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Tricuspid atresia with total anomalous pulmonary venous connection: An uncommon finding

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CASE

- A 33-year-old woman underwent fetal echocardiography at ~28 weeks gestation for evaluation of congenital heart disease revealing: a hypoplastic, thick echogenic tricuspid valve annulus with trivial antegrade flow and insufficiency, moderately hypoplastic RV (infundibulum), moderately hypoplastic pulmonary valve, main and branch pulmonary arteries (Fig 1).

- A left superior vena cava (LSVC) was noted and additional concerns for anomalous pulmonary venous connection possibly to the coronary sinus was noted (CoS).

- A scheduled repeat cesarean was performed at 38 weeks gestation resulting in the delivery of a female neonate weighing 2.87 Kg.

- Prostaglandins were initiated soon after delivery for maintaining adequate pulmonary blood flow.

- Post natal echocardiography confirmed the fetal diagnosis and defined the anomalous pulmonary venous connection to the LSVC close to its junction with the coronary sinus (Fig 2).

- A CT angiogram was performed for better delineation of the pulmonary venous anatomy and size (Fig 3).

- An initial palliation with a BT shunt was undertaken to ensure adequate pulmonary blood flow, coming off prostaglandins.

- Post-operatively, she developed signs of pulmonary over circulation and subsequently underwent main pulmonary artery banding to balance her systemic and pulmonary circulation.

- Planned stage II repair: Bilateral bidirectional Glenn procedure, TAPVC repair, atrial septectomy and MPA band tightening.

BACKGROUND

- Tricuspid atresia: 3-4% of all congenital heart disease.

- No true connection between the right atrium (RA) and right ventricle (RV) with concomitant findings of a ventricular septal defect (VSD), right ventricular hypoplasia, abnormalities of right ventricular outflow tract, pulmonary valve and pulmonary arteries.

- Common associated lesions have been described including transposition of great vessels, hypoplasia of aortic arch, double outlet ventricle or a common arterial trunk.

DECISION MAKING

- Prenatal diagnosis of tricuspid atresia with TAPVC is a very rare association but should be considered when there is failure to connect the pulmonary veins to the left atrium and when a left atrial to descending aorta gap is present on fetal echocardiogram.

- Diligent interrogation for a confluence or vertical vein should be undertaken in order to identify the abnormal pulmonary venous connection.

- Multi-modality imaging compliments echocardiography in diagnosing and assisting in management decision making.

- Important post-natal considerations include: Balancing pulmonary blood flow and unmasking potential presence of obstruction to pulmonary venous return.

- Repair of TAPVC can be deferred for the second stage repair of this single ventricle physiology when there is no clinical evidence of obstruction to the pulmonary venous flow.