Persistent leak after ligation of tracheoesophageal fistula – where is the air coming from?

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Objectives:

- Discuss the incidence of major anomalies associated with tracheoesophageal fistula/esophageal atresia, such as laryngotraceoesophageal cleft
- Review the clinical presentations of tracheoesophageal fistula/esophageal atresia
- Discuss different approaches to airway management for repair of TEF/EA in setting of comorbid airway anomalies
- Assess the utility of preoperative tracheobronchoscopy prior to TEF/EA repair
- Discuss implications of surgical planning

Case History:

A two-day-old term male infant weighing 4.1 kg is scheduled for repair of tracheoesophageal fistula. Presenting symptoms included increasing oxygen requirement and worsening reflux with feeds. Preoperative xray and feeding study consistent with TEF/EA type C.

Questions:

What preoperative workup is required prior to TEF repair? Imaging? What are the other subtypes of tracheoesophageal fistulas? What are the other anomalies associated with TEF?

Case history and physical exam (continued):

No concern for additional anomalies, and normal echocardiogram. Patient has been NPO and adequately prepared for surgery. Vital signs are within normal. Oxygen saturation 97% on 1.5 L nasal cannula, BP 68/45 (50), HR 148, 24 gauge PIV in right hand infusing 10% dextrose at 13 ml/hr. Physical exam reveals swaddled child, comfortably sleeping in no acute distress. Cardiac and lung exam were normal.

Questions:
How best is the airway managed for repair of a presumed TEF? Is the oxygen requirement concerning? Is direct laryngoscopy alone sufficient for intubation? How useful is preoperative bronchoscopy? Would you give paralytic for endotracheal tube placement?

**Intraoperative care – induction:**

Induction of anesthesia was performed using sevoflurane with spontaneous ventilation. Endotracheal intubation was accomplished after a propofol bolus. Direct laryngoscopy with a miller 0 blade revealed a Cormack Lehane grade II view with easy intubation. Airway was noted to be erythematous and swollen, which was attributed to worsening reflux. A 3.0 microcuff endotracheal tube was advanced into the right mainstem bronchus and then withdrawn until bilateral breath sounds could be auscultated. Patient was then paralyzed with rocuronium and placed on positive pressure ventilation.

**Questions:**

How useful is preoperative bronchoscopy? Is it beneficial for positioning the endotracheal tube? How does it aid occlusion of the fistula with a catheter? Who provides this service – anesthesiologist or surgeon? How likely is bronchoscopy to identify additional airway anomalies? How should you ventilate this child? Positive pressure ventilation or spontaneous respiration?

**Intraoperative care:**

After an arterial line and second IV were placed, the patient was positioned in left lateral decubitus position and equal breath sounds were heard bilaterally. At several points during the operative portion of the case, ventilation became more challenging, with occasional hypercapnia or altogether loss of end tidal carbon dioxide monitoring that resolved with minor adjustments in endotracheal tube depth. Approximately two hours into the case, during dissection and lung compression by the surgeon, the patient became hypoxic to 85-88%. End-tidal CO2 was lost despite adequate tidal volumes and maintained arterial blood pressure 68/45 (51) by arterial line. ETT was suctioned and surgical lung retraction released. ETCO2 waveform returned after mucus plug was removed and pulse oximeter rebounded into the high 90s with hand ventilation, 100% FiO2 and recruitment breaths. After ligation of the TEF, there was concern for a residual proximal fistula due to the presence of a persistent air leak. The surgeons continued to explore, looking for a second fistula. Approximately five hours into the case, after turn over to 2\textsuperscript{nd} provider, ETCO2 was completely lost and the patient was unable to be adequately ventilated. Extubation was presumed & the surgeons notified. The baby was easily reintubated in the lateral decubitus position with breath sounds again heard equally bilaterally. The 2\textsuperscript{nd} provider noted the airway to be extremely erythematous and swollen. After an hour of dissection, ENT surgery consultation was obtained intraoperatively, with subsequent direct laryngoscopy and rigid bronchoscopy. A grade IV laryngotracheoesophageal cleft was identified, with the cleft extending to one centimeter above the carina.
Questions:
Are there any clinical signs that differentiate TEF from laryngeal cleft? And how likely are they to occur together?

Would TEF repair have been postponed or canceled given gravity of airway anomaly in this particular patient? What is the outcome of complicated TEF repairs or high grade LTEC?

Postoperative care:
The patient remained intubated, sedated and paralyzed in NICU until his trachea grew large enough for ENT to attempt surgical repair of his laryngeal cleft at 6 weeks old & 5.25 kg.

Discussion:
Esophageal atresia occurs in approximately 1: 3,000-4,500 live births and 85% have a fistula from the distal esophagus to the trachea, a tracheoesophageal fistula. Clinical manifestations of esophageal atresia include: maternal polyhydramnios, excessive oral secretions, choking or coughing with feeding, abdominal distension and inability to pass a catheter into the stomach.

Congenital esophageal atresia (EA)/tracheoesophageal fistulas (TEF) have an incidence of approximately 1:2,500-3,000 live births, and coexisting congenital abnormalities exist in over half of EA/TEF which are collectively known as VACTERL or VATER.1 These anomalies include cardiovascular malformation, skeletal anomalies such as hemivertebrae or abnormal radius, imperforate anus, renal and urogenital anomalies. One such comorbid congenital anomaly is the laryngotracheoesophageal cleft (LTEC). The LTEC is a fissure between the laryngotraheal and pharyngoesophageal systems caused by a lack of separation between the laryngotraheal axis and the esophagus.2 The incidence of LTEC is reported as 1 in 10,000-20,000 live births. Clinical manifestations of LTEC are similar to TEF with an additional symptom of aphonia. LTEC is commonly seen (60% of cases) with other congenital abnormalities with the two most common associations being TEF (20-37%) and tracheomalacia.2

Traditional workup for TEF would not include direct laryngoscopy to examine for LTEC, although the presentation may be similar,3 as symptoms of LTEC vary in severity but generally correlate with severity of the cleft.4 Traditional airway management for surgical repair of TEF/EA would not necessitate bronchoscopy; however, preoperative bronchoscopy can be a useful intervention, offering an assessment of location and size of TEF, exclusion/inclusion of multiple TEF sites, and assessment for other potential airway problems, such as LTEC.5,6 Knottenbelt et al. found that bronchoscopy was only utilized in 40.5% of cases during a three year audit from 2005-2008.5 Endoscopic assessment is essential to LTEC diagnosis, although it is possible to miss a low-grade malformation if the examiner is not experienced.7

References