

The 2nd Contemporary Morphology Course
Congenital Heart Disease in Your Hands
December 6-7, 2019

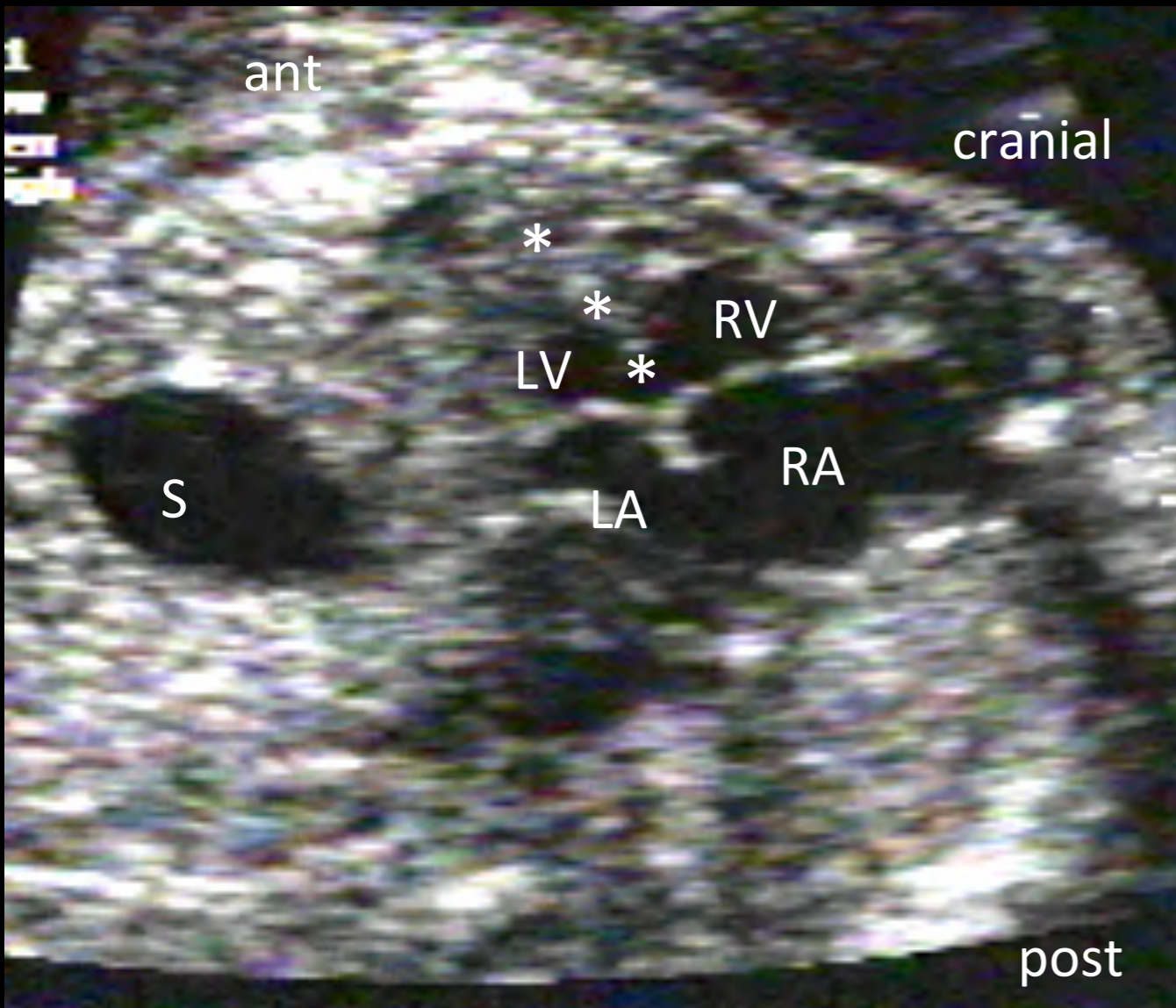
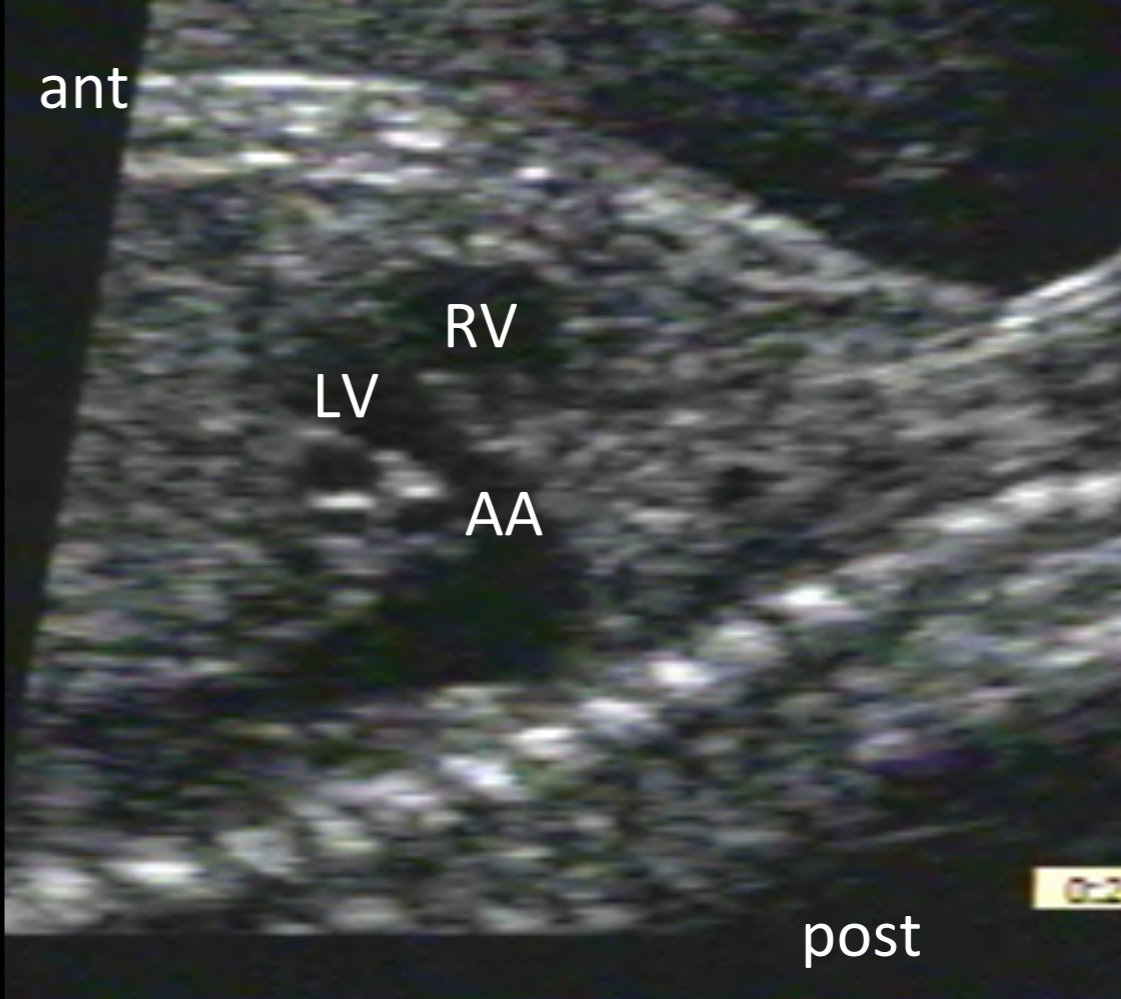
TWISTED, CRISS-CROSS,
SUPERIOR-INFERIOR, TOPSY-TURVY, ETC.
WHAT DO THEY ALL MEAN?

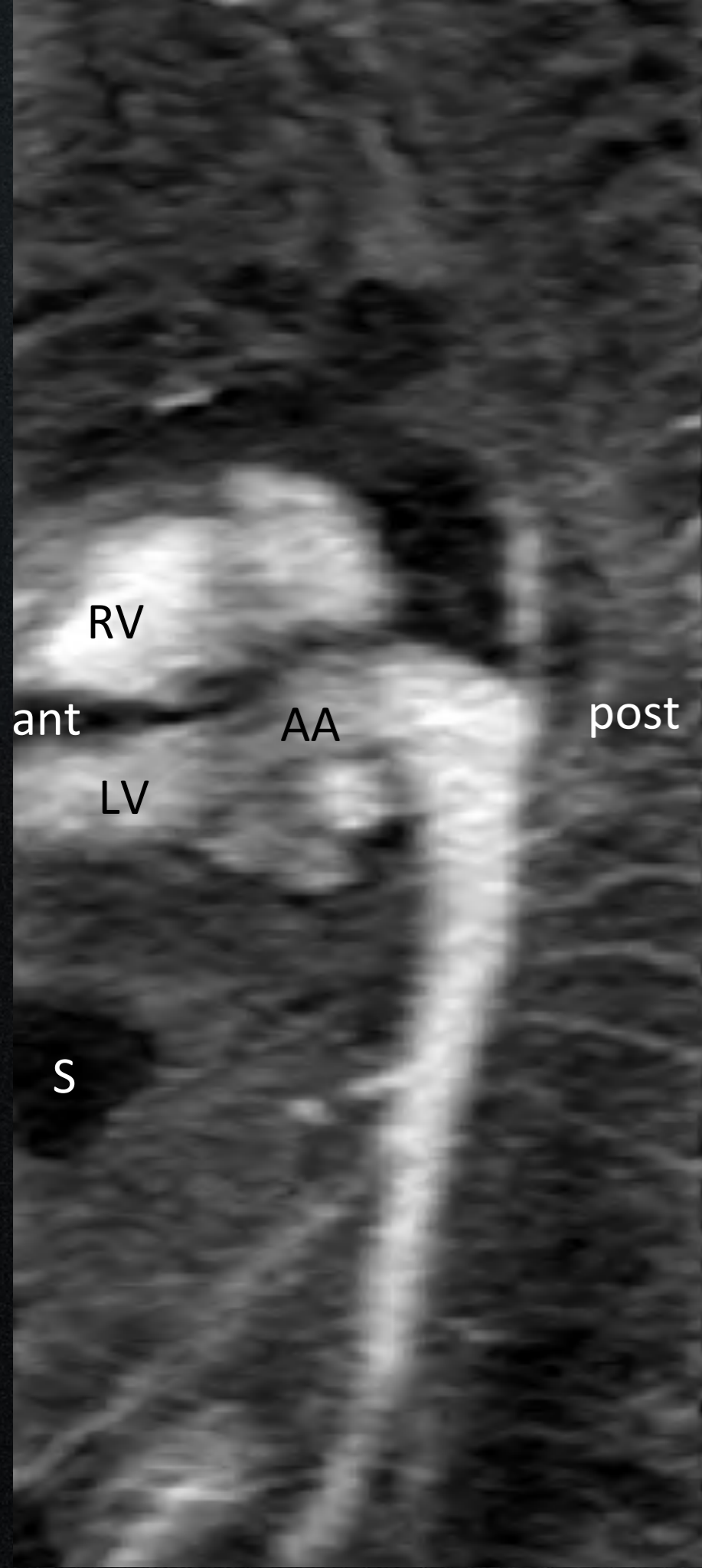
Shi-Joon Yoo, MD

Department of Diagnostic Imaging
Division of Cardiology, Department of Paediatrics
Division of Cardiovascular Surgery, Department of Surgery
University of Toronto

Canada

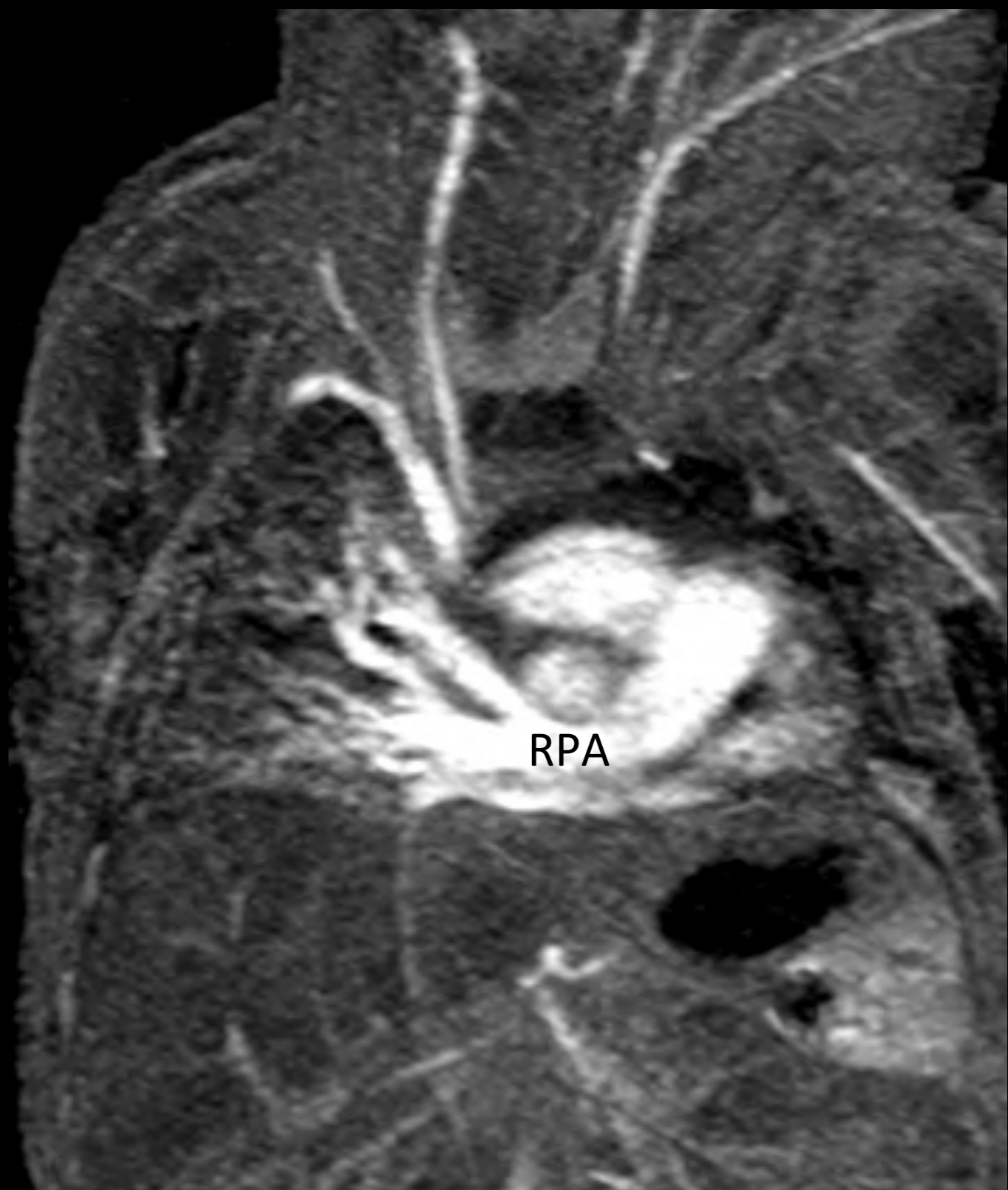
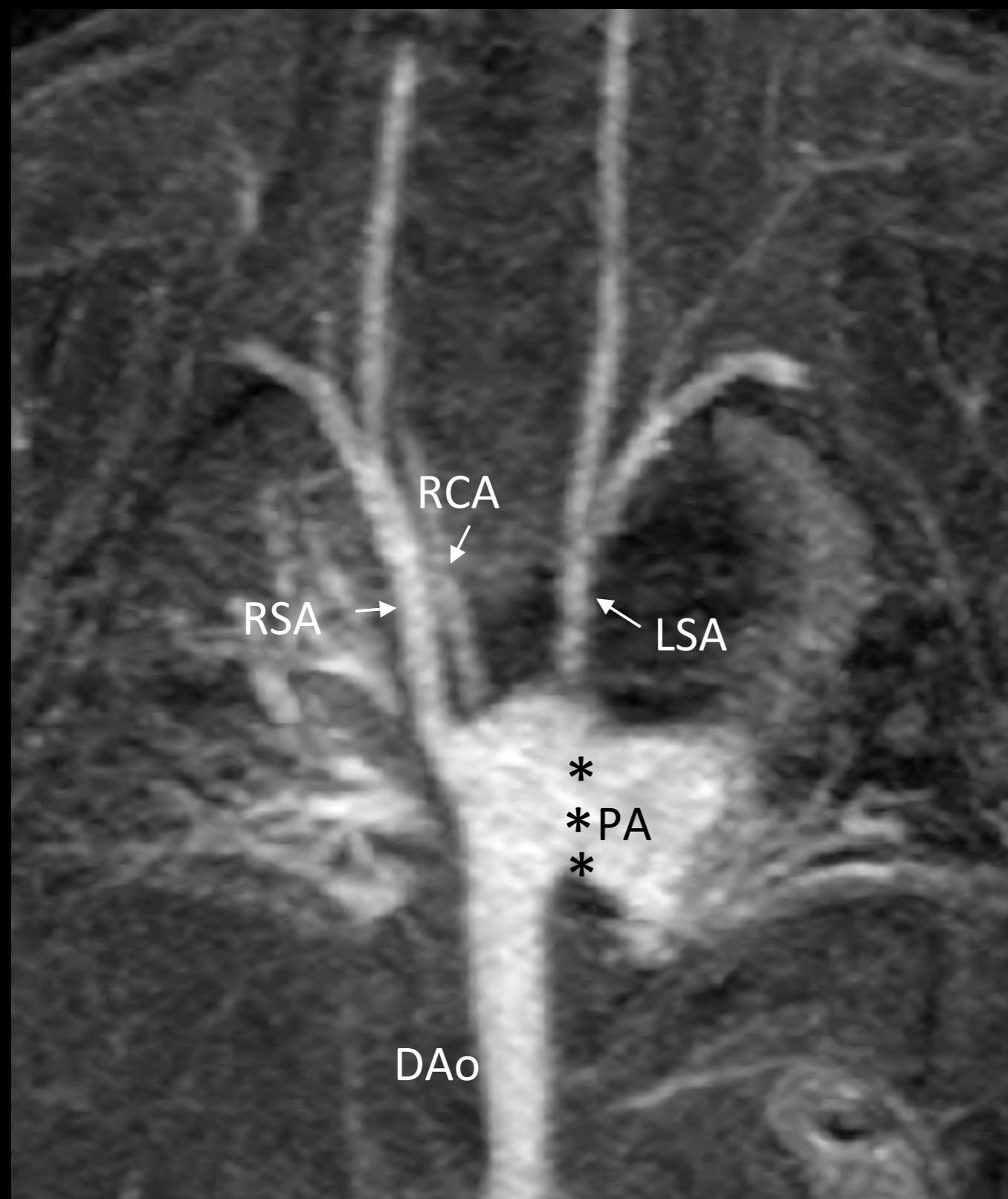
SickKids

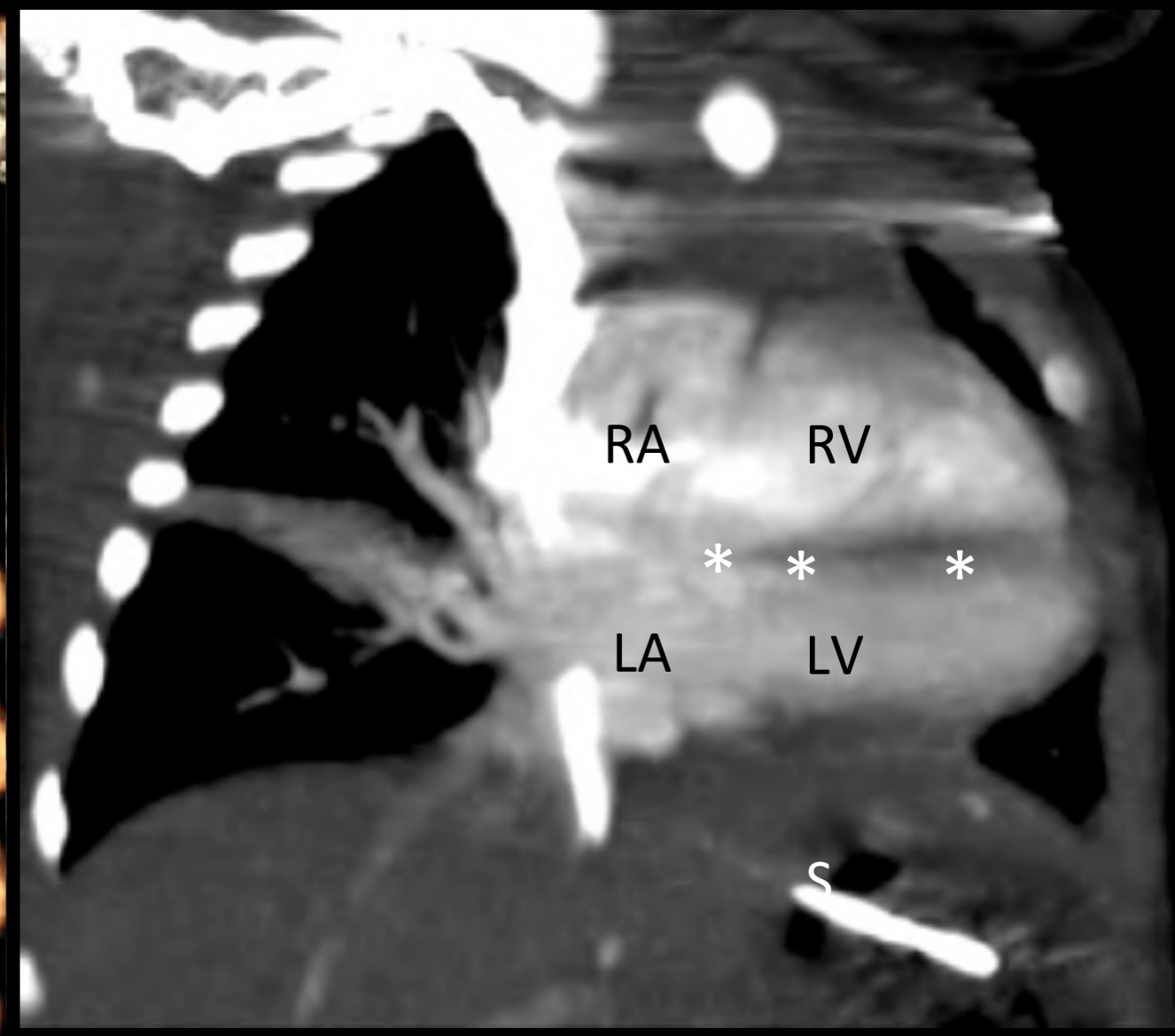
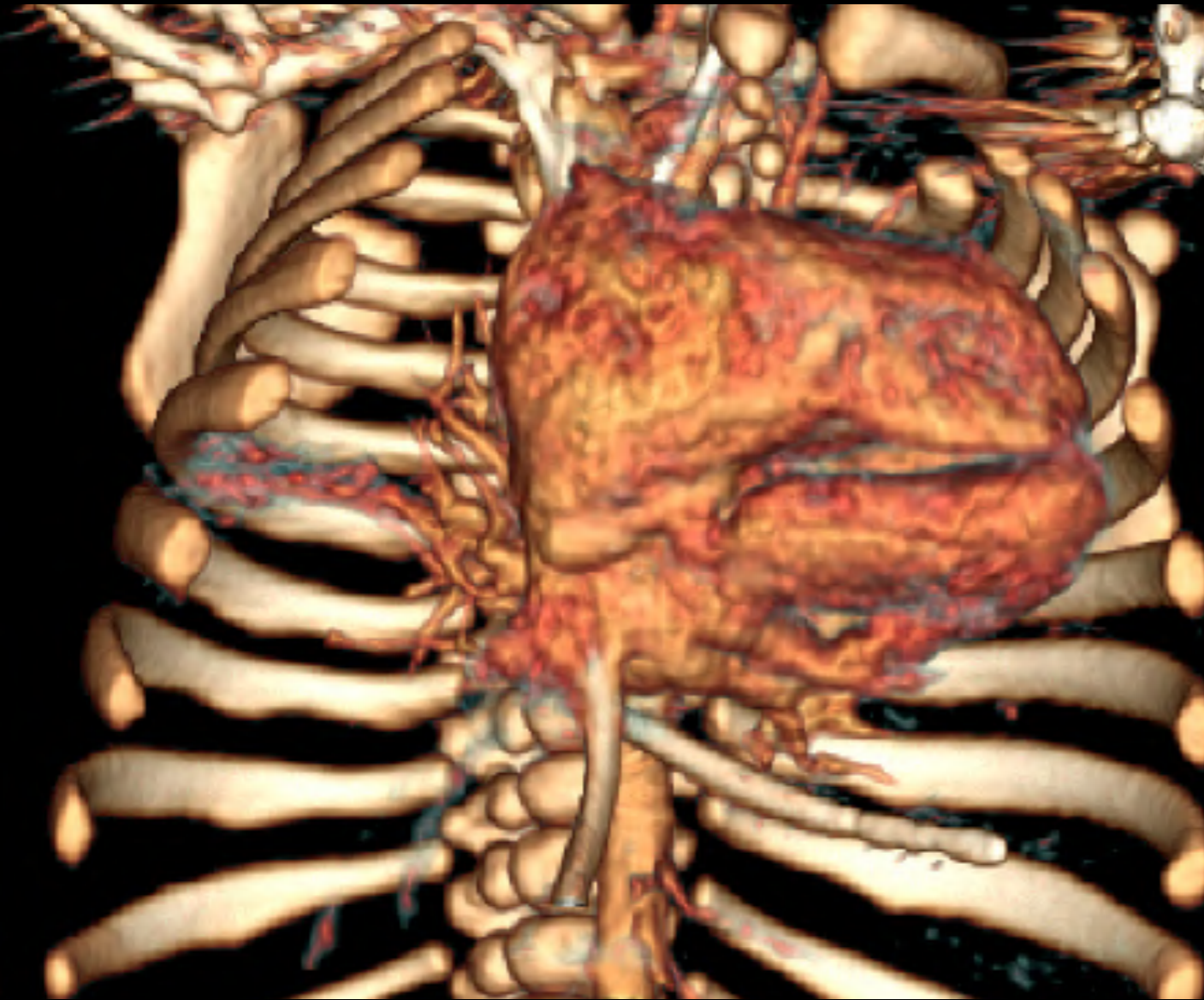


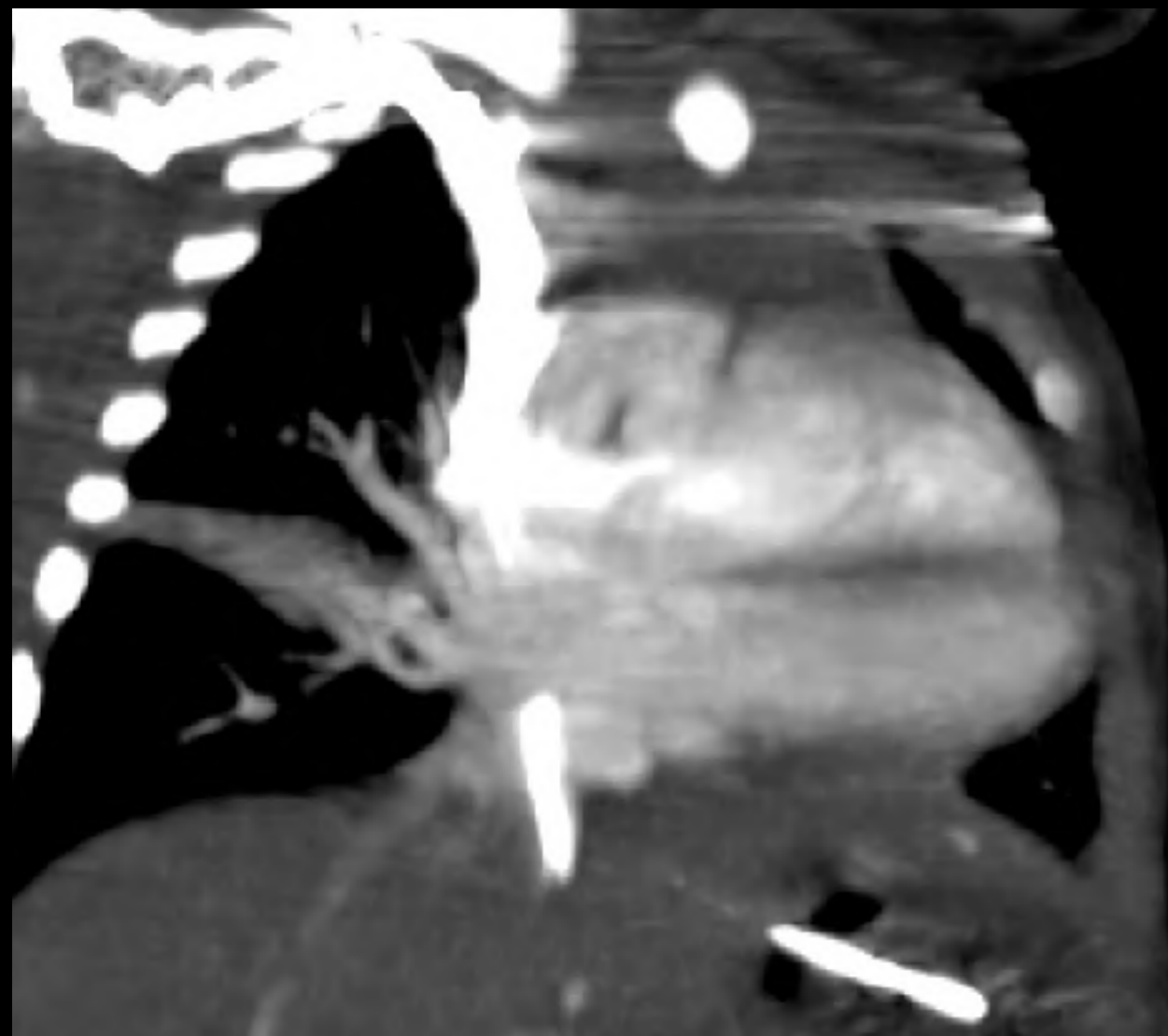


“What is unique in “topsy-turvy” hearts is the bizarre topography of the arterial outlets. The arterial pole of the heart is rotated posteroinferiorly. Thus, the great arteries seem to exit from the diaphragmatic aspect of the ventricular mass. This peculiar postseptational anomaly results in marked elongation of the brachio-cephalic arteries.”

Robert M. Freedom, Gordon Culham, and Fred Moes.
In *Angiography of Congenital Heart Disease*,
Macmillan Publishing Co., 1994







Topsy-Turvy Heart

The Paediatric Cardiology Hall of Fame

Robert Mark Freedom MD, FRCPC, FACC, O. Ont

Lee N. Benson,¹ Robert H. Anderson²

¹Division of Cardiology, The Hospital for Sick Children, The University of Toronto School of Medicine, Toronto, Ontario, Canada; ²Cardiac Unit, Institute of Child Health, University College, London, United Kingdom

"I can resist everything except temptation."
Oscar Wilde (1854-1900). Lady Windermere's Fan, act 1 (1893).

IN OCTOBER 2000, BOB FREEDOM RETIRED AS THE head of the Division of Cardiology at the Hospital for Sick Children, having served 3 five-year terms (Figs 1, 2). He had succeeded the late Richard D Rowe in 1986, carrying forward a tradition of clinical and academic excellence fostered by John Keith, the first head of the division. Bob joined the staff at Sick Kids in July 1974, when Dick Rowe, who had just become the head of cardiology in Toronto, enticed him to leave Johns Hopkins Hospital in Baltimore, where he was the director of the pediatric diagnostic cardiovascular laboratory, and developed the cardiovascular pathology registry. Just prior to moving to Toronto, Dick himself had been the director of the unit of pediatric cardiology at Johns Hopkins, having succeeded Helen Taussig, and recognized early on that Bob was far from the average young paediatric cardiologist just out from training. Indeed, Bob had already published 20 peer-reviewed papers, spanning topics from angiography and clinical outcomes to anatomy. In 1974, he had authored 2 landmark papers on pulmonary atresia with intact ventricular septum,^{1,2} a lesion which would form the basis of a lifelong study, culminating in the appearance of a monograph that remains today the authoritative work on the subject.³

An idea of his impact on the unit during his time in Toronto can be gauged by the memories of George Trusler, his long-standing colleague in paediatric cardiac surgery. George writes:

"Bob Freedom was a tremendous asset to both cardiology and cardiac surgery. He was a combination of many talents: a brilliant clinician and morphologist, an accomplished



Figure 1.
Robert Freedom photographed at the time of his retirement from the Hospital for Sick Children.

speaker, a prolific writer and a prodigious worker. He was in the forefront in studies of cardiac morphology and his wise counsel and superb teaching were of immense value to us as surgeons. He had a remarkable memory and his knowledge of the literature, both medical and surgical, was truly encyclopaedic. Frequently, to contribute to a discussion, he would refer to a paper, often in the surgical literature, and a quote the authors, the journal and even the date of publication as well as the salient content.

Like his predecessors, John Keith and Dick Rowe, Bob was surgically oriented, encouraging advances in surgical methods

Correspondence to: Lee N. Benson MD, Division of Cardiology, The Hospital for Sick Children, The University of Toronto School of Medicine, Toronto, Ontario, Canada, M5G 1X8. Tel: +1 416 813 6141; Fax: +1 416 813 7547; E-mail: lee.benson@sickkids.ca

Accepted for publication 8 November 2004



1997 at Dr. Rowe Award Ceremony



At his retirement in 2000



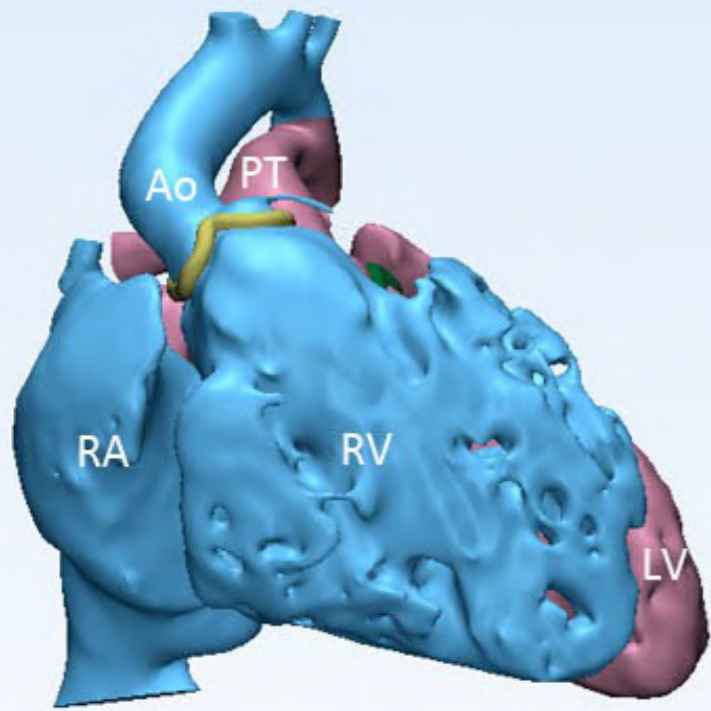
2004 in his home in Nova Scotia

- Criss-cross or twisted heart
- Superoinferior ventricles
- Topsy-turvy heart

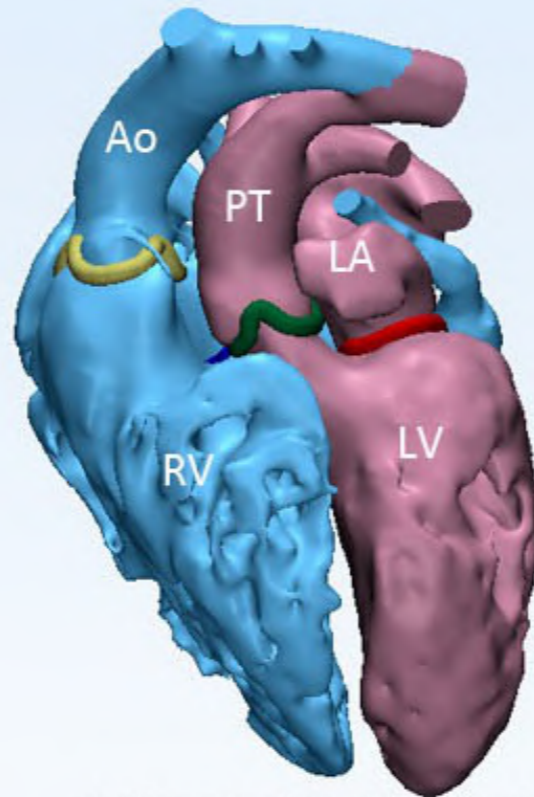
QUESTION:

Are 'criss-cross heart' and 'superoinferior ventricles' different entities?

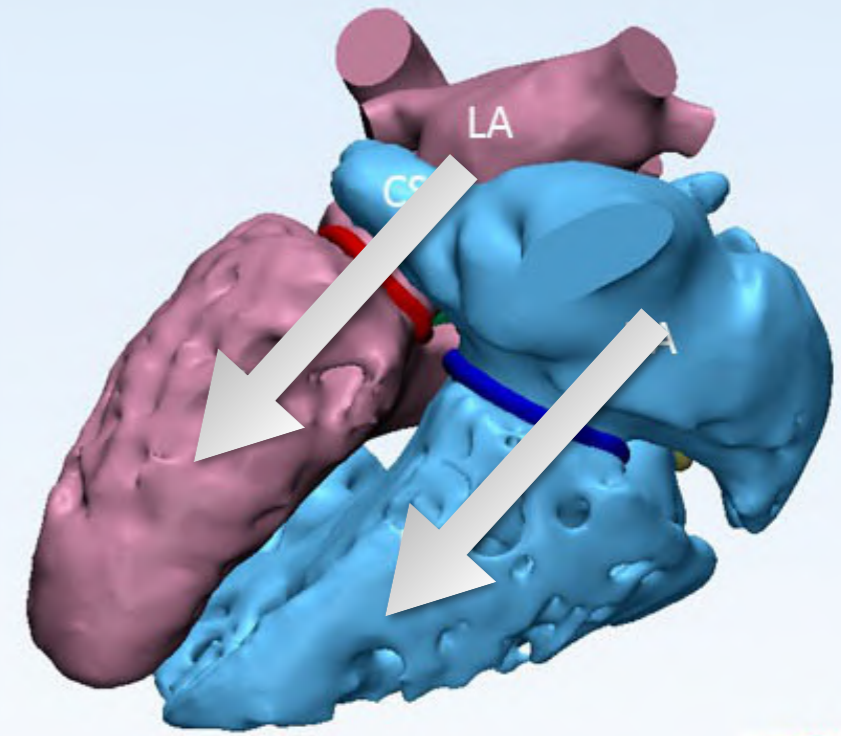
1. Yes
2. No
3. Yes and no



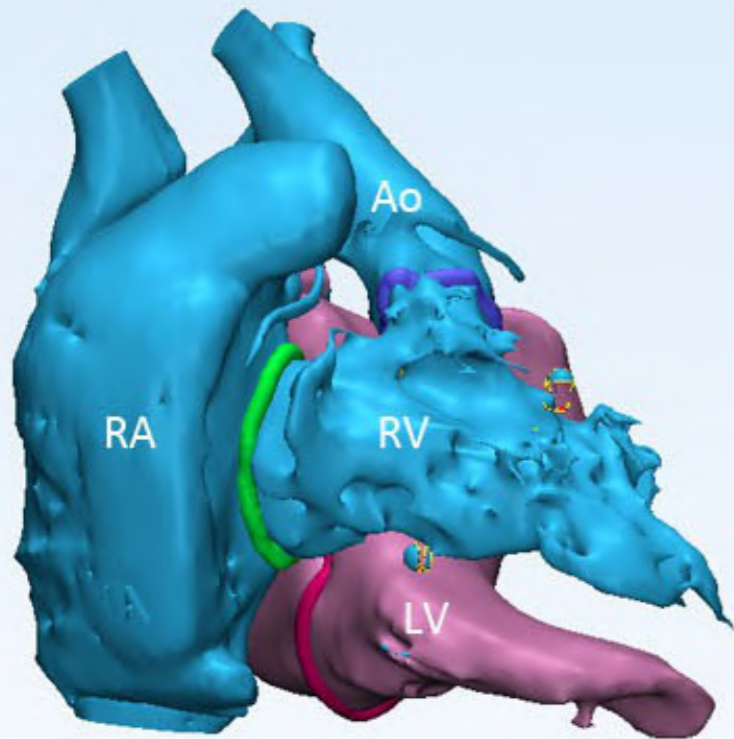
Frontal view



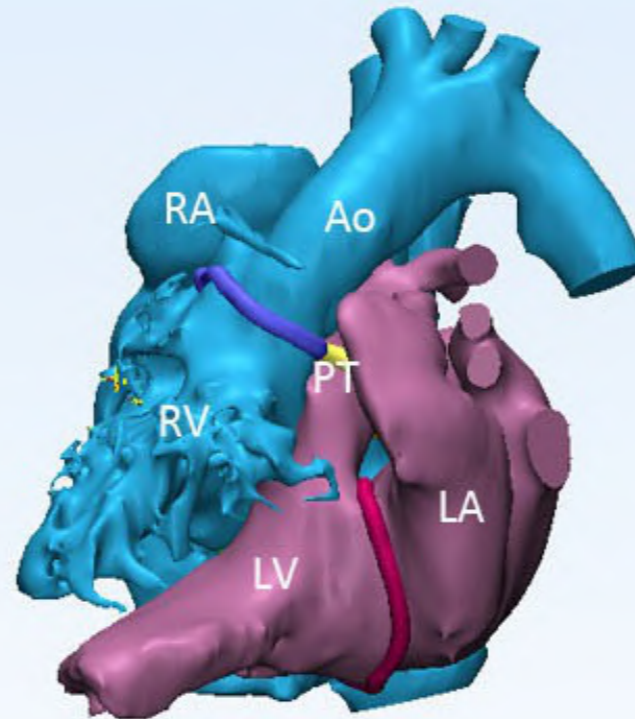
Left anterior oblique view



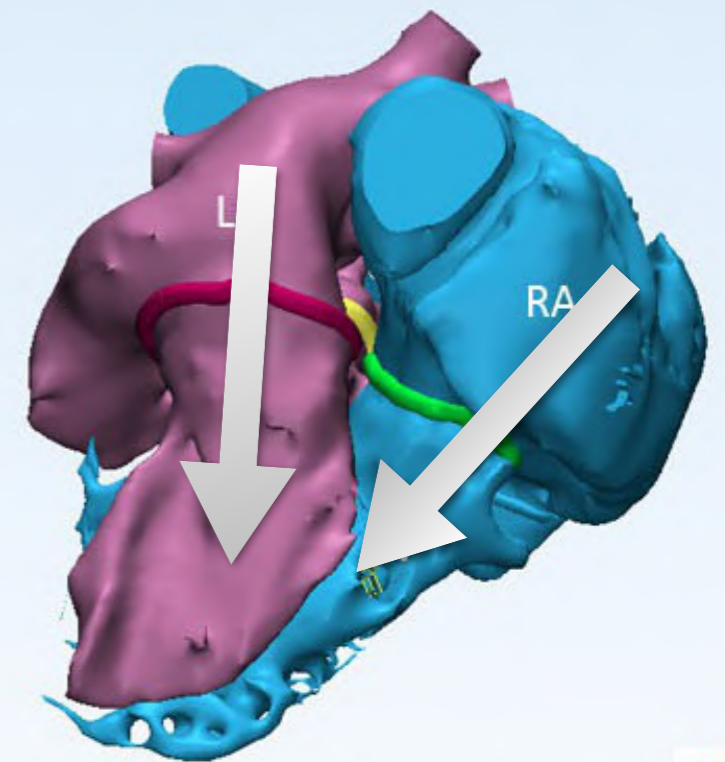
Posterior view



Frontal view



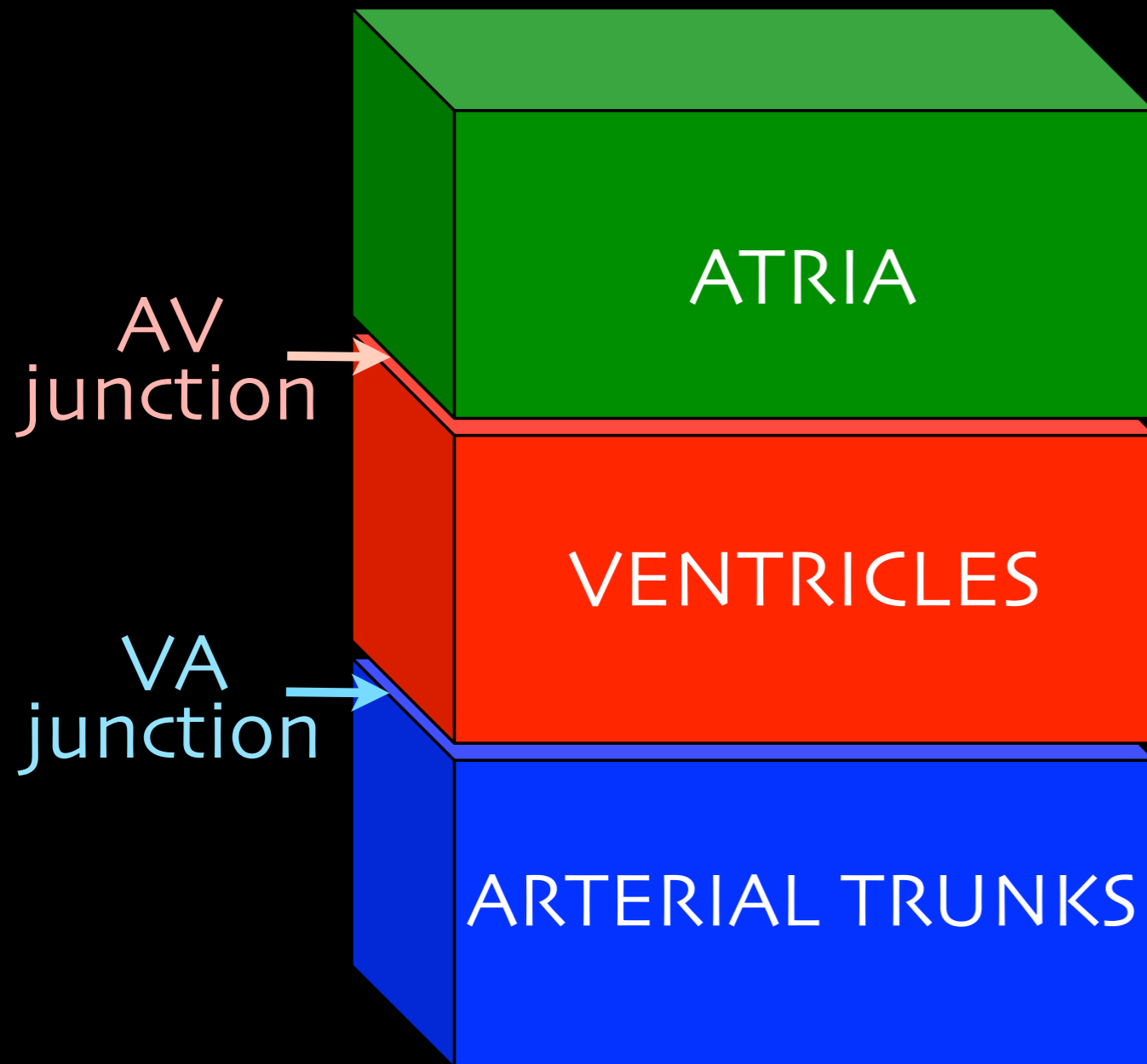
Left lateral view



Posteroinferior view



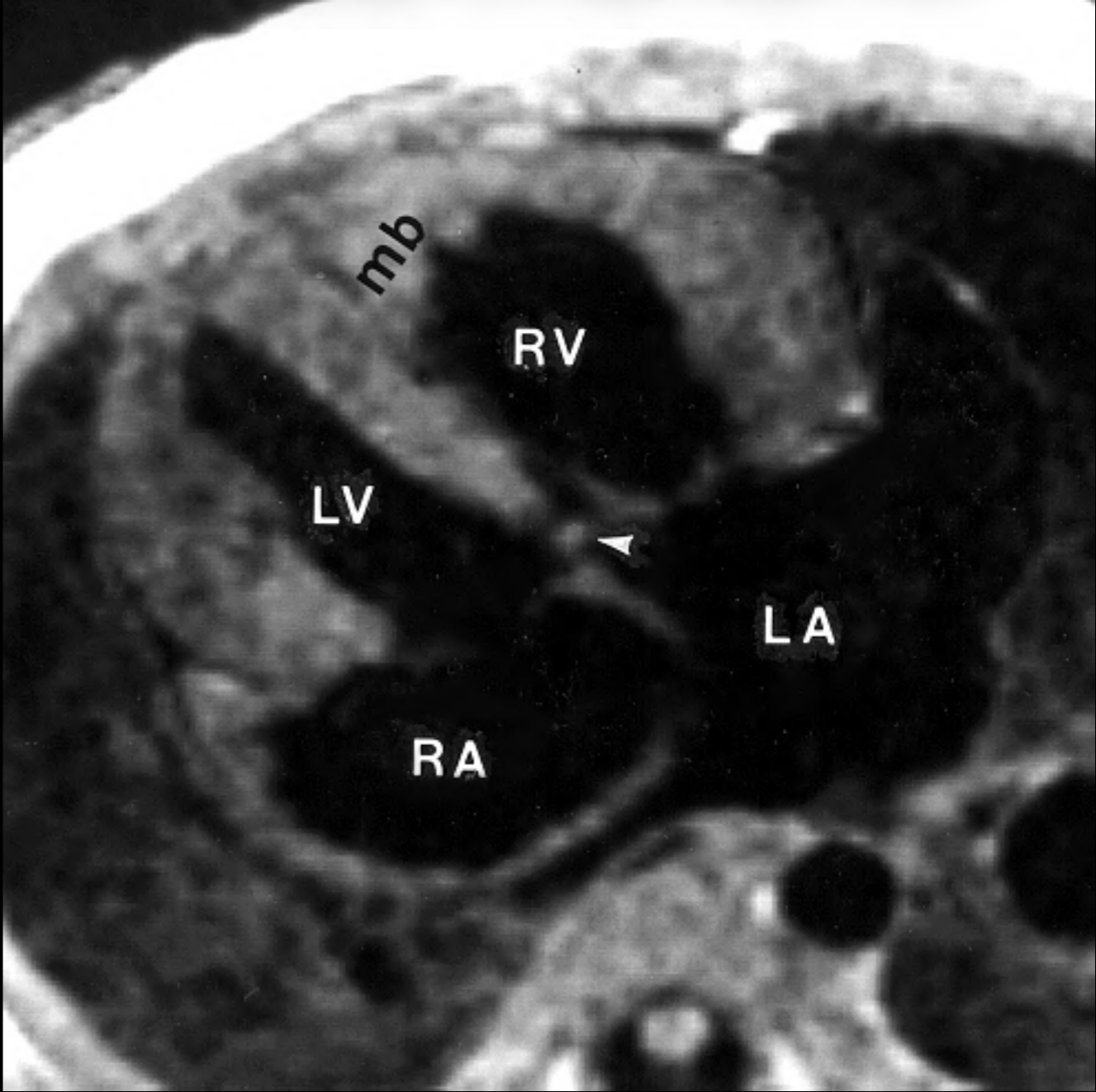
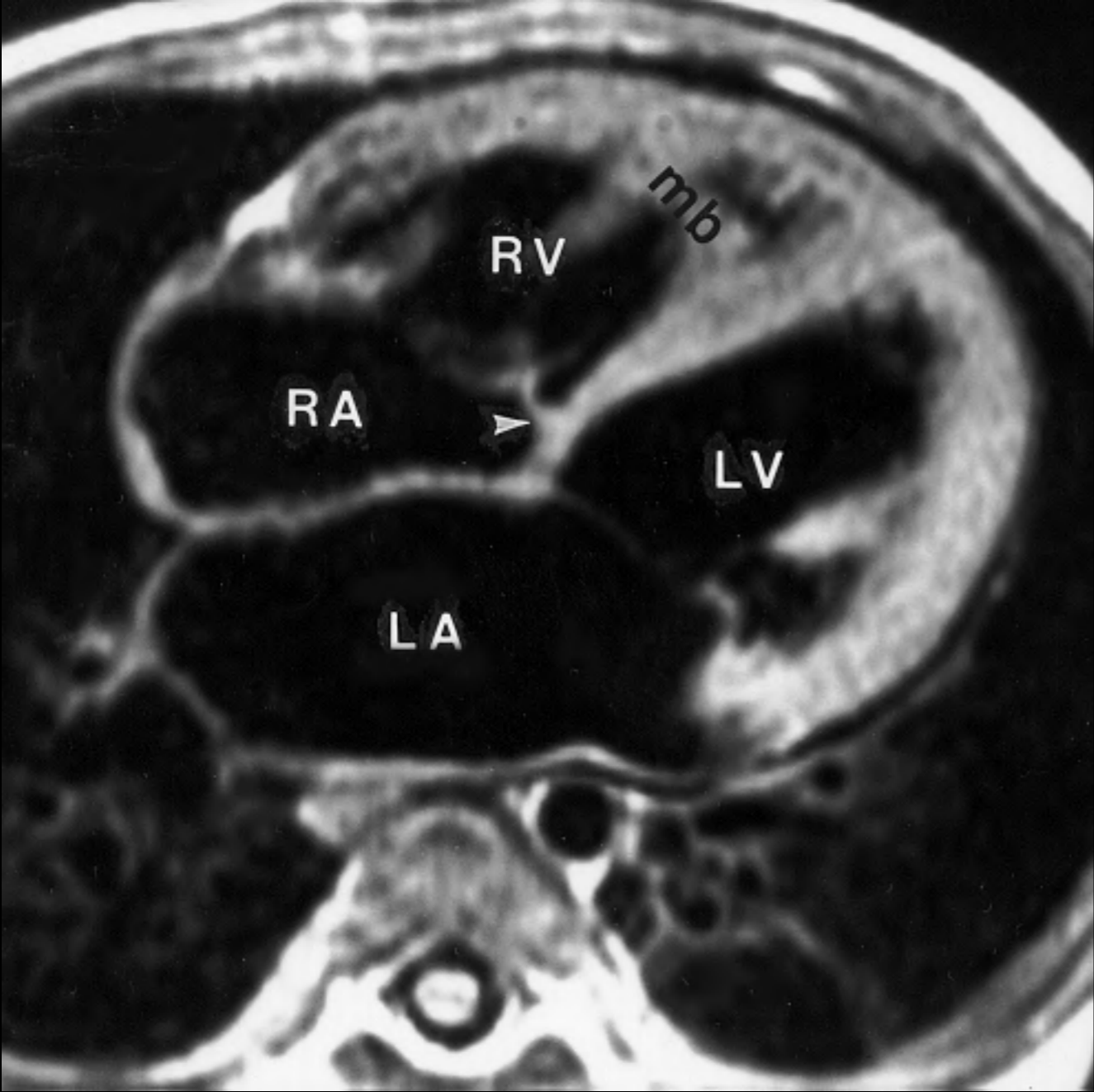
3 Segments and 2 Junctions



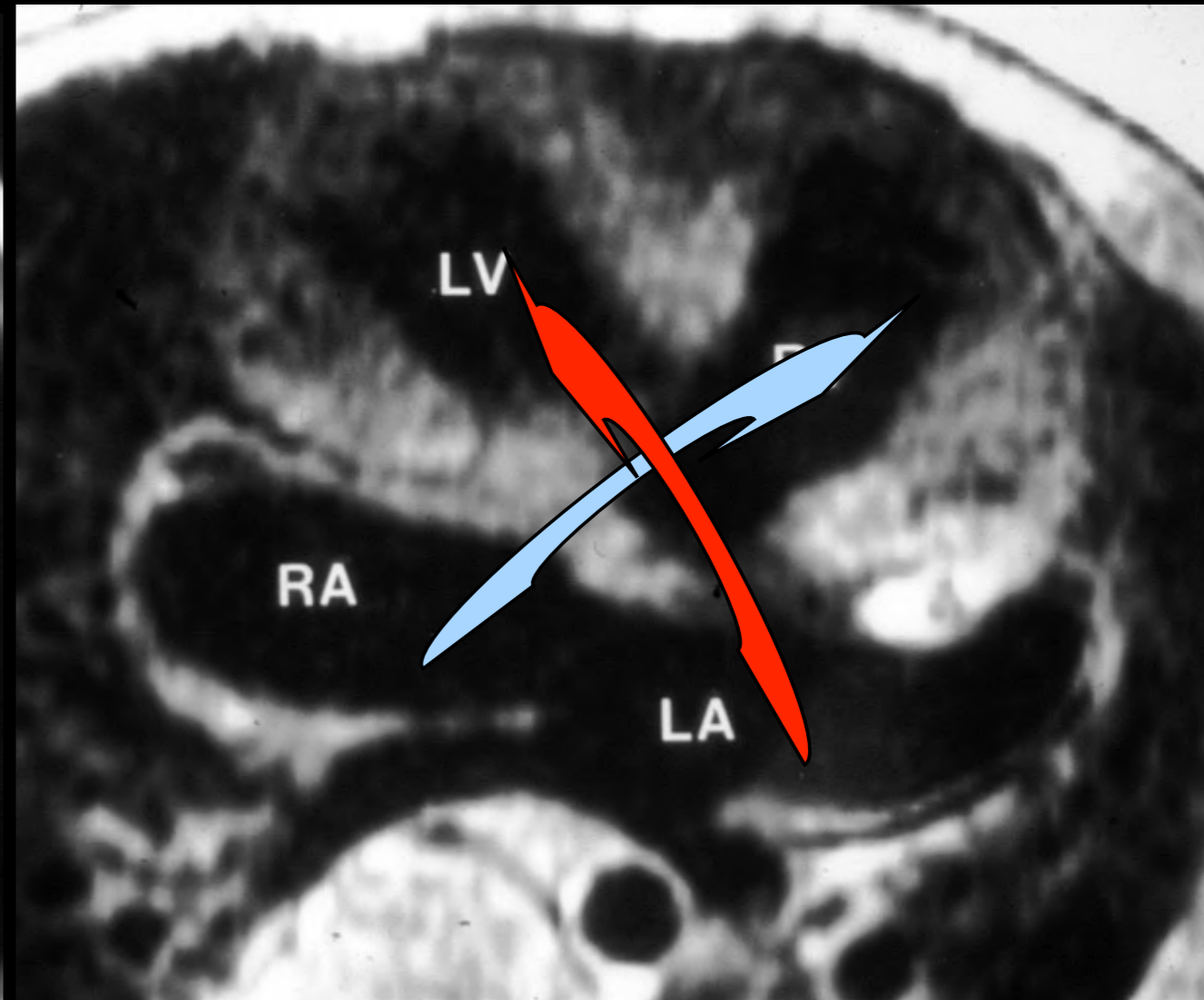
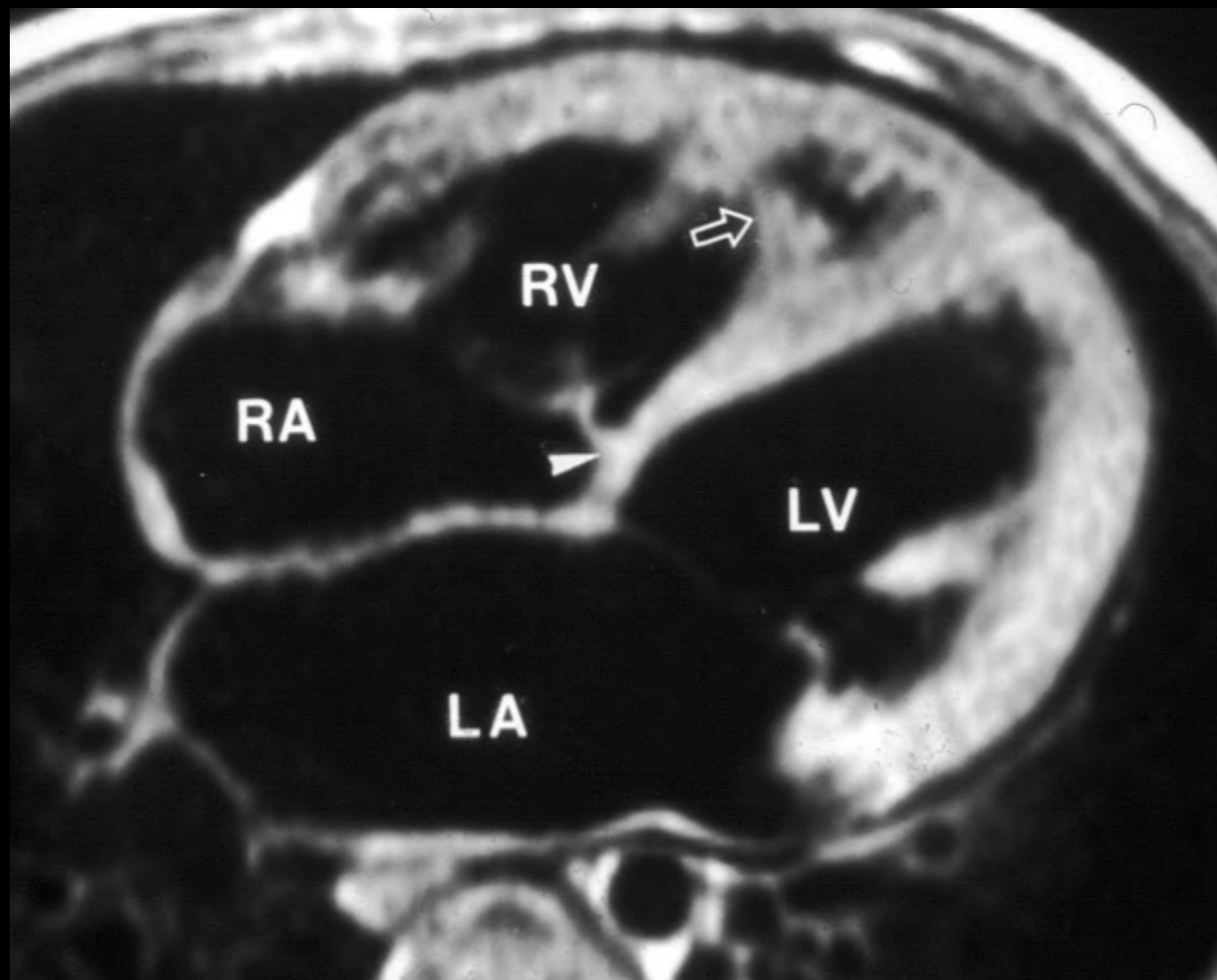
3 Facets

- Morphology
- Relationship
- Connection

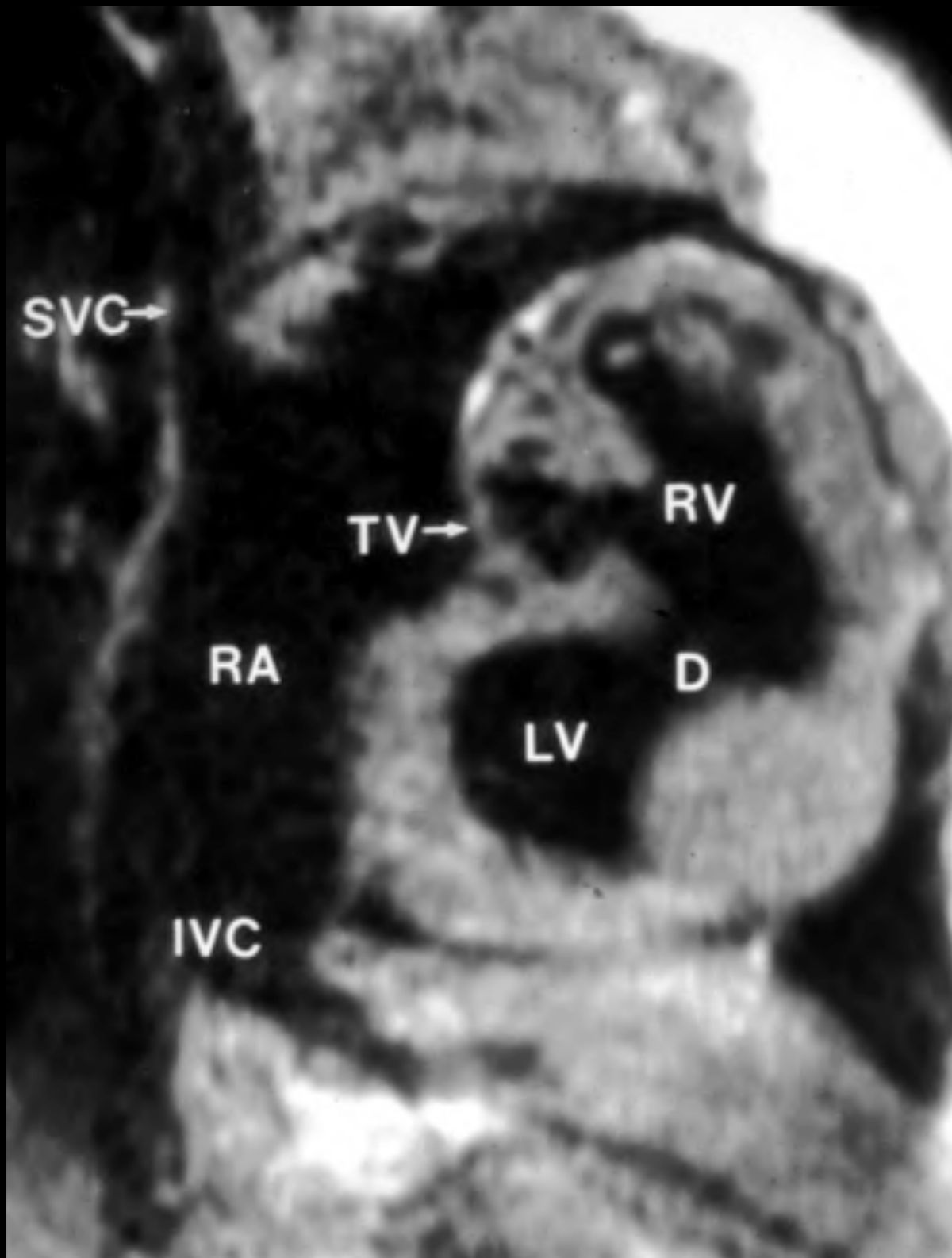
CONNECTION VERSUS RELATIONSHIP

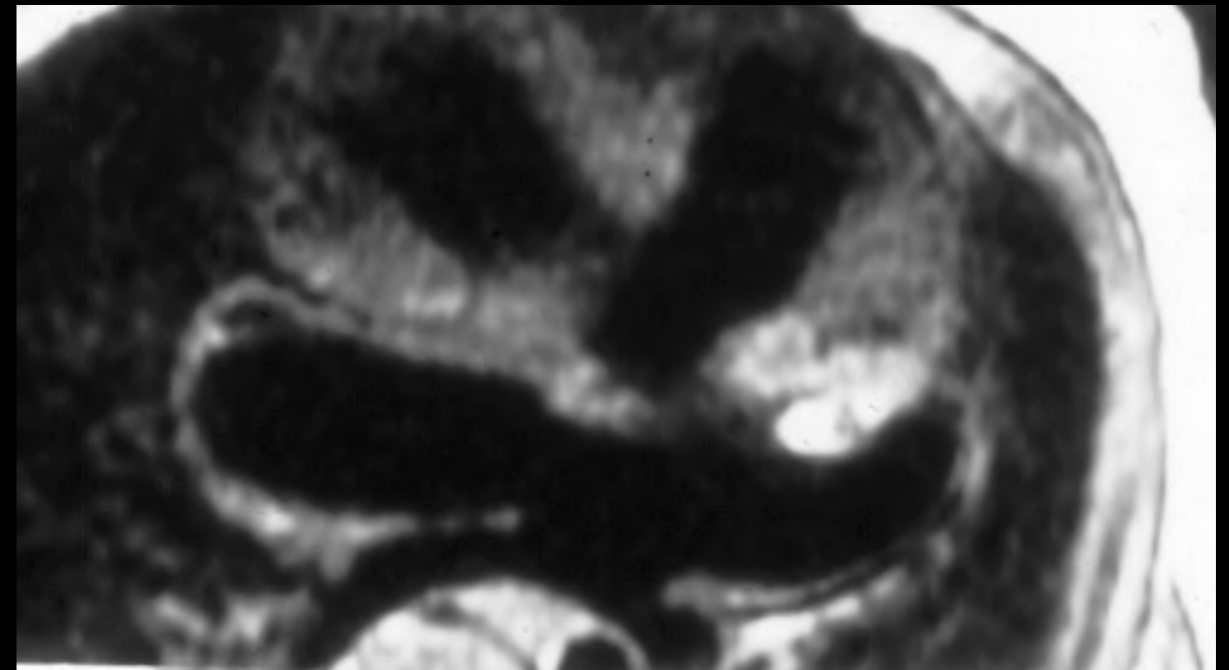
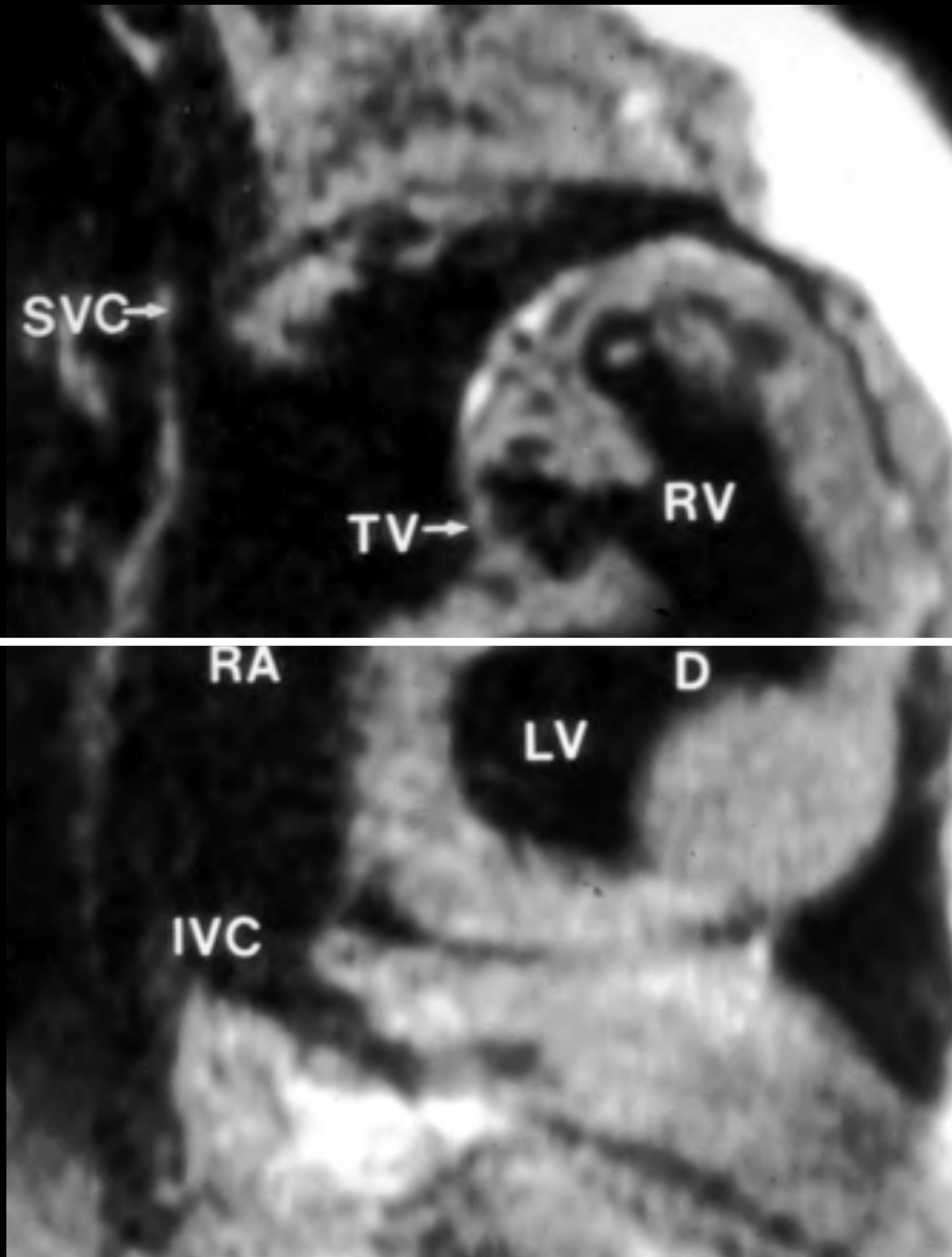


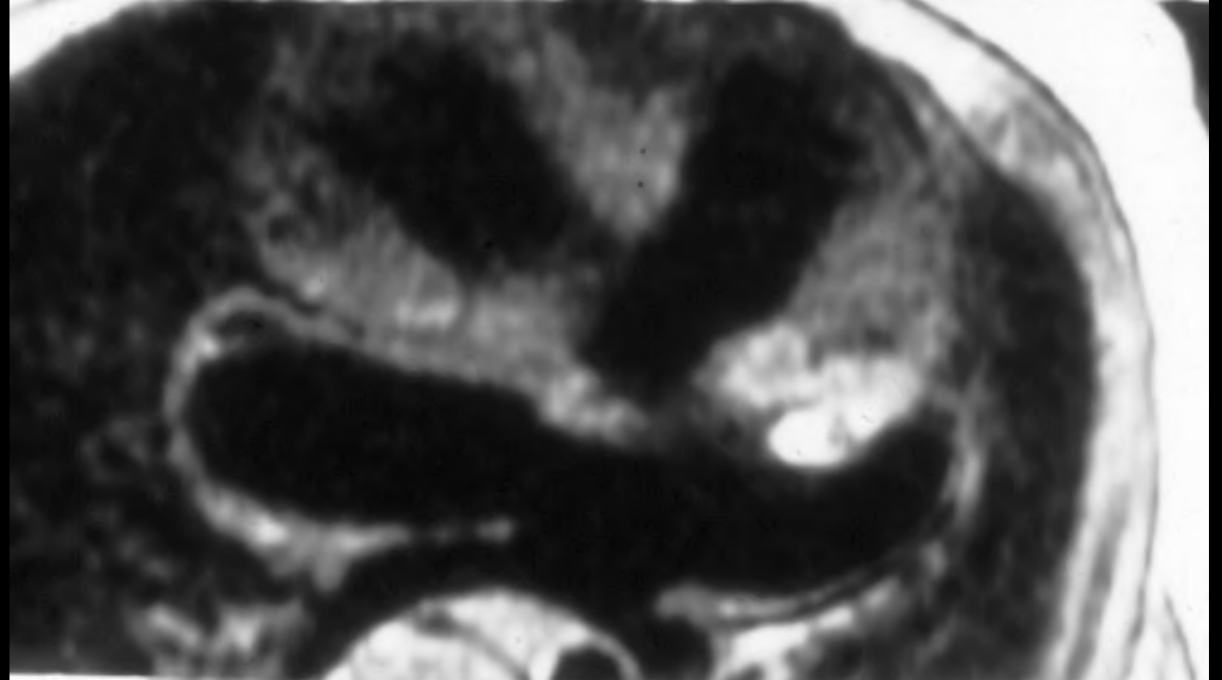
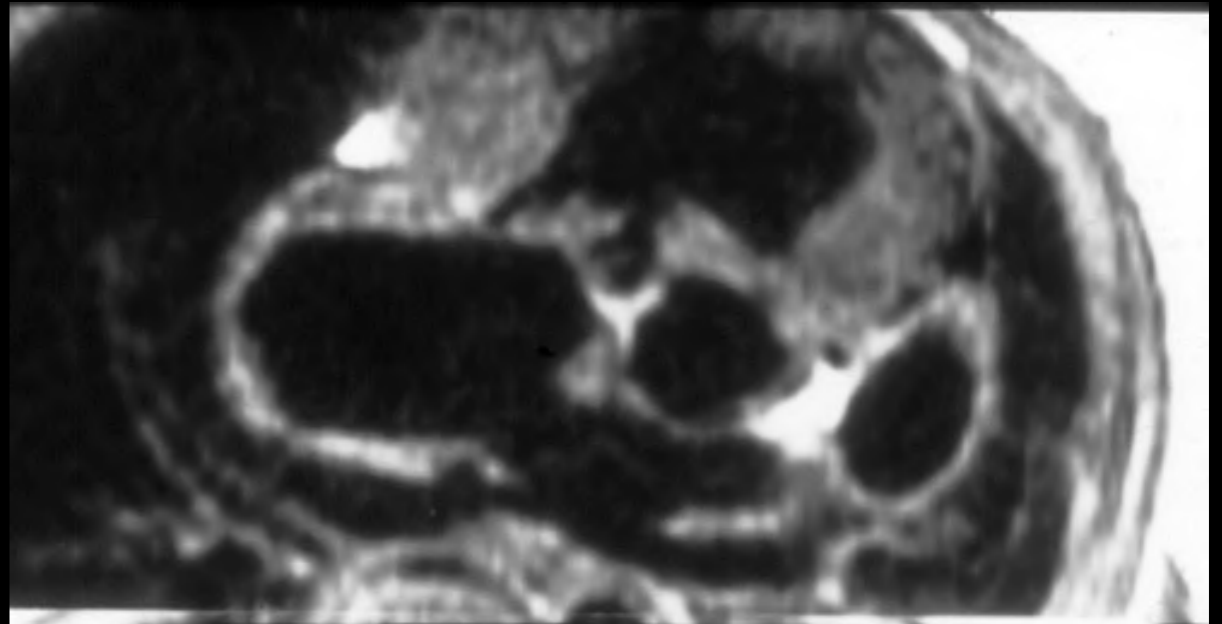
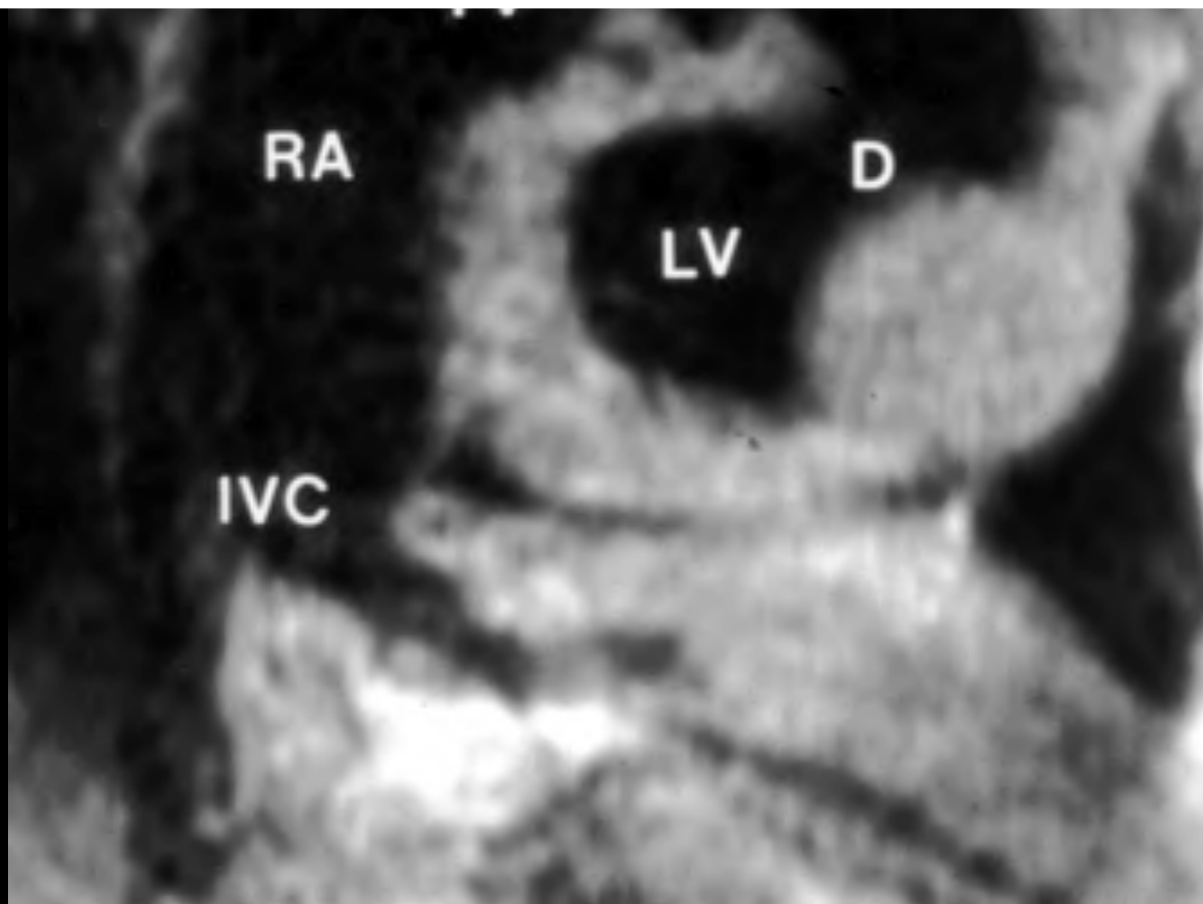
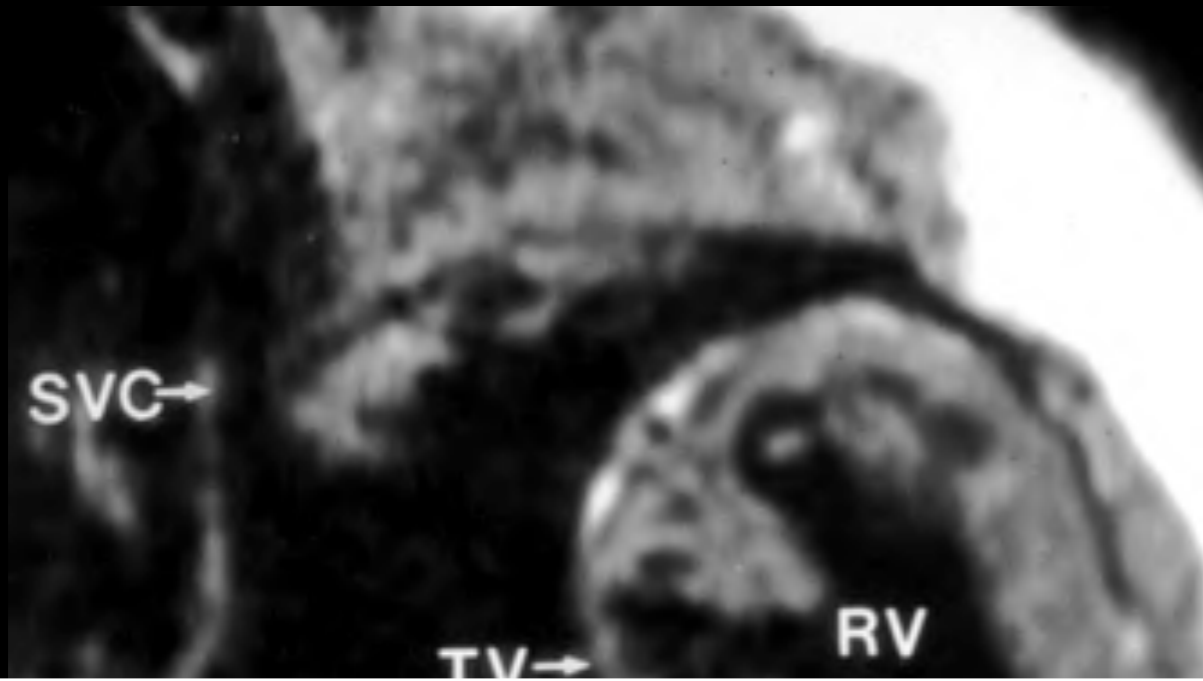
CONNECTION VERSUS RELATIONSHIP

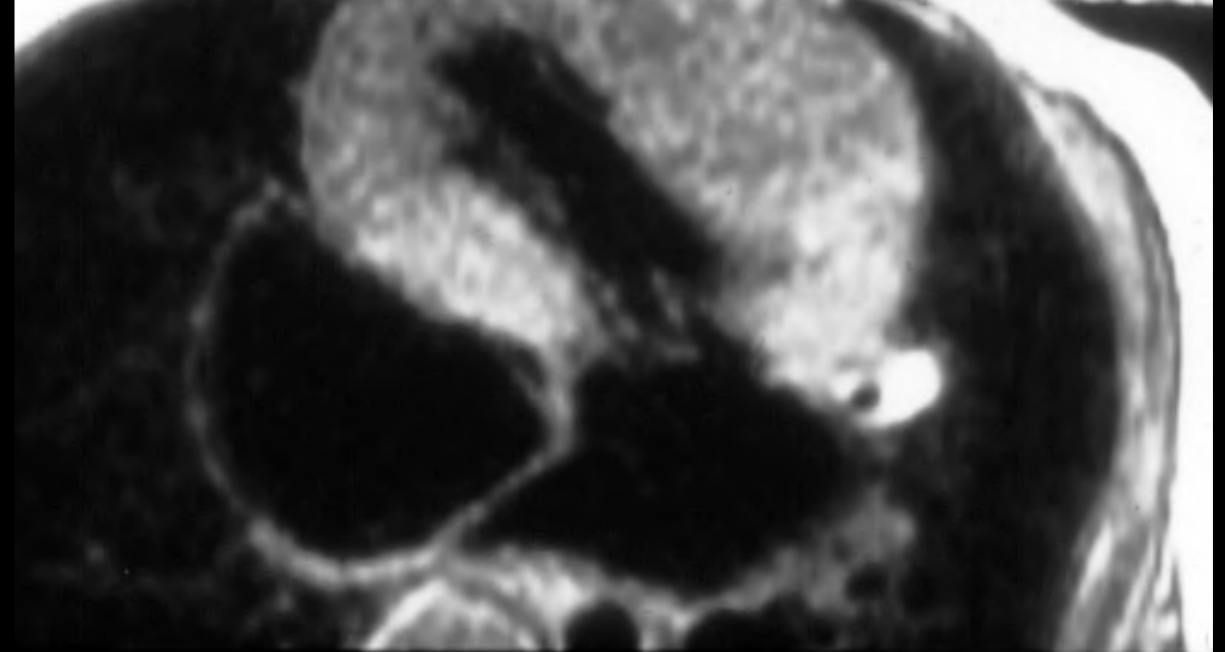
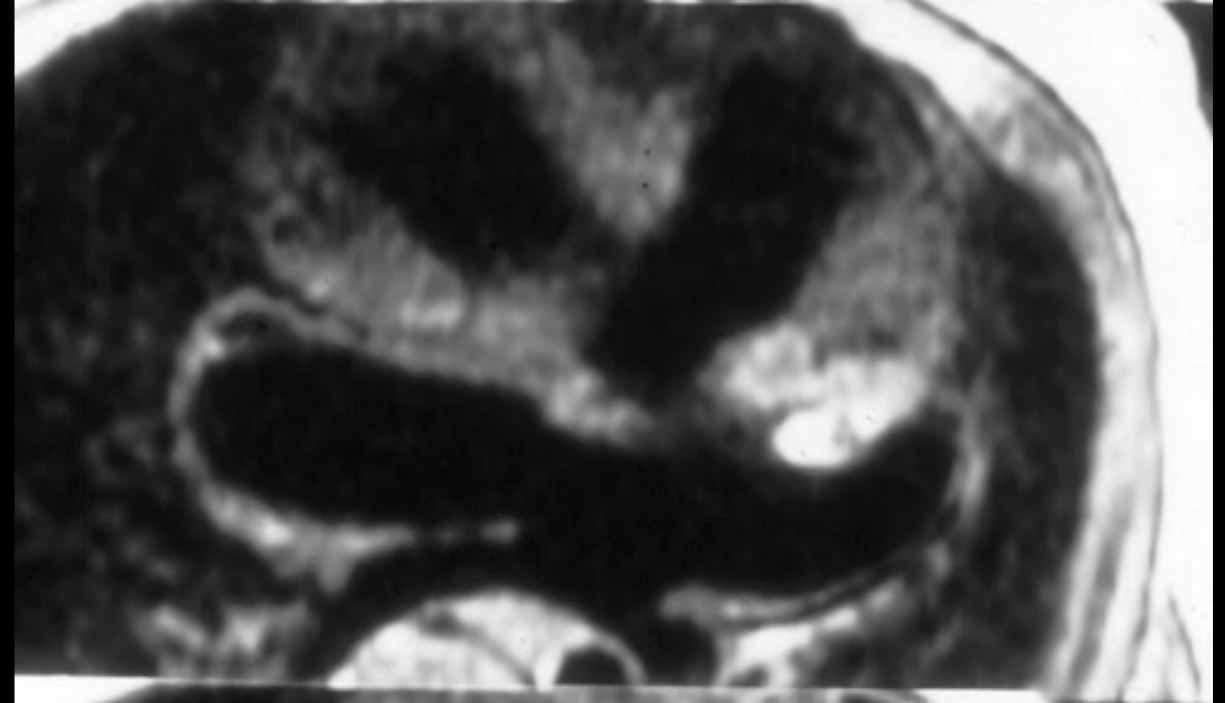
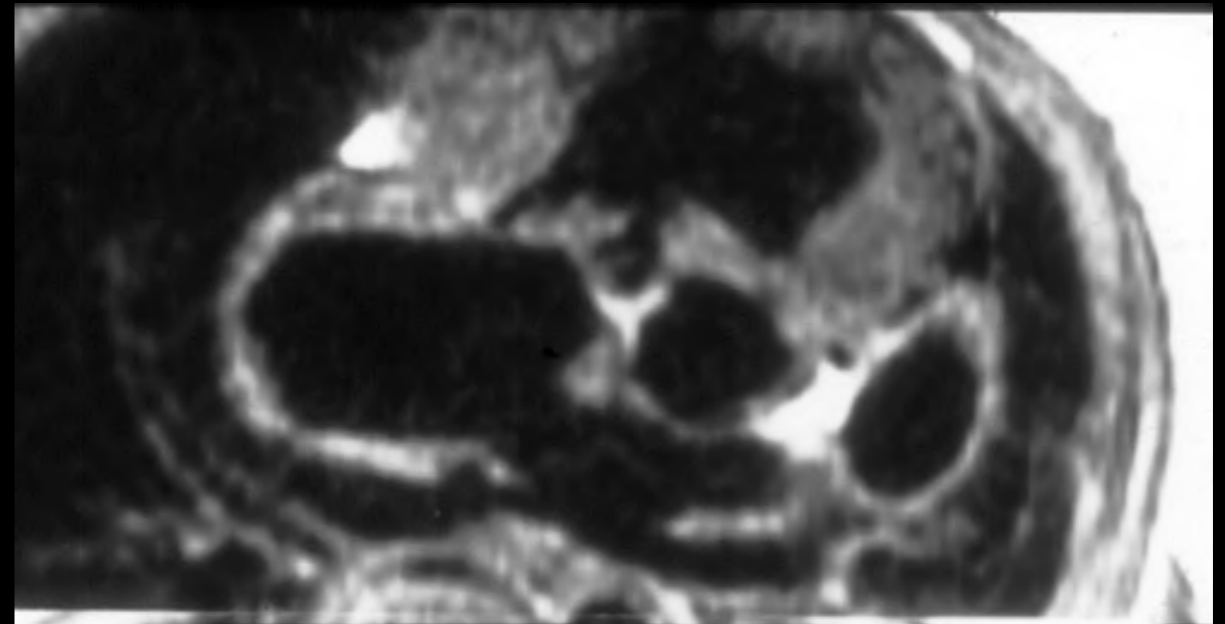
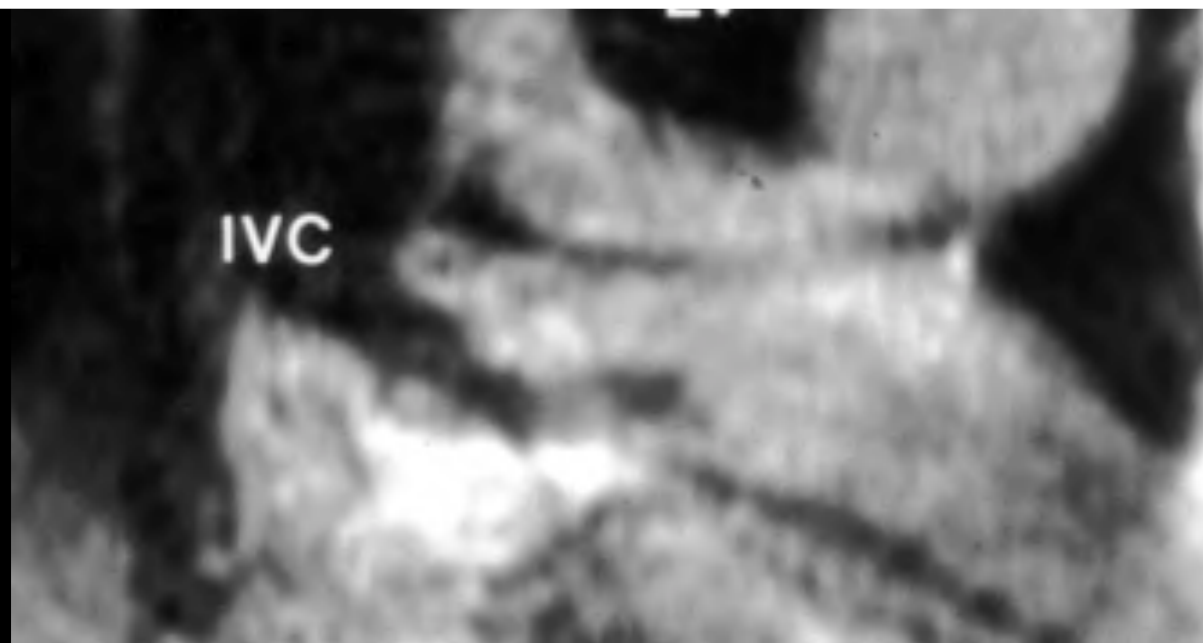
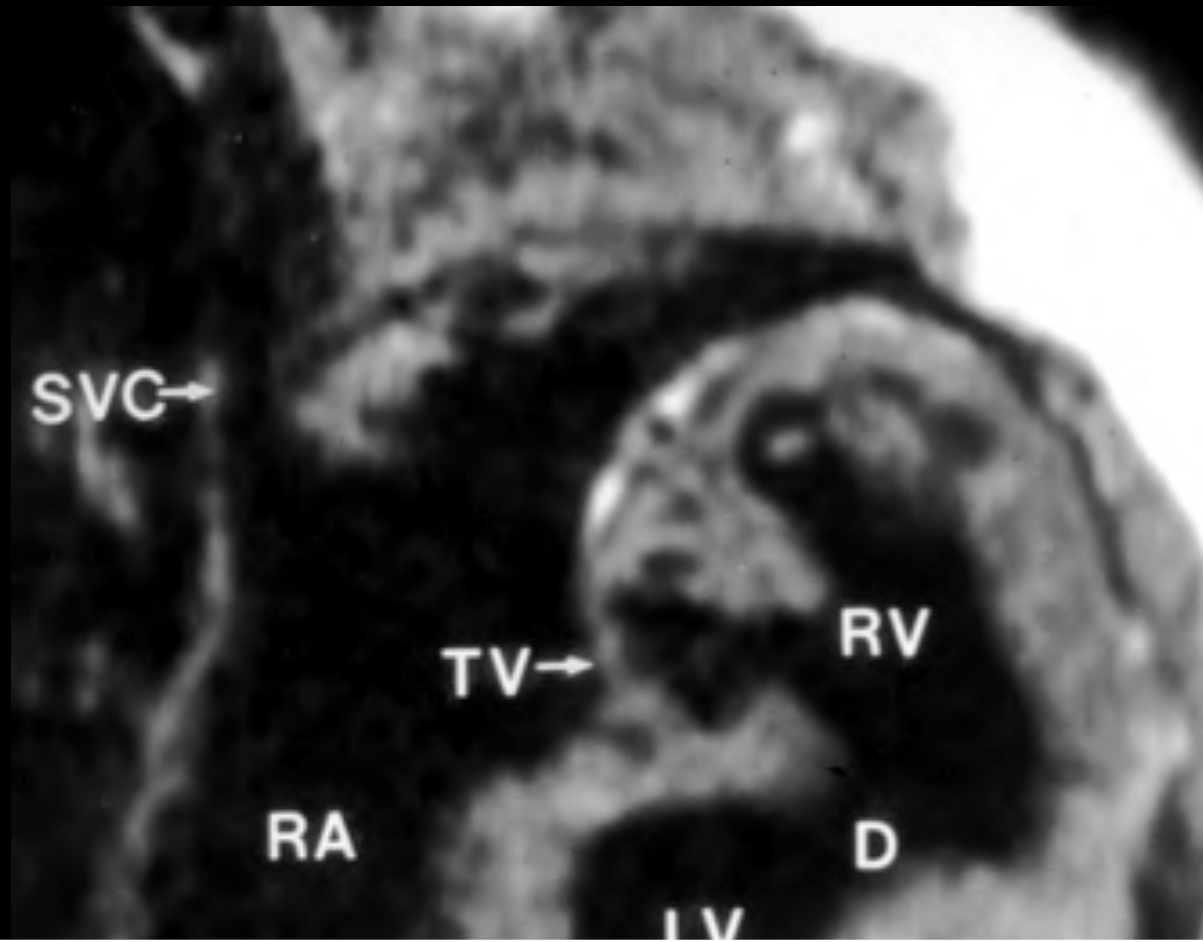


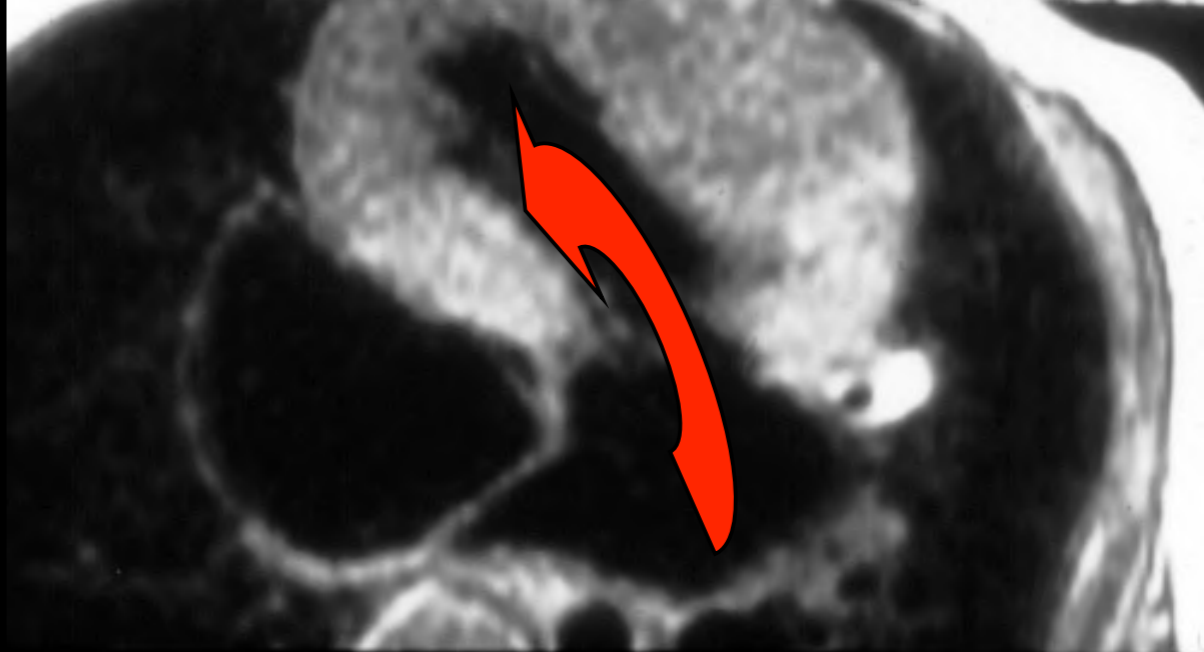
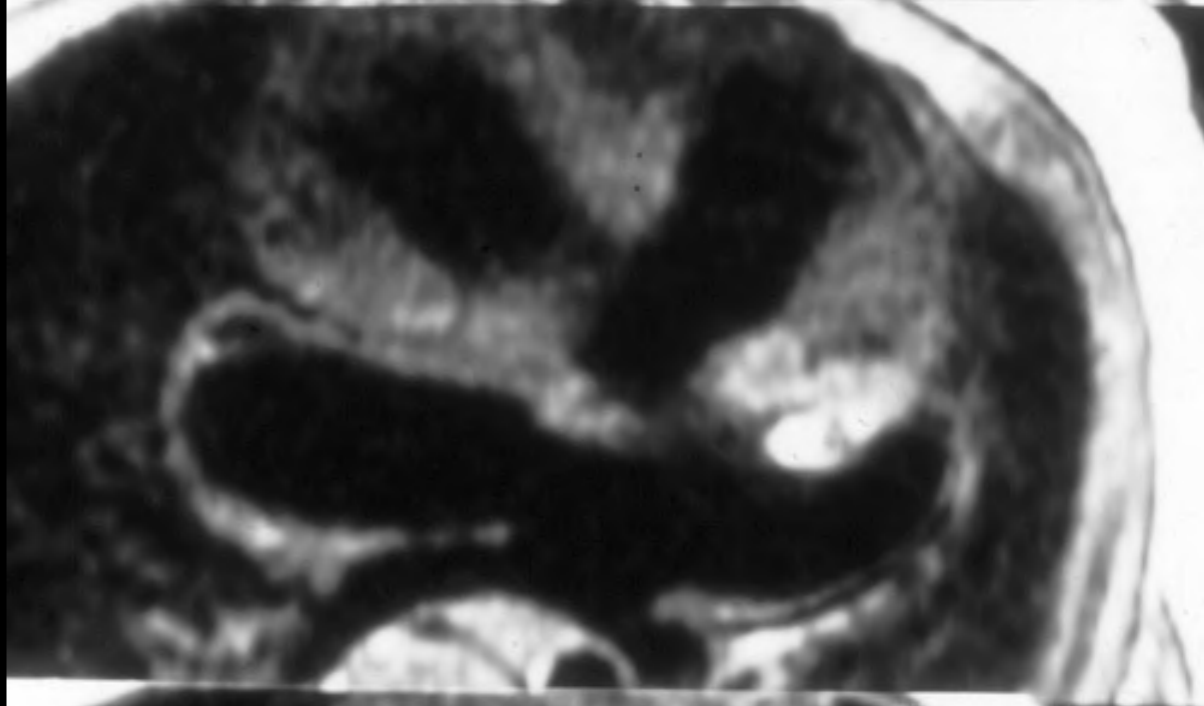
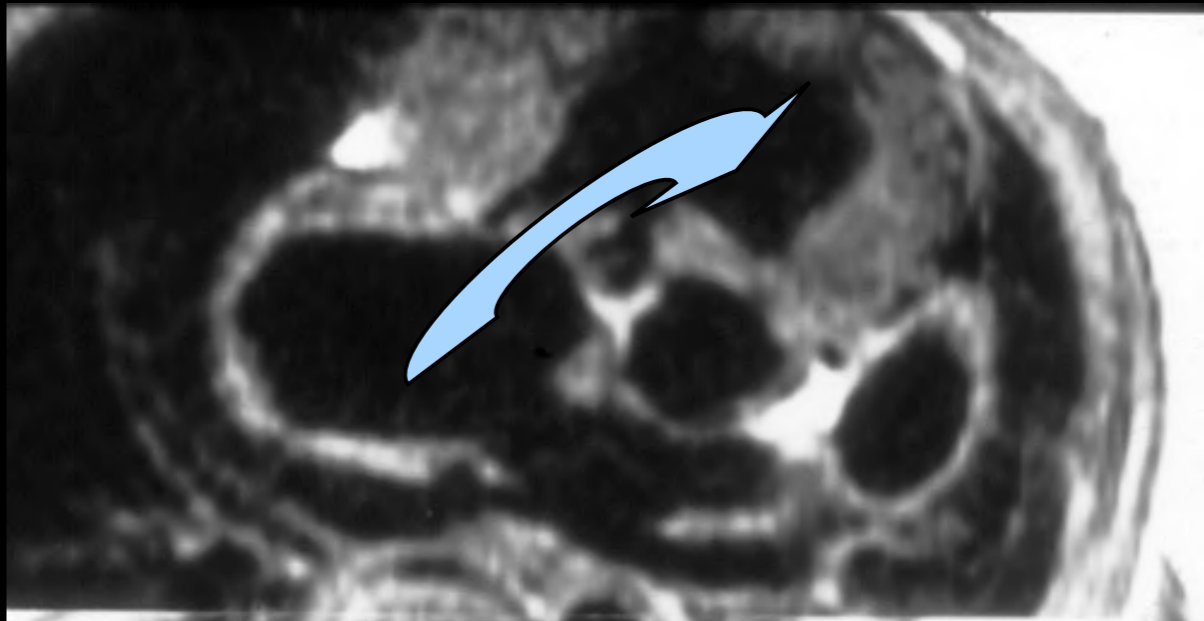
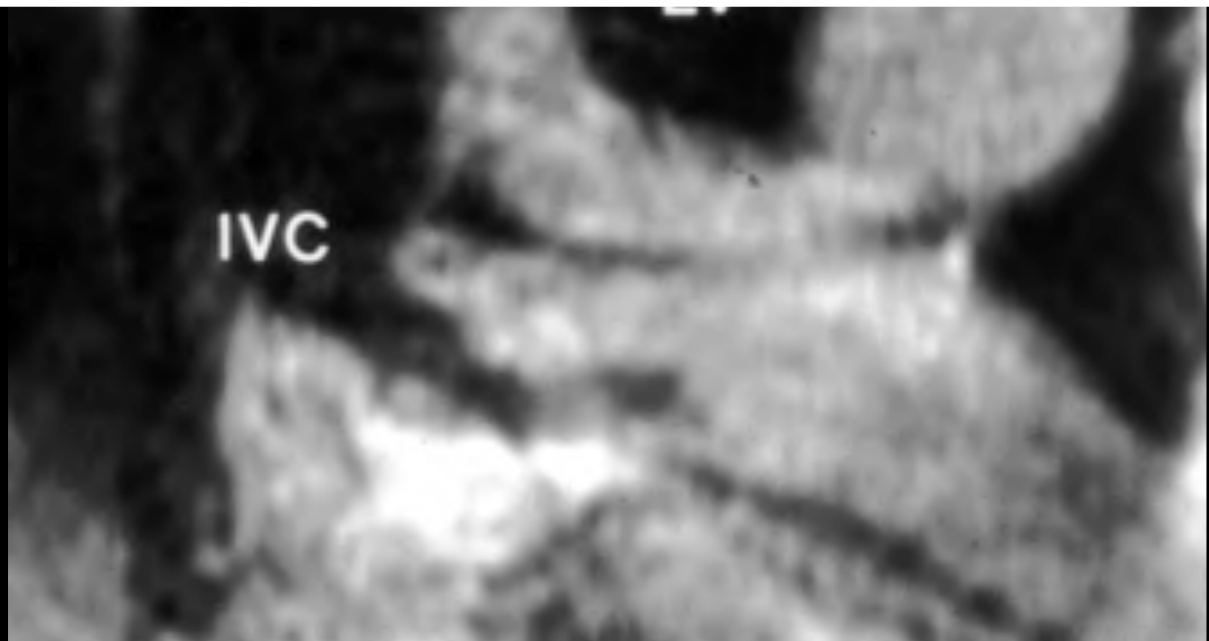
Disharmony between connection and relationship

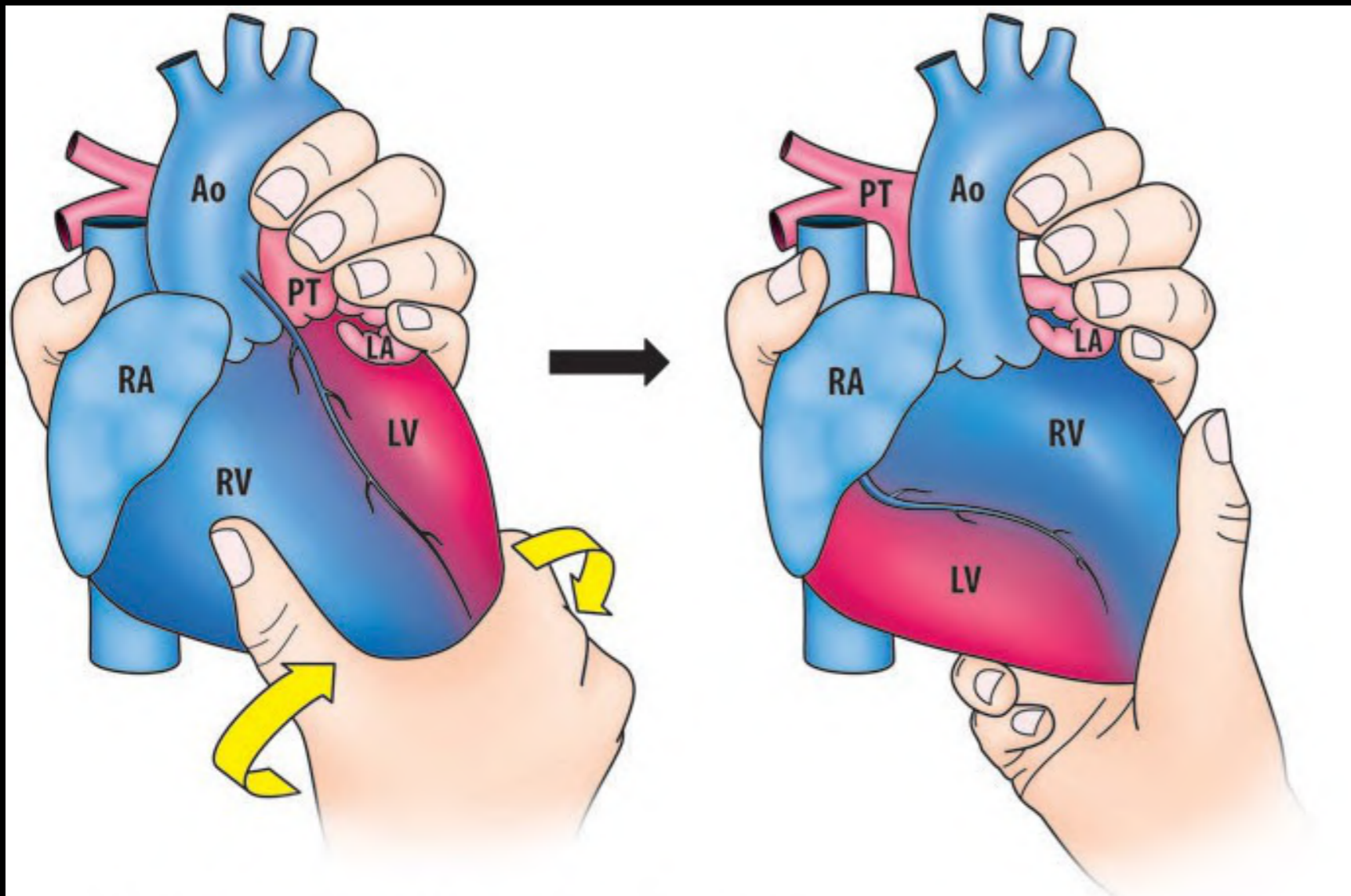


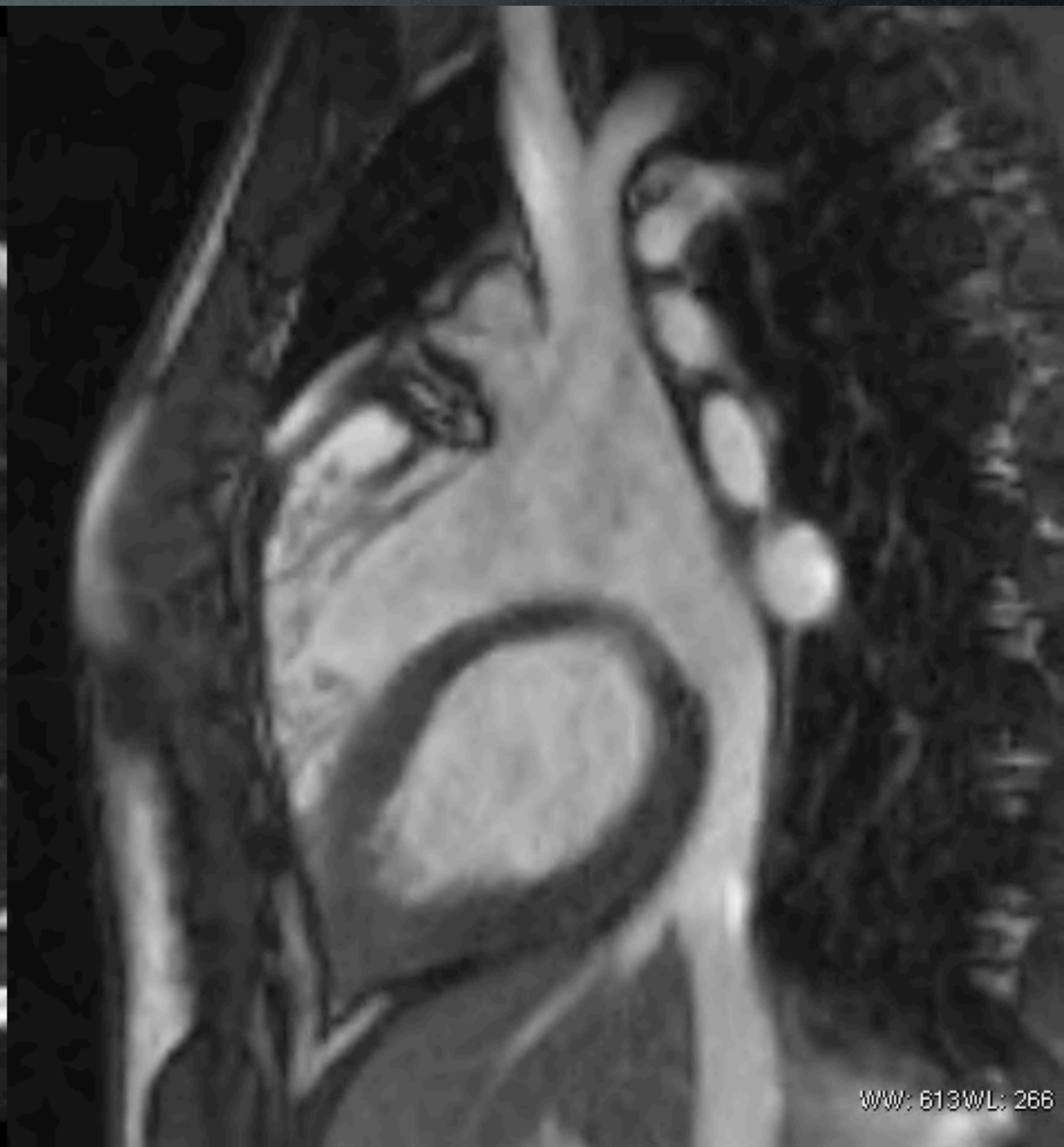
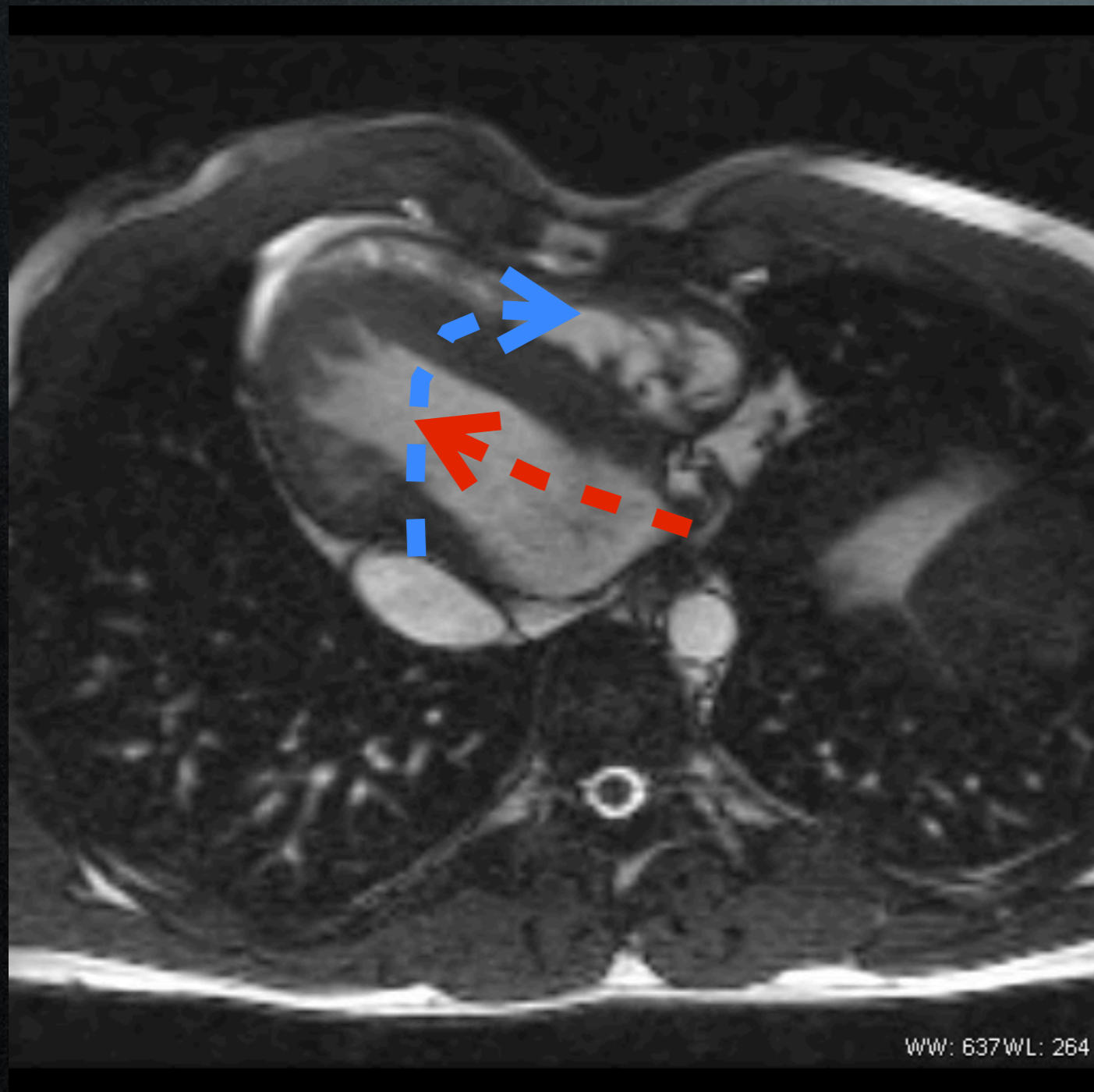






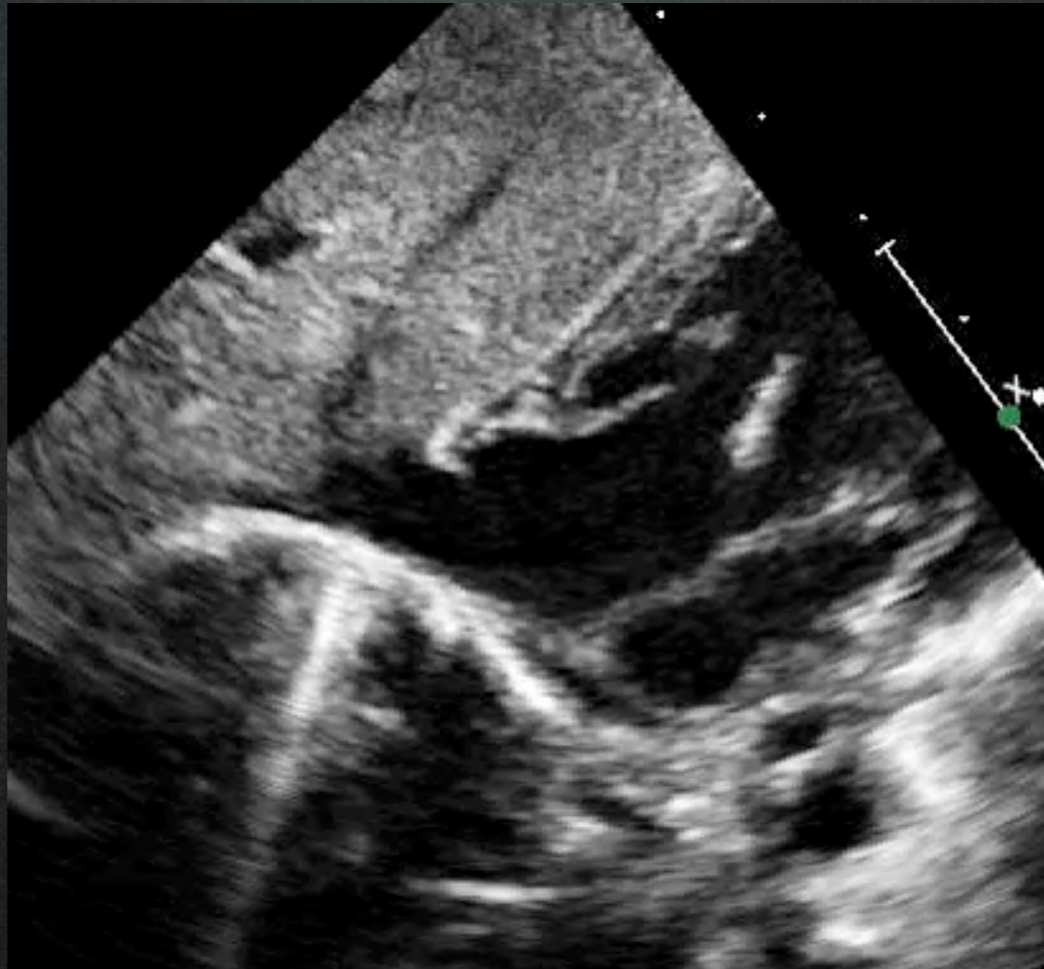
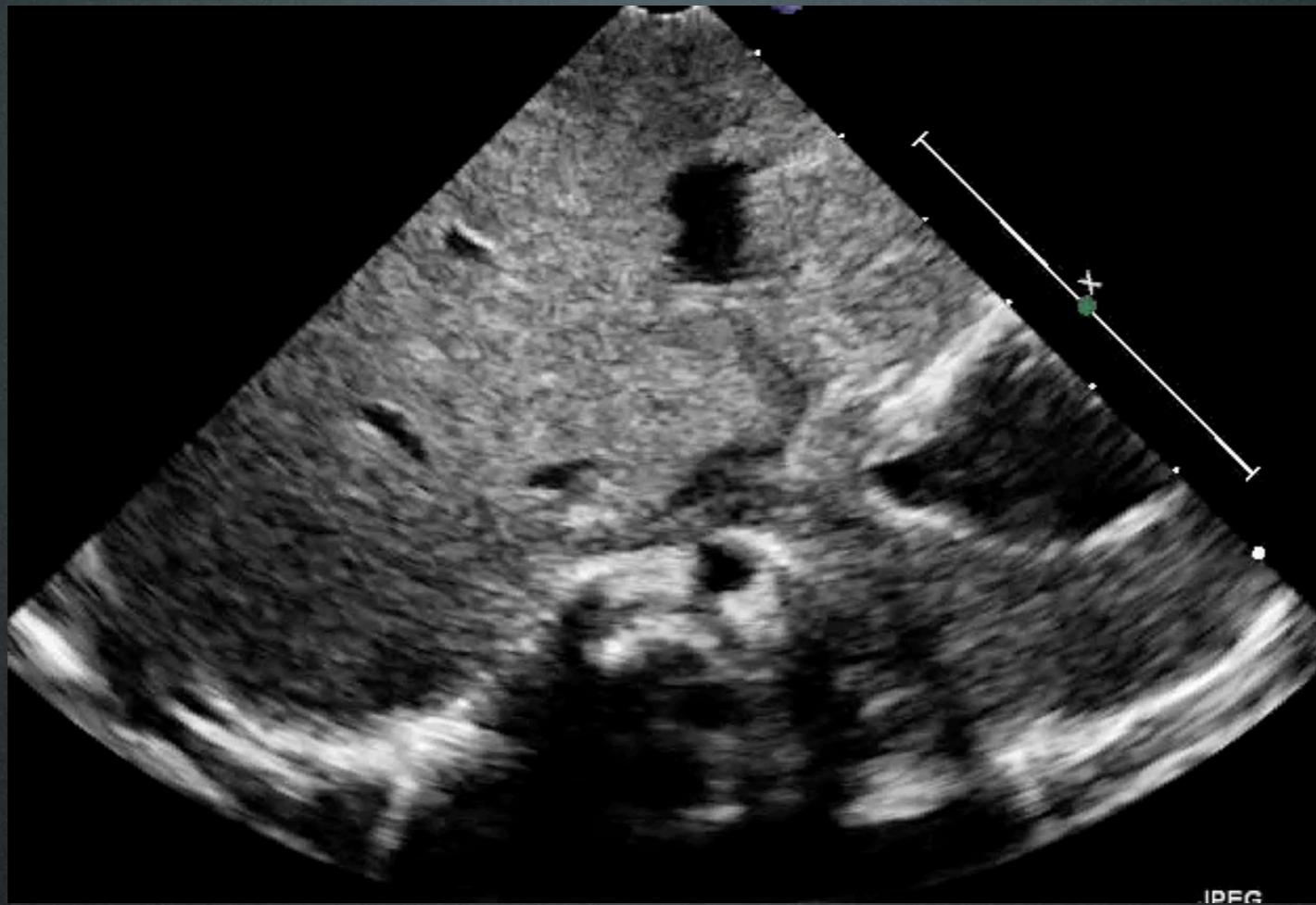


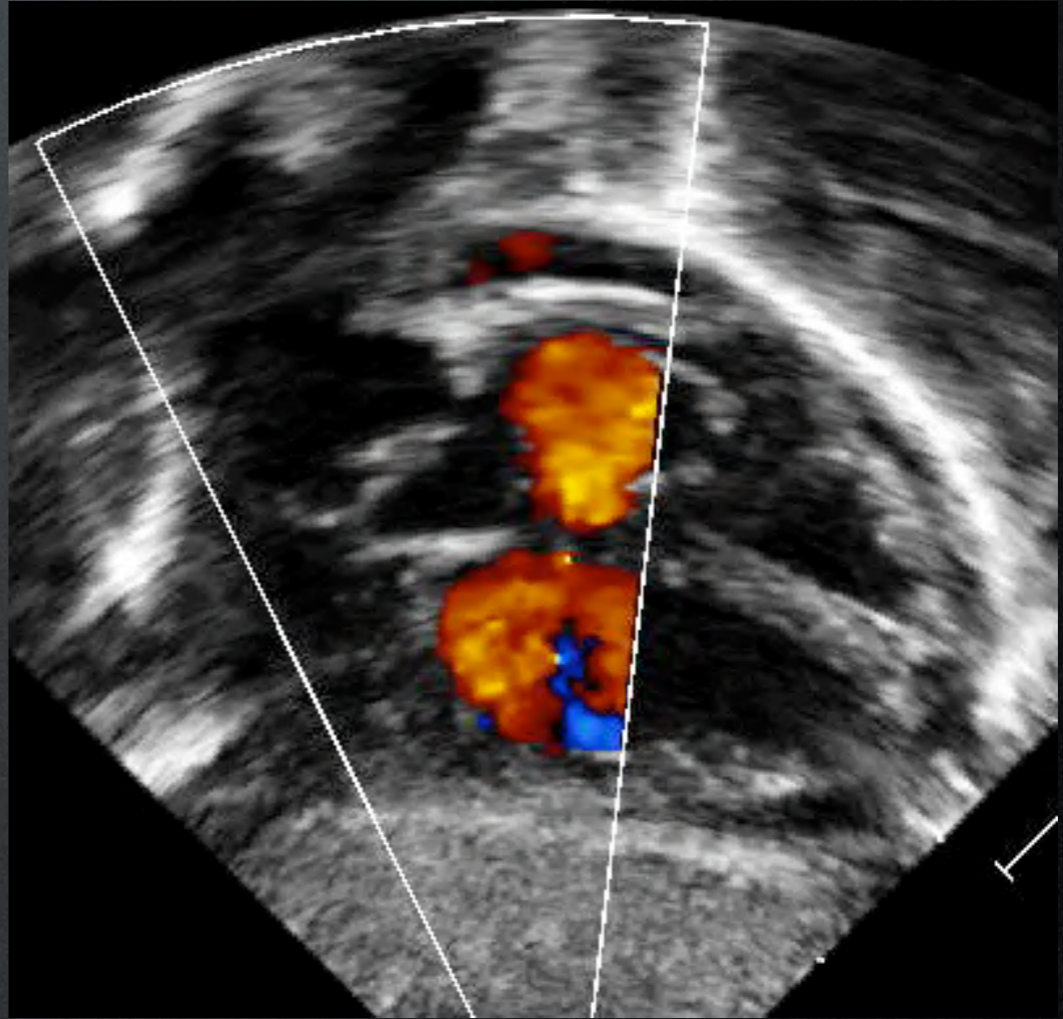
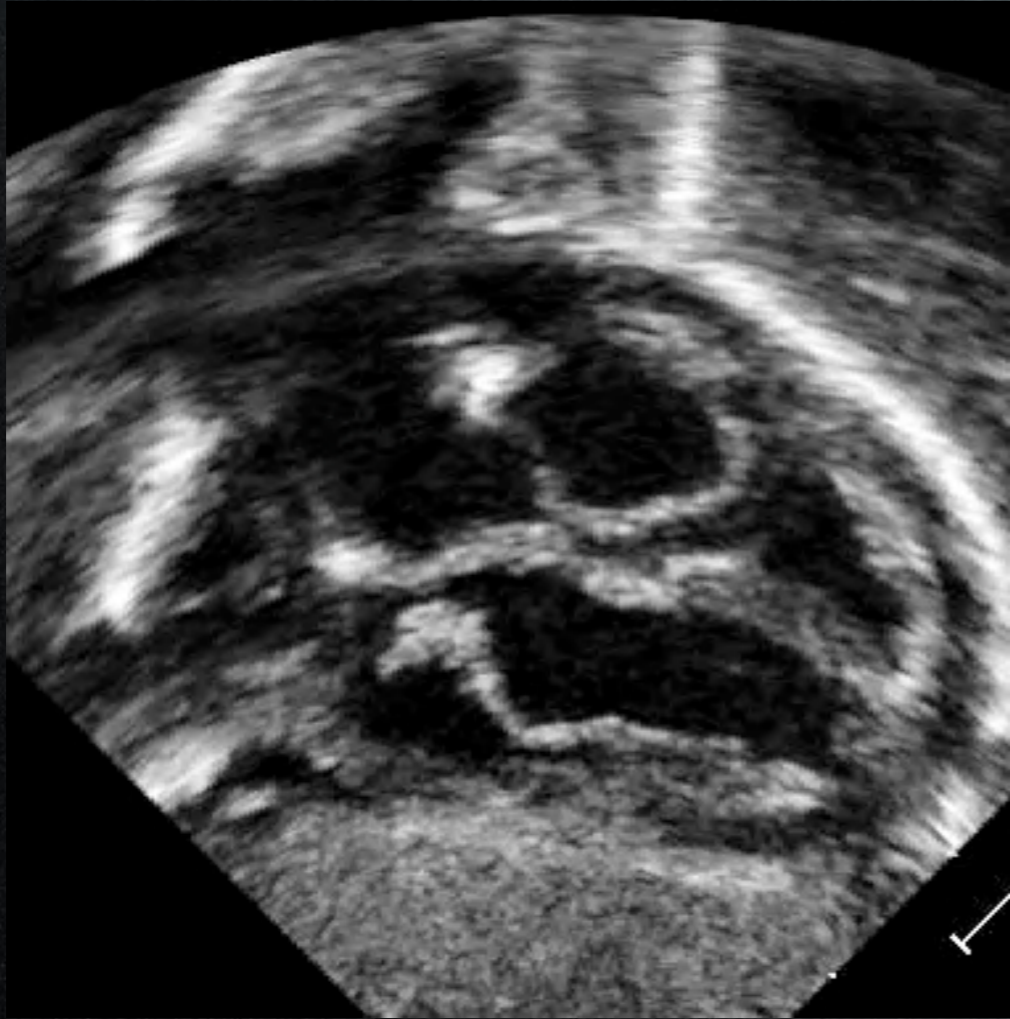
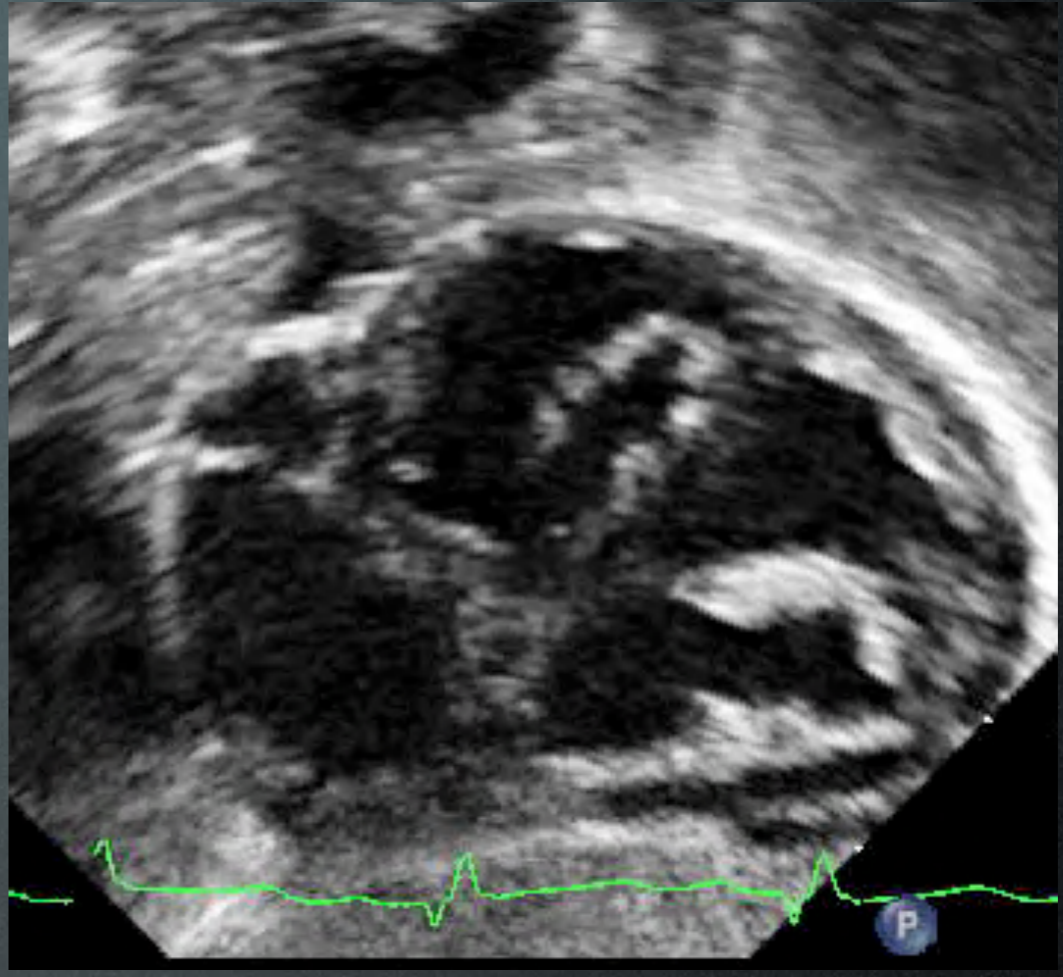




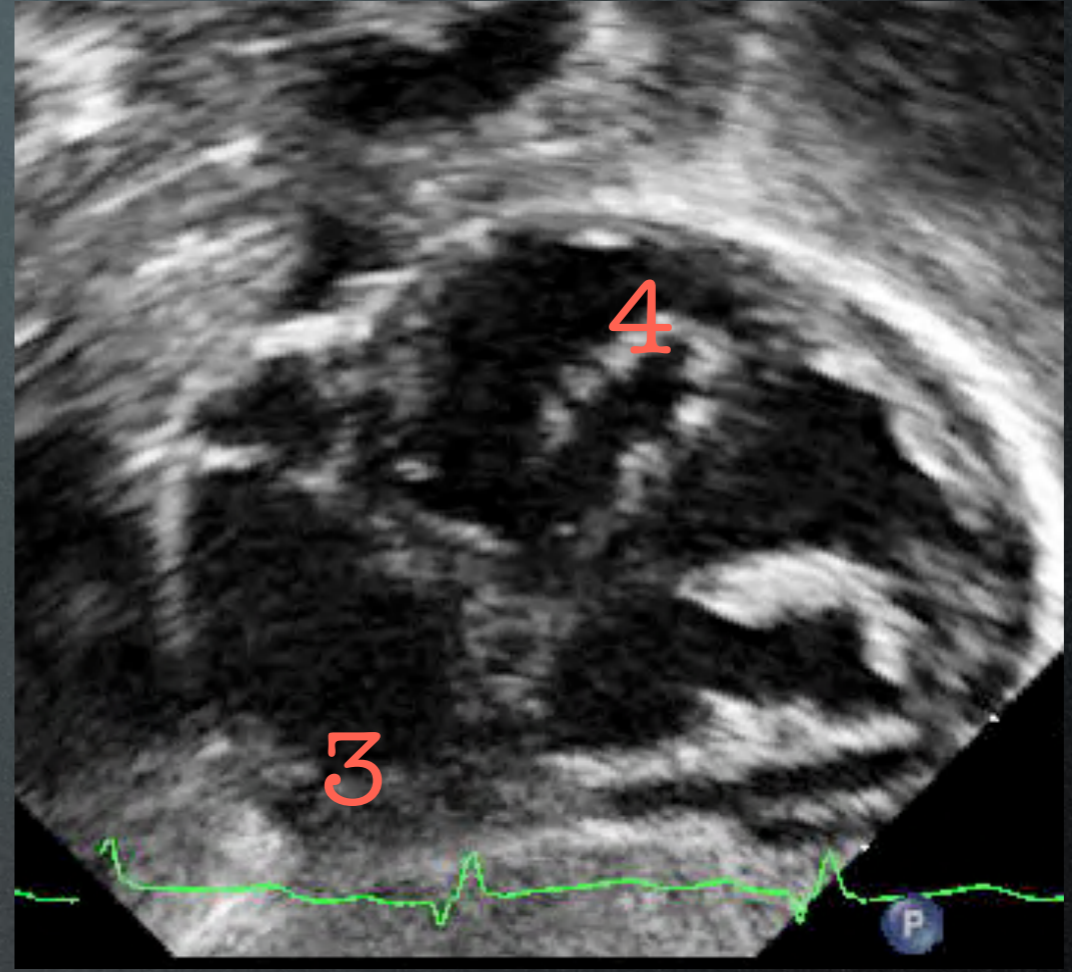
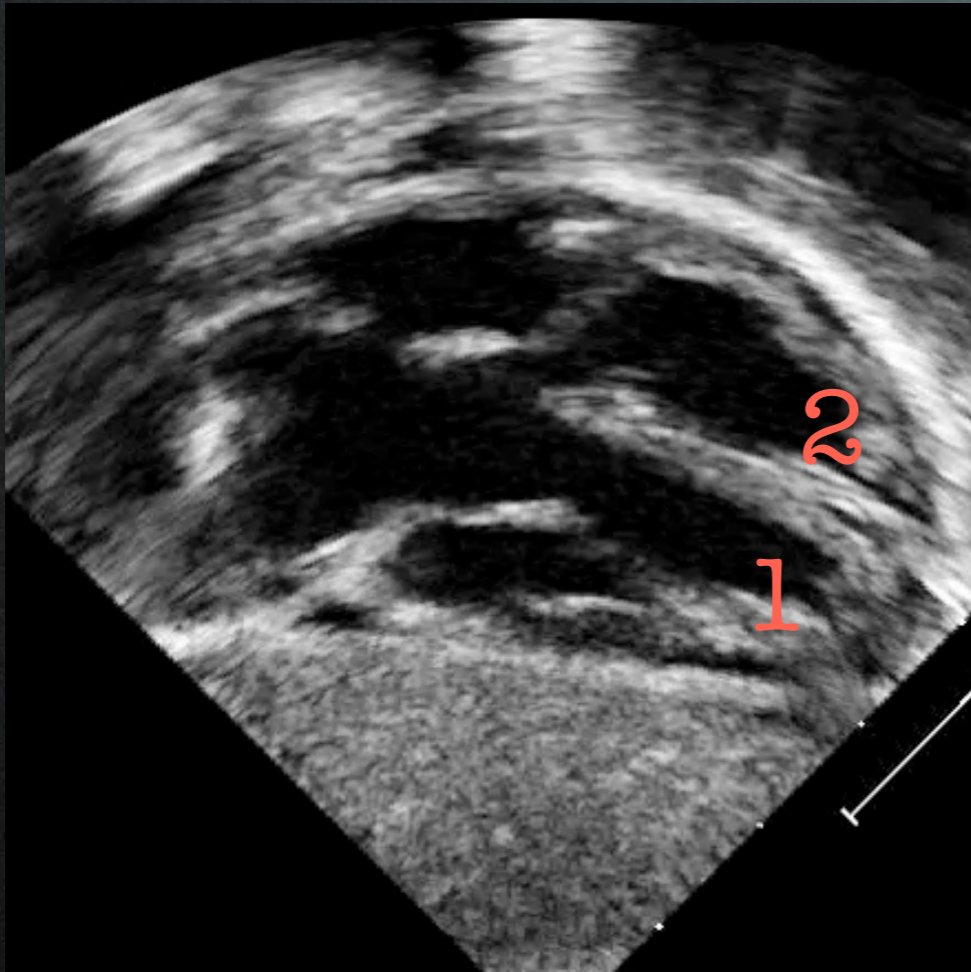
Case: Clinical history

- Born at term through a c-section
- Apgar scores: 9/9, vigorous crying
- Started PGE 6 min after birth
- Stable cardiac output on PGE
- SpO₂: pre 99%, post 87%
- Distally cyanotic

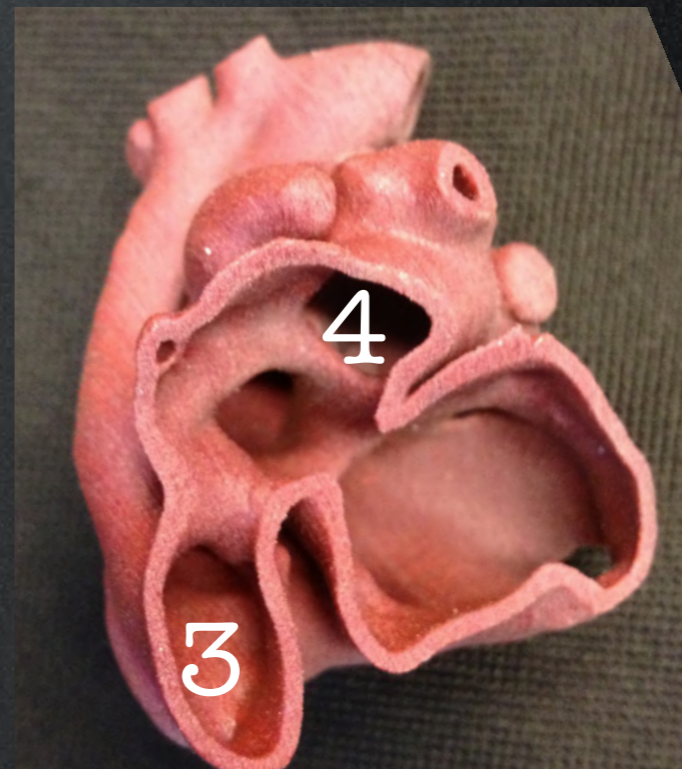
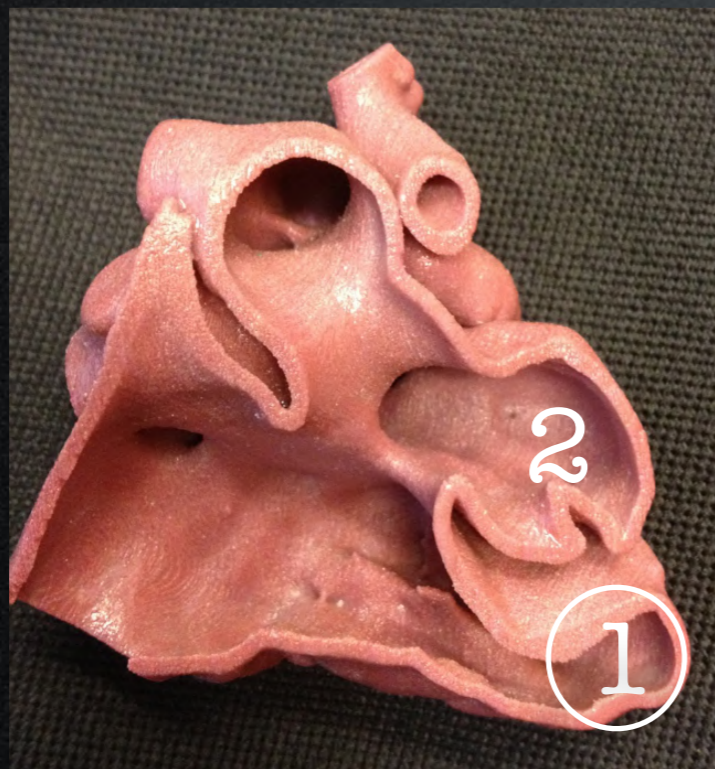
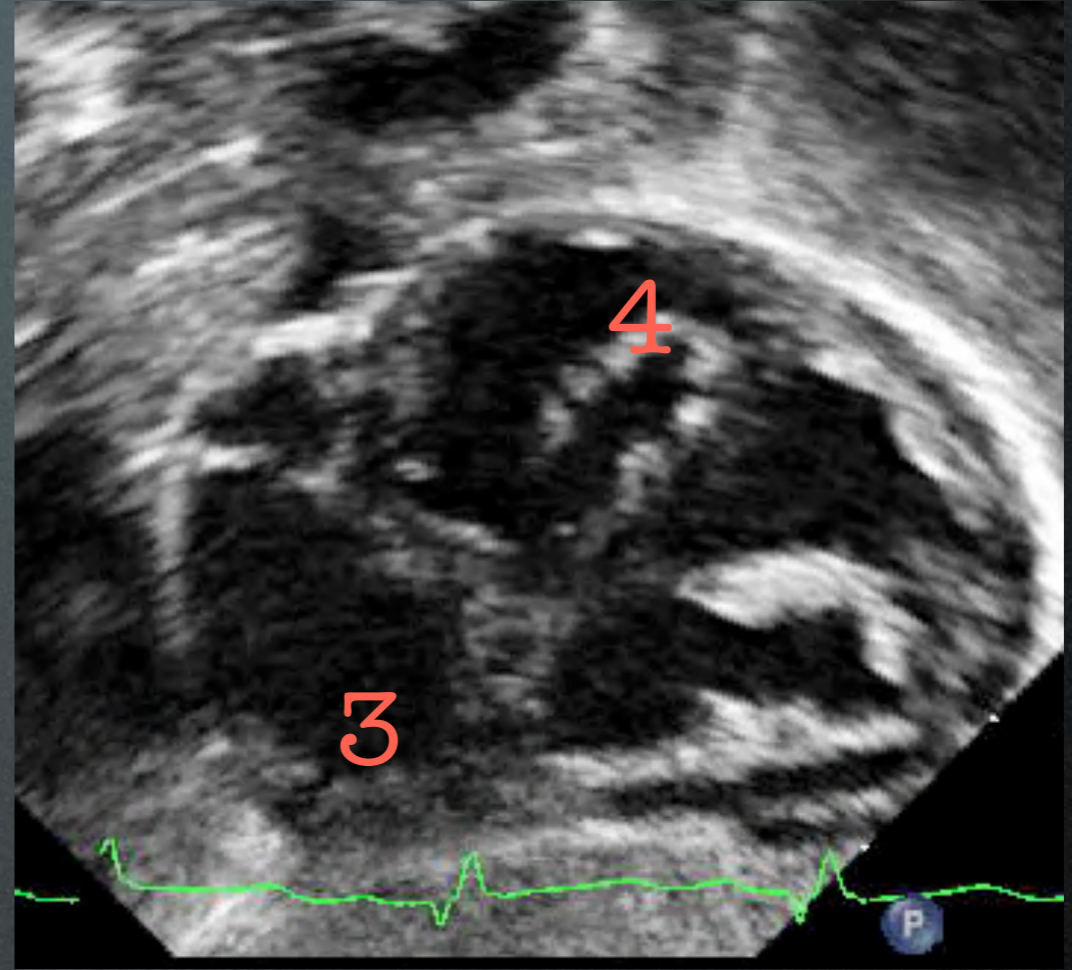
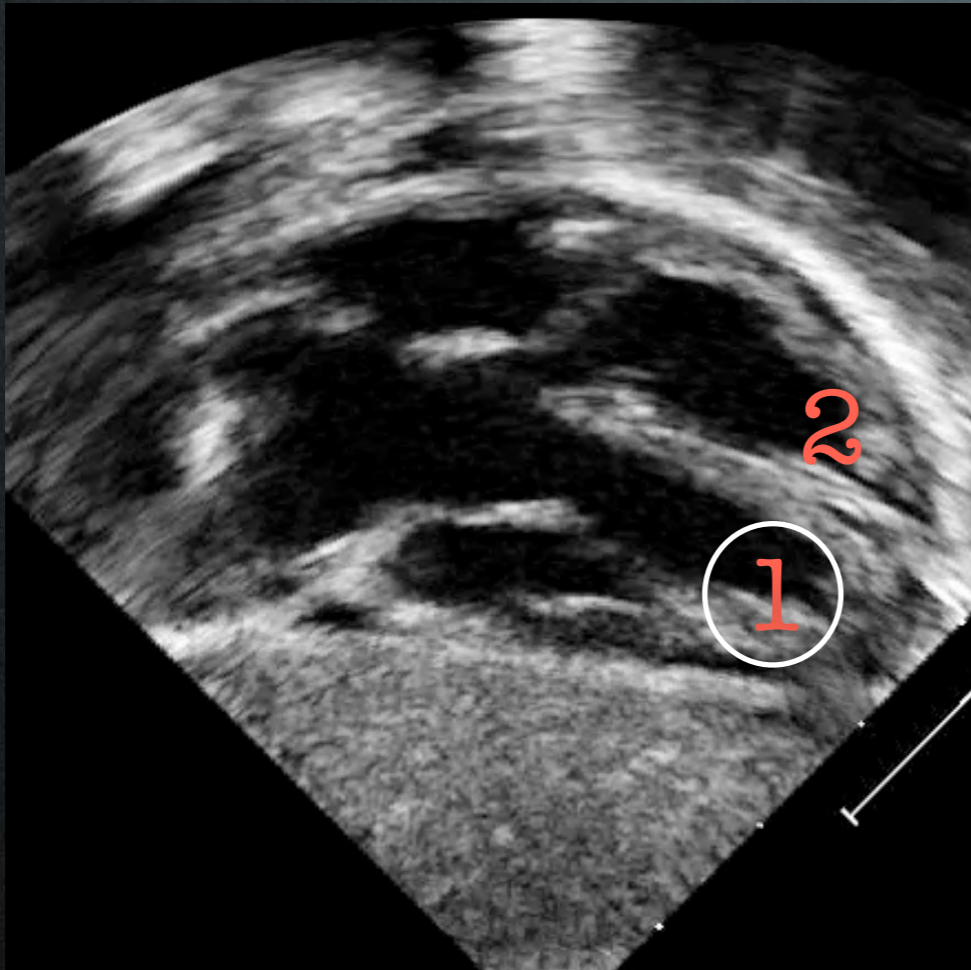




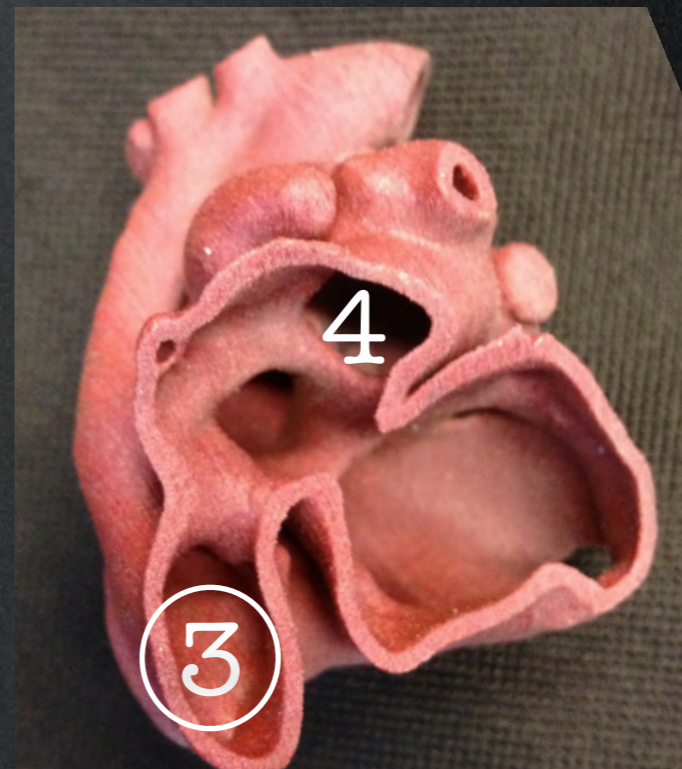
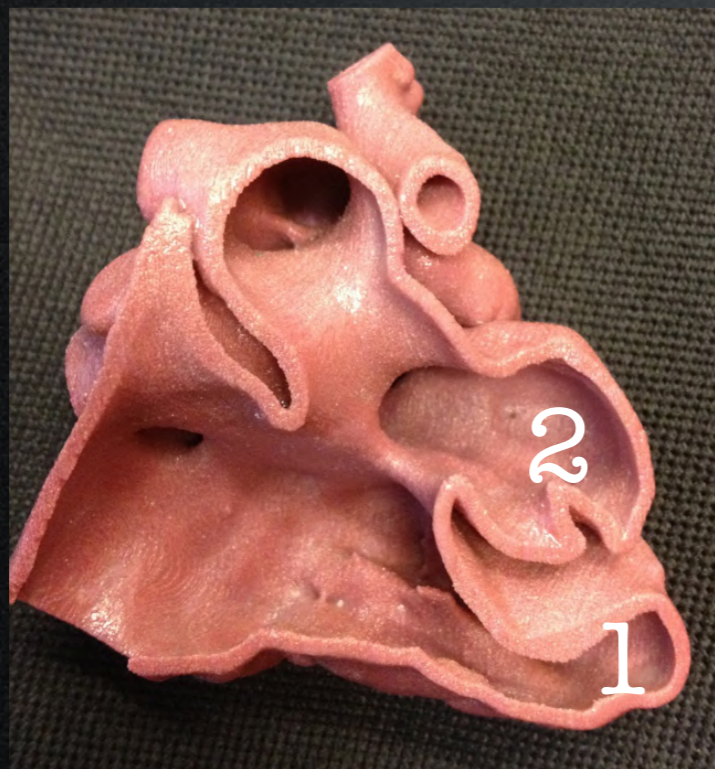
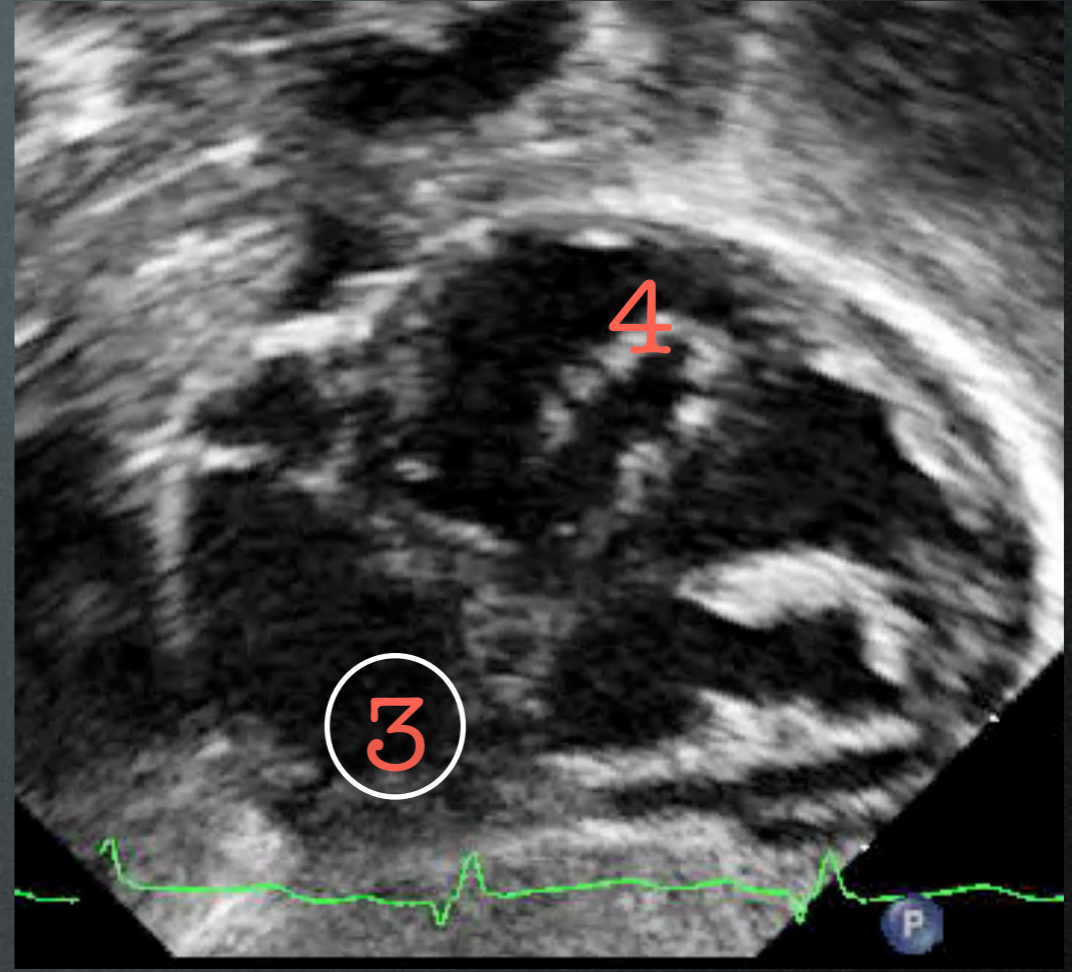
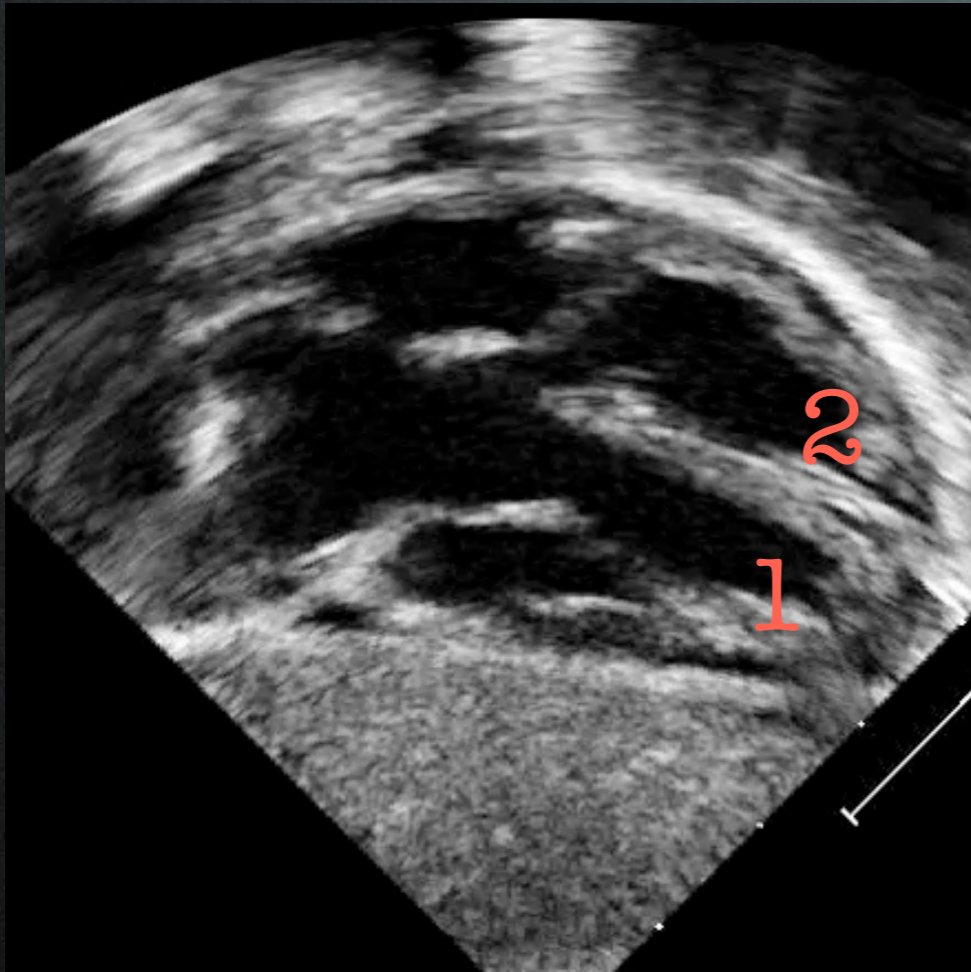
Q. Where are the left and right ventricular apices?



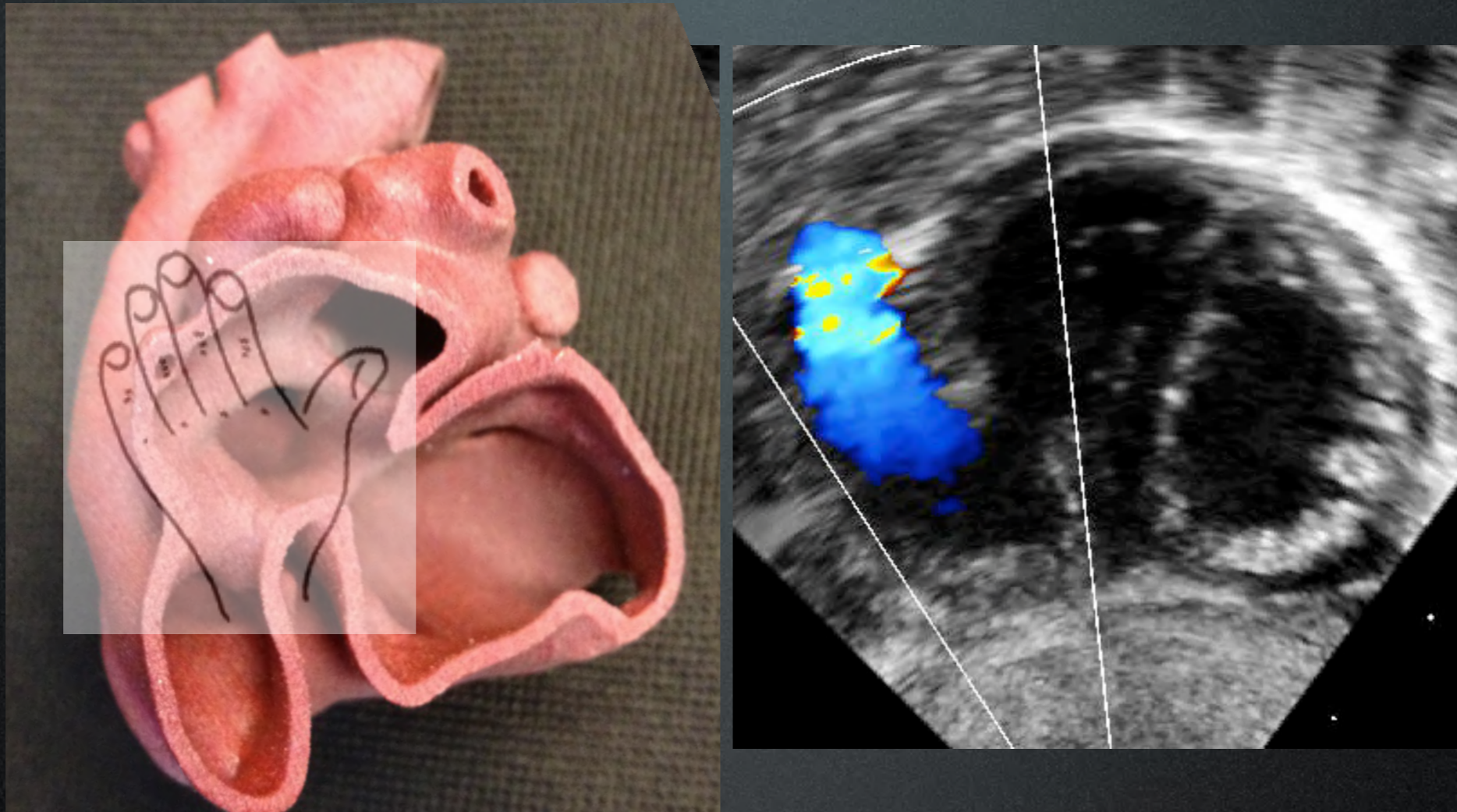
Q1. Where is the left ventricular apex?



Q2. Where is the right ventricular apex?

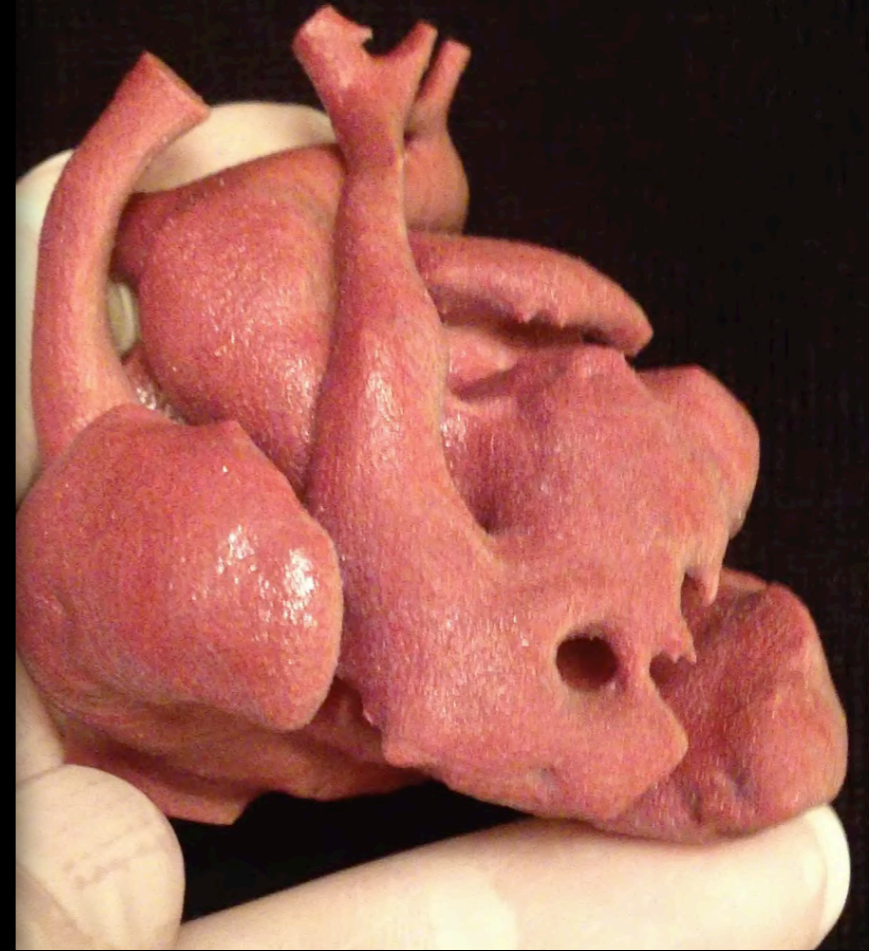
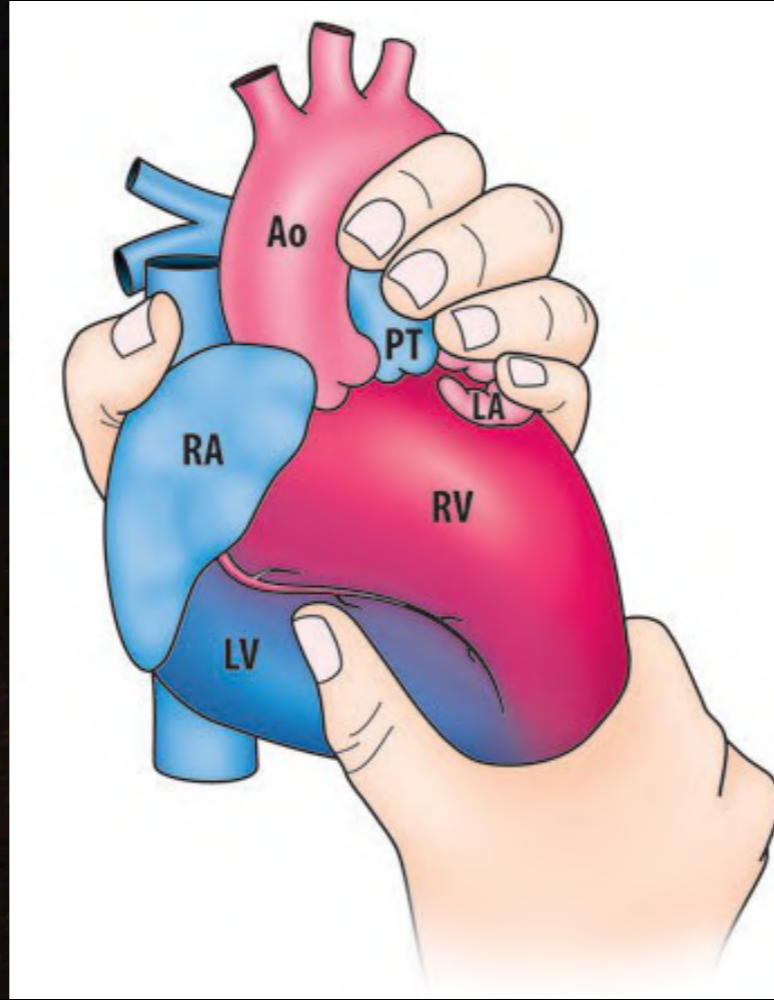


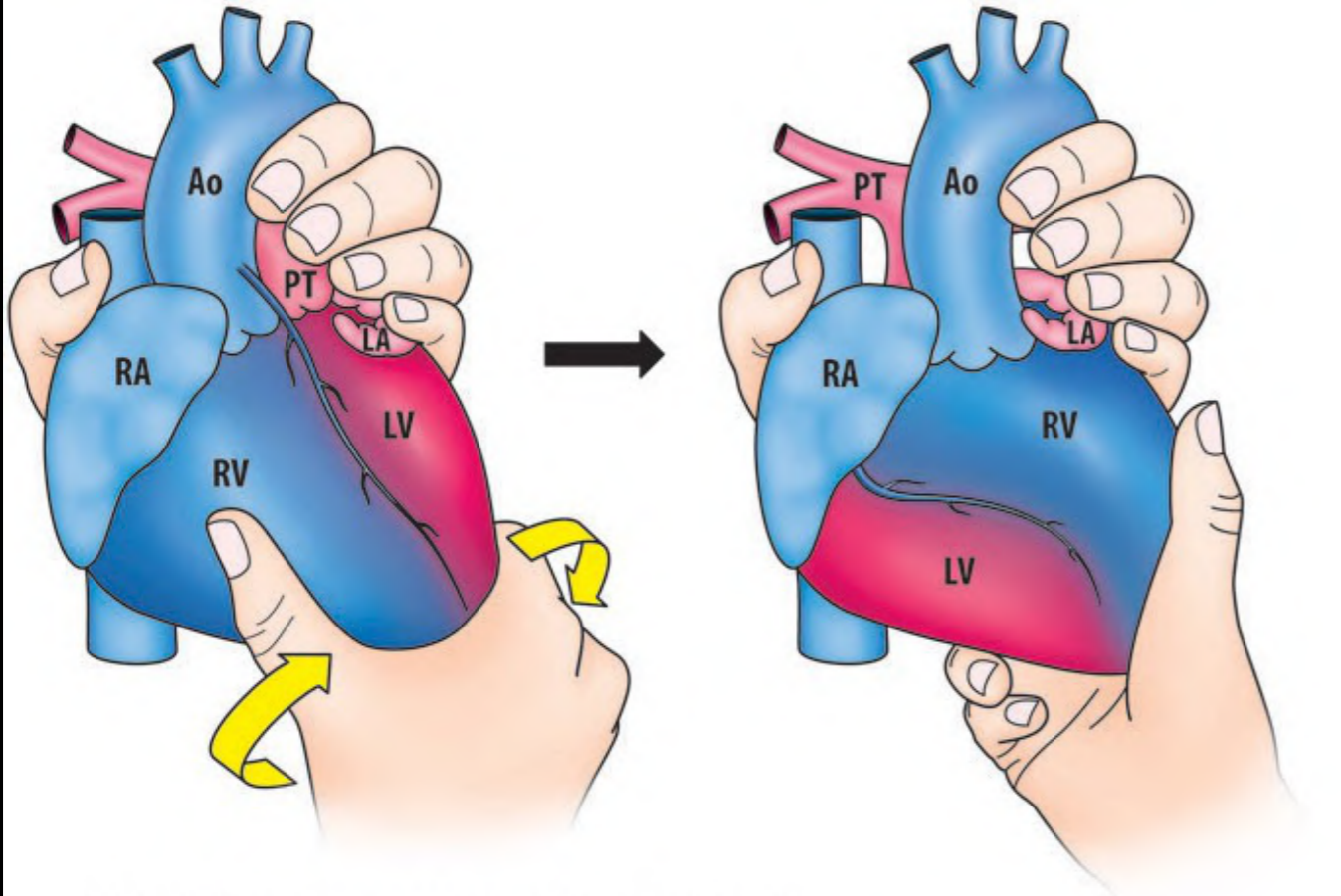
Q3. What is the ventricular topology or situs in this case?



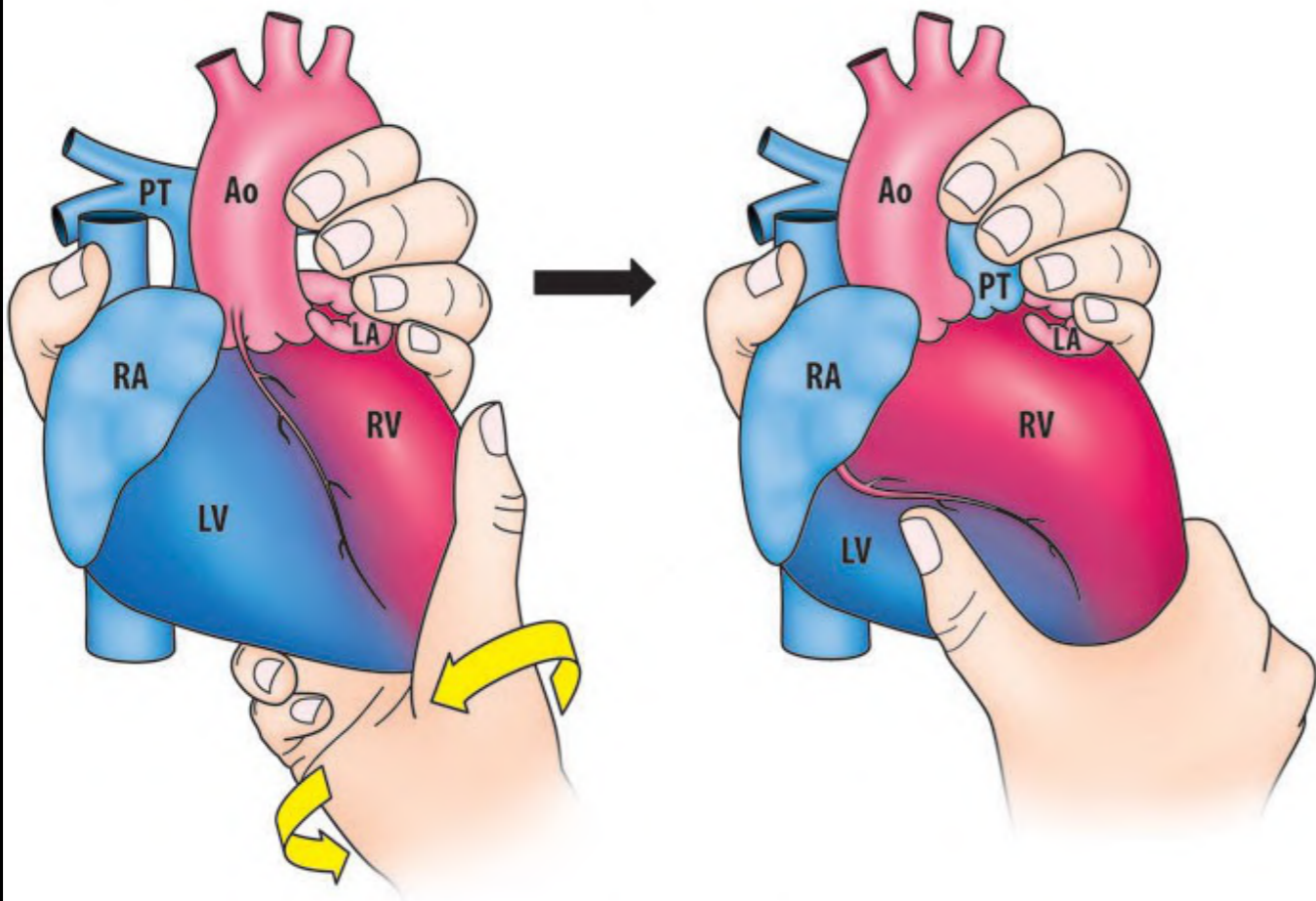
1. Right-hand pattern or D-loop
- ② Left-hand pattern or L-loop
3. Two-hands pattern or A-loop
4. None of the above

A newborn with congenitally corrected TGA (Case 5)





A. Classic Complete Transposition of the Great Arteries



B. Congenitally Corrected Transposition of the Great Arteries

Twisted Heart

Non-parallel AV connection axes and unexpected chamber position and orientation

NATURE OF TWISTING

- Along the base-apex axis
- To place the RV inlet superior and anterior to LV inlet in most (not all) cases
 - ➔ Clockwise twist in D-loop ventricles
 - ➔ Counterclockwise twist in L-loop ventricles

ESSENTIAL FINDINGS

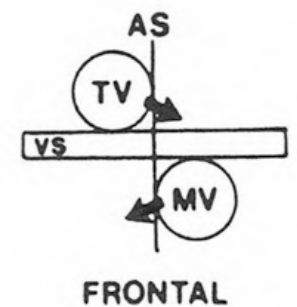
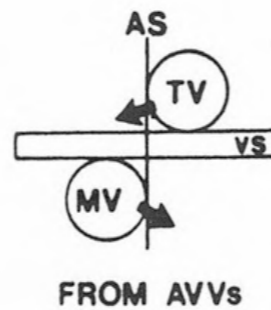
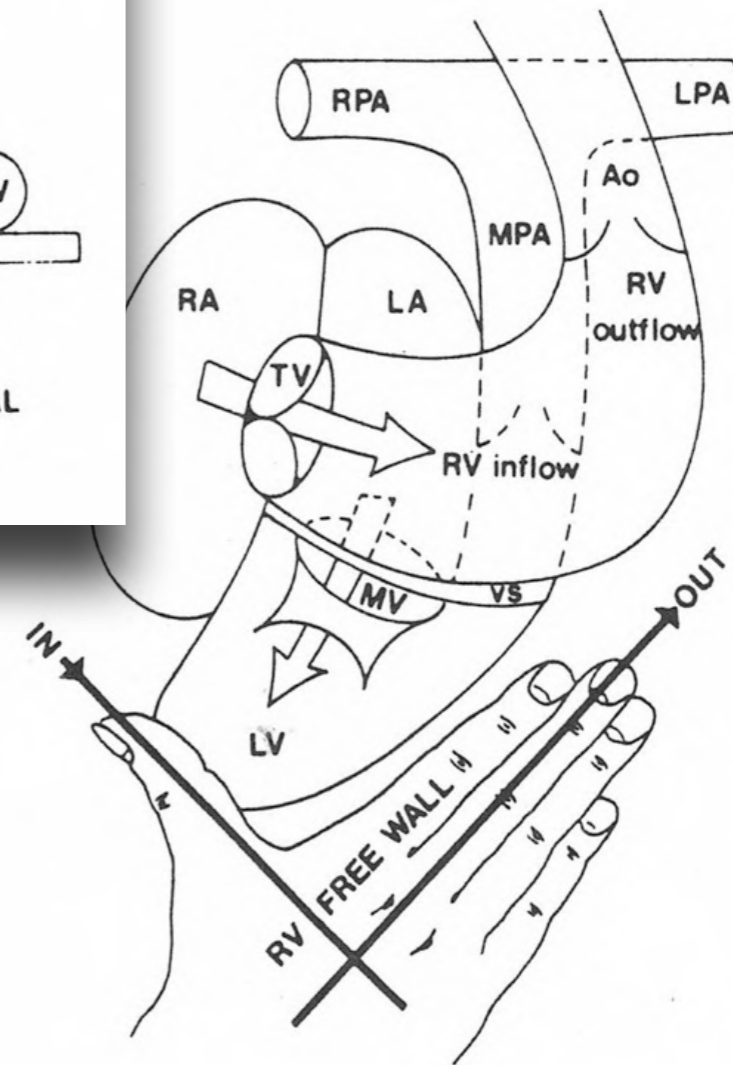
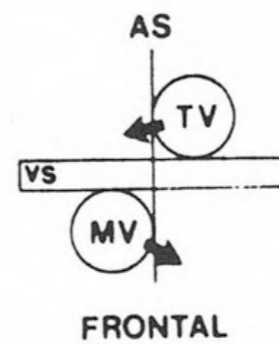
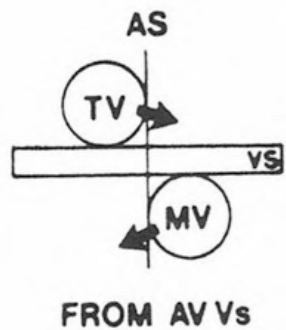
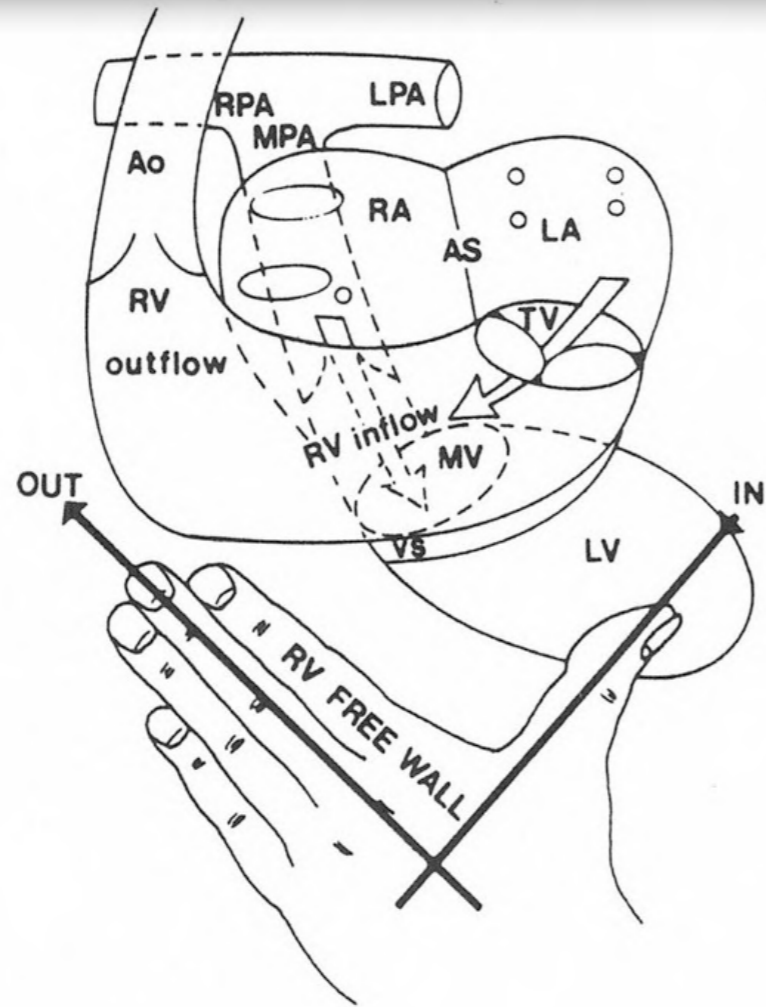
- Non-parallel opening axes of AV valves
- Unexpected ventricular relationship for the given AV connection
- Angled atrial septum
- Curved ventricular septum
- Unexpected great arterial relationship

FEATURES OF SURGICAL IMPORTANCE

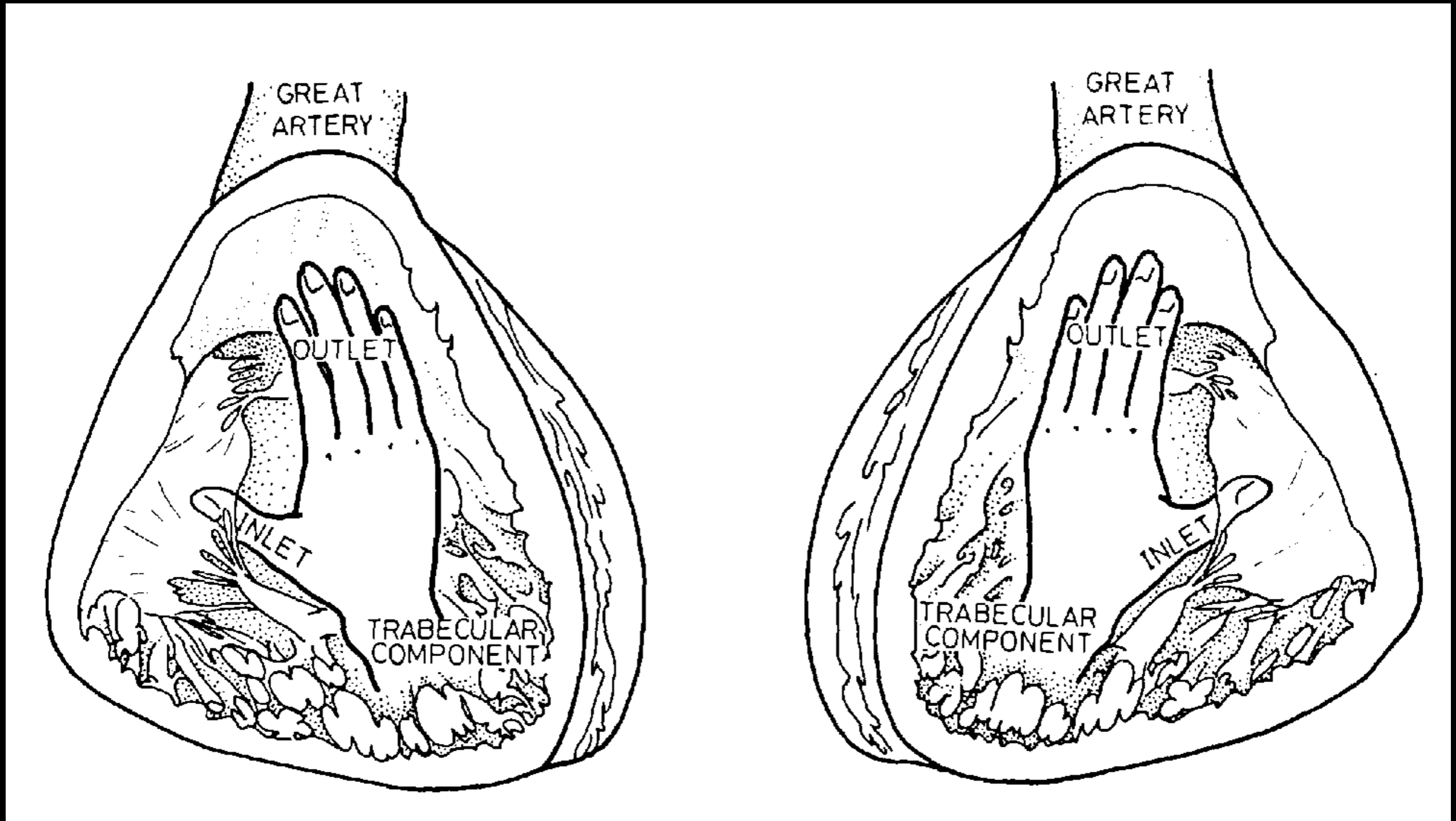
- Juxtaposition of the atrial appendages
- VSD, usually inlet
- AV valve abnormalities:
 - Hypoplasia
 - Straddling, overriding
- Ventricular hypoplasia, usually RV
- Abnormal AV conduction axis
- Abnormal VA connection
- Ventricular outflow tract obstruction, more often pulmonary outflow

VENTRICULAR CHIRALITY

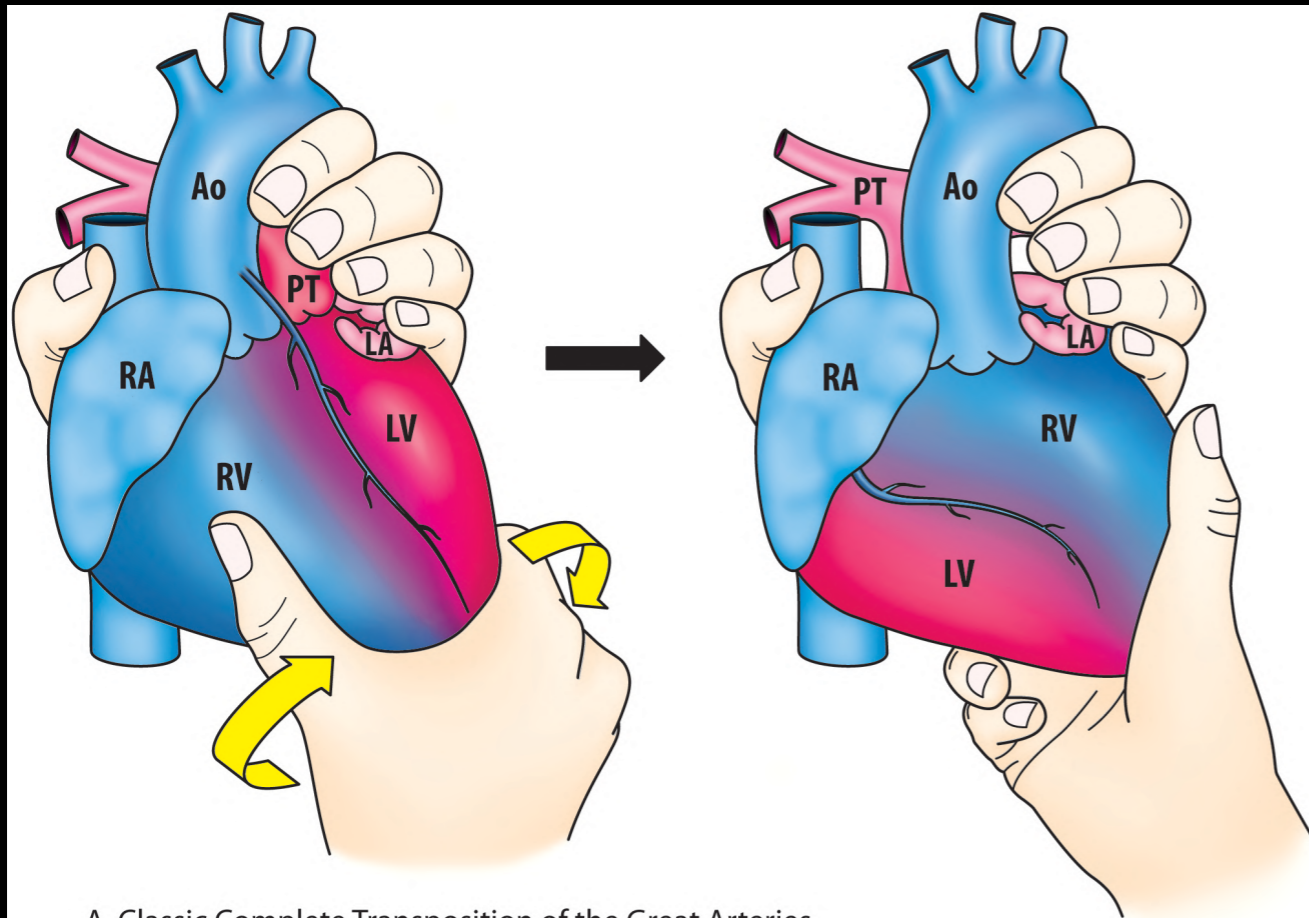
Van Praagh S, et al. In Van Praagh R and Takao A eds. Etiology and morphology of congenital heart disease. Futura,



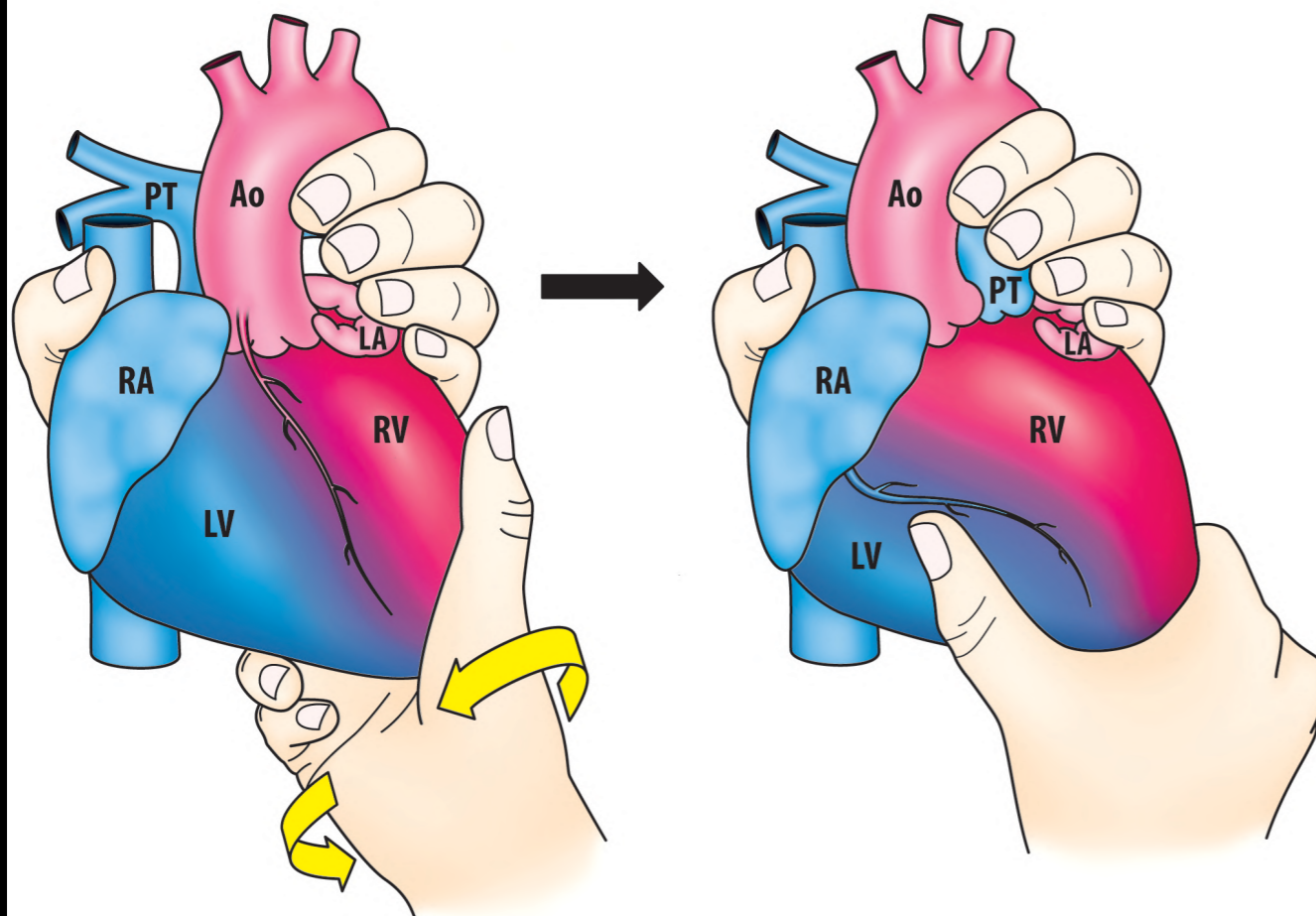
VENTRICULAR ARCHITECTURE OR TOPOLOGY



Anderson RH. A question of definition. Criss-cross heart revisited. *Pediatr Cardiol* 1982;3:305-313



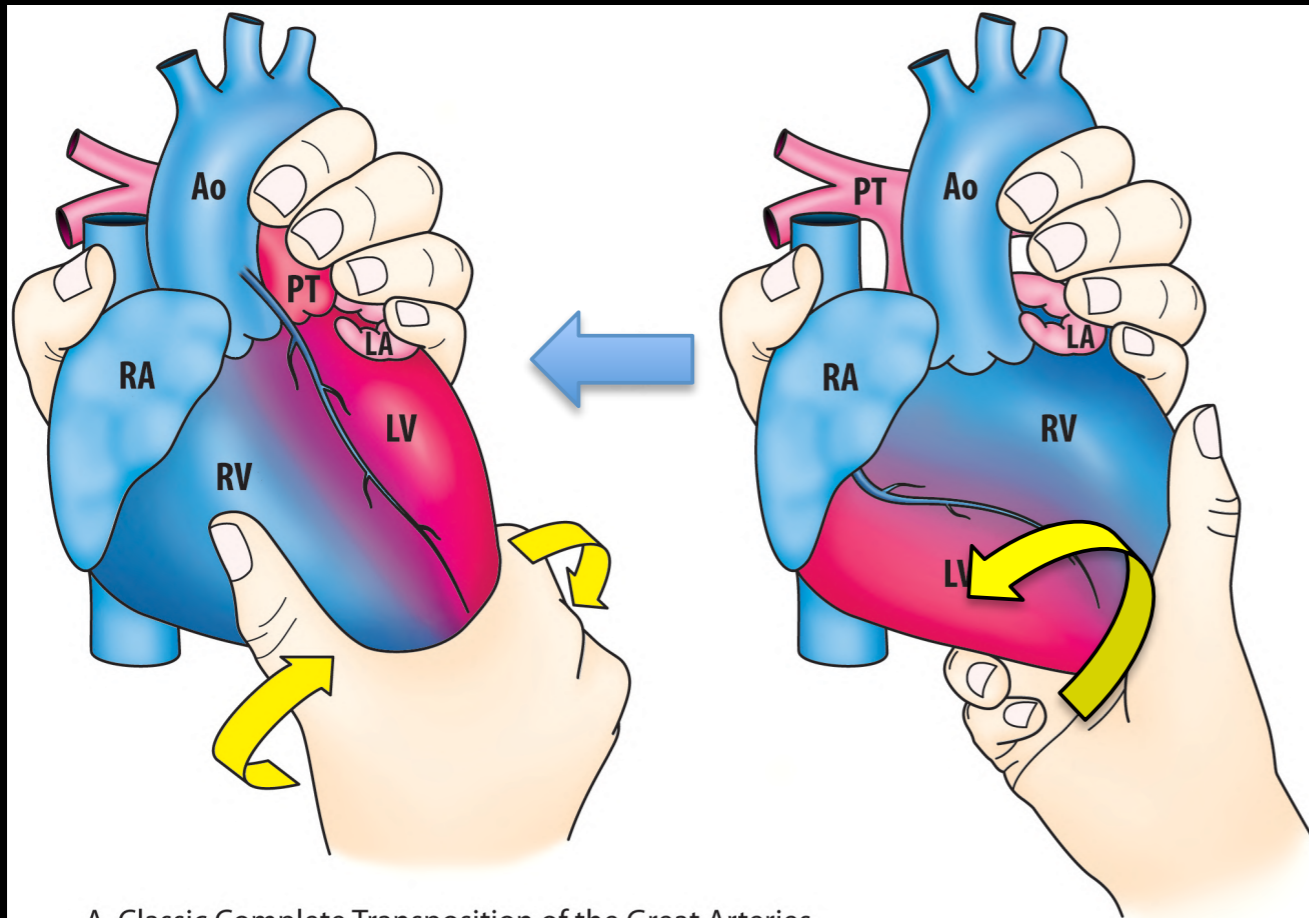
A. Classic Complete Transposition of the Great Arteries



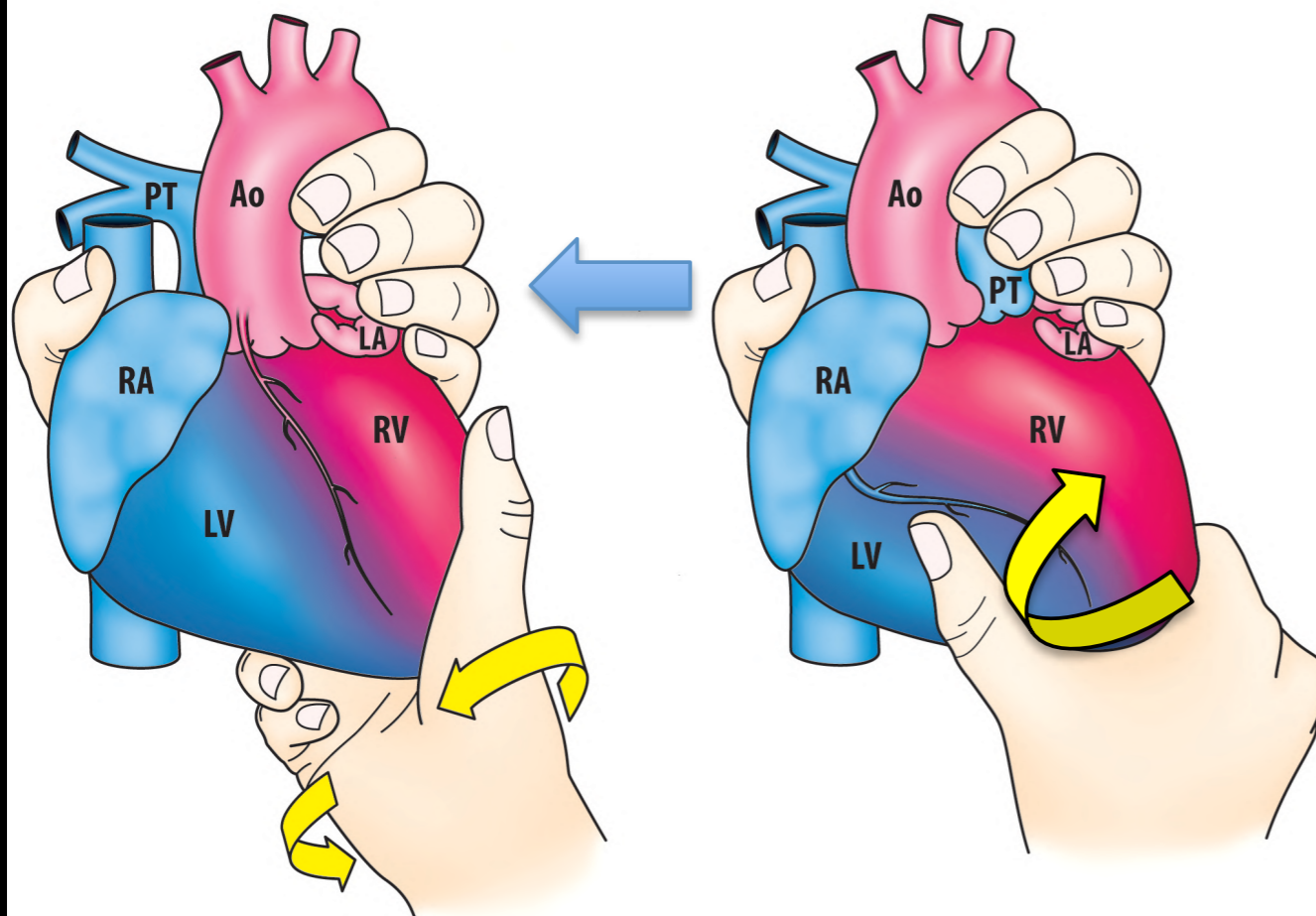
B. Congenitally Corrected Transposition of the Great Arteries

Twisted Heart

Non-parallel
AV connection
axes



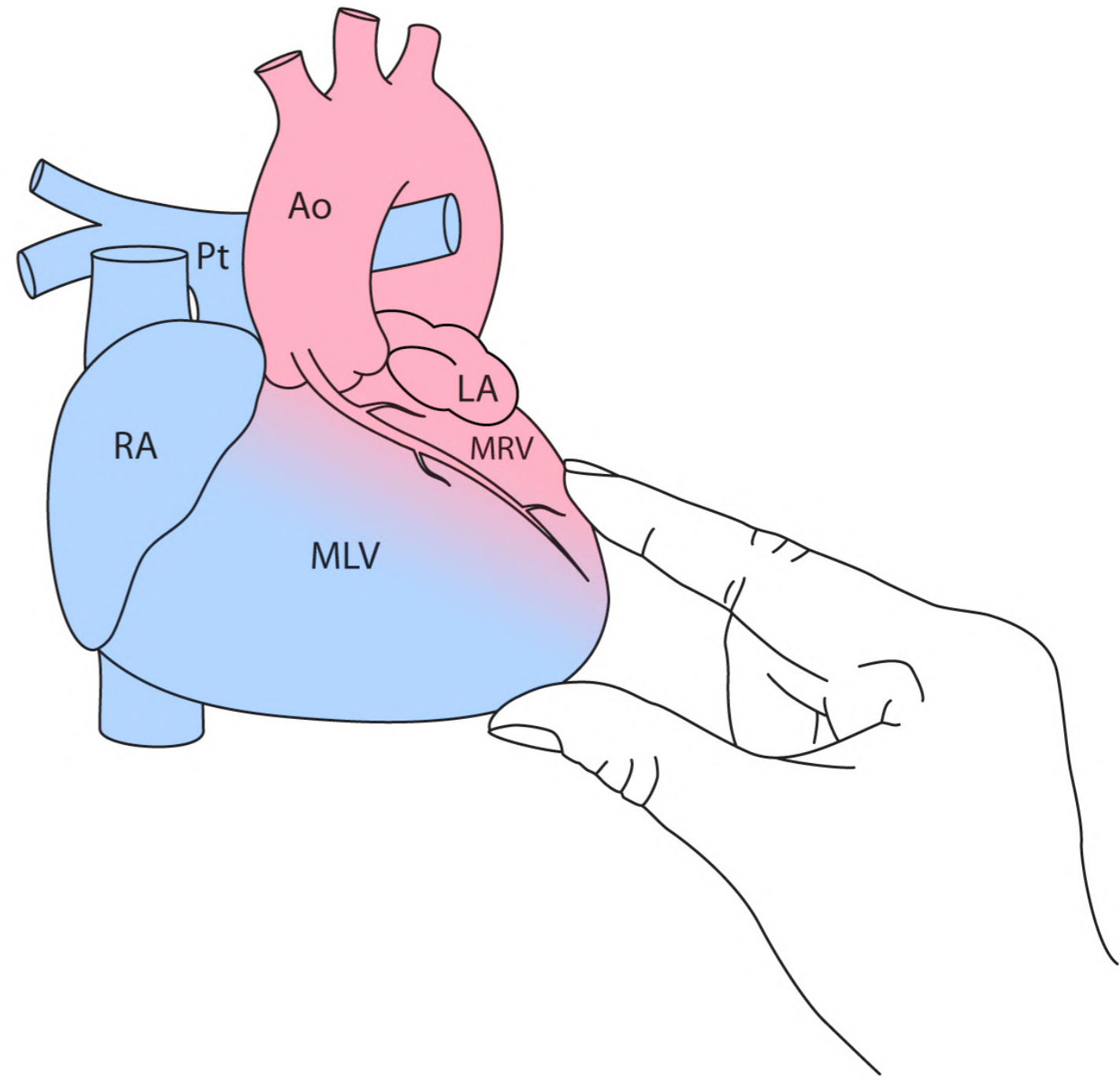
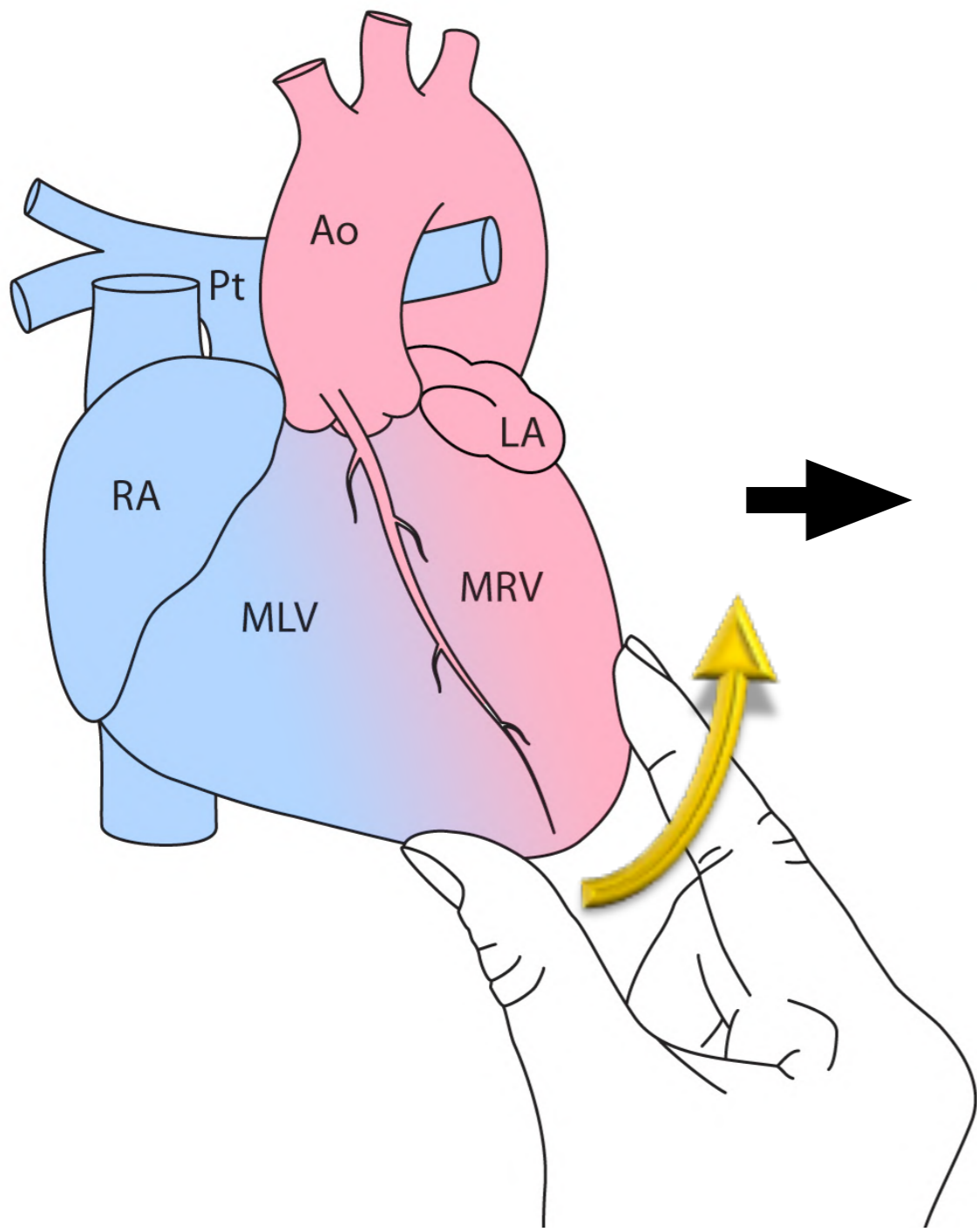
A. Classic Complete Transposition of the Great Arteries



B. Congenitally Corrected Transposition of the Great Arteries

Untwist the Heart

Stop using your
busy hands!



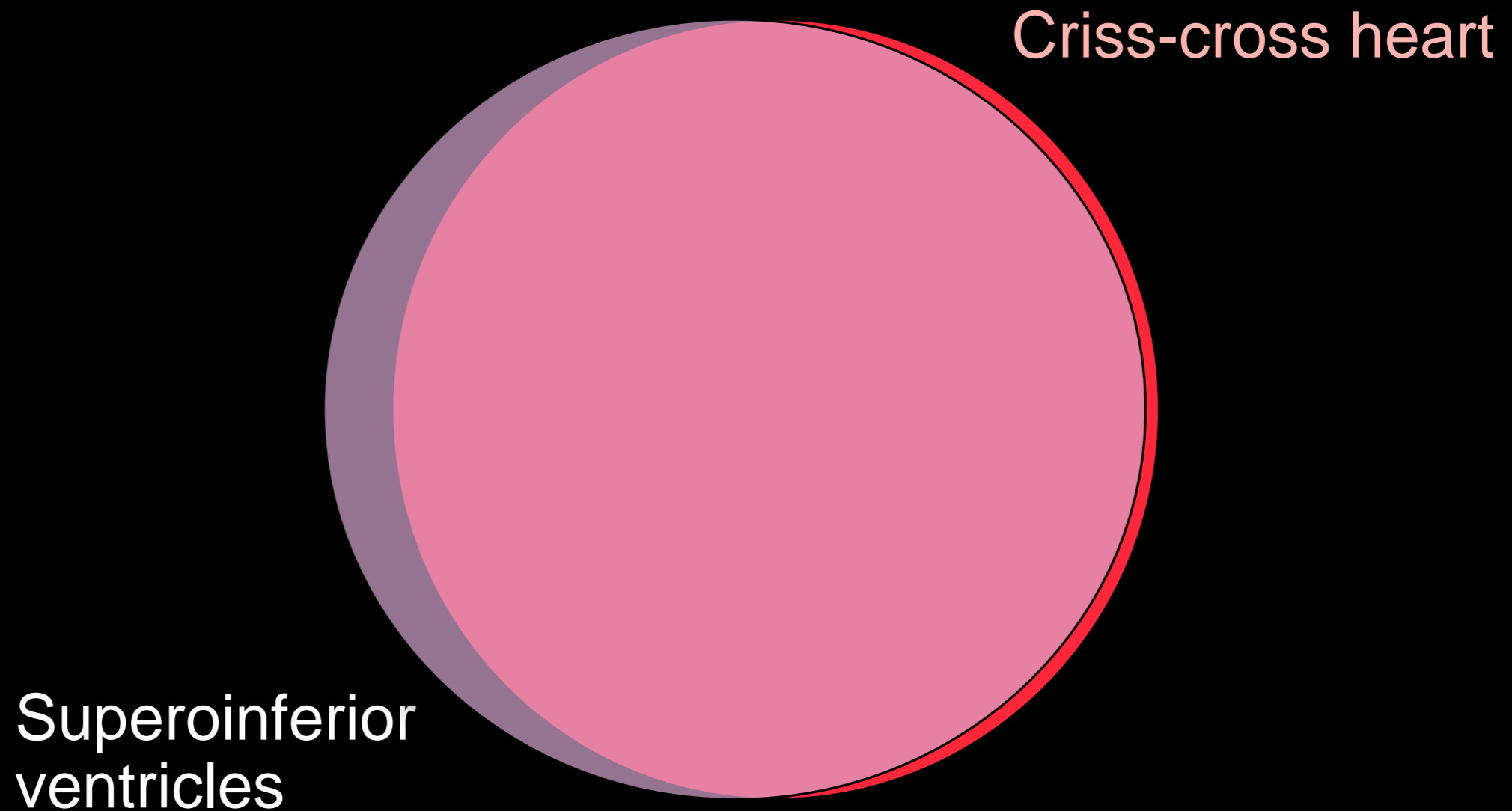
QUESTION:

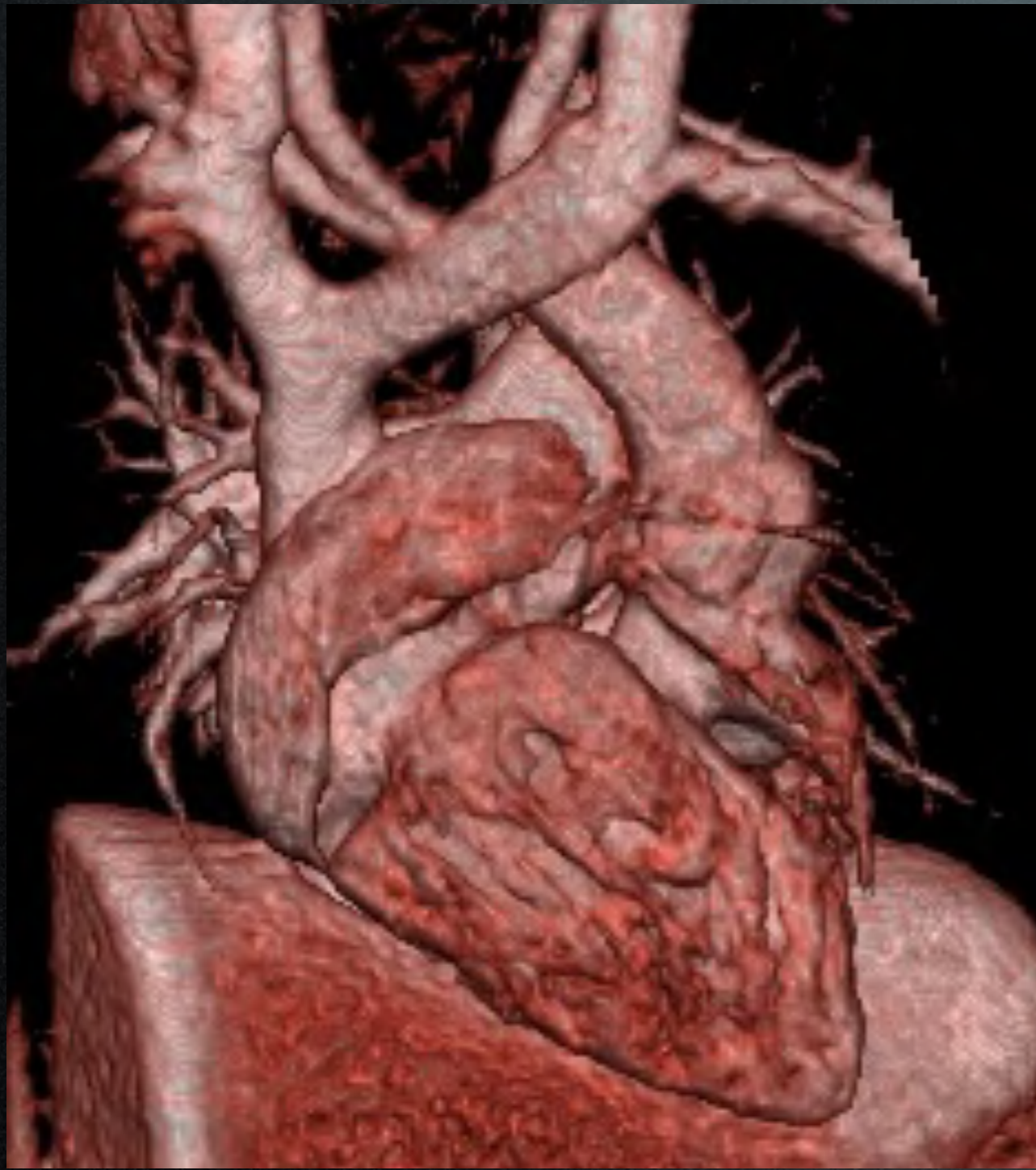
Are 'criss-cross heart' and 'superoinferior ventricles' different entities?

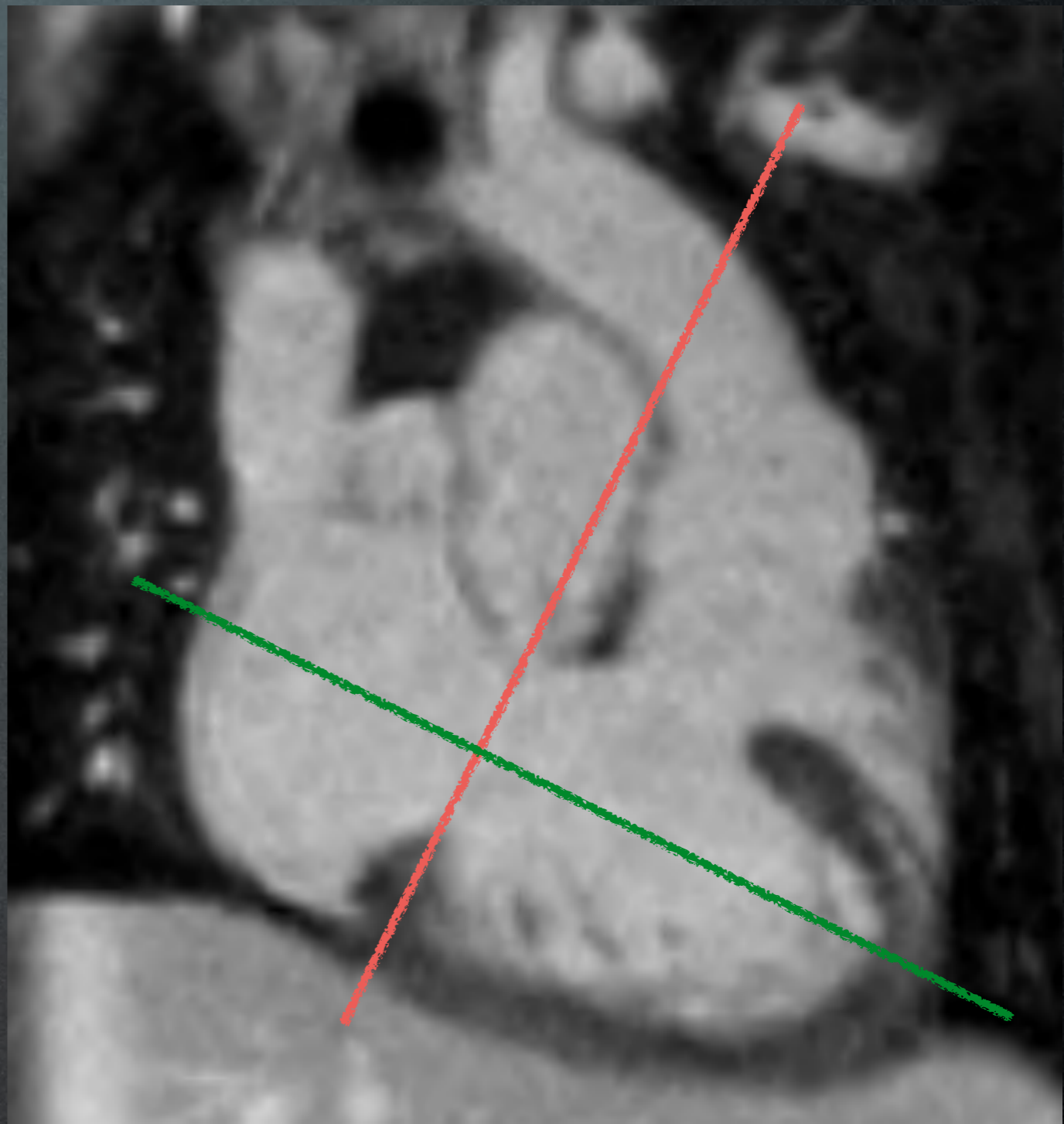
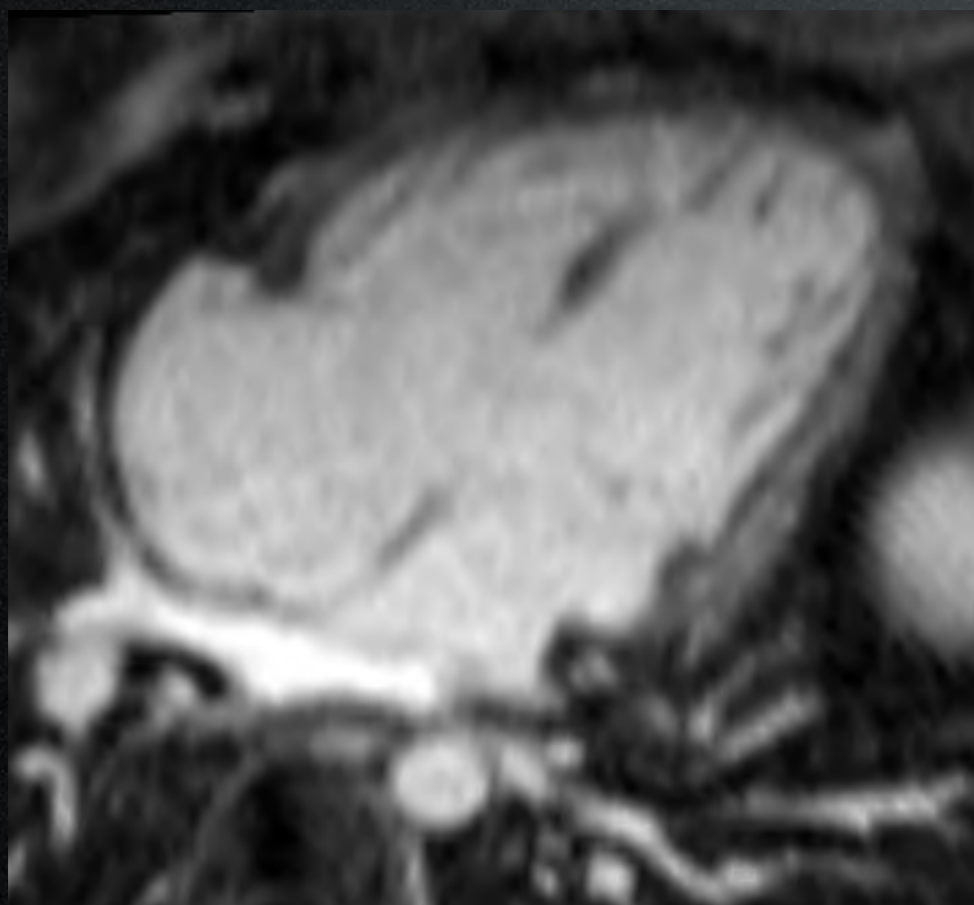
1. Yes
2. No
3. Yes and no

QUESTION:

Are 'criss-cross heart' and 'superoinferior ventricles' different entities?

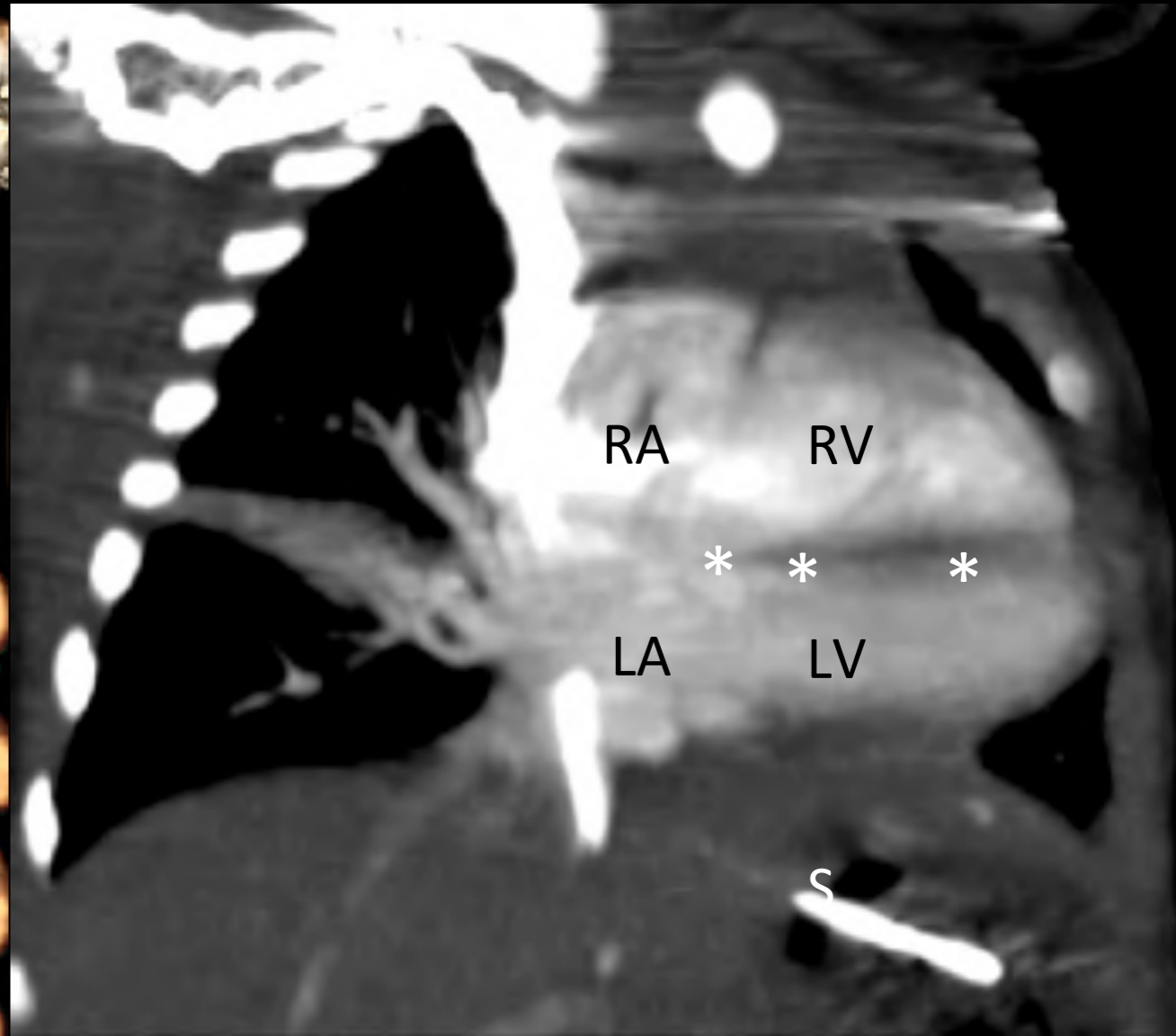
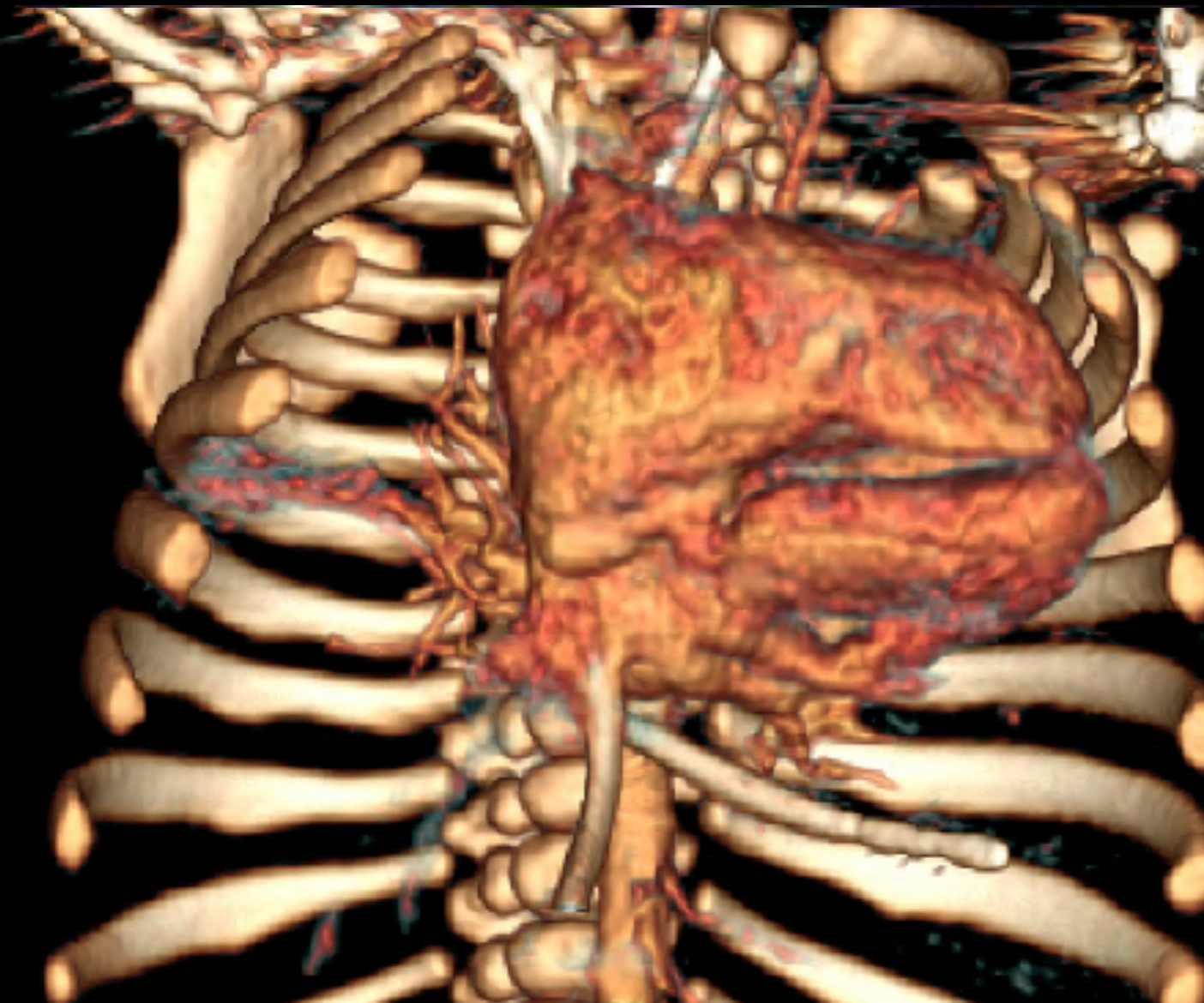


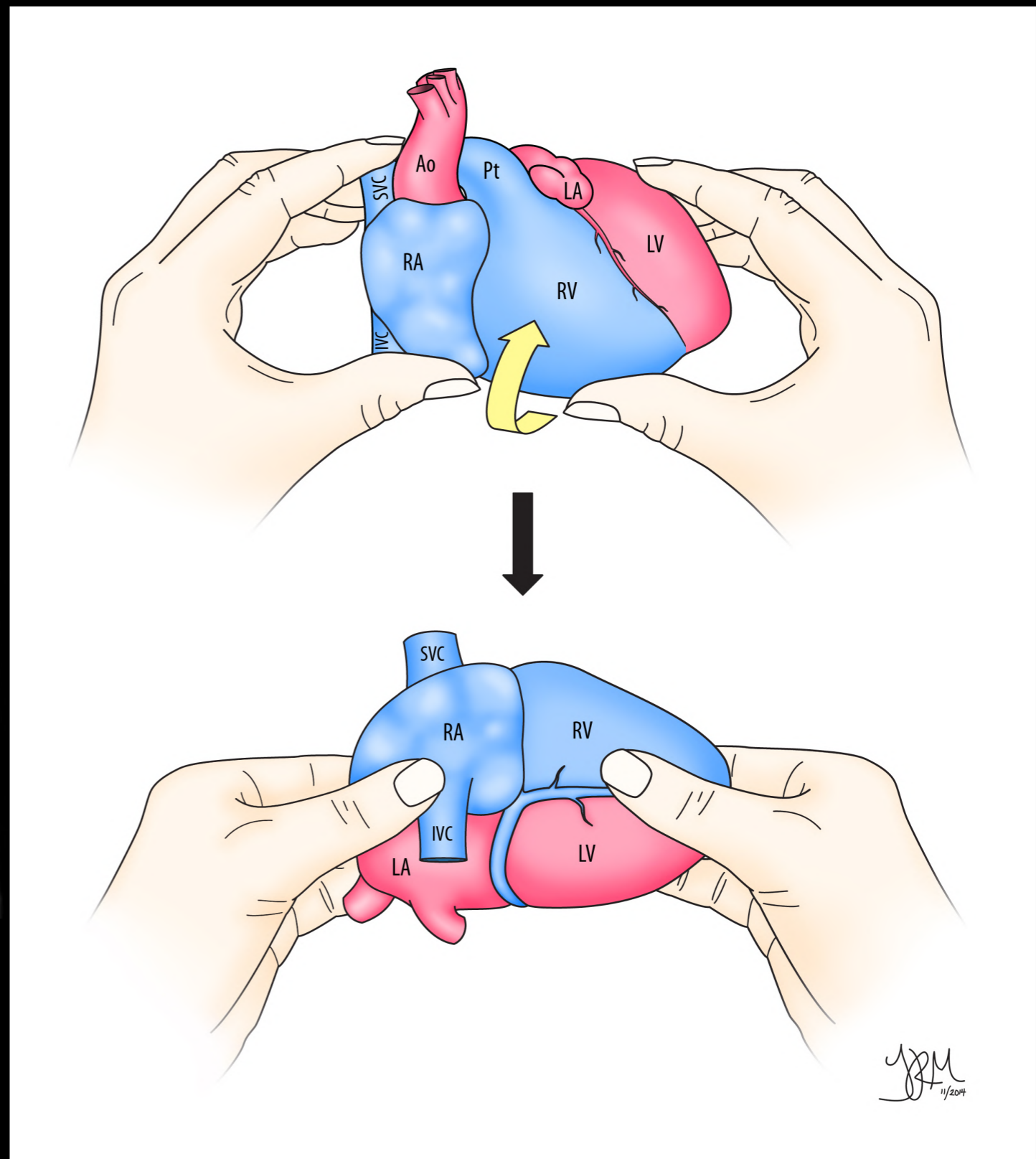
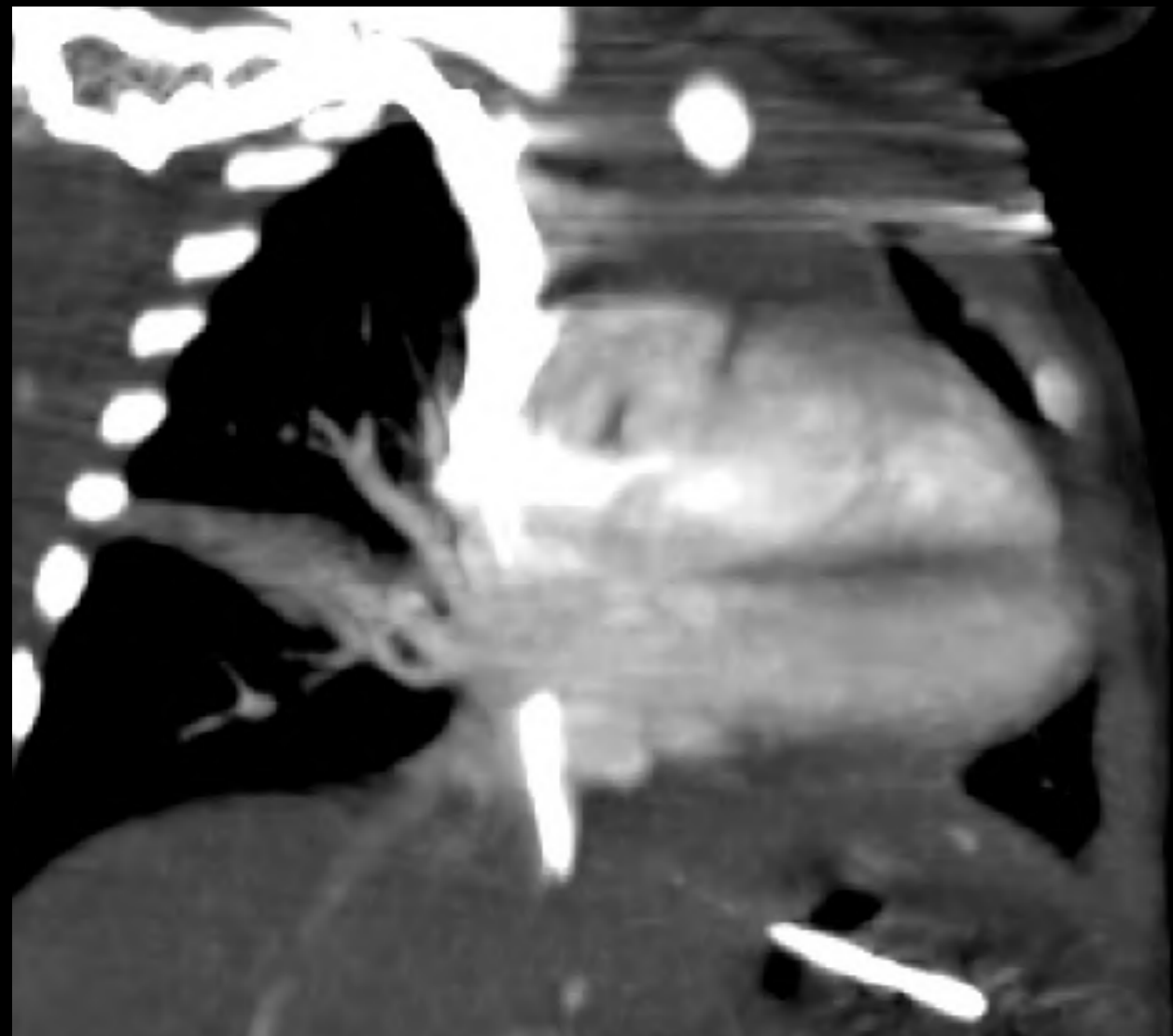


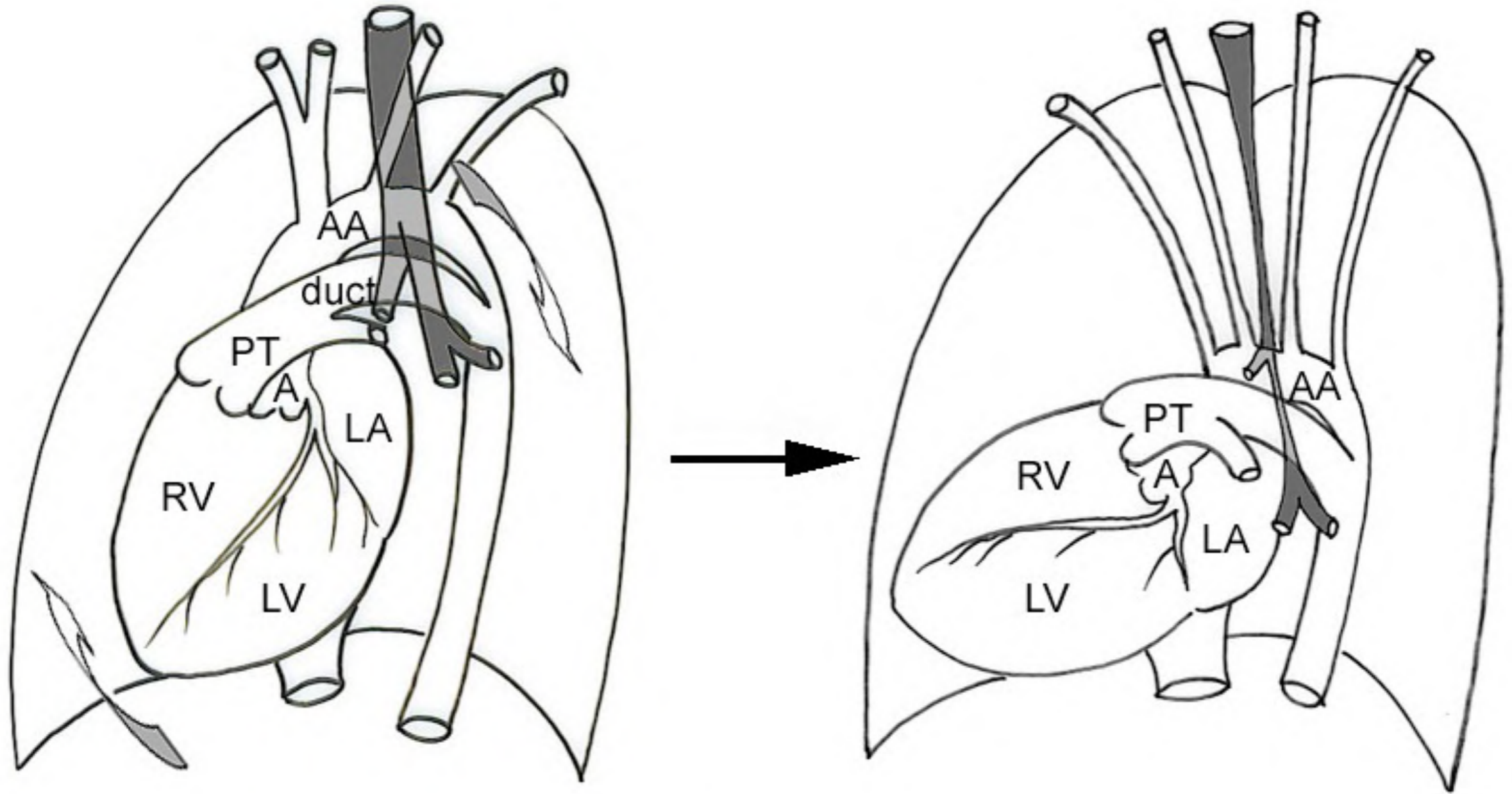


Topsy-Turvy Heart

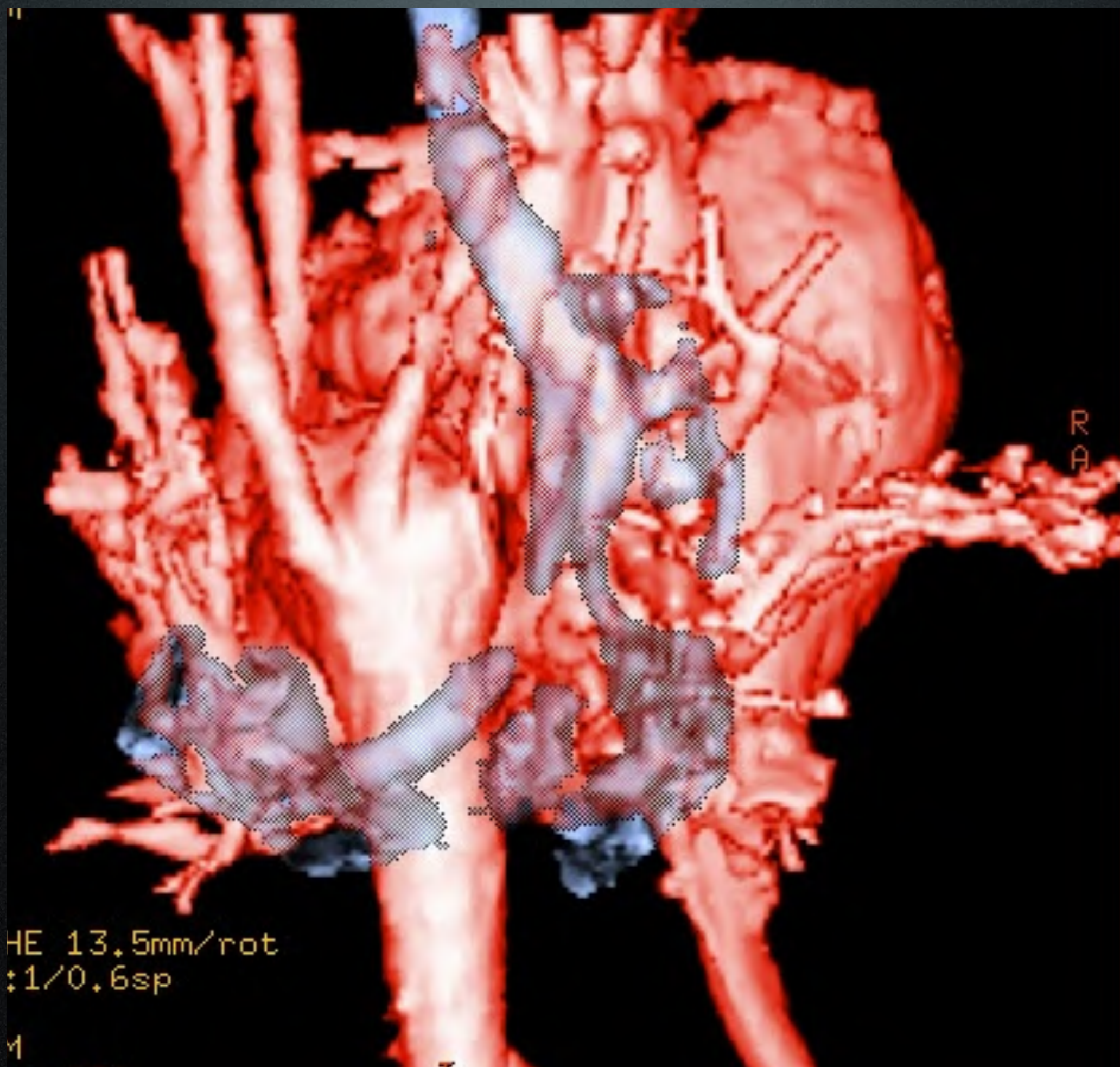
Case: Consanguineous marriage. 1st pregnancy terminated because of suspected HLHS





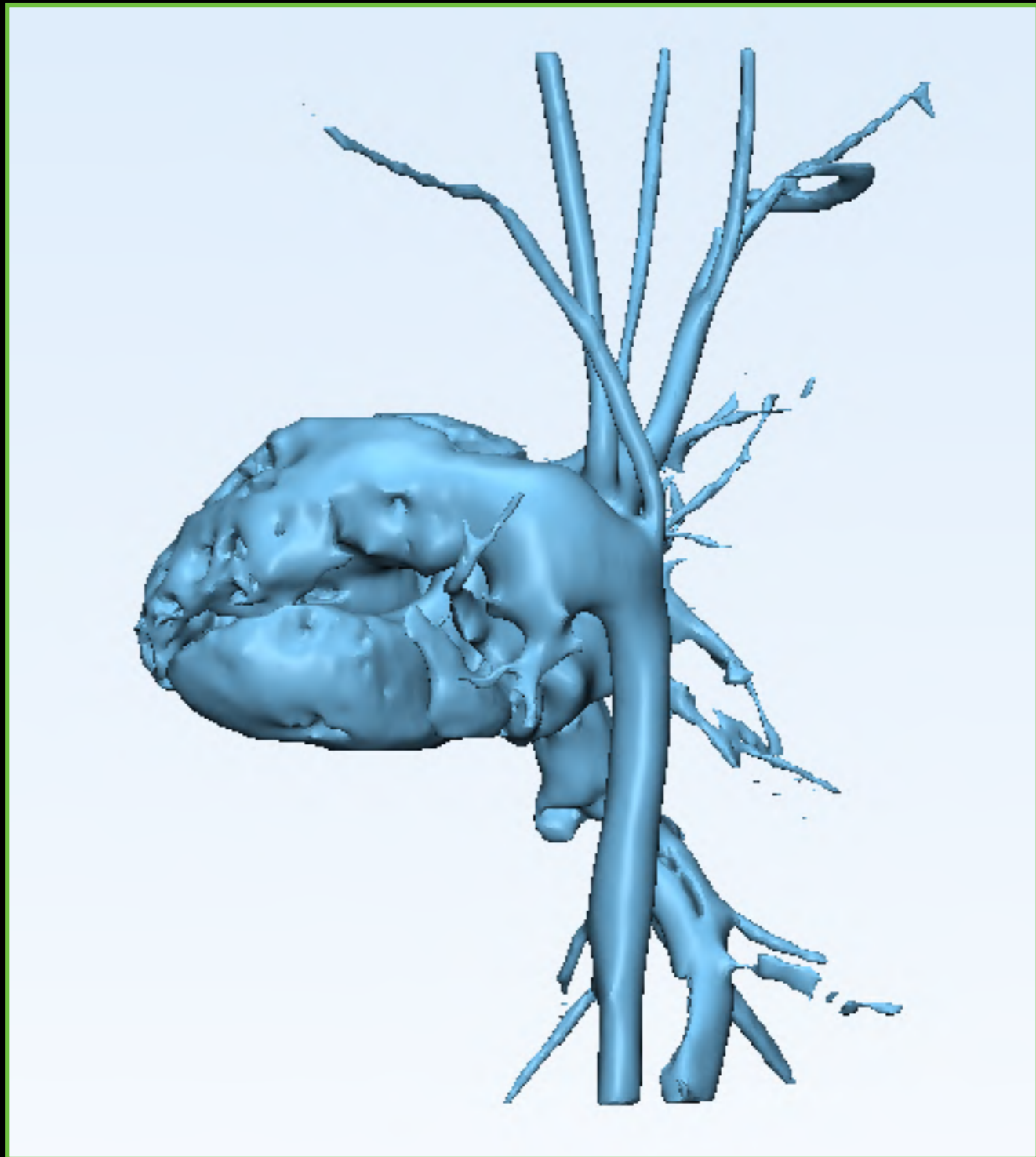
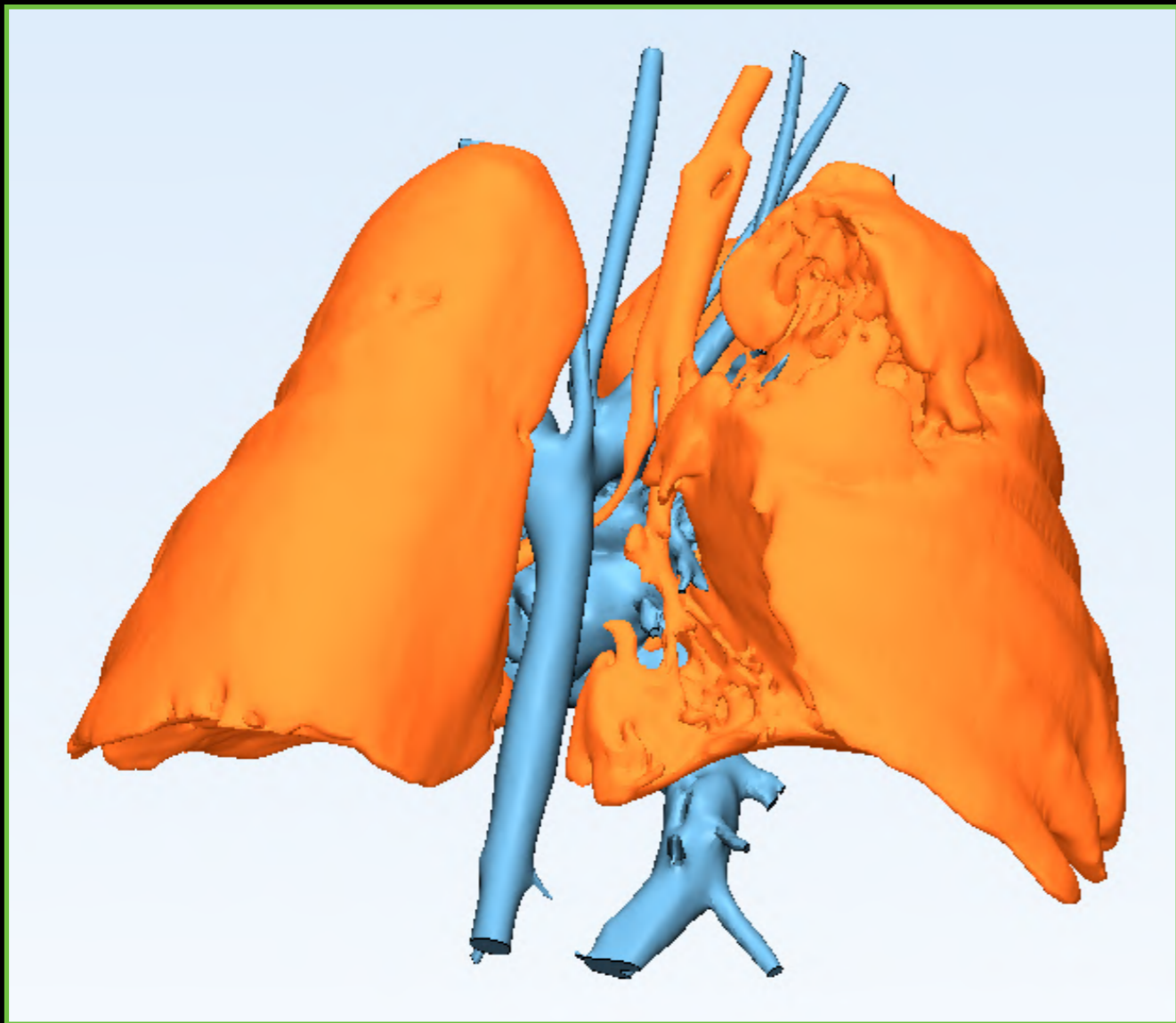


Topsy-Turvy Heart



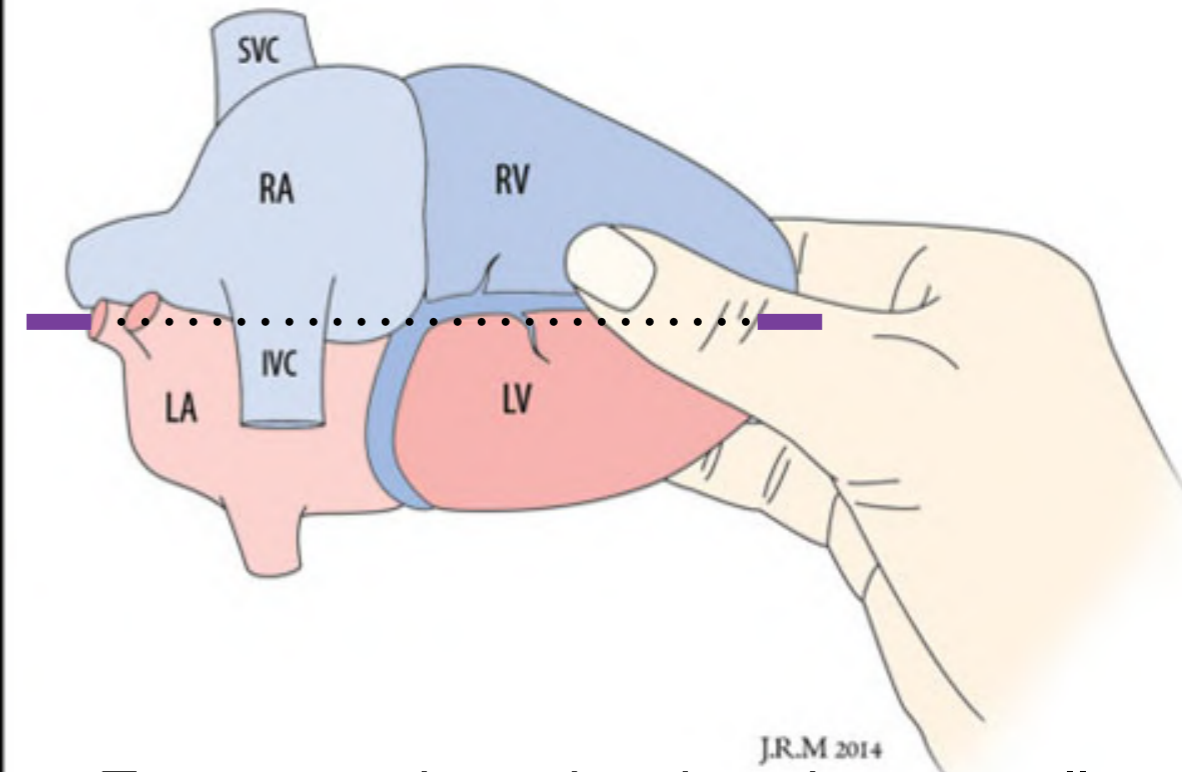
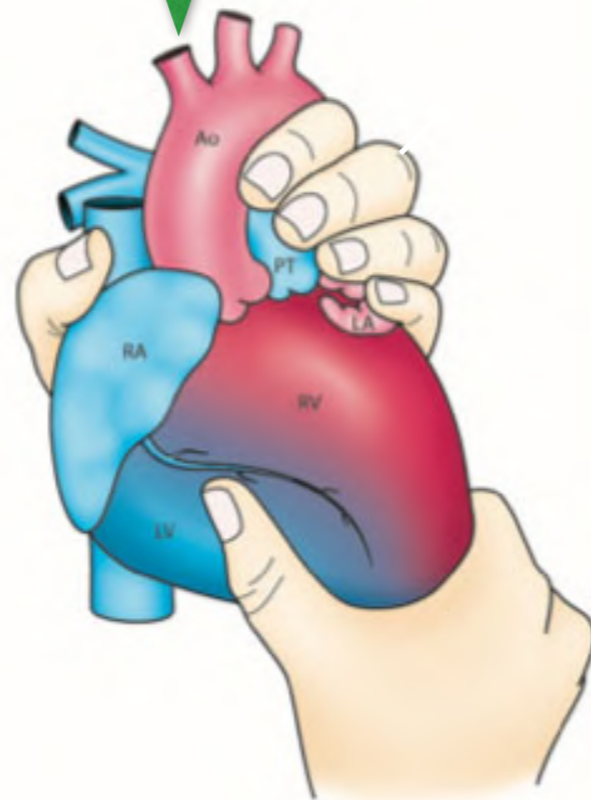
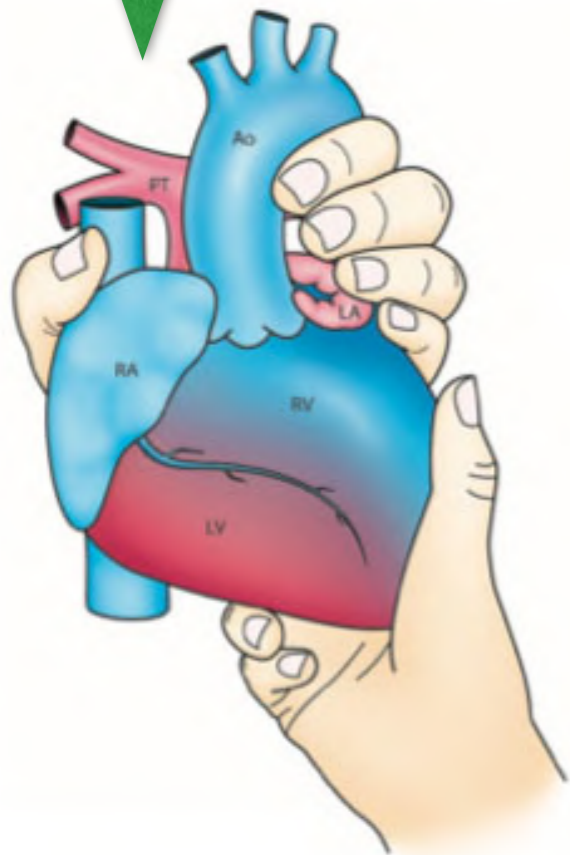
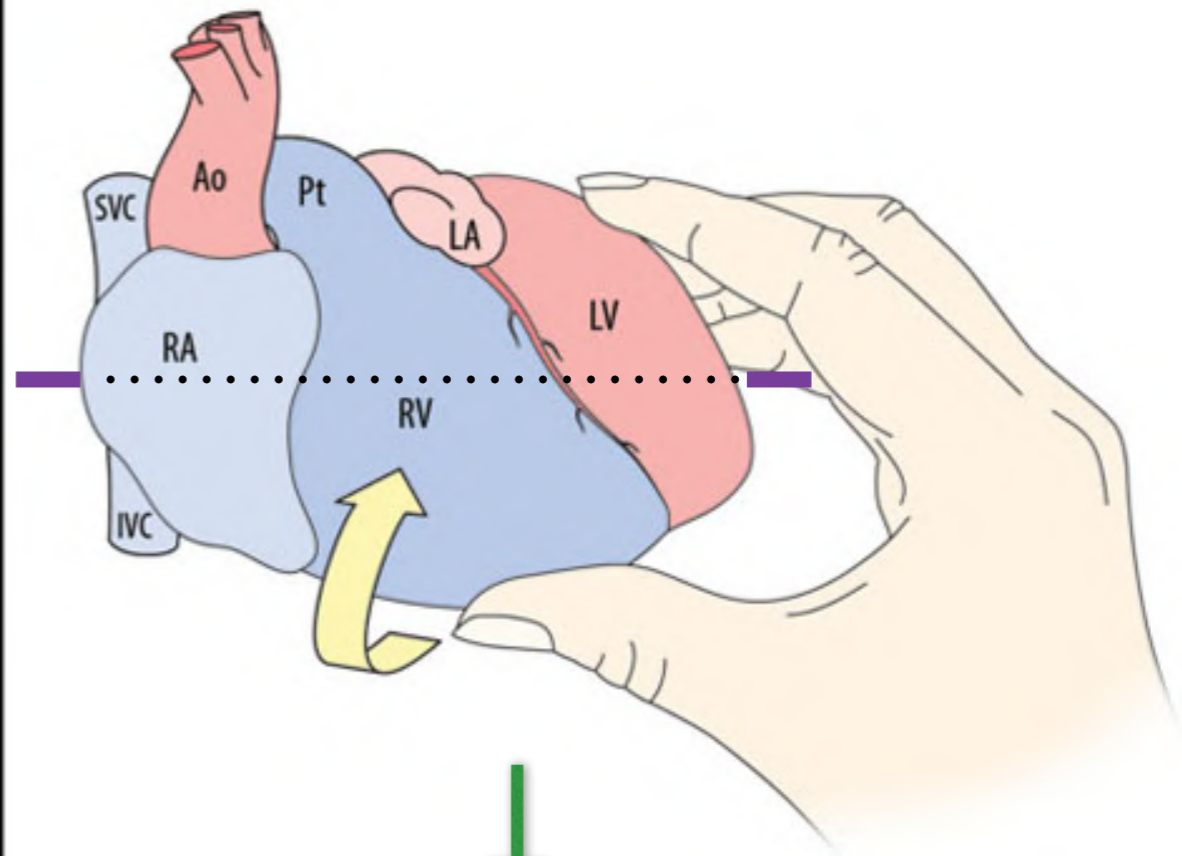
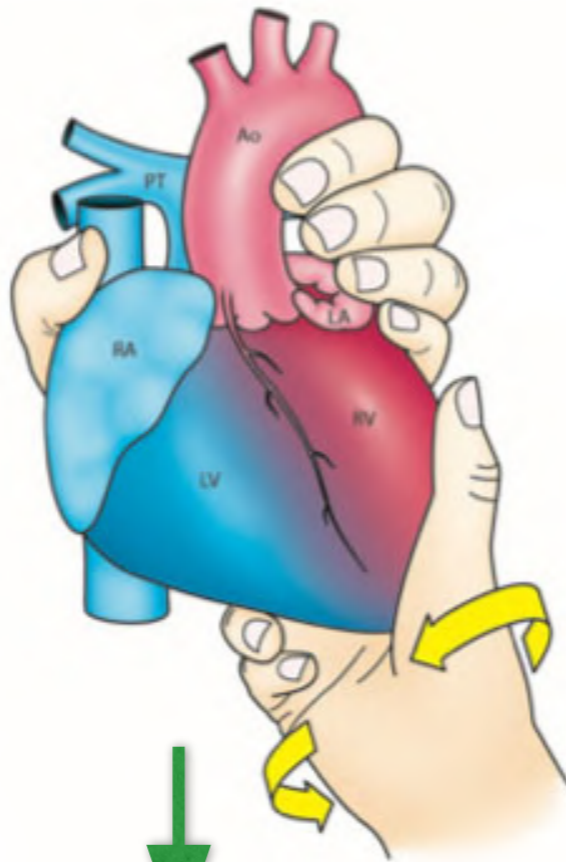
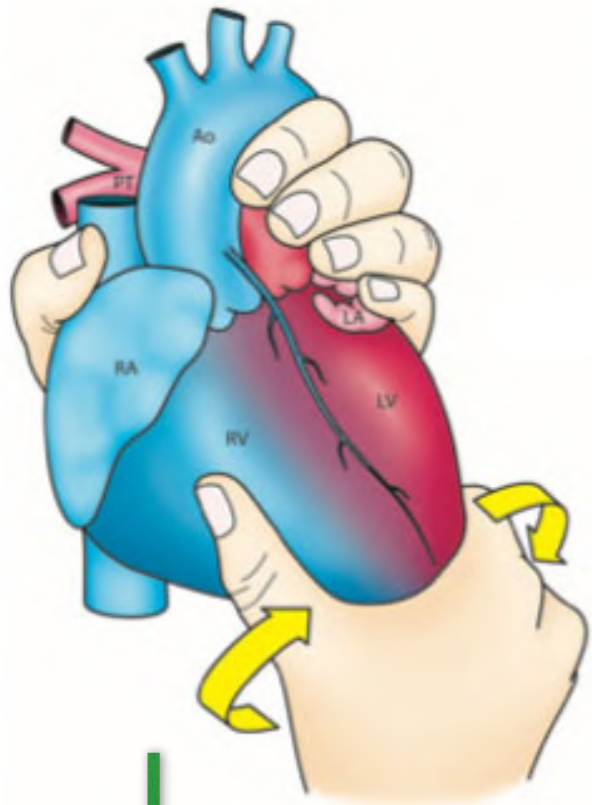
HE 13.5mm/rot
*1/0.6sp

4



What's in topsy-turvy heart?

- Organo-axial rotation (not twisting) around the base-apex axis.
- Four chambers arranged in a coronal plane.
- Elongated head and neck branches of the aorta.
- Elongated and stretched trachea and compression of the left main bronchus.



Twisted heart in complete transposition

Twisted heart in corrected transposition

Topsy-turvy heart in otherwise normally formed heart



TORONTO

Thank you!