A 4-hour-old term male infant has persistent drooling. The infant attempted to bottle-feed and immediately choked and spit. The pregnancy was complicated by polyhydramnios. A tracheoesophageal abnormality is suspected.

Of the following, the MOST likely tracheoesophageal abnormality is:

1. esophageal atresia with a distal and a proximal tracheoesophageal fistula
2. esophageal atresia with a distal tracheoesophageal fistula
3. esophageal atresia with a proximal tracheoesophageal fistula
4. esophageal atresia without a tracheoesophageal fistula
5. tracheoesophageal fistula without esophageal atresia

You selected 3, the correct answer is 2.

Congenital esophageal atresia (EA) with or without a tracheoesophageal fistula (TEF) is a common congenital anomaly with an incidence of 1 in 3,000 live births. Newborns with EA may present in the delivery room with either a sonorous "seal-bark" cry because of associated tracheomalacia or within the first few hours after birth with excessive oral secretions. Feeding an infant with EA will cause spitting and choking, and aspiration pneumonia can occur. Reflux of gastric secretions through a distal TEF also can cause aspiration pneumonia. Diagnosis of EA is suspected by failure to pass an orogastric tube beyond 10 cm to 11 cm from a term infant's lips. Chest radiography confirms the position of the orogastric tube in the proximal esophageal pouch.

From 30% to 60% of infants with EA and TEF have associated anomalies, including cardiac (25%), genitourinary (15%), skeletal (14%), and intestinal atresias (13%). The VACTERL association (vertebral defects, anorectal abnormalities, cardiac defects, TEF, renal abnormalities, limb defects) occurs in approximately 10% to 25% of cases.

Embryologic development of the trachea and esophagus is a complex process. During week four of gestation, the embryo is C-shaped, and the primitive (primordial) gut is divided into the foregut, midgut, and hindgut. The trachea and esophagus are formed from the foregut. The trachea develops from the laryngotracheal tube, which buds off the ventral surface of the foregut. The tracheoesophageal septum separates the foregut into tracheal and esophageal tubes. The esophagus rapidly elongates with growth of the embryo. The lumen of the esophagus becomes obliterated by the proliferation of endodermal lining cells. During week eight of gestation, endodermal cell death re-establishes the esophageal lumen. Failure of the tracheoesophageal septum to divide into the esophagus and trachea at week four of gestation, or failure of recanalization of the esophagus during week eight of gestation results in various types of EA and TEF.

Polyhydramnios may develop because the fetus with EA cannot swallow amniotic fluid. Significant polyhydramnios may lead to premature delivery in approximately 30% of cases. Because the fetus may derive some nutritional benefit from swallowed amniotic fluid, newborns with EA may be small for gestational age.

The most common tracheoesophageal abnormality (86%) is EA with a distal TEF (Fig. 1). The proximal esophagus ends blindly in the superior mediastinum at the third or fourth thoracic vertebra. The distal esophagus usually enters the posterior wall of the trachea 1 cm to 2 cm above the carina. The proximal esophageal pouch and the distal TEF may overlap or be
because the distal TEF allows some amniotic fluid to flow from the trachea to the gastrointestinal tract, polyhydramnios only occurs in approximately 33% of pregnancies with this type of EA.

EA with distal and proximal TEF, also known as a double TEF, is a rare (<1%) tracheoesophageal abnormality (Fig. 2). This type of malformation may be misdiagnosed as the more common EA with a distal TEF. If the small proximal TEF is unrecognized, then recurrent respiratory infections will occur. Preoperative endoscopy permits recognition of the double fistula and complete repair at the initial operation.

EA with a proximal TEF is another rare (2%) tracheoesophageal abnormality (Fig. 3). The TEF usually is located 1 cm to 2 cm above the distal end of the esophageal pouch. Polyhydramnios occurs nearly 100% of the time because no distal fistula is present.

Isolated EA without a TEF (Fig. 4), occurs in 7% of tracheoesophageal abnormalities. The proximal esophageal segment usually ends in the posterior mediastinum near the second thoracic vertebra. Unlike EA with distal TEF, infants without a distal TEF have a flat, gasless abdomen. A wide gap usually divides the upper and lower esophageal segments, making primary anastomosis difficult. Isolated EA without a TEF may be the result of failure of recanalization of the esophagus during week eight of gestation.

TEF without EA, also known as H-type TEF (Fig. 5), comprises 4% of tracheoesophageal abnormalities. Infants with H-type TEF may have intermittent choking episodes in the newborn period. More commonly, patients with H-type TEF present later in life, even into adulthood, with chronic cough, recurrent pneumonia, or reactive airway disease. This form of TEF is the most difficult to diagnose because the fistula may not be identified by routine contrast swallow studies. Esophagoscopy or bronchoscopy may be necessary to visualize the TEF.

References:


Content Specifications:
Know the morphogenesis of the gastrointestinal (GI) tract and factors that lead to congenital malformations
Know how to recognize and evaluate an infant with excessive gastric contents and hydramnios
Know how to diagnose polyhydramnios, its significance, and the management of pregnancy when polyhydramnios is diagnosed

Plan appropriate management for an infant with airway obstruction, such as vascular rings, choanal atresia, and tracheal abnormalities

Recognize the clinical features of VATER association
In the delivery room, you begin resuscitation of a term female infant who has apnea, bradycardia, and hypotonia. No meconium was present in the amniotic fluid. You have positioned, dried, and suctioned the nose and mouth as well as provided tactile stimulation. However, her heart rate is 50 beats per minute. You begin positive pressure ventilation (PPV) with a bag and mask.

Of the following, the MOST important clinical indicator of adequate ventilation is

- chest rise
- color
- heart rate
- muscle tone
- skin perfusion

You selected 2, the correct answer is 3.

The most important step in resuscitation of the depressed, newly born infant (heart rate <100 beats per minute, apnea or gasping respiration, hypotonia) is ventilation of the lungs. The most important response to positive pressure ventilation (PPV) is an immediate rise in heart rate. The infant in the vignette is expected to have a rapid rise in heart rate after ventilation is established. Evidence for interventions during neonatal resuscitation often is limited to comparative animal studies and consensus of opinion. Evidence that supports the heart rate response as the most important clinical indicator of response to PPV is based on animal experiments performed during the early 1960s (Figure). In these cardiorespiratory studies, changes in heart rate, breathing, and blood pressure were recorded. A rapid heart rate increase after initiation of bag and mask ventilation is followed by a gradual blood pressure increase and subsequent spontaneous respiration. Heart rate response as the most important clinical indicator of adequate ventilation is different from the frequently taught concept that chest rise is most important. Avoiding the risks associated with large tidal volume ventilation (pneumothorax and bronchopulmonary dysplasia) is an important goal that favors heart rate increase as the preferred indicator of response to PPV.

Chest rise during PPV is an indication that ventilation of the lungs is occurring. With inadvertent overventilation, the risks of volutrauma and barotrauma causing pneumothorax or initiating bronchopulmonary dysplasia in very preterm infants has caused clinicians to reassess the physiologic responses to, and technique of, ventilation with a resuscitation bag. Therefore, these risks of PPV and data from animal experiments indicate that a rapid heart rate increase, rather than chest rise, is a better indicator of adequate ventilation.

Mucus membrane color is an immediate clinical indicator of oxygenation. It follows that color will change from cyanotic to pink during the first minutes after birth in the healthy, spontaneously breathing newborn. Likewise, during PPV of an infant with bradycardia, apnea, and hypotonia, this transition to pink occurs only after ventilation of the lung with gas and establishment of cardiac output to the pulmonary and systemic circulations, both primary factors in oxygen delivery to tissues. In the neonate, heart rate appears to be more important than stroke volume to increase cardiac output.

Improved muscle tone is a sign that oxygen delivery to the brain has improved. Improved skin perfusion, on the other hand, is a sign that oxygen delivery to other organ systems also has improved. Resolution of hypotonia and improved skin perfusion are expected to follow.
improvements in heart rate, establishment of lung volume and ventilation of the lung in the infant in the vignette.

References:


Dawes GS. *Birth asphyxia, resuscitation and brain damage.* Chicago, IL: Year Book Medical Publisher Inc. 1968:141-159


Heart Rate, Blood Pressure and Breathing after Asphyxia

- **Gasp/min**
  - Primary Apnea
  - Last Gasp
  - Secondary or Terminal Apnea
  - Onset of Gasping

- **Heart Rate**
  - 1

- **Blood Pressure**
  - 2

- **Time From Onset of Asphyxia**
  - Resuscitation