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COMBINED CONGENITAL MALFORMATION OF ORGAN DEVELOPMENT: DIAGNOSIS AND MANAGEMENT

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Introduction

The coexistence of congenital cardiovascular anomalies and major aortic arch malformations represents an extremely rare and complex clinical challenge in pediatric practice. The Aortopulmonary Window (APW) is a rare congenital defect accounting for **less than 0.5%** of all congenital heart disease. Similarly, the aberrant right subclavian artery (*Arteria Lusoria*/ARSA) is the most common aortic arch anomaly (with a population prevalence of 0.5%–1.8%); however, its combination with APW and the resulting severe obstructive and aspiration syndromes in infancy is exceptionally sporadic in medical literature.

Clinical Case

We report the complex management of a pediatric patient who was diagnosed with APW in the neonatal period following necrotizing enterocolitis (NEC). Primary surgical correction was performed at 3 weeks of age, followed by a prolonged postoperative course (1.5 months) requiring mechanical ventilation, tracheostomy, recurrent sepsis, and pneumonia.

Post-discharge, the patient experienced severe recurrent respiratory failure and right-sided pneumonia. Exertion and feeding induced marked tachypnea, diaphoresis, and noisy breathing. The NT-proBNP level was 300 pg/mL. Multimodal imaging via chest CT-angiography revealed an *artery lusoria* exerting extrinsic compression on the esophagus. Barium contrast studies failed to identify the anomaly, whereas upper esophagoscopy definitively demonstrated pulsatile vascular compression nearly obliterating the esophageal lumen, which served as the primary pathophysiological cause of chronic aspiration and recurrent pneumonias. The patient underwent successful surgical intervention—reimplantation of the aberrant vessel into the right common carotid artery. The patient was successfully extubated within 24 hours postoperatively.

Pathophysiology and Epidemiology

While aspiration pneumonias and severe respiratory distress caused by aberrant vessels in infants are frequently compounded by soft tracheal cartilages and tracheobronchomalacia, persistent micro- and macro-aspiration driven by severe esophageal compression dramatically increases the incidence of recurrent pulmonary infections and mortality. APW induces a massive left-to-right shunt and pulmonary hyperperfusion, whereas *Arteria Lusoria* establishes an extrinsic vascular ring compression that disrupts normal swallowing mechanics.

Diagnostic Challenges and Management

Diagnosing such complex anomalies in infants is fraught with difficulties, as standard barium contrast examinations often fail to visualize dynamic obstruction. A multidisciplinary approach is mandatory: alongside **CT angiography**, **high-resolution endoscopic evaluation** plays a definitive diagnostic role. Management requires highly specialized pediatric intensive care and cardiothoracic surgical expertise.

Conclusion

This unique clinical case emphasizes that in cases of refractory respiratory failure and recurrent pneumonia, clinicians must actively screen for rare vascular anomalies (*Arteria Lusoria*) in addition to primary cardiac defects. Early multimodal diagnosis and prompt surgical correction are critical for patient survival and long-term prognosis.

Keywords: APW, ARSA, NEC, Aspiration Pneumonia.