A Congenital Two-For-One: A Rare Case of Right-Sided Heart Failure Secondary to Atrial Septal Defect and Partial Anomalous Pulmonary Venous Return

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- Congenital heart defects (CHDs) are a significant cause of right-sided heart failure (RHF)
- While the secundum type atrial septal defect (ASD) is the most common manifestation in adults, the presence of concomitant partial anomalous pulmonary venous return (PAPVR) is a relatively rare combination
- Here, we present a case of RHF in a patient with secundum type ASD and PAPVR

Case

A 30-year-old, Spanish-speaking female presented to the hospital for reoccurring right upper quadrant abdominal pain. She was previously evaluated at an outside facility for similar symptoms, but she was lost to follow-up.

On presentation, she was in no acute distress and hemodynamically stable. Cardiopulmonary examination was benign. Basic laboratory workup was only remarkable for troponin 170 and 168, but CT angiography of the chest and abdomen demonstrated severe dilation of the pulmonary arteries and right heart chambers.

She was admitted to the hospital with concern for RHF secondary to a left-to-right shunt. Suspicion for an ASD was confirmed with echocardiography, which revealed a secundum ASD. Additional findings included a severely dilated right ventricle (RV) with septal flattening, RV systolic pressure 31 mm Hg, and pulmonary-to-systolic blood flow ratio (Q_p:Q_s) 2.6 to 1.

She was deemed appropriate for ASD closure, so pre-operative evaluation with MRI and right heart catheterization was completed to expedite surgical repair. At this time, PAPVR with right superior and inferior pulmonary veins draining into the right atrium was incidentally discovered. Afterwards, she was scheduled for outpatient surgical repair with thoracic surgery and discharged home.

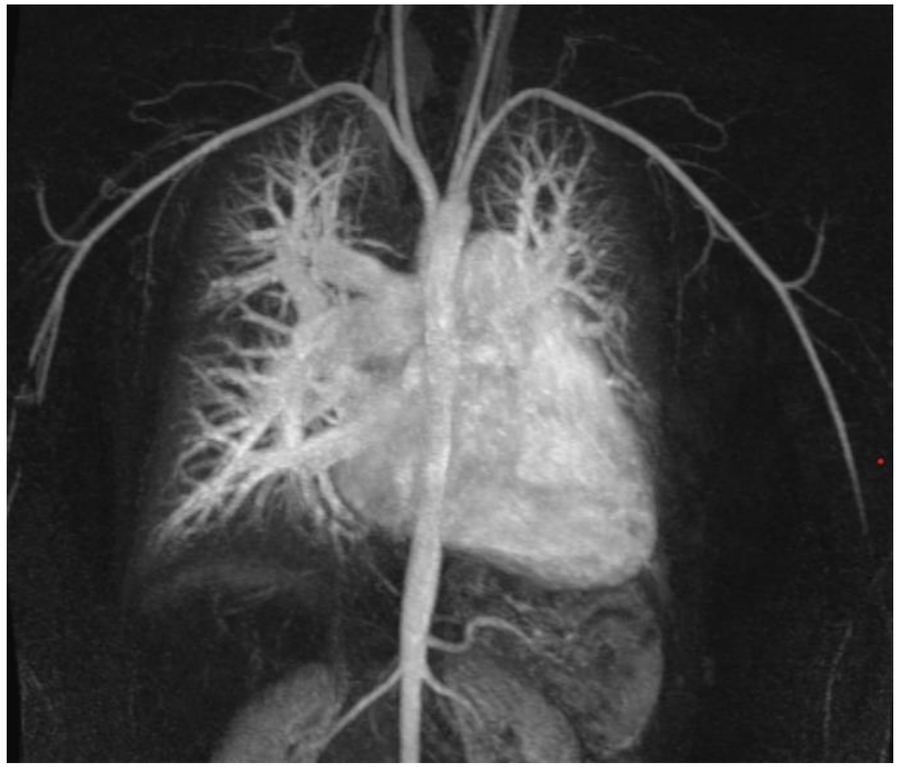

Image 1. *Cardiac MRI showing PAPVR with right superior and inferior pulmonary veins draining into the right atrium*

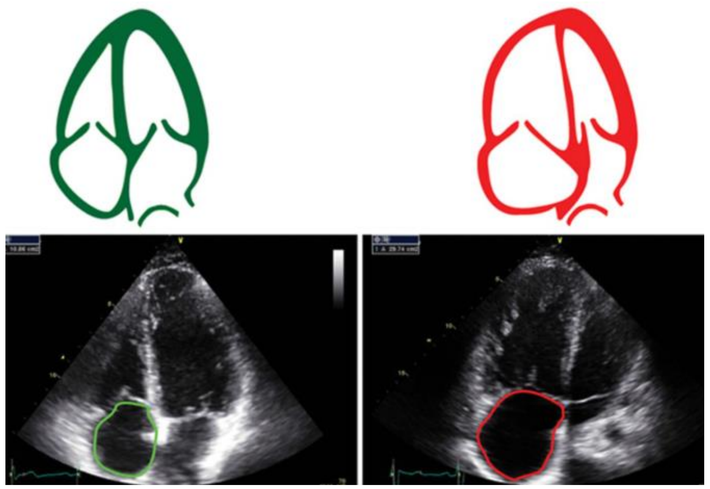

Image 2. *Schematic and echocardiogram highlighting the findings of RHF*

Discussion

- Differential diagnosis for RHF consists of intrinsic myocardial disease, pressure overload, and volume overload
- CHDs are potential causes of RHF from progressive volume overload if left-to-right shunting is untreated
- ASD is the most common CHD while PAPVR is less prevalent, accounting for 0.4-0.7% of cases [1,3]
- ASD is present in 51.6% of PAPVR cases, predominately as the sinus venosus subtype [2]
- Secundum ASD is a rare variant of ASD presentation in the setting of PAPVR, accounting for 10% of manifestations [2]
- Echocardiography is the first-line imaging modality for ASDs to assess defect size and location
- Cardiac magnetic resonance imaging provides accurate right ventricular size and function, locations of pulmonary veins, and Q_p:Q_s quantification
- Definite treatment is surgical repair, especially when the Q_p:Q_s is greater than 1.5:1 and evidence of right atrial or ventricular enlargement is present [4]

- CHDs with left-to-right shunting may cause RHF
- Secundum ASD with PAPVR is a rare manifestation
- Surgical repair is indicated in the following scenarios:
  - Q_p:Q_s > 1.5:1 with right atrial or ventricular dilation
  - Pulmonary hypertension
  - Paradoxical embolism
  - Symptomatic or complication prevention

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