A 5-month-old with un palliated Tetralogy of Fallot for bronchoscopy and cardiac catheterization after an Apparent Life Threatening Event

Walker, K Karisa MD - Kandiah, Nishanthi MD
Yale-New Haven Hospital, Yale University School of Medicine

CASE BACKGROUND

FM is a 5-month-old female born at 36 weeks with neonatal abstinence syndrome, who also suffers from Tetralogy of Fallot (ToF) (with a large VSD and minimal right-ventricular outflow obstruction (RVOTO)) and has a history of wheezing unresponsive to albuterol. She had maintained on 5mg furosemide daily for pulmonary overcirculation. She presented to our clinic for pre-operative anesthetic evaluation for repair of her ToF. Upon examination, the infant became upset suffered a spell of prolonged cyanosis and was transported immediately to the emergency department, where she was found to be profoundly hypoxic to 35%. SpO2, which resolved with O2 administration, morphine, and knee-to-chest position. She was started on prednisone and bronchodilators and ehaled on 5mg furosemide titrated off over the following month. Her 6 month medical evaluation revealed an Apparent Life Threatening Event (ALTE) with spontaneous ventilation and the patient was discharged home by post operative day five.

ANESTHETIC TECHNIQUE

This patient presented with ToF and episodes of cyanosis without RVOTO. Preoperatively the patient was treated for pulmonary overcirculation so we suspected chronic lung disease. We anticipated that any increase in PVR could reverse the intracardiac shunt and cause hypoxia. Bronchoscopy would likely involve oxygen deprivation, hypoventilation, and bronchoscopy, all of which would increase PVR. We maintained preload with IV hydration and used a general-endotracheal anesthetic with muscle relaxation to prevent movement and coughing. Midazolam and nitric oxide were readily available to reduce PVR, though ultimately this was not necessary. We kept the patient sedated and intubated post-procedure for transport to the ICU to maintain her airway.

BRONCHOSCOPY AND CARDIAC CATHETERIZATION FINDINGS

A right tracheal bronchus (bronchus sui) was noted, along with severe left bronchial collapse. Oxygen saturation was sustained above 92% throughout. Cardiac catheterization was performed on both room air and 100% FiO2. Qp/Qs was calculated to be 1.6:1 on room air and 3:1 on 100% FiO2. Additional findings included a large anterior-sub-aortic VSD with significant left-to-right shunt and enlarged LA/main pulmonary artery compressing the left mainstem bronchus. There was no evidence of pulmonary hypertension.

IMAGING

SURGICAL AND MEDICAL INTERVENTIONS

Five days later, FM underwent uncomplicated repair of her VSD and LPA imbrication. She was extubated to 100%-non-rebreather on post-operative day one, complicated by desaturation requiring frequent chest PT and bronchodilator treatments, as was expected given abnormal airway anatomy. Oxygen therapy was titrated down to room air and the patient was discharged home by post-operative day five. After discharge FM was admitted two more times with ALTEs. Awake airway floroscopy with spontaneous ventilation revealed that she additionally suffers from tracheomalacia. Echocardiography demonstrated a small residual VSD. She was started on 1L O2, which was titrated off over the following month. Her 6-month postoperative echocardiogram showed no residual VSD.

DISCUSSION

Although we initially suspected chronic lung disease and pulmonary hypertension, this child, in fact, had neither. Her condition, bronchus sui, is so named because this is the normal anatomy of a pig. This airway anomaly is found in 0.1% of the population and is often associated with frequent infection, bronchiectasis, and tracheobronchomalacia. As previously mentioned, severe tracheobronchomalacia was eventually diagnosed.

The mechanism of this patient’s cyanosis was then attributable to collapse of the upper airways, leading to hypoxia, increased PVR, shunt reversal and resultant profound hypoxia. The fact that the patient was maintained on beta-agonist bronchodilators for presumed airway reactivity may have paradoxically worsened her condition by relaxing bronchial smooth muscle. Indeed, these episodes began after the institution of albuterol therapy. Also, since part of the left bronchial compression was due to enlarged LPA, the intraoperative use of NO may have precipitated hypoxia had we used it for presumed pulmonary hypertension. When caring for children with incompletely diagnosed disease, it is important to fully consider all possible underlying diagnoses and the potential complications of the associated treatments. The treatment of one presumed condition, if the presumed diagnosis is incorrect, may worsen the patient’s true pathologic. FM is now nearly one year removed from her surgical intervention and is doing well at home with her parents.

REFERENCES