

A 3-day-old Term Newborn With an Unusual Cause of Weight Loss

Jorge M Lopez Da Re, MD

Abstract:

Esophageal atresia (EA) associated with Tracheoesophageal fistula (TEA) is a rare congenital anomaly caused by abnormal septation of the foregut into the esophagus and trachea. Although almost all Newborns with EA/TEF present symptoms immediately after birth, with excessive secretions (drooling, choking, and resp distress), inability to feed, gastric distention, and reflux with aspiration pneumonia, some can present later (Type E- TEF). This case describes a 3-day-old term newborn with an unusual cause of weight loss (>10%) caused by esophageal atresia (EA) associated with trachea esophageal fistula (TEF). The newborn had a normal exam, vital signs, glucose, and respiratory symptoms on admission. The presumptive diagnosis was based on an x-ray with an OG located on the upper inlet of the anterior-posterior chest radiograph. The confirmatory diagnosis was made by a water-soluble contrast study, using 5 ml of contrast through a Replogle tube. The study showed the esophageal pouch without aspiration, penetration, or fistula. VACTERL association screening was negative. We hypothesize that the absence of symptoms was related to exclusively breastfeeding with a low milk supply.

“A 3-day-old term female newborn was admitted to the inpatient pediatric floor due to hyperbilirubinemia requiring phototherapy and weight loss above 10%. Newborn history: Infant born at 39 weeks gestation to an otherwise healthy 27-year-old gravida 3, para 1 woman via vaginal delivery.”

Case Report:

A 3-day-old term female newborn was admitted to the inpatient pediatric floor due to hyperbilirubinemia requiring phototherapy and weight loss above 10%. Newborn history: Infant born at 39 weeks gestation to an otherwise healthy 27-year-old gravida 3, para 1

woman via vaginal delivery. Maternal laboratory results: O positive, Coombs negative, COVID-19 positive, rest negative. Maternal history: unremarkable and negative for diabetes, hypertension, and polyhydramnios. The mother denied taking drugs during the pregnancy. Routine fetal ultrasound during the second trimester was normal. She received routine resuscitation at birth and Apgar scores: of 7 and 9 at 1 and 5 minutes, respectively. Growth parameters were: weight of 3270 grams at the 53rd percentile; height of 52 cm at the 85th percentile. Head circumference was 35 cm at the 73rd percentile. Newborn blood type was B+, Coombs +. Newborn laboratories were normal. The infant was sent to the nursery for routine care. After two days of life, she went home with a follow-up appointment with a pediatrician within 24 hours after discharge. At home, she was exclusively breastfeeding, voiding, and stooling normally.

“Newborn laboratories were normal. The infant was sent to the nursery for routine care. After two days of life, she went home with a follow-up appointment with a pediatrician within 24 hours after discharge. At home, she was exclusively breastfeeding, voiding, and stooling normally.”

On admission, the newborn physical exam was normal except for weight loss above 10 % from birth and jaundice of the face and trunk. There were no signs or symptoms of dehydration. Vital signs: heart rate 154 bpm, respiratory rate 40 bpm, blood pressure 80/45 mmHg, MAP 57 mmHg, weight 2,828 grams (14% below birth weight), and glucose 83 mg/dl. She continued to breastfeed ad libitum. Laboratory: CBC: 9.8×10^3 /microliter, hemoglobin 10.9 g/dL, Hematocrit 32 %, platelets 344×10^3 /microL normal differential. Bilirubin 16.3 mg/dl at 71 hours of life (high-risk zone), meeting criteria for phototherapy. The basic metabolic profile was unremarkable except for mild hypernatremia (Sodium 149 mmol/L, mild hypokalemia Potassium 3.3 mmol/L). Blood gas was normal. The provider recommended formula supplementation due to dehydration, hyperbilirubinemia, and weight loss. On the first attempt of formula feeding, the newborn presented

NEONATOLOGY TODAY is interested in publishing manuscripts from Neonatologists, Fellows, NNPs and those involved in caring for neonates on case studies, research results, hospital news, meeting announcements, and other pertinent topics.

Please submit your manuscript to: LomaLindaPublishingCompany@gmail.com

with projectile vomiting (mouth and nose) associated with cyanosis and bradycardia. The infant required positive pressure ventilation with a good response requiring noninvasive respiratory support nC-PAP. Initial chest and abdomen x-ray (Fig. 1) showed clear lungs without acute infiltrate and dilated stomach with a normal bowel pattern. She was transferred to a warmed isolate with orders of n.p.o (nothing by mouth), OG tube placement, IV fluids, sepsis screening, ampicillin, and gentamycin. An attempt to pass an OG tube was unsuccessful. Follow-up x-ray after OG placement showed OG is at the proximal esophageal pouch, suggesting esophageal atresia (arrow); a dilated stomach, with air and fluid-filled, indicating a suspected distal tracheoesophageal fistula (figure 2).



Figure 1. Chest and abdomen x-ray one view—findings: Clear lungs with normal, no opacity, effusion, or pneumothorax. No acute bony abnormality was visualized. The cardio mediastinal silhouette is negative.

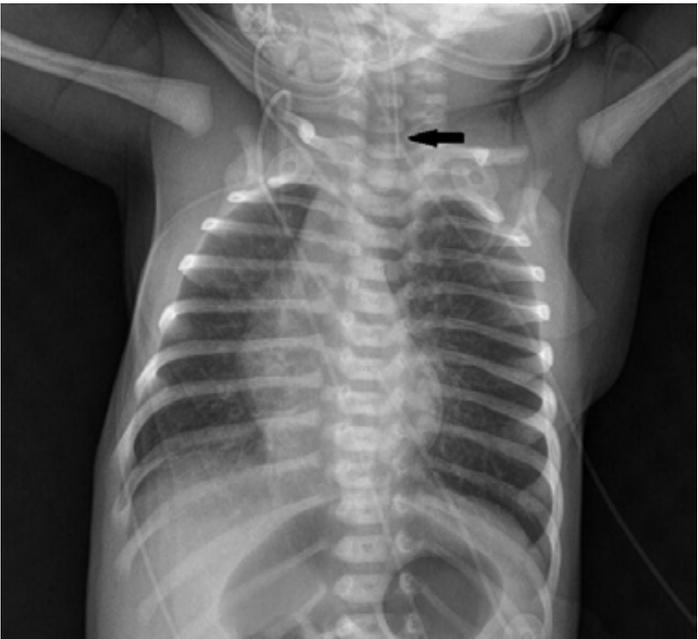


Figure 2. Chest and abdomen x-ray one view. Findings: OG is at the proximal esophageal pouch, suggesting esophageal atresia (arrow); the abdomen is distended, with air and fluid-filled, indicating a distal tracheoesophageal fistula.

Patient Course:

The newborn was transferred to a Level III unit. Type I EA/TEF was confirmed by a water-soluble contrast study using 5 ml of contrast through a Replogle tube (Fig. 3 A, B, and C). This study showed the esophageal pouch without aspiration, penetration, or fistula. The infant underwent surgery with dissection of the upper pouch and anastomosis with the lower esophagus and close of the fistula. The TEF was associated with mild to moderate tracheomalacia. The surgery was uncomplicated; the newborn was extubated and reached ad-lib feeding on days 3 and 6 post-operatory, respectively. VATERED association screening was negative, and echocardiography, vertebral studies, and renal ultrasound findings were normal.

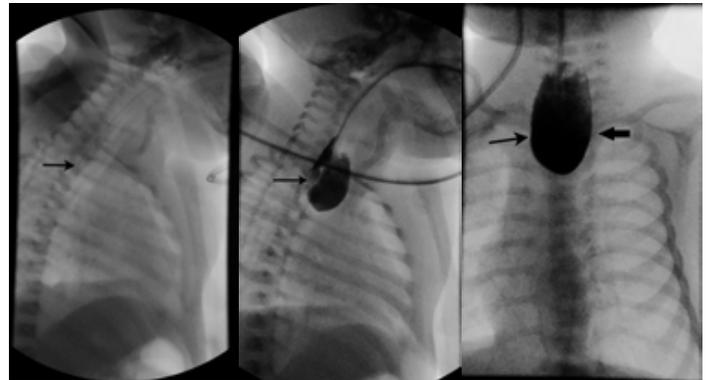


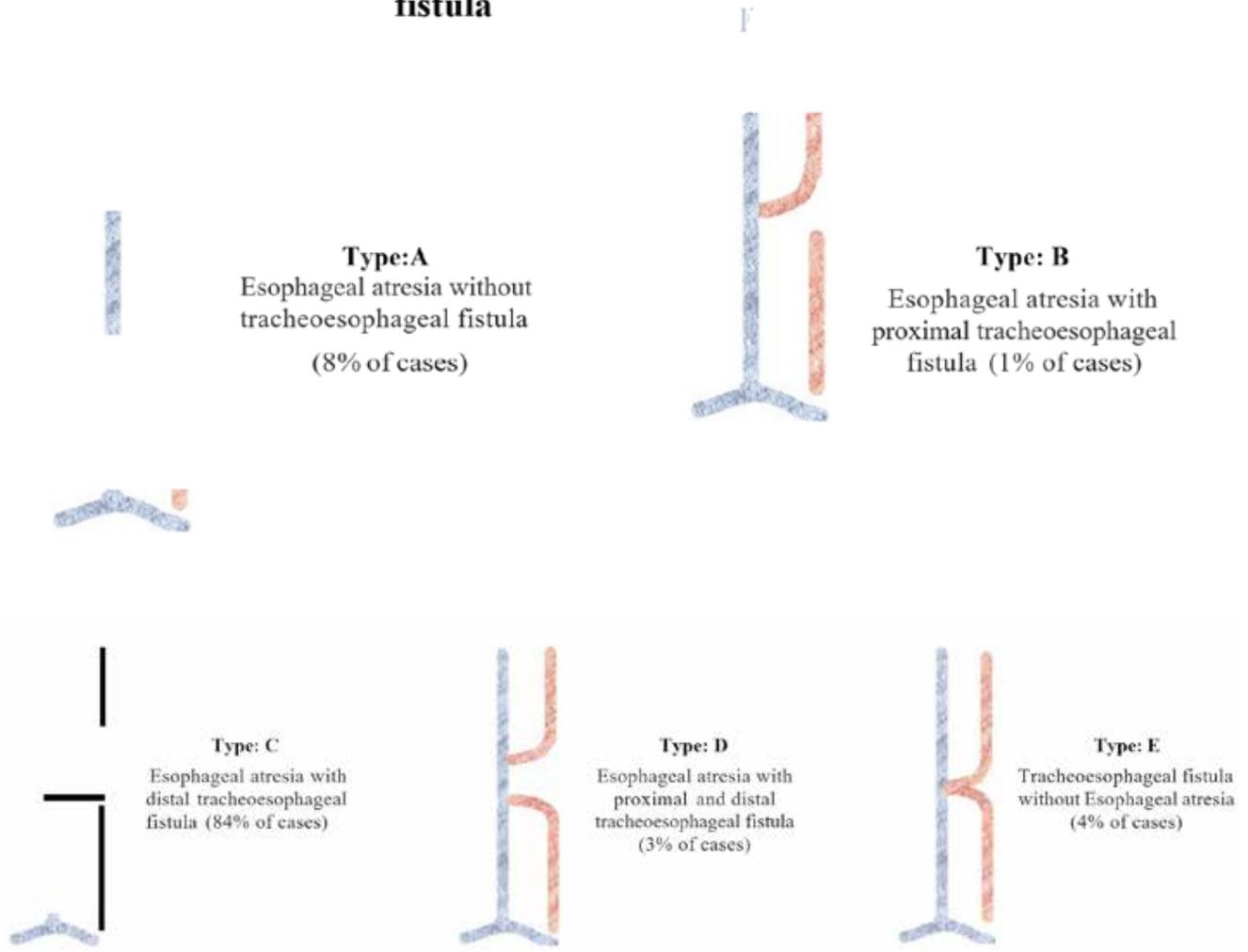
Figure 3 Upper GI study. Contrast x-ray. Fig. 3 A (Lateral no contrast), Fig. 3 B (lateral Contrast), Fig. 3 C (postero-anterior contrast). Findings: The proximal esophagus distends with contrast demonstrating a blind-ending pouch terminating about 3.5 cm thoracic vertebral body weights above the carina—impression: Proximal thoracic esophageal atresia. Given the bowel gas, the finding suggests type C esophageal atresia with tracheoesophageal fistula.

Discussion:

Esophageal atresia (EA) is the failure of the recanalization of the esophagus during the first eight weeks of development, and tracheal fistula is due to abnormal embryonic lung bud development that fails to undergo branching and remains connected to the esophagus. Esophageal atresia (EA) and tracheoesophageal fistula (TEF) have an incidence of 1 in 3500 to 1 in 4500 live births, which increases with maternal age. In the United States, the prevalence is 2.3 per 10,000 live births (1-2).

“Esophageal atresia (EA) is the failure of the recanalization of the esophagus during the first eight weeks of development, and tracheal fistula is due to abnormal embryonic lung bud development that fails to undergo branching and remains connected to the esophagus.”

Types of Esophageal Atresia/ tracheoesophageal fistula



Jorge M Lopez Da Ra, MD

Tracheoesophageal fistula types were classified according to the scheme developed by EC Vogt[1] in 1929, as modified by Gross in 1953.

Copyrights apply

Figure 4: Tracheoesophageal fistula types classified according to the scheme developed by EC Vogt[1] in 1929, as modified by Gross[2] in 1953.

Differential diagnosis:

The main differential diagnosis of projectile vomiting includes esophageal atresia, choanal atresia, tracheoesophageal fistula, esophageal stenosis, laryngo-tracheo-esophageal cleft, esophageal webs, esophageal rings, esophageal strictures, tubular esophageal duplications, congenital short esophagus, and pyloric stenosis.

Clinical presentation:

The most common prenatal finding is current or worsening polyhydramnios. At birth, the clinical presentation depends on the presence of esophageal anomaly "atresia (A, B, C, D) Vs. no atresia (E) Figure # 4" and/or the type of tracheoesophageal fistula "A, B no fistula CDE fistula" (3).

Newborns with EA/TEF are symptomatic in more than 95% at birth, and this presentation is independent of the type of tracheoesophageal fistula. The most common type of EA/TEF is type C (85%), with EA with distal TE fistula.

At birth, EA symptoms include excessive secretions that cause drooling, choking, respiratory distress, inability to feed, and, if TEF is present gastric distention. The only type of TEF that does not usually present at birth is the type E or H-type EA/TEFs, symptoms commonly seen after the newborn period due to delayed enteral feeding and no early placement of an NG/OG tube (4). These include a history of mild respiratory distress associated with feeding or recurrent episodes of pneumonia.

Diagnostic imaging:

Prenatal diagnosis of EA/TEF by ultrasound (US) is possible in the late trimester (mid-trimester 4.3% vs. late trimester 33%) (5). The most common signs in the last trimester by fetal ultrasound used to determine the need for further screening are small/absent stomach, esophageal pouch, and current or worsening polyhydramnios.

“(A, B, C, D) can be made by attempting to pass a catheter into the stomach, that usually does not advance beyond 10 cm (average of 10 to 15 cm) and remain high and sometimes curled on the Anterior-posterior chest radiograph in the upper esophageal pouch. It is crucial to remember to avoid forcing the advance of the NG/OG”

Postnatally, the initial presumptive diagnosis in most of the EA/TEF (A, B, C, D) can be made by attempting to pass a catheter into the stomach, that usually does not advance beyond 10 cm (average of 10 to 15 cm) and remain high and sometimes curled on the Anterior-posterior chest radiograph in the upper esophageal pouch. It is crucial to remember to avoid forcing the advance of the NG/OG/Repleg to prevent perforation of the upper esophagus pouch. Anterior-posterior and lateral views X-rays may aid in making the diagnosis of TEF. These will show a coiled catheter in the upper chest or esophagus and a gas-filled stomach. An air contrast or water-soluble contrast study confirms the diagnosis. A barium contrast study is contraindicated due risk of aspiration pneumonia.

“TEF/EA is often associated with CHARGE syndrome or VACTERL association. The management should include echocardiography, renal ultrasonography, and, as clinically indicated, contrast enema and limb radiographs.”

Type E or H-type EA/TEF diagnosis is challenging due to unspecific or late symptoms. A contrasted swallow study is the diagnosis of choice in this type. The contrast will help to visualize the fistula and passage of the contrast to the lungs. Other diagnosis methods include water-soluble contract material with two-step contrast and lower contrast, followed by the upper esophagus looking for the fistula.

CT scan is an additional study but not commonly done due to the high radiation and high sensitivity and specificity of the upper GI contrast studies.

TEF/EA is often associated with CHARGE syndrome or VACTERL association. The management should include echocardiography, renal ultrasonography, and, as clinically indicated, contrast enema and limb radiographs (6).

This case describes a 3-day-old term newborn with an unusual delay presentation of Type C EA/TEF. Per the mother's report, we attribute the delayed onset of symptoms to exclusive frequent breastfeeding with low milk supply, mostly colostrum, with a volume of less than 5 ml. Symptoms were evident only after higher volume formula supplementation.

We cannot explain how the glucose levels remained normal. A small, not visualized fistula between the upper and lower pouch can be an explanation.

References:

1. Cassina M, Ruol M, Pertile R, et al. Prevalence, characteristics, and survival of children with esophageal atresia: A 32-year population-based study including 1,417,724 consecutive newborns. *Birth Defects Res A* 118 Clin Mol Teratol 2016; 106:542.
2. Depaepe A, Dolk H, Lechat MF. The epidemiology of tracheo-oesophageal fistula and esophageal atresia in Europe. EURO-CAT Working Group. *Arch Dis Child* 1993; 68:743. [Abstract]
3. Clemens BS. Congenital malformations of the lung and airway. *Pediatric Respiratory Medicine*, 1st ed, Taussing LM, Landau (Eds), Mosby, St Luis 1999. Vol 1, p2 1115.
4. Corne De Vos, Charlie Kohler, Natash Fourie, et al. Delayed presentation of a baby with an esophageal atresia on day 14 of life *BMJ Case Rep* 2021;14.
5. E Kassif, T Weissbach, A Kushnir, et al. I. Esophageal atresia and tracheoesophageal fistula: a prenatal sonographic manifestation from early to late pregnancy. *Ultrasound Obstet Gynecol*. 2021 Jul;58(1):92-98. 125 doi: 10.1002/uog.22050.
6. Shaw-Smith C. Esophageal atresia, tracheo-oesophageal fistula, and the VACTERL association: Review of genetics and epidemiology. *J Med Genet* 2006; 43:545.

Disclosures: There are no reported disclosures

NT

Corresponding Author:



Jorge M Lopez Da Re, MD
Neonatologist
Adventhealth Medical Group, Orlando Florida.
Department of Pediatrics, Neonatology Division
1500 SW 1st Ave, Ocala, FL 34471
Email Jorge.lopezdare.md@adventhealth.com
Phone: 352-402-5090