Unusual cause of pneumoperitoneum in a spontaneously breathing 1-day-old term infant
This female infant was born to a healthy 37-year-old G3/P3 by spontaneous vaginal delivery at 40 5/7 weeks of gestation out of an incomplete footling breech presentation at a birth center. Pregnancy had been unremarkable with the exception of moderate polyhydramnios noted in the third trimester. Her birth weight was 3440 g (P 25-50), her head circumference was 36 cm (P 75) and her length was 52 cm (P 50). Postnatal adaptation was normal with Apgar scores of 5, 9, and 10 at 1, 5, and 10 minutes, respectively (umbilical cord-pH values were not measured). The family history was remarkable for esophageal atresia (EA) in the infant’s father.

At 12 hours of life, there was reportedly some yellowish emesis (felt to be possibly bilious) and a nasogastric tube (NG) could not be advanced into the stomach. The abdomen, however, was soft and non-distended. Six hours later she was examined by a pediatrician who noted a grossly distended abdomen and because of the reported bilious emesis referred the infant to our center with suspected intestinal atresia.

On admission, her heart rate was 150 beats per minute, her blood pressure 68/34 (mean 40) mmHg, and her oxygen saturation while breathing room air was 97%. At this point, she had mild respiratory distress with tachypnoea (72 breaths per minute) and intercostal retractions. Her abdomen was markedly disten-
ded and no bowel sounds were heard. Blood gas analysis, electrolytes, lactate and WBC were all within normal limits. A first babygram revealed an impressive pneumoperitoneum (Fig. 1) and a distended stomach and distended loops of bowel. In preparation for surgery, umbilical catheters were placed. An NG tube could not be advanced more than 8 cm and projected over the upper third of the esophagus (Fig. 2).

Esophageal atresia (EA) with tracheo-esophageal fistula (TEF) and intestinal perforation was felt to be the likely diagnosis. In the OR, the pneumoperitoneum was drained under local anesthesia and nasotracheal intubation was accomplished without difficulty. Bronchoscopy revealed a TEF approximately 1.5 cm above the carina. The endotracheal tube was advanced beyond the TEF to minimize air entry into the distal esophagus. At laparotomy, there was an area of the posterior gastric wall which was very thin with multiple perforations. Following partial resection of the affected area, a gastrostomy tube was placed to allow continuous drainage of air that might enter the stomach through the TEF during mechanical ventilatory support. Postoperative chest X-ray showed near complete evacuation of the pneumoperitoneum. Interestingly, resolution of the pneumoperitoneum lead to a dramatic change in umbilical catheter position (Fig. 3), highlighting the importance of repeating X-ray examinations after significant interventions.
Babygram on admission to the NICU at the age of 18 hours: massive pneumoperitoneum, gastric distension and dilated loops of bowel.
Babygram following insertion of umbilical catheters and attempted placement of an NG tube: the UVC projects over the 11th vertebral body, the UAC is at the 7th intercostal space, and the NG tube cannot be advanced beyond 8 cm.
Postoperative chest X-ray following repair of the gastric perforations and gastrostomy tube placement: the pneumoperitoneum has resolved; note the changed umbilical catheter positions (UVC now displaced into the right hepatic vein, UAC now projecting over 4th intercostal space).
Three days later, the TEF was divided and an anastomosis of the esophagus was performed without difficulties. The girl was extubated 2 days later. A contrast study on the 7th postoperative day revealed no leakage. Unfortunately, early stricture formation occurred and the patient underwent four dilatation procedures before being discharged at the age of one month with the gastrostomy still in place to allow for partial tube feeding. Three additional dilatations of the anastomosis were performed in the outpatient setting. The gastrostomy button was removed at the age of two months and, at four months of age, esophagoscopy and contrast studies revealed a widely patent esophagus, and the patient was taking full feeds and thriving.
Postoperative chest X-ray following division of the TEF and anastomosis of the esophagus.
The incidence of EA with/without TEF ranges between 1:3000 and 1:4000 live births. EA with distal TEF is the most common form and accounts for 86% of all EA cases (Fig. 5). EA is often associated with other congenital anomalies, most commonly cardiac abnormalities such as ventricular septal defect, patent ductus arteriosus or tetralogy of Fallot, but also more complex malformations (e.g., VATER, VACTRL associations).

Gastric perforation is a potentially lethal complication of EA with distal TEF (1, 2). One institution reported a perforation rate of approximately 1% (6 of 623 patients with EA and distal TEF) (2). The complication is most likely to occur in preterm infants that require mechanical ventilation for hyaline membrane disease or some other lung pathology. When lung compliance is poor, the TEF may act as a low-resistance vent through which air escapes from the trachea into the esophagus leading to potentially dangerous gastric distension.

Various methods of preventing this complication have been described: early surgical repair within 12 hours of birth (3), high frequency ventilation (4), water-seal gastrostomy (5), bronchoscopic placement of a Fogarty balloon catheter (6), distal placement of an ETT beyond the origin of the fistula (7), silastic banding of the lower esophagus (8), gastric division (9), and retrograde placement of a balloon catheter into the lower esophagus (10).
Our case is remarkable for several reasons. First, there was considerable delay in establishing the correct diagnosis. Given the history of polyhydramnios, episodes of “emesis” coupled with the inability to place an NG tube should have lead to prompt referral to a NICU. Second, the fact that there was a history of EA in the father of the patient is noteworthy. Although most cases of EA occur sporadically, familial cases have been reported in the literature (11). Third and most interestingly, gastric perforation occurred in our term infant in the absence of lung disease, respiratory distress and mechanical ventilation. Until very recently, to the best of our knowledge, this had not been reported in the literature. However, in 2011, Rathod et al. reported a case series from Chandigarh, India. They described 6 spontaneously breathing term infants with EA and TEF (out of a total of 856 patients with this anomaly treated between 2004 and 2010 at their center) who presented with gastric perforations. Their report appears to be the first description of this complication in non-ventilated infants with EA and distal TEF (12).
Fig. 5

Relative frequencies of occurrence of the various types of esophageal atresia (EA) with and without tracheoesophageal fistula (TEF) (Gross classification (1953), from medscape).

<table>
<thead>
<tr>
<th>Type</th>
<th>Frequency</th>
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<tbody>
<tr>
<td>Type A</td>
<td>4%</td>
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<tr>
<td>Type B</td>
<td>8%</td>
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<tr>
<td>Type C</td>
<td>86%</td>
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<td>Type D</td>
<td>1%</td>
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<tr>
<td>Type E</td>
<td>1%</td>
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1. Jones TB, Kirchner SG, Lee FA, Heller RM. Stomach rupture associated with esophageal atresia, tracheoesophageal fistula, and ventilatory assistance. AJR 1980;134:675-677


