

Right Lung Volvulus after Tracheoesophageal Fistula Repair

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Introduction: Tracheoesophageal Fistula (TEF) is a rare congenital anomaly associated with a high risk of aspiration and other congenital disorders. We present the case of a patient who underwent primary TEF repair hours after birth who subsequently developed continued pulmonary failure due to a right lung volvulus and consequent right upper lobe infarction.

Case Report: A 16 day-old male born at 35 weeks EGA presented to the OR emergently for a right thoracotomy s/p primary TEF Repair and Colostomy on the day of birth. The neonate was diagnosed at birth with a distal TEF, esophageal atresia, and an imperforate anus and was subsequently taken to the OR for repair. At the conclusion of the uncomplicated primary repair, the neonate was transported intubated to the NICU with a thoracic epidural. The epidural catheter was removed on postoperative day 1, because an epidurogram showed cephalad dispersment of local anesthesia to the cervical spine. Over the following two weeks, the neonate developed progressive pulmonary failure leading to high frequency oscillatory ventilation (HFOV) to facilitate adequate oxygenation. A chest CT scan on day of life (DOL) 15 showed obstruction of the right upper and middle lobes with a possible volvulus near the location of the primary TEF repair and interstitial emphysema of the entire left lung, and therefore the neonate was taken back to the OR for an emergent right thoracotomy while still on HFOV. Anesthesia was initially maintained with fentanyl and pancuronium. A head ultrasound prior to the OR showed a possible Grade 1 intraventricular hemorrhage (IVH), but the decision was made collectively to place the neonate on extra-corporeal membrane oxygenation (ECMO) in order to safely oxygenate the patient during the thoracotomy. Anticoagulation with heparin was therefore administered as the surgeons proceeded with cannulation for ECMO via the right internal jugular vein and the right common carotid artery. Adequate flows were easily achieved on VA-ECMO in the supine position and HFOV was discontinued, but continuous positive airway pressure (CPAP) was provided via a conventional ICU ventilator. Decreased VA-ECMO flow rates in the left lateral decubitus position responded well to colloid boluses, and the mean arterial pressure (MAP) was maintained ≤ 60 mmHg throughout the surgery. A right thoracotomy was performed at the fifth intercostal space where a nonviable, consolidated, and infarcted right upper lobe was discovered. Despite adhesiolysis, the lobe remained underperfused and a right upper lobectomy had to be performed. The chest was closed and the patient was taken back to the NICU on ECMO.

Discussion: The morbidity and mortality s/p primary TEF repair is high in patients with multiple other congenital anomalies; however, in patients with isolated TEF and EA, the one-year survival can be greater than 95%. Postoperative complications such as stricture and leak can occur in 10-40% of patients. Persistent pulmonary failure can also occur, with potential causes including pneumonia, atelectasis, or another proximal TEF. In this case, right lung volvulus was the cause of persistent postoperative pulmonary failure. Lung volvulus is an extremely rare and unusual postoperative complication of a primary TEF repair which proved difficult to diagnose. Initially, radiographic evidence suggested that our patient's pulmonary failure was due to either pneumonia or atelectasis, and fluoroscopic contrast studies confirmed the absence of an anastomotic leak. When our patient's condition did not improve despite HFOV, we were forced to consider other causes of persistent pulmonary failure. In this case, a CT scan with three-dimensional reconstruction was able to detect pathology that led to surgical exploration. A right thoracotomy revealed that the right upper lobe was infarcted requiring lobectomy. Perhaps an earlier diagnosis, by consideration of this unique pathology,

may have avoided an unnecessary lobectomy in our patient and decreased the duration of mechanical ventilation and its associated risks. Prior to the exploratory thoracotomy, we chose to employ VA-ECMO with anticoagulation early. Despite the presence of a Grade 1 IVH by cerebral ultrasound preoperatively and the known risks of cerebral hemorrhage with anticoagulation and ECMO initiation, our patient's ventilatory requirements were too excessive to oxygenate our patient by any other means. After discussion with the surgical team, we felt that early VA-ECMO cannulation and initiation was our only option to provide adequate oxygenation during the perioperative period. Early ECMO also provided the added benefit of postoperative pulmonary rest after two weeks of aggressive and escalating ventilatory support.

Conclusion: TEF and EA repair is not without associated risks such as persistent postoperative pulmonary failure. Lung volvulus is another potential and serious cause of respiratory failure that can be difficult to diagnose. With early consideration, high suspicion, and adequate imaging such as CT scan with three-dimensional reconstruction, earlier surgical intervention may be possible. This may help to avoid unnecessary pulmonary resection and reduce the duration of mechanical ventilation. ECMO initiation may be the only option for oxygenation in a patient with pulmonary complications after undergoing primary TEF and EA repair.

Refs:

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2. Kovesi T, *et al.* Chest 2004; 126; 915-925