

PBLD SPA 2015

Title: Lost on the road less traveled - Managing the complications from a prior TEF Repair and a Challenging Airway

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Goals& Objectives:

1. Understand the presentations of tracheoesophageal fistulas (TEF) and review the complications and morbidities associated with TEF repairs.
2. Review the airway algorithm with emphasis on the failed intubation.
3. Review and discuss the anesthetic considerations for intrathoracic surgery in an infant including monitoring. Be able to list three different monitors beyond the recommended ASA monitors that would benefit your anesthetic management of these cases.
4. Review options for post-operative pain management of an infant after a major surgical procedure. Compare and contrast regional versus narcotic pain management options.
5. Evaluate risks and discuss options for an anesthetic outside of the OR, in interventional radiology.

Presentation:

You are scheduled to anesthetize a 6-month-old infant for an outpatient EGD. The infant was born at term and has a history of tracheoesophageal fistula (TEF) repair, for which she underwent an uneventful anesthetic. The child also has a left vocal cord paralysis and recurrent emesis with feeds which leads to cyanotic spells. Two days prior to the scheduled EGD, the infant was taken to an outside hospital because of a cyanotic episode that required bag mask ventilation and approximately 30 seconds of chest compressions per the mother. The gastroenterologist suspects an esophageal stricture as the cause of these cyanotic episodes and will perform an EGD with possible dilatation.

The infant has a 24g PIV in place and has been NPO for 8hrs.

*Is there any additional information you would like to know prior to anesthetizing this infant? What is your plan for induction and maintenance of the anesthesia? Do you have any concerns about this patient? What size endotracheal tube would you utilize? Would you choose a rapid sequence induction? Would you use cricoid pressure? What is the right amount of cricoid pressure? Would you confirm correct ETT position with a fiberoptic scope?*

The patient's vital signs are as follows:

Wt. – 5.9 kg, HR – 93, BP – 116/69, RR – 40, SpO2 – 100% on room air

The patient was induced with 20 mg IV Propofol and light cricoid pressure was applied. She was noted to be an easy mask airway. On direct laryngoscopy, she had a grade 1 view of the vocal cords. A 3.5 cuffed ETT was placed easily. No EtCO<sub>2</sub> or chest rise was noted after intubation and the endotracheal tube was removed. She continued to be an easy mask ventilation with cricoid pressure. With a repeated direct laryngoscopy, the tube was again easily passed through the vocal cords under direct visualization. Again, there was no EtCO<sub>2</sub> noted, no chest rise, and no breath sounds were auscultated.

*What is in your differential diagnosis? How deep should the ETT be? What is your next plan of action? Do you want to call for any assistance or equipment?*

After removing the endotracheal tube a second time, the anesthetic was maintained with Sevoflurane and mask ventilation. Vital signs were within normal limits throughout the induction. The ENT surgeon working in the next room was consulted and after switching to TIVA (total intravenous anesthesia) with a Propofol infusion, a rigid bronchoscopy was performed. The rigid bronchoscopy revealed a large remnant cleft/ tracheal diverticulum from the previous TEF repair and some tracheomalacia that preferentially directed the scope (and previously the ETT) into the diverticulum. The diverticulum was noted at 12 cm from the lips (measured on the bronchoscope) and the carina was identified at 14 cm from the lips. The ENT surgeon, utilizing the rigid bronchoscope, intubated the patient with a 3.5 cuffed ETT, which was secured at 13 cm at the lips. Appropriate ventilation with chest rise, breath sounds and EtCO<sub>2</sub> was noted and the gastroenterologist proceeded with the EGD.

The endoscopist noted an esophageal stricture below the previous site of the tracheoesophageal fistula and an esophagram was performed. A balloon catheter was passed beyond the stricture and dilation of the stricture was attempted. The catheter was then removed and a second endoscopy revealed a large perforation of the esophagus. A surgery consult was called emergently. The surgeon declared that an immediate repair of the rupture was warranted due to the severity of the perforation and the patient was prepared for an open thoracotomy.

*What immediate complications can arise from the perforation of the esophagus? How does this affect your anesthetic plan? Is placement of a bronchial blocker indicated or feasible in this case? Would you like any additional monitors? Would you place an arterial line and/or a central venous line? What is your postoperative plan? Would you leave the patient intubated postoperatively? How will you control the patient's pain? Would you place a thoracic epidural? Where should the patient receive postoperative care – in the PACU, the PICU, on the floor?*

Prior to beginning the thoracotomy, a thoracic epidural was placed for postoperative pain control and to facilitate extubation. Near infrared spectroscopy (NIRS) monitors were placed to monitor tissue perfusion in the patient (head and somatic/renal saturations). Surgery was performed without the need for one lung ventilation. At the end of the procedure the child is transferred to the PICU intubated and sedated. The main concern

regarding extubation was the possible need for emergent reintubation that might prove difficult. The child was monitored in the PICU overnight and was extubated the next morning without difficulty.

The infant's vital signs after extubation:

T – 36.2, HR – 100, BP – 105/48, RR – 44, SpO<sub>2</sub> – 95% on high flow NC (FiO<sub>2</sub> 35%)

Three days later, the infant developed a pneumothorax after the chest tubes were removed. You are asked to take the patient to the Interventional Radiology Suite for placement of a chest-tube under fluoroscopy. The patient is currently on 10 L/min high flow nasal cannula.

*What are your considerations when planning anesthesia outside the operating room? Would you perform a general anesthetic and intubate the child? Would you plan a fiberoptic intubation or have ENT available? Are you concerned about an esophageal intubation and an esophageal tear? Is there a risk to performing the procedure with the child breathing spontaneously? Can the procedure be performed with sedation alone? Can the procedure be performed with local anesthetic alone? Do you need to do anything if the epidural is still working well?*

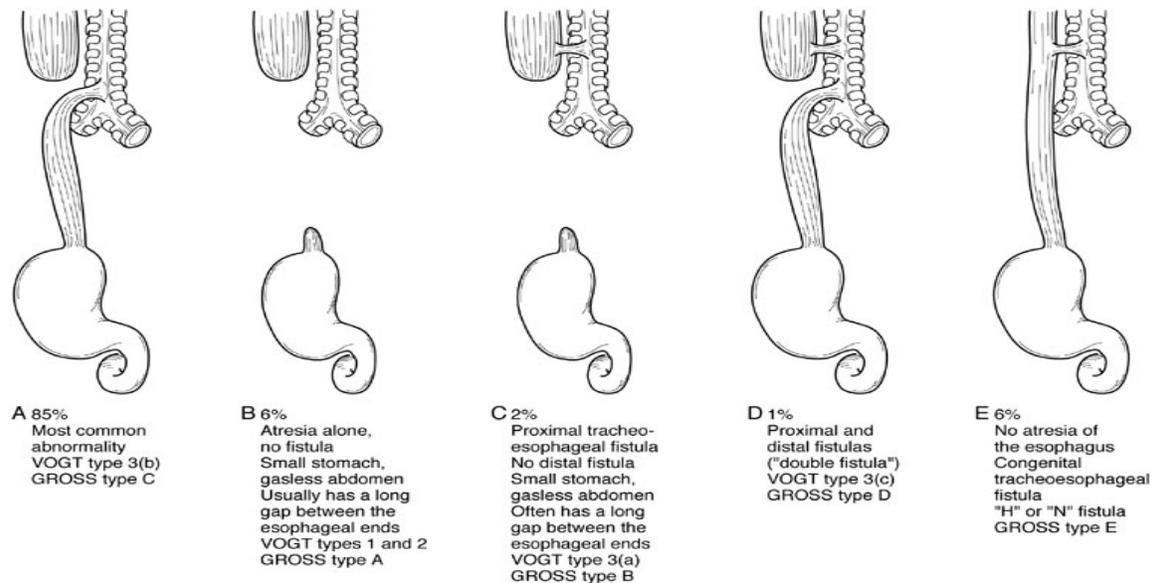
The patient was transported to the Interventional Radiology Suite on a Dexmedetomidine infusion at 0.5 mcg/kg/h and appeared well sedated. The patient was maintained on 2 L/min oxygen via nasal cannula. The epidural was bolused with an additional 1.5 ml of 1% Lidocaine and the interventional radiologist infiltrated subcutaneously at the operative site an additional 3 ml of 0.5% Lidocaine. As the procedure was started, the patient was given incremental doses of intravenous Propofol for a total of 15 mg to minimize movement of the infant. The patient tolerated the procedure well and was transferred back to the PICU.

*Retrospectively, would you have changed anything about this anesthetic technique?*

Problem Based Learning Discussion:

Tracheoesophageal fistula is a congenital anomaly that occurs in 1:3,500 births<sup>2</sup>. It usually involves some malformation of the esophagus and frequently a connection between the esophagus and trachea. There are five different variants of TEF, the most common variant being Type C. This malformation results in a blind end esophageal pouch arising from the oral pharynx and the lower esophagus connecting to the trachea via a fistula. TEF Type A is the only variant that does not include a fistula between the structures of the esophagus and trachea but instead involves only two blind end pouches of the esophagus. TEF Type E (also known as the H type) maintains a continuous esophagus with a fistula from the esophagus to the trachea. TEF Type B involves two blind end pouches of the esophagus and the fistula to the trachea arriving from the

superior esophageal pouch. TEF Type D involves a disruption in the esophagus and the two pouches both connecting to the trachea via two fistulas.<sup>2</sup>



TEF Diagram - Principles and Practice of Pediatric Surgery. Volume 2. 2005. Figure 65-1

TEF can be associated with other congenital malformations, such as in the VACTERL syndrome (vertebral anomalies, anal/rectal malformations, cardiac defect, tracheoesophageal fistula, renal abnormalities  $\pm$  radial limb abnormalities), CHARGE, Goldenhar Syndrome, Feingold Syndrome and Trisomy 18.<sup>2</sup> Surgical correction typically involves ligation of the fistula from the trachea and anastomosis of the esophageal ends. During the surgical repair, it is standard practice to leave a few millimeters of fistula attached to the trachea to minimize stricture of the trachea.

Many TEF patients continue to suffer from sequelae of the malformation or complications of the surgical repair like esophageal dysmotility, vocal cord paresis secondary to injury to the laryngeal nerve, recurrent gastroesophageal reflux (40-70%)<sup>3</sup>, esophageal stricture (30-40%)<sup>3</sup>, anastomotic leak (14-34%)<sup>3</sup> and occasionally rupture of the esophagus.<sup>4</sup> More common are pulmonary complications such as recurrent aspiration pneumonias either from gastroesophageal reflux and esophageal dysmotility or from a recurrence of the tracheoesophageal fistula. Tracheomalacia, secondary to weakening of the mucosa and cartilaginous structures surrounding the previous fistula often requires CPAP therapy or aortopexy<sup>1</sup>. Other tracheal complications include tracheal stricture, occasionally rupture of the trachea, and less commonly a tracheal diverticulum or false passageway. Case reports describe tracheomalacia and tracheal diverticula, which resulted in respiratory failure, apneic events and occasionally in cardiac arrests<sup>1</sup>. The diverticulum and resulting false passageway can result in a difficult intubation and ventilation because the ETT may catch on the mucosa of the diverticulum resulting in the passage of the ETT into the diverticulum.<sup>1</sup> The ASA difficult airway algorithm suggests in cases of ineffective ventilation after successful intubation to consider returning to spontaneous ventilation, mask ventilation or placement of an LMA.

In situations of “cannot ventilate, cannot intubate” it is advisable to proceed down the emergency airway pathway and consider placement of an invasive airway<sup>6</sup>. Similar to our case, in which intubation was followed twice by the inability to ventilate, other authors have also reported successful reestablishment of ventilation by removing the ETT. Bag mask ventilation stents open the tracheomalacia and allows effective ventilation past the diverticulum.<sup>1</sup>

Although still rare, there are now several case reports of tracheal diverticulum that resulted from the surgical correction of a TEF. While many diverticula are corrected during the initial surgical repair, the tracheomalacia and weakened tracheal wall may allow these false passageways to become more prominent with time. Case reports describe obstructive apneic events, chronic infections, aspirations, lobar collapse and respiratory failure with failed intubation. If the experienced practitioner is unsuccessful in establishing effective ventilation despite passing the ETT past the vocal cords under direct visualization, it is reasonable to consider the presence of a false passage or tracheal diverticulum. Intubation may be achieved with placement of the ETT over a fiberoptic or rigid bronchoscope. In all instances reported, ventilation could be maintained successfully with bag mask ventilation.<sup>1&3</sup>

Intubation of a six month old infant requires the use of appropriately sized equipment. The expected tube size for this infant would be a 3.5 to a 4.0 uncuffed ETT. The addition of a cuff on the ETT is the equivalent of an additional half size, so a 3.5 cuffed ETT has the approximate outer diameter of a 4.0 uncuffed ETT. A rough estimate of where the tube should be secured at the lip is to multiply the size of the ETT by three. For example, a 3.5 ETT would be appropriately positioned at around 10.5 cm at the lip. This formula is an estimate and the appropriate positioning should still be confirmed by auscultation and clinical judgment. True rapid sequence induction is not commonly performed in infants. There is a black box warning concerning Succinylcholine at this early age because an infant may not have had enough time to be diagnosed with a muscular dystrophy or other condition that could be detrimental when given Succinylcholine. More commonly, a modified version of rapid sequence induction is utilized in infants including the use of Rocuronium and sometimes small ventilatory breaths because of the infants’ high metabolic demands and tendency to desaturate quickly. Cricoid pressure is commonly applied to infants and children, but unlike adults, their pharyngeal/laryngeal structures are still pliable and too much pressure can result in the inability to ventilate and intubate.

As described earlier, esophageal strictures at the site of the esophageal anastomosis are common after tracheoesophageal fistula repairs<sup>3&4</sup>. These are monitored via Barium or Gastrografin esophagrams if symptomatic and endoscopies. Frequently these strictures require serial dilatations to maintain patency. In our institution, this is accomplished by placing an ultra-thin SDS 4 mm to 8 mm balloon catheter under fluoroscopic guidance to dilate these areas. One of the possible complications of this procedure is the rupture of the esophagus. Associated morbidities of an esophageal tear include pneumomediastinum, mediastinitis, infection of the mediastinum and pneumothorax. In

our patient, the tear was large enough to warrant immediate surgical correction via an open thoracotomy.

The anesthetic management of thoracotomies in infants can prove to be challenging. There is no appropriately sized double lumen tube for a 6-month old patient. This child has a 3.5 cuffed ETT in place. Alternatively a bronchial blocker can be placed to assist in one lung ventilation but this can easily slip and possibly block ventilation of the trachea. Another option is to advance the ETT into the right main bronchus; this maneuver can result in significant pulmonary shunt flow, elevated airway pressures and can significantly impair intraoperative oxygenation. Given the small size of the child, the small ETT and the very tenuous and precise placement of the ETT, we decided to leave the ETT at the initial position. The surgeons were able to optimize their surgical field by packing and retracting the lung.

Intraoperative monitoring is crucial during an infant thoracotomy. The utilization of the standard ASA recommended monitors can be complimented by NIRS (Near Infrared Spectroscopy) to monitor both cerebral and somatic (renal) tissue saturations. NIRS saturations allow conclusions regarding oxygen delivery to the underlying tissues and thus assist in monitoring cardiac output, compression of major vascular structures and a decrease in hematocrit/ blood loss. An arterial line is also useful to monitor intraoperative blood gases and hemodynamic stability of the infant. A central line can help with volume assessment but can be challenging to place in a 6 kg infant. Also, if the line is placed into the thorax, the lateral positioning, open chest, and surgical manipulation may make the information assessed from the central line unreliable. A breathing circuit appropriate for the patient's size and age that minimizes dead space and limits compliance is recommended in small infants to allow close monitoring of EtCO<sub>2</sub> and optimize ventilation.

Intraoperative volume assessment via a combination of perfusion monitoring (NIRS), urine output monitoring, hemodynamic stability and estimated blood loss is crucial. In an infant of almost 6 kg the estimated blood volume is only 480 ml. As in our case, it is imperative to have PRBCs available when operating around large blood vessels in a confined space due to the possibility of brisk blood loss.

At the end of the surgery, the decision must be made whether to extubate. Factors that should be taken into consideration include: possible large fluid shifts, atelectatic lung and inflammation after surgical manipulation, the presence of a pneumothorax, and splinting from surgical pain. All these factors can contribute to respiratory compromise. An emergent reintubation, possibly by inexperienced hands, may result in esophageal intubation and disruption of the suture lines, or the intubation may yet again prove difficult and lead to cardiac arrest. In our case, the decision was made to take the child to the PICU (pediatric intensive care unit) intubated and allow her to recuperate overnight before extubating her under controlled circumstances on the next day. A thoracic epidural was placed to minimize post-operative pain, avoid splinting, reduce or avoid the need for narcotics, and to optimize the infant's respiratory effort.

The infant was successfully extubated 24 h after the surgery to high-flow nasal cannula and was doing well. On the third day, the operative chest tubes were pulled and a second pneumothorax appeared that caused the child significant respiratory distress. The decision was made to replace a chest tube in interventional radiology. The option to perform this procedure under a general anesthetic with intubation was considered risky because of the previously discussed potential complications of intubation. Also, there was a concern of causing prolonged postoperative intubation for a short procedure. These concerns warranted other anesthetic considerations. The infant still had a very well working epidural that covered the proposed area for chest tube placement. The decision was made to provide surgical anesthesia by bolusing the epidural with 1% Lidocaine. The radiologist was given a small additional dose of 0.5% Lidocaine to cover the immediate operative site, and the infant was given small titrated boluses of Propofol to minimize patient movement but allowed continued spontaneous ventilation. The chest tube was successfully placed; the infant never required any airway manipulation and recovered quickly from her procedure.

#### Conclusion:

The anesthetic management of infants after tracheoesophageal fistula repair requires a thorough knowledge of the potential complications and associated comorbidities and their impact on your anesthetic. Long term sequela of TEF repairs includes GERD, aspiration pneumonia, tracheomalacia, esophageal and tracheal stricture and respiratory compromise. The anesthesiologist must also be prepared to handle the less common but more complicated tracheal diverticulum and esophageal rupture and be comfortable with the difficult airway algorithm. Management of these patients requires significant vigilance and flexibility.

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