

ANESTHETIC CONSIDERATIONS FOR THE NEONATE WITH TRACHEOESOPHAGEAL FISTULA

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Abstract

Tracheoesophageal fistula (TEF) and associated esophageal atresia (EA) in the neonate, presents during the first week of life. This congenital defect can be complicated by aspiration, respiratory distress, and other congenital anomalies. The knowledge and ability of the anesthesiologist to anticipate the challenges in managing neonates presenting for repair, plays an important role in their treatment and survival. Also, it is not uncommon for anesthesiologist to care for a patient later in life following repair of TEF. A familiarity with the immediate complications and long term outcome and sequelae after TEF repair is important to ensure the best patient outcome.

Introduction

Tracheoesophageal fistula manifests in the neonate within hours to days of life. Considered a surgically correctable anomaly of the

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gastrointestinal and respiratory systems, TEF and the perioperative anesthetic considerations are of acute importance to the anesthesiologist. Prior to the first successful staged repair in 1939, esophageal atresia and associated TEF were uniformly fatal. Advancements in pediatric anesthetic techniques and monitoring, neonatology, and pediatric surgery have reduced mortality figures and survival is now higher than 90%¹. Prematurity and severe associated congenital abnormalities continue to be the biggest contributors to mortality associated with TEF².

Several classification systems of EA and TEF have been developed based on the presence of atresia and the relation of the fistula location to the atresia. The Gross classification system describes EA with and without TEF, types A through F³. Another well known classification system describes five types of TEF including types I, II, IIIA, IIIB, and IIIC. Regardless of classification type, the most common form of this anomaly is esophageal atresia with distal TEF.

Epidemiology

Tracheoesophageal fistula occurs in about 1 in every 3000 to 1 in 4500 births and remains one of the major challenges in neonatal surgery. With surgical repair, the rate of survival exceeds 90% even in low birth weight infants. Significant mortality is now limited to infants with severe coexisting congenital or chromosomal abnormalities². Congenital heart disease is the most common co-morbidity and can be a major determinant in survival³.

Neonates with TEF and EA frequently have associated anomalies described by the acronym VACTERL. These anomalies and their associated incidences include *Vertebral* 17%, *Anal* 12%, *Cardiac* 20%, *Tracheoesophageal fistula* and *Esophageal atresia*, *Renal* 16%, *Limb* 10% and other midline defects (cleft lip and palate 2%, sacral dysgenesis 2%, urogenital abnormalities 5%)⁴ (Table 1).

Table 1
Congenital Anomalies Associated with EA and TEF^{4,5,6}

Type	Incidence	Examples
Vertebral	17%	Scoliosis, vertebral defects
Anal	12%	Imperforate anus malrotation, duodenal atresia
Cardiac	20%	VSD, PDA, tetralogy of Fallot, ASD, right-sided aortic arch
Renal	16%	Renal agenesis/dysplasia, hypospadias, polycystic/horseshoe kidney
Limb	10%	Radial anomalies, polydactyly, lower-limb defects

Embryology

During embryologic development, the trachea and esophagus begin as a ventral diverticulum of the foregut. Around the third week of intrauterine life, a proliferation of endodermal cells appears on the lateral aspect of the growing diverticulum. The cell masses then divide the foregut into tracheal and esophageal tubes⁵. Tracheoesophageal abnormalities occur as a result of interruption of this normal event. However, the primary mechanism of EA and fistula formation is unknown. Neonates with TEF and EA have a 50% chance of having one of the associated VACTERL embryologic anomalies. Rarely, EA and TEF may be associated with DiGeorge syndrome, Pierre-Robin syndrome, Holt-Oram syndrome, and polysplenia³.

Pathophysiology

The two main pathological entities in the neonate with TEF are dehydration and aspiration pneumonitis. Saliva and secretions accumulate in the upper esophageal pouch and normal swallowing is disturbed. Contamination of the lungs as a result of spillage from the pouch and/or aspiration of gastric contents through distal TEF results in atelectasis and pneumonitis⁶.

Clinical Features and Manifestations

Excessive amniotic fluid or polyhydramnios on prenatal ultrasound arouses suspicion of EA or some obstruction of the gastrointestinal tract. After delivery, the presence of atresia is usually confirmed by the inability to pass a nasogastric tube into the stomach. Clinical features after birth include excessive salivation, coughing, gagging, and choking, cyanosis and regurgitation associated with attempted feeding. Pulmonary aspiration of gastric contents results in atelectasis and pneumonitis in neonates with EA and TEF. Because birth before term occurs in 30% to 40% of these neonates, respiratory distress of prematurity may also contribute to pulmonary impairment⁷.

The Gross classification of EA and TEF describes 6 types of defects. Type A represents esophageal atresia without fistula. Type B illustrates esophageal atresia with proximal fistula. Esophageal atresia with distal fistula is classified as type C and is the most common type occurring in 80% to 90% of cases (Fig. 1).

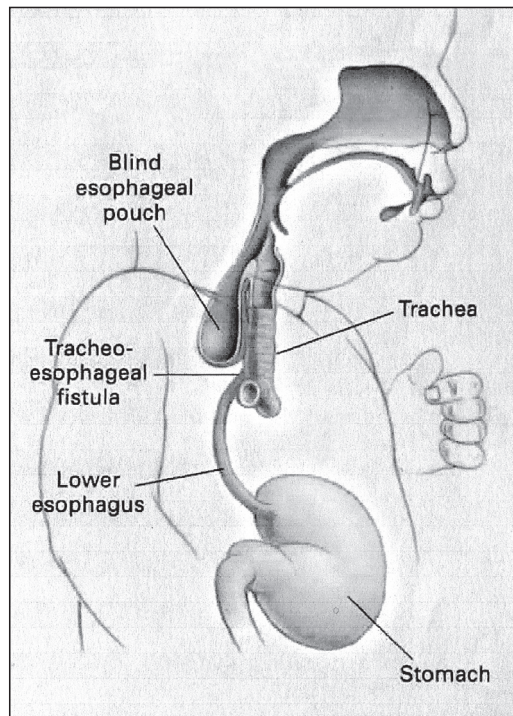
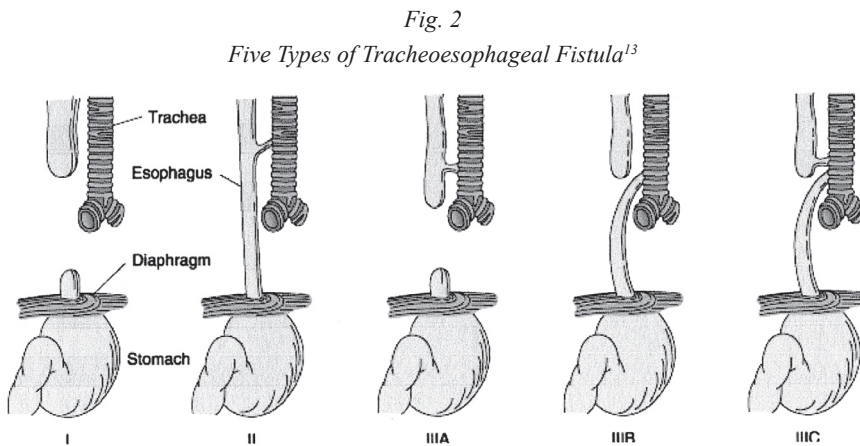


Fig. 1
Type C Tracheoesophageal
fistula¹

Type D represents EA with proximal and distal fistula. TEF without atresia is classified as Type E. Type F represents esophageal stenosis.

Five types of TEF have been described based on the anatomic characteristics of the esophagus and the trachea (Fig. 2). Type I represents esophageal atresia with no fistula. In type II, there is no atresia and a communication between the trachea and esophagus (H-type fistula) is present. Type IIIA has esophageal atresia and a communication between the upper portion of the esophagus with the trachea. In Type IIIB (Type C in the Gross classification system), esophageal atresia occurs with a blind upper pouch and the lower segment communicates with the trachea. This is the most common form of TEF. Type IIIC has atresia with both upper and lower segments communicating with the trachea⁶.



Source: Morgan GE, Mikhail MS, Murray MJ: *Clinical Anesthesiology*, 4th

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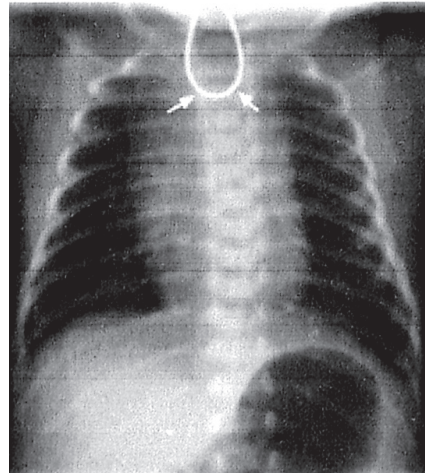
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Major co-existing cardiac anomalies are present in 20% of neonates with EA and TEF including ventricular septal defect, tetralogy of Fallot, patent ductus arteriosus, coarctation of the aorta, and atrial septal defect⁴. Post-natal echocardiogram is generally performed to identify such anomalies.

Diagnosis

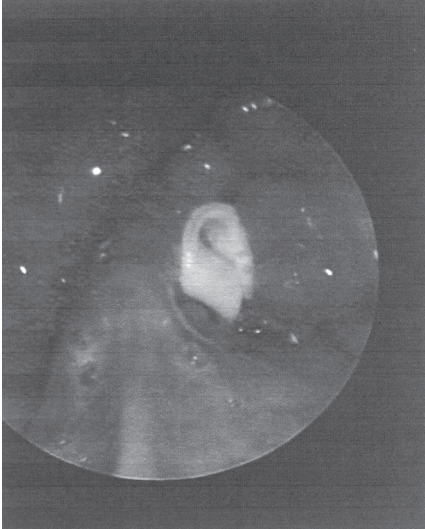
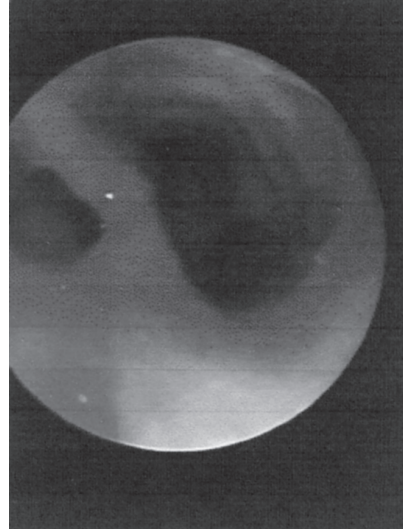
Diagnosis of EA is usually made shortly after delivery by the inability to pass a nasogastric tube beyond 8 to 10 cm⁶. Polyhydramnios is diagnosed prenatally and when no swallowing or stomach contents are seen, suspicion of EA and TEF is high. Confirmation of the diagnosis at birth by chest x-ray shows a nasogastric tube curled up in the upper chest or neck (Fig. 3).

Fig. 3
Nasogastric Tube Coiled in Upper Chest¹⁴



In neonates not diagnosed at birth, coughing, cyanosis, or vomiting with onset of feeds, as well as an association with the VACTERL anomalies should raise suspicion of EA and TEF.

Prenatal ultrasound has limited reliability in diagnosis of EA and TEF. Prenatal ultrasonographic suspicion of EA is usually based on the presence of polyhydramnios and a fetal stomach that is either absent or shows reduced filling. When the diagnosis is based on these two signs, outcome cannot be predicted prenatally. However, prenatal ultrasonographic diagnosis of EA and/or TEF enables parents to be prepared for the birth and treatment of their affected child (including transfer to a neonatal center), and prompt neonatal management and earlier identification of associated anomalies is possible⁸.

*Fig. 4**Gore-tex protruding from left side of larynx**Fig. 5**Tracheoesophageal Remnant*

Treatment

Surgical repair is the definitive treatment for EA and TEF. Surgery is generally performed within 24 to 72 hours in otherwise healthy neonates. Delay in surgical correction increases the risk of aspiration of saliva as a result of accumulation in the upper esophageal pouch. Reflux of gastric acid through the lower pouch and a TEF can cause pneumonitis³. A primary repair involves isolation and ligation of the fistula followed by primary anastomosis of the esophagus. A staged repair is an alternative for neonates that are unable to tolerate surgery due to pneumonia and/or other congenital anomalies.

Early diagnosis and aggressive treatment of associated anomalies, particularly cardiac malformations, have resulted in significant decrease in mortality rates. The Waterson classification allows for a risk evaluation to predict outcome and determine surgical timing. Three main factors contribute to this evaluation including birth weight, the presence of additional congenital anomalies, and pneumonia. Birth weight less than 1500 grams and the presence of associated congenital heart disease are

significant predictors of increased morbidity and mortality⁶. Neonates in category A have a birth weight greater than 2500 grams and undergo prompt surgical repair. Neonates in category B have a birth weight of 1800 to 2500 grams or present with pneumonia and congenital anomaly requiring a short term delay in surgical repair. Neonates in category C have a birth weight less than 1800 grams or have severe pneumonia and congenital anomaly. These patients require a staged repair (Table 2).

Table 2
*Waterson Classification*¹³

Category	Weight/Comorbidities	Surgical Timing
Category A	> 2500 grams	Can undergo surgery
Category B	1800-2500 grams or pneumonia or congenital anomaly	Short term delay, needs stabilizing treatment prior to surgery
Category C	< 1800 grams or severe pneumonia or congenital anomaly	Requires staged repair

Except for the most severely ill neonates, most undergo early complete repair. The presence of a birth weight less than 1500 grams and congenital heart disease decrease survival in TEF from 97% to 22%⁹.

Anesthetic Management

The neonate who presents for repair of EA and TEF represents a significant challenge to the anesthesiologist. Some of the difficulties encountered during anesthetic management include ineffective ventilation due to the endotracheal tube being placed in the fistula, massive gastric dilation, severe pre-existing lung disease from previous aspiration of gastric contents and/or respiratory distress syndrome of prematurity, and associated anomalies, particularly cardiac.

Anesthetic and surgical management focuses on ventilating the lungs without ventilation of the fistula. Techniques include awake tracheal intubation and avoidance of muscle relaxants and excessive positive

pressure ventilation until the fistula has been controlled. Special attention to placement of the endotracheal tube is warranted, and gastrostomy, either preoperatively under local anesthesia or soon after induction is sometimes used to decompress the stomach and prevent gastric distentions⁷.

Preoperative preparation of the neonate undergoing repair of EA and TEF involves stabilization of the patient prior to surgery. Preoperative intravenous fluids should be given to avoid dehydration and hypoglycemia. An isotonic fluid (normal saline) should be used to correct hypovolemia followed by maintenance fluids containing glucose (5% dextrose in $\frac{1}{4}$ normal saline) at 4 ml/kg/hour. Acid-base abnormalities should be corrected and respiratory impairments treated appropriately. Prophylactic antibiotics are administered to reduce the risk of perioperative respiratory infection³. Standard monitoring and arterial line placement are indicated

Suction is applied to the upper esophageal pouch and oropharynx, and the neonate may be kept semiupright. A precordial stethoscope is fixed to the left axilla. Either awake intubation or inhalation induction with spontaneous ventilation may be used to facilitate airway management. If awake intubation is performed, careful demonstration of adequate ventilation without gastric distension is achieved before induction of general anesthesia⁷. Proper positioning of the endotracheal tube is facilitated by inserting the tube as far as possible and slowly withdrawing until bilateral ventilation is auscultated. If the fistula is large and just above the carina, the tip of the endotracheal tube may enter the fistula. The endotracheal tube may have to be gradually adjusted to avoid ventilation of the fistula. A Fogarty catheter may be used to occlude the fistula until it is ligated or a neonatal cuffed tube may also be used to occlude the TEF⁴. Preoperative rigid bronchoscopy is generally performed in neonates with TEF to define the position of the TEF and detect other airway abnormalities.

Once the surgeon has ligated the fistula, muscle relaxation and gentle positive pressure ventilation can be initiated⁷. Narcotics are used for

analgesia with volatile anesthetic agent for maintenance. Positioning is left lateral for a right thoracotomy to ligate the fistula and perform esophageal anastomosis. An extrapleural approach to the posterior mediastinum is used by the surgeon whenever possible. Desaturation may occur when the surgeon packs the lung in order to mobilize the distal segment of the esophagus for anastomosis. Expansion of the lung may be required to correct a low oxygen saturation⁵. Hypoxemia may also result from intubation of the right mainstem bronchus, endotracheal tube obstruction by secretions or purulent drainage, bleeding, kinking of the bronchus or trachea, and atelectasis.

Inhalation induction is an alternative to awake intubation. After the neonate is deeply anesthetized, intubation may proceed without muscle relaxation followed by gentle positive pressure ventilation. The endotracheal tube is taped at a location below the fistula and above the carina. The location of the fistula is identified first by listening over the lungs and stomach. Alternatively, prior to inserting the endotracheal tube, rigid bronchoscopy may be performed by the surgeon to locate the fistula. This allows the surgeon to define the position of the TEF and to detect any other airway abnormalities. Intubation of the TEF with a catheter can help the surgeon identify the fistula and evacuate air from the stomach. After rigid bronchoscopy and catheter placement, the endotracheal tube is placed with the tip below the fistula. Muscle relaxant may be administered if ventilation can be achieved without gastric inflation. A caudal catheter advanced to T6-T7 provides an excellent supplement to general anesthesia, as well as post operative analgesia. Avoidance of opioids facilitates early extubation. However, neonates less than 2000 grams may require post operative ventilation⁹.

Managing the neonate with a large fistula, especially one that is near the carina, can be difficult. The goal of airway management is to ventilate the lungs adequately without ventilating the fistula. Ineffective ventilation, gastric distention or rupture, hypotension, or gastric reflux can all result from ventilation of the TEF. Current strategies to meet this goal include proper positioning of the endotracheal tube and catheter occlusion of the

TEF. Problems may arise in maintaining the position of the endotracheal tube when the fistula is just above the carina. Patient movement or surgical manipulation may lead to subtle changes in the position of the tube and problems with ventilation. Furthermore, occlusion of the fistula with a Fogarty embolization catheter through the trachea (anterograde occlusion) or gastrostomy site (retrograde occlusion) may or may not be effective. Ventilation must be interrupted to pass the catheter through a rigid bronchoscope and the size of the bronchoscope may limit use of the catheter. Moreover, retrograde occlusion through a gastrostomy is not always an option because gastrostomy is not routinely performed in patients with no other complications. The catheter may occlude the trachea if dislodged making ventilation impossible. Also, the catheter can damage the esophageal mucosa at the balloon site. Vigilance on the part of the anesthesiologist allows for early recognition and correction of intra-operative complications.

Postoperative management includes admission to the neonatal intensive care unit whether or not the neonate is extubated. The need for postoperative ventilator support is based on the degree of respiratory impairment due to previous aspiration and/or respiratory distress of prematurity and associated anomalies. Controversy exists over whether the risk of reintubation is greater in infants less than 2000 grams than the risk of continued intubation. Abrasion to the site of the fistula may be more likely if the trachea remains intubated. However, if laryngoscopy and reintubation is necessary, trauma to the fistula site and traction on the esophageal repair may occur⁹. Surgical postoperative complications include anastomotic leak, stricture, gastroesophageal reflux, tracheomalacia, and recurrent TEF.

Future anesthetic management may involve becoming more familiar with endoscopic surgical procedures as they are becoming an attractive alternative to open procedures¹⁰. Newer airway tools such as the proseal laryngeal mask airway which supports positive airway pressure better than its predecessors and also allows for drainage of gastric fluid and air, thereby decreasing the chance of gastric insufflation¹¹.

Prognosis and Long Term Sequelae

Prognosis in otherwise healthy neonates is good following repair of EA and TEF. The Spitz system is an outcome-based classification system based on birth weight and the presence or absence of major congenital heart disease (Table 3). The mortality rate for EA and TEF is less than 1.5% for patients without major cardiac anomalies and a birth weight greater than 1500 grams³.

Table 3
Spitz Classification¹³

Group	Features	Survival (%)
I	Birth weight > 1500 grams, no major cardiac anomaly	98.5
II	Birth weight < 1500 grams or major cardiac anomaly	82
III	Birth weight < 1500 grams and major cardiac anomaly	50

Respiratory and gastrointestinal complications may persist lifelong. Respiratory sequelae include tracheomalacia, recurrent pneumonia, obstructive and restrictive ventilatory defects, and airway reactivity. In children and adults with a history of EA and TEF, aspiration may manifest as respiratory symptoms and/or recurrent lower respiratory infections¹².

Following repair of EA and TEF, gastroesophageal reflux occurs in 35% to 58% of patients probably due to intrinsic esophageal dysfunction. Postoperative esophageal motility disorders include abnormal peristalsis and impaired lower esophageal sphincter tone. In some patients with severe esophageal motility disorders and poor esophageal emptying, dysphagia may be a long term problem.

Summary

The neonate who presents for repair of esophageal atresia and tracheoesophageal fistula can be especially challenging for the anesthesiologist. Anticipating potential perioperative problems and communicating with the surgeon are essential in the treatment of the

neonate with these congenital defects. Although patients with associated VACTERL anomalies have a poorer prognosis, survival rate of postsurgical repair is greater than 90%. Most children have good long term quality of life but are likely to return to the operating room later in life. Therefore, the anesthesiologist must be familiar with the peri-operative management of the neonate in need of TEF repair and the long term sequelae following repair. Life long problems such as gastroesophageal reflux, tracheomalacia, obstructive and restrictive ventilatory defects, airway reactivity, and recurrent pneumonia should be suspected in patients with a history of TEF repair.

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