Unusual Cardiac and Airway Anatomy in a Patient with TOF/PA/MAPCAs

Ana Cox MD¹, Lisa Wise-Faberowski MD MS¹, Glyn Williams MD¹, Andrew Shin MD², Doug Sidell MD³, Katsuide Maeda MD⁴

Departments of Pediatric Anesthesiology¹, Cardiology², Otolaryngology³, and Cardiothoracic Surgery⁴, Stanford University School of Medicine, California, USA

Introduction:
• 5 day old, ex 35 3/7 week, male infant presented with a history of severe hypoxemia after birth requiring intubation. High peak airway pressures were required for ventilation (>30mmHg).
• Cardiac MRI revealed distal MPA atresia with tiny residual left PA and five MAPCAs.
• CT angiogram showed small central airways and a hyperinflated right lower lung causing compression of the right atrium and SVC.

OR Course:
• Patient went urgently to the OR for laryngoscopy and bronchoscopy with ENT and pulmonology to further evaluate the airway and pulmonary tree.
• The ECMO team was on standby due to his complex cardiac pathology and possible cardiovascular instability or collapse during airway manipulation.
• Spontaneous ventilation was maintained and albumin was given to prevent further hyperinflation and to improve preload respectively.
• The OR course was uneventful.
• Patient was found to have moderate tracheomalacia with no evidence of tracheal rings, severe right bronchomalacia with complete collapse, and narrow left mainstem.

Discussion:
• Extracardiac anomalies have been estimated to occur in up to 45% of patients with CHD.
• Approximately 4% of patients with CHD have congenital airway anomalies.
• However, patients with with TOF/PA/MAPCAs have been shown to have a much higher incidence of airway anomalies. In one study by Maeda, 44% of patients had bronchomalacia.

Conclusion:
• Involvement of several disciplines was required to further define this patient’s cardiac and airway anatomy.
• Despite having uncomplicated intracardiac anatomy with preserved cardiac function, the large sequestration likely prevented normal airway development.
• The absence of multiple lung segments and hypoplastic distal airways became the limiting factors to surgical palliation, despite our center’s expertise for palliating TOF/PA/MAPCAs.

References: