Thoracoscopic repair of Tracheo-esophageal fistula in a newborn with tetralogy of Fallot and pulmonary atresia

Nicole Dobija, MD, DDS
Assistant Professor, Anesthesiology and Pediatrics
&
Michael A. Arnold, D.O.
Chief Resident and cardiac fellow
The University of Florida, Gainesville, Florida

Objectives:

- Understand the anatomy and physiology of tetralogy of Fallot with pulmonary atresia (TOF c PA).
- Understand the timing and order of surgical repair for patients with tracheoesophageal fistula (TEF) with TOF c PA.
- Understand the anesthetic considerations for the repair of a TEF in a patient with TOF c PA, both open and thoracoscopically.
- Understand the physiologic effects of thoracoscopy and how this affects the patient with single ventricle physiology.
- Evaluate methods of intraoperative and postoperative pain management in this patient population.

Case History:

A baby girl was born at 40 weeks to a 30 year old mother with no pregnancy related complications. Her Apgars were 8 and 9. She was intubated approximately three hours after delivery for respiratory distress and transferred to the level III NICU at our institution. On echocardiography, she was noted to have tetralogy of Fallot with pulmonary atresia. The pediatric surgery service was also consulted for management of her esophageal atresia, tracheoesophageal fistula, and imperforate anus.

Questions:

1. What is tetralogy of Fallot with pulmonary atresia and how does it differ from other types of tetralogy of Fallot?
2. How does the presence of a tracheoesophageal fistula complicate matters?
3. Is there an association between tetralogy of Fallot and tracheoesophageal fistula?

Preoperative studies:

A chest x-ray shows an orogastric tube in the mid-esophagus which cannot be advanced any further. Diffuse air is noted in the bowel. The transthoracic echocardiogram findings are noted above. The infant is completely ductal
dependent and has no other systemic to pulmonary shunts present. She also has a persistent left superior vena cava, draining directly into the coronary sinus. She is passing meconium through a presumed recto-vestibular fistula. Her oxygen saturations are mostly in the mid to high 80s. She is on prostaglandin to maintain ductal flow. She is also receiving TPN and lipids.

Questions:
1. Are there any additional studies warranted in a newborn with the above abnormalities?
2. If so, what additional tests/studies should be performed prior to surgery?
3. Which problem should be corrected first and why?
4. Are oxygen saturations in the 80's concerning?

Case Progression:
Extensive discussion took place between the Pediatric Surgery team and the Congenital Heart team as to which surgical repair should take place. It was decided that since the patient was stable on her prostaglandin that the Pediatric Surgery team would repair her tracheoesophageal fistula and imperforate anus. They also planned to place a Broviac catheter to allow for central access for the impending surgical repair of her congenital heart disease. The Pediatric Surgery team elected to repair the tracheoesophageal fistula thoracoscopically.

Questions:
1. In the pediatric population, are there specific concerns regarding thoracoscopic surgery and minimally invasive surgery in general?
2. What are the potential benefits of minimally invasive surgery in the pediatric population?
3. What are the options for achieving lung isolation in the pediatric population?
4. What, if any, additional monitors would be helpful?

Intraoperative Management:
The patient, now five days old, arrived to the operating room intubated. Induction of anesthesia, as well as maintenance, was achieved with volatile agent and fentanyl. She was chemically paralyzed with rocuronium. The case proceeded uneventfully until insufflation of carbon dioxide. Despite using insufflation pressures of 5 mmHg, the patient became hypotensive. An arterial blood gas showed a respiratory acidosis with a pH of 7.09 and a pCO2 of 79 mmHg. At that point, the thoracoscopic repair was abandoned in favor of an open thoracotomy. The remainder of the surgery was uneventful and included repair of her imperforate anus and placement of the Broviac catheter.

Questions:
1. What are the expected physiologic changes associated with CO2 insufflation for thoracoscopic surgery?
2. How might these expected changes affect a patient with a single functional ventricle?

Postoperative Course:
The patient was returned, intubated, to the NICU. She has some persistent atelectasis on the surgical side. She was extubated on post-operative day 3. She was started on enteral feeds, which she tolerated well. She was stooling appropriately. Her hospital course was uneventful and she returned to the operating room 3 weeks after her initial surgery for a complete repair of tetralogy of Fallot.

Discussion:
Minimally invasive surgery is increasing in the pediatric population. Complex congenital anomalies, such as tracheoesophageal fistulas (TEF), are amenable to endoscopically assisted techniques. The Annals of Surgery in 2005, showed similar results between open and thoracoscopic repair of tracheoesophageal fistula. A similar retrospective study in the Journal of Laparoendoscopic and Advanced Surgical Techniques showed equivalent outcomes between open and minimally invasive techniques.

There are limited reports of thoracoscopic repair of TEF in patients with cardiac abnormalities. The October 2005 issue of Anesthesia and Analgesia outlines the successful thoracoscopic repair of a TEF in a neonate with tricuspid atresia. Single ventricle pathology presents a unique challenge to procedures dependent on carbon dioxide insufflation. The deleterious effects on the pulmonary vasculature can disrupt the balance between pulmonary and systemic blood flow. Hypoxia can occur during single lung ventilation and further exacerbate this imbalance. The addition of single ventricle physiology can complicate this surgery and requires special preparation and attention to anesthetic technique.

References:


