted during the laryngoscopy and before the actual tracheoscopy (Figure 2A). Further examination revealed a hypopharyngeal perforation of about 1.5 cm in length (Figure 2B; Supplemental Digital Content, Video 1, http://links.lww.com/AACR/A416), which had been mistaken for esophageal atresia on chest radiograph examinations. The subsequent tracheoscopy and an esophagogastroscopy were unremarkable and confirmed normal anatomy of the upper gastrointestinal tract and trachea (Supplemental Digital Content, Videos 2 and 3, http://links.lww.com/AACR/A417, http://links.lww.com/AACR/A418). After placement of a 6 charrière nasogastric tube under endoscopic guidance, the child's further hospital course was uneventful with spontaneous healing of the perforation with broad-spectrum antibiotic prophylaxis with vancomycin and meropenem.

VIDEO+

VIDEO+

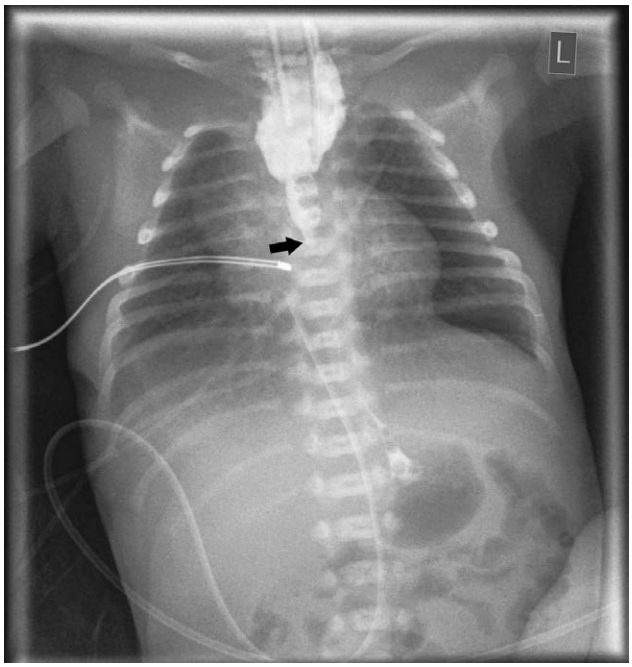

Figure 1. Chest radiograph showing the tip of a nasogastric tube (arrow) at the level of the tracheal bifurcation. A contrast agent was installed via this tube.

In summary, we present a case report of a hypopharyngeal perforation mimicking an esophageal atresia in an extremely low birth weight infant, emphasizing the necessity of endoscopic investigation before surgical repair. In addition to advancing knowledge beyond prior reports, we provide excellent images and videos of these procedures.

DISCUSSION

The preoperative verification of the diagnosis of an esophageal atresia is critical. Primarily, a thoughtful evaluation of the information provided about history of delivery, resuscitation, nasogastric tube placement, and radiographic examination is essential. Difficulties with endotracheal intubation and placement of a gastric tube can cause injury that must be considered. The surgical repair of an esophageal atresia is a challenging procedure with a risk of severe complications, especially in infants with extremely low birth weight[1] and unnecessary thoracotomies must be avoided.

However, achieving diagnostic certainty without the use of advanced techniques, such as endoscopy is difficult. The initial radiographic findings (Figure 1) at the referring hospital were not typical for esophageal atresia type Vogt IIIb for the following reasons: (1) the upper esophageal stump is usually a smoothly bounded pouch, instead of a structure with irregular edges and (2) passage of contrast agent into the lower esophagus and stomach requires passage through the tracheobronchial system, which was not visible on the chest radiograph. It is important that the preoperative chest radiograph be thoroughly evaluated.

Although repeated inability to pass a nasogastric tube into the stomach is commonly accepted to confirm the diagnosis, there are several etiologies that impede correct nasogastric tube placement and may subsequently mimic an esophageal atresia. These "false positives" include esophageal foreign body impaction,[6] pharyngeal pouching after a traumatic birth,[7] and pharyngeal or esophageal perforations due to traumatic attempts of endotracheal intubation[8] or nasogastric tube placement.[2]

The projection of a nasogastric tube in radiographic examinations may be unreliable to either confirm or rule out esophageal atresia. Apart from false-positive diagnoses, there are also false-negative radiographic findings, which usually

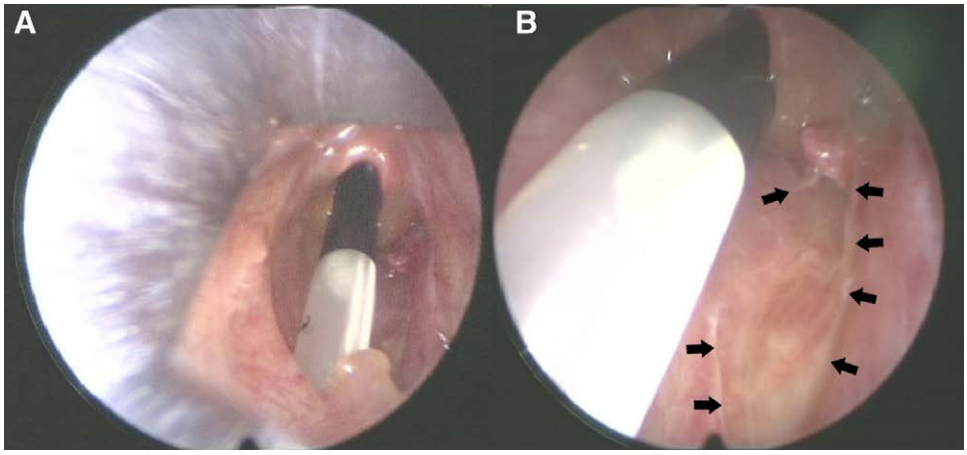

Figure 2. View of the larynx and pharynx during preoperative direct laryngoscopy with a rigid bronchoscope. The arrows mark the edge of the perforation.

result from a nasogastric tube passed through a false passage. Reported cases include the perforation of an upper esophageal pouch with the nasogastric tube and passage of the tube through the mediastinum into the abdomen.[9] Alternatively, it may pass through the larynx or an upper tracheoesophageal fistula into the trachea and exit through a lower tracheoesophageal fistula into the lower esophagus.[10–12]

Despite the diagnostic uncertainty associated with esophageal atresia, there is no consensus on whether endoscopic examinations should be mandatory before surgical interventions.[3–5] A review of 4 Australian hospitals revealed that only 40% of children who underwent a surgical repair of an esophageal atresia had a bronchoscopy performed before the operation. There was marked heterogeneity among the participating centers with regard to the rate of preoperative endoscopic examinations (4%–90%).[13] A recent European consensus statement recommends tracheobronchoscopy, but not to exclude false diagnoses, but to evaluate tracheolaryngeal pathologies.[14] All the cited recommendations for a tracheoscopy primarily address the need to detect pathologies, the localization of the fistula, and possible presence of multiple fistulae[15] but do not recommend esophagoscopy before surgery. This might be attributable to the fact, that in most cases of esophagus atresia, a fistula is present. This means that in most cases, the presentation of a fistula confirms the diagnosis. As an addition to the existing recommendations, we therefore strongly suggest that an additional esophagoscopy must be performed if there is no fistula proven by tracheoscopy. This could either demonstrate a continuity of the esophagus if no malformation is present or an occluded upper pouch with an isolated esophageal atresia (Vogt type 2).

An endoscopic examination including tracheoscopy, and esophagoscopy if no fistula is present, immediately before the surgical intervention is part of the routine diagnostic workup of every case of esophageal atresia in our pediatric hospital. Because the child was transferred to the operating room while endotracheally intubated, it otherwise would have been likely that no laryngoscopy was performed before the start of surgery. Hypopharyngeal perforation and other pathologies can be visualized much better with endoscopic optics than with the naked eye.

Tracheoscopy and esophagoscopy are quick, safe, and easily performed procedures for properly equipped and trained personnel. Our case report emphasizes the benefit of presurgical endoscopic investigations in children suspected of an esophageal atresia. The image quality of those investigations permits a reliable diagnosis, even in an extremely low birth weight infant. The endoscopy should be performed at the same time as the planned repair to limit the children's exposure to multiple anesthetics.

We expect that this report contributes to recommendations for wider implementation of mandatory endoscopic investigations before surgical interventions for esophageal atresia. As an addition to the existing recommendations, we strongly suggest that an additional esophagoscopy must be performed if there is no fistula proven by tracheoscopy. Apart from improving diagnostic certainty before a risky procedure, tracheoscopy may also help with optimal tracheal tube positioning and enable placement of tubes or balloons into the tracheoesophageal fistula.[4]

The mandatory inclusion of preoperative endoscopic investigations as part of procedural standard of care for the surgical repair of an esophageal atresia is a critical step to improve patient safety.

DISCLOSURES

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Contribution: This author participated in the treatment of the patient, contributed to the search of the relevant literature, helped in writing of the abstract, introduction, case description and discussion, and revised the final manuscript, and approved the final manuscript as submitted and agree to be accountable for all aspects of the work.
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