Lesson 273: PreAnesthetic Assessment of the Neonate With Tracheoesophageal Fistula

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NEEDS STATEMENT

Tracheoesophageal fistula (TEF) and associated esophageal atresia (EA) in the neonate present during the first week of life. These congenital defects can be complicated by aspiration, respiratory distress, and other congenital anomalies. The knowledge and ability of the anesthesiologist to anticipate challenges in managing neonates requiring repair play an important role in the treatment and survival of patients. Also, it is not uncommon for anesthesiologists to care for patients later in life following repair of TEF. A familiarity with immediate complications and long-term outcomes and sequelae after TEF repair is important. The management of neonatal anesthesia has been identified by committee as required information for anesthesiologists.

Target Audience

Anesthesiologists

Learning Objectives

At the end of this activity, the participant should be able to:
1. Summarize the special anesthetic problems associated with managing the neonate with EA and TEF.
2. Describe the generally accepted classification systems of EA and TEF.
3. List associated anomalies.
4. Describe the embryologic development of EA and TEF.
5. Identify the main pathophysiologic entities associated with EA and TEF.
6. Describe the clinical features of TEF.
7. Develop an anesthetic plan.
8. List potential perioperative complications.
9. Discuss the prognosis after TEF repair.
10. Describe the long-term sequelae of TEF repair.

Case History

An 11-year-old girl presented with shortness of breath, related to physical activity, that had been occurring for 2 months. For the previous 2 days she had also been experiencing dyspnea at rest. Her medical and surgical history was complicated, including repair of TEF, ventricular septal defect, patent ductus arteriosus during infancy, and anterior cricoid split and medialization thyroplasty at the age of 9 years. An examination with flexible fiber optics revealed laryngotracheal stenosis, immobility of the true vocal cord, and apparent granulation tissue or a foreign body causing an almost complete obstruction of the larynx. The patient was scheduled for direct laryngoscopy, bronchoscopy, and possible tracheostomy under general anesthesia.

CALL FOR WRITERS

If you would like to write a CME lesson for Anesthesiology News, please send an e-mail to Elizabeth A.M. Frost, MD, at ElizFrost@laol.com.

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Time to Complete Activity: 2 hours

Release Date: April 2008

Termination Date: April 30, 2009

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Tracheoesophageal fistula (TEF), which manifests in the neonate within the first hours to days of life, is considered a surgically correctable anomaly of the gastrointestinal and respiratory systems. The perinatal anesthetic considerations for the neonate with TEF are of acute importance to the anesthesiologist. Before the first successful staged repair in 1939, esophageal atresia (EA) and associated TEF were uniformly fatal. Advancements in pediatric anesthetic techniques and monitoring, neonatology, and pediatric surgery have decreased mortality, and the survival rate is now above 90%. Premature birth and associated severe congenital abnormalities continue to be the biggest contributors to mortality linked to TEF.

Several systems for classifying EA and TEF have been developed based on atresia and its location relative to the fistula. The Gross classification system describes EA with and without TEF, types A through F. Another well-known classification system describes 5 types of TEF, including types I, II, IIIA, IIIB, and IIIC. Regardless of the classification, the most common form of this anomaly is EA with distal TEF.

Epidemiology

TEF occurs in about 1 in every 3,000 to 4,500 births and continues to be a major challenge in neonatal surgery. With surgical repair, the rate of survival exceeds 90%, even in infants with a low birth weight. At present, significant mortality is limited to infants with severe coexisting congenital or chromosomal abnormalities. Congenital heart disease is the most common comorbidity and can be a major determinant of survival. The acronym VACTERL describes anomalies frequently observed in neonates with TEF and EA: vertebral (incidence, 17%), anal (12%), cardiac (20%), tracheoesophageal fistula, esophageal atresia, renal (16%), and limb (10%). Other midline defects include cleft lip and palate (2%), sacral dysgenesis (2%), and urogenital abnormalities (5%; Table 1).

Embryology

During embryologic development, the trachea and esophagus derive from 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 tubes. 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, or polyplenia.

Preanesthetic Assessment

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Pathophysiology

The 2 main pathologic 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 aspiration of gastric contents through distal TEF results in atelectasis and pneumonitis.6

Clinical Features and Manifestations

Excessive amniotic fluid (polyhydramnios) observed by prenatal ultrasound arouses suspicion of EA or an obstruction of the gastrointestinal tract. After birth, atresia is usually confirmed by the inability to pass a nasogastric tube into the stomach of the newborn. Clinical features include excessive salivation, coughing, gagging, choking, cyanosis, and regurgitation associated with attempted feeding. As mentioned, pulmonary aspiration of gastric contents results in atelectasis and pneumonitis in neonates with EA and TEF. Because 30% to 40% of these neonates are delivered before full term, the respiratory distress of prematurity may also contribute to pulmonary impairment.7

The Gross classification of EA and TEF describes 6 types of defects. Type A is EA without a fistula. Type B is EA with a proximal fistula. EA with a distal fistula is classified as type C and is the most common type, occurring in 80% to 90% of cases (Figure 1). Type D is EA with proximal and distal fistulae. TEF without atresia is classified as type E. Type F is esophageal atresia.

Five types of TEF have been described based on anatomic characteristics of the esophagus and trachea (Figure 2). Type I is EA without a fistula. In type II, there is no atresia, and a communication exists between the trachea and esophagus (H-Type fistula). In type IIA, there is EA and a communication between the upper portion of the esophagus and the trachea. In type IIB, a proximal fistula and multiple tracheal branches exist (Figure 2). Type III, there is a common fistula between the esophagus and the trachea; in type IV, there is a communication between the lower segment of the esophagus and the trachea. These patients require a staged repair. Except for those who are severely ill, most neonates undergo early, complete repair. The survival rate of neonates with TEF and congenital heart disease whose birth weight is less than 1,500 g drops from 97% to 22%.8

Table 1. Congenital Anomalies Associated With EA and TEF

<table>
<thead>
<tr>
<th>Type</th>
<th>Incidence, %</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vertebral</td>
<td>17</td>
<td>Esophageal atresia, vertebral defects</td>
</tr>
<tr>
<td>Anal</td>
<td>12</td>
<td>Imperforate anus, malrotation, duodenal atresia</td>
</tr>
<tr>
<td>Cardiac</td>
<td>20</td>
<td>VSD, PDA, tetralogy of Fallot, ASD, right-sided aortic arch</td>
</tr>
<tr>
<td>Renal</td>
<td>16</td>
<td>Renal agenesis/dysplasia, hypospadias, polyhydramnios</td>
</tr>
<tr>
<td>Limb</td>
<td>10</td>
<td>Radial anomalies, polydactyly, lower-limb defects</td>
</tr>
</tbody>
</table>

ASD, atrial septal defect; PDA, patent ductus arteriosus; VSD, ventricular septal defect

Anesthetic Management

Neonates who undergo repair of EA and TEF have severe lung disease caused by previous aspiration of gastric contents or respiratory distress syndrome of prematurity, and associated—particularly cardiac—anomalies. The anesthetic and surgical management of neonates with TEF focuses on ventilation of the lungs without ventilation of the fistula. Techniques include awake tracheal intubation and avoidance of muscle relaxants as well as excessive positive pressure ventilation until the fistula has been controlled. Special attention to placement of the endotracheal tube is warranted. Gastrostomy—either preoperatively under local anesthesia or soon after the induction of general anesthesia—is sometimes used to decompress the stomach and prevent gastric distention.

Preoperative preparation of the neonate undergoing repair of EA and TEF involves stabilizing the patient before surgery. I.V. fluids should be given preoperatively to avoid dehydration and hypoglycemia. An isotonic fluid (normal saline) should be administered to correct hypovolemia, followed by maintenance fluids containing glucose (5% dextrose in one-fourth normal saline) at 4 mL/kg per hour. Acid–base abnormalities should be corrected and respiratory impairments treated appropriately. Propylhexyl antibiotics are administered to reduce the risk for perioperative respiratory infection.7 The placement of standard monitoring devices and an arterial line is indicated. Suction is applied to the upper esophageal pouch and oropharynx; the neonate may be kept in a semi-upright position. A pre-cordial stethoscope is affixed to the left axilla.

Either awake intubation or inhalation induction with spontaneous ventilation may be used to facilitate airway management. If awake intubation is to be performed, adequate ventilation without gastric distention must be ensured before the induction of general anesthesia.7 Appropriate positioning of the endotracheal tube is facilitated by inserting the tube as far as possible and slowly withdrawing it until bilateral ventilation is confirmed on auscultation. If the fistula is large and just above the carina, the tip of the endotracheal tube may enter the fistula. The position of 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; a neonatal cuffed tube may also be used to occlude the TEF.4

Rigid bronchoscopy is generally performed preoperatively in these patients to define the position of the TEF and detect other airway abnormalities. After the surgeon has ligated the fistula, muscle relaxation and gentle positive pressure ventilation can be initiated.7 Opioids are administered for analgesia with a 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 to mobilize the distal segment of the esophagus for anastomosis. Expansion of the lung may be required to correct a low oxygen saturation.7 Hypoxemia may also result from intubation of the right main bronchus. Obstruction of the endotracheal tube by secretions or purulent drainage, bleeding, kinking of the bronchus or trachea, or 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, before insertion of 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 detect any other airway abnormalities. Intubation of the TEF with a catherer may
Surgical postoperative complications include birth weight >1,500 g; birth weight <1,500 g or <1,800 g; birth weight >2,500 g; and birth weight <2,000 g or <1,500 g. The anesthesiologist leads to early recognition and correction of intraoperative complications. Management of the neonatal patient who has a large fistula—especially a fistula near the carina—may 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. Occlusion of the fistula with a Fogarty embolectomy 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. The size of the bronchoscope may limit the use of the catheter. Furthermore, retrograde occlusion through a gastrostomy is not always an option because gastrostomy is not routinely performed in patients with no other complications. If the catheter is dislodged, it may occlude the trachea, making ventilation impossible. The catheter can also damage the esophageal mucosa at the balloon site. Vigilance by the anesthesiologist leads to early recognition and correction of intraoperative complications.

Postoperative management includes admission to the neonatal intensive care unit whether or not the patient is extubated. The need for ventilatory support postoperatively is based on the degree of respiratory impairment caused by previous aspiration, the respiratory distress of prematurity, and associated anomalies. Whether the risk of reintubation is greater than the risk of continued intubation in infants weighing less than 2,000 g is controversial. Abrasion of the site of the fistula may be more likely if the trachea remains intubated. However, if laryngoscopy and reintubation are necessary, trauma to the fistula site and tractions on the esophageal repair may occur. Surgical postoperative complications include anastomotic leak, stricture, gastroesophageal reflux, tracheomalacia, and recurrent TEF.

**Figure 1. Type C tracheoesophageal fistula.**
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**Figure 2. Five types of tracheoesophageal fistula.**

**Figure 3. Nasogastric tube coiled in upper chest.**

**Table 2. Waterston Classification System**
<table>
<thead>
<tr>
<th>Category</th>
<th>Weight/Comorbidities</th>
<th>Surgical Timing</th>
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<tbody>
<tr>
<td>A</td>
<td>&gt;2,500 g</td>
<td>Can undergo surgery</td>
</tr>
<tr>
<td>B</td>
<td>1,800-2,500 g or pneumonia or congenital anomaly</td>
<td>Short-term delay, needs stabilizing treatment before surgery</td>
</tr>
<tr>
<td>C</td>
<td>&lt;1,800 g or severe pneumonia or congenital anomaly</td>
<td>Requires staged repair</td>
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**Table 3. Spitz Classification System**
<table>
<thead>
<tr>
<th>Group</th>
<th>Features</th>
<th>Survival, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Birth weight &gt;1,500 g; no major cardiac anomaly</td>
<td>98.5</td>
</tr>
<tr>
<td>II</td>
<td>Birth weight &lt;1,500 g or major cardiac anomaly</td>
<td>82</td>
</tr>
<tr>
<td>III</td>
<td>Birth weight &lt;1,500 g or major cardiac anomaly</td>
<td>50</td>
</tr>
</tbody>
</table>


**Prognosis and Long-term Sequelae**
The prognosis in otherwise healthy neonates is good after the repair of EA and TEF. The Spitz system is an outcomes-based classification system based on birth weight and the presence or absence of major congenital heart disease (Table 3). The mortality rate from EA and TEF...
is less than 1.5% for patients without major cardiac anomalies and whose birth weight is greater than 1,500 g. Respiratory and gastrointestinal complications may persist throughout life. 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 recurrent lower respiratory infections.

Following the repair of EA and TEF, gastroesophageal reflux (probably due to intrinsic esophageal dysfunction) occurs in 35% to 58% of patients. Postoperative disorders of esophageal motility 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.

Management of the Case Presented

The patient was premedicated in the holding area with metoclopramide and ranitidine by in I.V. access. She was noted to have inspiratory stridor and sternal retractions. Oxygen saturation was 100% with use of a nonrebreather mask. She was transferred to the operating room and standard monitors were applied. She received an I.V. infusion of dexmedetomidine at a loading dose of 18 mcg over 10 minutes. I.V. ketamine, 15 mg, was titrated slowly to achieve anesthesia while spontaneous ventilation was maintained.

The surgeon evaluated the airway with a rigid bronchoscope. A strip of Gore-Tex membrane could be seen protruding from the left side of the larynx with a large amount of overlying granulation tissue (Figure 4). After further examination, the trachea was found to be normal except for a remnant of the previously repaired TEF (Figure 5). The bronchoscope was removed, and the surgeon inserted a 3.5-mm uncuffed endotracheal tube; correct placement was verified. Sevoflurane and dexmedetomidine at 0.5 mcg/kg per hour were administered for anesthesia. I.V. ketamine, 15 mg, was titrated slowly to achieve anesthesia and for a remnant of the previously repaired TEF (Figure 5). After further examination, the trachea was found to be normal except for the presence of overlying granulation tissue (Figure 4). After correct placement was verified, Sevoflurane and dexmedetomidine at 0.5 mcg/kg per hour were administered for anesthesia. The trachea was found to be normal except for a remnant of the previously repaired TEF (Figure 5). The bronchoscope was removed, and the surgeon inserted a 3.5-mm uncuffed endotracheal tube; correct placement was verified. Sevoflurane and dexmedetomidine at 0.5 mcg/kg per hour were administered for anesthesia. I.V. ketamine, 15 mg, was titrated slowly to achieve anesthesia while spontaneous ventilation was maintained.

The surgeon inserted an operating laryngoscope to assist the patient with severe esophageal motility disorders and poor esophageal emptying, dysphagia may be a long-term problem.

Summary

Repair of EA and TEF in the neonate can be especially challenging for the anesthesiologist. Anticipation of potential perioperative problems and communication with the surgeon are essential in treating these congenital defects. Although patients with associated VACTERL anomalies have a poorer prognosis, their survival rate is greater than 90% after surgical repair. Most children have a good long-term quality of life but are likely to return to the operating room later in life. Thus, the anesthesiologist must be familiar not only with the perioperative management of neonates undergoing TEF repair but also with the long-term sequelae after surgery. Lifelong problems such as gastroesophageal reflux, tracheomalacia, obstructive and restrictive ventilatory defects, airway reactivity, and recurrent pneumonia should be anticipated in patients with a history of TEF repair.

References


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For inquiries about course content only, send an e-mail to ram.roth@mssm.edu. Ram Roth, MD, is director of PreAnesthetic Assessment Online and assistant professor of Anesthesiology at The Mount Sinai School of Medicine, New York, NY.

4. The diagnosis of TEF after birth is confirmed by:
   a. barium swallow
   b. bronchoscopy
   c. chest radiography
   d. esophagography
5. Which of the following is true regarding the Waterston classification?
   a. It predicts survival based on birth weight and the presence or absence of congenital heart disease.
   b. It predicts mortality based on 3 main factors, including birth weight, the presence of additional congenital anomalies, and the presence of pneumonia.
   c. Neonates with a birth weight of at least 2,500 g should undergo a staged repair.
   d. It suggests that low-birth-weight neonates with or without congenital heart disease have increased mortality.
6. All of the following are potential difficulties encountered during the anesthetic management of the neonate with TEF except:
   a. ineffective ventilation as a result of placement of the endotracheal tube in the fistula
   b. severe preexisting lung disease from previous aspiration of gastric contents
   c. respiratory distress syndrome of prematurity
   d. inadequate ventilation as a result of the use of muscle relaxants being contraindicated
7. The goal of airway management in the neonate with esophageal atresia (EA) and TEF is:
   a. to secure the airway immediately by intubation and mechanical ventilation
   b. to ventilate the lungs adequately without ventilating the fistula
   c. achieved only with the use of a cathereter occluding the fistula
   d. to not interrupt ventilation at any time during repair of the fistula
8. Which of the following is true regarding postoperative management of the neonate after TEF repair?
   a. Admission to the neonatal intensive care unit is not usually necessary.
   b. Surgical complications include anastomotic leak and stricture.
   c. Failure to meet standard extubation parameters is the basis for determining the need for postoperative ventilatory support.
   d. Reinstitution of the neonate postoperatively poses no risk to the surgical repair.
9. Long-term sequelae of TEF repair are least likely to include:
   a. recurrent pneumonia
   b. obstructive and restrictive ventilatory defects
   c. disorders of esophageal motility
   d. hiatal hernia
10. The mortality rate for neonates with EA and TEF after repair is:
    a. consistent whether or not associated congenital anomalies are present
    b. high for the majority of neonates undergoing repair
    c. difficult to estimate because of various coexisting congenital anomalies
    d. less than 1.5% for those with no major cardiac anomalies and a birth weight greater than 1,500 g.