Tracheal Stenosis associated with Pulmonary Hypoplasia/Aplasia: Case Series and Literature Review

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Objective/Hypothesis
To describe our experience in the surgical management of Tracheal Stenosis Associated with Pulmonary Hypoplasia/Aplasia

Study Design
Case Series

Methods
A review of our cases with pulmonary hypoplasia/aplasia was undertaken. We further reviewed patients that had tracheal stenosis and underwent operative management. Details of presentations, surgical reports and post-operative management were reviewed

Results
All three patients presented with right lung hypoplasia/aplasia complete tracheal rings and associated tracheal stenosis. Tracheal stenoses in two of the patients were managed by anterior tracheofissure, one with rib graft augmentation and the other with a tracheal autograft. Slide tracheoplasty was performed in the remaining patient. Two of these patients required cardiopulmonary bypass during their surgeries. Subsequent evaluations have revealed normal healing graft sites in the patients who underwent tracheofissures with tracheoplasty. The patient who underwent the slide tracheoplasty has residual distal tracheal stenosis.

Conclusion
Pulmonary Hypoplasia/Aplasia is a rare entity, and can be associated with other airway abnormalities including complete tracheal rings and tracheal stenosis. These patients may be successfully managed surgically, but there are unique ventilatory and aesthetic considerations that must be recognized in each case.

Introduction
Congenital tracheal stenosis (CTS) has been recognized for over 100 years. It is characterized by narrowing of the trachea, and is often associated with complete tracheal rings. Accurate characterization of CTS, as well as identification of associated upper airway, bronchial, pulmonary and cardiac anomalies is paramount to successful management. Pulmonary anomalies such as pulmonary hypoplasia and aplasia can complicate the perioperative and intraoperative management of these patients as their ventilation is based on a single lung. We present three patients managed surgically with congenital tracheal stenosis, associated complete tracheal rings and single lung physiology. We also reviewed the literature on repair or tracheal stenosis pulmonary hypoplasia/aplasia within the last 20 years.

Patient 1
- Presented at 3 months old with respiratory distress
- Bronchoscopy showed, tracheal stenosis 4.3 cm in length
- CT scan showed severe right pulmonary hypoplasia and dextrocardia
- Underwent Autograft tracheoplasty
- At 3 years old required balloon dilation and now continues to thrive at age 5

Patient 2
- Diagnosed with right pulmonary aplasia and dextrocardia (Figure 1)
- Bronchoscopy showed complete tracheal rings 2.7 cm long
- Day of life 50 had transversal augmentation tracheoplasty with costal cartilage graft
- Required aortopexy at age 2 and thriving since

Patient 3
- Tracheal stenosis diagnosed via CT at birth with dextrocardia and right lung aplasia
- Bronchoscopy showed funnel shaped tracheal stenosis
- Recurrent Acute Life-Threatening Events (ALTE’S) through first few months of life
- At 19 months underwent slide tracheoplasty * under cardiopulmonary bypass and has done well since

Figure 1: Coronal Chest Computer Tomography showing Right Pulmonary Aplasia, Dextrocardia and Absence of the Right Mainstem Bronchus

Conclusion
- Tracheal stenosis can be managed by a variety of techniques including tracheoplasty with augmentation or autografts, or slide tracheoplasty
- Cardiopulmonary support is often needed intraoperatively
- Tracheal stenosis can be managed successfully even in cases with pulmonary hypoplasia/aplasia

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