Uncommon association of Tetralogy of Fallot, Absent Pulmonary valve (APV) and Pierre Robin sequence causing severe airway compromise.

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Introduction

- Tetralogy of Fallot (TOF) with absent pulmonary valve (APV) is an uncommon variant of tetralogy (3-6% of cases) characterized by combined pulmonary stenosis and insufficiency.
- Massive enlargement of the main and branch pulmonary arteries ensues and causes diffuse compression of the tracheobronchial tree.
- Pierre Robin causes upper airway obstruction and difficult laryngeal visualization.
- We present the case of a patient suffering from a combination of these airway compromising defects.

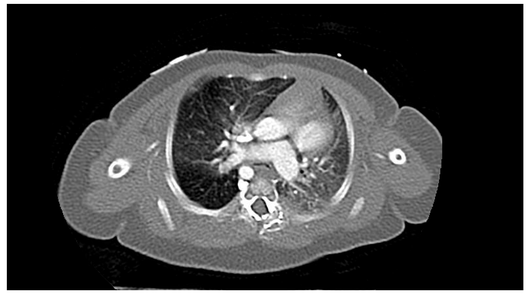
Case History

- A 2-week-old female with both TOF/APV and Pierre Robin Syndrome and acute systemic desaturation required an emergency tracheostomy when laryngeal visualization could not be accomplished by direct laryngoscopy.
- Subsequent flexible laryngoscopy revealed a small glottic opening and a laryngeal web.
- Ten weeks later she presented for laryngeal web resection with the interim progression ventilatory settings shown in the table below.

Vent Sett	POD 0	POD 45	POD 46	POD 49	POD 50	POD 72	POD 72
PEEP	4	8	10	12	15	15	15
PIP	19	22	25	28	30	28-30	18
FIO2	20-30	20-30	20-30	20-30	20-30	20-30	25

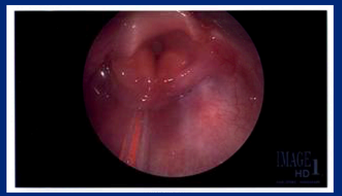
Case History

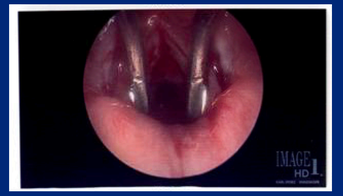
- A CT scan at this time revealed severe enlargement of pulmonary arteries causing complete effacement of the carina.



Intraoperative Course

- Anesthesia was induced with Sevoflurane via tracheostomy.
- Anesthesia was maintained with Sevoflurane and intermittent boluses of short acting opioids.
- Mechanical ventilation parameters were adjusted to optimize mean airway pressure and thus airway patency.
- Hemodynamic parameters remained stable throughout the case without vasoactive infusions.
- Suspension laryngoscopy was performed with visualization of normal supraglottic structures.
- A grade 3 laryngeal web and malacia of the mid-distal trachea and bilateral main stem bronchi were identified.
- Laryngeal-tracheal reconstruction with cartilage grafting was then undertaken.
- Postoperatively the patient was transferred to the cardiac ICU for monitoring and required prolonged mechanical ventilatory support.





Discussion

- This case illustrates the challenging combination of difficult intubation, laryngeal pathology, and intrathoracic airway obstruction.
- The upper airway pathology was successfully managed by surgical interventions.
- The intrathoracic airway obstruction caused by the dilated pulmonary arteries persisted with limited surgical options.
- These patients commonly have pulmonary segments of hyperinflation and lobar atelectasis that are minimally responsive to bronchodilators and pulmonary toilet.
- In these patients, management of ventilation strategy to optimize airway patency is often much more challenging than managing hemodynamics and intracardiac shunting.

References

1. Kauth AL, Max G. AM. Respiratory symptoms secondary to aorto-pulmonary collateral vessels in tetralogy of Fallot with Absent Pulmonary Valve syndrome. Am J Cardiol 2004,93:503-505.
2. Dodge-Khatami A, Mavroudis C. Complete repair of Tetralogy of fallot with Absent pulmonary valve including the roe of airway stenting. J card Surg 1999;14:82-91
3. Szwast A, Rychik J. Anatomic variability and outcome in prenatally diagnosed absent pulmonary valve syndrome. Ann thoracic Surg 2014;98:152-8.

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