# PBLD: Management of tracheoesophageal fistula (TEF) in a 29 week old, extremely low birth weight neonate with Tetralogy of Fallot (TOF)

**Moderators: Alina Bodas, MD and Elizabeth Jansen, DO**

## Objectives

1. Discuss incidence of EA/TEF and outcomes associated with congenital heart disease

2. Discuss respiratory physiology in neonates with emphasis on that seen in an anesthetized neonate with TEF

3. Discuss airway management strategies for isolation of TEF

4. Discuss pathophysiology of TOF and anesthetic management in a neonate with unrepaired TOF

## Case Report

Our patient was an 810 g female born at 29 weeks gestation. Due to respiratory failure, she was intubated within the first 90 seconds of life and transferred to the NICU. TOF had been diagnosed on prenatal ultrasound and she was found to have EA/TEF (Type C) shortly after birth. A Replogle tube was placed to decompress the esophageal pouch and surgical consultation obtained. The patient maintained stable hemodynamics without pharmacologic support. Her $SpO_2$ on 40% $FiO_2$ was consistently in the 80s and 90s.

## Questions

*What is the incidence of EA/TEF?*
*What is the impact of concurrent CHD and low birth weight in these patients?*

## Case Report (continued)

A staged surgical procedure was planned due to the patient's comorbidities. The goal for the first procedure was to place a gastrostomy tube and isolate the fistula by inserting a catheter retrograde and then ligate the fistula via open thoracotomy. Once the patient had grown, esophageal anastomosis would occur in a second stage procedure. General anesthesia was achieved with sevoflurane via a 2.5 uncuffed oral ETT and neuromuscular blockade with rocuronium. During retrograde fistula intubation with a Fogarty catheter via surgically placed gastrostomy, there were several desaturation episodes requiring 100% oxygen, 5% albumin, fentanyl, and phenylephrine boluses. Due to prolonged periods of desaturation in the initial phase of the procedure, it was determined that the patient would not tolerate an open thoracotomy. The decision was made to delay ligation. The gastrostomy tube and Fogarty catheter were sutured in place to improve ventilation and minimize aspiration. She remained hemodynamically stable and was extubated to CPAP on POD #4.

**Questions**
*What approaches exist for isolation of the fistula and airway management? How might they be affected by respiratory physiology in a neonate?*
*What is the etiology of hypoxia seen in infants with TOF? How are episodes of hypoxia or "Tet spells" managed?*

**Case Report (continued)**

On day-of-life 25, weighing 1.2 kg, the patient returned to the OR after serial CXRs revealed that the Fogarty catheter had dislodged and ventilation was becoming more problematic. The plan was to ligate the fistula via right thoracotomy. Spontaneous ventilation was maintained with a 0.4 mcg/kg/hr dexmedetomidine infusion as a 4Fr Fogarty catheter was placed into the fistula via an antegrade approach using fluoroscopic guidance. The patient was intubated with a 3.0 uncuffed ETT adjacent to the Fogarty catheter. The patient had several desaturation episodes which were treated with small boluses of albumin and phenylephrine. The ligation was successfully completed and the patient was transported to NICU where she remains on ventilator support.

**Questions**
*Describe techniques for airway management using a Fogarty catheter for TEF isolation.*
*Why was it important to maintain spontaneous ventilation throughout airway management?*
*What are some advantages of this approach?*

**PBLD: Management of tracheoesophageal fistula (TEF) in a 29 week old, extremely low birth weight neonate with Tetralogy of Fallot (TOF)**

## Discussion

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) occurs in approximately 1 in 3000 to 1 in 4500 live births.[1] Advancements in neonatology, anesthesiology and surgery have allowed for a shift from mortality rates of 70% in the 1940s to current survival rates over 90%.[1] Studies have identified birth weight <1500 g and concurrent congenital heart disease as predictors of increased morbidity and mortality in these patients.[1] Diaz et al, demonstrated that CHD in infants with TEF/EA conferred a perioperative mortality rate of 23% compared with no mortality in infants with EA/TEF in the absence of CHD.[1]

Various airway management techniques have been utilized for TEF repair, including mainstem intubation with one-lung ventilation, intubation distal to the fistula with ETT above the carina for two-lung ventilation, occlusion of the fistula from below with two-lung ventilation and occlusion of the fistula proximally with a Fogarty catheter allowing for two lung ventilation.[2]

Pulmonary factors that may complicate anesthetic management include poor lung compliance and respiratory distress syndrome associated with prematurity.[3] Approximately 30% of neonates with EA/TEF are premature.[3] Furthermore, pneumonitis caused by reflux of gastric contents into the lungs may contribute to pulmonary complications. EA/TEF is often seen in conjunction with other anomalies and tracheo-bronchomalacia, pulmonary hypoplasia, tracheal agenesis/stenosis or tracheal upper pouch may pose additional challenges.[3]

Neonates and infants have highly compliant chest walls and lungs (with minimal elastic recoil) secondary to increased cartilaginous content and underdeveloped elastic fibers.[4] These factors make it more difficult for neonates to maintain FRC.[4] Mechanisms to preserve FRC include glottic closure or laryngeal braking during expiration, diaphragmatic braking to minimize diaphragmatic activity in expiration and tonic contractions of the diaphragm and intercostals to stiffen the chest wall.[4] These compensatory mechanisms are lost under general anesthesia with muscle relaxation and make it difficult to maintain FRC in an anesthetized neonate.[4] Furthermore, increased airway resistance and closing volumes add to potential ventilatory difficulty in this group.[4]

Tetralogy of Fallot is the most common form of cyanotic CHD, and cyanosis from increased right to left shunting, also known as a “tet spell”, presents a significant challenge in anesthetic management.[5] The severity of the tet spell depends on the degree of RVOT obstruction, often caused by decreased SVR or pulmonary infundibular spasm.[5] Prompt recognition and treatment of these events helps improve efficacy and prevent further hemodynamic compromise. The goal of treatment is relief of infundibular spasm and reversal of right to left shunting.[5] This can be accomplished by increasing $FiO_2$ to

100%, abdominal compression to increase SVR, increasing preload with an IV fluid bolus, increasing SVR with phenylephrine, increasing anesthetic depth with fentanyl and esmolol to relieve infundibular spasm.[5]

In 1982, Filston published a case report describing isolation of TEF in a 1765 g premature neonate using a 4 Fr Fogarty catheter.[2] The neonate was kept breathing spontaneously as a Storz bronchoscope was used to intubate the fistula and then introduce a Fogarty catheter to occlude it.[2] The patient was then intubated following Fogarty placement.[2] The Fogarty balloon was inflated with contrast dye and x-ray utilized to confirm placement.[2] When the endotracheal tube was properly positioned, the infant could be successfully ventilated without continuous inflation of the Fogarty balloon, which reduced risk of pressure necrosis.[2] Inflation of the balloon successfully occluded the fistula and prevented escape of ventilatory gases into the low pressure gastrostomy.[2] Spontaneous ventilation was emphasized in this case because the TEF did not pose a significant ventilatory problem when the neonate was breathing spontaneously.[2] On the contrary, controlled or assisted ventilation causes major ventilatory impairment as the TEF became a low pressure vent that compromised ventilation to the lungs.[2]

**References**


1. Diaz LK et al. *Tracheoesophageal fistula and associated congenital heart disease: implications for anesthetic management and survival*. Paediatr Anaesth. 2005 Oct;15(10):862-9.

2. Filston HC et al. *The Fogarty balloon catheter as an Aid to management of the infant with esophageal atresia and tracheoesophageal fistula complicated by severe RDS or pneumonia.* J Pediatr Surg. 1982 Apr;17(2):149-51.

3. Broemling N and Campbell F. *Anesthetic management of congenital tracheoesophageal fistula.* Pediatric Anesthesia. 2011 (21) 1092-1099.

4. Davis PJ et al. *Smith's Anesthesia for Infants and Children.* Chapter 3: Respiratory Physiology in Infants and Children. 2011 43-49.

5. Twite MD et al. *Tetralogy of Fallot: Perioperative Anesthetic Management of Children and Adults.* Semin Cardiothorac Vasc Anesth. 2012 Jun;16(2):97-105.