Obstructed TAPVR, single ventricle physiology and single lung with tracheal rings: A fatal constellation

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BACKGROUND

Left ventricular outflow tract obstruction and TAPVR affect a small percentage of children diagnosed with congenital heart defects. Palliation of single ventricle defects depends on the presence of an adequate pulmonary circuit.

CASE

Background: A term 2.54 kg neonate with prenatal diagnosis (Figure 1) of unbalanced AV canal defect and abnormal pulmonary venous return developed bradycardic arrest 2 hours after delivery, concern raised for obstructed pulmonary venous return (Figure 2)

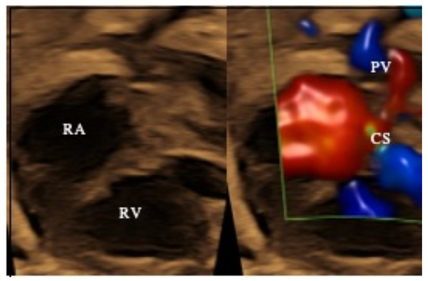

Figure 1: Fetal echo apical 4 chamber with color compare showing large RV, small LV, and at least 1 pulmonary vein entering the coronary sinus

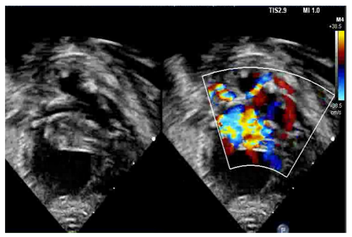

Figure 2: Initial transthoracic echo with concern for tortuous venous connection with intact atrial septum

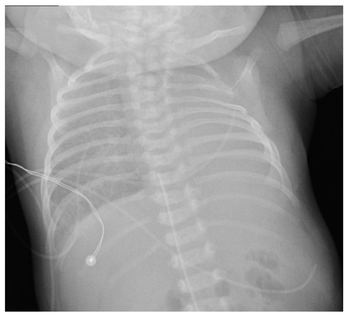

Figure 3: CXR at 2 hours of life concerning for obstructed pulmonary veins

Emergent Cardiac Cath:
- Need for LA decompression (Figure 2)
- Diagnosed TAPVR with tortuous venous connection draining to the coronary sinus with intact atrial septum and absent LPA
- Multiple attempts at balloon atrial septostomy failed, resulting in interatrial stent placement via hybrid approach (Figure 4)

Airway concerns:
- Grade 1 view with difficulty passing endotracheal tube through vocal cords, initial chest x-ray with ETT at thoracic inlet
- Intraoperative bronchoscopy by ENT visualized tracheal rings and absent left bronchus
- Median sternotomy views concerning for left lung aplasia

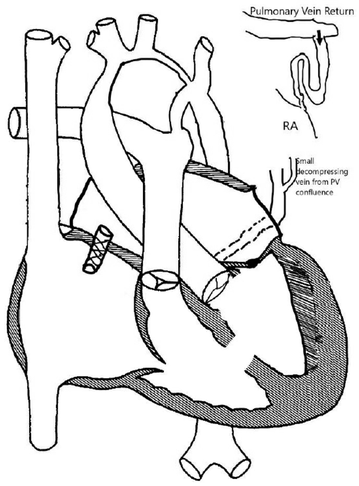

Figure 4: 2D representation of anatomy: Hypoplastic left heart, mitral atresia, aortic arch hypoplasia, absent LPA, muscular VSD, TAPVR with small connection to RA and decompressing vein to coronary sinus

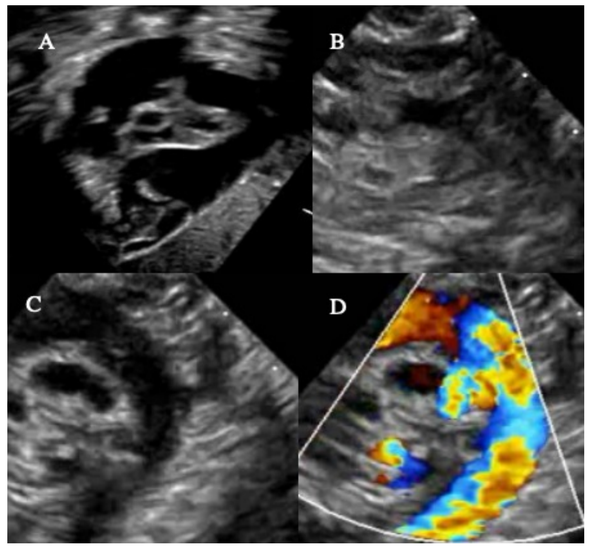

Figure 5: Transthoracic echo showing A. short axis view with dilated MPA, B. degraded suprasternal notch view of hypoplastic appearing transverse aortic arch, C. intact ductal arch with D. antegrade flow by color doppler

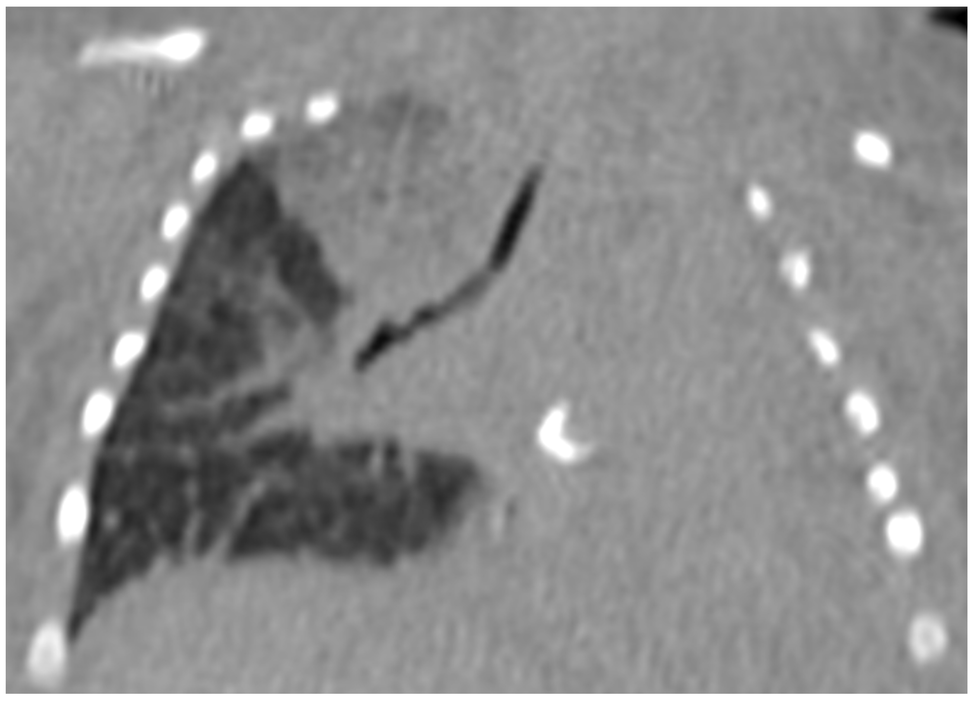

Figure 6: CT chest coronal view showing absence of left lung and left mainstem bronchus

CASE CONTINUED

Hospital Course:
- Admitted to PCICU on vasoactive and inotrope support with prostaglandin E1 for ductal dependent systemic blood flow with aortic arch hypoplasia (Figure 5)
- CT imaging confirmed congenital aplasia of left lung and bronchus (Figure 6)
- Cardiac surgical conference discussion: potential but extremely high-risk candidate for hybrid Norwood palliation given low birth weight and cardiopulmonary anomalies
- Multiple episodes of bradycardic and respiratory arrest during first week of life given unstable airway and ductal dependent single ventricle physiology

Family Meeting:
- Despite decompression of her obstructed pulmonary venous return, patient remained significantly compromised from a cardiac and ventilatory standpoint due to her congenital airway malformations.
- Ultimately, her family opted to transition to comfort care, and she died on day of life 8

CONCLUSIONS

The triad of left outflow tract obstruction, pulmonary venous obstruction and left lung aplasia with tracheal rings poses a likely lethal constellation.

Single ventricle heart disease with airway anomalies is associated with increased mortality and morbidity. Fetal diagnostics and advanced imaging offer crucial details that assist with family counseling regarding future prognostication and in multidisciplinary team decision-making for postnatal management.

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