Anesthetic considerations in the patient with tracheoesophageal fistula

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This article provides a review of the medical and surgical management of the infant/neonate with tracheoesophageal fistula, including the condition's diagnosis, the preoperative preparation of the patient, anesthetic considerations, the surgical correction, postoperative care, and finally prognosis. The authors point out that recent medical and surgical advances have greatly influenced the prognosis and enabled most children with this anomaly to lead normal lives.

Forty-five years ago all the infants with tracheoesophageal fistula died. Today the survival rate is nearly 100% in full term infants, free of other anomalies, who are treated prior to the development of pulmonary complications. Many medical and surgical advances have made the management of this complicated anomaly less chaotic and have enabled most of these children to lead normal lives.

The anomaly of tracheoesophageal fistula (TEF) with esophageal atresia (EA) was first described by Thomas Gibson in Anatomy of Humane Bodies Epitomized (London, third edition) in 1697. However, it was not until 1936 that the first surgical repair was attempted at Children's Hospital Medical Center in Boston by Mixter, who described an extrapleural division of the fistula. The first successful “staged” repairs were accomplished by Ladd and Leven in 1939.

Other surgical pioneers were Haight and Towsley who completed the first successful primary esophageal anastomosis with ligation of the TEF in 1941. Finally, in the 1950's and 1960's respectively, Gross classified the types of esophageal atresia and tracheoesophageal fistula (Figure 1), and Waterson, et al. assigned risk factors predicting morbidity and mortality (Table I).

The defect of TEF occurs between the 21st and 24th day of fetal development when the foregut fails to separate and divide into the larynx and trachea anteriorly and the esophagus posteriorly. Abnormal septation at any point leads to the various types of TEF described. Despite intensive investigation, no environmental or genetic factor has been identified as a cause of this malformation.

Incidence and diagnosis

The incidence of EA/TEF is approximately 1:3000 live births. The Boston area reports one of the highest occurrences of 1:500 live births. Most studies have shown that males are more often affected than females.

A maternal perinatal history often reveals polyhydramnios during pregnancy and premature labor. Although the Apgar scores are usually good, a neonate with excessive nasopharyngeal secretions, choking, dyspnea, or cyanosis should be suspect for EA/TEF. Inability to pass a no. 8-10
French catheter into the stomach and/or feeding difficulties with regurgitation of all ingested fluids is pathognomonic of esophageal atresia.

Abdominal distension, aspiration, and subsequent pneumonitis, as well as the aforementioned signs and symptoms are most likely indicative of TEF type C, the most common malformation.

The diagnosis of TEF can usually be made after a complete history and thorough physical examination. Clinical diagnosis of EA/TEF may be confirmed radiographically when an opaque catheter is passed and curls up in the proximal esophageal pouch. The length of the pouch can also be determined at this time. The use of contrast media, such as barium, is not required and is potentially hazardous without fluoroscopic guidance, as the pouch can overfill causing spillage into the trachea.

A chest radiograph demonstrating air in the stomach indicates that a fistulous connection exists between the trachea and the distal esophageal segment (types C,D,E). Therefore, absence of intestinal gas in the presence of EA generally indicates that a rare proximal TEF exists or that no TEF is present (type B,A).

The most difficult diagnosis to make is that of TEF without EA, termed the “H type” because of its anatomical appearance (type E). Gaseous distension in the newborn is often the presenting symptom, but the diagnosis may not be made for several months when the infant develops a chronic or recurrent pneumonia. Thus, the advantages of diagnosis before the first feeding are obvious, and, when the diagnosis is not made immediately, these infants usually develop a severe pneumonia. Two factors contributing to the development of pneumonia are (1) chronic aspiration of saliva from the upper pouch and (2) regurgitation of gastric juice up the distal esophagus into the lungs via the TEF.

Lastly, the incidence of pulmonary complications seems to be related to the time of diagnosis and the age at which surgical correction is undertaken. This greatly influences the infant’s chances for survival.

Associated anomalies occur in approximately 50% of patients with TEF and 34% with EA that weigh less than 2.5 kg. A list of these anomalies with their relative incidences is found in Table II. Neuroskeletal defects occur most often. Tracheoesophageal fistula is also seen in association with cardiovascular, gastrointestinal tract, and genitourinary tract malformations. Tracheoesophageal fistula is one of the major features of the VATER association described by Quan and Smith in 1962, which consists of vertebral defects, anal atresia, tracheoesophageal fistula with esophageal atresia, and

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<th>Table I</th>
<th>A, B, C, risk assignment</th>
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<tr>
<td>A. Over 5½ lb (2.5 kg) and well.</td>
<td>Predicted survival: 95%.</td>
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<tr>
<td>B. 1. Birth weight 4 to 5½ lb (1.8-2.5 kg) and well.</td>
<td>2. Higher birth weight, moderate pneumonia, and a congenital anomaly.</td>
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<tr>
<td>C. 1. Birth weight less than 4 lb (1.8 kg).</td>
<td>2. Higher birth weight, severe pneumonia, and a severe congenital anomaly.</td>
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renal or radial dysplasia. VATER is the acronym used to describe this association of anomalies.

Once the diagnosis of TEF has been confirmed, all feedings by mouth are discontinued and the infant is positioned upright to minimize regurgitation of gastric secretions through the fistula. The upper esophageal pouch is suctioned continuously with a double lumen catheter (Replogle tube) to prevent aspiration of secretions. Gastric distension with air may interfere with diaphragmatic excursion and result in respiratory embarrassment. Acute relief of gastric distension may be accomplished with needle aspiration followed by surgical gastrostomy under local or general anesthesia. Gastrostomy is an important aspect of treatment in the patient who has a fistulous communication between the distal esophageal segment and the trachea. The advantages of a gastrostomy include:

1. A decreasing of incidence of pneumonia secondary to regurgitation of gastric contents by decompression of the stomach.
2. The providing of a means of relieving severe gastric distension during the delivery of positive pressure ventilation.

### Table II
**Associated anomalies**

1. Neuroskeletal and musculoskeletal anomalies, specifically vertebral and spinal malformations (20-30%) including radial amelia, polydactyly, syndactyly, wrist anomalies and knee malformations.
2. Cardiovascular anomalies (15-20%) including ventricular and atrial septal defects, patent ductus arteriosus, coarctation of the aorta, truncus arteriosus, Tetralogy of Fallot, transposition of the great arteries, and tricuspid atresia.
3. Gastrointestinal anomalies (10-15%) such as anorectal agenesis, malrotation of the gut, duodenal atresia, annular pancreas, and Meckel’s diverticulum.
4. Genitourinary anomalies (10-15%) including renal agenesis, lobulation, malposition, hydronephrosis, ureteral anomalies, and hypospadius.
5. Craniofacial malformations (9.7%).
6. Central nervous system anomalies (7.2%).
7. Pulmonary anomalies (2.1%).
8. Chromosomal defects, especially Trisomy 13 (1.7%).


3. The offering of a means by which one may deliver feedings safely.

These infants should also be transferred to a facility equipped to care for neonatal surgical patients. Surgical repair is not urgent and is usually performed within 12-48 hours once the infant is stabilized. Intravenous access is established and any acid/base, electrolyte, or glucose abnormality is corrected. Routine newborn laboratory tests, such as hematocrit, calcium, bilirubin, and blood sugar are screened, as many of these infants are also premature. A complete physical examination is essential to detect any associated anomalies. An electrocardiogram and echocardiogram are also useful to uncover asymptomatic cardiac disease. Since the neonate’s pulmonary vascular resistance may remain elevated for several days after birth, left-to-right shunts may not become evident for some time.

**Anesthetic considerations**

Anesthesia for TEF ligation and primary repair of esophageal atresia

**Table III**

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<th>Monitoring devices should include:</th>
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<td>— Electrocardiogram.</td>
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<td>— Oxygen analyzer for regulation of N₂O/O₂/air admixtures.</td>
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<tr>
<td>— Blood pressure cuff with doppler over radial artery, or an automatic blood pressure device (Dinamap™), or an arterial line for direct blood pressure readings and blood sampling to achieve a PaO₂ between 50-70 torr.</td>
</tr>
<tr>
<td>— Stethoscope on the left anterior chest wall and/or an esophageal stethoscope if feasible.</td>
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<tr>
<td>— Rectal temperature probe.</td>
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**Ways to maintain normothermia:**

— Increase environmental temperature (26-28°C).
— Heating blanket.
— Radiant heat lamps (at proper distance) for use during induction and preparation of the patient before draping.
— Cap for infant’s head.
— Warm, humidified anesthetic gases with in-circuit temperature measurement and regulation to maintain an inspired temperature less than 35°C. (Humidity also helps to prevent depression of the mucociliary transport system.)
— Warm skin prepping solution.
— Clear plastic sterile surgical drape which is lighter in weight than the usual surgical drapes (also provides better visibility of the infant).
pair of esophageal atresia requires the usual considerations afforded the newborn. These include maintenance of normothermia, metabolic homeostasis, and careful monitoring of arterial oxygen tension to minimize the risk of retinal oxygen toxicity in infants less than 44 weeks gestation. (Table III.)

The infant is positioned in a semi-erect fashion and the proximal pouch is suctioned. A satisfactory intravenous route is established with a plastic cannula and D$_{2}$/4NS at 5 ml/kg is started for maintenance fluid requirements using a microdrip chamber. Atropine sulfate (0.1 mg/kg) IV or IM is administered prior to intubation to block vagal tone. The infant is then preoxygenated for 3 minutes and an awake intubation is performed. Usually a 3.0-3.5 mm (internal diameter) endotracheal tube without a side hole is required to effectively bypass the fistula. There is an abnormally high incidence of a small cricoid ring which may complicate the intubation.17

An assistant is needed to assess the infant's breath sounds while the endotracheal tube is advanced until the tip is at the level of the carina or actually down the right mainstem bronchus. It is then withdrawn 0.5-1.0 cm until breath sounds are heard over both lung fields and not the stomach.18

Although it is unusual to intubate the fistula, it has been reported and must be avoided. Keeping the bevel of the tube anterior and downward during intubation may reduce the risk of intubating the fistula which is usually above the carina.12,19 It is also important to permit an air leak at about 20-25 cm of H$_{2}$O around the endotracheal tube, as the smallest diameter of the infant larynx is at the cricoid ring. If the tube is too tight, subglottic edema may occur with post-extubation "croup" and subglottic stenosis.

The patient is positioned for a right or left thoracotomy depending on which side the aorta is found on echocardiogram. The breath sounds are again assessed and are continuously monitored with a left anterior chest wall stethoscope to detect the occurrence of an endobronchial intubation. Right endobronchial intubation may manifest itself by loss of breath sounds over the left lung field, followed by cyanosis, bradycardia, and hypotension. It is imperative that the endotracheal tube be repositioned promptly as this is a life-threatening situation.

Dried secretions and blood may also cause blockage of the endotracheal tube, and a spare tube should be readily available if reintubation becomes necessary. Intermittent tracheobronchial suctioning may help alleviate potential endotracheal tube obstruction. Esophageal stethoscopes are also useful monitoring devices but are not accepted by all surgeons. Anything passed into the blind esophageal pouch must be done carefully to avoid possible perforation. The esophageal stethoscope will need to be removed before the esophageal anastomosis is begun.

If a previous gastrostomy has not been performed, it should be done prior to thoracotomy, especially if gastric distension with air becomes a problem. Acute gastric distension can be relieved with needle aspiration and, indeed, cardiac arrest has been reported from severe gastric distension which has impinged on diaphragmatic excursion during positive pressure ventilation without a gastrostomy.20

Diffusion of nitrous oxide may aggravate gastric distension and should be avoided in some cases. Air/oxygen admixtures can be used to keep the inspired oxygen tension lower if the nitrous oxide is discontinued. Occasionally, in order to provide adequate ventilation, the gastrostomy tube may have to be partially or totally clamped intermittently. The anesthetist should add an extension to the gastrostomy tube so that the degree of occlusion can be readjusted to suit ventilatory needs.

Incision is usually made under the right scapula and across the midaxillary line at the fourth intercostal interspace allowing the surgeon to approach the vagus nerve, esophagus, trachea, and fistula by either the retropleural or transpleural route.21 Lung collapse, hypoxia, and hypercarbia usually occur if positive pressure ventilation is not instituted and are most likely to occur during lung retraction.8 Surgical retraction of the upper lung may kink the main bronchus, and it must be remembered the right lung will be at least partially collapsed during surgery because the trachea is approached extrapleurally or transpleurally through a right thoracotomy. Thus, periodic hyperinflation of the lungs will help expand atelectatic areas secondary to retraction and hypoventilation.

Muscle relaxants, such as pancuronium bromide (0.1 mg/kg), can be safely used once the fistula is ligated and are helpful when the esophageal anastomosis is being made to block diaphragmatic movements. Many anesthetists feel that muscle relaxants and controlled ventilation are desirable right from the start if controlled ventilation is employed gently.

Metabolic homeostasis is a vital aspect of any anesthetic care plan. The anesthetist must continuously assess blood loss through observation and weighing of surgical sponges. In one series, the
range of blood replacement was 10-200 ml with an average of 68 ml in the first series and 42 ml in the second. This was equivalent to approximately 30% of the estimated blood volume. Metabolic acidosis and hypoglycemia are also potential problems when large volumes of blood are administered. Third space requirements can be replaced with Ringer's lactate at 2-4 ml/kg/hr. The hematocrit should be followed serially and maintained at or greater than 40%.

Following an uncomplicated repair and anesthetic, most infants without other pre-existing problems can be reversed with neostigmine methylsulfate (0.06 mg/kg) and atropine sulfate (0.2 mg/kg) IV and extubated after gentle suctioning of the trachea and esophagus, taking care not to traumatize the suture line. The infant must exhibit return of normal neuromuscular function and be awake prior to extubation. A peripheral nerve stimulator is a useful guide to assessing the return of neuromuscular function. Infants with complicated preoperative or intraoperative courses should be mechanically ventilated until they are well enough to support their own respiratory function. Neck extension should be avoided to prevent tension on the esophageal anastomosis.

If primary esophageal anastomosis is not possible, it is delayed until 6-12 months of age and is accomplished using a segment of colon as a bridge. This procedure is called a "colon interposition." A proximal TEF with EA (type B) is managed with gastrostomy, cervical esophagostomy, and later colon interposition.

Summary

In summary, the choice of anesthetic technique is less important than performing it in a meticulous fashion.

It is essential that these infants be cared for postoperatively in an intensive care unit by staff familiar with this surgical lesion. The postoperative care of these infants is directed at prevention and management of pulmonary complications. Since pulmonary complications are the most common cause of death in these patients, one can expect that at least 25% of these infants will require postoperative ventilatory support.

If mechanical ventilation becomes necessary, a nasotracheal tube is preferred postoperatively as it is most easily secured and tolerated by the patient. The pharynx is suctioned frequently with a soft catheter which is measured and the length clearly marked so that the anastomosis site is not traumatized. Gastrostomy feedings are started 3-5 days postoperatively or a central venous line for total parenteral nutrition is established.

A pulmonary complication commonly reported is tracheal collapse secondary to tracheomalacia, which occurs in approximately 25% of patients. Tracheomalacia is produced by the dilated proximal esophageal pouch which compresses the trachea during intrauterine development. Disruption of the esophageal anastomosis is another common complication. Signs of anastomosis leak depend on whether a retropleural or transpleural surgical approach was used. If a transpleural approach was used, respiratory distress may be sudden and alarming.

A review of survival rates indicates that the fate of many of these infants is probably determined by prematurity, associated anomalies, and the degree of pulmonary insult before surgery is undertaken.

Advanced medical care of the premature neonate as well as a greater number of staged operations in poorer risk patients have increased survival rates. The overall survival rate is reported at 65-75%, and is nearly 100% in term infants free of other major anomalies who are diagnosed and treated prior to the development of aspiration pneumonitis. Advances in respiratory support and total parenteral nutrition have largely contributed to improved survival rates as well.

Long-term problems, such as abnormal esophageal motility leading to gastroesophageal reflux, chronic aspiration, bronchospasm, restrictive lung disease, and esophageal stricture occur in children who have undergone successful surgical repair of EA/TEF. Abnormal respiratory function has also been detected 7-18 years later in these children. A total of 65% also had reactive airways. However, a long-term study of 242 patients with successful repair of TEF at Children's Hospital Medical Center in Boston demonstrated that approximately 90% of these patients were asymptomatic 15-25 years later.

This uncommon and serious congenital anomaly presents a medical and surgical challenge to all those involved with gratifying results for these patients and families.

REFERENCES


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