

Tracheoesophageal fistula repair in a premature infant with VACTERL, infradiaphragmatic TAPVR, and pulmonary hypertension.

Moderators: Annette Schure, MD; Lauren Welsh, MD

Objectives:

1. Describe the preoperative evaluation and perioperative management of newborns with trachea-esophageal fistula (TEF).
2. Discuss the preoperative evaluation and perioperative management of newborns with total anomalous venous return (TAPVR).
3. Review timing and management strategies for newborns with coexisting TEF and TAPVR.
4. Describe the role of the anesthesiologist for the coordination of diagnostic and therapeutic procedures among the specialties involved.
5. Discuss the postoperative complications after TAPVR repair and the anesthetic implications for subsequent thoracotomies.

Case Description:

You are called to the NICU to evaluate a 1 day old baby girl with trachea-esophageal fistula for possible repair. The baby had been emergently transferred from another facility. She was born to a 36 year-old G2P1 at 34weeks, via emergency C-section for fetal distress, birth weight 1.4kg. The pregnancy had been closely followed for IUGR and possible anomalies. In the delivery room the baby required positive pressure ventilation, followed by facial CPAP for low saturations and heart rate. APGARS were 2, 7 and 8. Due to increasing respiratory distress the baby was intubated with a 3.0 uncuffed ETT and received 1 dose of surfactant prior to the transfer.

Questions: What do you expect to find? Are there any specific concerns in babies with TEF?

Further investigations reveal that the baby has also an imperforate anus, situs inversus totalis and total anomalous venous return (TAPVR).

Questions: How does this influence your management? What is TAPVR? Do you have any specific concerns?

The echo shows infradiaphragmatic TAPVR with mild obstruction: 4 pulmonary veins form a confluence to the right-sided anatomically left atrium. The confluence drains via a vertical vein into the liver, connecting with a hepatic vein anterior to the IVC/RA junction. There is a mean gradient of 5 mmHG in the vertical vein as it crosses the diaphragm. There is also a large ASD, large PDA, right aortic arch with aberrant left subclavian artery, dilated RV with depressed function, qualitatively good LV function.

The baby is currently intubated, saturating in the mid 90's, hemodynamically stable, without inotropic support. UAC and UVC are in situ. Hct 39%, ABG: 7.27/43/114/19/98%, Lactic acid 8mmol/l

Questions: Do you have any concerns regarding the ventilation? What about the vascular access?

Both, the general surgeon and the cardiologist call you to discuss the case.

Questions: Who else would you like to participate in the discussion? What are your management options? What will you tell the family?

Several hours later, the baby underwent an uneventful ligation of the TEF fistula via a left thoracotomy and placement of a gastrostomy tube. Postoperatively she was transferred to the Cardiac ICU for further management of the TAPVR.

Questions: What are your immediate concerns in the postoperative period? How do you treat them?

On postoperative day 8, she was extubated to CPAP. The pulmonary venous obstruction remained initially stable, then became worse and caused increasing respiratory problems. So at the age of 5 weeks and a weight of 1.52kg, the TAPVR was repaired under deep hypothermic cardiac arrest. The ASD was closed and the PDA ligated. The postoperative course was prolonged and complicated.

Question: What are the typical complications after TAPVR repair?

After several weeks in the Cardiac ICU, followed by a long “feed and grow” period in the medical ICU, she finally presents at the age of 5 months for repair of her trachea-esophageal fistula. Weight 3.4kg.

Questions: How do you evaluate her pre-operative status? What complications do you expect? Are there any specific diagnostic tests that you would like to see?

The last cardiac catheterization a month ago showed no evidence of pulmonary vein stenosis, low to normal LV function (EF 55%), mild RV dilation and qualitatively good function. RVp 1/2 - 2/3rds of systemic pressure. The patient tolerated FiO₂ of 30-35% during the procedure and after changing to 100% O₂, the mean PAP went from 40 to 35 mmHg. After adding Nitric Oxide at 80 ppm, mean PAP went from 35 to 30 mmHg. Contradicting the recent cath results, a CT angio demonstrated pulmonary vein stenosis, especially left middle and lower pulmonary veins, and narrowing of the right pulmonary veins. In addition, the narrow parts of the pulmonary veins are in extremely close proximity to the proximal esophageal pouch. Recent lung perfusion scan reveals 61% of blood to the left lung. And finally, one of your colleagues recalls significant respiratory instability after intubation with desaturation and bradycardia during the cath.

Questions: How does this information influence your management? What do you tell the surgeon? What do you tell the family?

After a long discussion with the general surgeon, the cardiologist and the cardiac surgeon, the decision is made to proceed with the surgery.

Questions: How do you plan your anesthesia? What kind of intraoperative monitoring would you like to use? Would you be able to use intraoperative cardiac echo to monitor right ventricular function? Do you plan to extubate at the end? How do you plan to manage the postoperative pain?

The labs you asked the team to run on Sunday afternoon for tomorrow's first case start come back at 4pm. Arterial Blood Gas results are pH 7.316, pCO₂ 81.2, pO₂ 43.9, bicarb 40 and the chemistry and CBC are within normal limits. The patient remains looking comfortable in the ICU on nasal cannula with a replegole in place.

Questions: Do you still proceed with the surgery? Would you consider electively intubating or starting bipap or CPAP overnight? Would you then repeat the labs in the morning?

Discussion:

Tracheo-esophageal fistula (TEF) and esophageal atresia (EA) are congenital malformations of the foregut which occur with an incidence of about 1:3000 live births. Depending on the location of the fistula and the presence of EA several anatomic variations have been described (Figure 1). The most common (85%) is type C with a blind ending proximal esophageal pouch and a fistula connecting the distal esophagus with the trachea, often just above the carina.

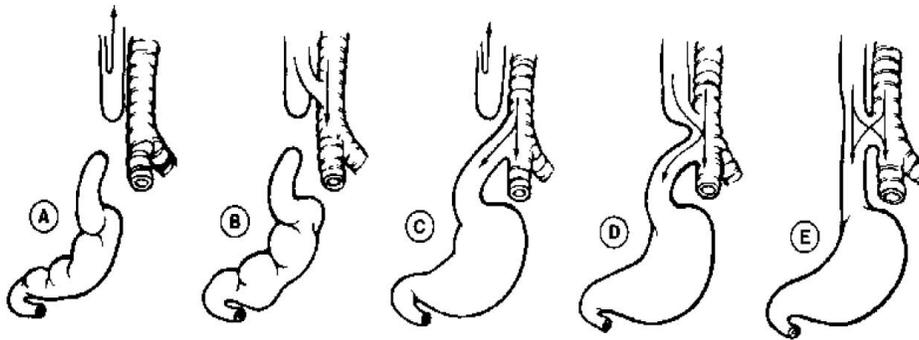
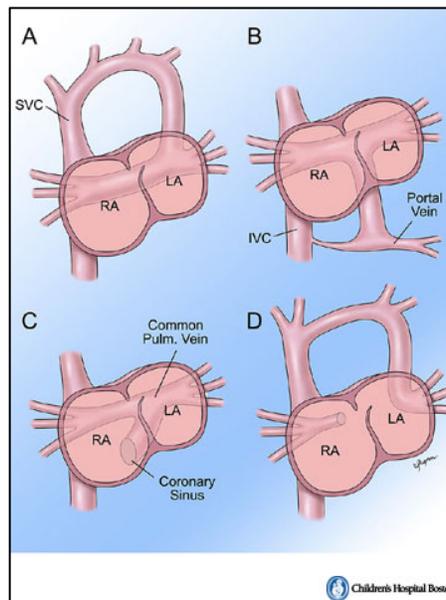


Figure 1: Anatomic variations of TEF/EA. www.waybuilder.net

TEF/EA is often associated with additional congenital defects (20-25% for TEF, 50-70% for isolated EA), particularly vertebral, anal, cardiac, renal and limb anomalies (VATER/VACTERL association) which can complicate the management significantly. Prenatal diagnosis is difficult because the most common ultrasound findings (polyhydramnios and small or absent gastric air bubble) are relatively nonspecific. The diagnosis is often made in the delivery room by the inability to advance an orogastric tube, and later confirmed with a CXR. The primary concern is the risk for aspiration of saliva and secretions from the upper pouch but also from the stomach via the fistula. Keeping the baby NPO, nursing in the upright position with intermittent suction of the upper pouch and appropriate treatment of aspiration pneumonia are important considerations prior to any surgical intervention. In newborns who require positive pressure ventilation due to prematurity or lung injury, inadequate gas exchange is another major problem: A significant percentage of the tidal volume can be lost via the fistula and inflate the stomach leading to abdominal distention and further respiratory compromise. Bag mask ventilation or facial CPAP should be avoided and careful positioning of the endotracheal tube with the tip just **beyond** the opening of the fistula is of utmost importance. Several induction techniques have been described, ranging from awake intubation to inhalational induction followed by intubation without the use of muscle relaxants to maintain spontaneous ventilation until the fistula is ligated. Associated anomalies and the preoperative evaluation of the respiratory status will determine if the newborn is able to tolerate a thoracotomy with single lung ventilation for the total repair (open or thoracoscopic) or will require a staged repair with an initial gastrostomy and ligation of the fistula, followed by a definitive repair later in life.

Total anomalous pulmonary venous return (TAPVR) is a description for congenital heart defects where all 4 pulmonary veins fail to connect to the left atrium but drain instead via remnants of the fetal venous system (vertical veins) into the SVC, IVC or RA, resulting in a left-to-right shunt. Obviously an intra-cardiac right-to-left shunt is necessary to maintain cardiac output. There are several anatomic variations of TAPVR (Figure 2): supracardiac ($\approx 40\%$), cardiac ($\approx 25\%$), infracardiac ($\approx 25\%$) and mixed types ($\approx 5\text{-}10\%$), depending on the location of the anomalous connection. TAPVR is an isolated cardiac defect in about 60%, but 1/3 are associated with complex anomalies, especially heterotaxy syndromes. The clinical presentation is often complicated by some degree of pulmonary venous obstruction, particularly in the infracardiac type, leading to significant pulmonary hypertension, poor cardiac output and increasing acidosis. Placement of a “high lying” umbilical venous catheter can further impair venous return. Since the introduction of prostaglandin E₁, TAPVR with severe obstruction is one of the last remaining “true” cardiac surgical emergencies. Peri-operatively, these patients are at risk for a pulmonary hypertensive crises with cardiac decompensation. Timing of the surgical repair depends on the anatomy of the defect and severity of the symptoms. Anastomosis of the pulmonary venous confluence to the left atrium, ligation of the vertical vein and closure of the ASD are done on cardiopulmonary bypass with periods of deep hypothermic arrest or low flow perfusion.



Given the high risk for this premature, IUGR newborn with a significant congenital heart defect, a preoperative team discussion (neonatologist, cardiologist, general surgeon, cardiac surgeon and pediatric (cardiac) anesthesiologist) with a careful risk/benefit analysis of all the management options is extremely important. As best as possible, the parents should be fully informed and included in the decision making. Based on birth weight and co-existence of major cardiac defects, 3 risk categories have been described:

1. Birth weight > 1500g, no major cardiac defect: 98% survival
2. Birth weight < 1500g or major cardiac defect: 82% survival
3. Birth weight < 1500g **and** major cardiac defect: 50% survival

The following treatment options could be discussed:

- ECMO for temporary stabilization in case of severe hemodynamic and respiratory instability
- Stenting of obstructed veins in the cardiac cath lab
- Staged repair of TEF with initial ligation of fistula, followed by final repair later in life
- Full repair with ligation of fistula and end-to end anastomosis of esophagus

After ligation of the fistula and gastrostomy the primary concerns are the respiratory status (premature lungs, possible tracheomalacia, aspiration risk, post thoracotomy lung injury etc) and the potential for worsening of the pulmonary venous obstruction with increased risk for pulmonary hypertensive crisis and cardiac failure. Adequate postoperative pain control is also important.

The postoperative period after TAPVR repair is often complicated by persistent pulmonary hypertension (PHT), sudden pulmonary hypertensive crisis and prolonged periods of low cardiac output. In order to minimize the risk for PHT, many centers treat these patients prophylactically with high dose opioids, neuromuscular blockade, mild hyperventilation with high FiO₂ and aggressive correction of metabolic acidosis. Echocardiography is an invaluable tool to rule out residual pulmonary venous obstruction and assess ventricular function. Pulmonary hypertensive crises often require inhaled nitric oxide and additional inotropic support. In cases of postcardiotomy syndrome with low systemic vascular resistance the successful use of vasopressin has been described. In animal models and adult case series vasopressin has been shown to be a selective pulmonary vasodilator and systemic vasoconstrictor.

The preoperative evaluation for the second stage of a TEF repair several months after TAPVR repair should clearly focus on the possibility of postoperative pulmonary venous obstruction, a classic complication that occurs in about 15% of patients. In addition to the history and physical exam looking specifically at weight gain, respiratory and hemodynamic status, medications and possible effects on electrolyte balance; several diagnostic studies are useful: Echocardiography to assess pulmonary vein obstruction, pulmonary hypertension and ventricular function, CT Angiography to delineate exact anatomy of pulmonary veins in relation to the esophageal pouch, a lung perfusion scan to specify differences in lung perfusion and possibly cardiac catheterization to evaluate and treat pulmonary vein stenosis.

Once again, a thorough preoperative discussion with the entire team followed by a meeting with the family is extremely important to review the timing and extent of the procedure, clarify treatment goals and debate potential turning points, e.g. the need to abort the surgery.

References:

1. Brett C, Davis PJ: Chapter 18: Anesthesia for General Surgery in the Neonate. In: Davis PJ, Cladis FP, Motoyama EK, eds. *Smith's Anesthesia for Infants and Children*. 8th ed. Elsevier Mosby 2011, p. 574-579.
2. Knottenbelt G, Skinner A, Seefelder C.: Tracheo-oesophageal fistula (TOF) and oesophageal atresia (OA). *Best Pract Res Cl Anaesth* 2010; 24: 387-401
3. Lopez PJ, Keys C, Pierro A: Oesophageal atresia: improved outcome in high-risk groups. *J Pediatr Surg* 2006; 41:331-334.
4. Trinkhaus PM, Hordof AJ, Murphy AM, Greeley WJ: Chapter 32: Total Anomalous Pulmonary Venous Return. In: Nicholas DG, Ungerleider RM, Spevak PJ, Greeley WJ, Cameron DE, Lappe DG, Wetzel RC, eds. *Critical Heart Disease in Infants and Children*. 2nd ed. Elsevier Mosby 2006, p 699-713.
5. Rothman A, Galindo A, Evans WN: Temporary transumbilical stenting of the ductus venosus in neonates with obstructed infra-diaphragmatic total anomalous venous return. *Pediatr Cardiol* 2011; 32:87-90.
6. Friesen RH, Williams GD: Anesthetic Management of children with pulmonary arterial hypertension. *Ped Anesth* 2008; 18: 208-216
7. Scheurer MA, Bradley SM, Atz AM: Vasopressin to attenuate pulmonary hypertension and support systemic blood pressure after correction of obstructed total anomalous pulmonary venous return. *J Thorac Cardiovasc Surg* 2005; 129:465-466.
8. Husain SA, Maldonado E, Rasch D et al: Total anomalous pulmonary venous connection: Factors associated with mortality and recurrent pulmonary venous obstruction. *Ann Thorac Surg* 2012; 94: 825-832.

