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

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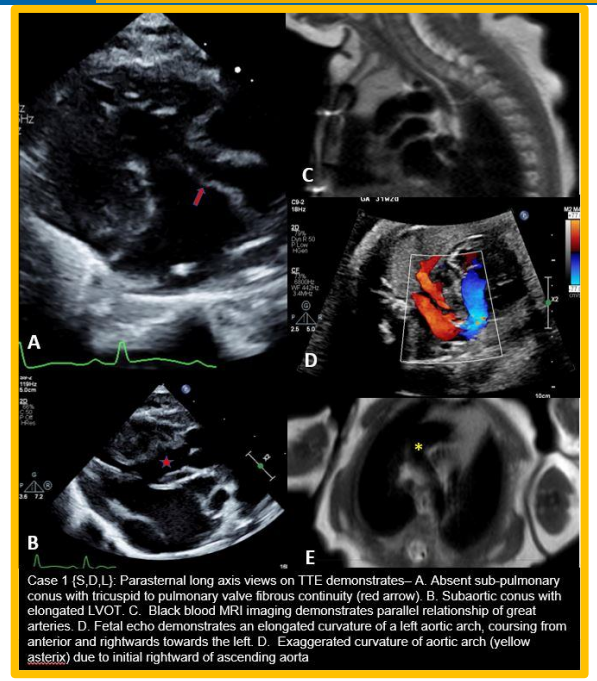
Clinical Presentation

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 1 {S,D,L}: Parasternal long axis views on TTE demonstrates– A. Absent sub-pulmonary conus with tricuspid to pulmonary valve fibrous continuity (red arrow). B. Subaortic conus with elongated LVOT. C. Black blood MRI imaging demonstrates parallel relationship of great arteries. D. Fetal echo demonstrates an elongated curvature of a left aortic arch, coursing from anterior and rightwards towards the left. D. Exaggerated curvature of aortic arch (yellow asterix) due to initial rightward of ascending aorta

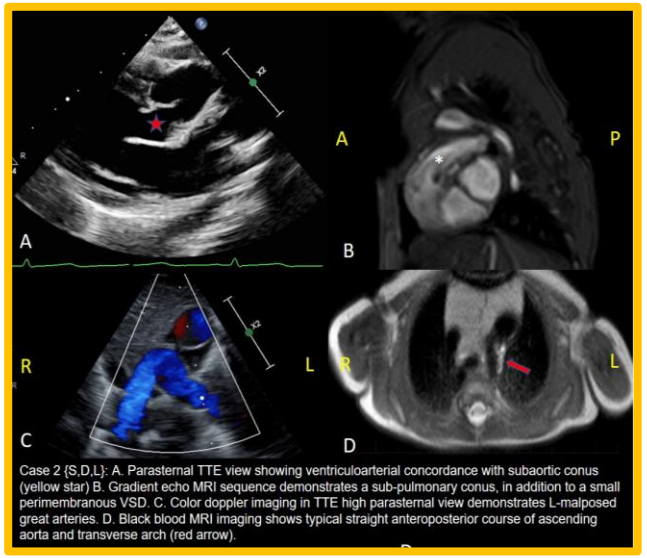
Case 1:

- This rare ACMGA type posed the most diagnostic challenges.
- A more anterior rotation of the left ventricle, the horizontal orientation of the ventricular septum due to absence of a sub-pulmonary conus and an elongated subaortic conus led to the misconception of an overriding aorta and DORV prenatally.
- Pulmonary to tricuspid valve fibrous continuity was present.
- The unusual position of the aortic valve resulted in an elongated curvature of the aortic arch, with a proximal rightward course of the ascending aorta but eventually crossing leftward of the trachea on top of the left bronchus towards a left thoracic and descending aorta.

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.



Case 2 {S,D,L}: A. Parasternal TTE view showing ventriculoarterial concordance with subaortic conus (yellow star) B. Gradient echo MRI sequence demonstrates a sub-pulmonary conus, in addition to a small perimembranous VSD. C. Color doppler imaging in TTE high parasternal view demonstrates L-malposed great arteries. D. Black blood MRI imaging shows typical straight anteroposterior course of ascending aorta and transverse arch (red arrow).

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.