Chapter

20 Esophageal Atresia and Tracheoesophageal Fistula

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A six-day-old male presents with excessive drooling and tachypnea with feeding. When drinking his bottle, he coughs and desaturates to the high 80s. A nasogastric tube was placed in the nursey with postplacement X-ray (Figure 20.1). He was born via spontaneous vaginal delivery at 38 weeks. There is no additional medical or surgical history.

His current vital signs are: blood pressure 60/37 mmHg; heart rate 150/min; respiratory rate 32/min; temperature 37.1°C; SpO_2 100% on room air. Weight is 3.45 kg.

How Is the Diagnosis of Esophageal Atresia Made?

Esophageal atresia (EA) often presents soon after birth with inability to eat. Children with EA often choke and become cyanotic with feeding. Food that is unable to enter the GI tract enters the trachea and lungs. Esophageal atresia may be diagnosed by failure to pass an oro/nasogastric tube (OGT/NGT). X-ray may demonstrate an OGT/NGT coiled in the chest or a blind stomach air pocket. Esophageal atresia may be full or partial and may occur in association with other congenital anomalies such as cardiac anomalies or trachea-esophageal fistulae (TEF). Prenatal ultrasound may show polyhydramnios as the infant is unable to swallow amniotic fluid.

What Is the Most Likely Age for Presentation?

Generally, the diagnosis is made shortly after birth; however, a variant (H-type fistula) exists with a small fistula between the trachea and esophagus which can result in a delayed diagnosis. These children can often present with recurrent aspiration prior to diagnosis of a fistula.

What Is the Incidence of EA?

Esophageal atresia has an incidence of 1 in 3,000–4,000. The incidence of EA/TEF is greater in patients with trisomy 21 and 18.

Idenify the Fistula Variants Presents in Patients with EA/TEF

Five main variants exist for congenital malformations of the esophagus (Figure 20.2).

In What Percentage of Patients with EA/TEF Are Other Anomalies Present?

More than 50% of patients with EA/TEF have associated congenital anomalies.

What Anomalies Are Most Commonly Associated with EA/TEF?

The most common association is that of VACTERL/VATER syndrome. VACTERL/VATER is really an association in which certain congenital anomalies are seen grouped together with a significant degree of frequency.

The following anomalies are known to be associated and therefore require diagnostic testing when a patient presents with EA/TEF.

- **V**: Vertebral
- **A**: Anal defects
- **C**: Cardiac anomalies
- **TE**: Tracheoesophageal fistulae
- **R**: Renal
- **L**: Limb deformities (often radial absence)

Additionally, numerous other associated anomalies may be present (i.e., cleft lip and palate, gastric wall defects)

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What Are the Classic Vertebral Defects in VACTERL Syndrome?

Generally, the defects involve the vertebral bodies which may be absent or incompletely formed. The defects may occur anywhere but are most common in the cervical and lumbar regions. These defects can result in instability of the neck due to incompletely formed vertebral bodies. These children also have an increased incidence of scoliosis as they mature.

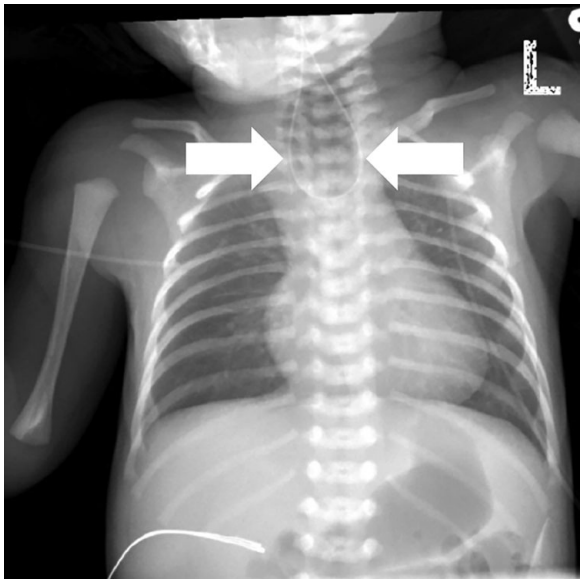

Figure 20.1 Chest X-ray of infant with tracheoesophageal fistula revealing coiling of gastric tube in the upper gastric pouch. Reproduced from: Adler AC, et al. A A *Case Rep* 2017;8(7):172–174. Copyright © 2017 International Anesthesia Research Society

What Are the Classic Anal Defects in VACTERL Syndrome?

Most commonly, these defects include anal atresia or imperforate anus. This necessitates the need for early surgical intervention generally with a multistaged procedure requiring an ostomy, recreation of an anus, and a colonic pull-through to reestablish continuity. The repair of the EA/TEF precedes the anal surgical intervention.

In What Percent of Patients with EA/TEF Is a Cardiac Defect Present?

Approximately one-third of patients with EA/TEF are found to have an associated congenital heart defect.

The most common defects are atrial and ventricular septal defects, patent ductus arteriosus, and Tetralogy of Fallot.

What Are the Classic Renal Defects in VACTERL Syndrome?

Renal defects can include hypoplasia or aplasia of the kidneys and issues related to ureteral valves. If left untreated, these unrelated valve defects can progress to hydronephrosis and/or renal failure.

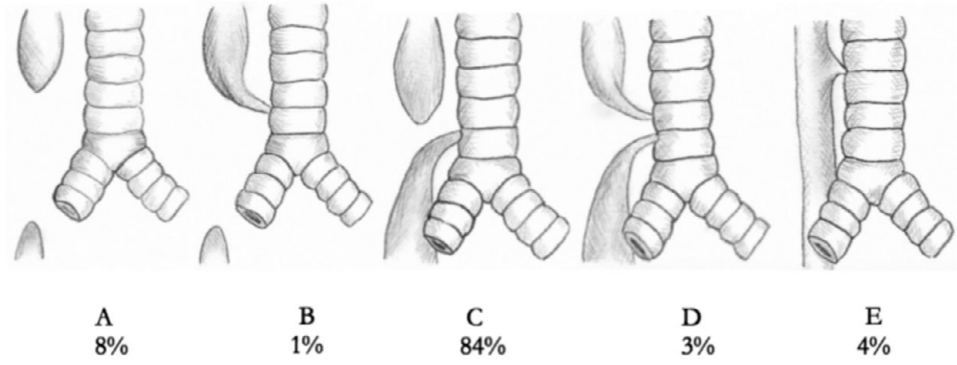

Figure 20.2 Schematic of the types of five main variants of congenital esophageal malformations and their incidences. Courtesy of Adam C. Adler, MD. Drawn by J. Daynes

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What Are the Classic Limb Defects in VACTERL Syndrome?

Digital issues including: hypoplastic thumb, polydactyly, and syndactyly, as well as disorders of the forearm including absent radius. Interestingly, limb and renal defects are often ipsilateral and patients with bilateral limb deformities often have bilateral renal issues.

Is Neurocognitive Function Altered in Patients with VACTERL Syndrome?

Most patients with VACTERL have normal intelligence.

What Is the Expected Mortality in Patients with EA/TEF?

Outcomes after EA/TEF repair have been excellent in recent years with minimal mortality. Patients with cardiac defects, low birth weight (<1,500 grams) and those suffering postoperative respiratory complications have a significantly increased mortality.

Describe the Preoperative Evaluation in Patients with EA/TEF

Aside from the general evaluation of the neonate, patients with suspected EA/TEF should undergo a comprehensive work-up aimed at identifying commonly associated pathologies. In particular, a cardiac anomaly should be excluded as this both contributes to increased mortality and may have significant anesthetic considerations based on the cardiac lesion. A transthoracic echocardiogram should be performed to identify the majority of the pathologies as well as presence of a right aortic arch (2–3%) which has implications for the surgical approach.

Other evaluations include ultrasound of the kidneys for renal anomalies, and radiographs of the spine if concern for vertebral anomalies exist.

Upon diagnosis, a catheter can be placed (with fluoroscopy if needed) in the esophageal blind pouch to remove secretions. These patients are kept NPO with maintenance IV fluid and regular glucose checks.

What Is the Preferred Method of Induction in Patients with EA/TEF?

Classically, the patient is induced with inhalational agents to maintain spontaneous ventilation and avoidance of muscle relaxants prior to fistula ligation. Positive pressure ventilation should be minimized to avoid insufflation of the gastrointestinal tract.

What Is the Benefit of Performing a Bronchoscopy Prior to Intubation?

Many surgeons will perform a flexible or rigid bronchoscopy after induction to allow for confirmation of diagnosis, localization of fistula, and to exclude the presence of multiple fistulae or other pathology.

Describe the Proper Positioning of the Endotracheal Tube (ETT)

The ETT should be positioned (under vision, Figure 20.3) distal to the fistula to avoid gastric insufflation. At times, the fistulous portion may involve the carina, in which case the ETT is generally placed in the main stem bronchus with deliberate one lung ventilation.

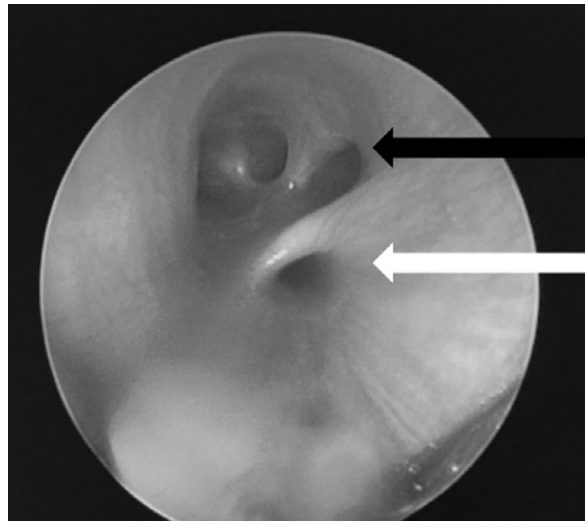

Figure 20.3 Intraoperative rigid bronchoscopic view of EA/TEF showing the fistula arising from the carina (black arrow); also present is a tracheal take-off of the right upper lobe bronchus (pig bronchus) (white arrow). Reproduced from: Adler AC, et al. A A *Case Rep* 2017;8(7):172–174. Copyright © 2017 International Anesthesia Research Society

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What Is the Most Concerning Consequence of Continued Gastric Insufflation?

Continuous gastric insufflation can result in significant gastric/abdominal distension significantly impacting ventilation. Conversely, gastric insufflation, if severe, can result in gastrointestinal rupture and pneumoperitoneum. Treatment relies on early recognition and decompression.

What Is the Intraoperative Approach to EA Repair and TEF Ligation?

The approach is often via an open thoracotomy or a minimally invasive thoracoscopic approach. In cases of two lung ventilation (absent or high fistula) the ipsilateral lung can be manually retracted or is deflated by the pressure of insufflation in cases using a thoracoscopic approach.

The approach to this procedure is dictated by the type of fistula. With most being type C (blind, proximal esophageal pouch with distal tracheal/esophageal fistula), the repair includes ligation of the fistula and anastomosis of the esophageal pouches. In some cases, the distance between the proximal and distal esophageal pouches is too long to perform a primary anastomosis without placing the esophagus under tension.

Aside from standard ASA monitoring, placement of an arterial line may be considered in certain patients. IV access should be sufficient to allow for for rapid administration of blood products if significant bleeding is encountered.

What Is the Postoperative Approach to EA Repair and TEF Ligation?

This is strongly dictated by the type of fistula and presence of associated anomalies (especially cardiac anomalies). Depending on the type of TEF and difficulty of the esophageal repair, the patient may be extubated. In general, low birth weight babies, premature infants, co-existing cardiac anomalies, and those with a "tight" anastomosis should remain intubated.

What Head/Neck Maneuver Should Be Avoided in Patients Post-EA Anastomosis?

In patients following esophageal anastomosis and especially one under some degree of tension, neck extension should be avoided as this can exacerbate the degree of tension on the suture line.

What Regional Anesthetics Can Be Offered to These Patients?

Post thoracotomy patients may benefit from either a thoracic epidural or a caudally placed thoracic epidural. Paravertebral blocks (and catheters) and the newer erector spinae block can be performed as well. Intercostal blocks are usually avoided due to their high absorption, short duration of action, and concern for local anesthetic toxicity.

Suggested Reading

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