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Key Points in the Echocardiographic Diagnosis of Supracardiac Total Anomalous Pulmonary Venous Connection to the Vertical Vein

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Clinical Image description

A female term newborn was admitted to the neonatal intensive care unit in her first 24 hours of life due to cyanosis and respiratory difficulty. On physical exam, she weighed 3.8 kg, was 50 cm long, had a heart rate of 170 beats per minute, a respiratory rate of 48 breaths per minute, central cyanosis with 82% saturation on room air, a hyperdynamic right sternal impulse, an intensified split second sound, and a systolic murmur at the right sternal border. A chest x-ray showed cardiomegaly and increased pulmonary flow. The echocardiogram diagnosed non-obstructive supracardiac total anomalous pulmonary venous connection to the vertical vein as well as severe pulmonary hypertension. She was referred for surgical repair.

Total anomalous pulmonary venous connection (TAPVC) is an infrequent Congenital Heart Disease (CHD), accounting for 1-3% of all CHDs (1). Without surgical repair, mortality is greater than 50% in the first three months of life, and almost 90% at age one. Death occurs earlier when the anomaly is obstructive (as evidenced by respiratory failure, severe heart failure and cyanosis), than when it is non-obstructive (2). The development of Pulmonary Arterial Hypertension (PAH) leads to a deterioration in functional class and death in survivors (3).

The key echocardiographic elements in the diagnosis of supracardiac TAPVC are:

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1. The absence of connected pulmonary veins in the LA
2. Incomplete LA (the venous portion is lacking)
3. Identification of the collector (venous convergence and pulmonary veins)
4. Identification of the exact connection site (the vertical vein in this case)
5. Marked dilation of the right chambers
6. Identification of the atrial septal defect with right to left flow (in a venous-arterial direction)
7. Severe pulmonary hypertension
8. A smaller left ventricle (incomplete filling volume)
9. Evaluation of the presence and degree of obstruction
10. Associated lesions with their degree of hemodynamic impact

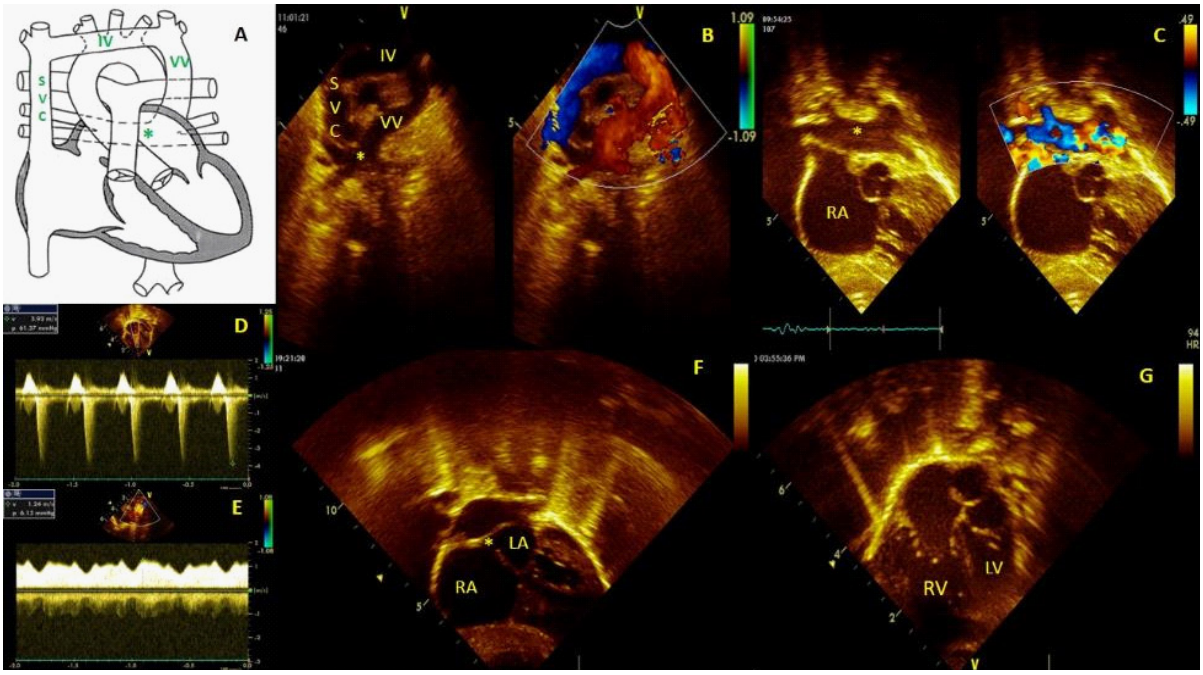

Figure 1: A: Ilustration image of supracardiac total anomalous pulmonary venous connection to the vertical vein.
VV: Vertical vein; IV: Innominate Vein (venous brachiocephalic trunk); SVC: Superior Cava Vein; (*) Collector.
B: Dual comparative doppler color echocardiographic supraesternal view, shows the same in A.
C: Dual comparative doppler color subcostal view, shows retrocardiac collector (*).
RA: Right Atrium.
D: Doppler spectral tricuspid reurgitation shows pulmonary hypertension.
E: Doppler image into VV.
F: Subcostal view shows atrial septal defect (*).
LA: Left Atrium.
G: Apical four chambers view, show dilated RV and small LV.
RV: Right Ventricle, LV: Left Ventricle.

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