

Children's Mercy Kansas City

SHARE @ Children's Mercy

Fetal Cardiology Education Series

Fetal Health Center

2-2024

Congenitally Corrected Transposition of the Great Arteries: Imaging Pearls for Fetal Assessment

Rita France

Let us know how access to this publication benefits you

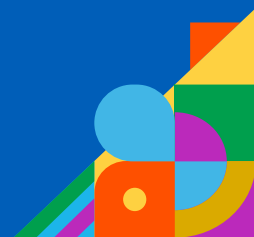
Follow this and additional works at: https://scholarlyexchange.childrensmercy.org/fetal_cardiology_series

Congenitally Corrected Transposition of the Great Arteries Imaging Pearls for Fetal Assessment

Rita France, RDCS, RDMS, RT, FASE
Director, Echocardiography



No disclosures



Objectives

- Discuss how to identify morphology and anatomic connections utilizing a segmental approach when imaging Congenitally Corrected Transposition of the Great Arteries
- Highlight the associated defects and imaging required to identify these defects

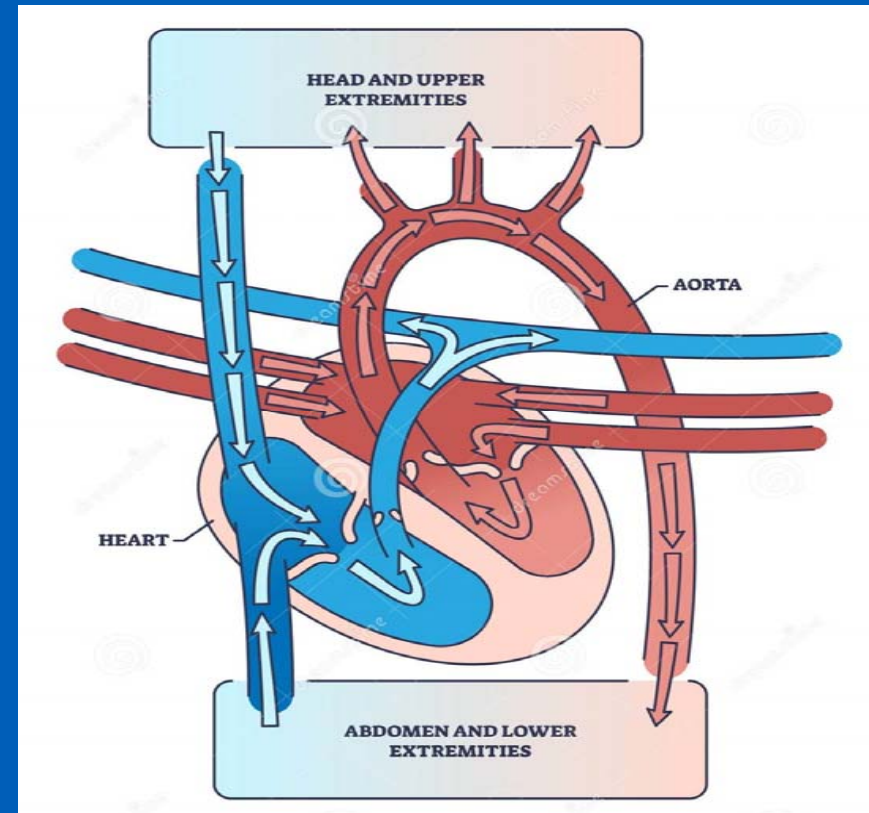
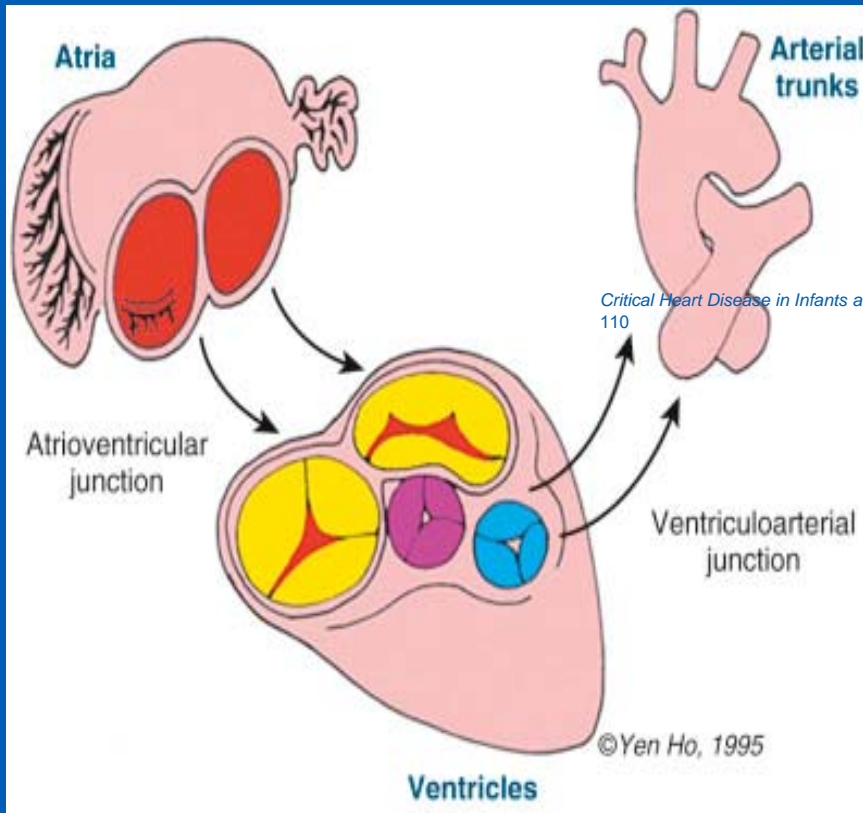


Introduction

- Characterized by atrioventricular and ventriculoarterial discordance
- 0.02 to 0.03 per 1000 live births
- 0.5 to 1% of all Congenital Heart Defects



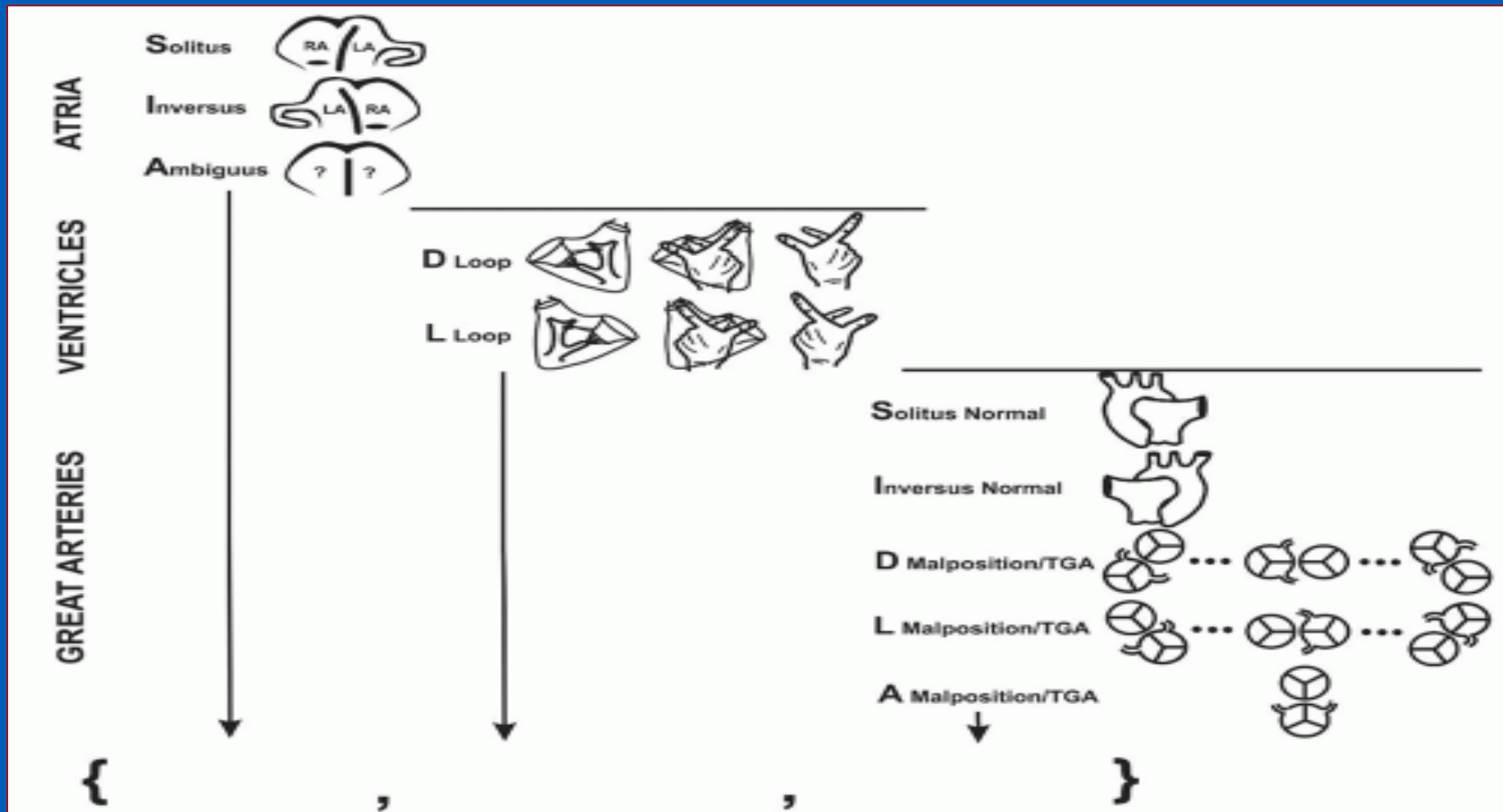
The Segmental Approach



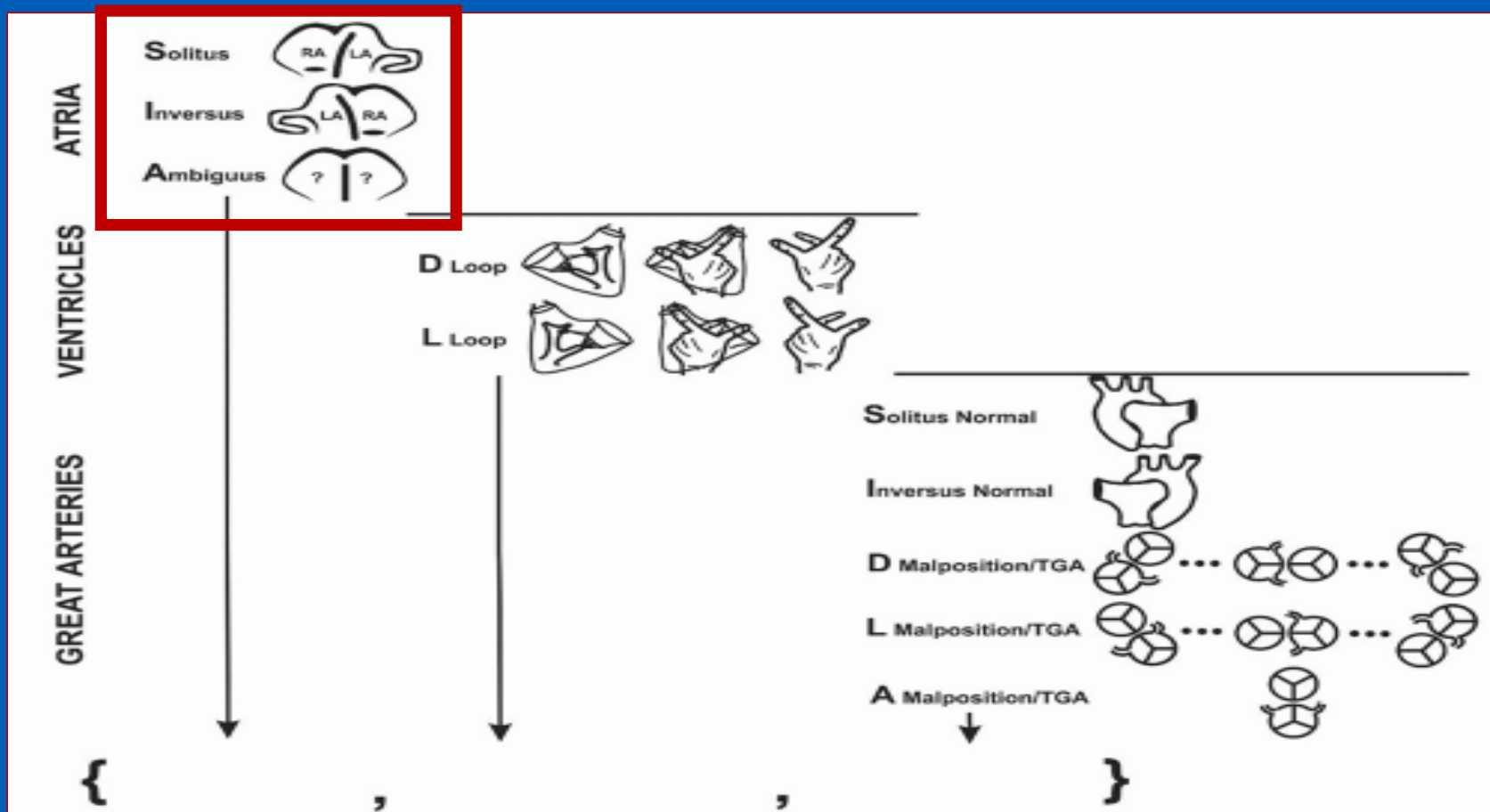
Critical Heart Disease in Infants and Children. 3rd ed. Elsevier 2019; 100-110

<https://www.dreamstime.com/systemic-circulation-heart-aorta-blood-system-outline-diagram-labeled-educational-scheme-upper-lower-extremities-supply-image296707851>

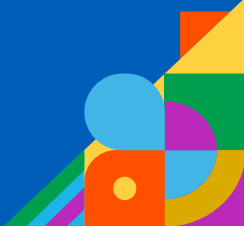
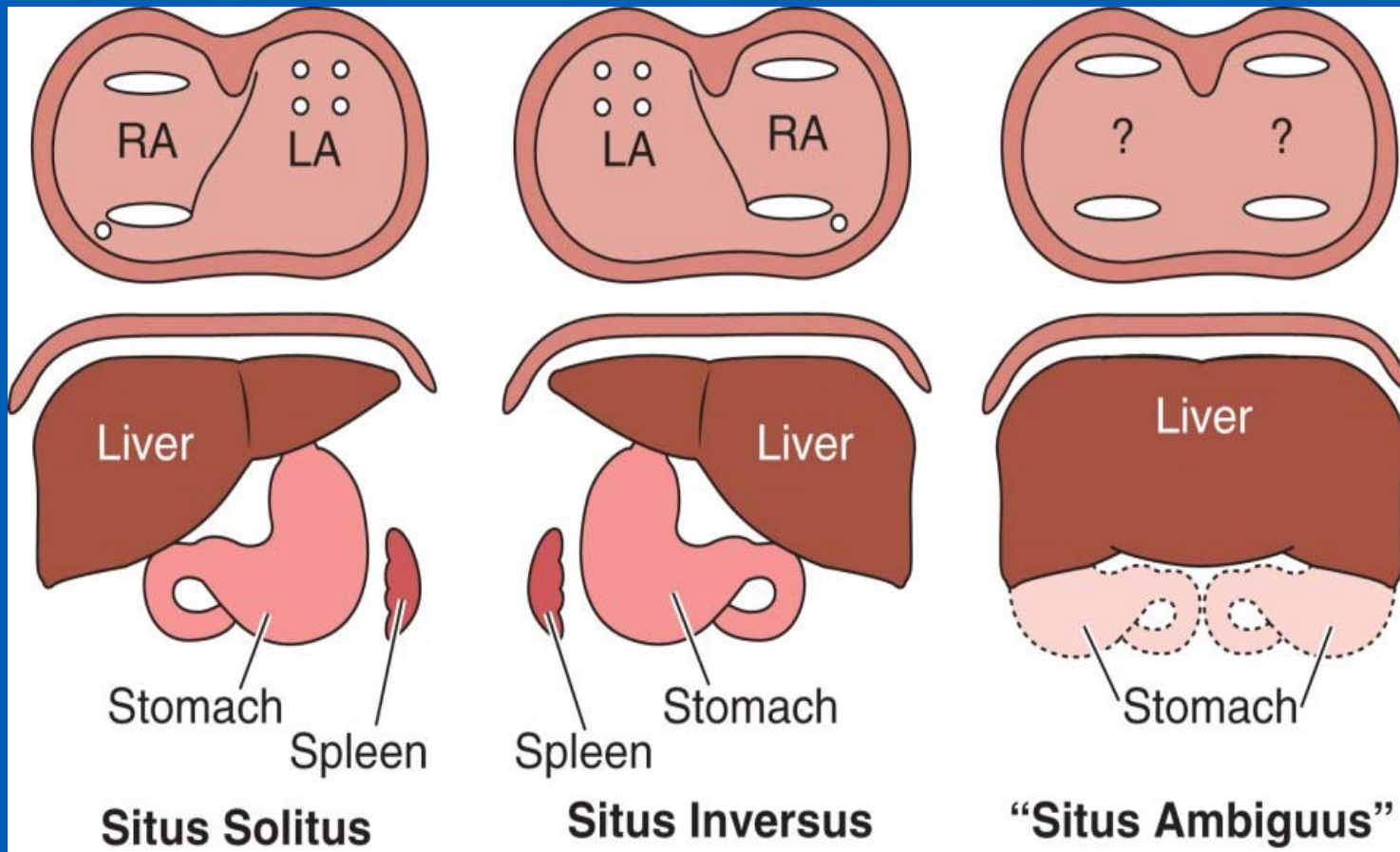
Segmental Notation



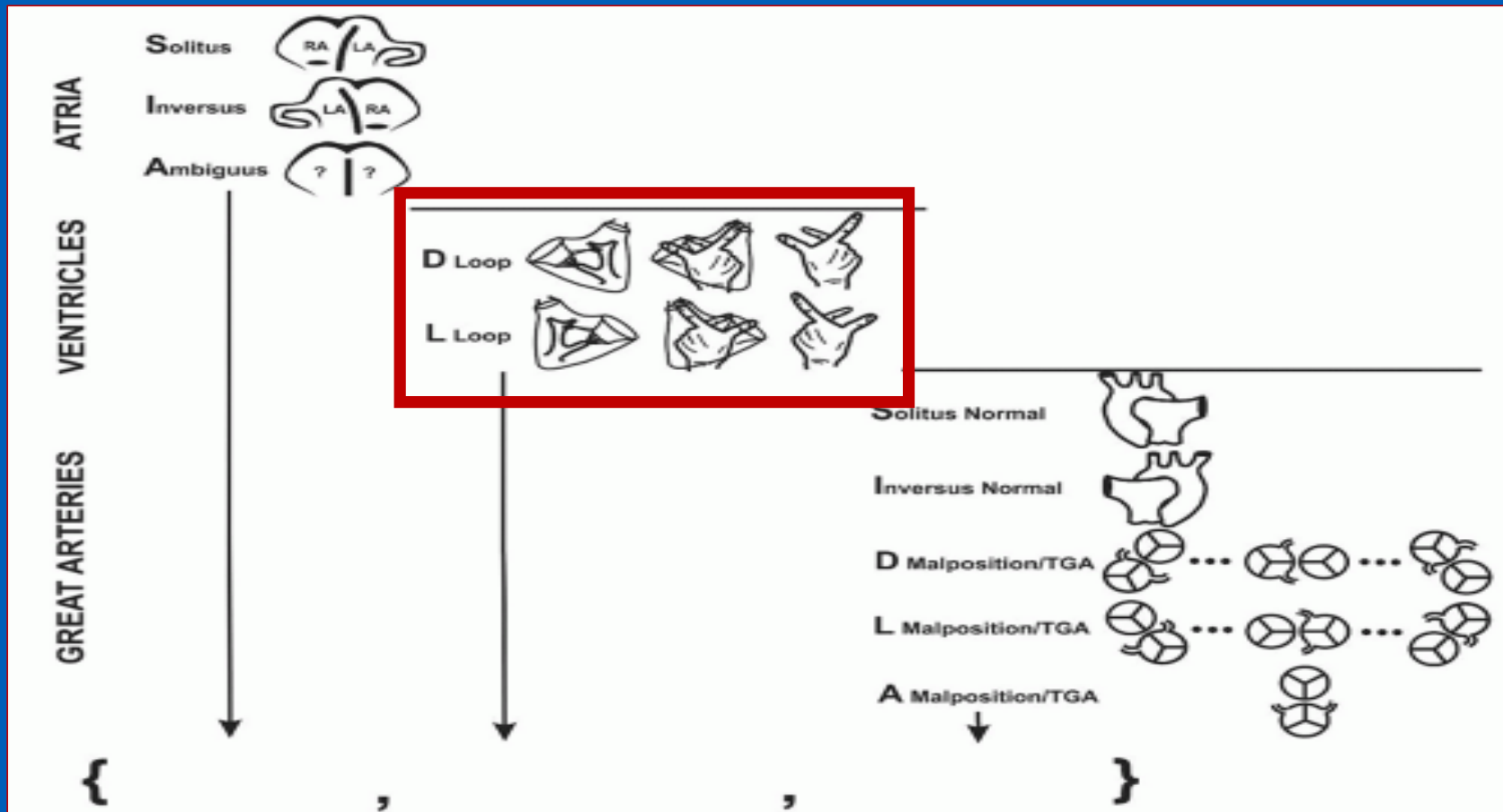
Segmental Notation



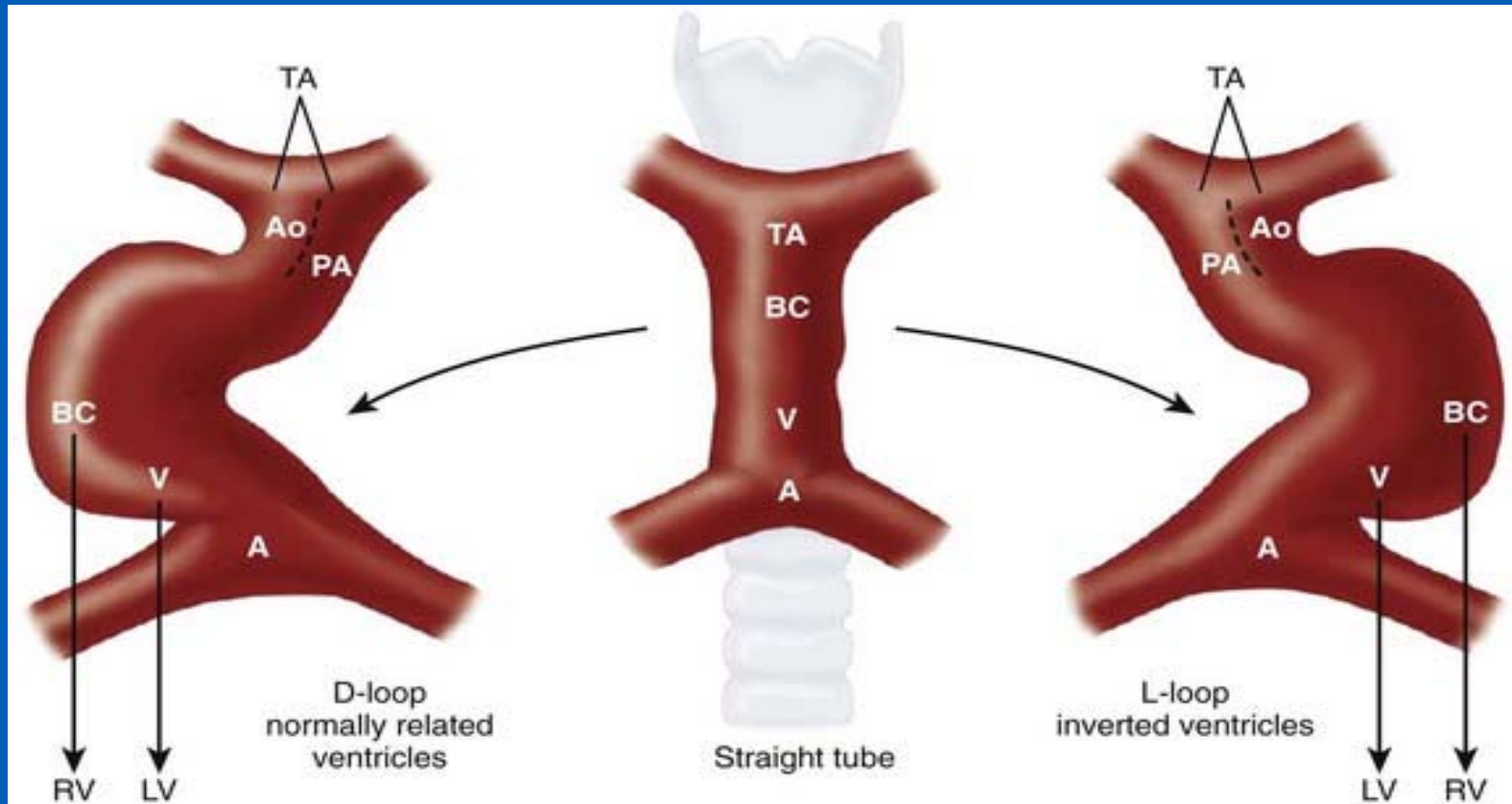
Atrial Situs



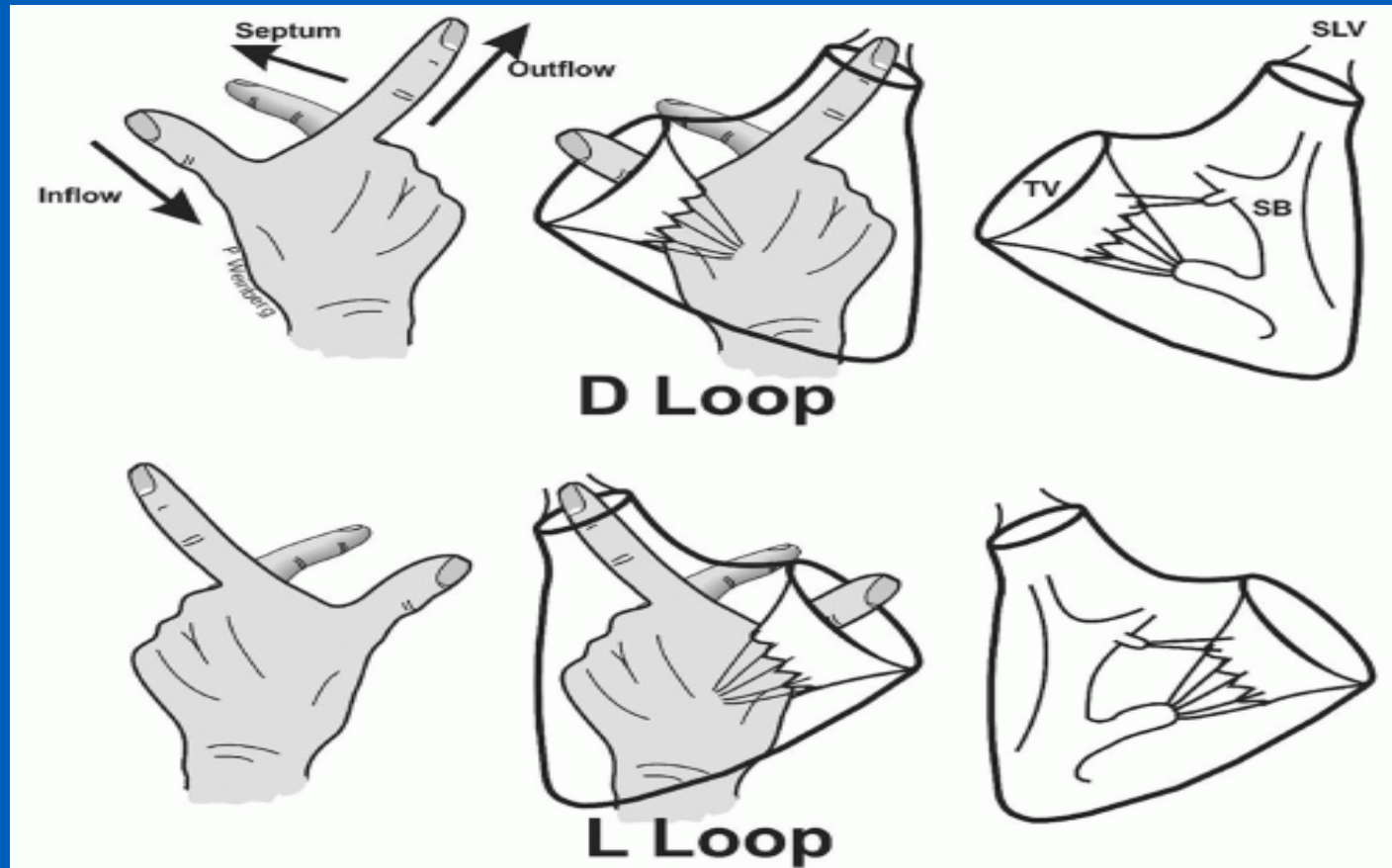
Segmental Notation



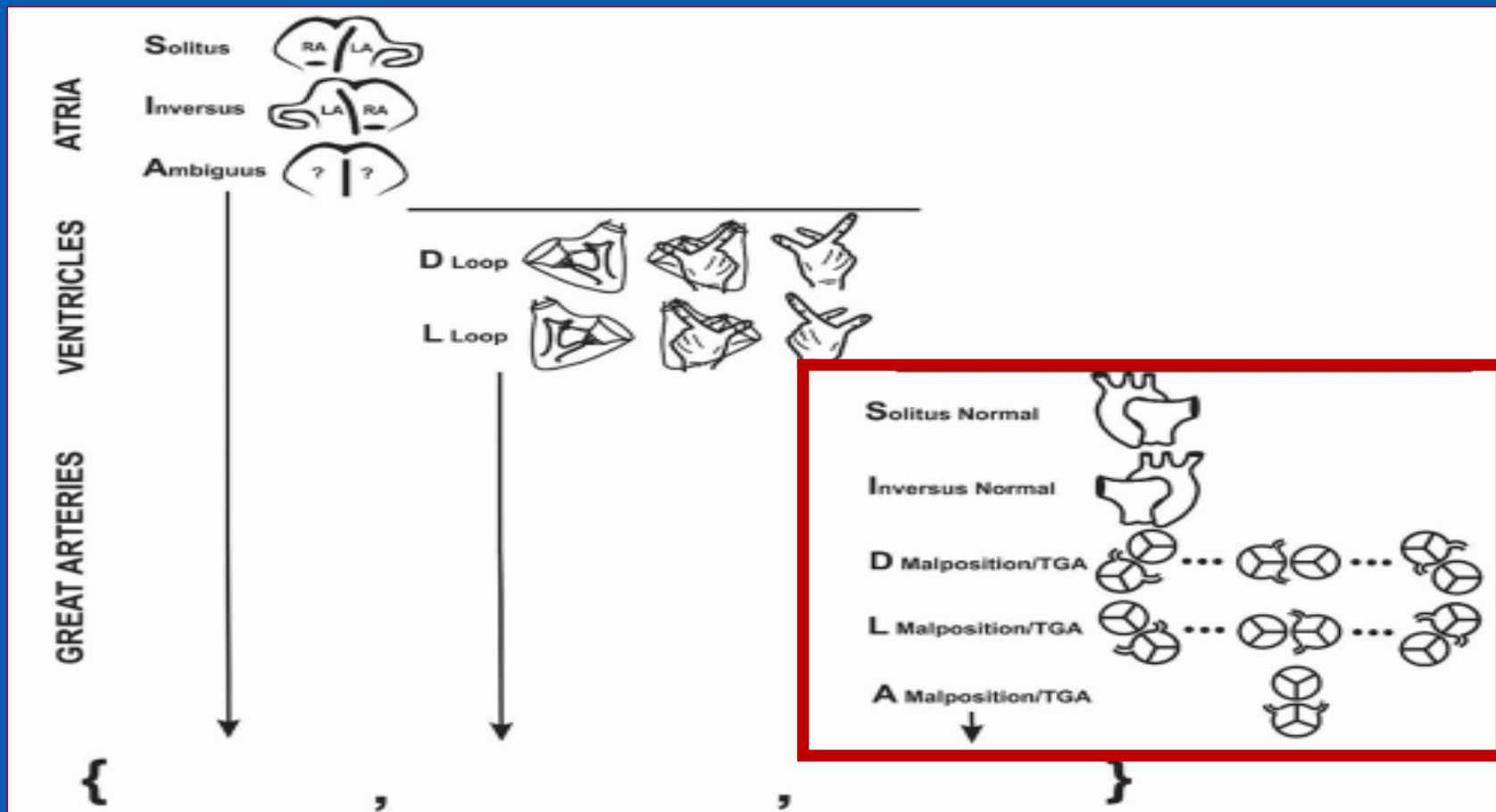
Looping



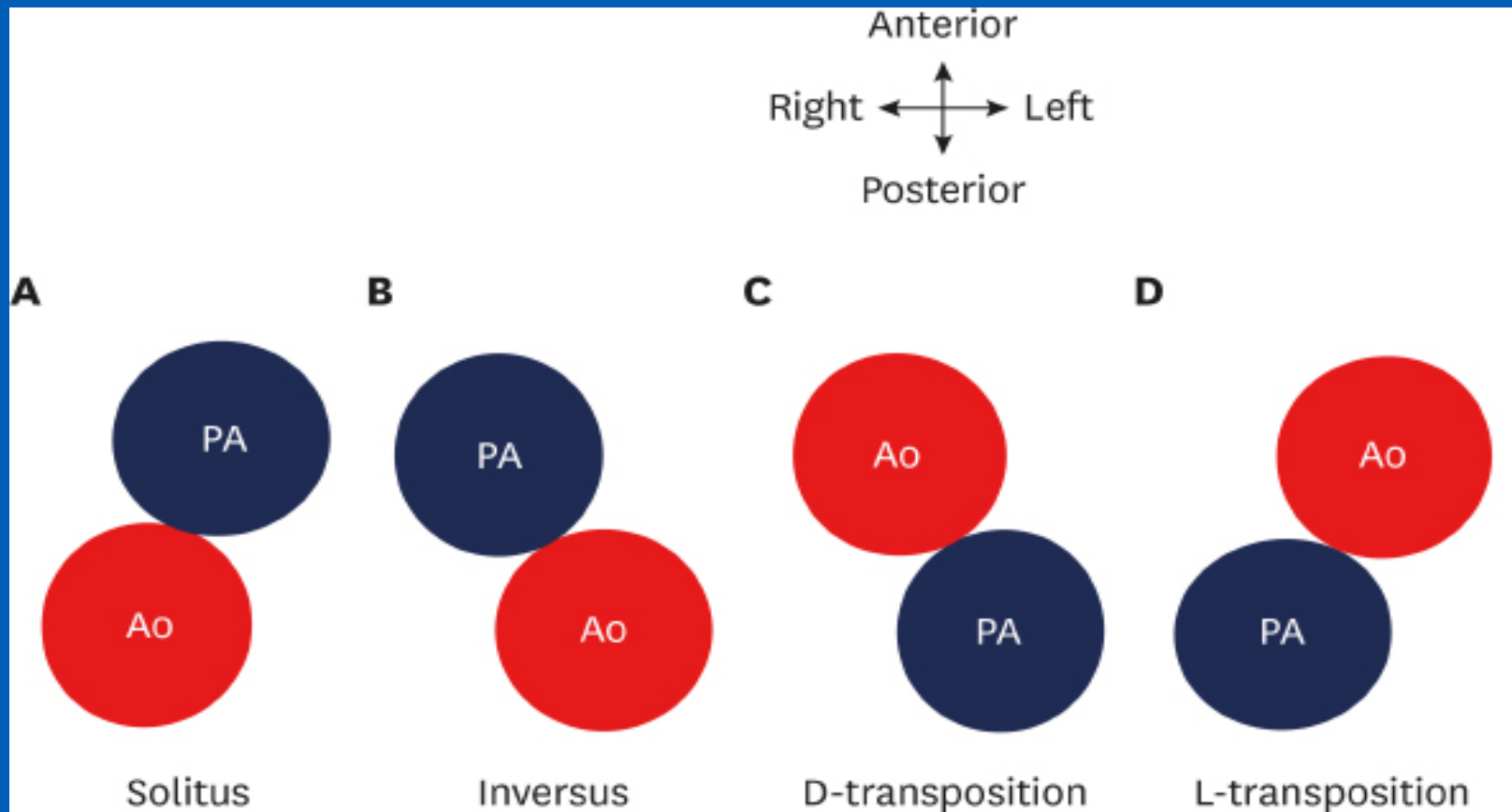
Looping



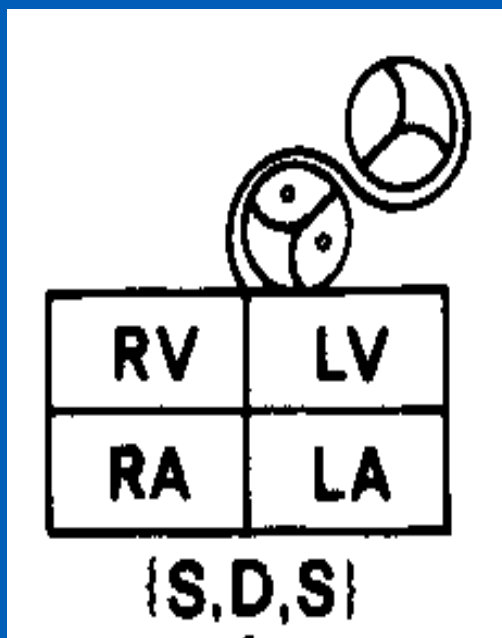
Segmental Notation



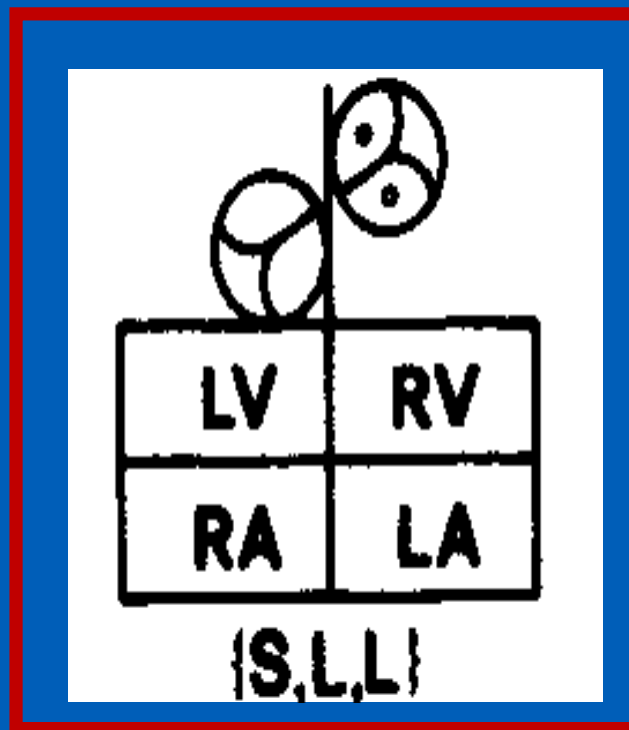
Great Artery Position



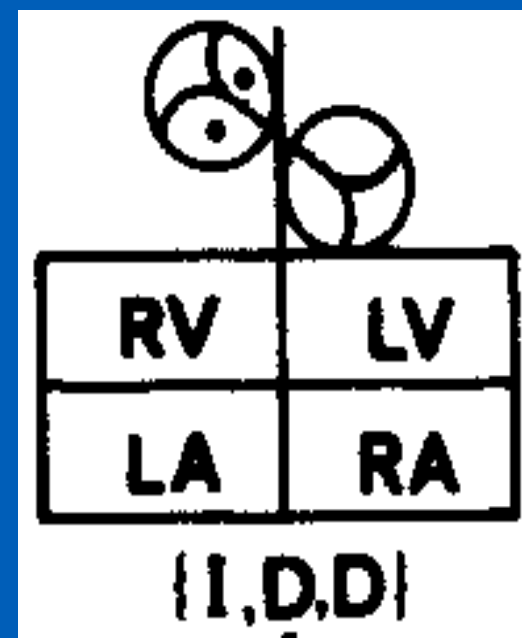
Segmental Classification



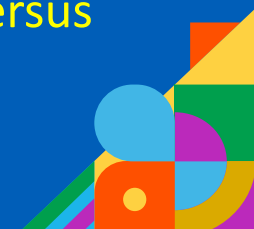
NORMAL



CCTGA



CCTGA Situs Inversus



Path of Blood Flow in a CCTGA

Systemic veins



Morphologic RA



Mitral Valve



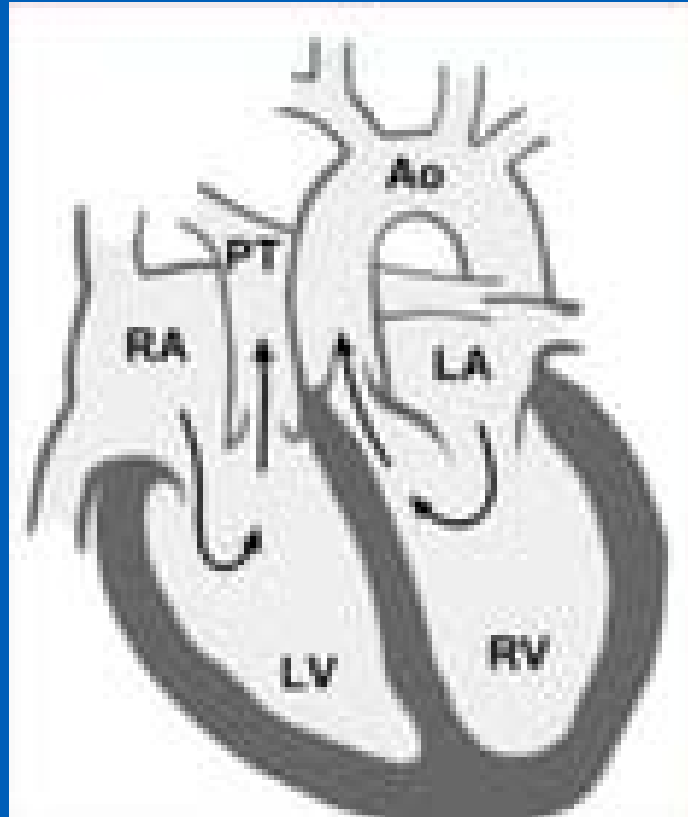
Left Ventricle



Pulmonary Valve



Pulmonary Artery



Pulmonary Veins



Morphologic LA



Tricuspid Valve



Right Ventricle



Aortic Valve



Aorta



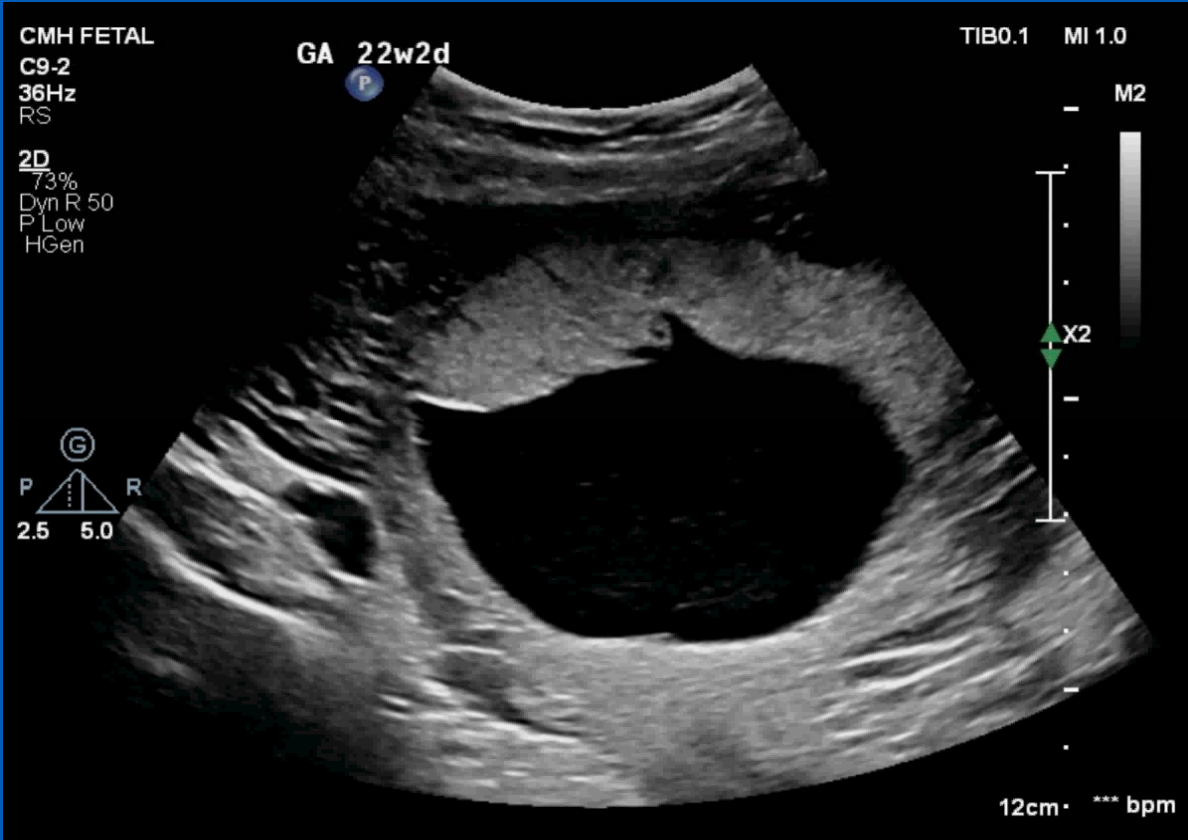
CCTGA Associated Lesions

Associated Lesions can occur in over 90% of CCTGA

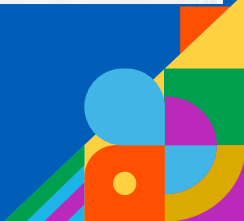
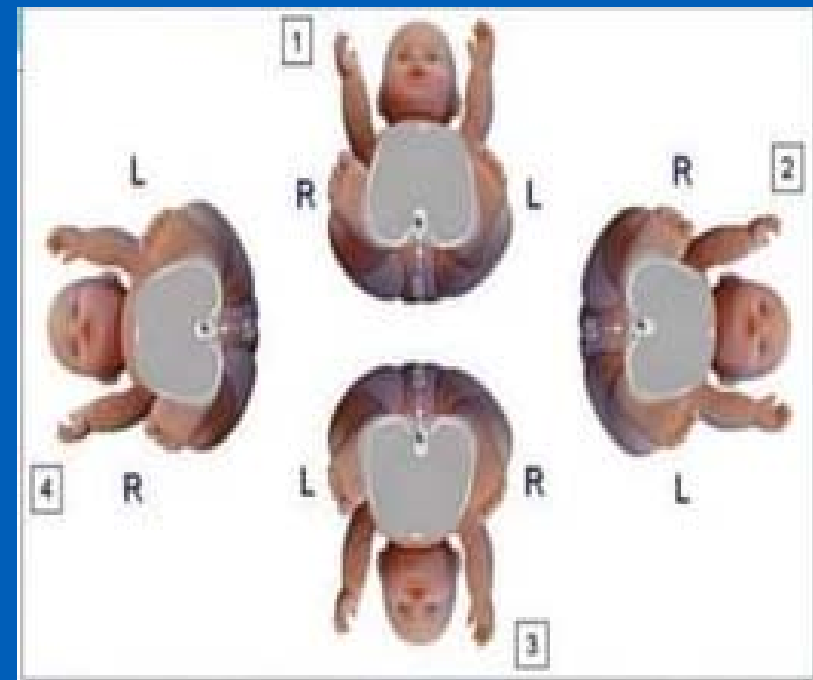
- **VSD in 60-80%, most commonly perimembranous**
- **LVOT or subpulmonary obstruction in approximately 50%**
- **Tricuspid valve issues are common – up to 90% in autopsy findings, less frequently identified clinically**
- **Situs inversus in approximately 5%**
- **Right arch can occur in 18%**
- **AV node abnormalities which can result in Complete Heart Block due to the abnormal course and location of the AV node and bundle of His**

Wyman, et al Echocardiography in Pediatric and Congenital Heart Disease; From Fetus to Adult. Spain: Wiley-Blackwell; 2011;444-447

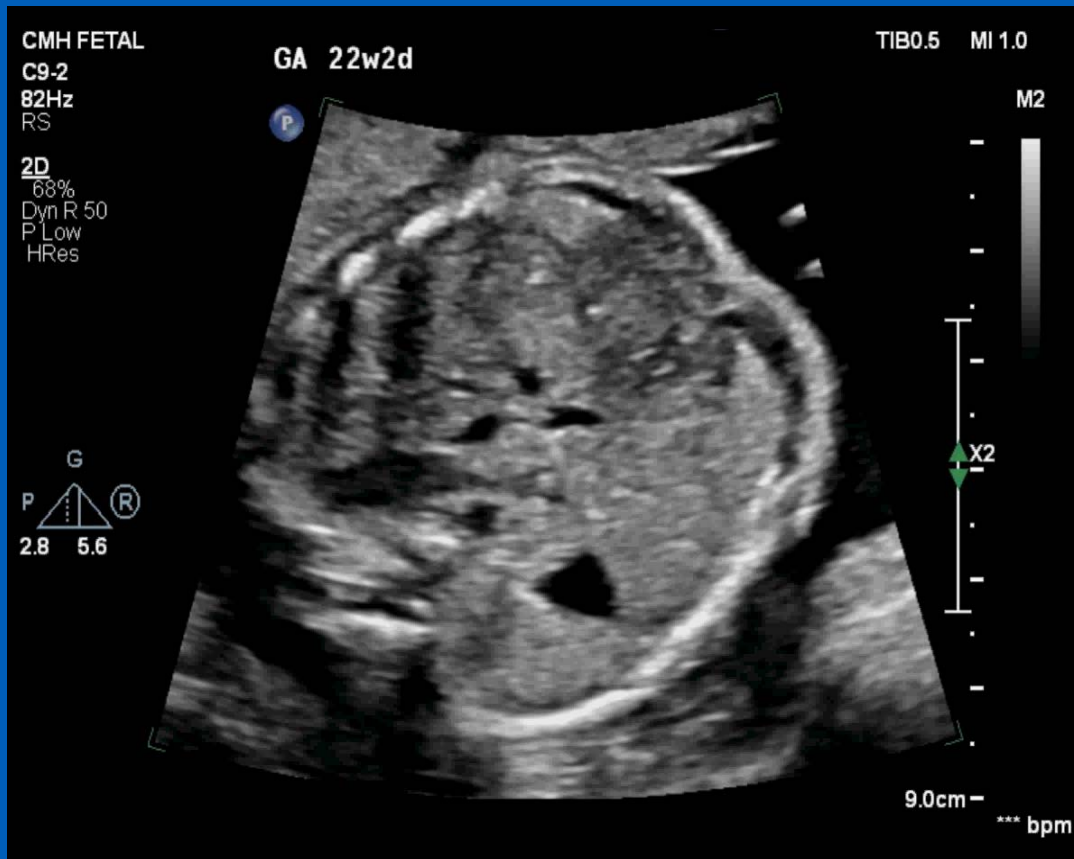
Fetal Presentation



CORDES



Focused 2D Sweep with Breech Presentation



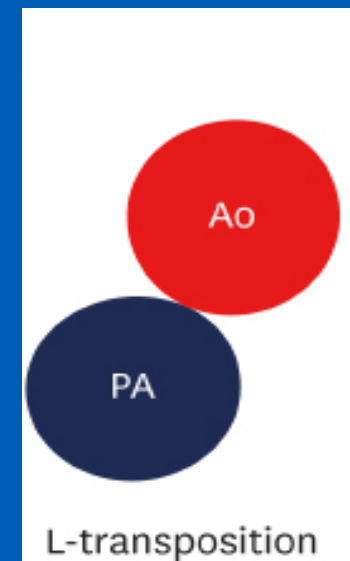
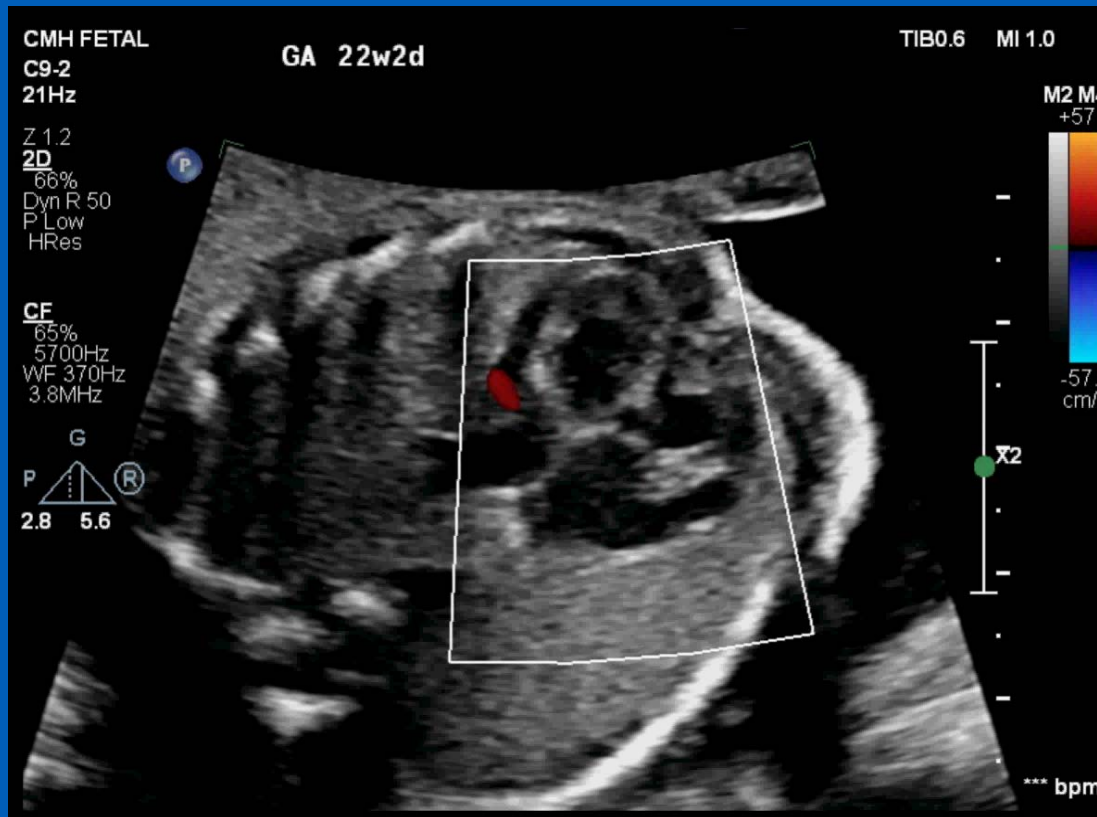
Visceral Situs



Mesocardia



Focused Color Sweep in Breech Presentation



Atrioventricular Valve Attachments

Right sided LV/MV



Left-sided RV/TV



Tricuspid Valve Abnormalities

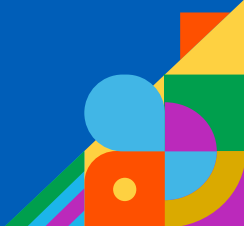
- Dysplasia with/without displacement
- Ebstein-like malformation
 - Atrialized portion usually small and myocardium is not thinned
 - Typically no rotational displacement of the leaflets
 - Anterior leaflet is not usually large and “sail like”



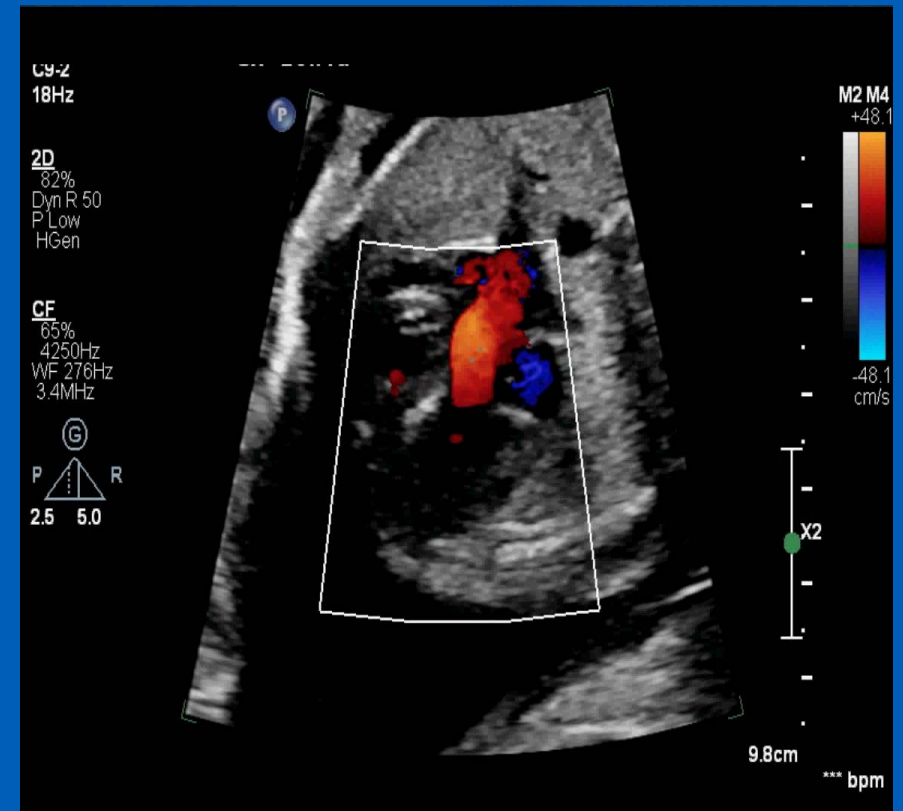
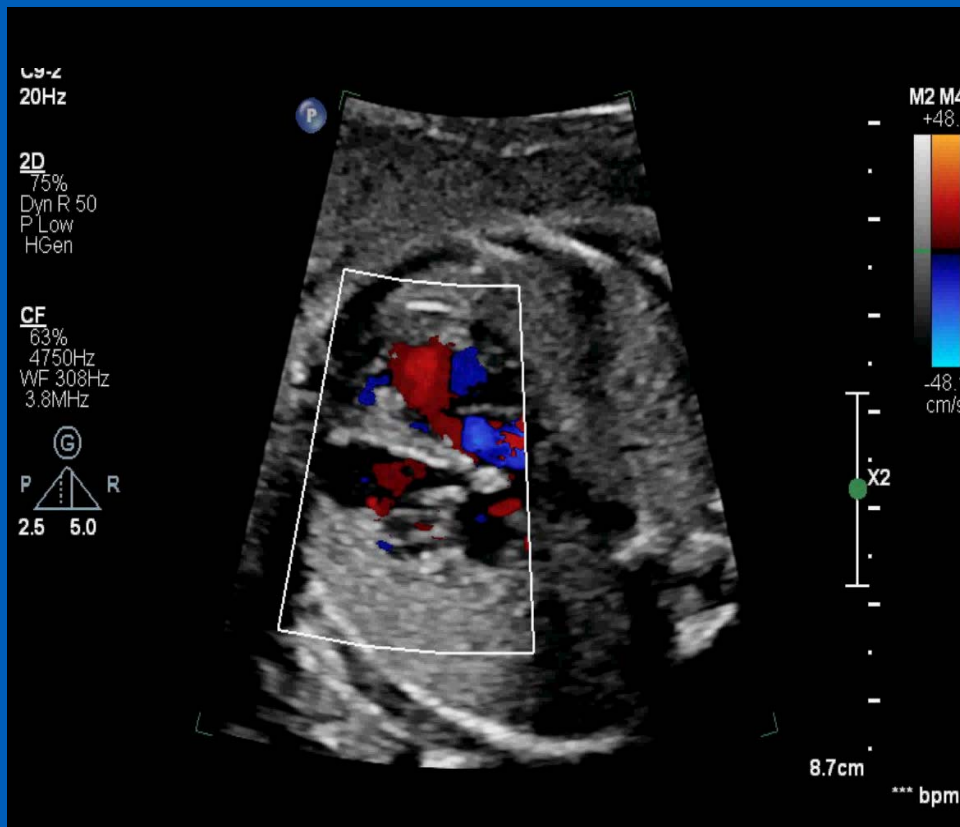
4 Chamber



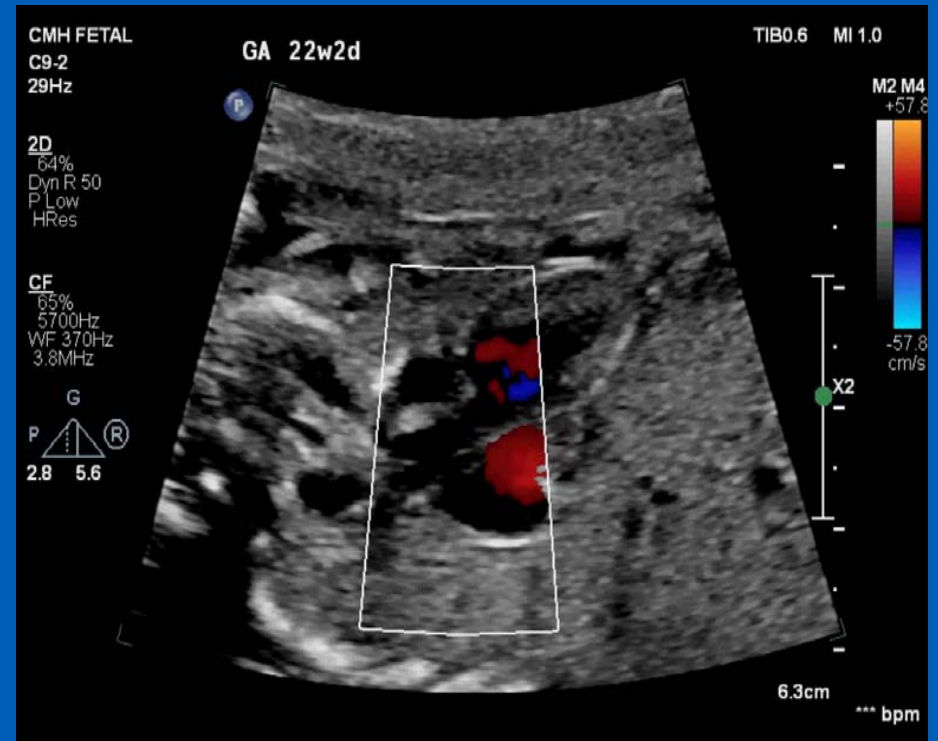
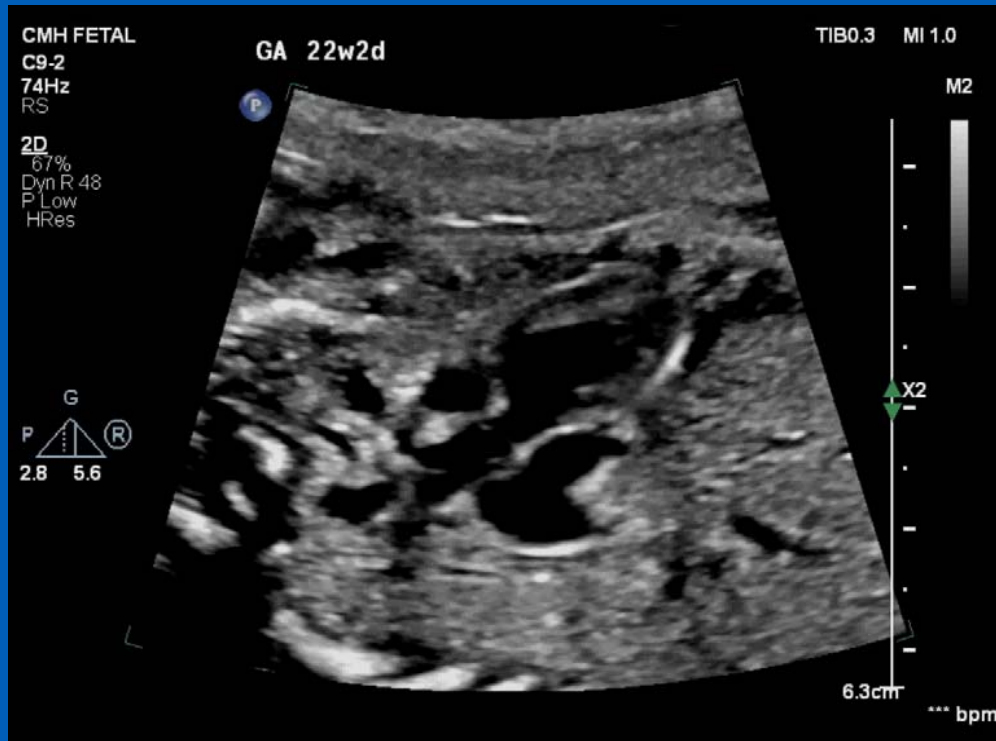
Coronal



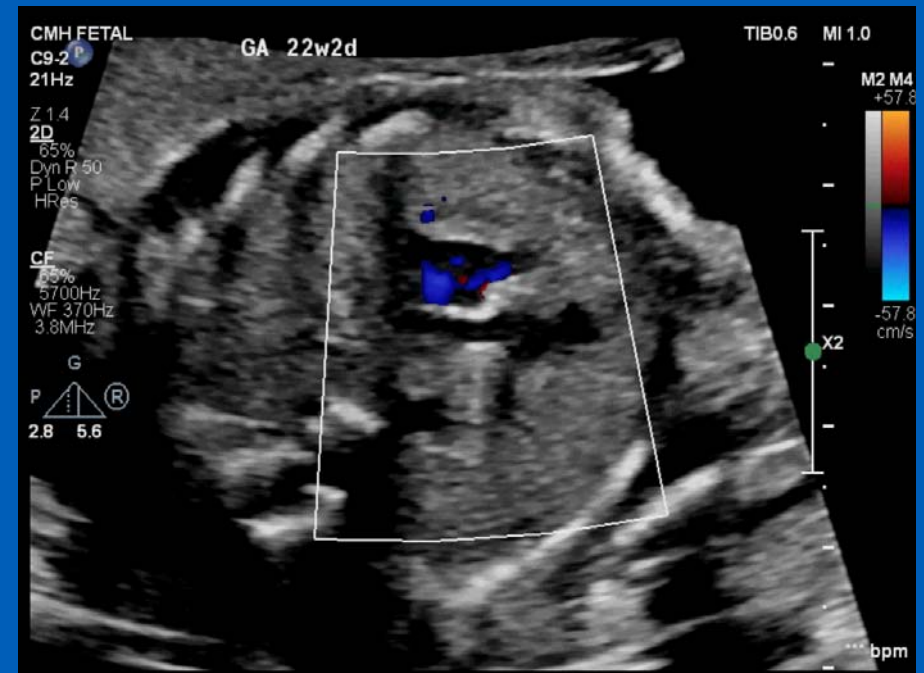
AV inflows



Left Ventricle to Pulmonary Artery



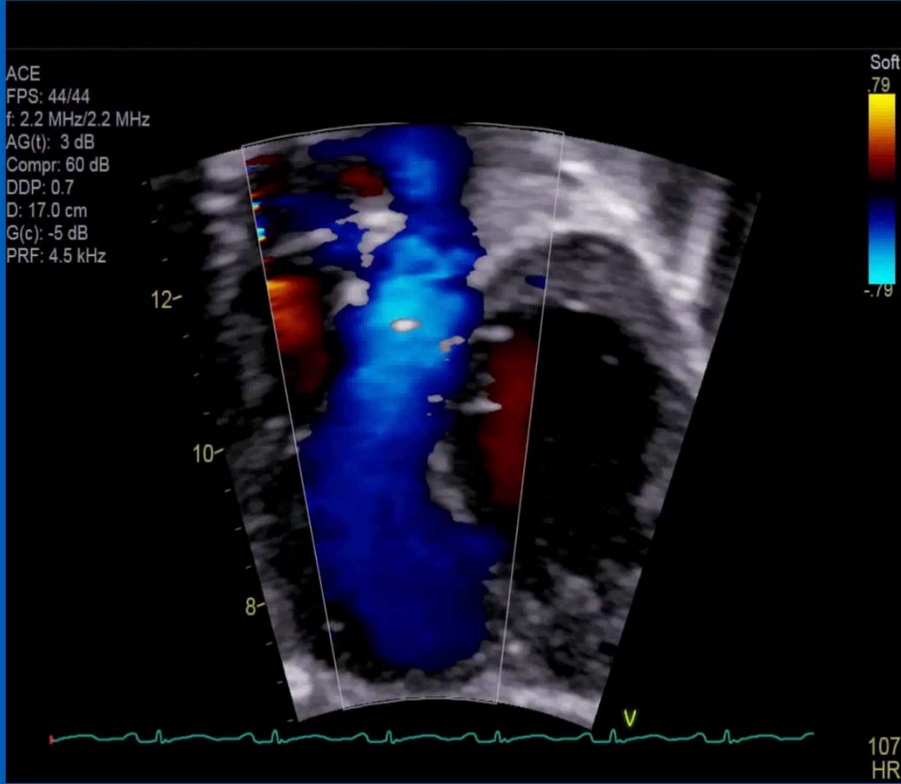
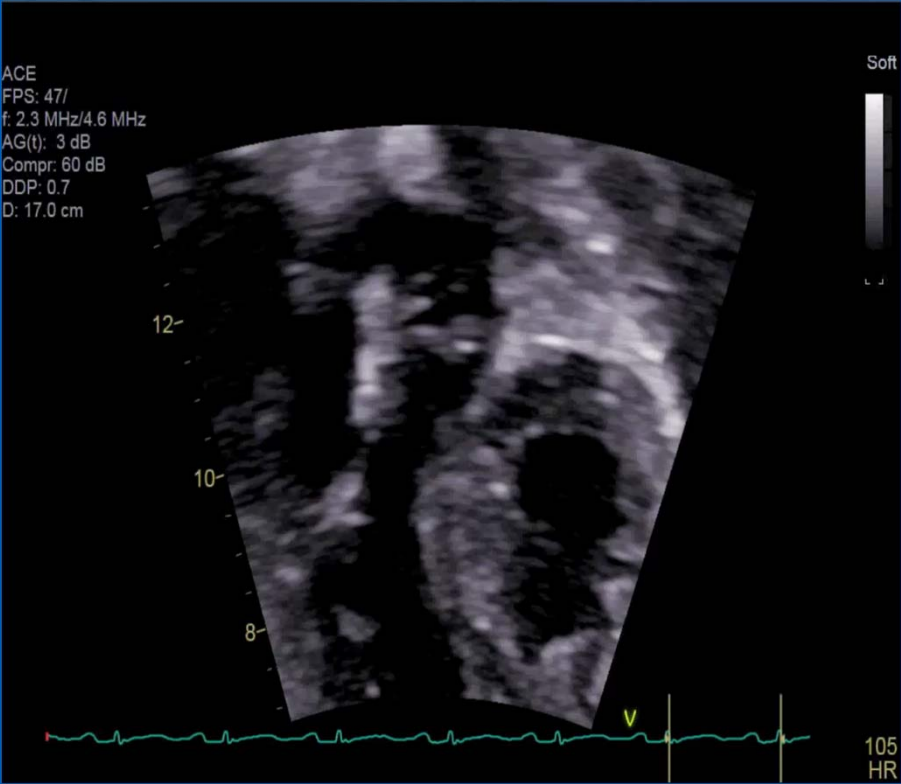
PA branches



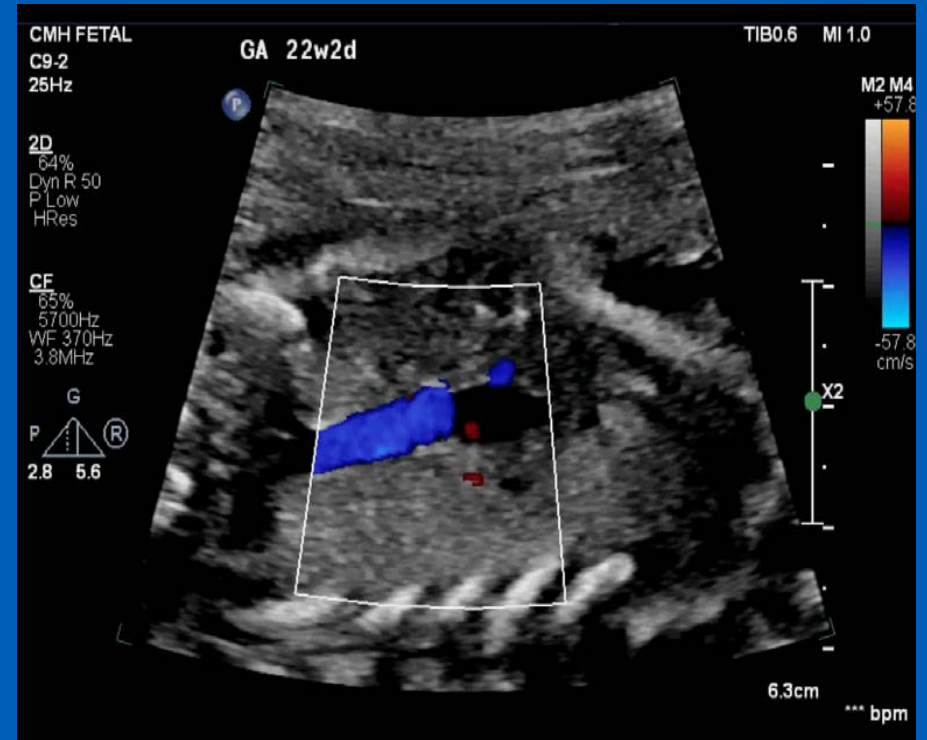
Dynamic and Fixed Pulmonary Outflow Obstruction



Postnatal LVOT/PA

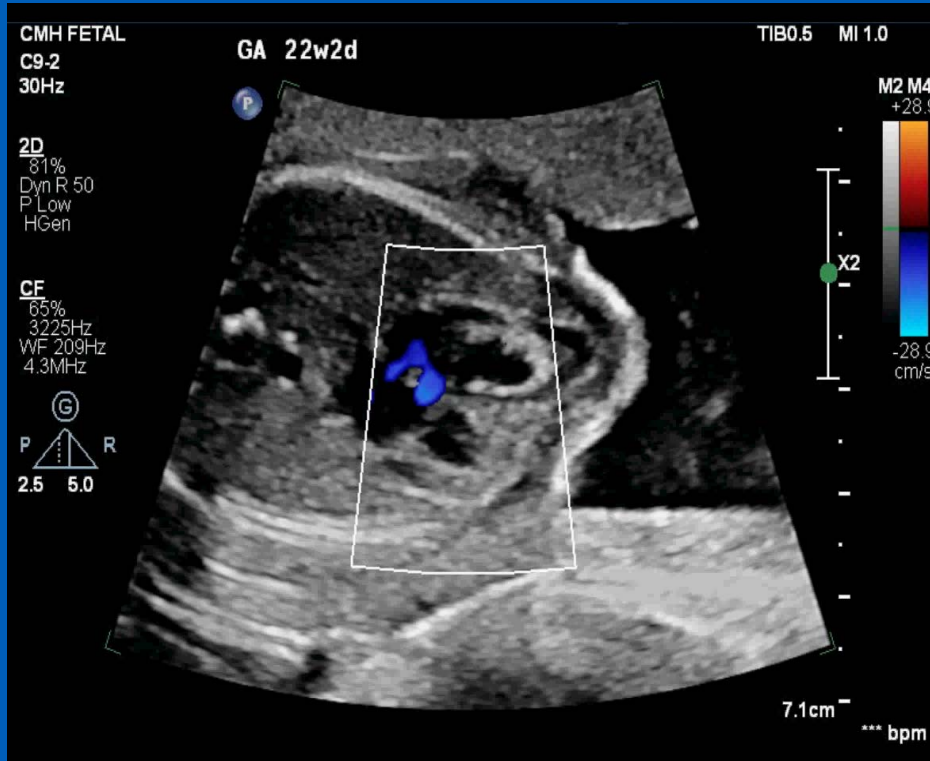


Right Ventricle to Aorta

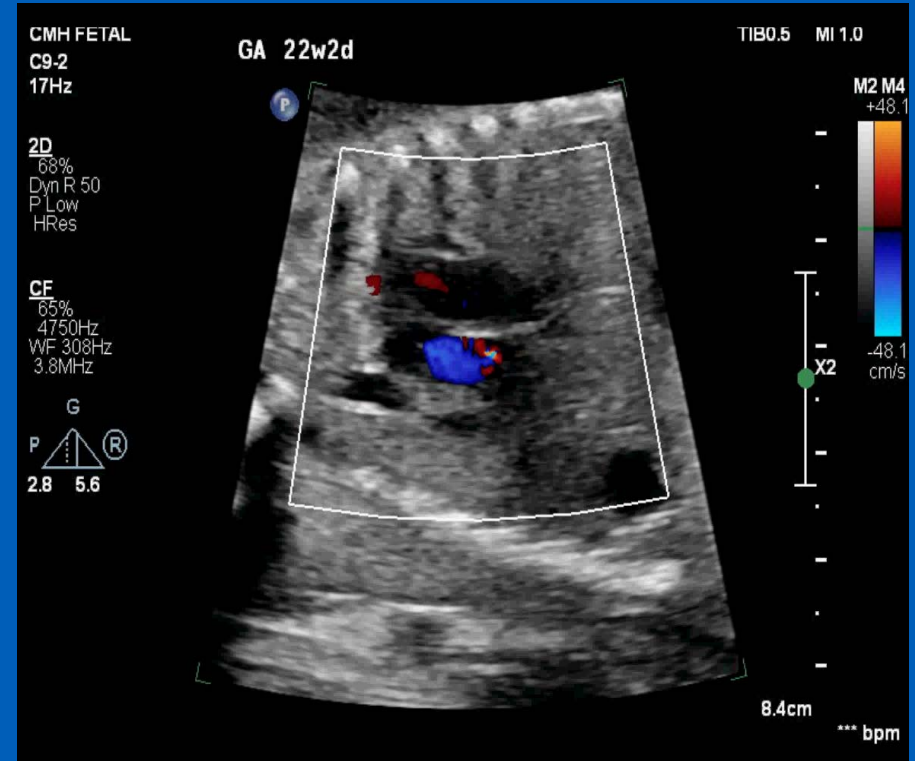


VSD Sweeps

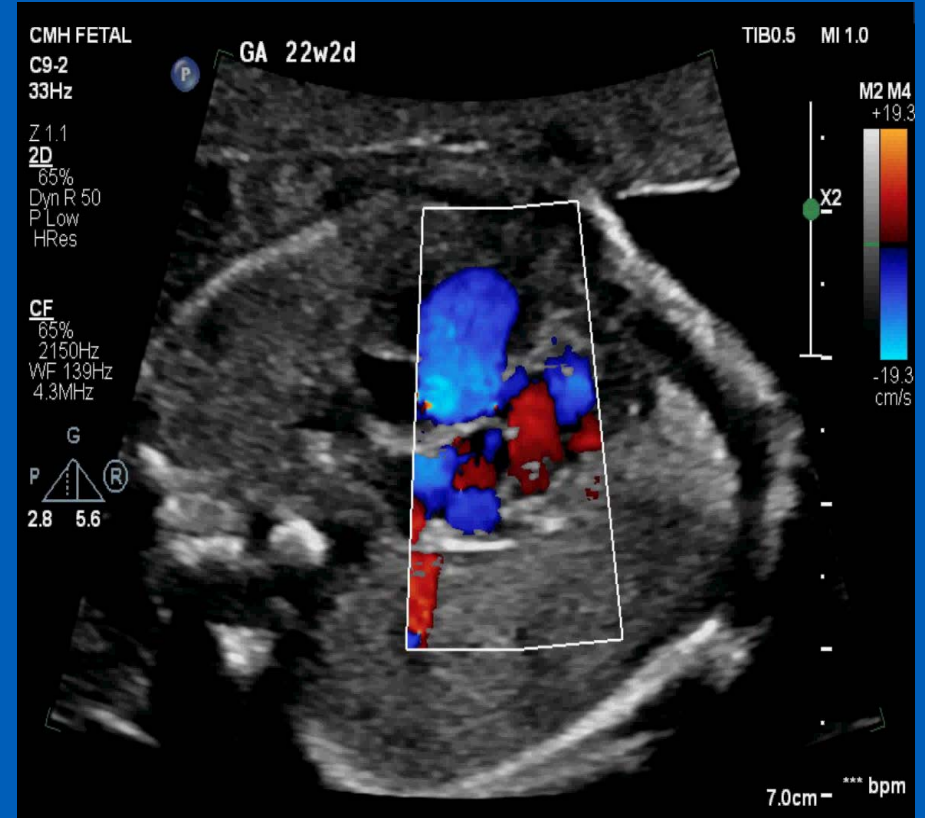
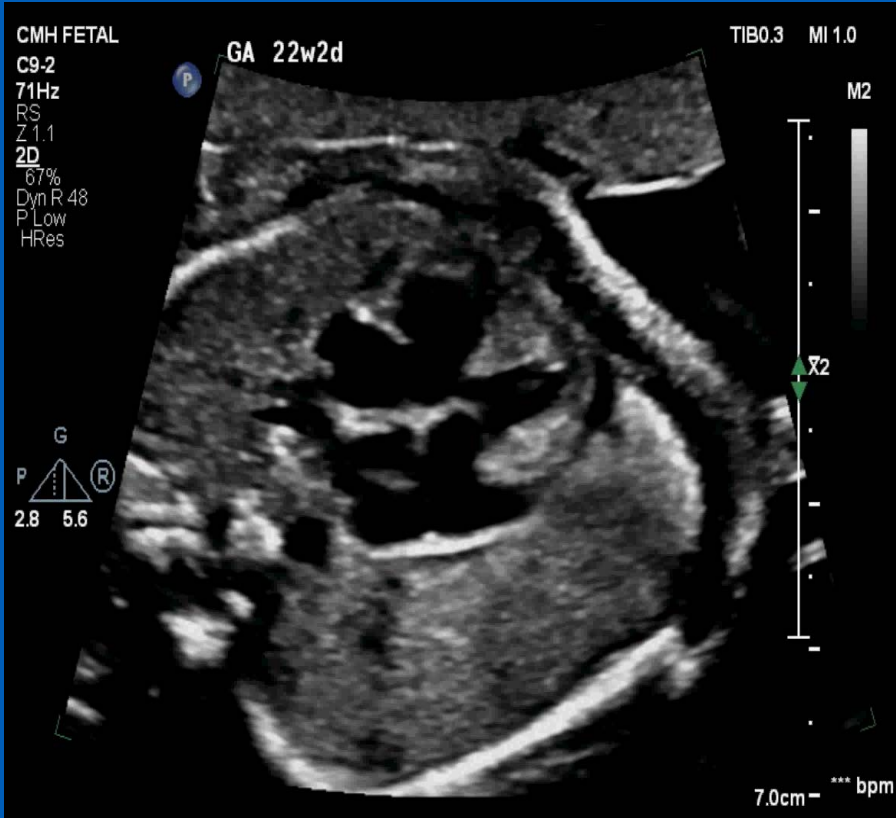
Coronal



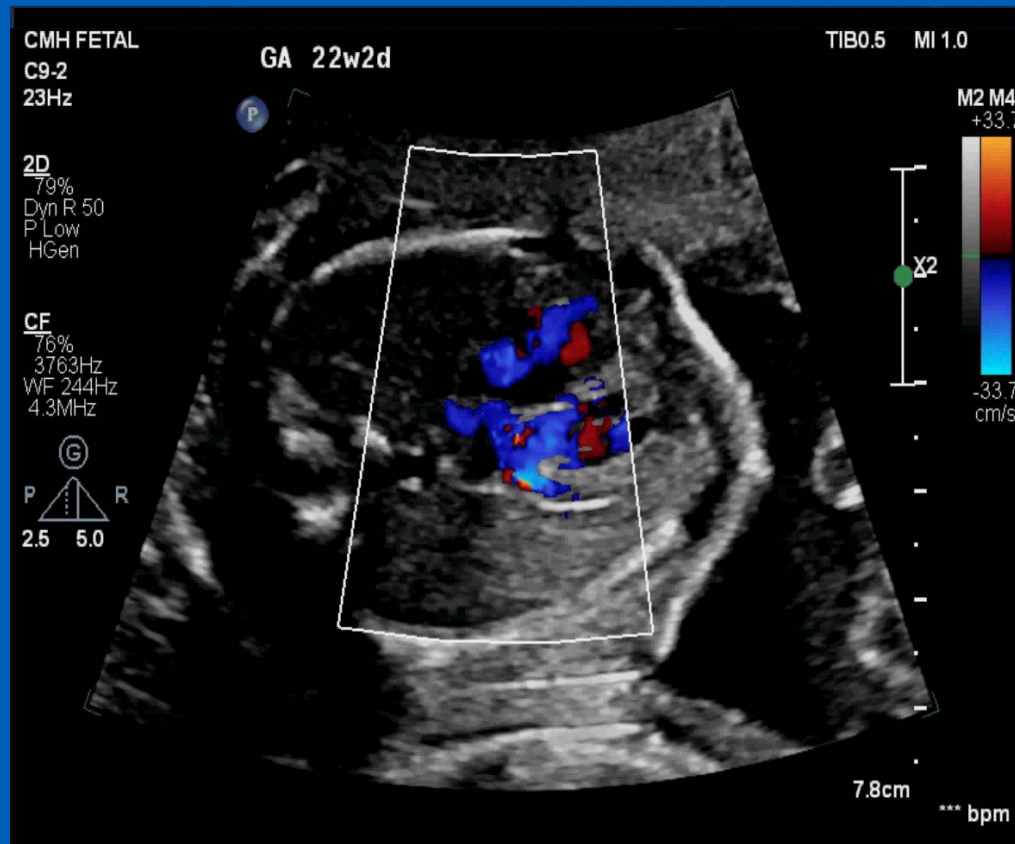
Short axis – Base to Apex



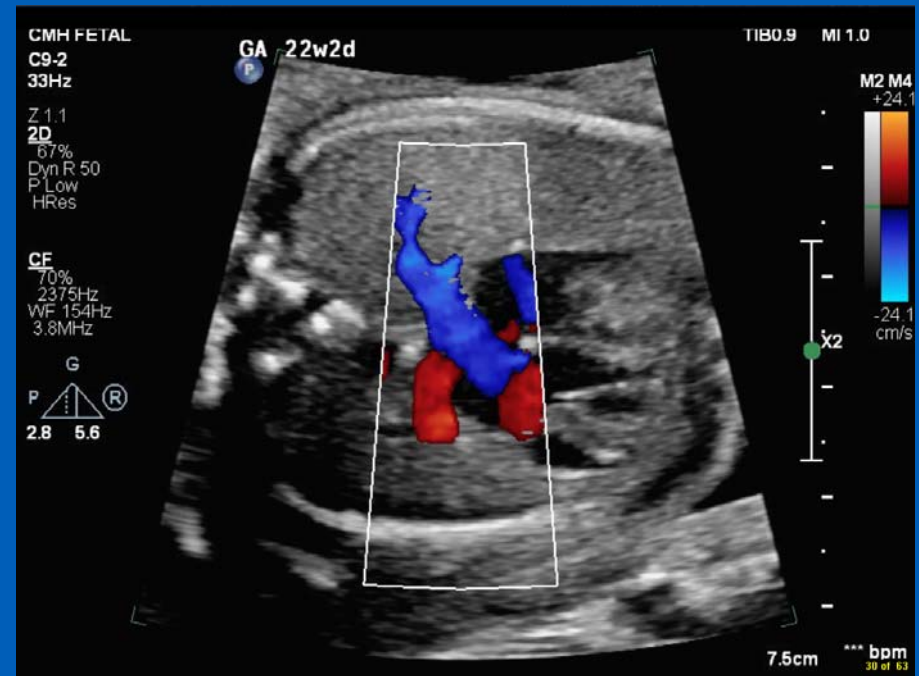
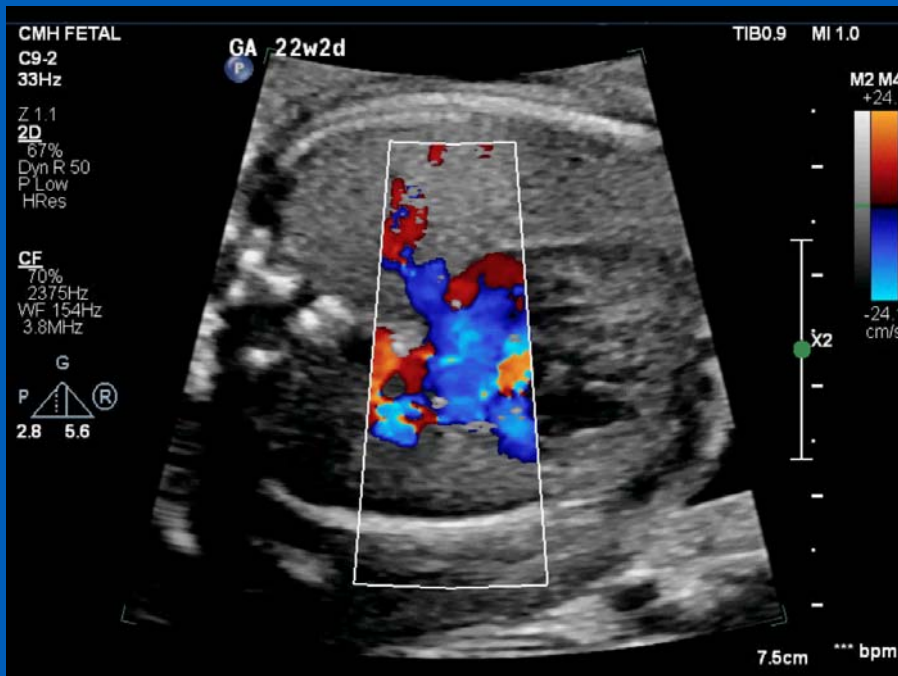
VSD



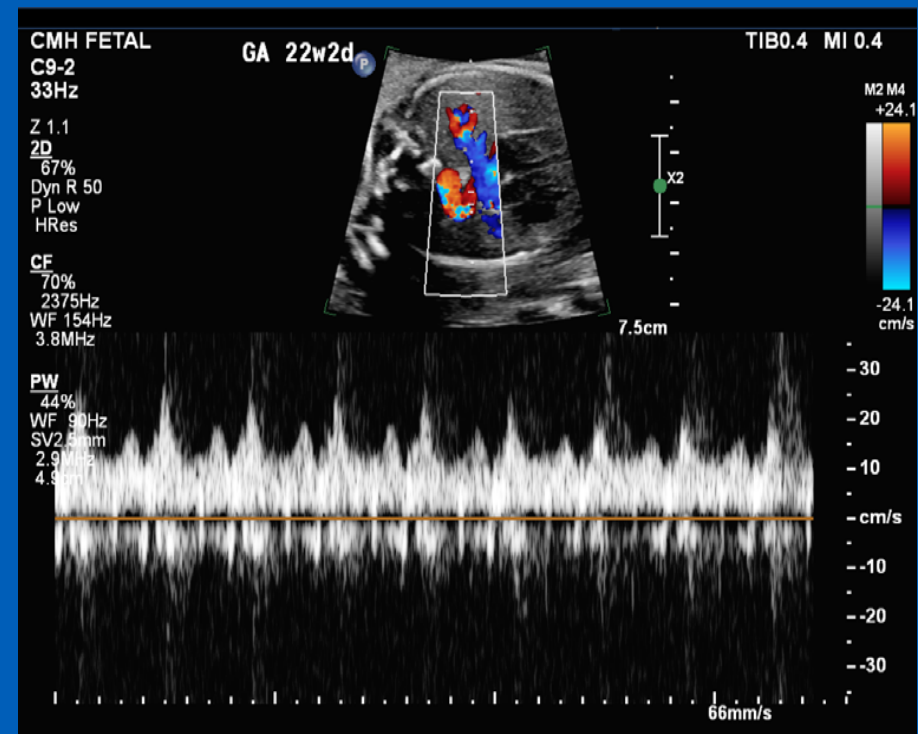
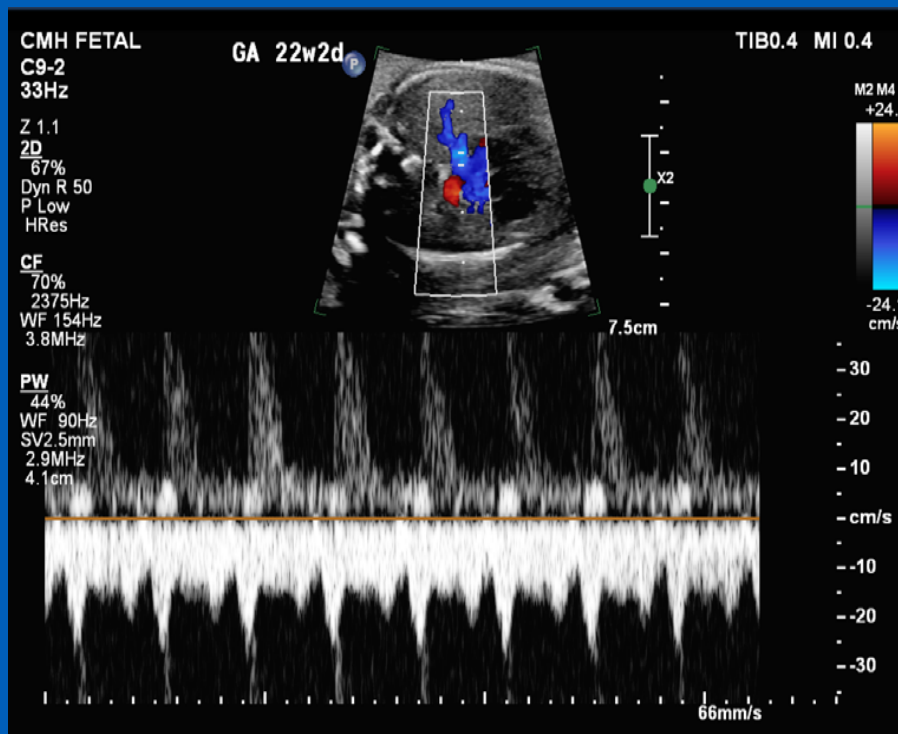
PFO



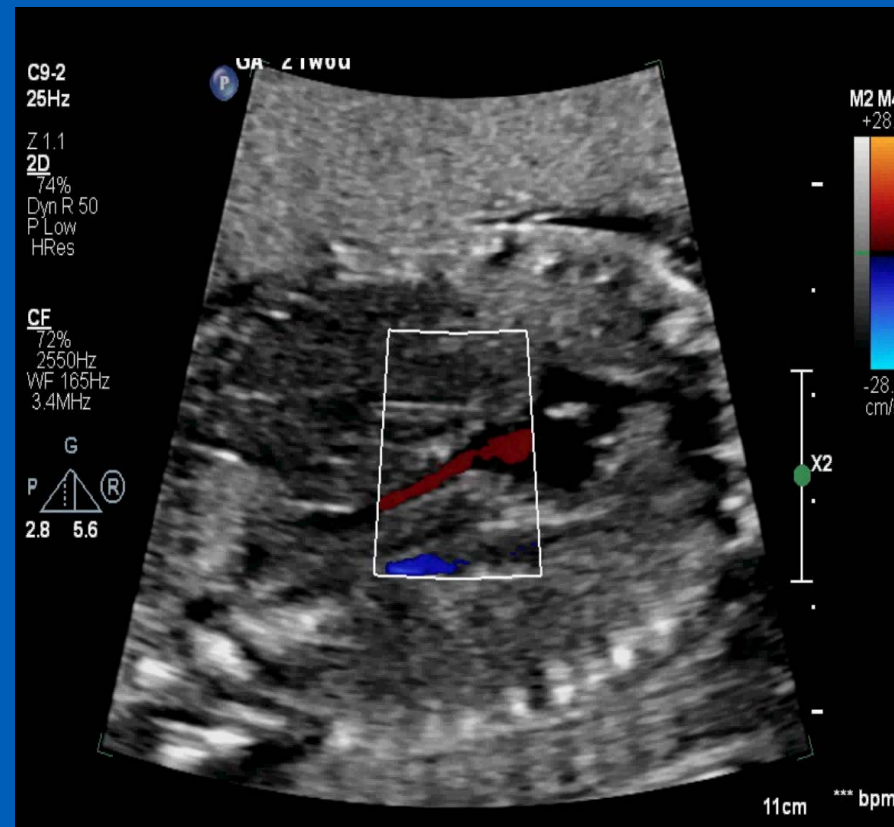
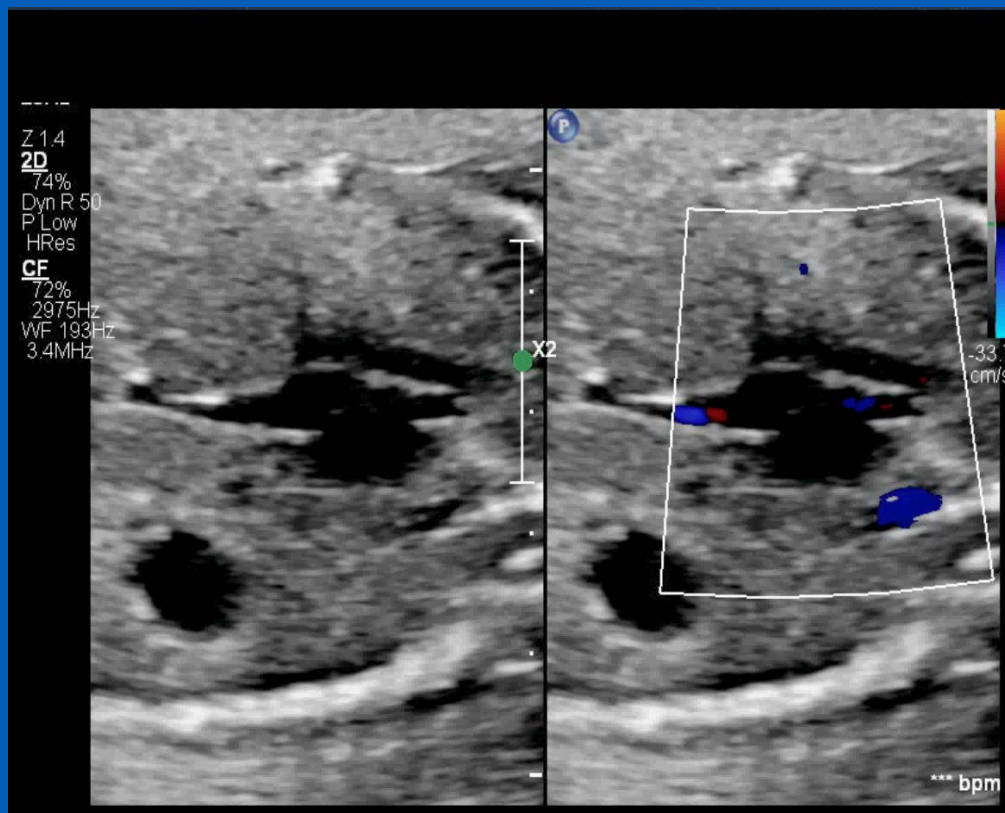
Pulmonary Veins to Left Atrium



Pulmonary Venous Dopplers



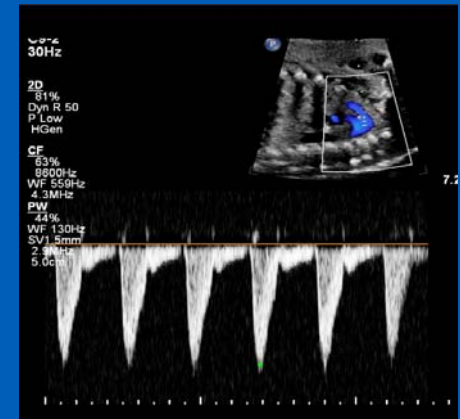
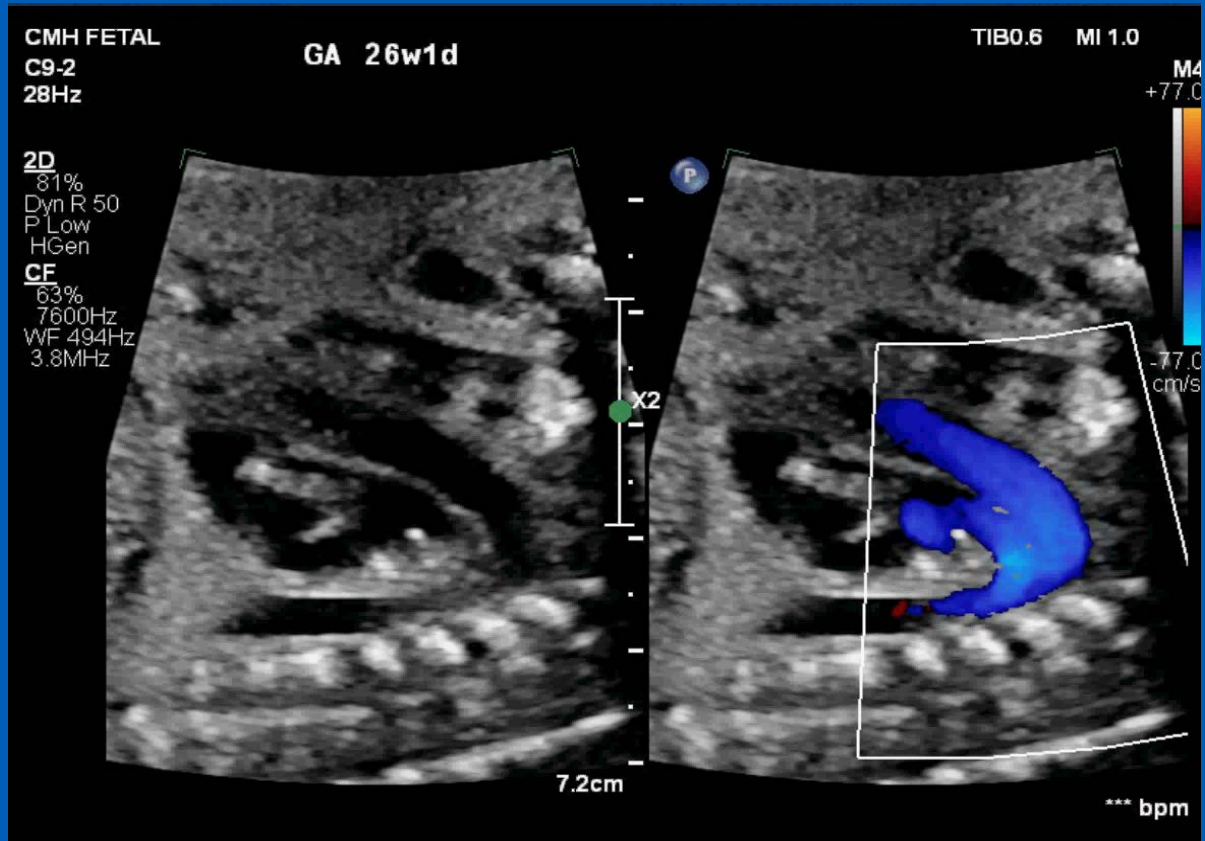
Bi Caval View with PFO



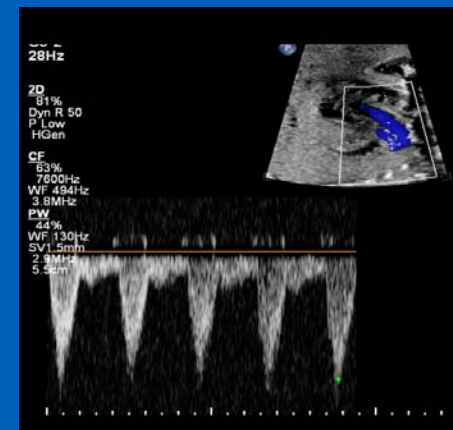
Aortic and ductal arches



Aortic and ductal arches

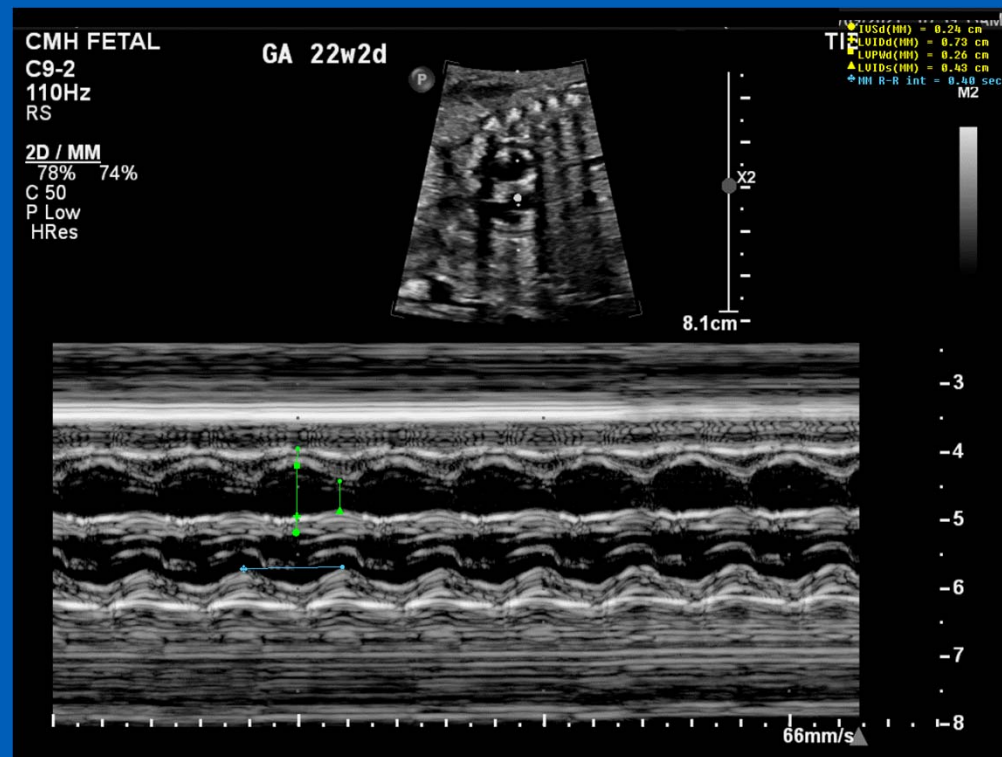


Isthmus

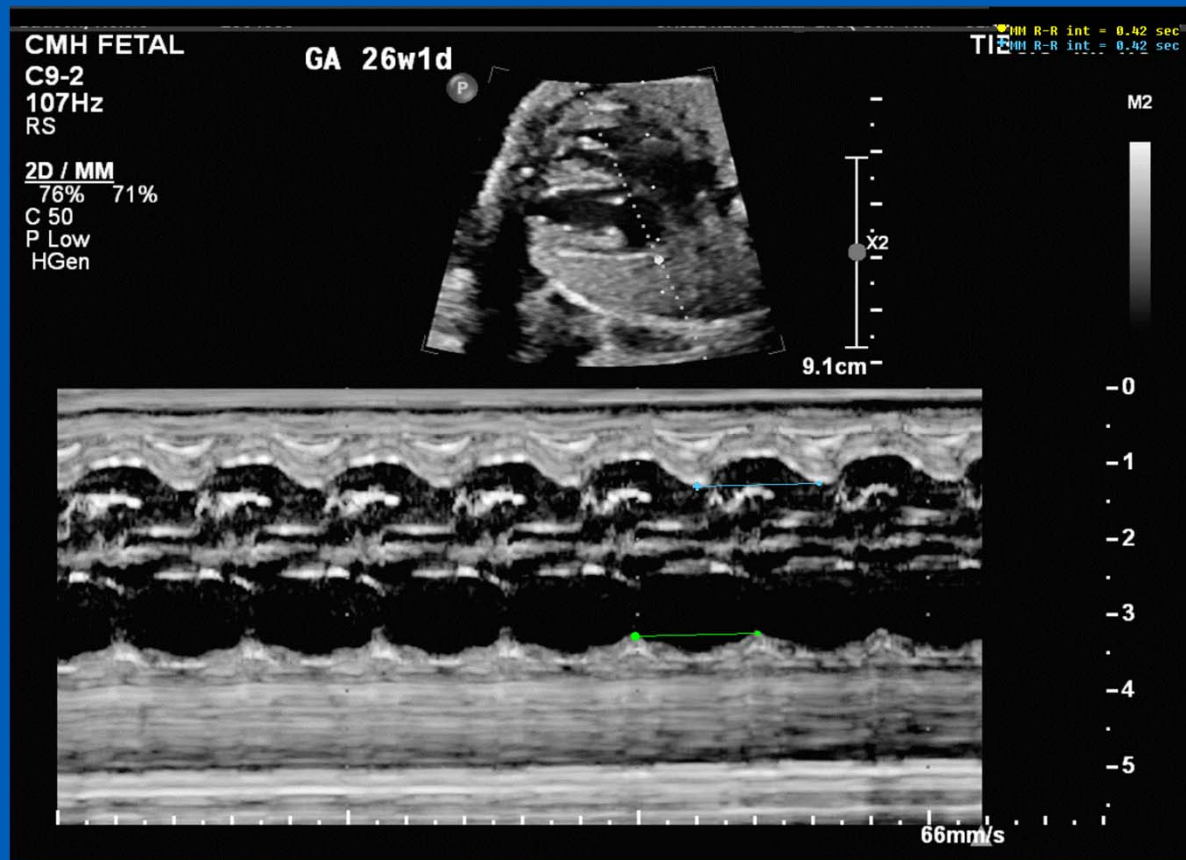


Ductus Arteriosus

M-Mode for heart rate and fractional shortening



AV Conduction



Summary

- Careful segmental evaluation of the cardiac structures and their relationships utilizing 2D, color flow and doppler imaging.
- Performing well-optimized sweeps through different planes with and without color helps to build the understanding of these connections
- Identify the presence of associated anomalies.



References

- Anderson RH, Becker AE, Freedom RM, Macartney FJ, Quero-Jimenez M, Shinebourne EA, Wilkinson JL, Tynan M. Sequential segmental analysis of congenital heart disease. *Pediatric Cardiology* 1984. 5 281–287. (10.1007/BF02424973) [[PubMed](#)] [[CrossRef](#)] [[Google Scholar](#)]
- Cohen et al. Congenitally Corrected Transposition of the Great Arteries: FetalDiagnosis, Associations, and Postnatal Outcome: A Fetal Heart SocietyResearch Collaborative Study. 6/2023 JAHA 2023;<https://doi.org/10.1161/JAHA.122.029706>
- Cordes et al. Distinguishing Right From Left: A Standardized Technique for Fetal Echocardiography. *JASE* Volume 7, Issue 1 1994 pages 47-53
- Congenitally Corrected Transposition of the Great Arteries, 1/2022 Adult Congenital Heart Association, <https://www.achaheart.org/your-heart/educational-qas/types-of-heart-defects/cctga/>
- Weinber, P. Anatomy and Classification of Heart Disease , <https://thoracickey.com/anatomy-and-classification-of-congenital-heart-disease/>
- Amikam S, Lemer J, Kishon Y, Riss E, Neufeld HN. Complete heart block in an adult with corrected transposition of the great arteries treated with permanent pacemaker. *Thorax*. 1979;34:547–9. [[PMC free article](#)] [[PubMed](#)] [[Google Scholar](#)]



Suggested Reading

Journal of the American Heart Association












Volume 12, Issue 11, 6 June 2023

<https://doi.org/10.1161/JAHA.122.029706>



ORIGINAL RESEARCH

Congenitally Corrected Transposition of the Great Arteries: Fetal Diagnosis, Associations, and Postnatal Outcome: A Fetal Heart Society Research Collaborative Study

Jennifer Cohen, MD ; Bhawna Arya, MD; Richard Caplan, PhD ; Mary T. Donofrio, MD ; Dina Ferdman, MD; Jamie K. Harrington, MD; Deborah Y. Ho, MD, MPH ; Whitnee Hogan, MD; Lisa K. Hornberger, MD ; Simone Jhaveri, MD; Stacy A. S. Killen, MD, MSCI; Christopher L. Lindblade, MD ; Erik Michelfelder, MD ; Anita J. Moon-Grady, MD ; Sheetal Patel, MD, MSCI ; Emilio Quezada, MD; Christina Ronai, MD, MEd ; Aura A. Sanchez Mejia, MD ; David N. Schidlow, MD, MMus; Corey Stiver, MD; Varsha Thakur, MD, MSc; Shubhika Srivastava, MD



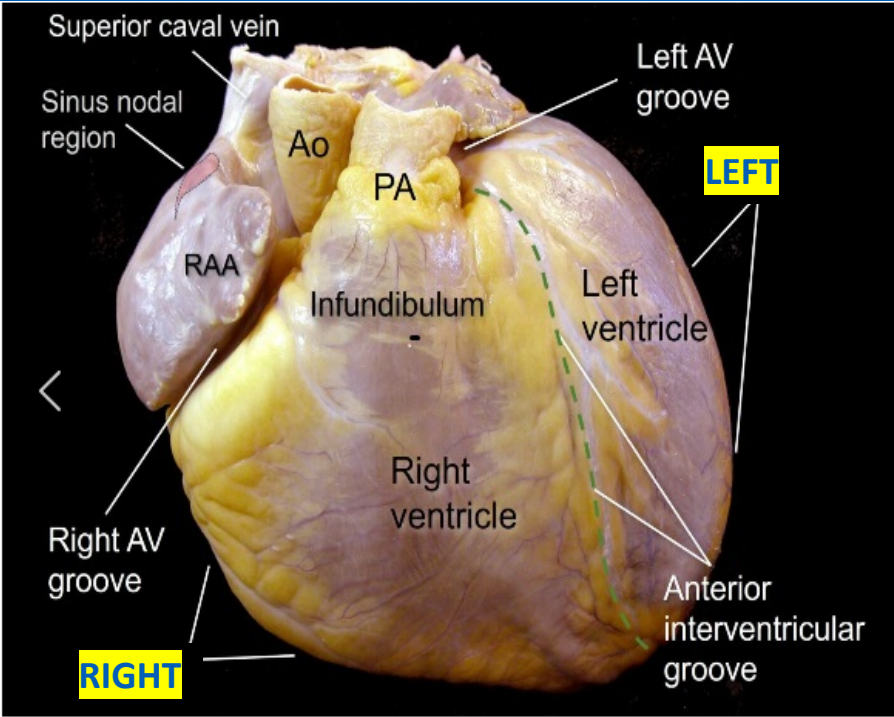
References

- Anderson RH, Becker AE, Freedom RM, Macartney FJ, Quero-Jimenez M, Shinebourne EA, Wilkinson JL, Tynan M. Sequential segmental analysis of congenital heart disease. *Pediatric Cardiology* 1984. 5 281–287. (10.1007/BF02424973) [[PubMed](#)] [[CrossRef](#)] [[Google Scholar](#)]
- Cohen et al. Congenitally Corrected Transposition of the Great Arteries: FetalDiagnosis, Associations, and Postnatal Outcome: A Fetal Heart SocietyResearch Collaborative Study. 6/2023 JAHA 2023;<https://doi.org/10.1161/JAHA.122.029706>
- Cordes et al. Distinguishing Right From Left: A Standardized Technique for Fetal Echocardiography. *JASE* Volume 7, Issue 1 1994 pages 47-53
- Congenitally Corrected Transposition of the Great Arteries, 1/2022 Adult Congenital Heart Association, <https://www.achaheart.org/your-heart/educational-qas/types-of-heart-defects/cctga/>
- Weinber, P. Anatomy and Classification of Heart Disease , <https://thoracickey.com/anatomy-and-classification-of-congenital-heart-disease/>
- Amikam S, Lemer J, Kishon Y, Riss E, Neufeld HN. Complete heart block in an adult with corrected transposition of the great arteries treated with permanent pacemaker. *Thorax*. 1979;34:547–9. [[PMC free article](#)] [[PubMed](#)] [[Google Scholar](#)]

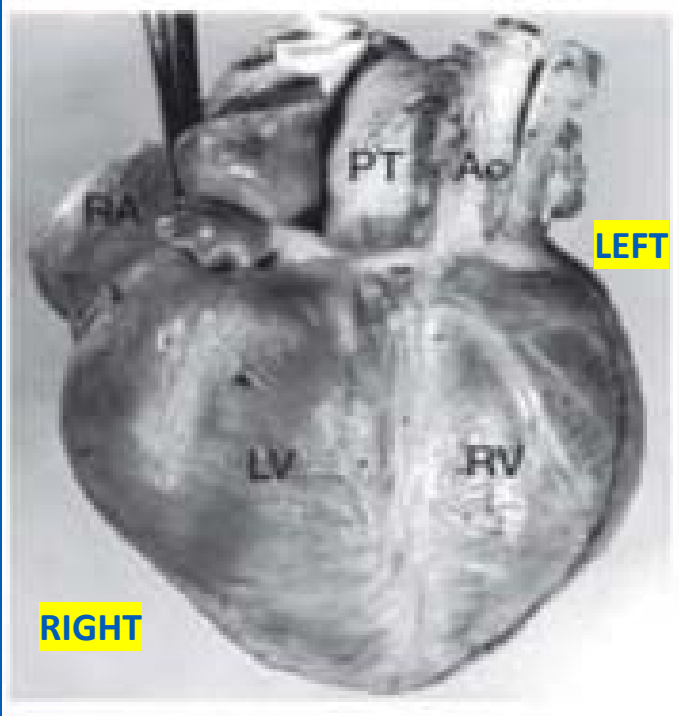




Normal vs CCTGA



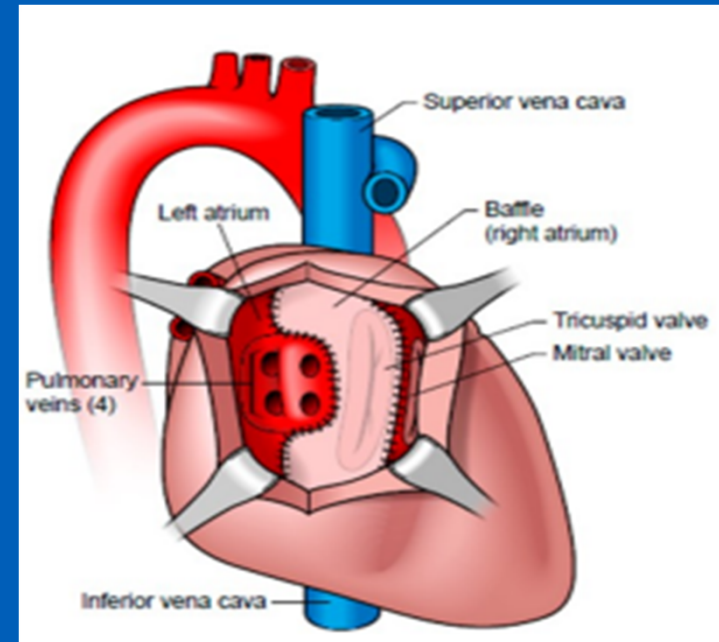
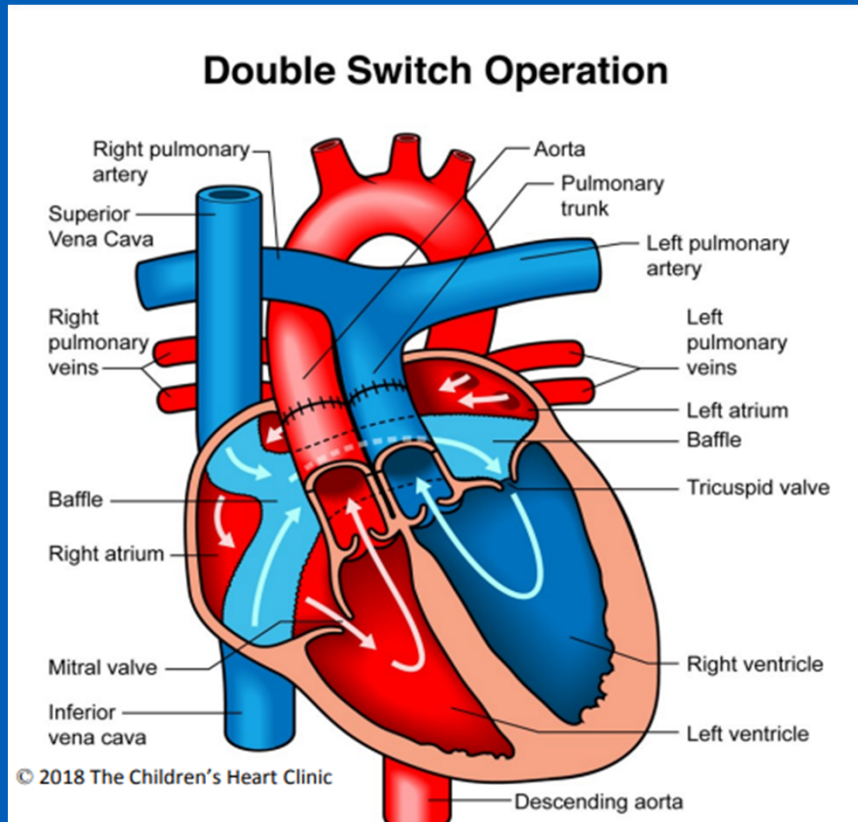
NORMAL



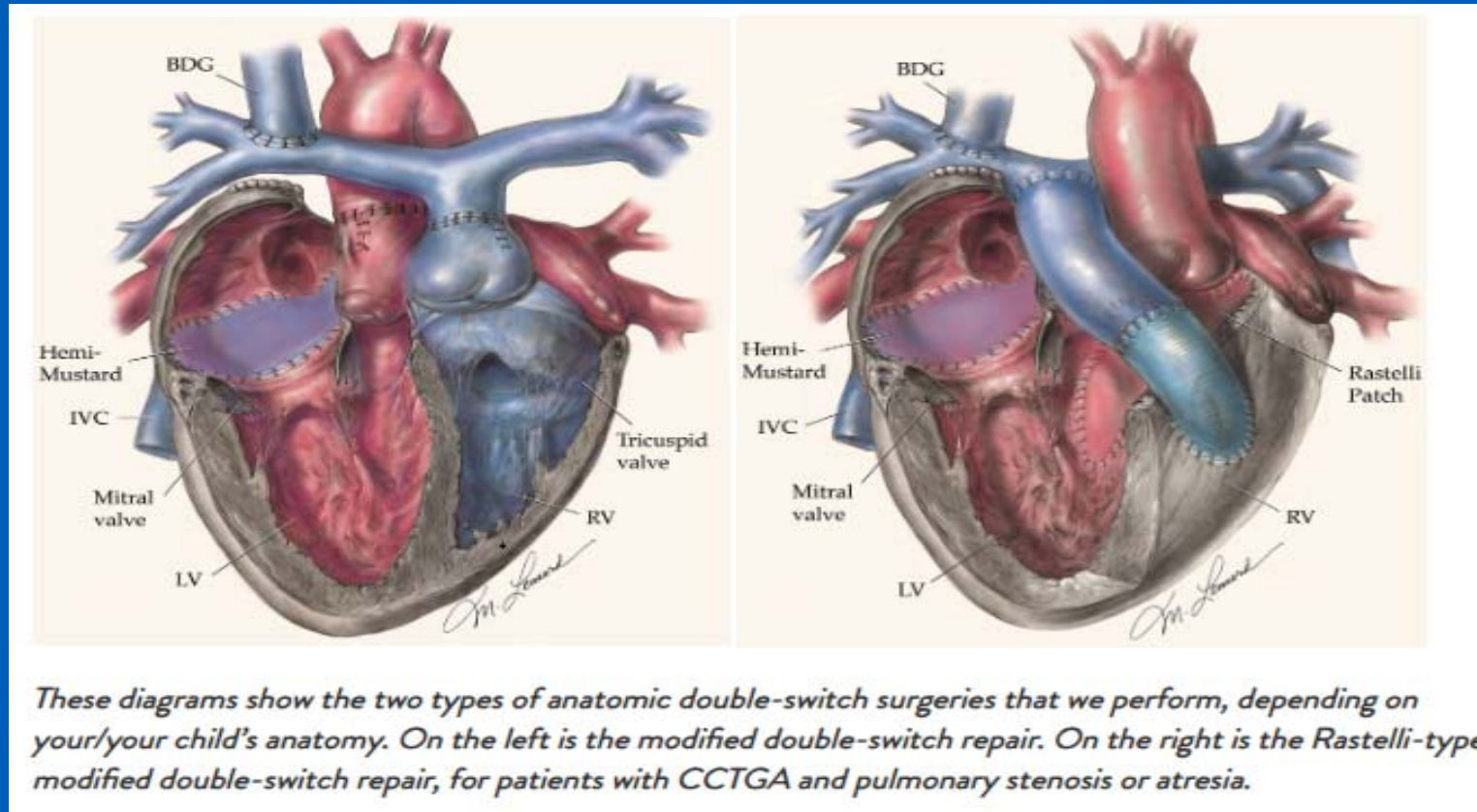
CCTGA



Double Switch



Modified Double Switch



Segmental Comparison

