

PBLD – Table #4

Loss of End-tidal CO₂ during repair of Tracheoesophageal Fistula: Things never happen the same way twice.

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Objectives:

1. Review the anatomy of tracheoesophageal fistula (TEF) and its associated syndromes.
2. Review the preoperative evaluation and preparation of TEF repair in neonates.
3. Discuss the anesthetic induction and intraoperative management in neonates undergoing TEF repair.
4. Discuss the etiology and management of difficult ventilation and loss of end-tidal carbon dioxide during surgical repair of TEF.

Case history:

A 2 day old, full term, 3.5 kg baby presents with increased salivation and choking after feeding. Chest x-ray demonstrates coiling of a suction catheter at the level of the neck. He is stable on 0.5 L oxygen by nasal cannula with a SpO₂ saturation of 98%. The patient is scheduled for thoracotomy with TEF repair.

Questions:

What are the different types of TEF and esophageal atresia (EA)? What other associated anomalies should be evaluated in a newborn with EA/TEF? What preoperative studies would be required before surgery? What are the options for surgical timing in isolated EA/TEF?

Case history and physical examination:

The patient was delivered by uncomplicated vaginal delivery. His physical exam at birth was normal. At one hour of life, excessive secretions and respiratory distress were noted, and the infant was transferred to the NICU. Attempt to pass a NG tube was not possible; a CXR demonstrated it coiled in the esophagus. He was subsequently transferred to your hospital for management of suspected EA/TEF. Prior to transport a PIV was placed with D10W infusion. During transport, his oxygen saturation dropped to the 80's and HR dropped to 70s. His nasal cannula oxygen flow was increased to 1L. His vital signs improved and the remainder of his transport was uneventful.

Upon arrival to NICU, the patient had an oxygen saturation of 98%, and was normothermic and euglycemic. His NG was on continuous suction. A PICC line was placed on his left brachial vein for TPN. The patient had a CXR with the following findings:

An esophageal stump is seen projecting over a proximal air-filled structure likely a distended esophageal pouch. Air is visualized in the bowel loops. This could suggest a proximal esophageal atresia with a distal tracheoesophageal fistula.

Pelvis X-Ray: Normal radiographic examination of the pelvis.

Renal U/S: Normal renal ultrasound including the bladder.

Spine U/S: Incidental note is made of a tiny filar cyst, a normal variant, otherwise normal infant spine ultrasound.

ECHO:

- 1) Small patent ductus arteriosus, left-to-right shunt with a peak gradient 21 mmHg
- 2) Patent foramen ovale, left-to-right shunt
- 3) Normal chamber sizes and biventricular systolic performance
- 4) Left sided aortic arch

The surgeon schedules surgery for a suspected C-type TEF as an add-on the following day, or on Saturday depending on the OR schedule. The patient is typed and cross-matched, and preoperative antibiotics are ordered. ENT is consulted to perform a ML&B to evaluate the fistula at the time of TEF ligation.

Questions:

What do you think about the surgeon's plan? Do you need invasive monitoring? Is an arterial line necessary? How will you induce anesthesia? What about postoperative pain control? Is regional anesthesia appropriate? If so, when and how will you perform the block? Is rigid bronchoscopy prior to intubation an appropriate request? How would you manage anesthesia and ventilation in this patient during bronchoscopy?

Intraoperative care:

An uneventful MLB is performed by ENT with the following findings:

- 1) The laryngeal exposure was easy and grade I, and the supraglottis was normal;
- 2) the glottis was normal;
- 3) bronchoscopy demonstrated a normal subglottis;
- 4) the trachea demonstrated a TEF less than 1 cm above the carina ;
- 5) the airway was sized with a 3.0 endotracheal tube with no leak at 25 cm water.

Questions:

How will you place the endotracheal tube and verify position? How will you manage ventilation if you cannot prevent gastric insufflation? What are the pros and cons of intentional main stem intubation in this patient? What are your thoughts on using a Fogarty catheter? Will you use a cuffed or un-cuffed endotracheal tube?

Intraoperative care (continued):

Following intubation, confirmed by the presence of end-tidal CO₂ and auscultation, the patient's SpO₂ is stable. He is placed in the right lateral decubitus position, then prepped and draped. Surgery begins, but initial lung retraction results in his SpO₂ dropping to 80%. The inspired concentration of oxygen is increased, and with hand ventilation the SpO₂ can be maintained in the low 90's.

Question:

How would you manage desaturation during lung retraction?

After the TEF site is identified and ligated, attention is turned to the upper pouch. The pouch is dissected with difficulty due to the distance between the upper and the lower pouches. Suddenly the end-tidal CO₂ waveform disappears and the SpO₂ drops to 85%.

Question:

What is the differential diagnosis? How will you manage this situation? Will you call a code? At your hospital, who responds to an inter-operative code? What equipment will you need in this situation?

The patient is stabilized and surgery progresses with an end-to-end esophageal anastomosis. A chest tube is placed, the thoracotomy incision is closed, and the drapes are removed. While turning the patient supine, his end-tidal CO₂ waveform disappears again. The patient is difficult to ventilate and minimal chest movement is noted.

Question:

What is happening? How will you refine your diagnosis? How will you manage the patient? After correcting the problem, will you extubate the patient or will you keep him intubated?

The patient is then transferred back to the NICU. Due to the surgical tension on the esophagus the patient remained intubated and sedated. He was subsequently extubated on post-op day 3.

Discussion:

Tracheoesophageal fistula (TEF) and associated esophageal atresia (EA) occurs in 1 out of every 2500-3000 live births and is described by two major anatomic classification systems: Gross and Vogt (Figure 1). The most common type is a blind-ending upper esophageal pouch with a distal fistula (type C/IIIb). Co-existing congenital anomalies occur in 50% of patients with EA/TEF (Table 1), more commonly in isolated EA (65%) than in isolated TEF (10%).

Figure 1: Anatomic classification and incidence

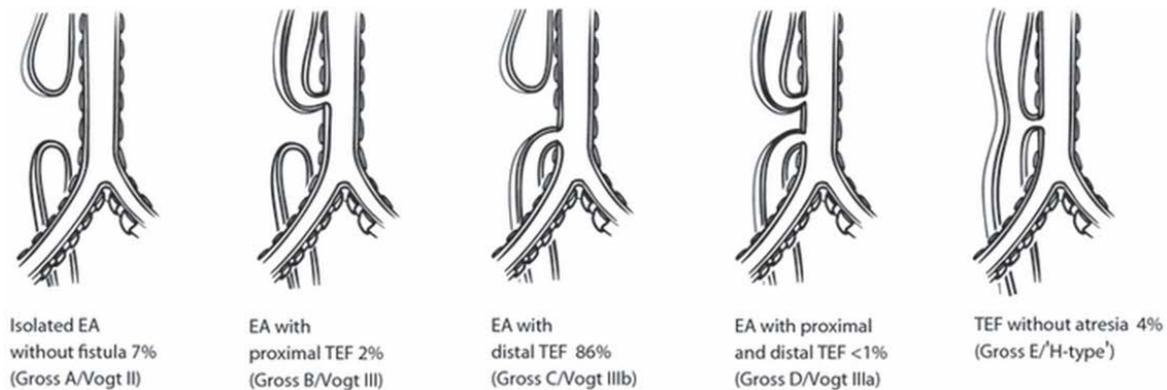


Table 1: Congenital abnormalities associated with EA/TEF

Organ System	Incidence (%)	Examples
Cardiac	29	VSD, PDA, tetralogy of Fallot, ASD, right-sided aortic arch
Gastrointestinal	14	Duodenal atresia, imperforate anus, malrotation, pyloric stenosis, omphalocele
Genitourinary	14	Renal agenesis, hypospadias, horseshoe/polycystic kidney, ureteric/urethral abnormalities
Musculoskeletal	10	Radial limb abnormalities, polydactyly, lower limb defects, hemivertebrae, rib defects, scoliosis
VATER syndrome (VACTERL)	10	Vertebral, anorectal, tracheoesophageal, renal or radial anomalies (expanded to include cardiac and limb defects) other associated syndromes include: CHARGE (coloboma, heart defects, atresia choanal, retarded growth and development, genital hypoplasia and ear deformities), Potter's (pulmonary hypoplasia, bilateral renal agenesis, characteristic facies of intrauterine compression), SCHISIS (omphalocele, cleft lip and/or palate, genital hypoplasia)
Respiratory	6	Tracheo-bronchomalacia, pulmonary hypoplasia, tracheal agenesis/stenosis, tracheal upper pouch
Genetic	4	Trisomy 21, Trisomy 18, 13q deletion

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

TEF is difficult to diagnose prenatally; it may be associated with polyhydramnios and an absent or small stomach bubble. Clinical presentation following delivery includes excessive salivation, choking/cyanosis with feeds, and the inability to pass a suction catheter into the stomach. An X-ray of the thorax and abdomen is required to determine if there is gas below the diaphragm; the presence of abdominal gas indicates a fistula.

All neonates with TEF/EA should undergo preoperative echocardiogram due to the high prevalence of congenital heart disease. Delayed diagnosis of TEF contributes to the development of pneumonia. Aspiration can be minimized by continuous suction of the upper esophageal pouch.

The surgical management of TEF includes right thoracotomy or thoracoscopic primary repair. Rigid bronchoscopy is performed to characterize airway anatomy and to assist in perioperative decision making. Staged repair with preoperative gastrostomy is sometimes performed in the case of long-gap atresia (greater than 2.5 cm or 2-6 vertebral bodies between upper and lower esophagus), or for very low birth weight and severely ill neonates. The presence of a birth weight less than 2 kg and congenital heart disease reduce survival rates from nearly 100% to 27%.

Traditionally, the maintenance of spontaneous ventilation and avoidance of gastric distension from positive pressure ventilation are advocated. Due to potential of two fistulas and complications from an unrecognized, unrepaired fistula, many surgeons begin with rigid bronchoscopy to define the fistula anatomy, prior to surgical repair. Rigid bronchoscopy can identify the size, location and number of fistulas, as well as concomitant airways anomalies including tracheo-bronchomalacia, tracheal agenesis or stenosis. These findings may alter the decision to proceed with muscle relaxation. Maintaining spontaneous ventilation during rigid bronchoscopy can be challenging, and the addition of a propofol infusion to an inhalational technique may be helpful. Many pediatric anesthesiologists routinely paralyze these patients following demonstrated adequacy of bag-mask ventilation and position the endotracheal tube (ETT) well between the fistula and carina without compromising ventilation. Patients with poor pulmonary compliance, due to their greater risk of gastric insufflation, should be approached conservatively. Other airway management strategies including one-lung ventilation and occlusion of the fistula with a Fogarty catheter have been successfully used in this setting.

In addition to standard noninvasive monitoring, arterial line placement is indicated in patients with complex congenital heart disease or pulmonary disease, and in patients undergoing thoracoscopic surgery. A caudal catheter advanced to T6-T7 is utilized in some centers for postoperative pain control and to facilitate early extubation from the avoidance of opioids. Post-operative extubation is determined by airway anatomy, respiratory impairment, prematurity, and associated anomalies. Enteral nutrition typically begins soon after surgery via a trans-anastomotic feeding tube. Re-intubation of infants following repair of TEF should be performed with great caution.

Infants with EA/TEF may go on to have long term medical issues. Esophageal dysmotility commonly occurs following surgery, and GERD is a frequent chronic problem. One third of patients will develop esophageal strictures that may require serial dilatations. Tracheomalacia is also found in this patient population.

In summary, this case illustrates the challenges of peri-operative care in infants requiring surgical repair of TEF.

References:

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