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Anesthetic Management of Tracheal Esophageal Fistula with Distal Tracheal Stenosis

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Esophageal atresia with tracheal esophageal fistula (TEF) is a rare anomaly of the aerodigestive tract. Although closure of the TEF is usually successful, postoperative morbidity and mortality may occur from perioperative reflux and pulmonary aspiration of gastric contents; therefore, the TEF should be closed as soon as the infant's condition permits operative intervention. Congenital anomalies of the respiratory tract are rare, although tracheal stenosis and atresia proximal to the tracheal esophageal fistula have been reported.[1–6]

We report a case of severe distal tracheal stenosis in a child with esophageal atresia (EA) with TEF and associated duodenal atresia.

CASE REPORT

A 12-h-old 2.0-kg male delivered at 32 weeks because of placenta previa was scheduled for emergency gastrostomy and TEF ligation. The apgar score was 4 and 8 at 1 and 5 min, respectively. Respiratory distress occurred immediately, and necessitated tracheal intubation. During transport to our hospital, the trachea was extubated accidentally, but the infant remained stable with only mild respiratory distress. Therapy included supplemental oxygen and constant suction of the blind proximal esophageal pouch.

Prior to induction of anesthesia, electrocardiographic leads, arterial blood pressure cuff, precordial stethoscope, and pulse oximeter were applied to the infant. Systemic arterial blood pressure was measured with an existing umbilical arterial cannula connected to a transducer and displayed. Atropine, 0.10 mg iv, and oxygen preceded intubation of the trachea. Laryngoscopy was uneventful, and the trachea was intubated with a 3.0-mm i.d. endotracheal tube (ETT). The bevel of the ETT was rotated posteriorly to avoid accidental intubation of the TEF. Breath sounds were bilaterally equal, and oxygenation was maintained. Gallamine, 10 mg, was administered iv.

With the onset of positive-pressure ventilation, the infant became intensely cyanotic (pH, 6.8; Pa_{O_2}, 2 mmHg; Pa_{CO_2}, 121 mmHg; BE, −17 mEq/l) and bradycardic (40 bpm). Equipment malfunction was ruled out, and repeated laryngoscopy twice confirmed translaryngeal placement of the ETT. After emergent percutaneous decompression of the stomach and placement of a 3.5 mm ETT, oxygenation improved immediately. *Via* a right thoracotomy with retropleural dissection, the distal tracheoesophageal fistula was ligated and divided, and primary esophagoesophagostomy was performed without difficulty. Correction of the duodenal atresia was postponed.

Neuromuscular blockade was reversed with neostigmine, 70 μg/kg, and atropine, 0.10 mg iv. After clinical evaluation and train-of-four determination, the trachea was extubated. Respiratory distress and cyanosis recurred immediately, but were relieved by positive-pressure ventilation by mask. Endotracheal reintubation was unsatisfactory because the ETT appeared to enter the remnant of the TEF. On the third attempt, laryngoscopy, bronchoscopy, and intubation with a rigid fiberoptic light (Storz-Hopkins Zero-Angle Rod Light; Karl Storz, Inc., Culver, CA) allowed placement of an ETT beyond the orifice of a large blind pouch into the proximal end of an extremely narrowed trachea.

Abnormal findings during diagnostic laryngoscopy and bronchoscopy with a 3.0-mm rigid ventilating bronchoscope were confined to the distal trachea. A circumferential stenotic congenital segment of trachea with a fibrous band in the posterior wall was seen (lumen diameter estimated at 2 mm). Immediately cephalad and posterior to this stenosis was a large, pouch-like remnant of the TEF. The carina could not be visualized. After bronchoscopy, a 3.5-mm ETT was placed in the correct intratracheal position, as previously described. The right thorax was again opened, and tracheoplasty was accomplished by rotating a portion of the wall of the TEF pouch onto the trachea. Because the ETT could still not be advanced, it was exchanged using a guide-wire technique for a 2.0-mm ETT which was left across the tracheoplasty as a stent.

Initially, the infant was mechanically ventilated (FI_{O_2} = .30–.35;

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positive inspiratory pressure (PIP) = 24–29 cm H_2O; RR = 30–40 bpm; PEEP = 3 cm H_2O) without difficulty (Pa_{O_2} = 61–98 mmHg, Pa_{CO_2} = 32–45 mmHg; *p*Ha = 7.30–7.28; BE = 0–10 mEq/l). During the second postoperative week, bronchoscopy demonstrated a healing suture line with a small area of granuloma formation, and the trachea was successfully extubated.

Peripheral and central hyperalimentation was instituted, but the infant developed anasarca and leaky-capillary syndrome (presumedly as a result of sepsis) which limited fluid and, therefore, caloric intake. Inappropriate secretion of anti-diuretic hormone also was documented. Echocardiographic examination of the heart revealed a clinically insignificant right atrial thrombus, but no intrinsic intracardiac defect. Twenty-four hours prior to the infant's death at 6 weeks of age, ventilation and oxygenation deteriorated. Death occurred from respiratory failure secondary to sepsis and malnutrition. Autopsy showed healing of the esophageal anastomosis and the tracheal suture line. Gross abnormalities of cartilage formation, numerous malformed tracheal rings, and a 1-cm length of tracheal stenosis at the site of the tracheoplasty were present (fig. 1).

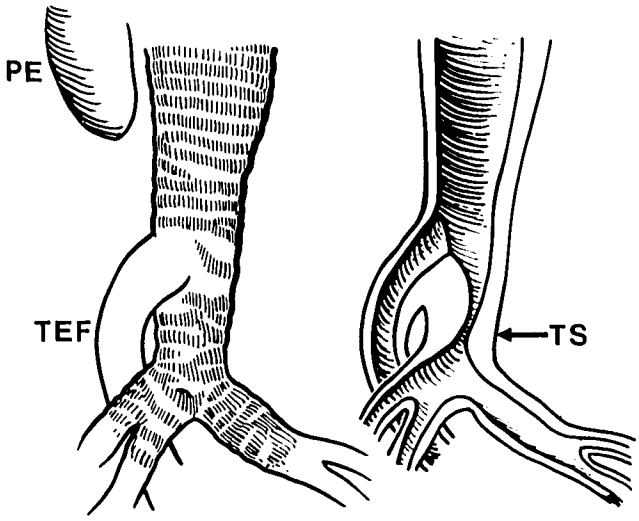

FIG. 1. Preoperative anatomy. Artist's rendition from the intraoperative surgical description and autopsy material including silver stain for cartilage. PE = proximal esophageal pouch; TEF = tracheal esophageal fistula; TS = tracheal stenosis.

DISCUSSION

Although closure of a TEF is usually successful, postoperative morbidity and mortality can occur from perioperative reflux and pulmonary aspiration of gastric contents; therefore, the TEF should be closed as soon as possible after diagnosis. While decreased compliance has been reported as a result of gastric aspiration or from respiratory distress syndrome of prematurity,[7,8] in this instance, the infant's preoperative chest roentgenogram had not shown significant pulmonary disease.

Since the TEF is usually immediately proximal to the carina on the posterior aspect of the trachea, we felt that accidental endotracheal intubation of the TEF was the most likely reason for oxygen desaturation. This was, in fact, the case, but only because the trachea distal to the fistula was too stenotic to permit passage of the ETT. Simultaneous with emergency decompression of the stomach with a large bore cannula, we successfully improved oxygen and ventilation by inserting a 3.5-mm ETT assuming that this size ETT would be too stiff and large to accidentally deflect into a TEF. In all likelihood, the ETT remained at the level of the stenosis for the same reasons. After the initial TEF repair was completed, the infant's trachea was electively extubated. The infant again developed respiratory distress, and direct visualization with a rigid fiberoptic bronchoscope was necessary to successfully place an ETT. During this procedure, the stenotic trachea immediately distal to the TEF pouch was discovered.

In summary, this infant had severe distal tracheal stenosis associated with an obvious TEF. The association of these two defects has not been previously reported. It should be considered in any patient with severe ventilatory problems during evaluation and operation for TEF and esophageal atresia.

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