Challenges in Echocardiographic Diagnosis of Corrected Malposition of Great Arteries: The Segmental Approach coming Handy

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**Challenges in Echocardiographic Diagnosis of Corrected Malposition of Great Arteries: The Segmental Approach coming Handy**

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### Clinical Presentation

Two cases of Anatomically Corrected Malposition of Great Arteries (ACMGA) \(\{S,D,L\}\) are presented with distinct differences in conal anatomy.

**Case 1:**
A fetal echocardiogram (echo) performed at 28 weeks gestation due to multiple fetal anomalies revealing: atrial situs solitus, D-loop ventricles, a large conoventricular ventricular septal defect (VSD) and an overriding, anterior and leftward aorta. The diagnosis of double outlet right ventricle (DORV) vs ACMGA was entertained. Postnatal echo revealed an \(\{S,D,L\}\) segmental anatomy, with atrioventricular and ventriculoarterial concordance consistent with ACMGA and a muscular VSD.

**Case 2:**
A 3-week-old boy presented to clinic for the evaluation of a murmur with no associated cardiac symptoms. Transthoracic echocardiogram revealed ACMGA \(\{S,D,L\}\) and a small membranous VSD.

### Imaging Findings

**Case 2:**
- The most typical anatomic characteristics of ACMGA \(\{S,D,L\}\) were identified with the commonly found bilateral sub-arterial conus and parallel outflow tracts.

Echocardiographically subcostal, parasternal and suprasternal sweeps will help diagnose ACMGA by delineating ventriculo-arterial alignment, ventricular and outflow relations and conal anatomy. Magnetic Resonance Imaging as used in both cases can be performed for evaluation of the Qp/Qs given the VSDs and helps confirm the diagnosis of ACMGA.

### Role of Imaging in Patient Care

Accurate prenatal diagnosis of ACMGA is essential for appropriate counseling and postnatal management. Postnatally, standard echocardiographic views and multimodality imaging will elucidate ventriculoarterial connections, conal anatomy, and severity of associated anomalies.

### Summary

ACMGA is a rare congenital heart disease occurring from failure of involution and rightward rotation of the subaortic conus. This results in parallel spatial relationship of the great arteries while maintaining ventriculoarterial concordance. Subtypes of segmental anatomy include the most common \(\{S,D,L\}\) (usually with bilateral conus) and \(\{I,L,D\}\) both with normal physiology, and the rarer \(\{S,L,D\}\) and \(\{I,D,L\}\) with transposition physiology. Identifying the segmental anatomy and relations will result in accurate diagnosis of this rare entity and appropriate management.