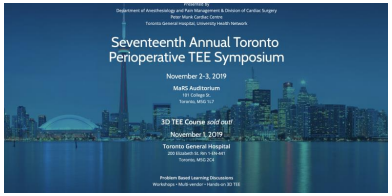

CONGENITAL HEART DISEASE: *NOMENCLATURE*

Erwin Oechslin, MD, FRCPC, FESC
Professor of Medicine, U of T
The Bitove Family Professor of ACHD
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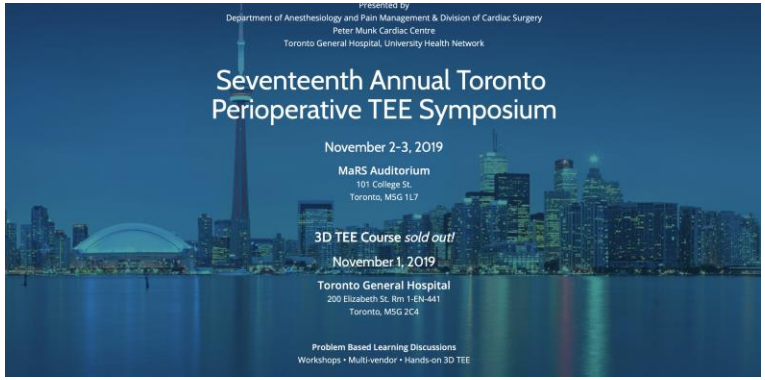
University Health Network / Toronto General Hospital
Toronto, ON, Canada



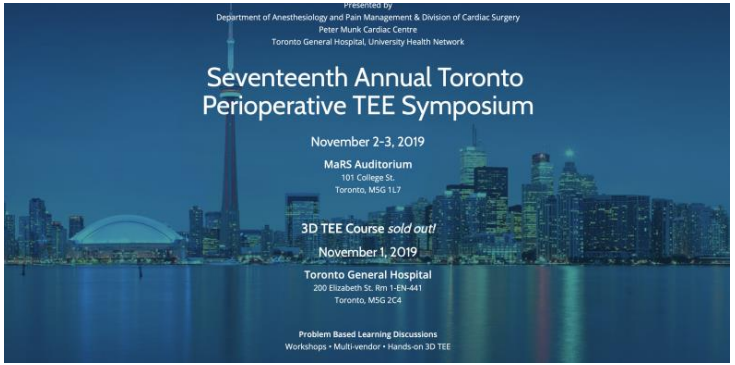
CONGENITAL HEART DISEASE:
NOMENCLATURE
"FEAR NOT – MAKE IT SIMPLE"

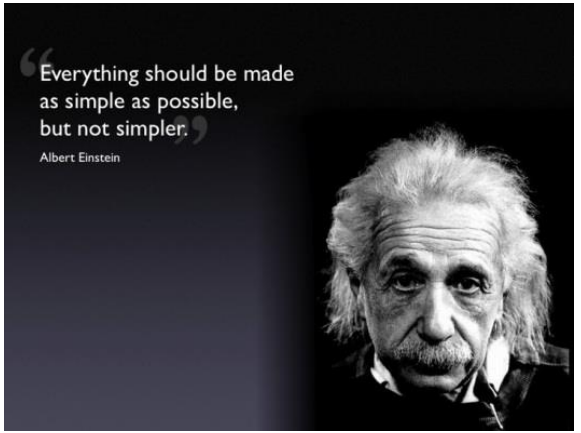
UHN Peter Munk Cardiac Centre

Toronto Congenital Cardiac Centre for Adults

SickKids The Labatt Family Heart Centre

University of Toronto





CONGENITAL HEART DISEASE:
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University of Toronto

DISCLOSURES

- **NOTHING TO DISCLOSE**

OBJECTIVES

- To recognize the i**mportance and understanding of basic terminology** in CHD.
- To formulate the **philosophy of the segmental analysis** to describe cardiac anatomy.
- To identify the **three segments of the heart** and to describe **their connections.**

OUTLINE

- **Introduction**
- **Segmental approach**
  - Atrial arrangement / cardiac position
  - The three segments
  - Connections
- **Looping**
- **Summary**

CHALLENGES

- **Congenital heart defects – very wide spectrum:**
  - Various forms and combinations
  - Modification of the underlying anatomy / pathophysiology by the congenital heart surgeons!
  - Complex anatomy and morphology
- **Terminology / language in CHD!**

CHALLENGES

- **Surgical / interventional procedures**
  - Named after surgeons / physicians
  - Different types / modification of procedures for the same CHD
- **Combination of different CHD**
- **Syndromes**


CHALLENGES AHEAD
Blalock Hanlon procedure
Senning / Mustard procedure
Kawashima procedure
Scimitar syndrome
Waterston shunt
Arterial switch procedure
Yasui procecure
Sterling Edwards procedure
Fontan procedure
and its modifications
Blalock Taussig
Thomas shunt
Shone complex
Eisenmenger syndrome
Potts shunt
Holt-Oram syndrome
Konno procedure
Williams syndrome
Baffes procedure
LEOPARD syndrome
Isomerism
Microdeletion 22q11

BEFORE YOU START……

- **Surgical history**
  - Surgical notes!
- **Morphology / anatomy**
- **Pathophysiology**

BEFORE YOU START……

- Surgical history
  - Surgical notes!
- Morphology / anatomy
- Pathophysiology

To understand the anatomy / connections and surgical procedures

To understand the specific long-term complications of each CHD and procedure

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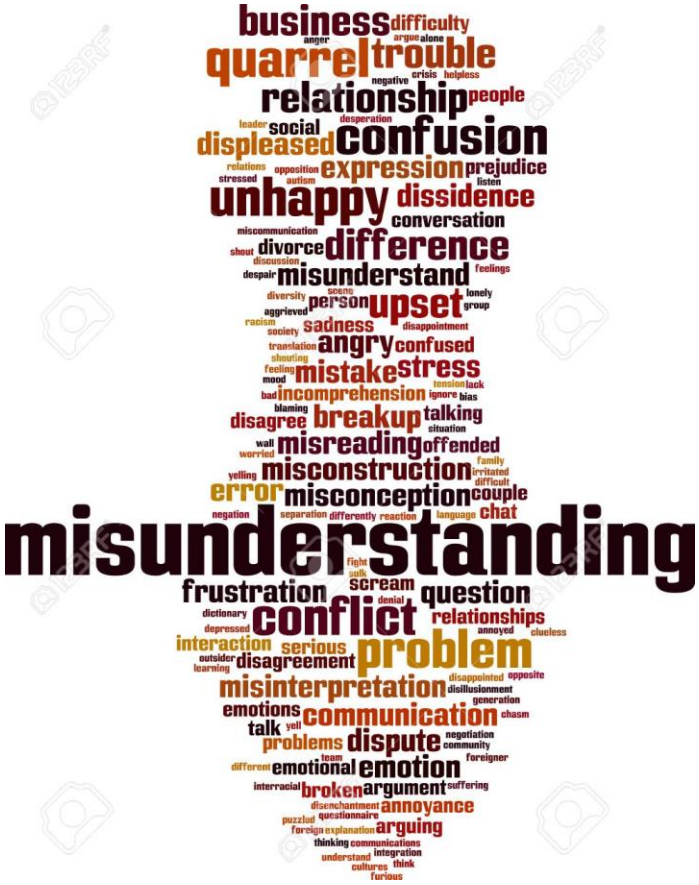
What Are You Looking For?

Sequential Segmental Analysis

Clear language



By Frits Ahlefeldt

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There is a Congenital in the ER …. ?????

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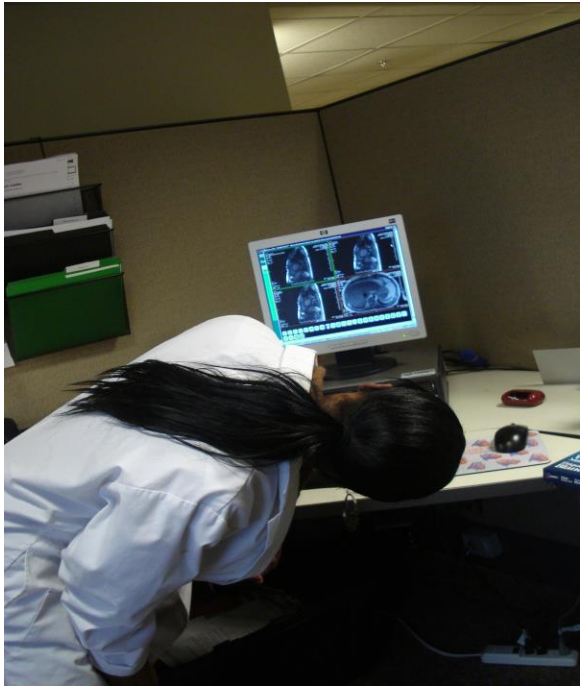
Everybody is confused…..



those who work with them every day

Philosophy of Segmental Analysis is Founded on MORPHOLOGY

- **Chambers** are recognized according to their **morphology**
  - Each chamber has **intrinsic features**
- The chambers are not in their anticipated location!

Segmental Approach

Atrial Arrangement / Position

↓

Identify The Three Segments:

- (Atria) – Ventricles – Great Arteries

↓

Define the Connections

- Atrio-ventricular / ventriculo-arterial

Cardiac Position

Position of the heart within the chest

Cardiac Position

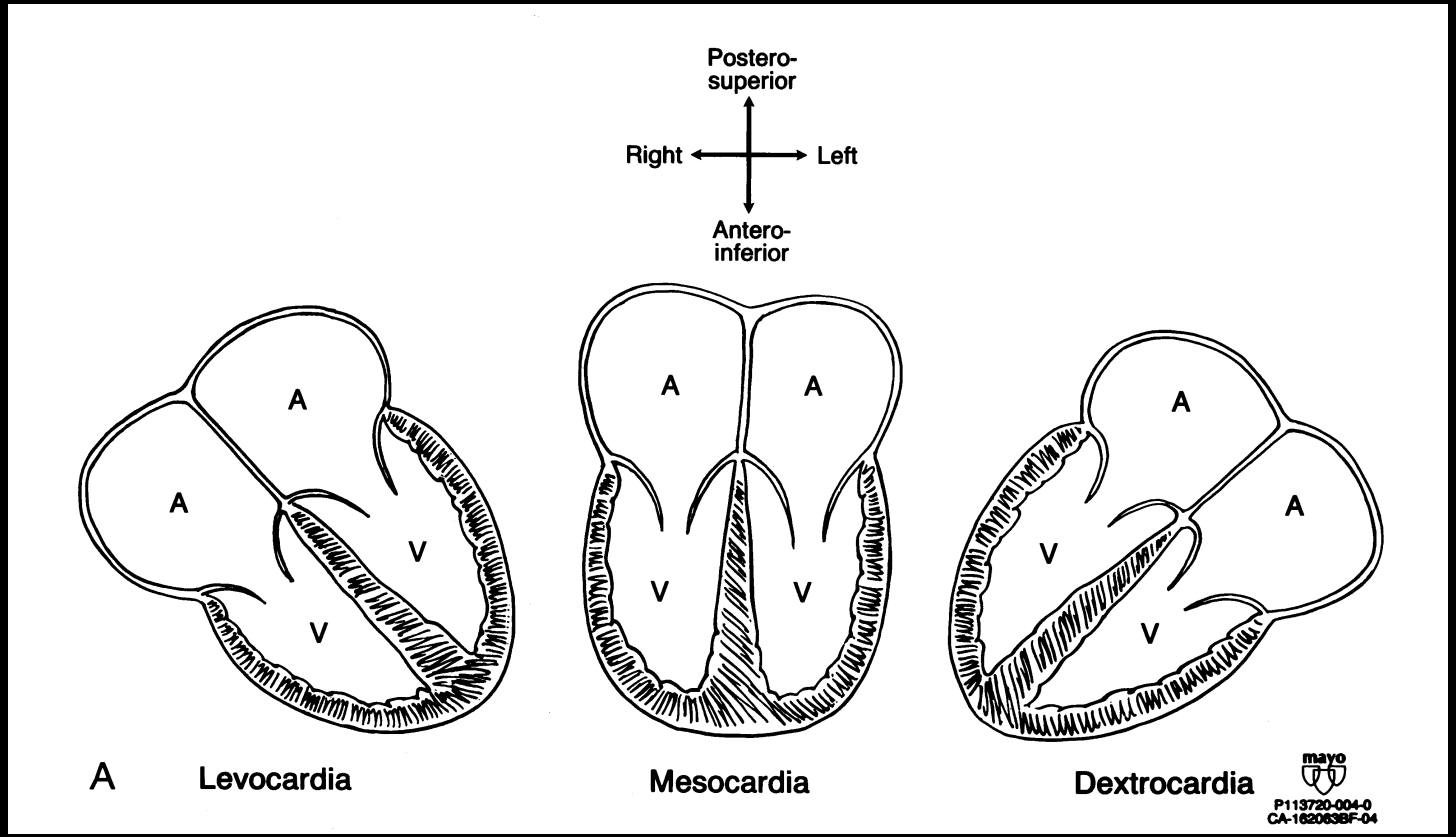
- Levoposition
- Mesoposition
- Dextroposition

Dependent from many factors

- Cardiac malformation
- Medistinal/thoracic structures

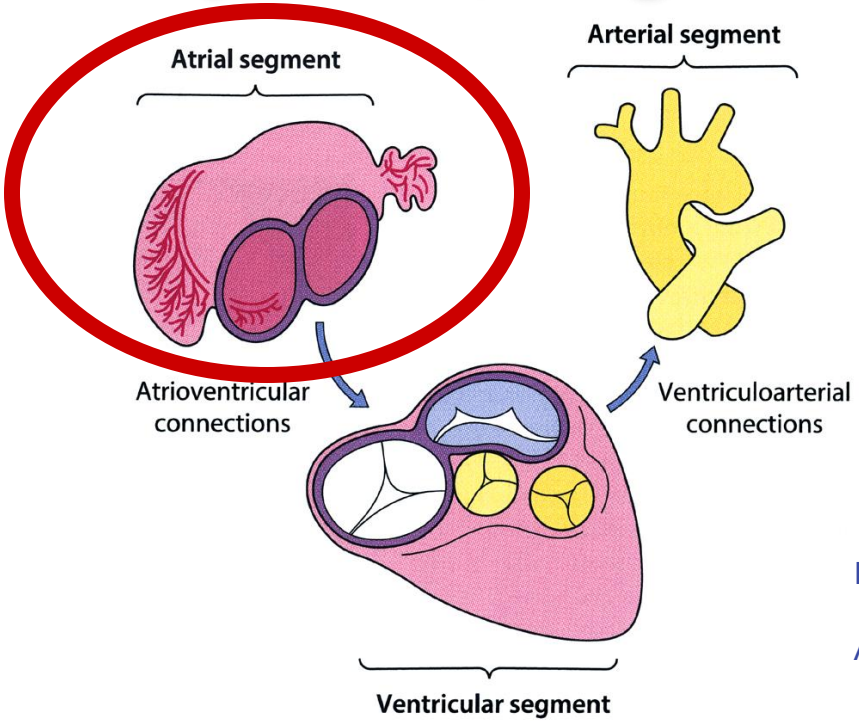
Cardiac Orientation

- Levocardia
- Mesocardia
- Dextrocardia



Edwards WD. In: Heart Disease in Infants, Children, and Adolescents.
Moss and Adams (eds.), 1995, page 108

The *UNKOWN* Patient: Sequential, Segmental Approach



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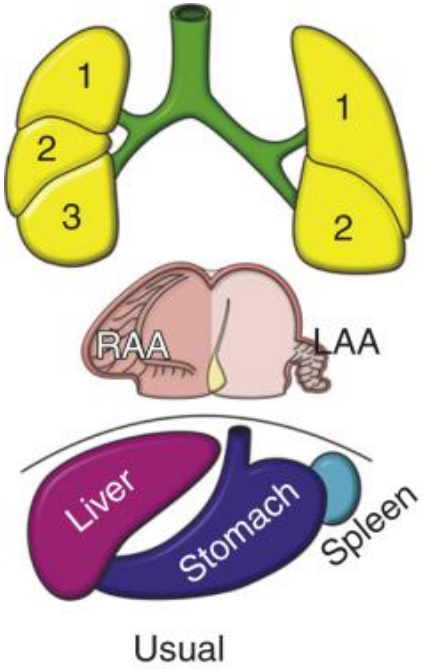
S I T U S - SIDEDNESS

- ***Cardiac*** Situs
- ***Pulmonary*** Situs
- ***Abdominal*** Situs

Atrial Arrangement (Cardiac Situs)

- ***Position*** **of the morphologic *RIGHT ATRIUM*, <u>independent</u> from:**
  - Cardiac position
  - Cardiac orientation
  - Position of the ventricles / great arteries

Atrial Arrangement

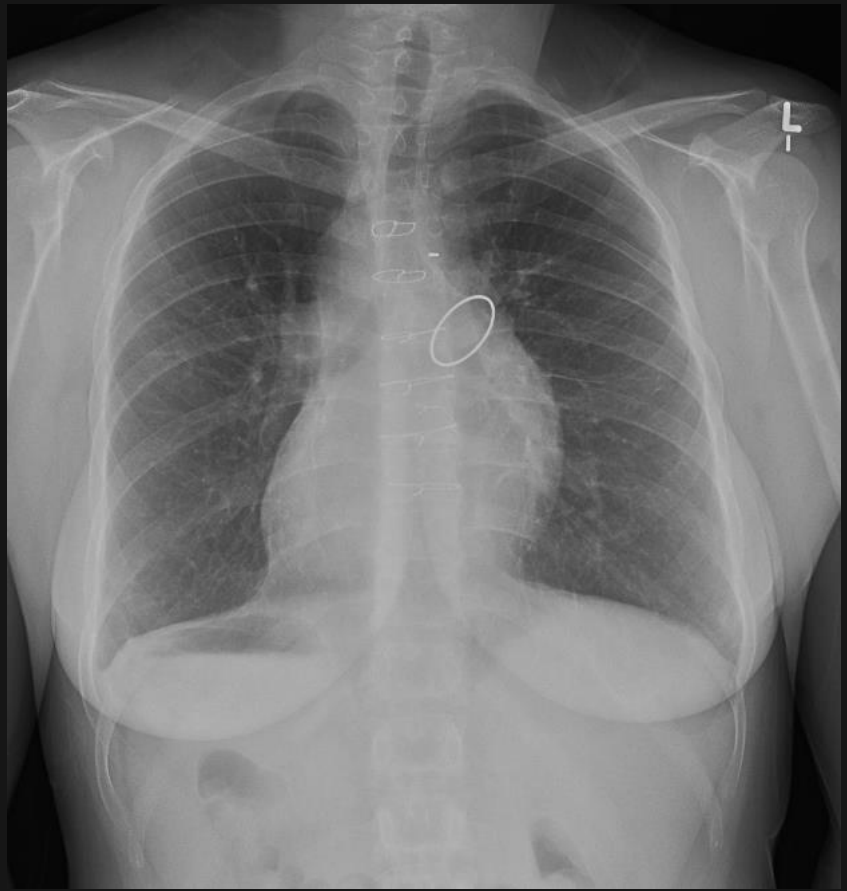


Usual

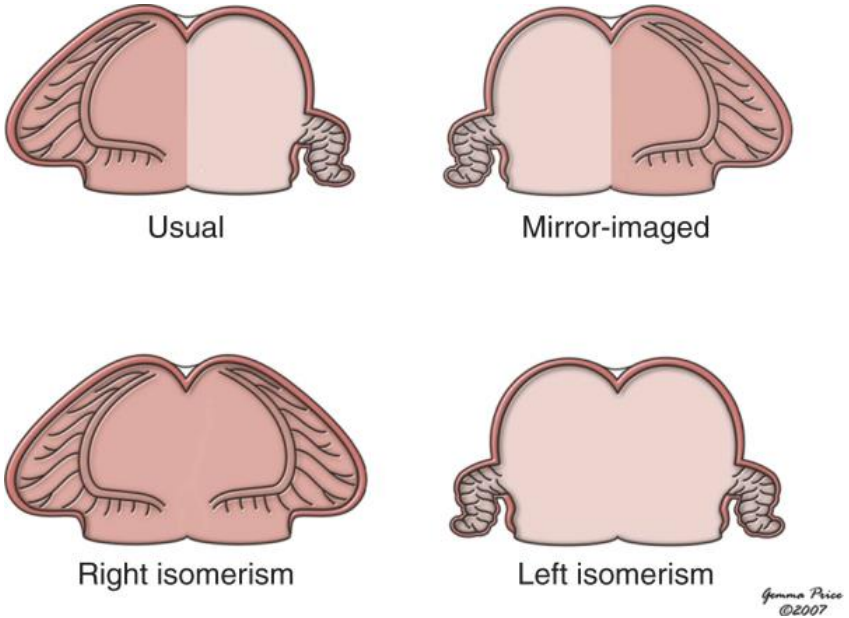
Harmony between arrangement of the atrial appendages and thoraco-abdominal organs

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16

Situs Inversus, Dextrocardia



Atrial Arrangement



- Situs **solitus**:
  - Morphological RA is on the right of the morphological LA
- Situs **inversus**:
  - Morphological RA is on the left of the morphological LA
- Situs **ambiguus**:
  - Indeterminate sidedness in the setting of isomerism

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16

Atrial Arrangement

- Situs **solitus**:
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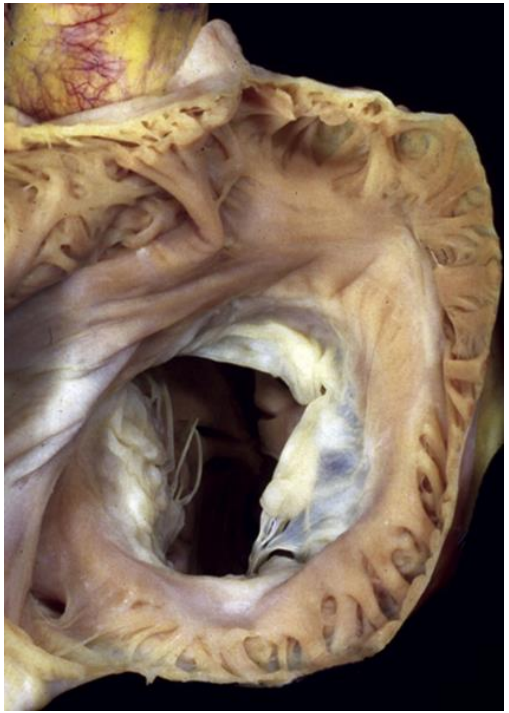
Appendages:
the landmark for morphologic
RIGHTNESS and ***LEFTNESS***

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16

RIGHT ATRIAL APPENDAGE



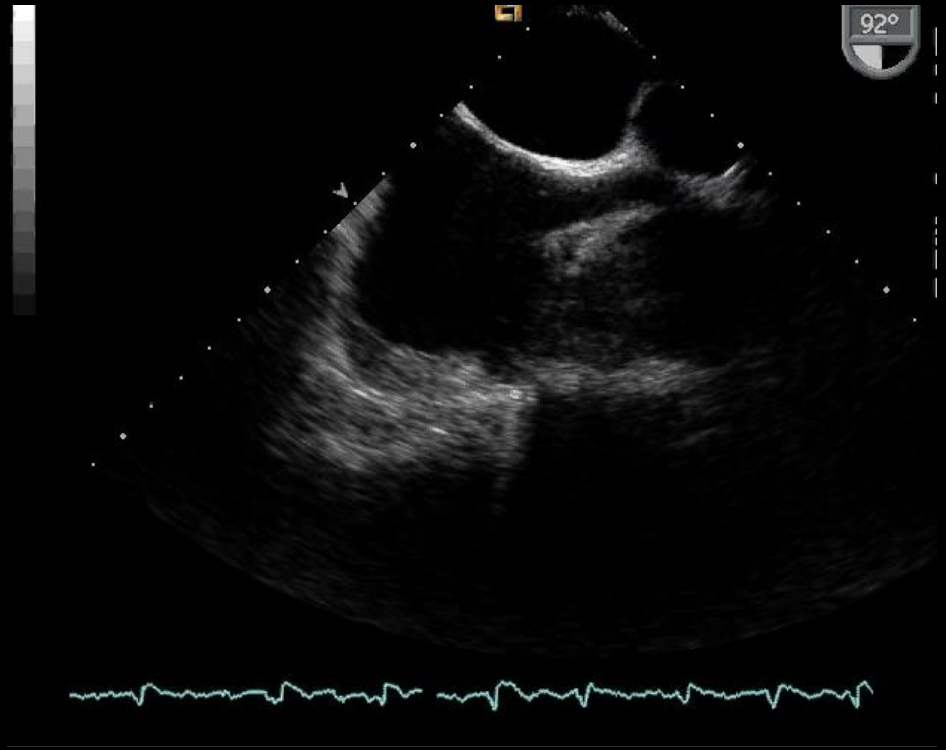
- Triangular
- Broad base
- Pectinate muscles within the appendage extend all round the vestibule of the tricuspid valve!

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16

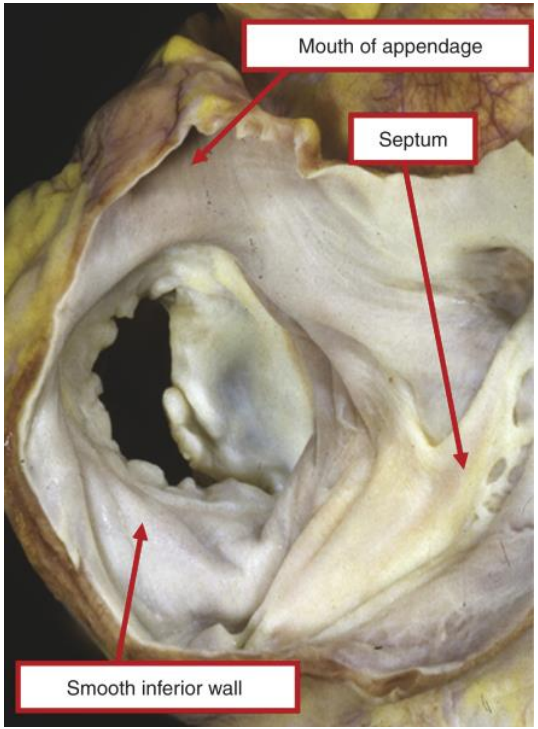
Right Atrial Appendage

Right Atrium

- Appendage
  - Triangular
  - Broad base
- Terminal crest



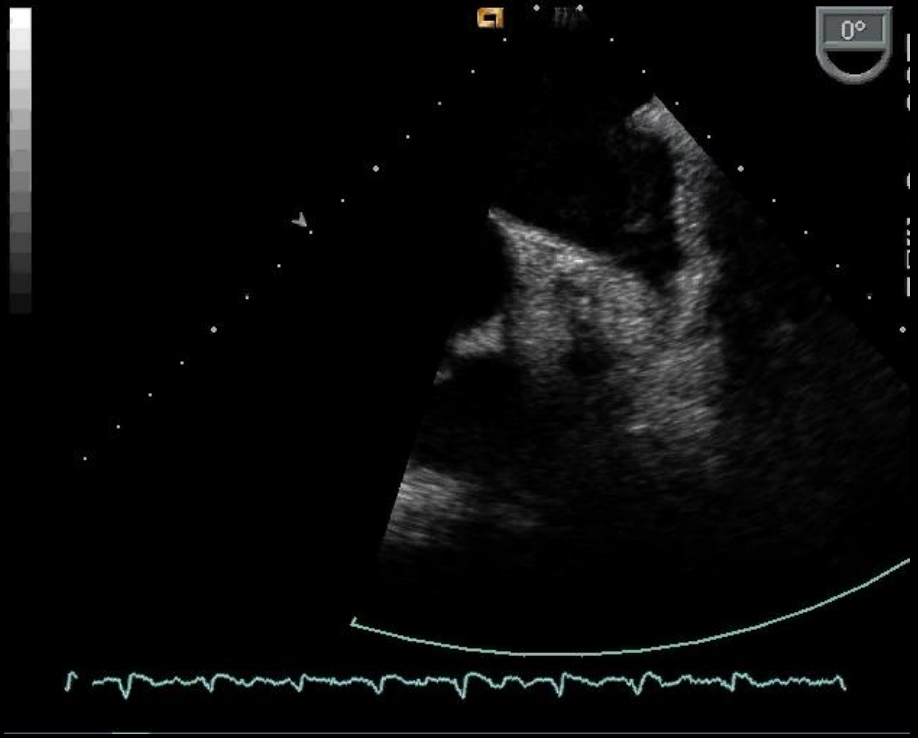
LEFT ATRIUM



- **Left Atrium – Appendage**
  - Hook-shaped
  - Narrow entrance
  - Pectinate muscles are confined within the appendage
- **No terminal crest**

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16

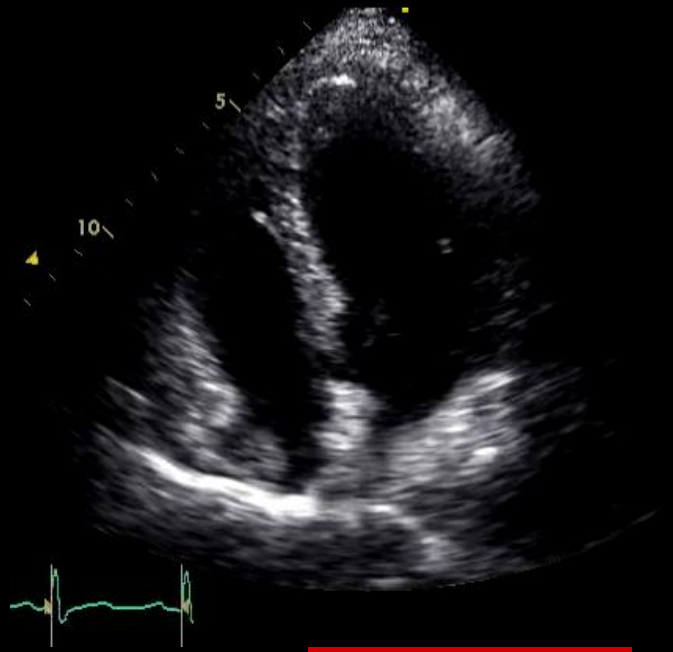
Left Atrial Appendage



Left Atrium

- Appendage
  - Hook-shaped
  - Narrow entrance
- No terminal crest

Anatomic Landmarks of the Right / Left Atrium



Terminal Crest

LA Appendage

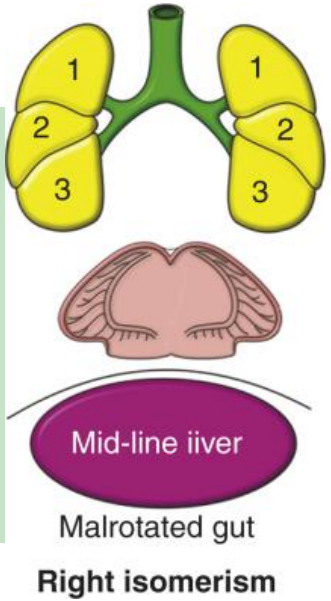
ISOMERISM

- Paired, **mirror-image** sets of normally single, **non-identical** organ systems
  - Atria
  - Lungs
  - Viscera

Isomerism

Paired morphologically ***right*** structures:

- Bilateral right bronchi
- Bilateral morphologic right atria
- Asplenia / transverse liver
- Other malformations



Right isomerism

Isomeric lungs and atrial appendages abdominal organs are jumbled up

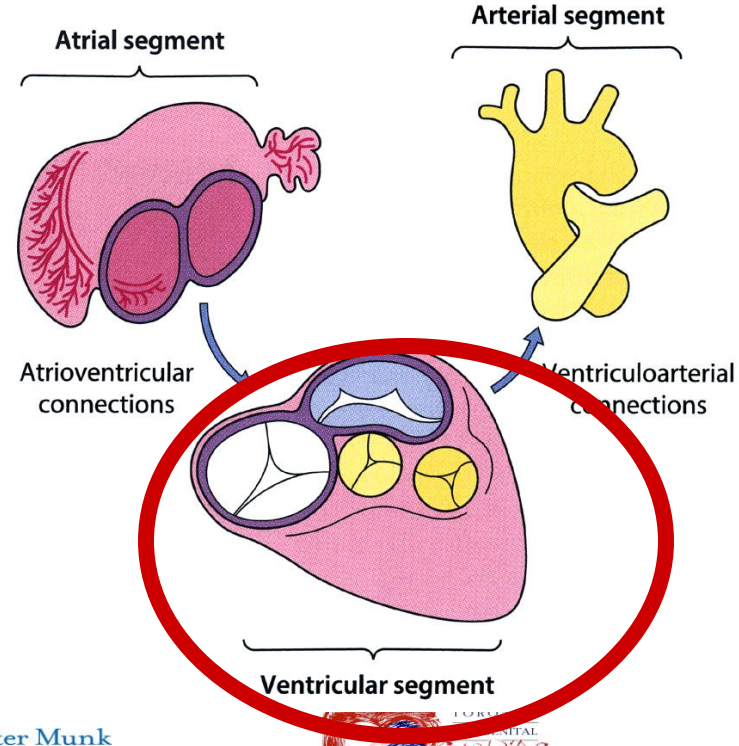
Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16

Left Isomerism

Anatomic Features

Anatomic Feature	Right Atrium	Left Atrium
Veins	**IVC: constant** SVC/CS: variable	Pulmonary veins: variable
Appendage	Broad, triangular	Narrow, finger-like
Musculi pectinati	Many	Few
Terminal Crest	Present	Absent
Septal Surface	Septum secundum	Septum primum
Conduction system	Sinoatrial node	

The *UNKOWN* Patient: Sequential, Segmental Approach



S.Y.Ho,Cardiac Morphology and Nomenclature, p. 7-18
In: Gatzoulis, Webb, Daubeny (eds.) Diagnosis and Management of Adult Congenital Heart Disease 2003

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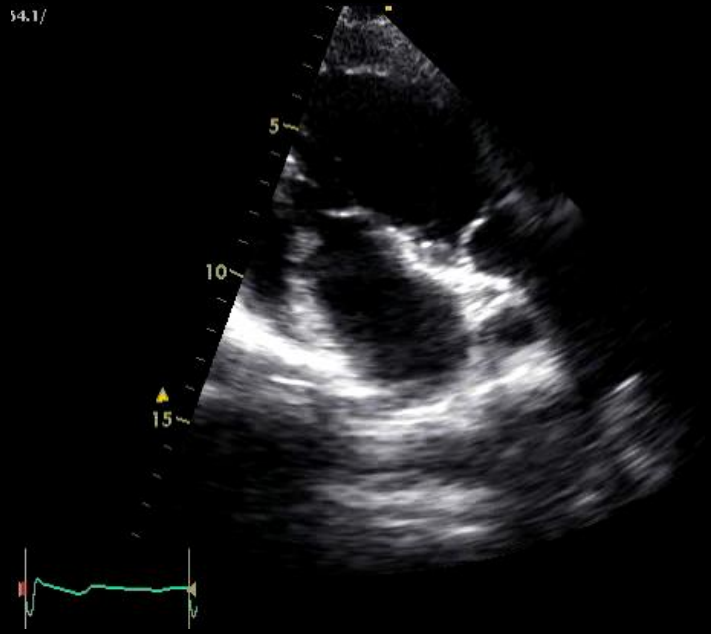
Left Ventricle

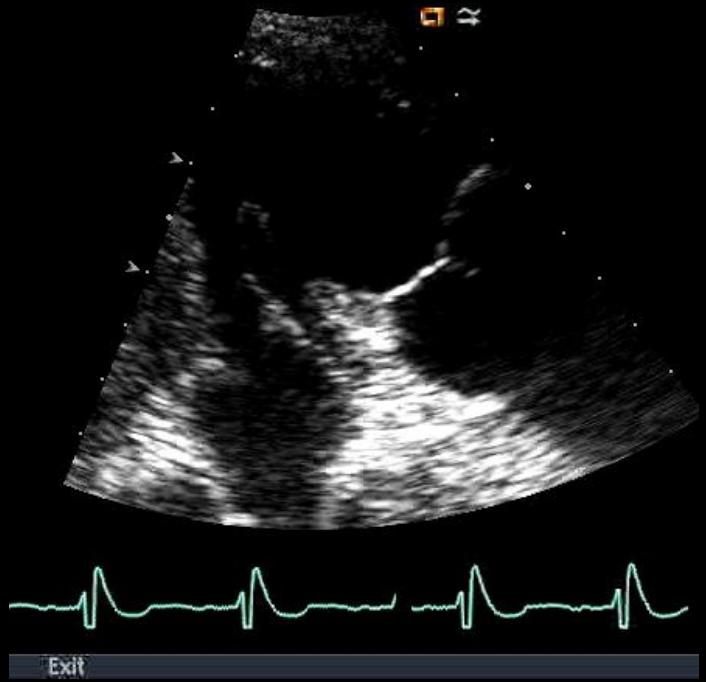
Fibrous Continuity

Right Ventricle

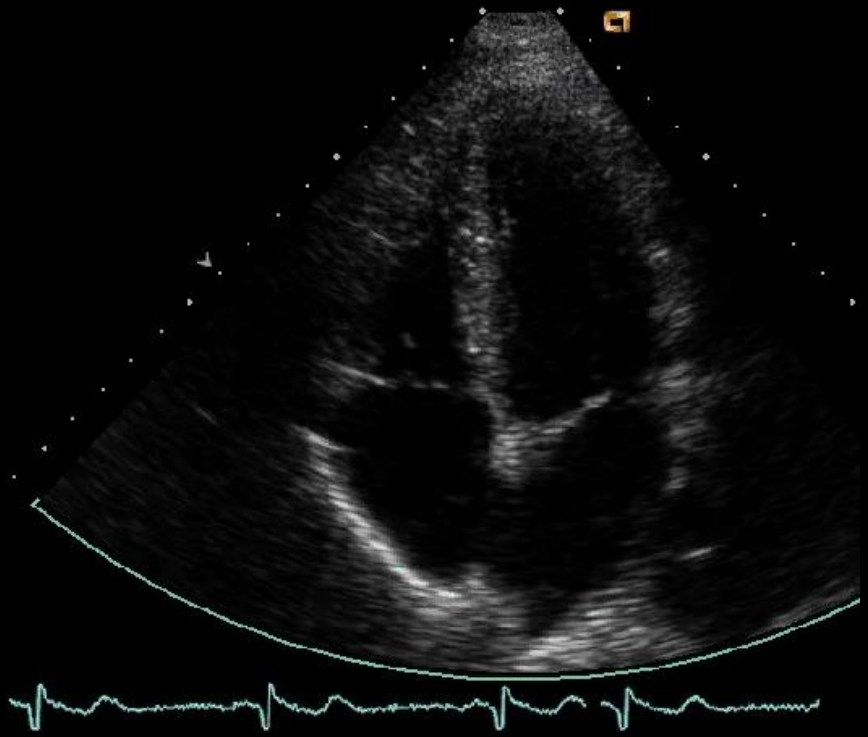


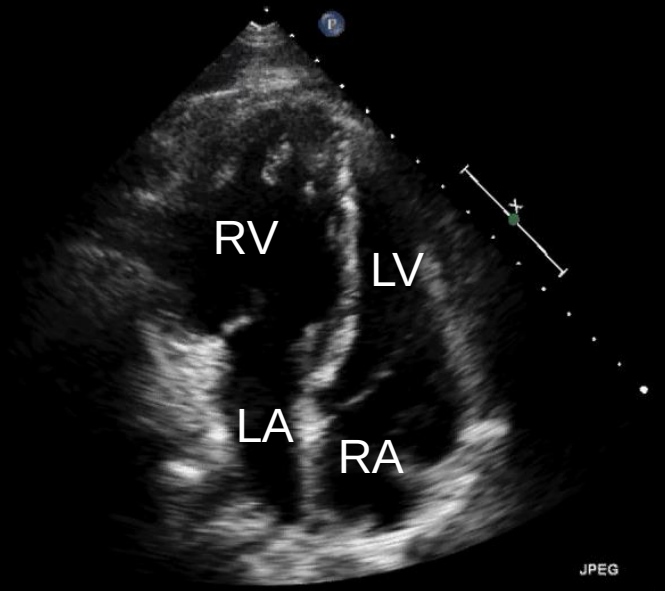
Ventricular Crest

Ventricular Crest
Landmark for RV Morphology



- Muscular crest in the RVOT intervening between tricuspid and semilunar valve
- Demarcation the junction between the outlet septum and the pulmonary infundibulum





Normal heart

Situs inversus
ccTGA

Apical attachment

of the septal tricuspid valve leaflet

VENTRICULAR SEGMENT

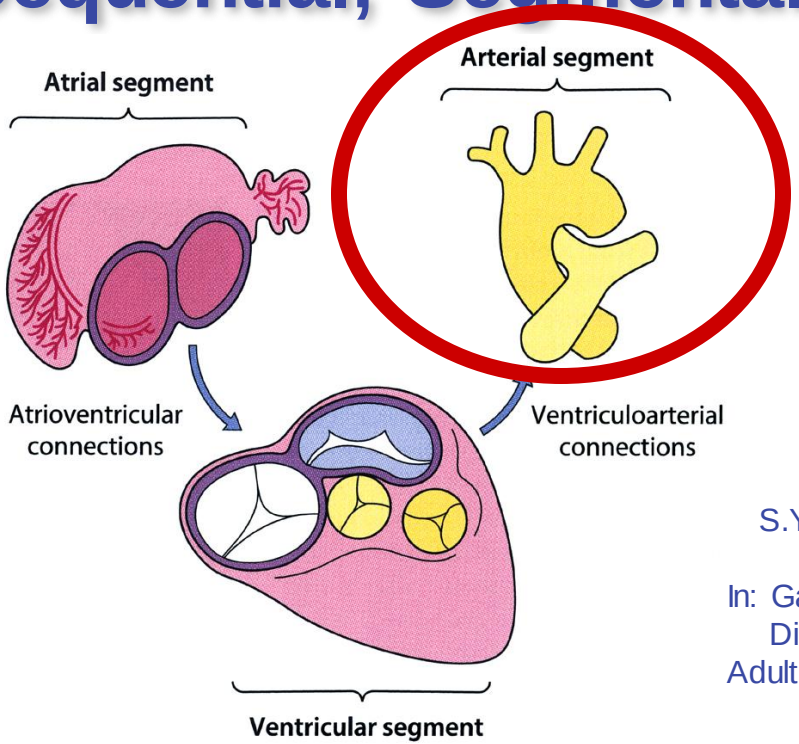
Tricuspid Valve / RV

- Apical attachment of the septal leaflet
- Ventricular crest
- Extensive septal leaflet tethering to the septum
- Moderator band
- Course trabeculations

Mitral Valve / LV

- No tendinous chords tethering to the septum
- Fibrous continuity to the semilunar valve

The *UNKOWN* Patient: Sequential, Segmental Approach



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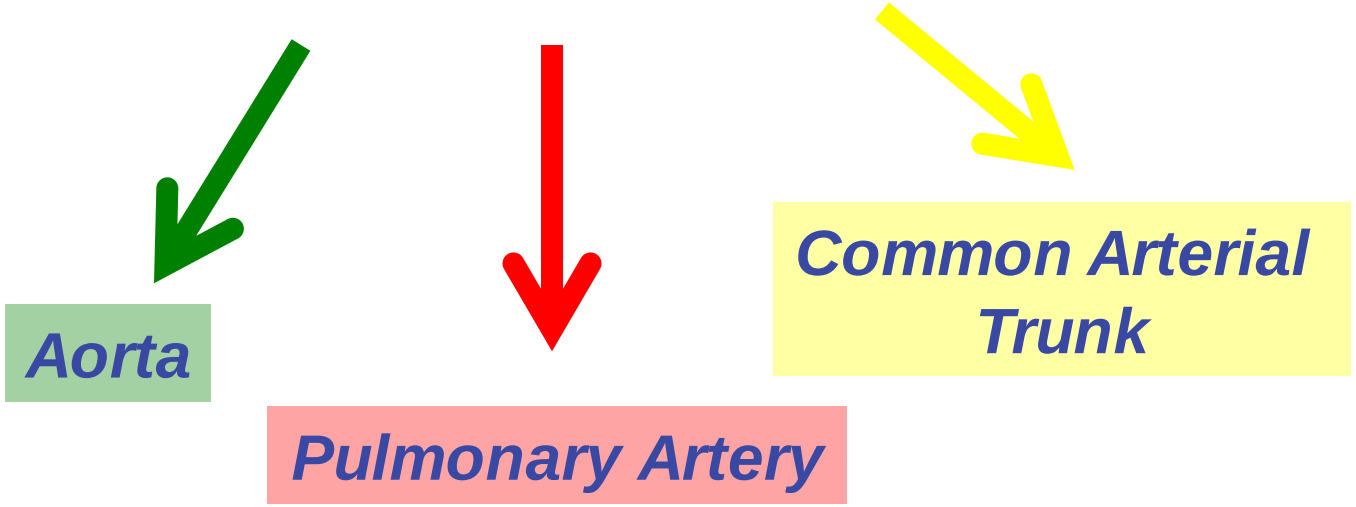
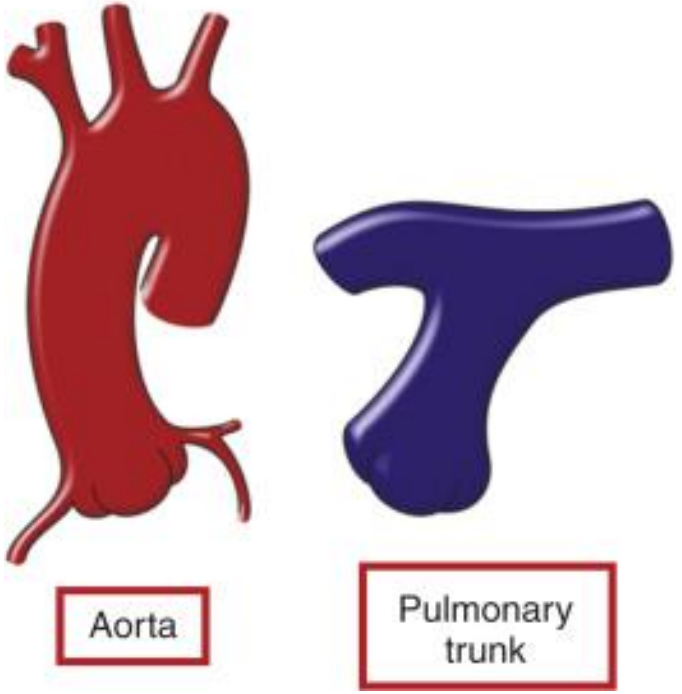
Diagnosis and Management of Adult Congenital Heart Disease 2003

The ARTERIAL Segment



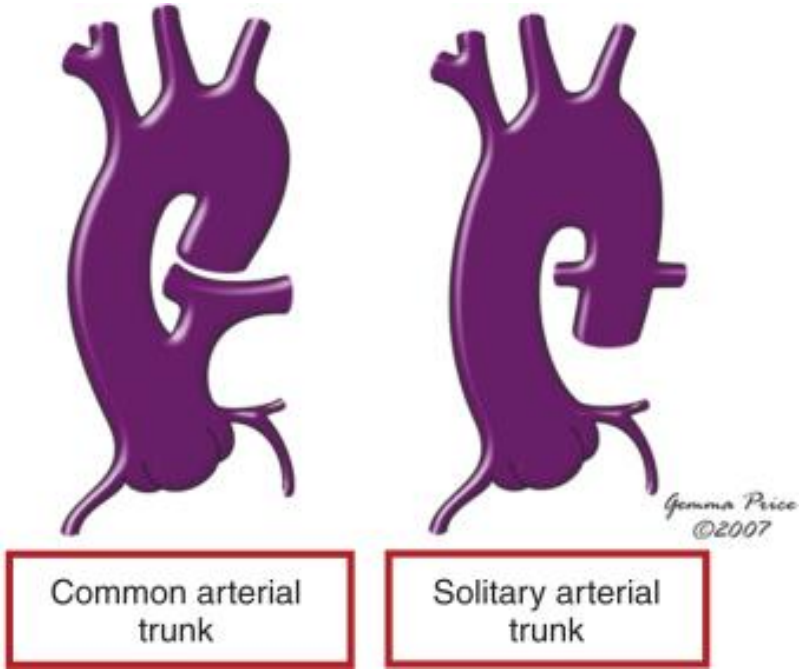
The ARTERIAL Segment

- **Aorta**
  - Coronary arteries
  - Branches to the head
- **Pulmonary arteries**
  - Bifurcation to the ***left*** and ***right*** lung



Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16

The ARTERIAL Segment



- **Common arterial valve**
- **Blood supply to:**
  - Coronary arteries
  - Pulmonary arteries
  - Systemic arteries

Anderson RH, Paediatric Cardiology, 3rd edition, 2010; pages 3-16

Segmental Approach

Atrial Arrangement / Position

↓

Identify The Three Segments:

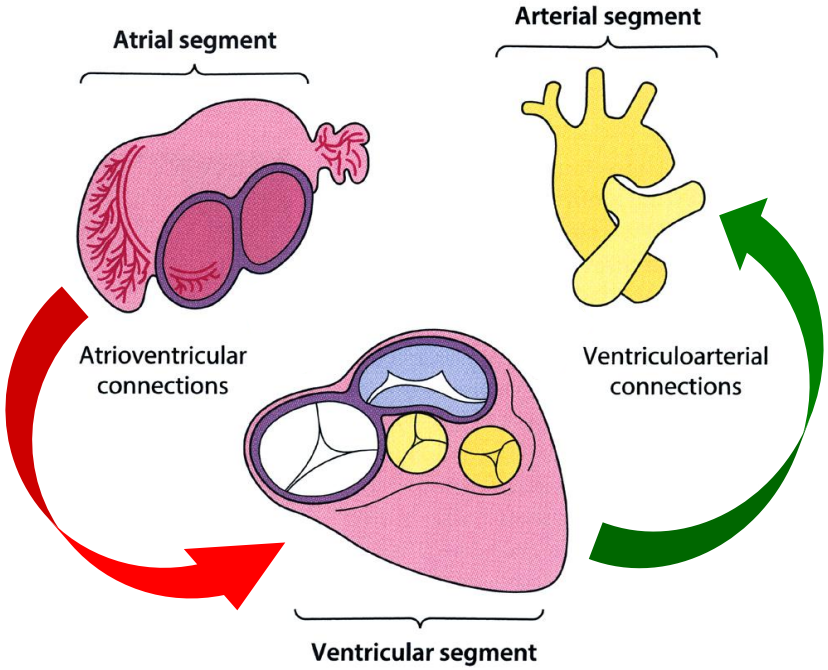
- (Atria) – Ventricles – Great Arteries

↓

Define the Connections

- Atrio-ventricular / ventriculo-arterial

The *UNKNOWN* Patient: Sequential, Segmental Approach



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CONNECTION *vs* DRAINAGE

CONNECTION

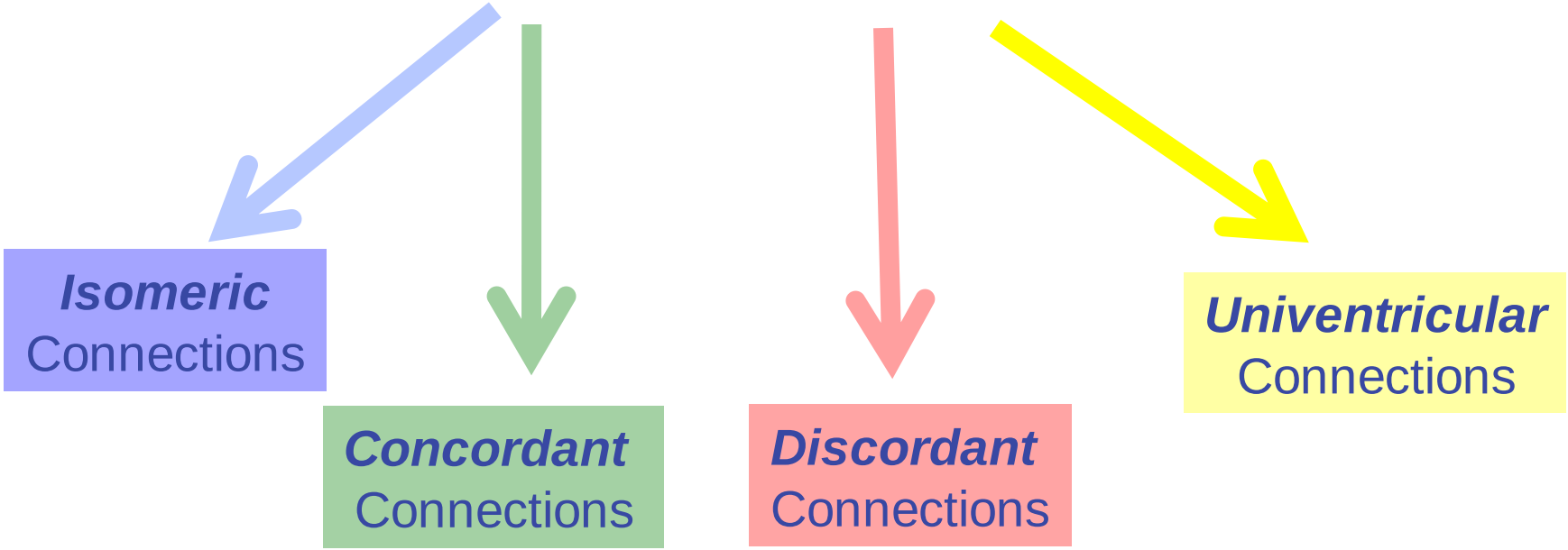
- *Anatomic* term
- Link between two structures
  - Veno-atrial
  - Atrio-ventricular
  - Ventriculo-arterial

DRAINAGE

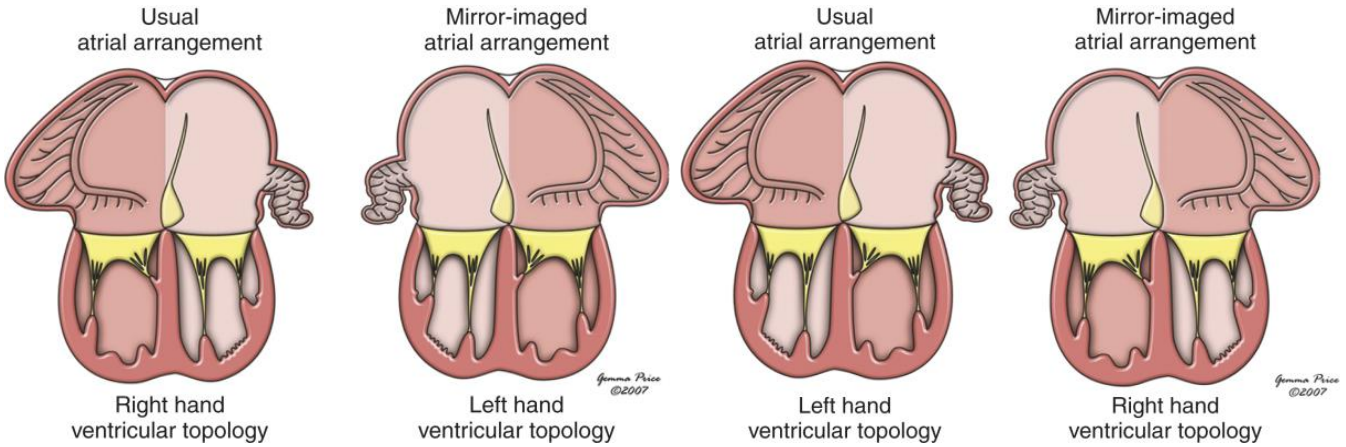
- *Hemodynamic* term
- Blood flow direction

Left Isomerism

CONNECTIONS



AV CONNECTIONS



Concordant Connections

Discordant Connections

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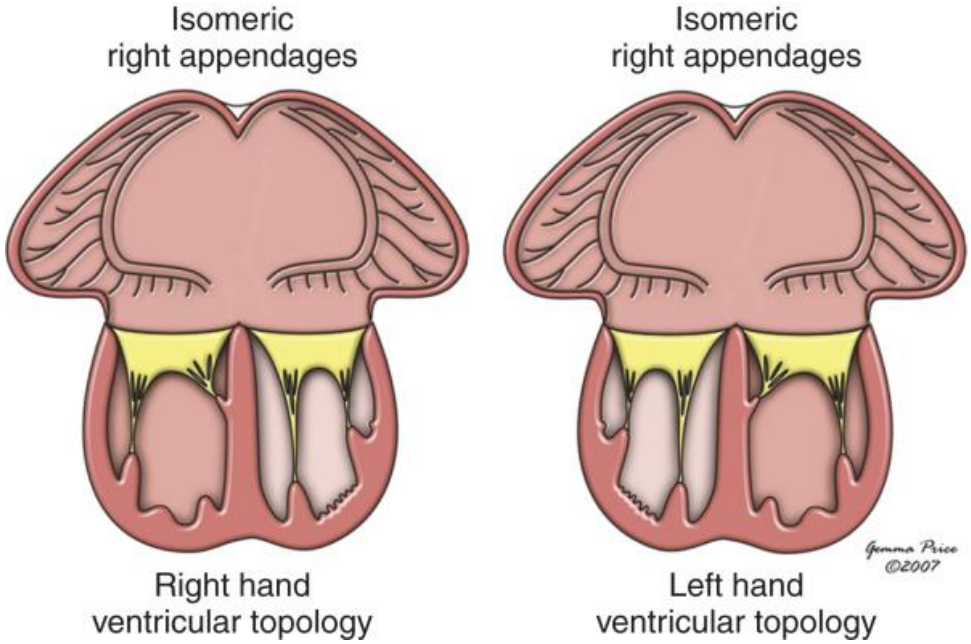
Anderson RH Paediatric Cardiology, 3rd edition, 2010; pages 3-16

ISOMERIC CONNECTION



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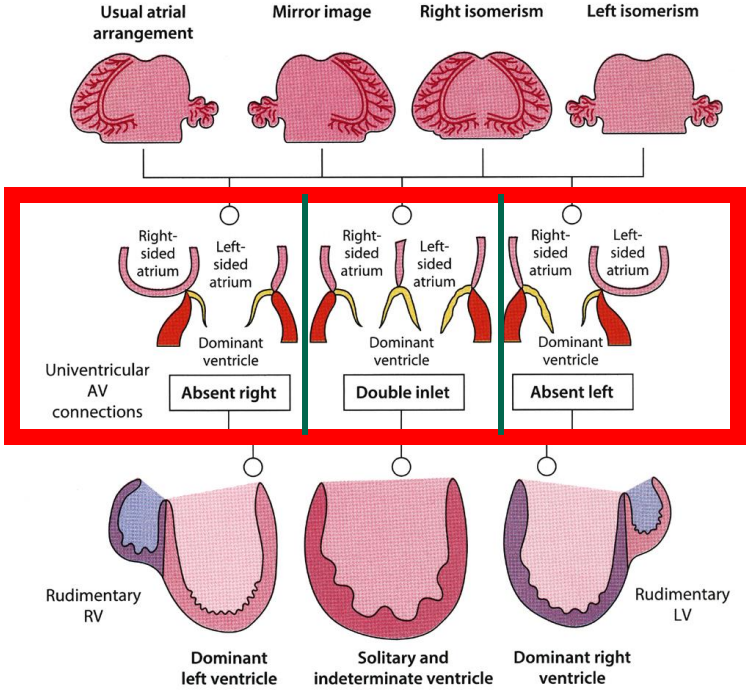
UNIVENTRICULAR Connections
THREE TYPES



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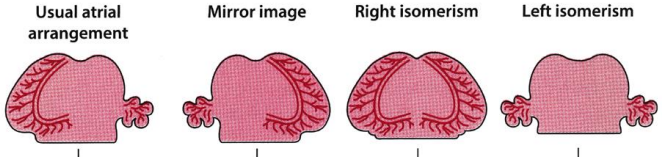

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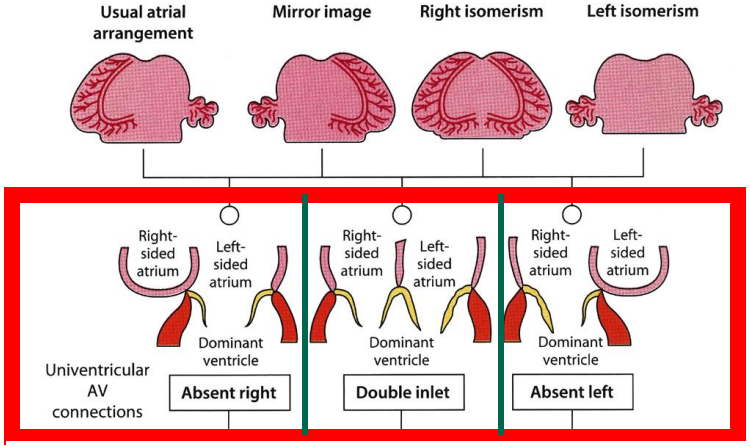

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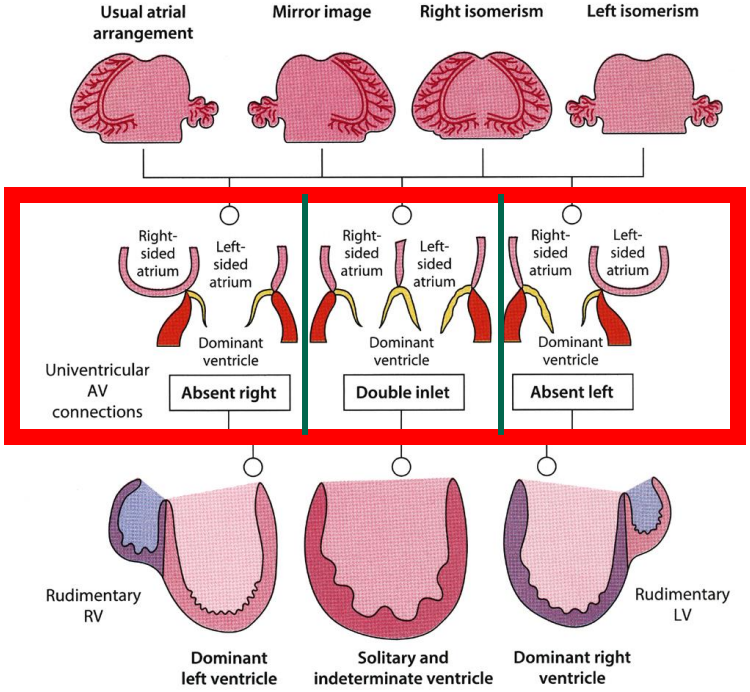

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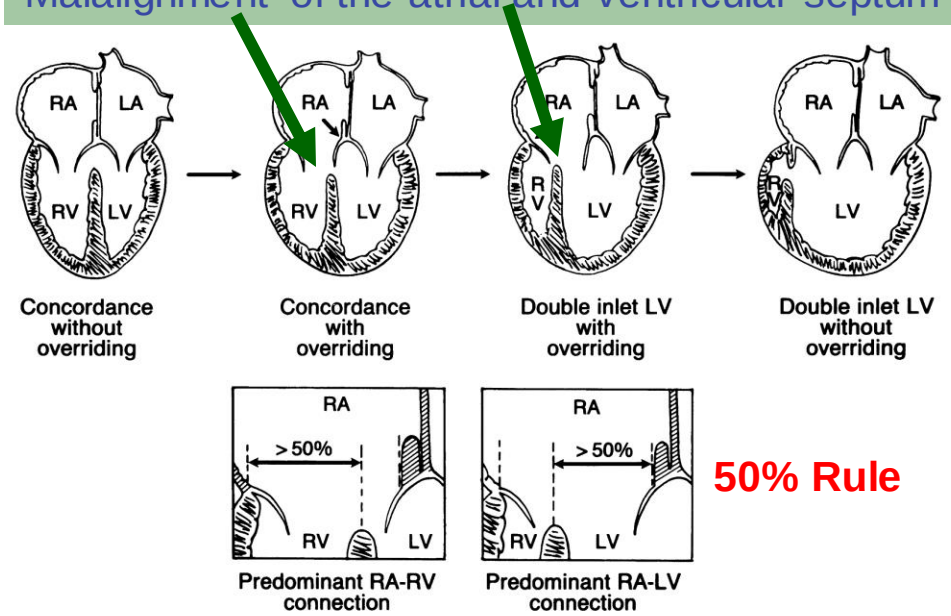
Double Inlet Left Ventricle

Malalignment of the atrial and ventricular septum



50% Rule

Edwards WD. In: Heart Disease in Infants, Children, and Adolescents. Moss and Adams (eds.), 1995, page 125

Double Inlet Left Ventricle

OVERRIDNG

VS

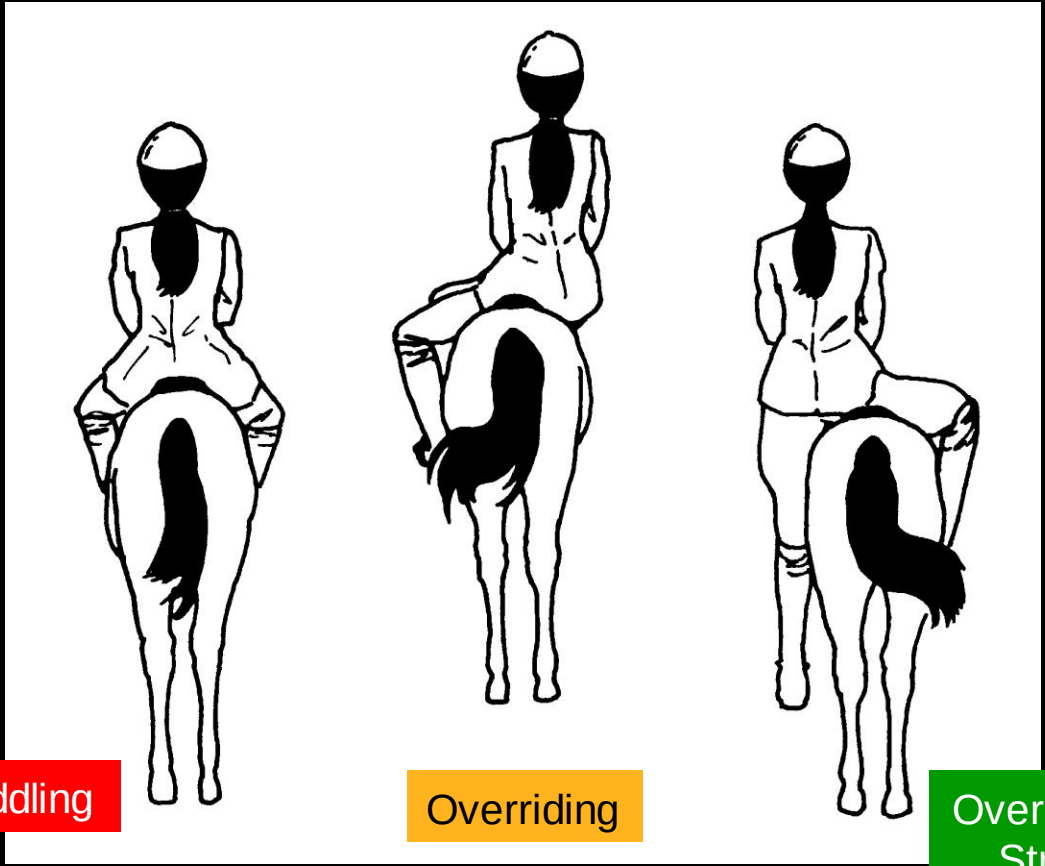
STRADDLING

STRADDLING AV-valve

- ***Anomalous insertion*** **of tendinous cords of papillary muscles into the** ***contralateral ventricle (VSD!!!)***

OVERRIDING

- ***Malalignment*** **of the annulus of one AV-valve or semilunar valve relative to the ventricular septum**

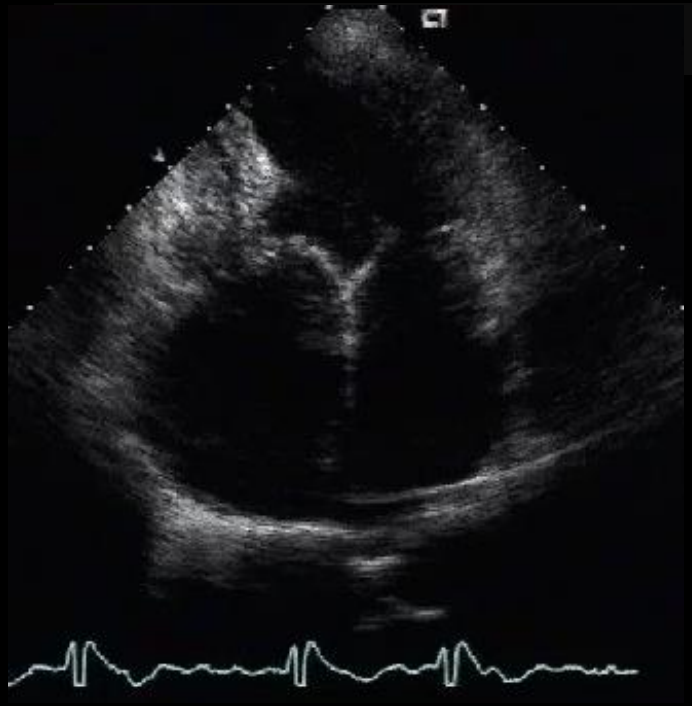


Straddling

Overriding

Overriding and Straddling

Freedom, RM, Mawson JB, Yoo SJ, Benson LN (eds.)
In: Textbook of Angiocardiography, 1997, page 110



DILV

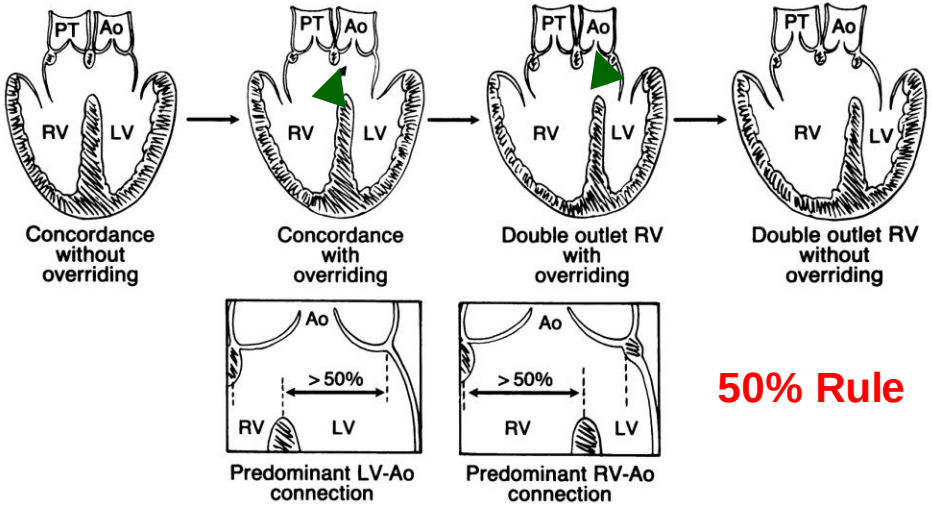
Left Isomerism
AVSD

DOUBLE OUTLET RIGHT VENTRICLE

- **Both great arteries arise predominantly from the one ventricle**
  - DORV

DOUBLE OUTLET RIGHT VENTRICLE

Malalignment of the outlet septum relative to the remainder of the interventricular septum

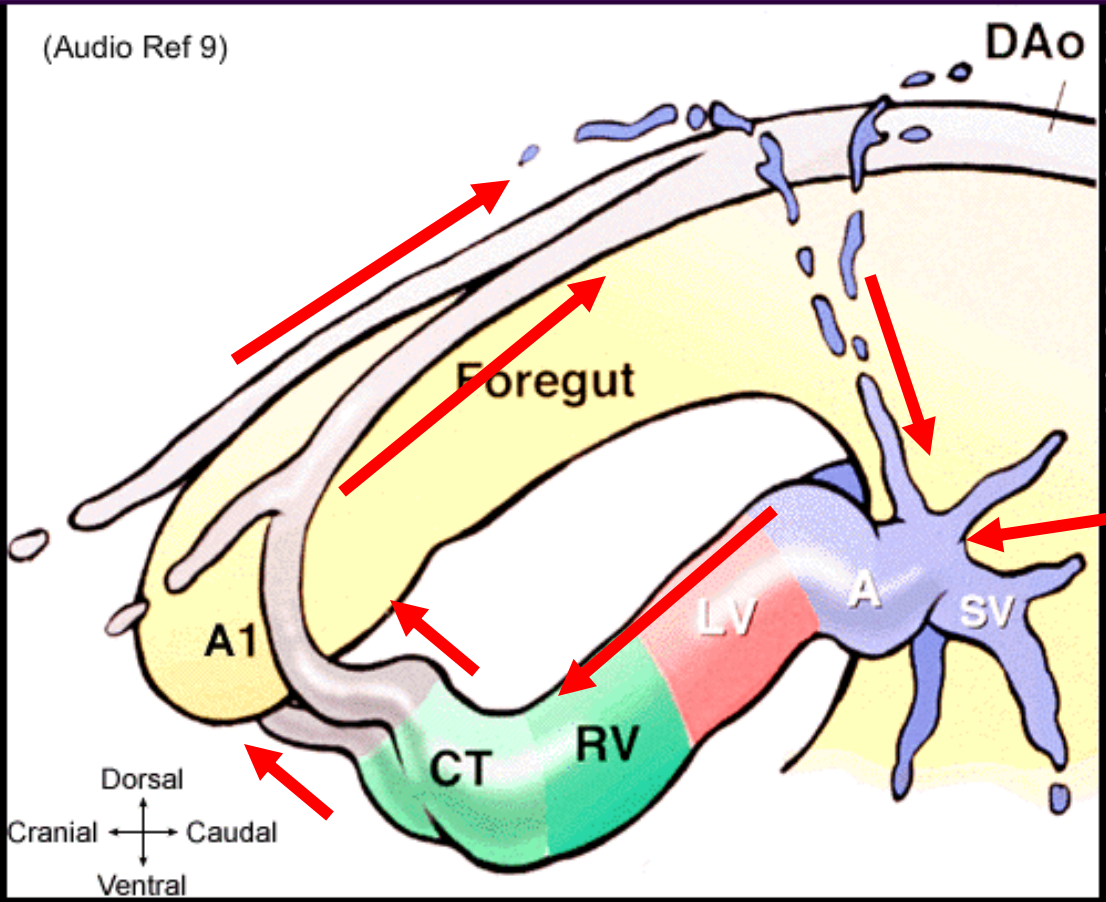


50% Rule

Edwards WD. In: Heart Disease in Infants, Children, and Adolescents. Moss and Adams (eds.), 1995, page 126

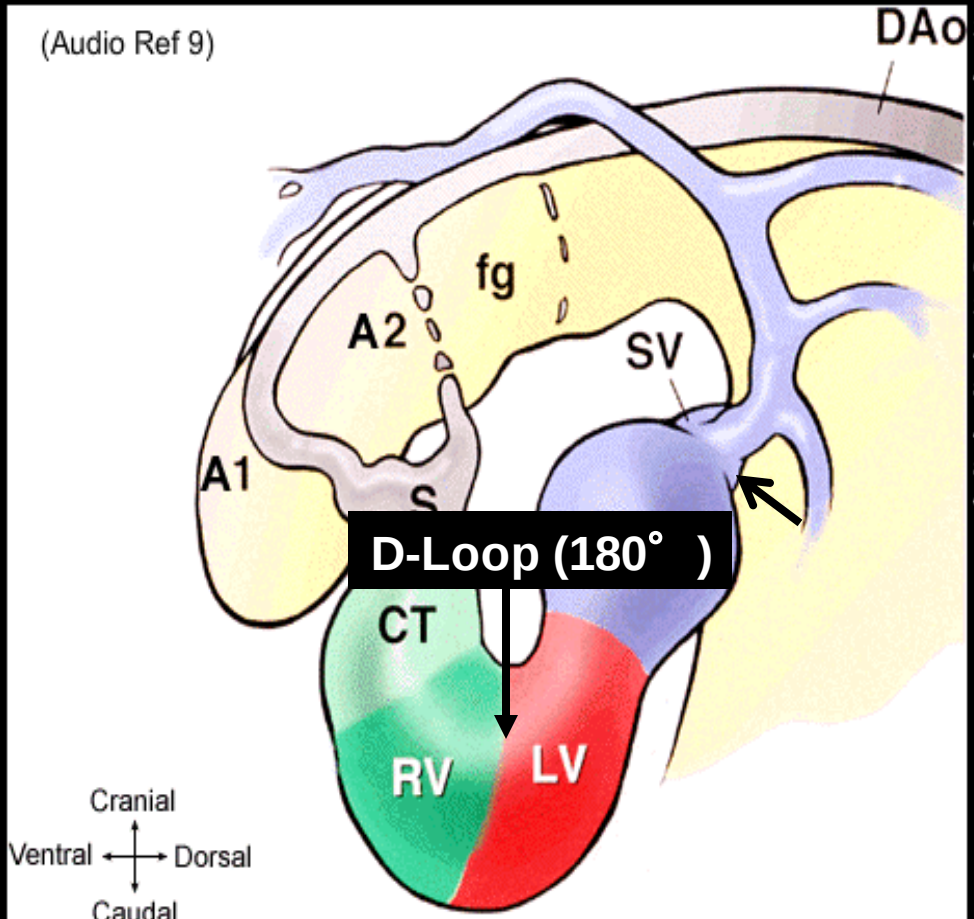
LOOPING

(Audio Ref 9)



SV, Left horn sinus venosus
A, Future atrium
LV, Future left ventricle
RV, Future right ventricle
CT, Future conotruncus
A1, First aortic arch arteries
DAo, Dorsal aorta

D – Loop of the Heart Tube



SV, Sinus venosus
A, Primitive atrium
LV, Presumptive left ventricle
RV, Presumptive right ventricle
CT, Conotruncus
S, Aortic sac
A1, First aortic arch arteries
A2, Second aortic arch arteries forming
fg, Foregut
DAo, Dorsal aorta

LOOPING

D-Loop

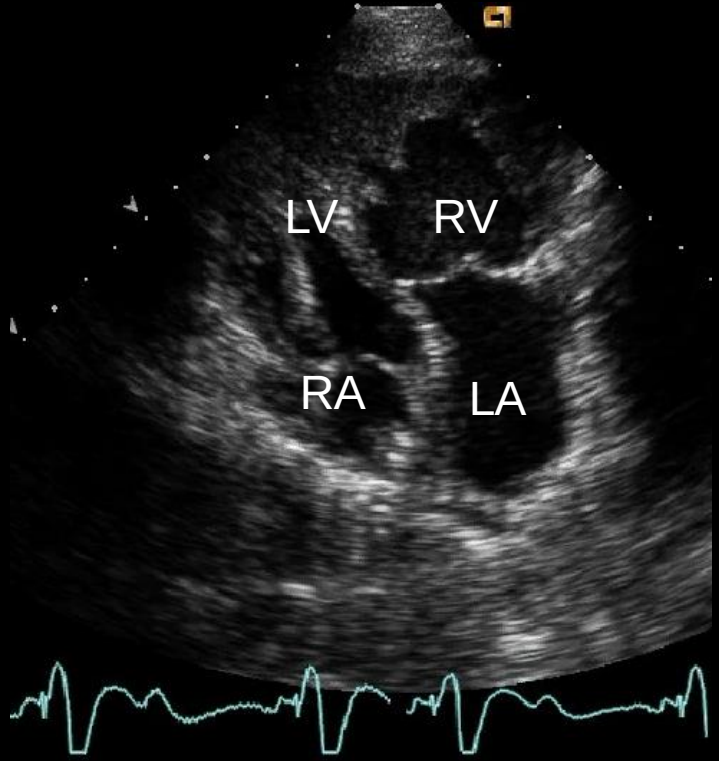
- *Inflow portion* of the morphologic *RV* lies to the *right* of the morphologic *LV*

L-Loop

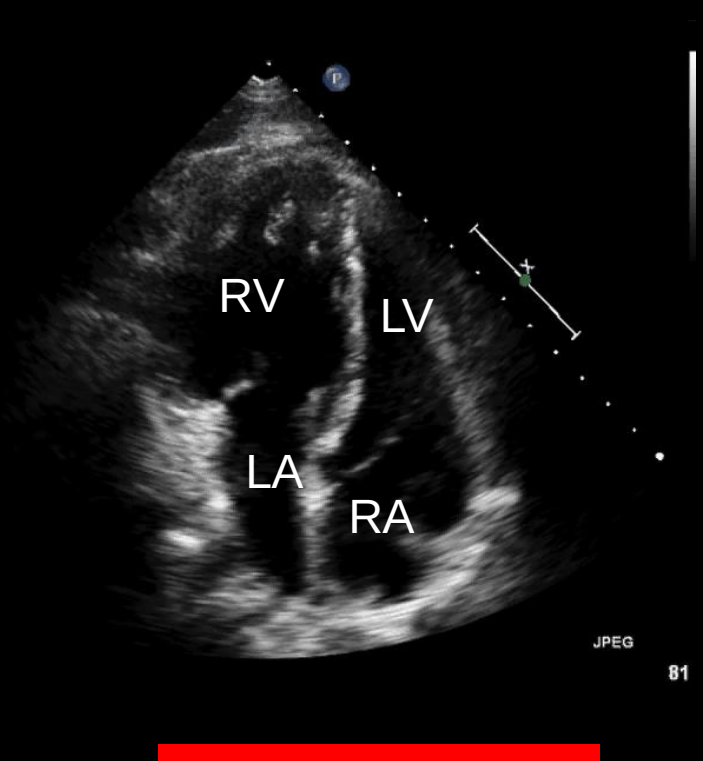
- *Inflow portion* of the morphologic *RV* lies to the *left* of the morphologic *LV*

Normal Heart
D - LOOP

Mustard Procedure
D - LOOP

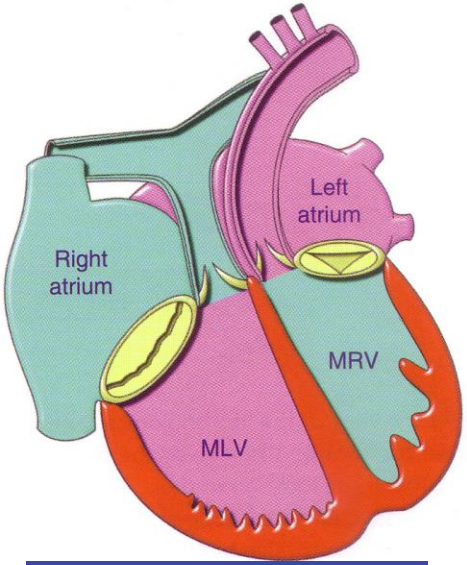


Situs solitus
ccTGA
L – LOOP
(95%)

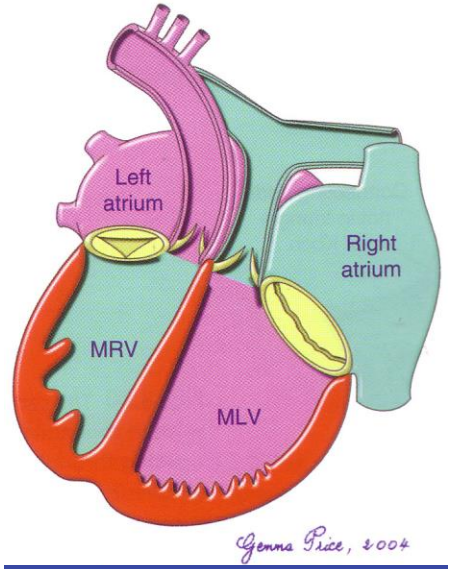


Situs inversus
ccTGA
D – LOOP
(5%)

ccTGA



Situs solitus – 95%



Situs inversus – 5%

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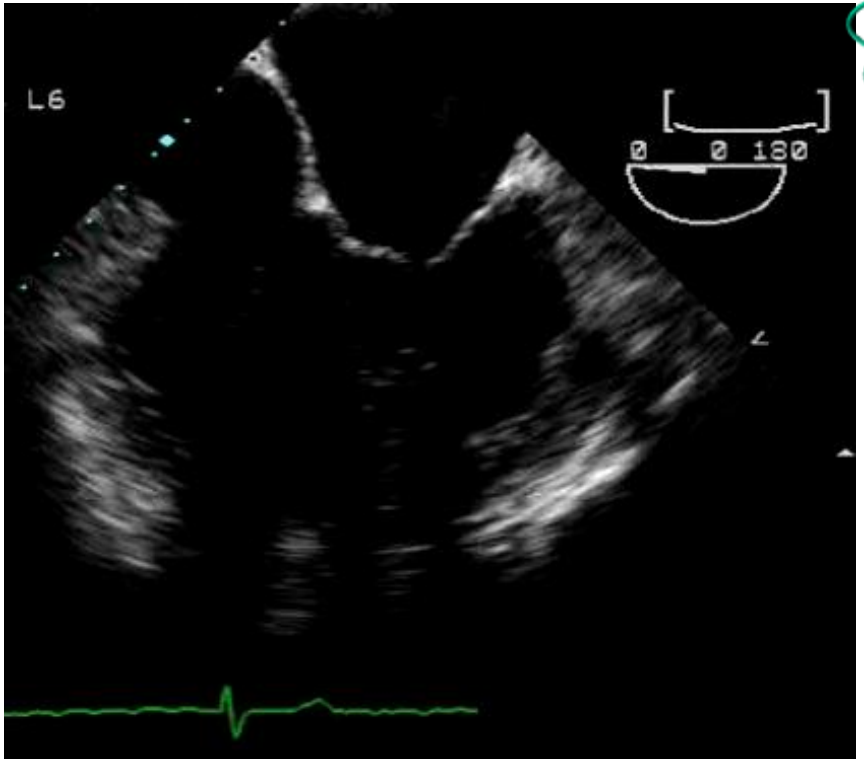
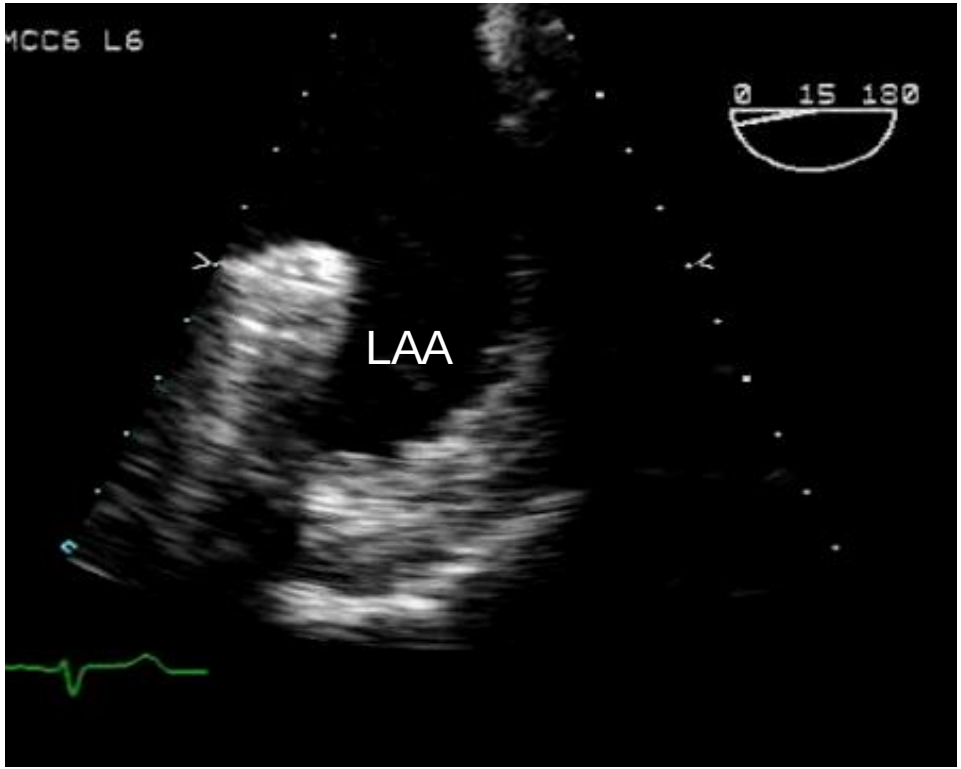
Wilcox B, et al. Surgical Anatomy of the Heart; 2005; p. 251

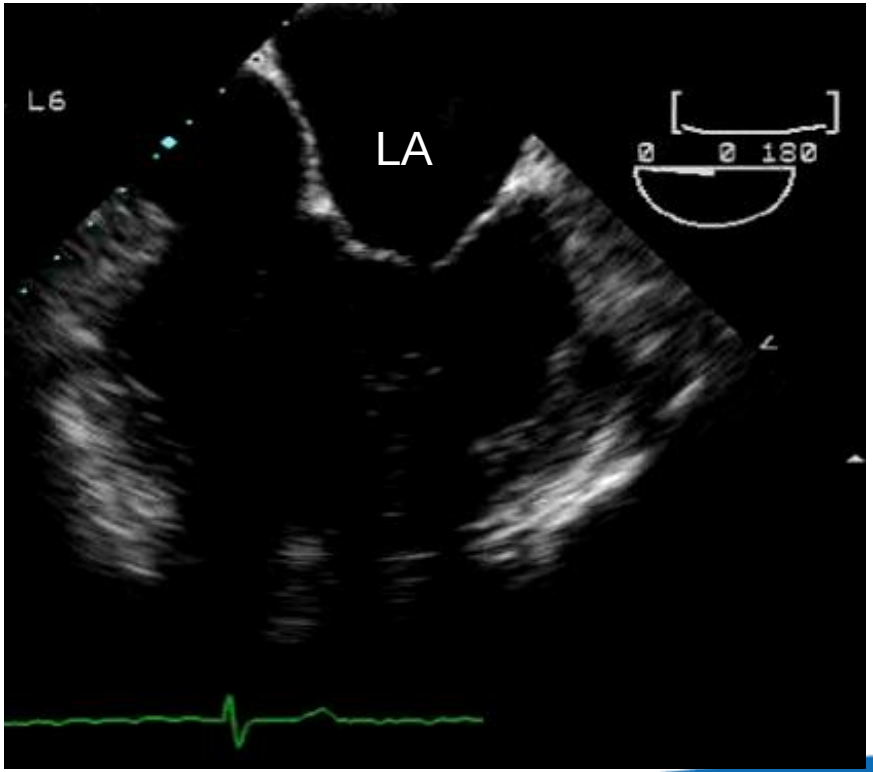
PUT IT TOGETER!



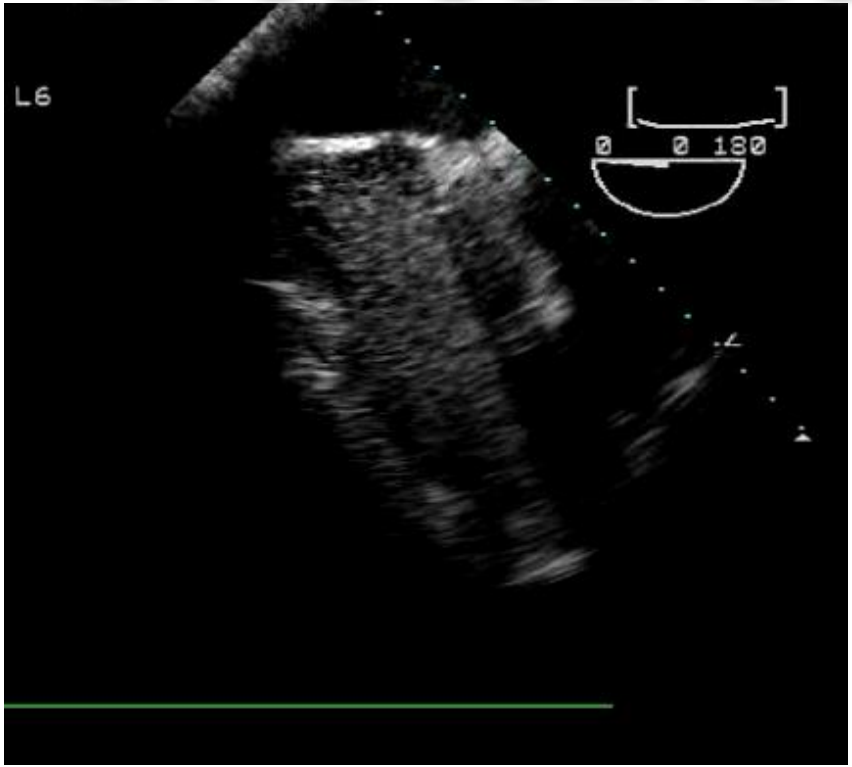
Atrial Arrangement?

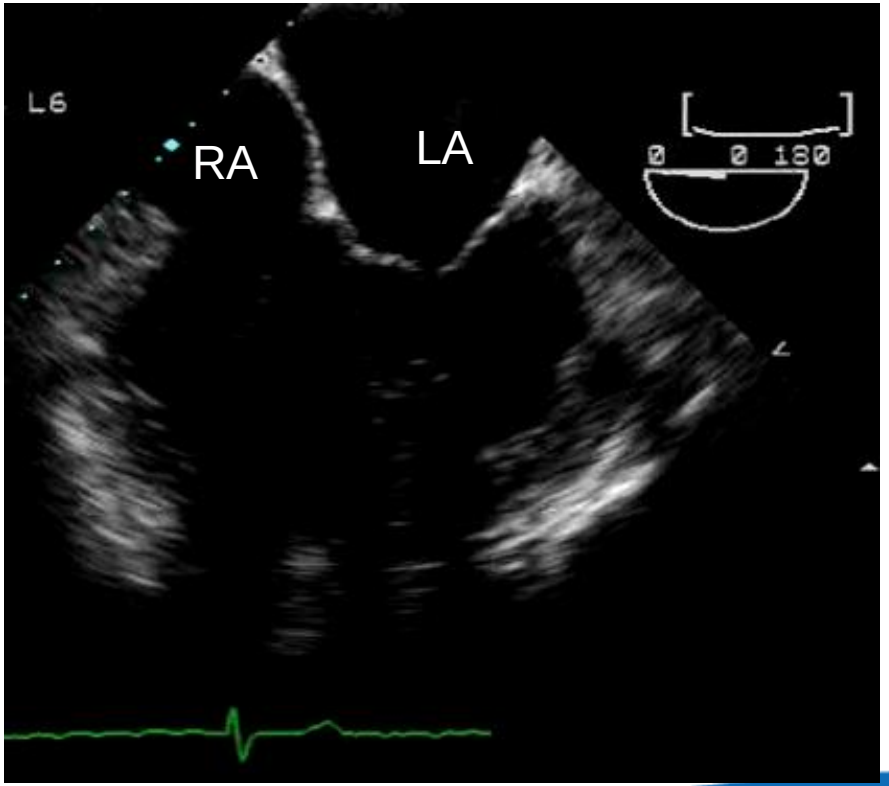
SITUS SOLITUS



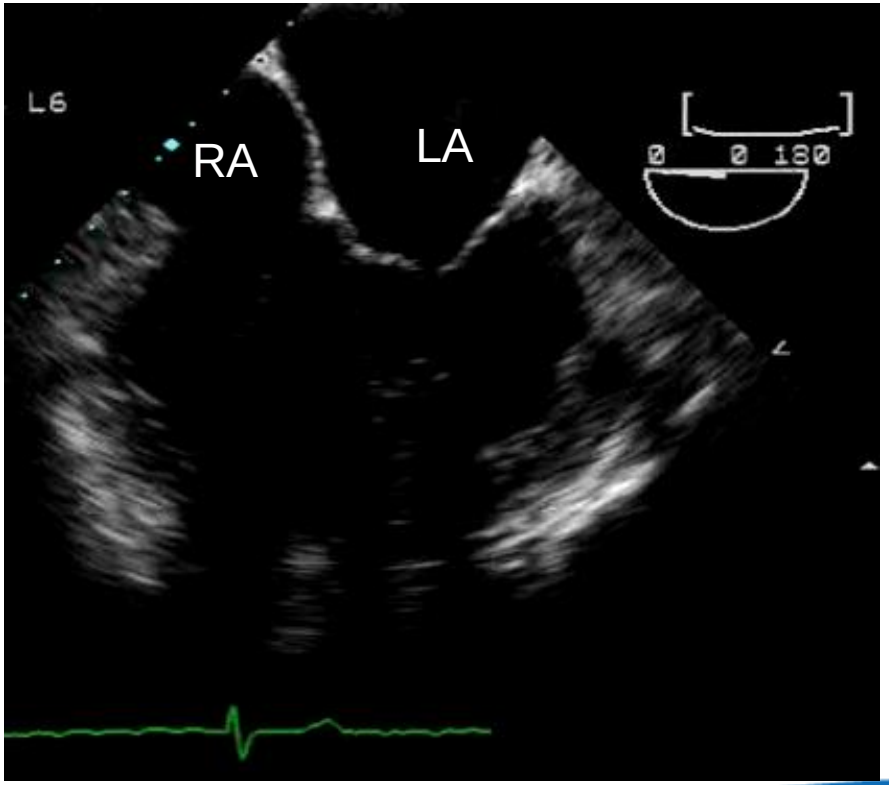


SITUS SOLITUS

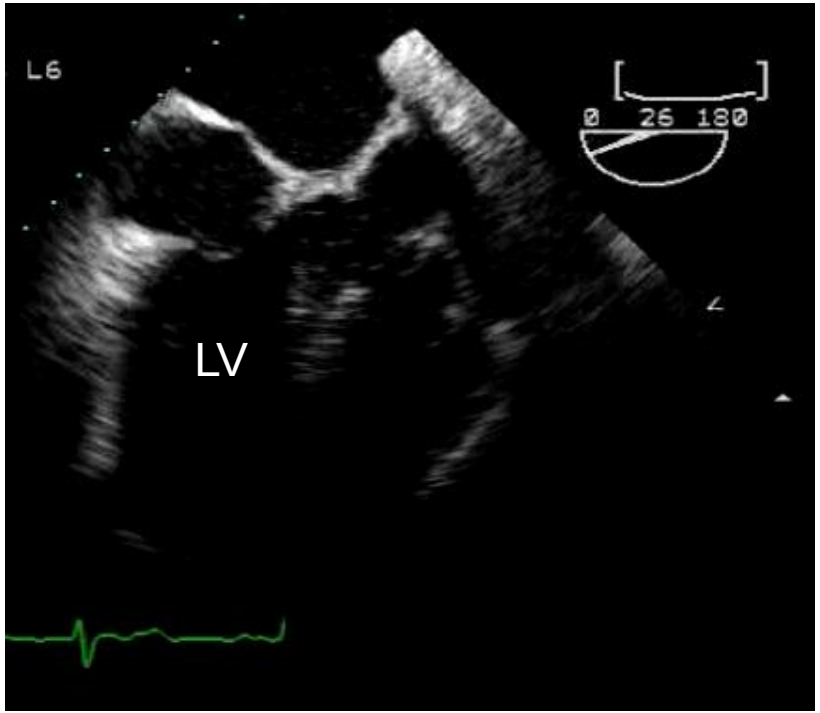


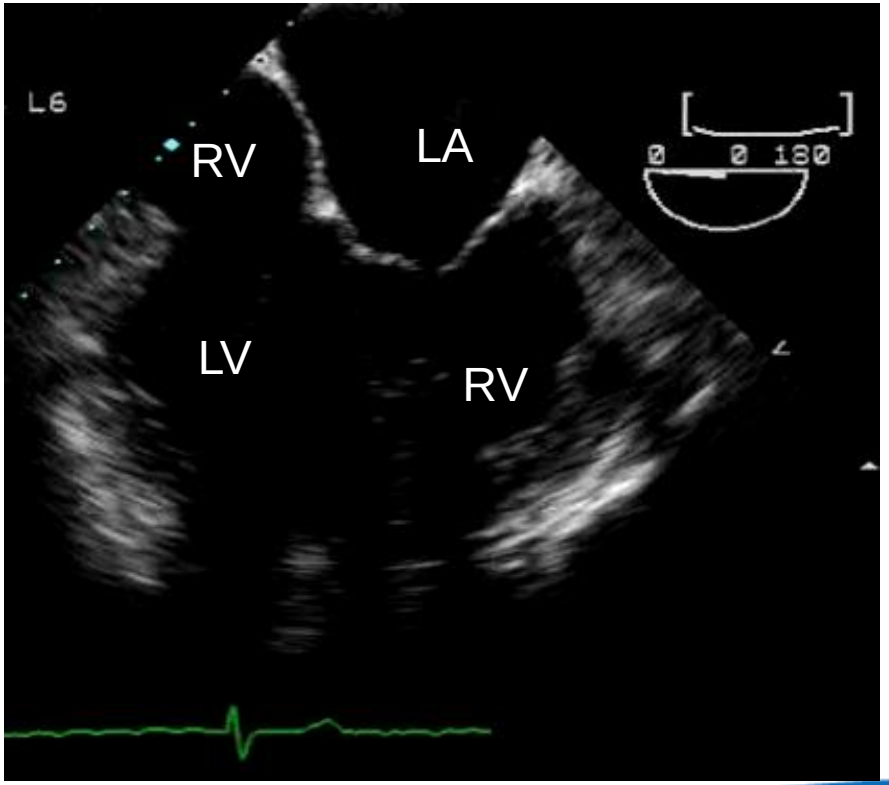


Ventricular Segement?

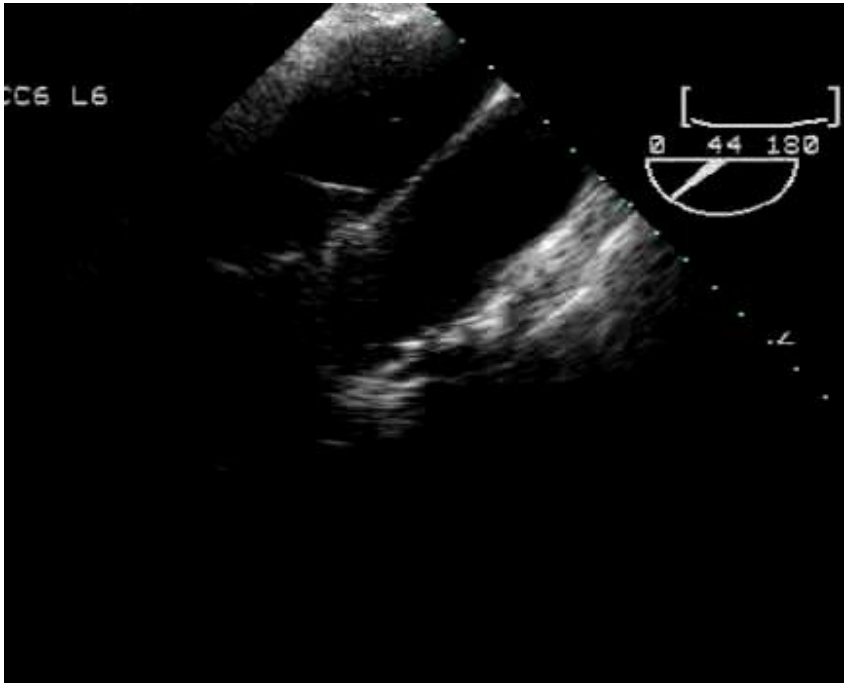


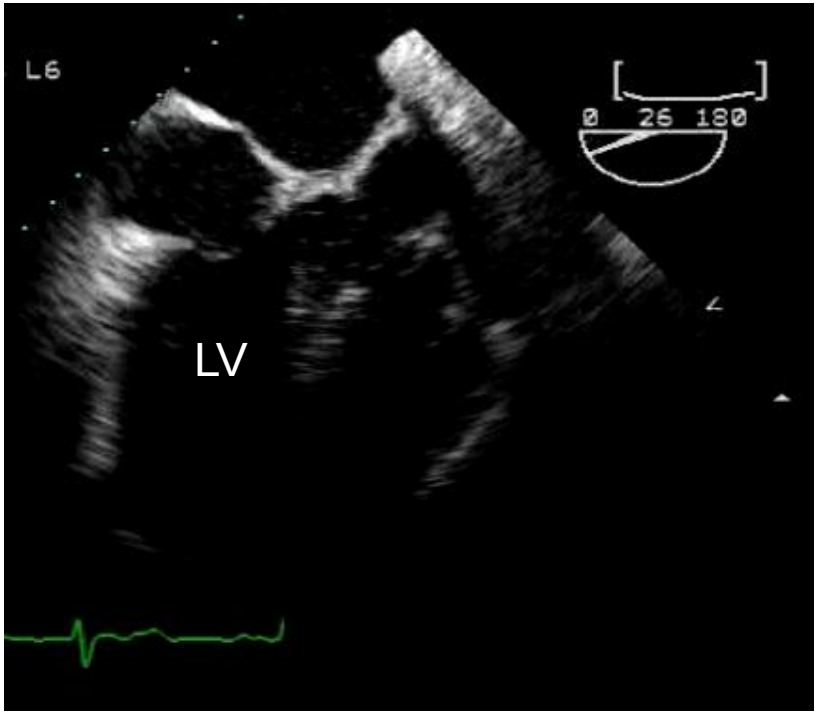
DISCORDANT AV CONNECTION



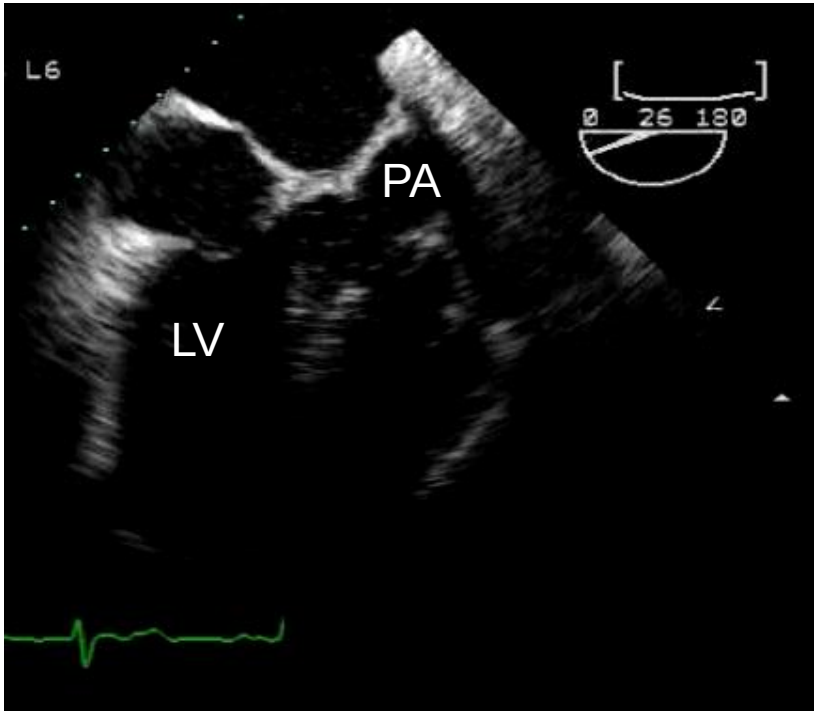


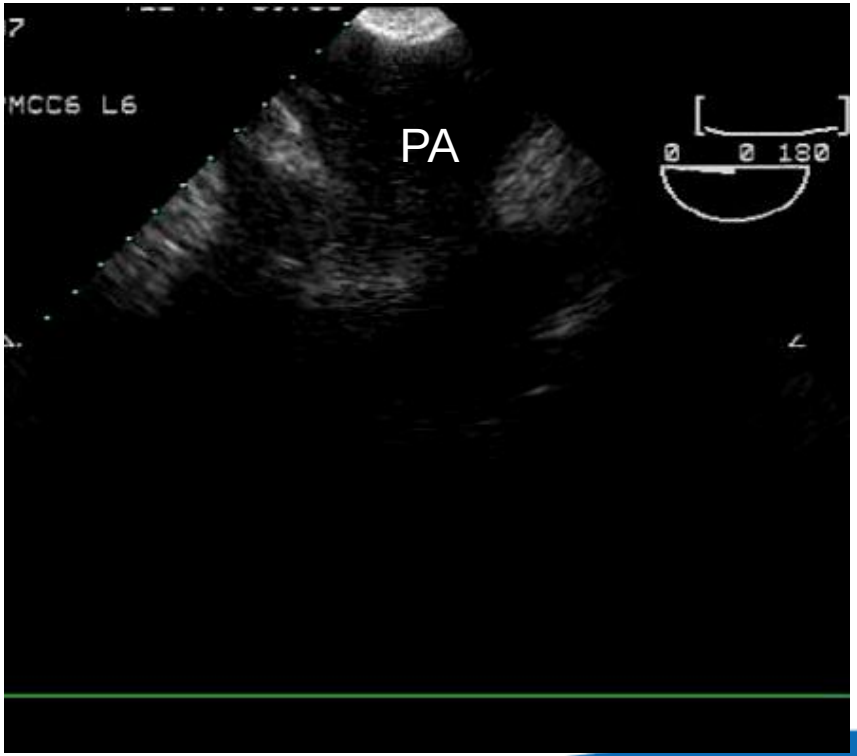
ARTERIAL SEGMENT?



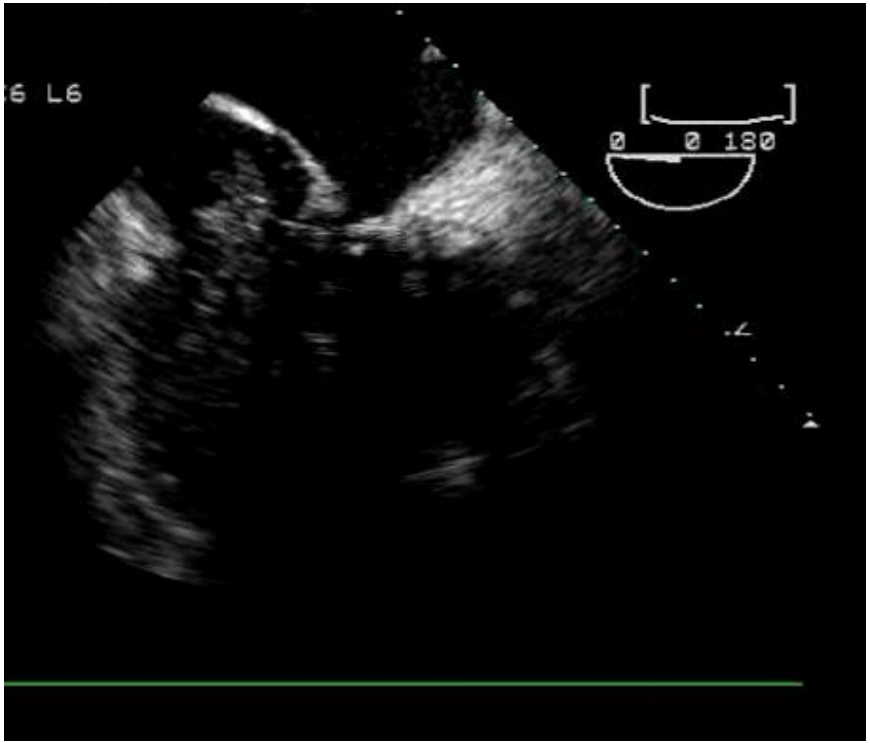


ARTERIAL SEGMENT?





SITUS SOLITUS - CCTGA



SUMMARY

Review Surgical Notes!

Knowledge and and expertise

- Terminology / Anatomy / Morphology
- Surgical Procedures

Long-term complications

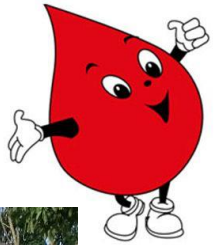
Segmental analysis makes CHD simple

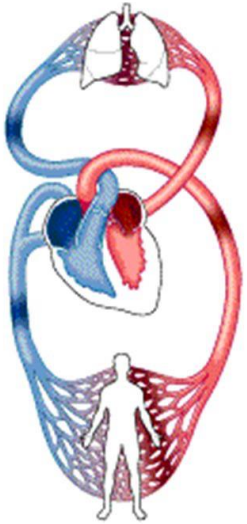
W Y S I W Y D

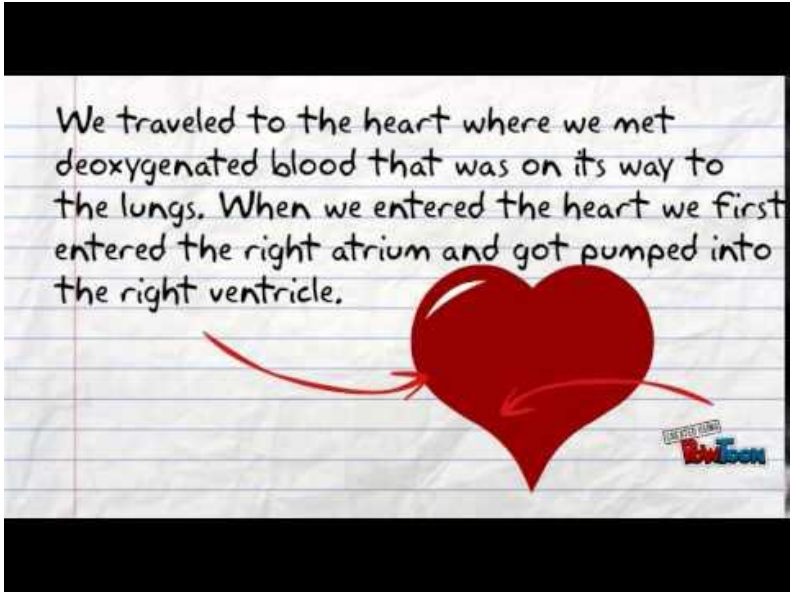
What ***Y***ou ***S***ee ***I***s

What ***Y***ou ***D***escribe

The journey of a red blood cell







Dr. Jane Heggie

"You need to know the journey of the red blood cells"

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ACKNOWLEDGEMENT
CARDAIC ANESTHESIA TEAM

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