6-2024

Post-Operative Diagnosis of Anomalous Left Coronary Artery From The Right Pulmonary Artery via Transthoracic Echocardiogram

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ASE2024 Global Echocardiography

June 14-16, 2024 Oregon Convention Center Portland, Oregon

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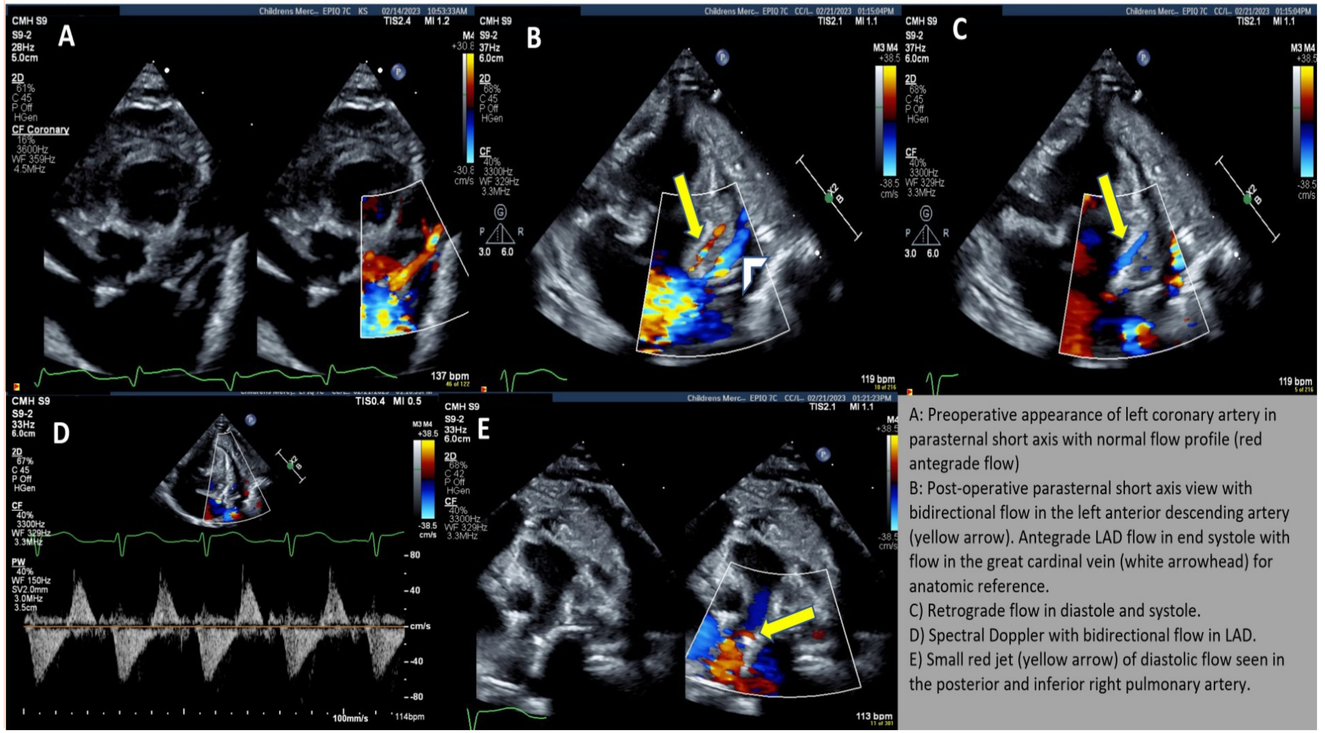
CASE PRESENTATION

- A term infant female was diagnosed with a ventricular septal defect (VSD) and aortic coarctation.
- Preoperative echocardiography and computed tomography angiography (CTA) showed a normal right coronary artery origin, but inconclusive left coronary artery (LCA) origin.
- There was a normal flow profile in the LCA by echocardiography. She underwent repair with extended end-to-end anastomosis and VSD closure during which surgical inspection revealed a usual appearing, retro pulmonary path of the LCA towards the left sinus of Valsalva.
- Postoperative echocardiogram revealed mild global dysfunction but no regional wall motion abnormalities.
- Repeat echocardiogram revealed severe dysfunction most prominent in the anterolateral and posterolateral segments from base to apex.
- There was new, bidirectional flow in the LCA and abnormal diastolic flow in the posterior right pulmonary artery (RPA) raising suspicion for anomalous origin of the LCA from the RPA (ALCARPA). Cardiac catheterization confirmed the diagnosis.

DIAGNOSIS OF ALCARPA

- Echocardiography plays a vital role in detection of ALCARPA.
- Commonly observed clues for diagnosis are flow reversal in the LCA, abnormal diastolic flow in the pulmonary artery and sequelae of left ventricular ischemia
- The challenge is that these signs vary with age and pulmonary artery pressure.
- Other imaging modalities such as Computed tomography angiography (CTA) are valuable in diagnosis but may suffer from improper contrast timing and motion artifact.
- When the LCA originates from the RPA, the origin appears normal due to its proximity to the left sinus of Valsalva, even by visual inspection.
- Meticulous coronary imaging and clinical suspicion are required for timely detection. Invasive angiography is the gold standard for diagnosis.



A: Preoperative appearance of left coronary artery in parasternal short axis with normal flow profile (red antegrade flow)
B: Post-operative parasternal short axis view with bidirectional flow in the left anterior descending artery (yellow arrow). Antegrade LAD flow in end systole with flow in the great cardinal vein (white arrowhead) for anatomic reference.
C) Retrograde flow in diastole and systole.
D) Spectral Doppler with bidirectional flow in LAD.
E) Small red jet (yellow arrow) of diastolic flow seen in the posterior and inferior right pulmonary artery.

DISCUSSION

- There are few cases associating ALCARPA with aortic coarctation.
- This case is unique because the coronary anatomy was interrogated before surgery, but ALCARPA was not demonstrated due to a low index of suspicion for this rare anatomy and associated pulmonary hypertension.
- Presence of a coarctation and VSD lead to sufficient pulmonary hypertension for antegrade LCA flow which hinders the preoperative diagnosis.
- This case highlights the diagnostic challenge of this association of defects and reiterates the importance of definitive coronary imaging preoperatively.
- Persistent myocardial dysfunction following coarctation and VSD repair should prompt careful evaluation for ALCARPA

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