



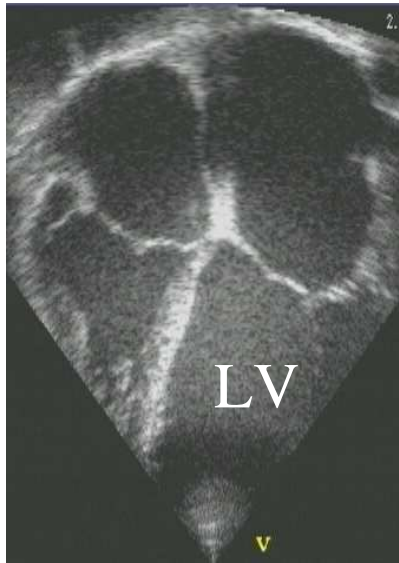
Segmental approach to normal and abnormal situs arrangement - Echocardiography -



Jan Marek
Great Ormond Street Hospital
London

How the heart should be imaged? “European Chaos”

- “Transatlantic”
way

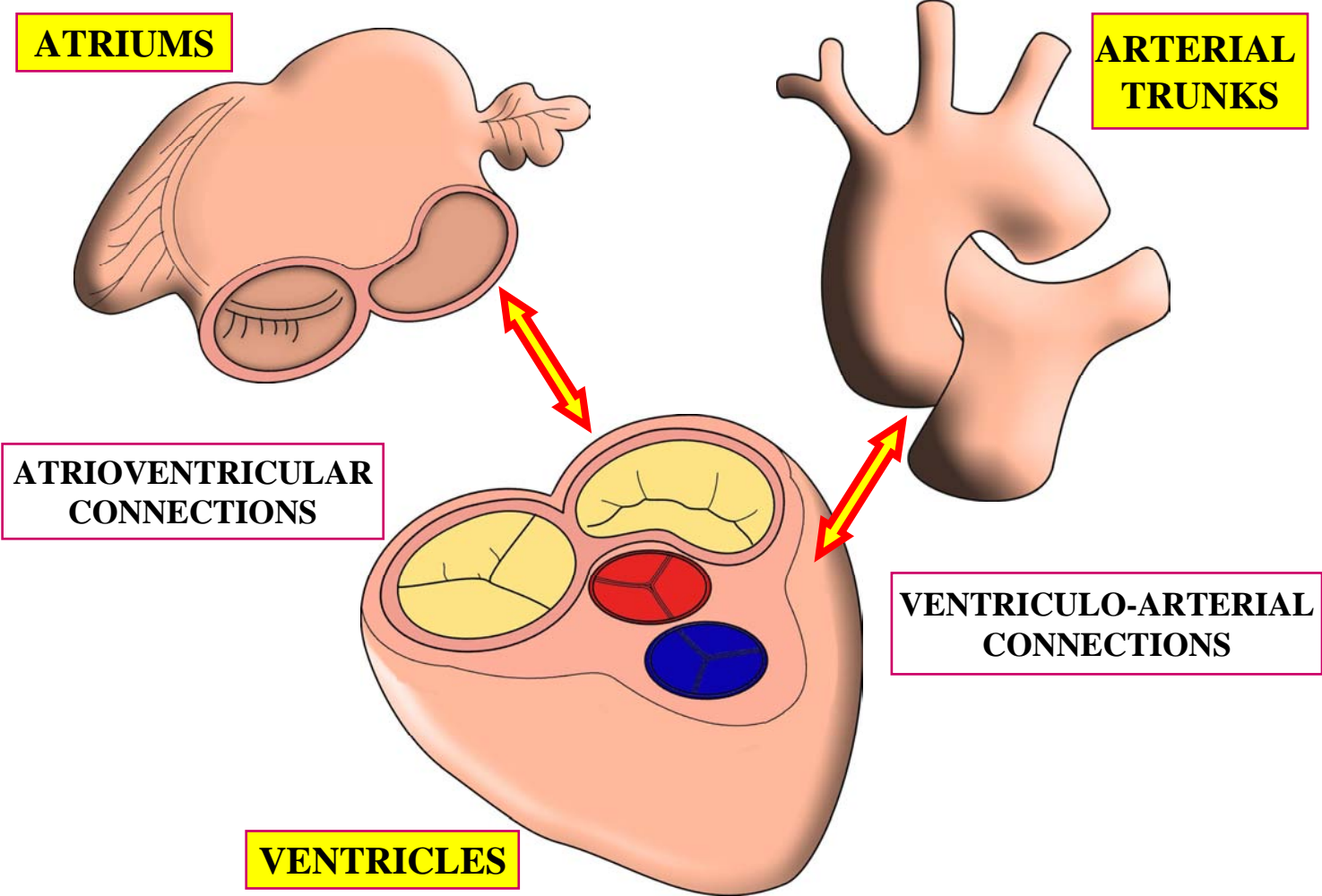


- “Adult” way

- “Dutch” way



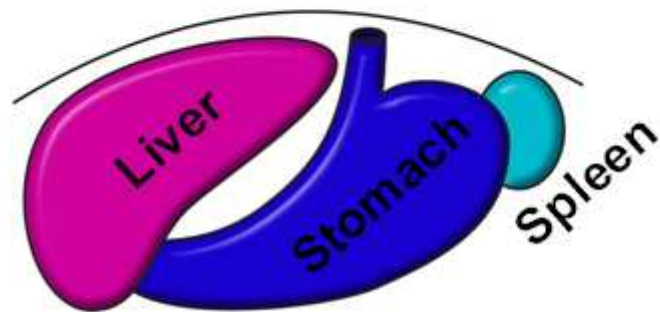
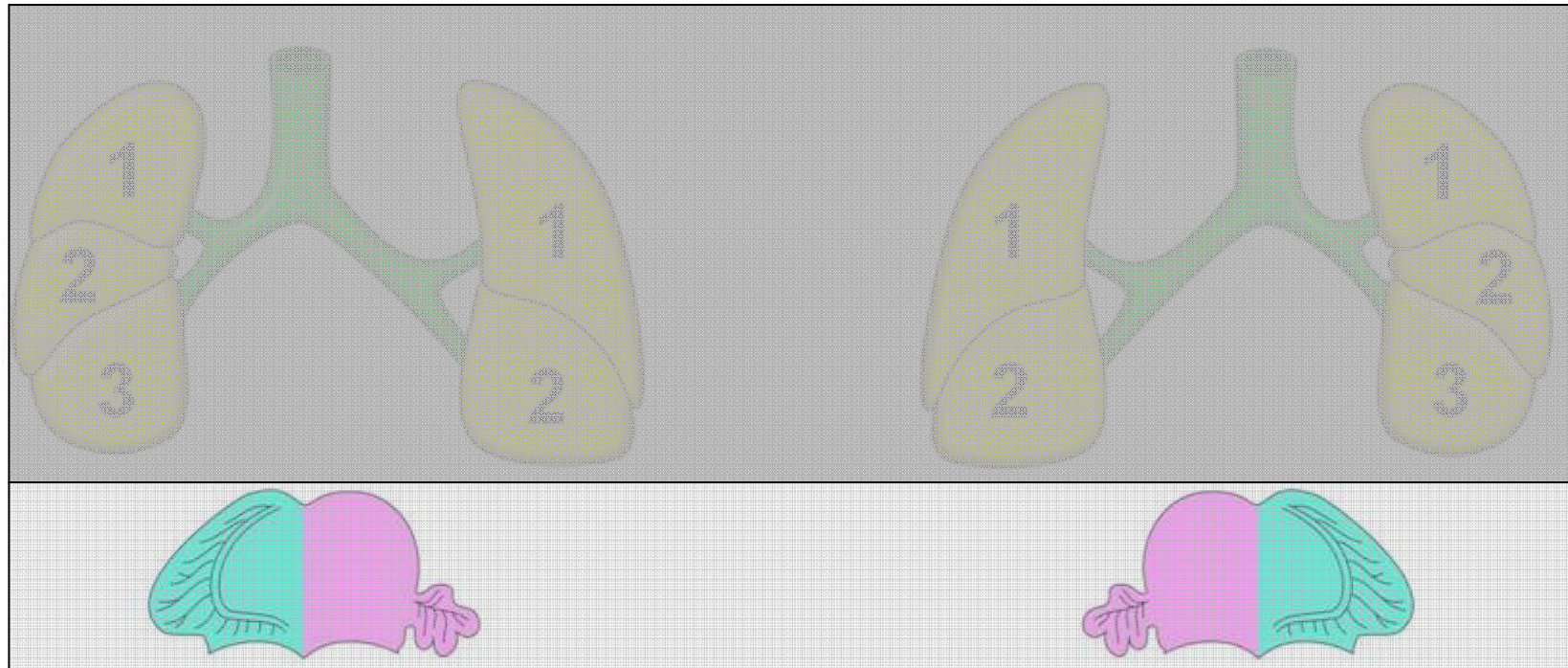
SEQUENTIAL SEGMENTAL ANALYSIS



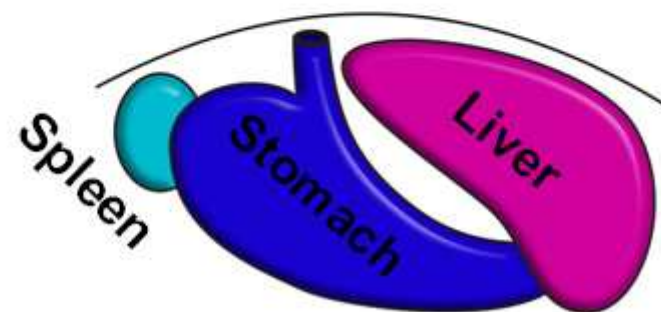
Courtesy RH Anderson

- Abdominal situs !
- Atria - Atrial situs (systemic and pulmonary venous connections)
- AV connections
- Ventricles
- VA connections
- Aorta, PA trunk (AO arch and PA branches)

LATERALIZED BODILY ARRANGEMENT

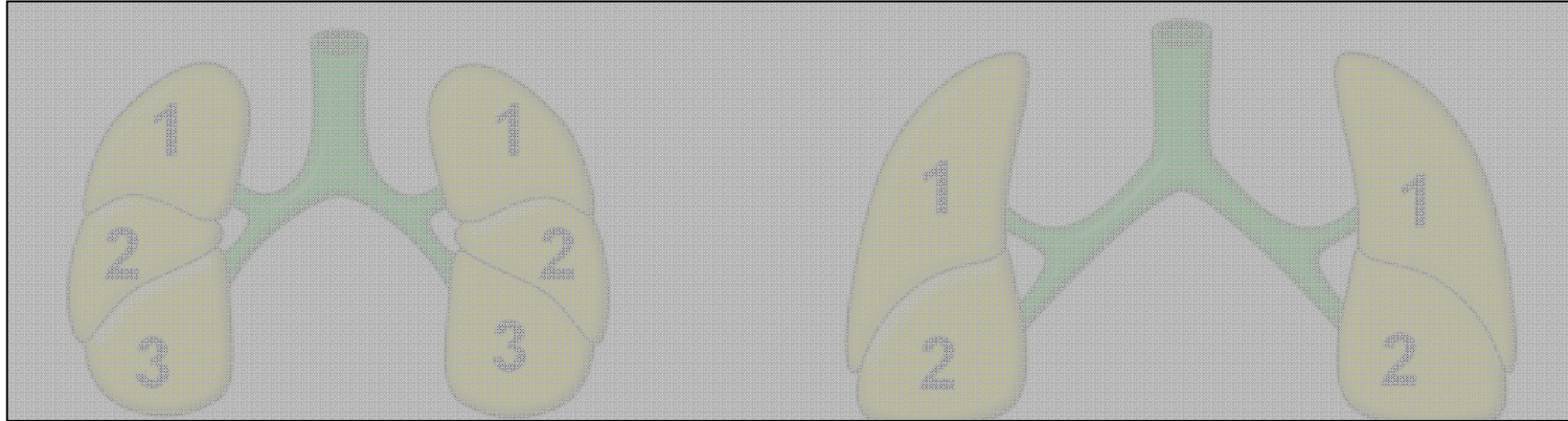


Usual



Mirror-imagery

VISCERAL HETEROTAXY

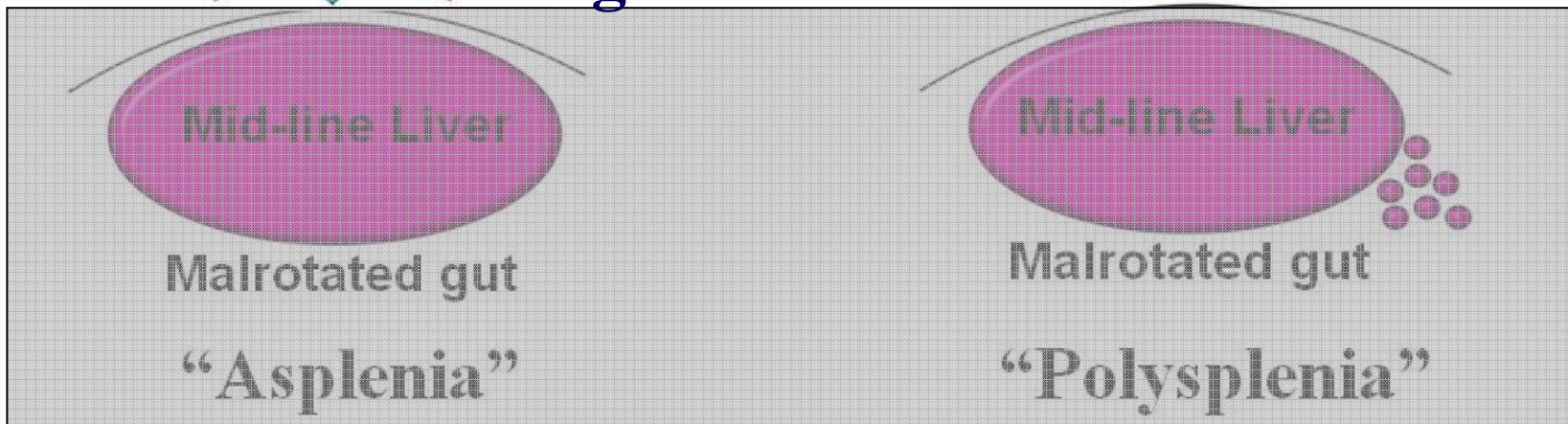
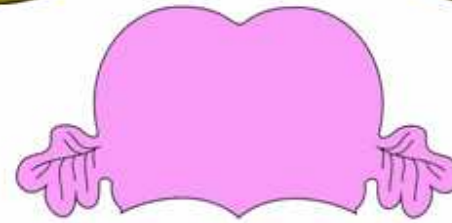


Atrial

Isomerism

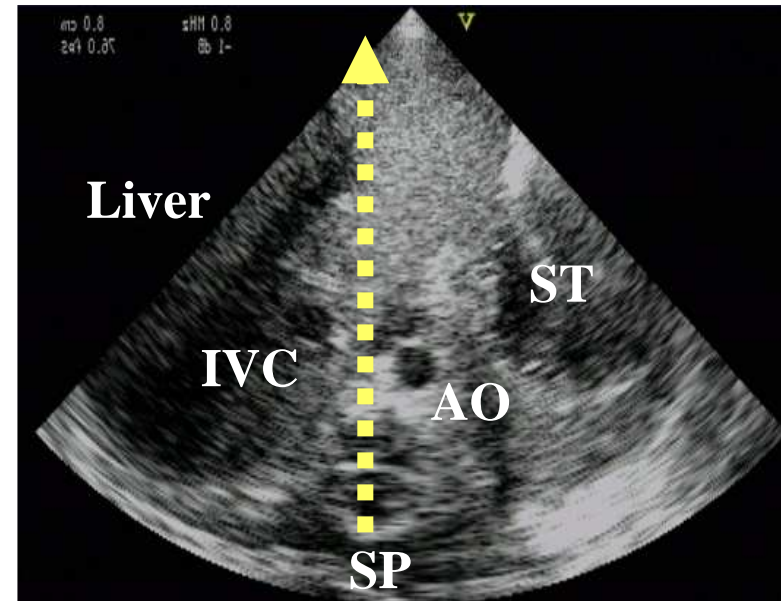
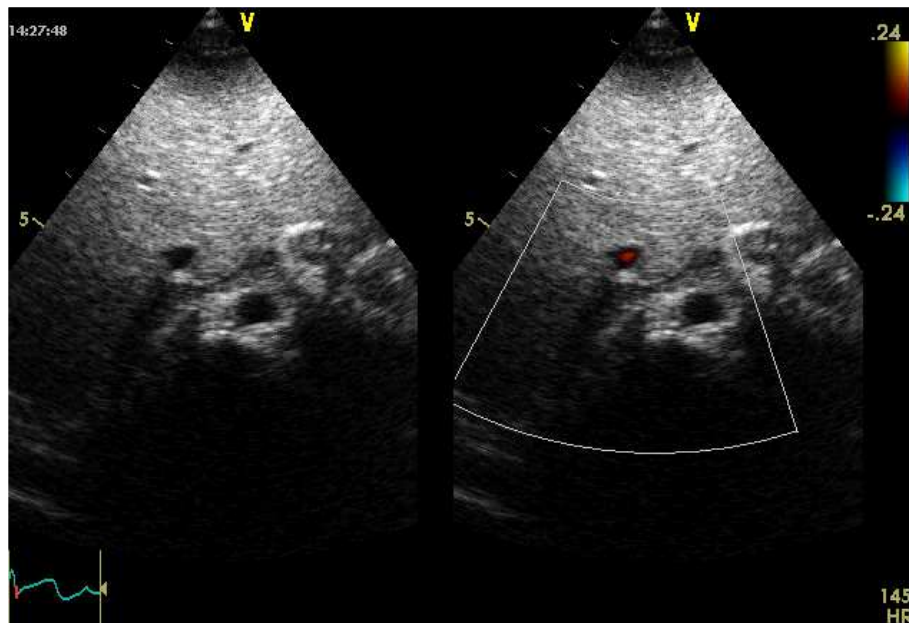
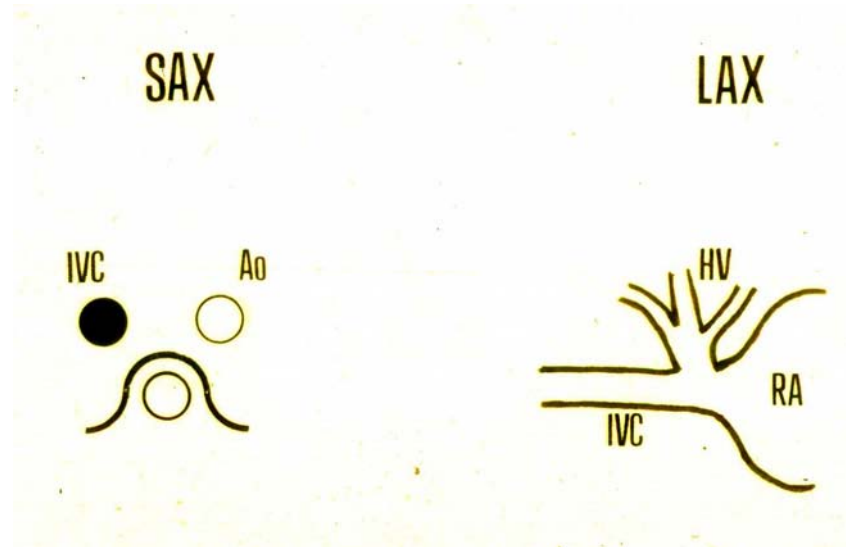
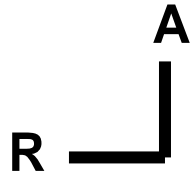
Right

Left

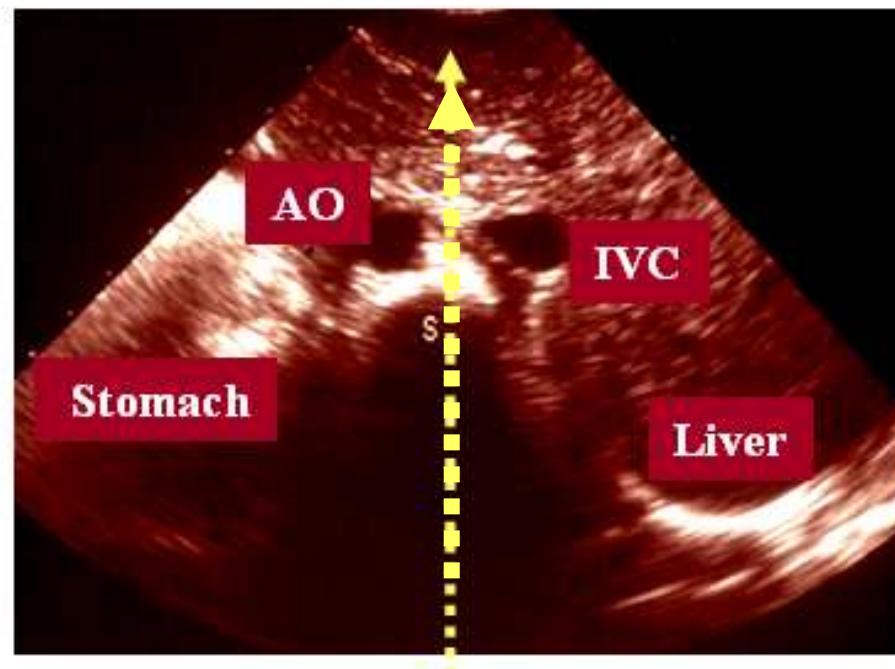
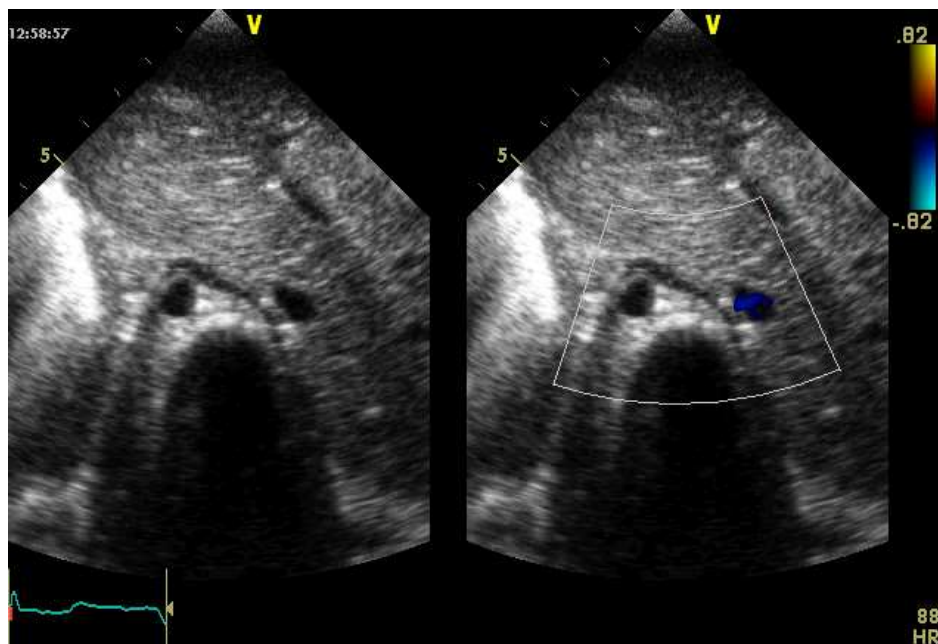
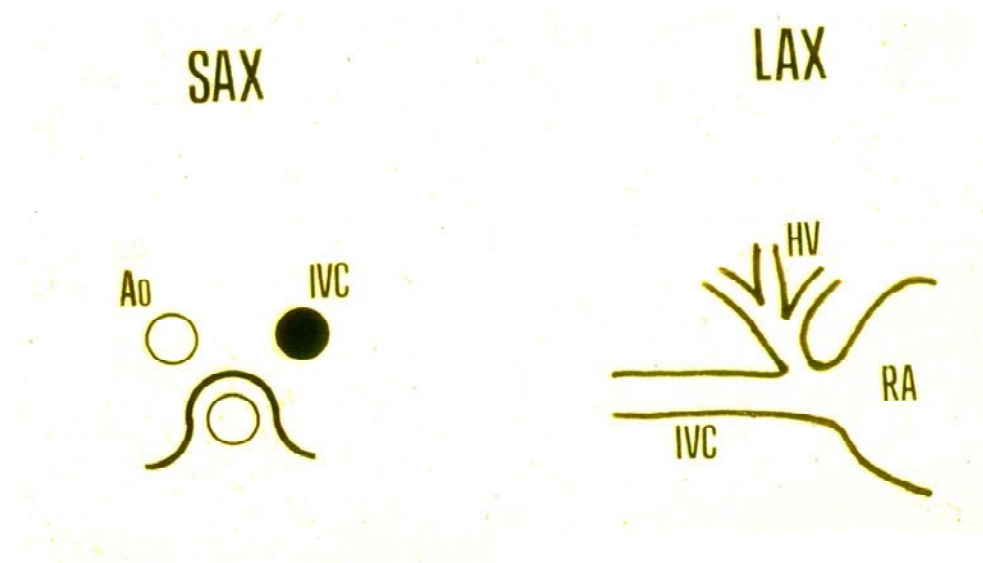
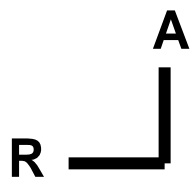


- **Abdominal situs**
- **Atria - Atrial situs** (systemic and pulmonary venous connections)
- **AV connections**
- **Ventricles**
- **VA connections**
- **Aorta, PA trunk** (AO arch and PA branches)

Usual Arrangement (Solitus)

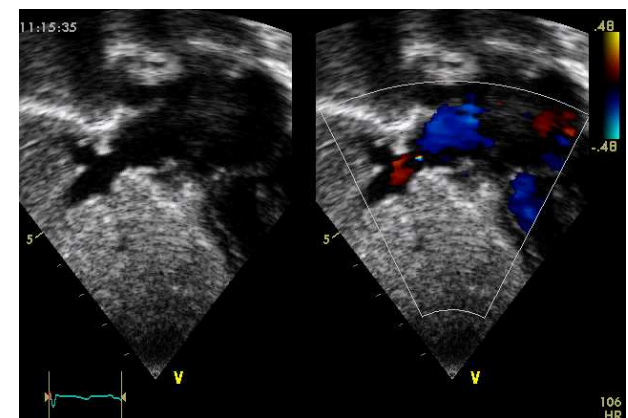
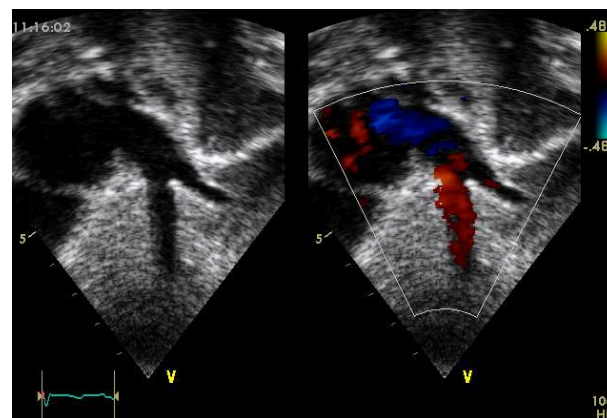
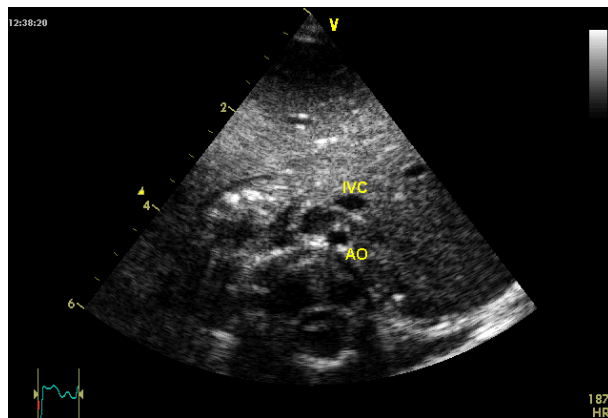
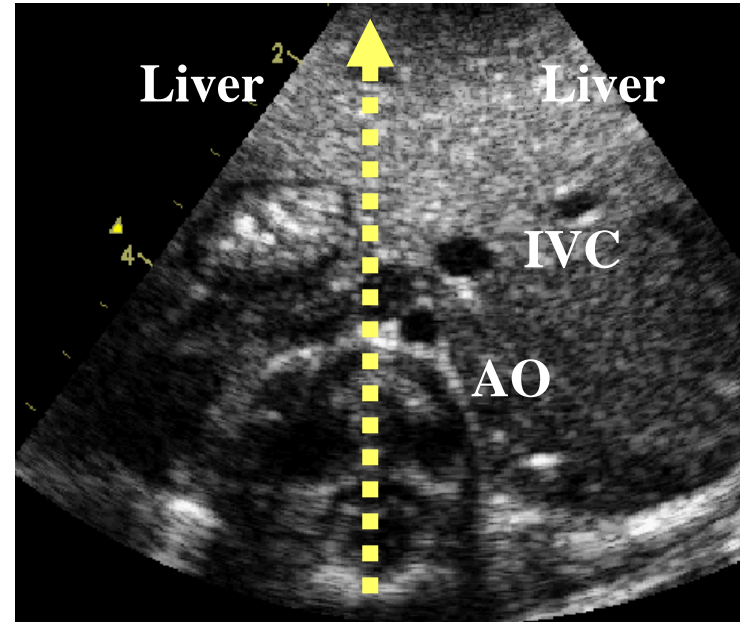
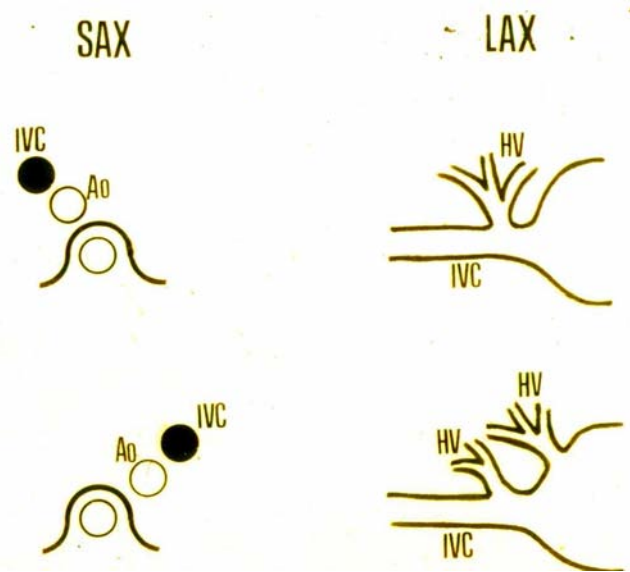


Mirror Imagery (Inversus)

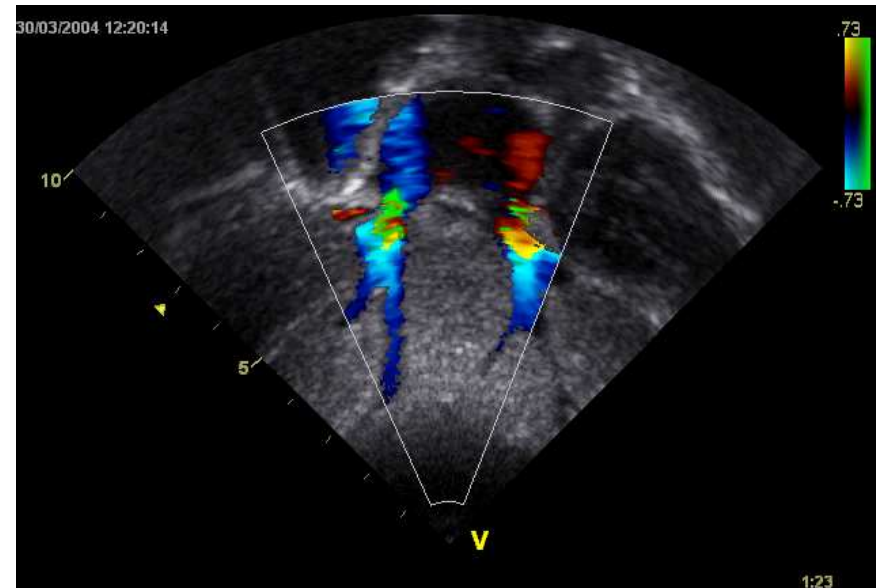
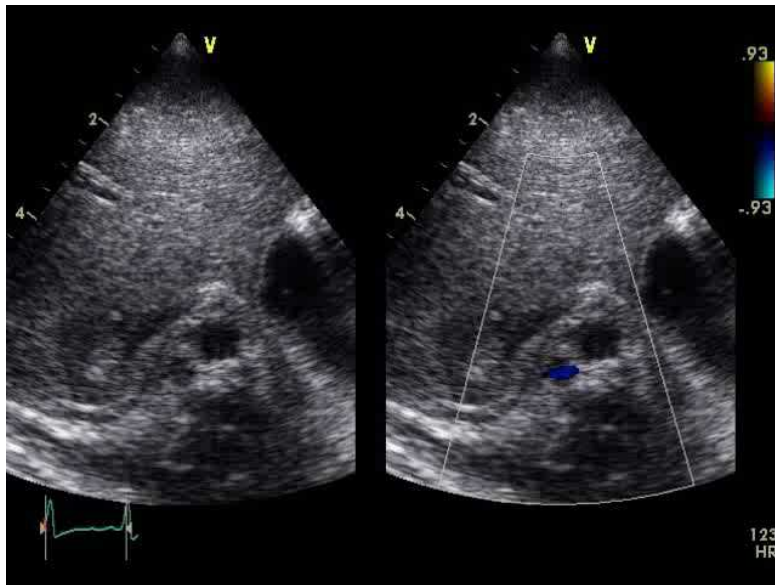
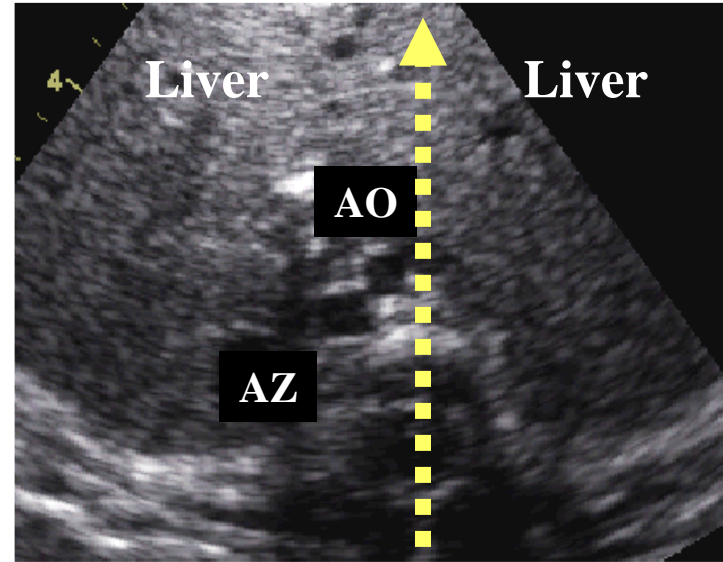
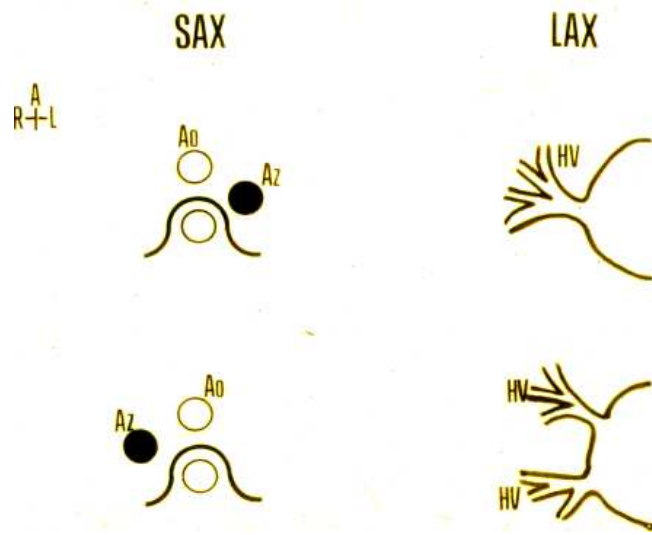
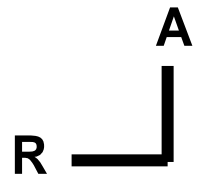


Right Isomerism (Asplenia)

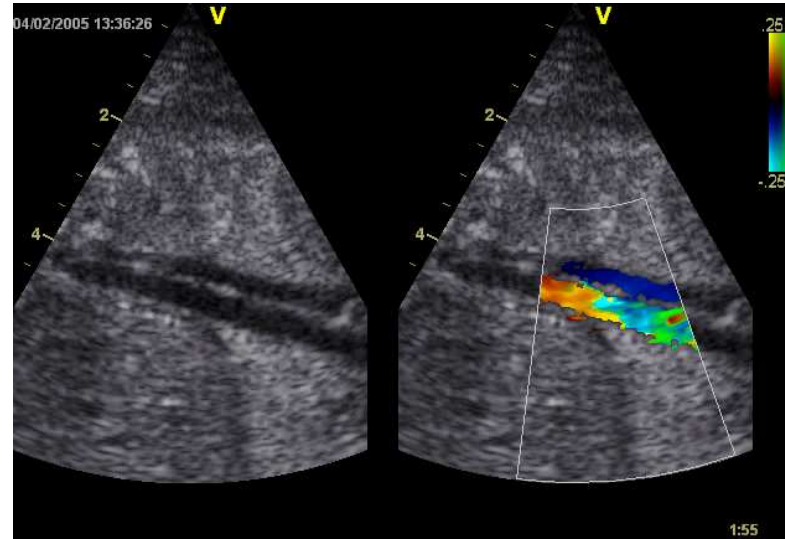
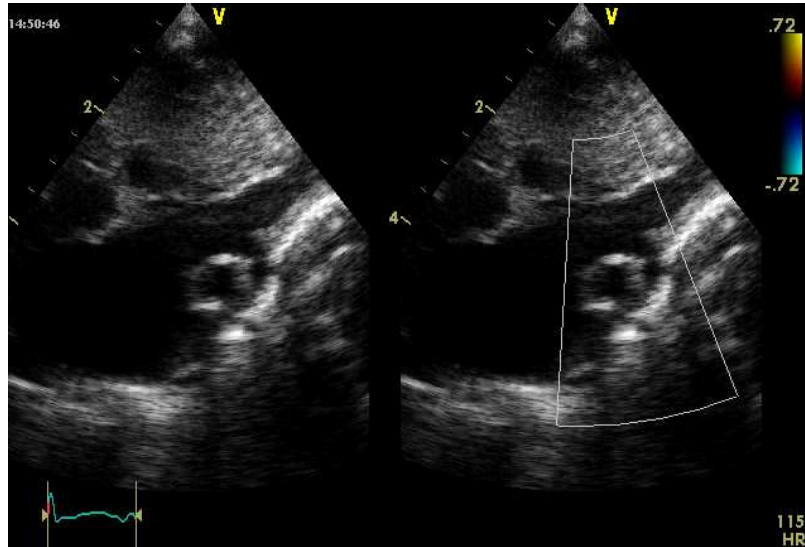
R
A



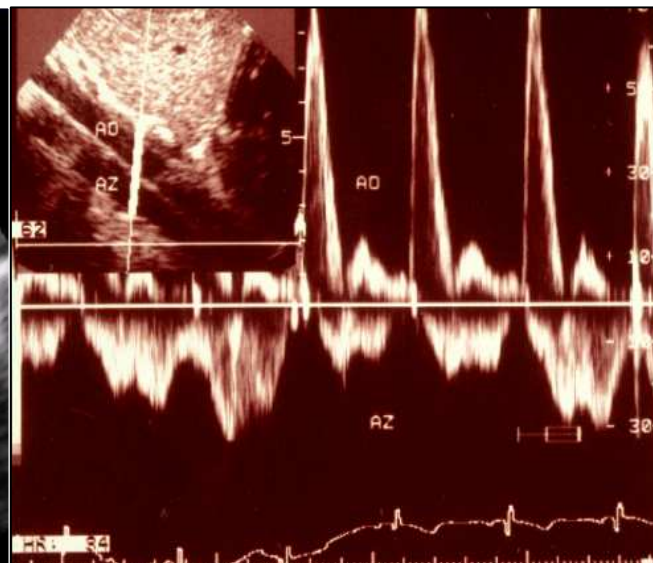
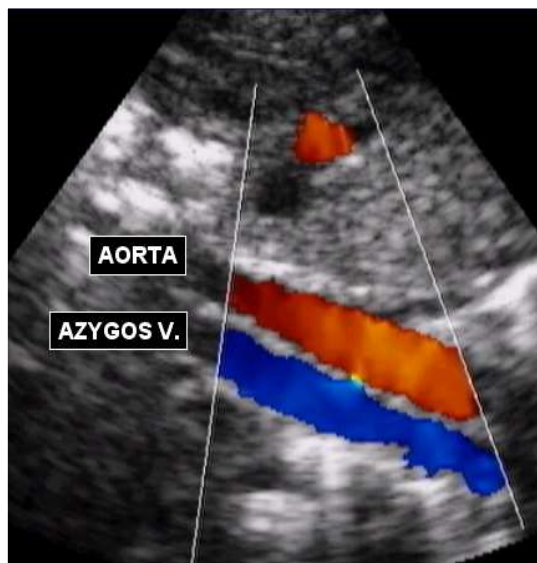
Left Isomerism (Polysplenia)



Left Isomerism (Polysplenia)



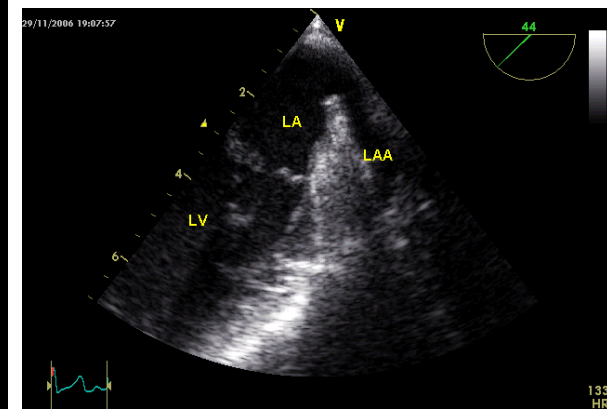
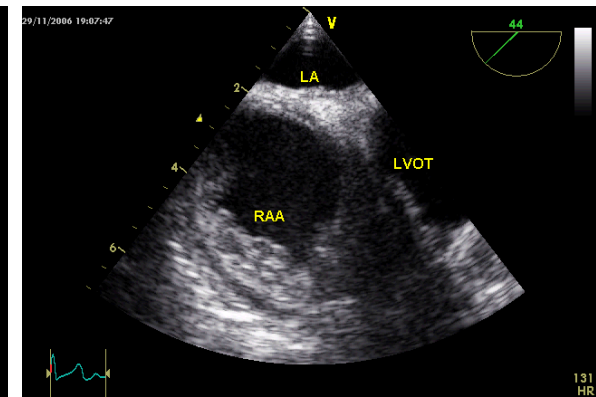
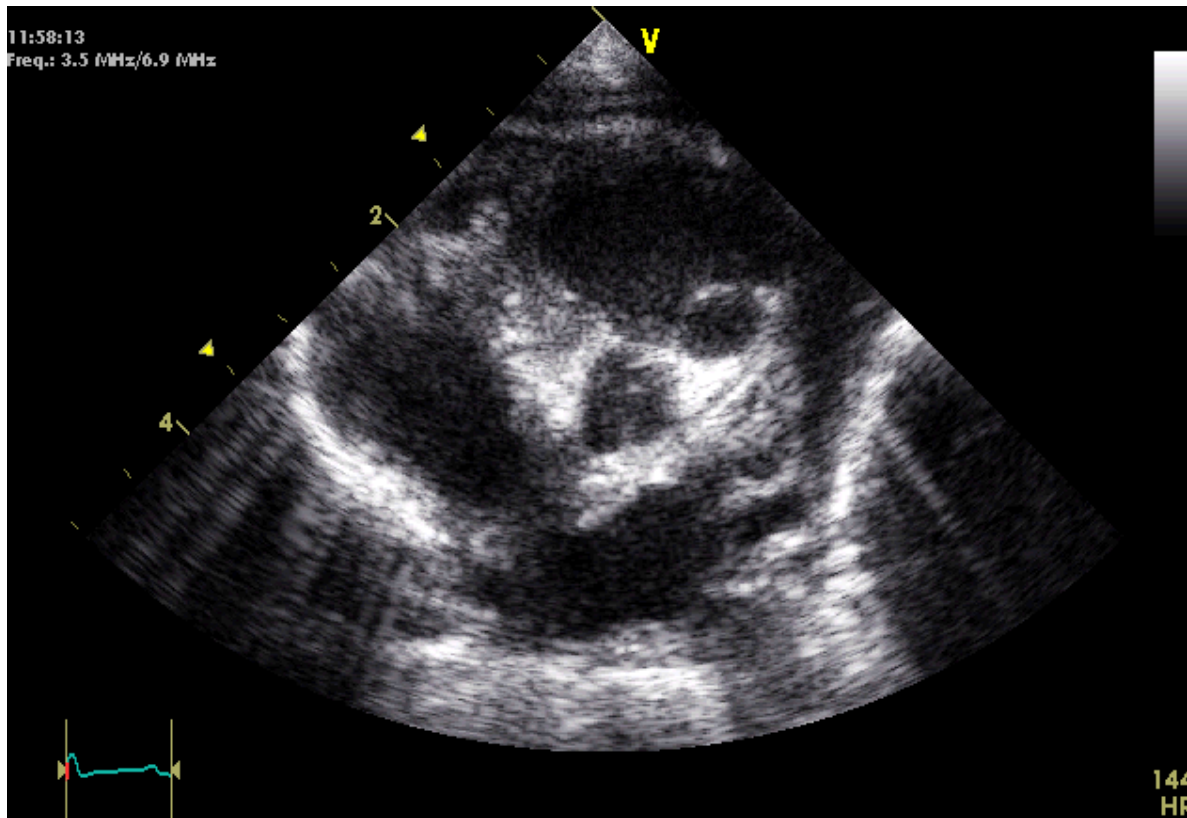
Fetus
32 WoG



- Abdominal situs
- **Atria - Atrial situs** (systemic and pulmonary venous connections)
- AV connections
- Ventricles
- VA connections
- Aorta, PA trunk (AO arch and PA branches)

Usual Arrangement (Solitus)

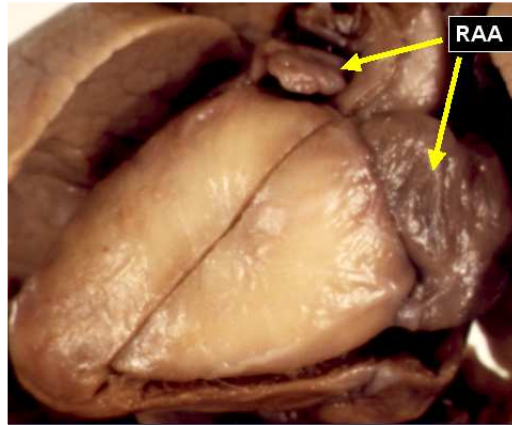
- Atrial appendages -



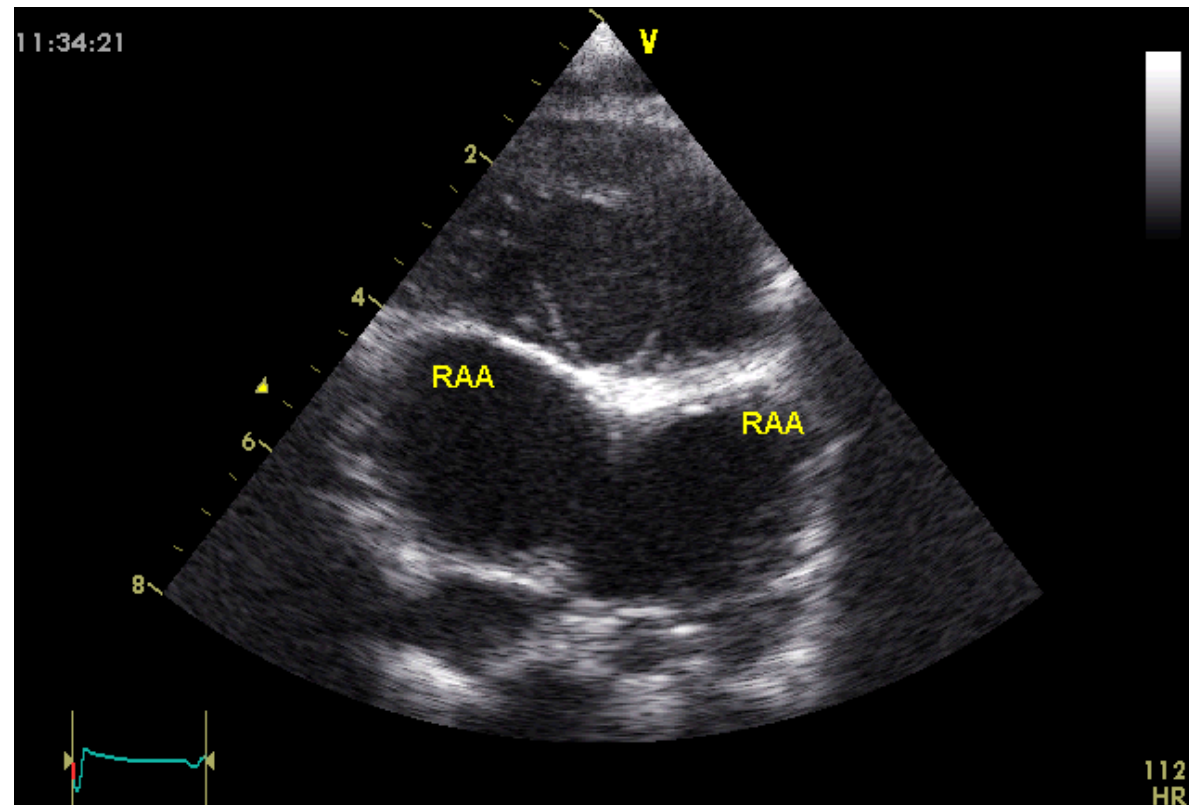
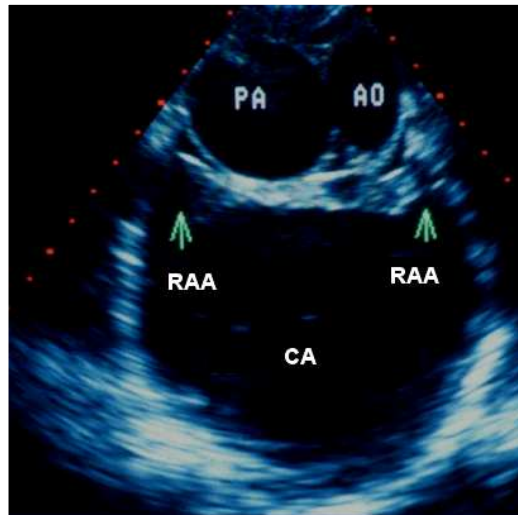
Right Isomerism (Asplenia)

- Atrial appendages -

Fetus 20. WoG

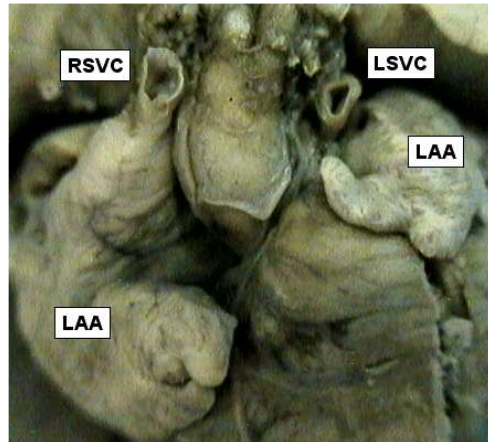


Courtesy V.Povysilova

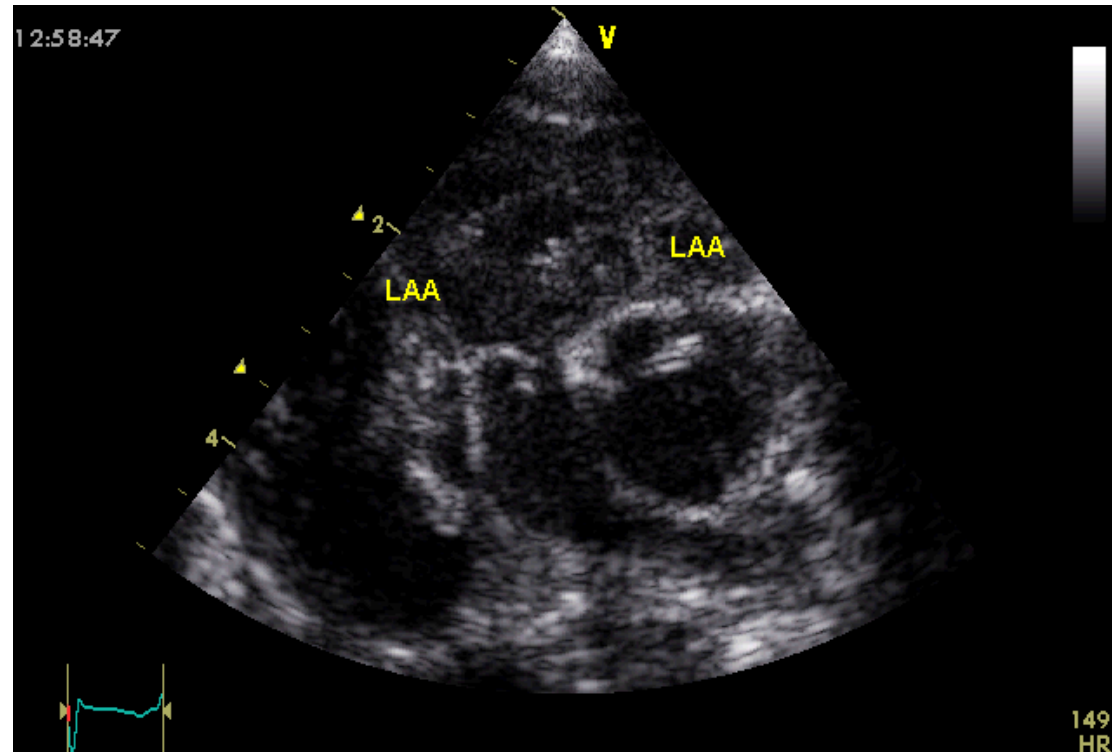
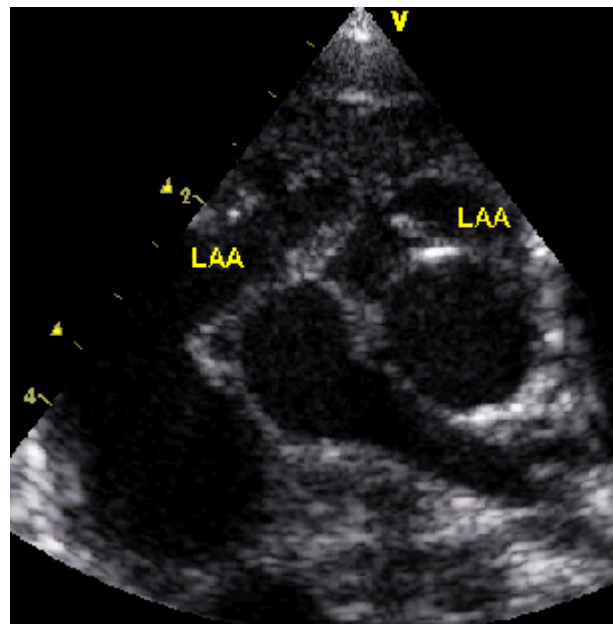


Left Isomerism (Polysplenia)

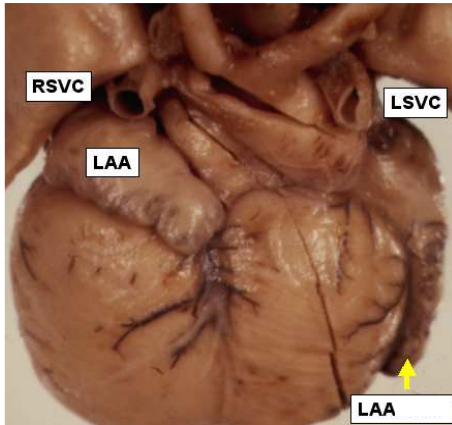
- Atrial appendages -



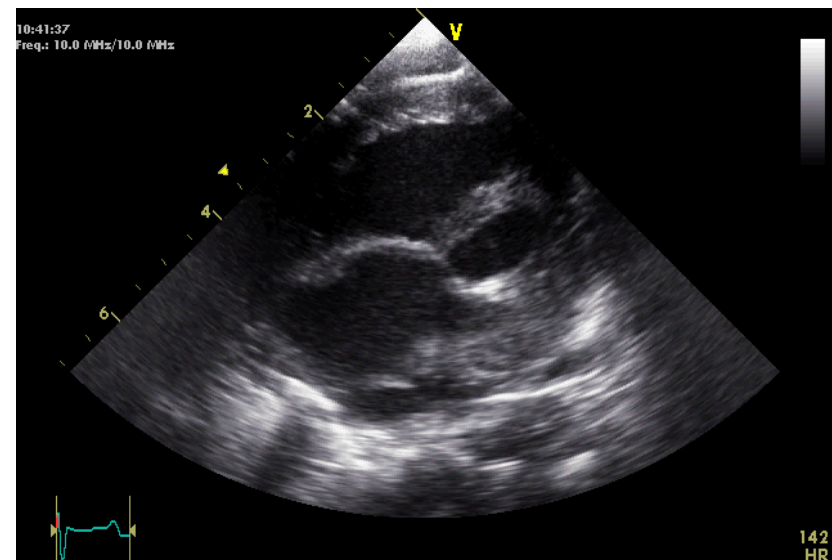
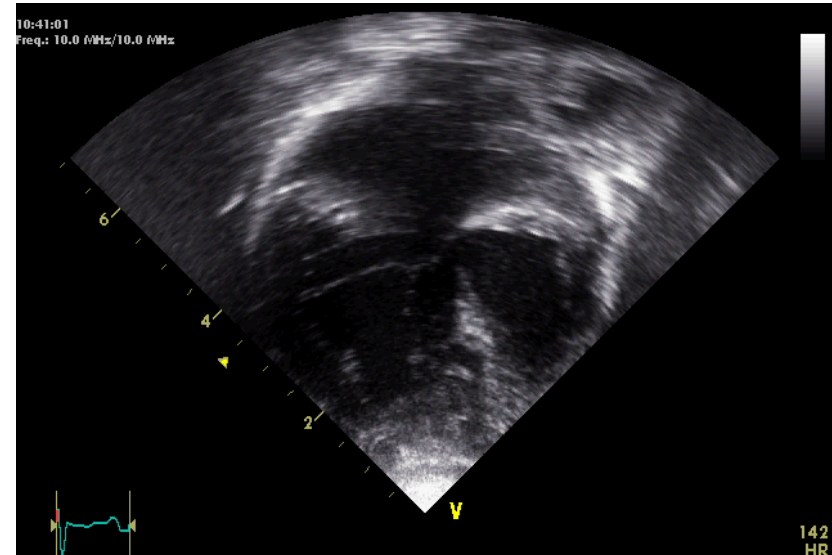
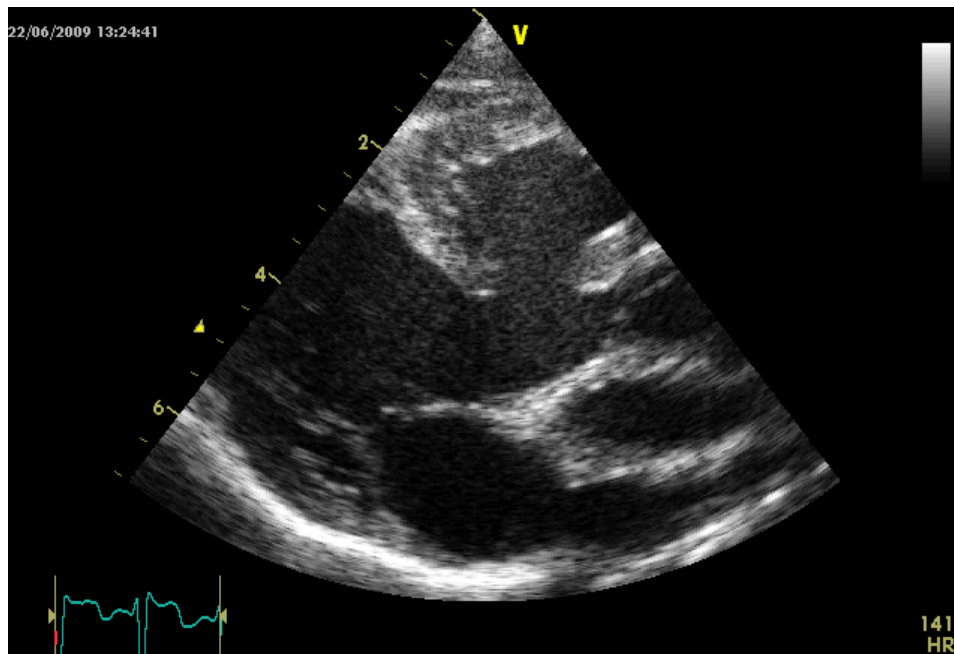
Courtesy V. Povysilova



Atrial appendages - Juxtaposition -



Courtesy V. Povysilova



“Usual” arrangement
SOLITUS



“Unusual arrangement”
ISOMERISMUS



Courtesy Ian D Sullivan

- Abdominal situs
- Atria - Atrial situs (systemic and pulmonary venous connections)
- **AV connections**
- Ventricles
- VA connections
- Aorta, PA trunk (AO arch and PA branches)

Atrio-ventricular Connections

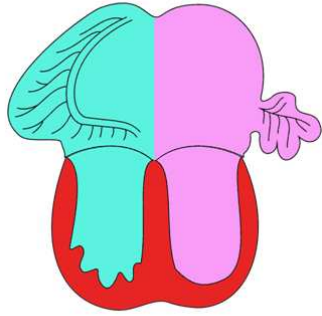
- Concordant
- Discordant
- Univentricular AV connections

Absent AV connection

Common AV valve

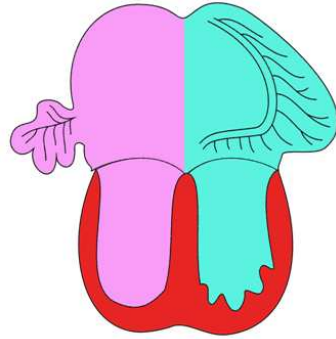
Double inlet connections

Usual Atrial Arrangement



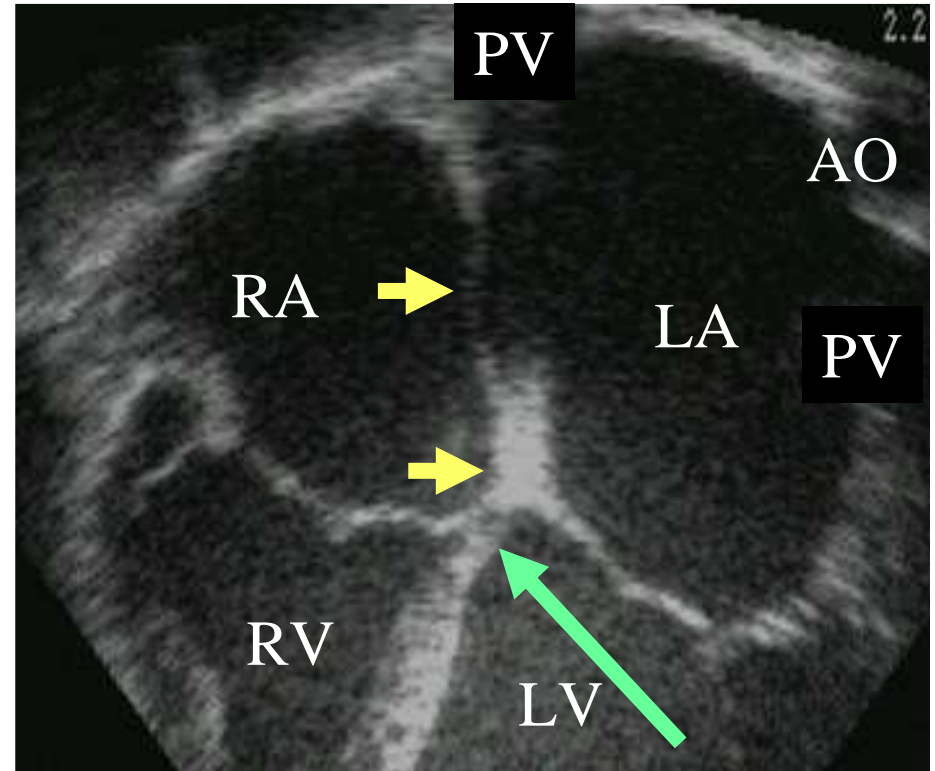
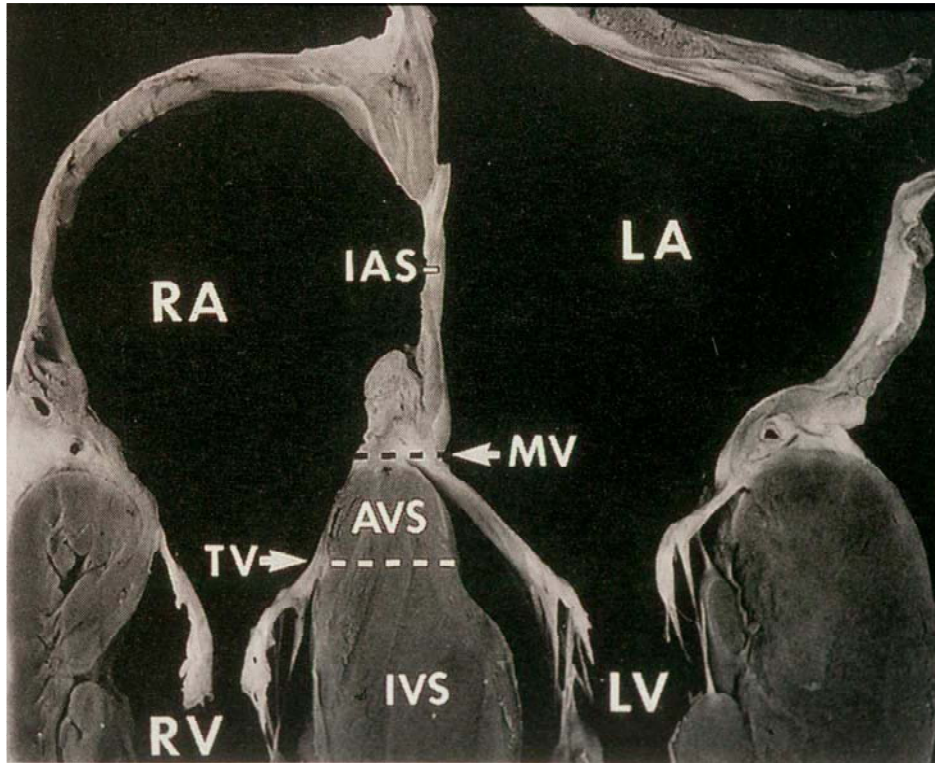
Right Hand Topology

Mirror-imagery



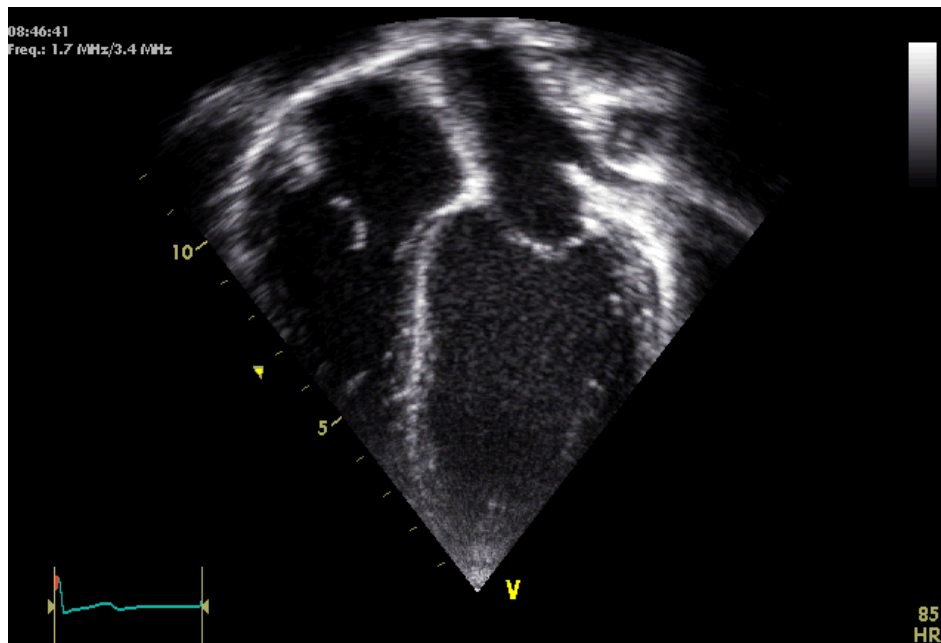
Left hand topology

Concordant Connections

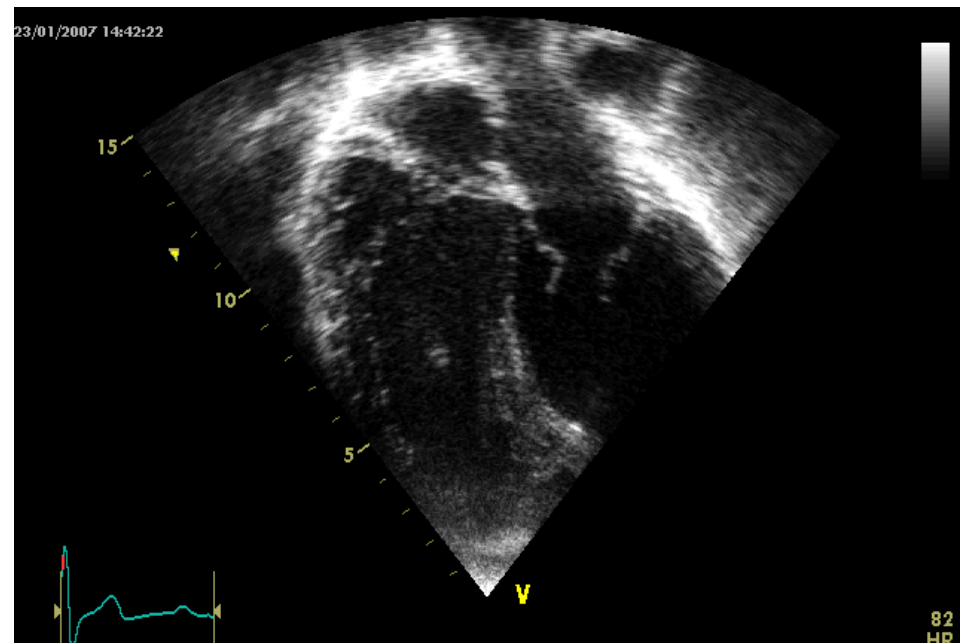


Offsetting

Concordant AV connection



Discordant AV connection

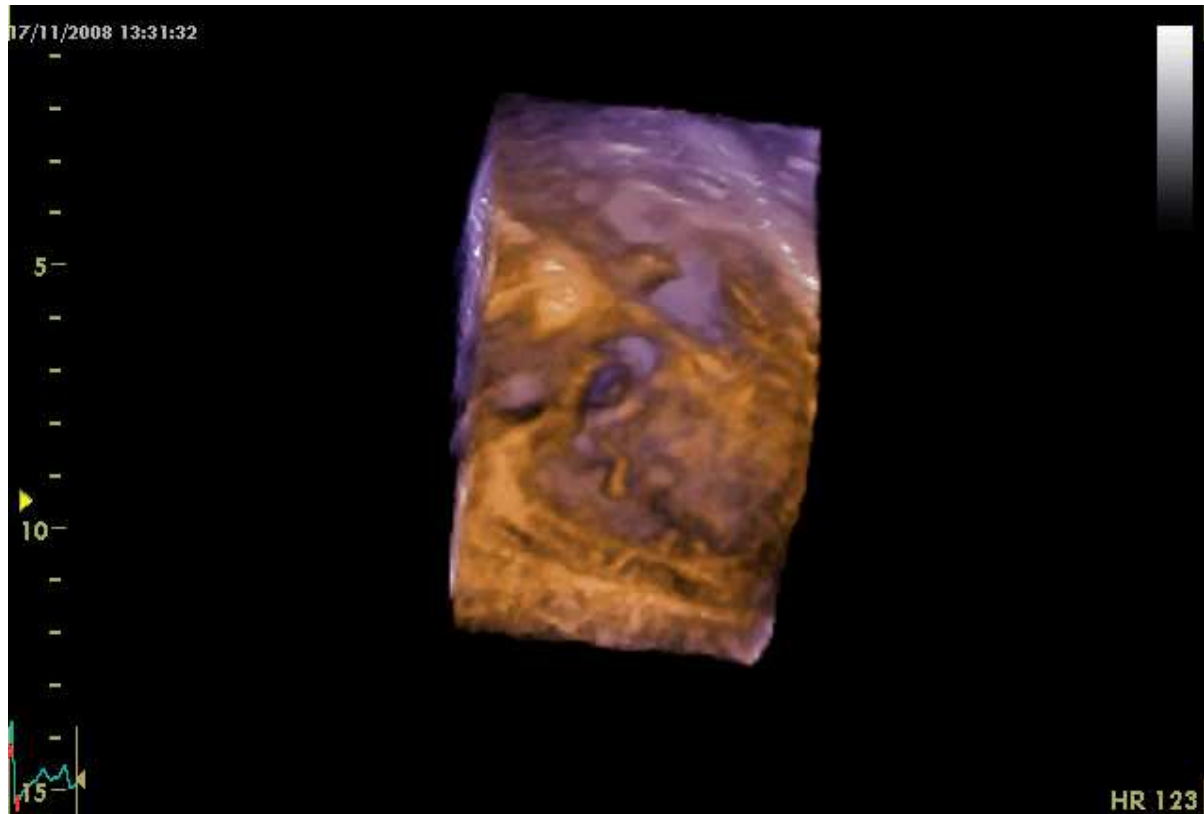


Discordant Connections

CCTGA

DORV/VI

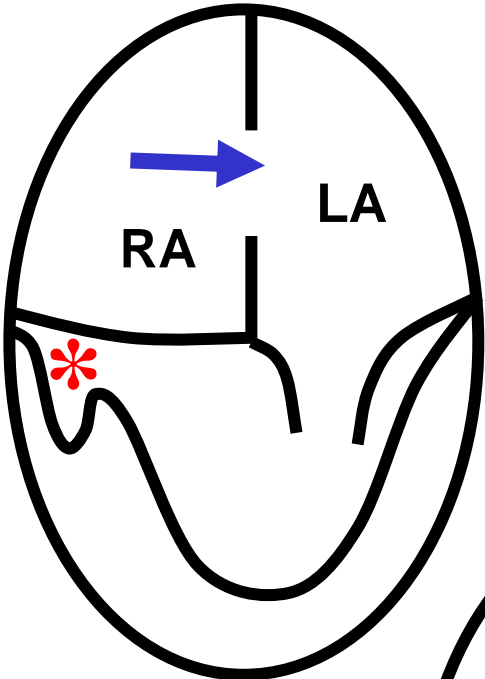
PA/VSD/VI



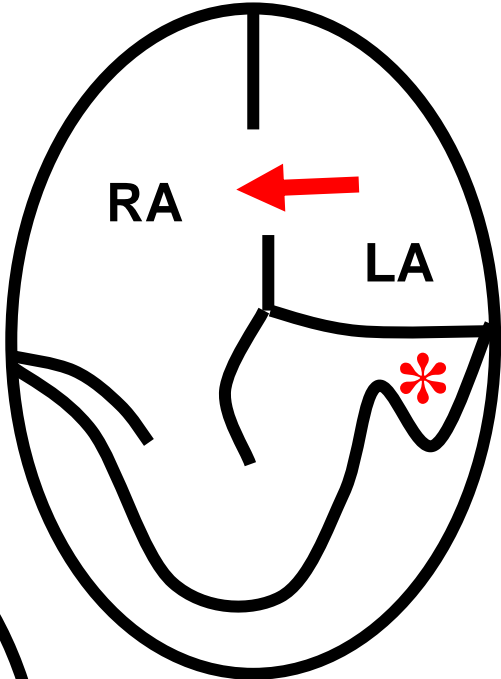
Univentricular AV Connections

- Absent AV connection (right / left)
 - Tricuspid / mitral atresia
- Common AV valve
 - With single ventricle
- Double inlet ventricle
 - Connection of both AV valve to the same ventricle:
 - Anatomically left
 - Anatomically right
 - Undetermined
 - (rudimentary chamber always present)

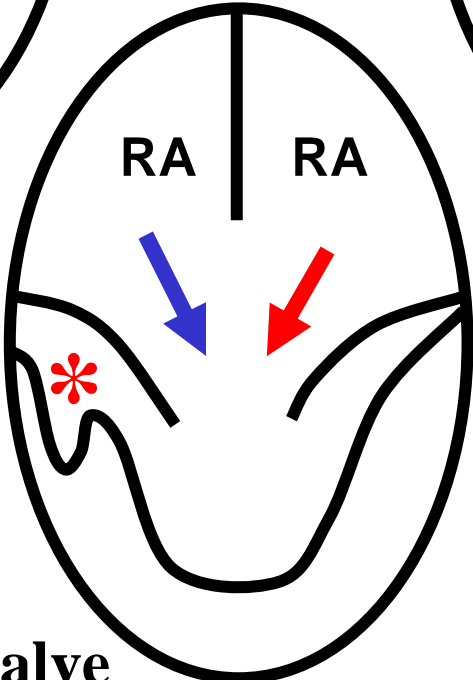
Univentricular AV connections



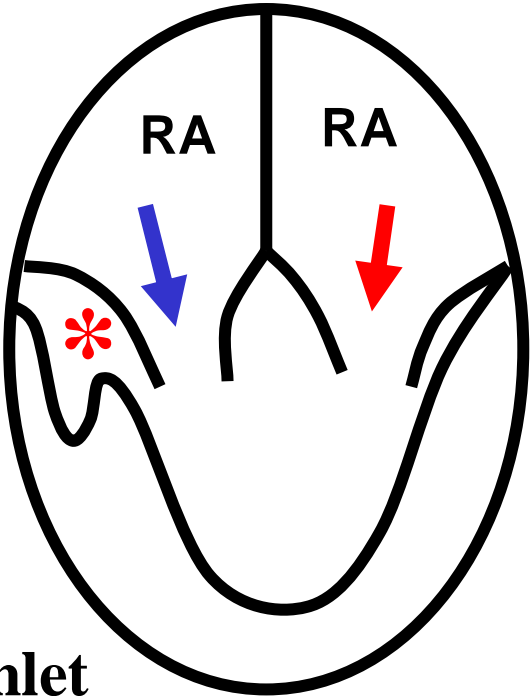
Absent right AV connection



Absent left AV connection

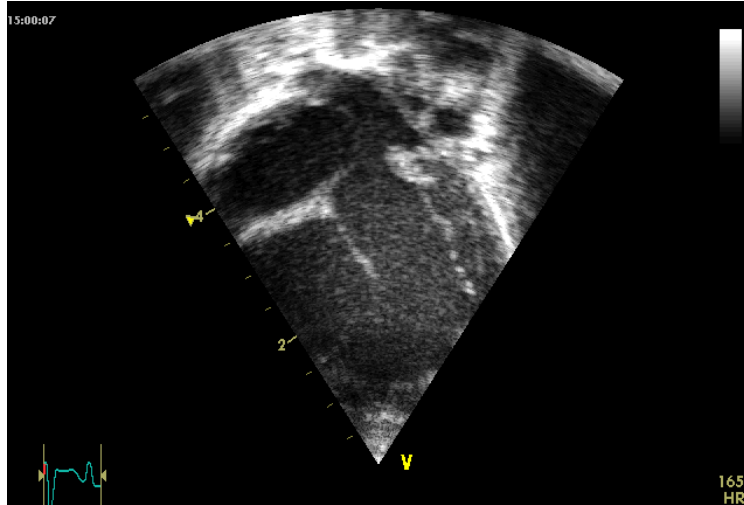


Common AV valve

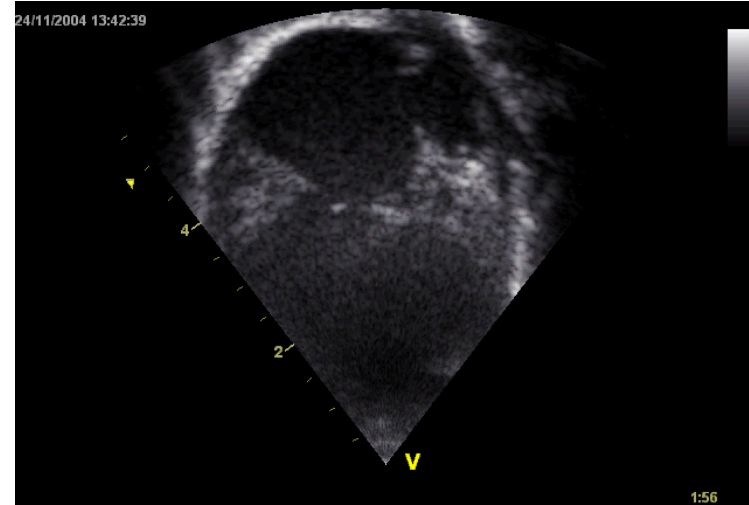


Double inlet

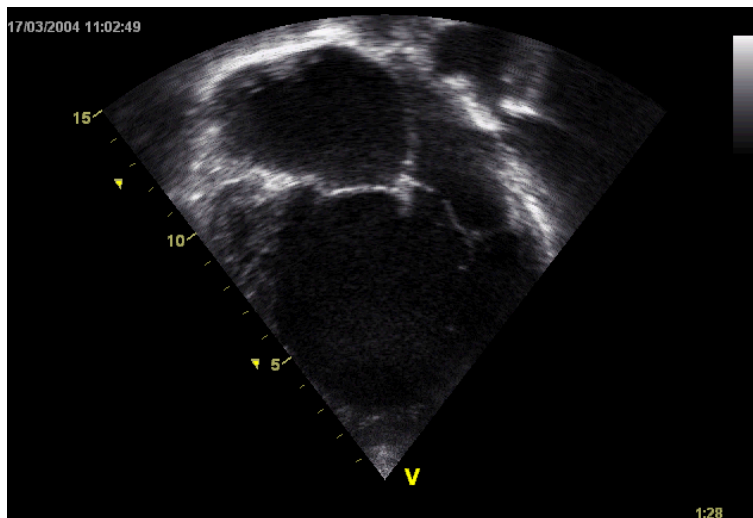
Absent right AV connection



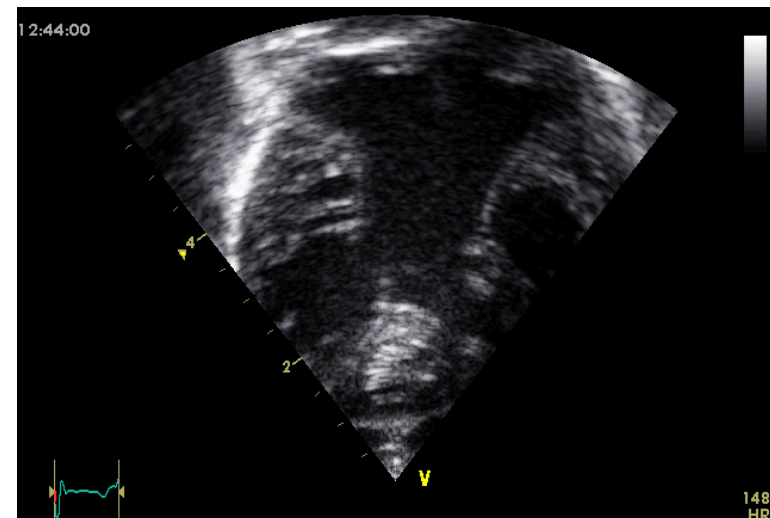
Absent left AV connection



Double inlet



Common AV valve



- Abdominal situs
- Atria - Atrial situs (systemic and pulmonary venous connections)
- AV connections
- Ventricles
- **VA connections**
- Aorta, PA trunk (AO arch and PA branches)

Ventriculo-arterial connections

- Concordant (=normal)

Ventricular septal defect, tetralogy of Fallot...

- Discordant

Transposition

- Double outlet ventricle

With subaortic VSD

subpulmonary VSD

doubly committed VSD

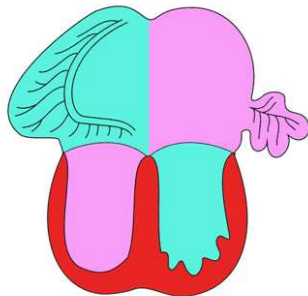
non-committed VSD

- Common arterial trunk

Discordant AV / VA connections

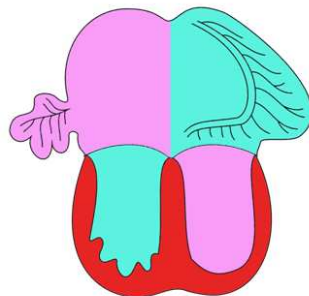
(AV & VA discordance = Congenitally corrected TGA)

Usual Atrial Arrangement

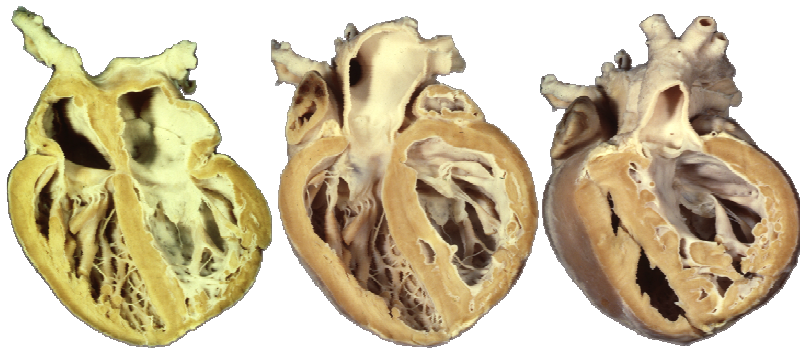


Left Hand topology

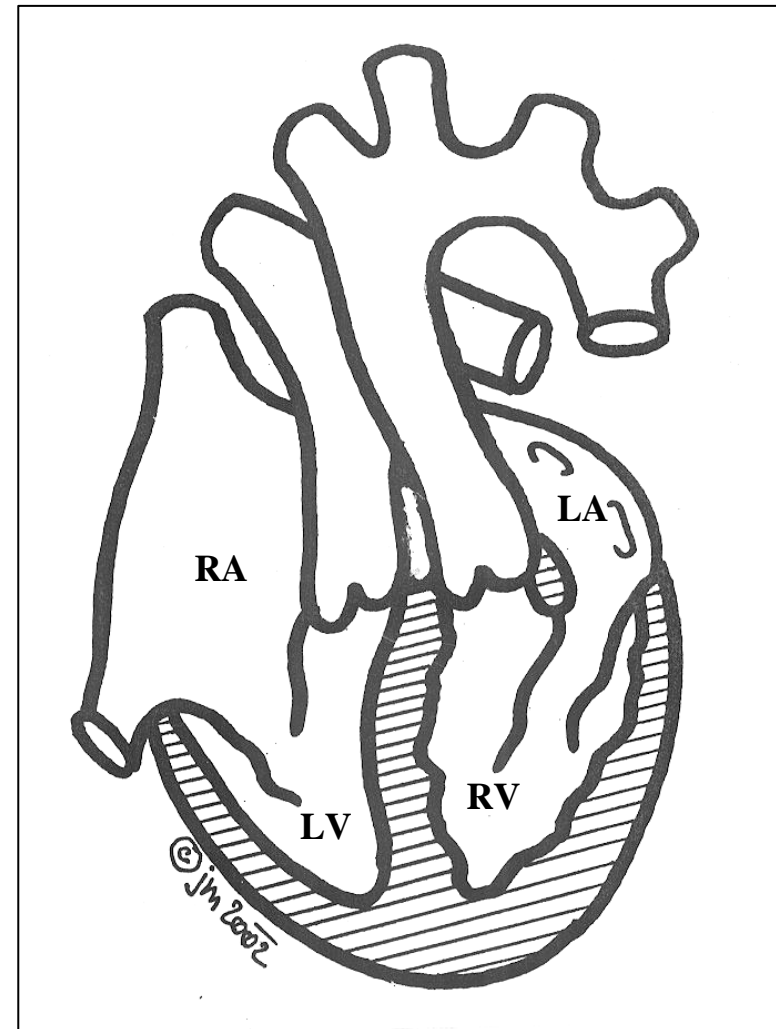
Mirror-imagery



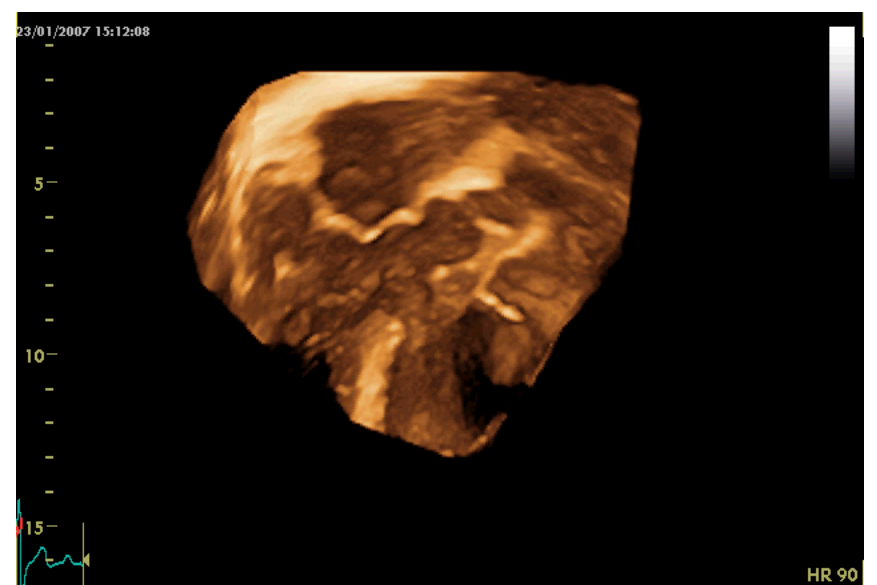
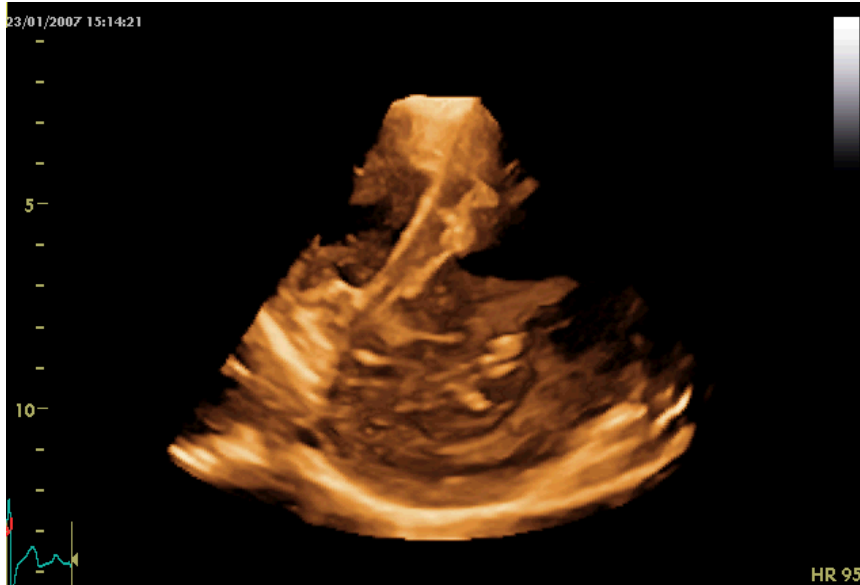
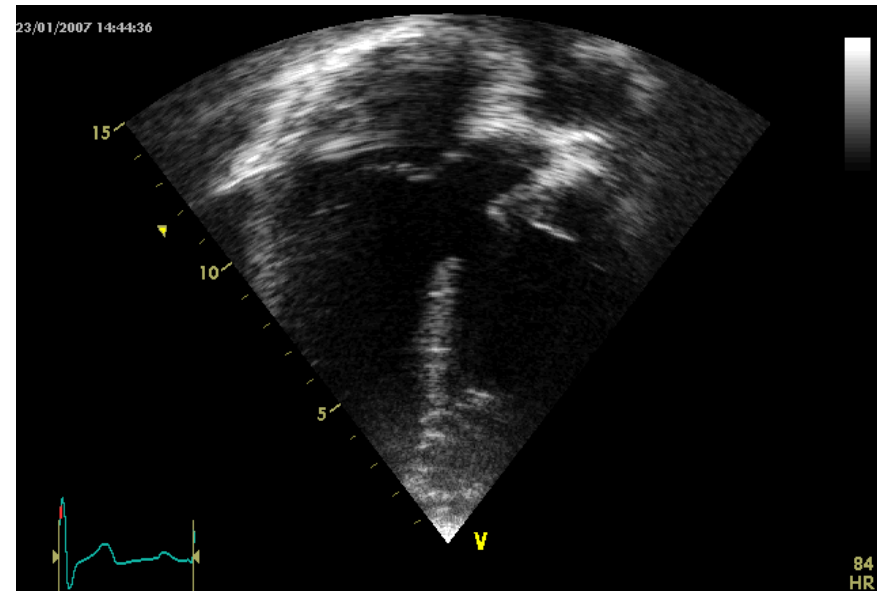
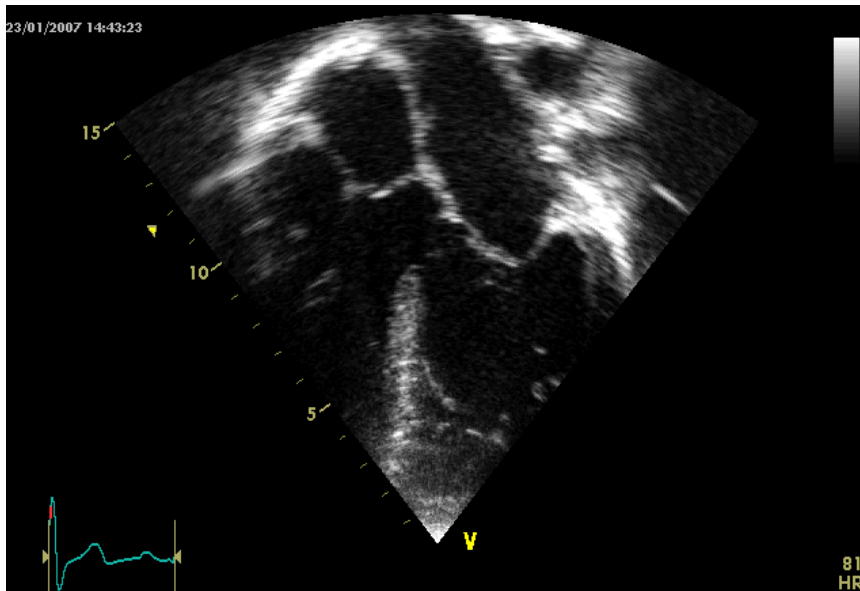
Right Hand Topology



Courtesy RH Anderson

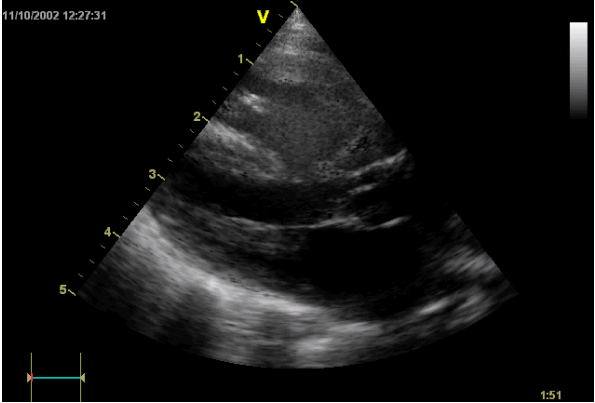


Discordant AV / VA connections



Overriding aortic valve: 50% rule

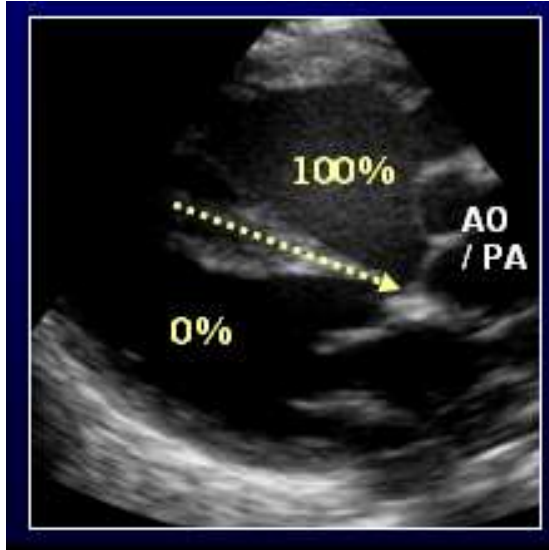
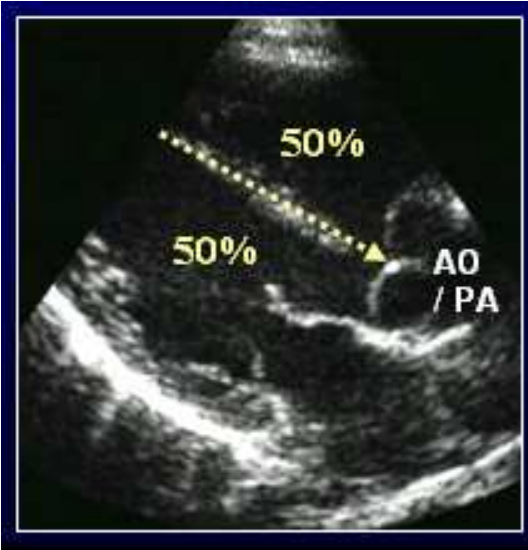
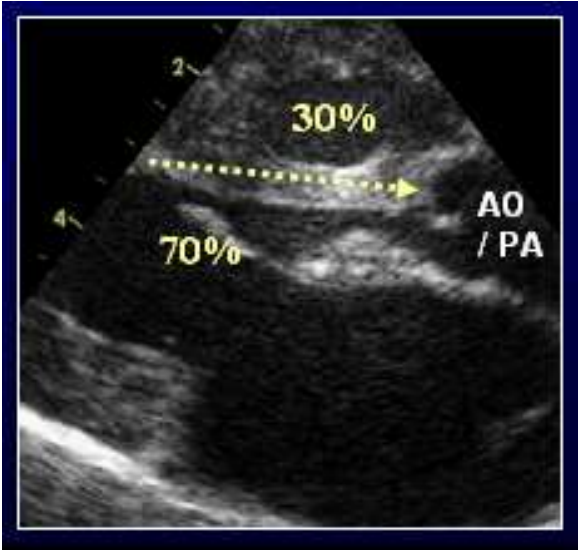
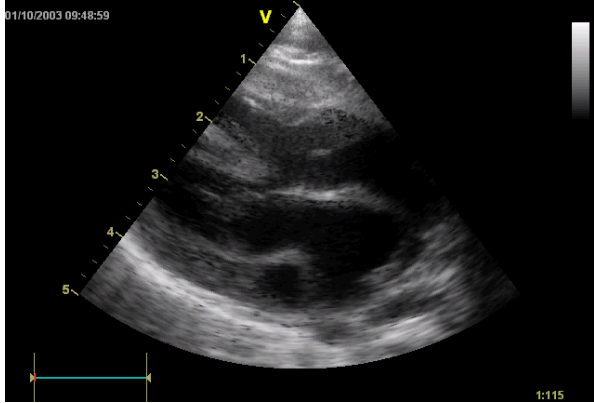
VSD



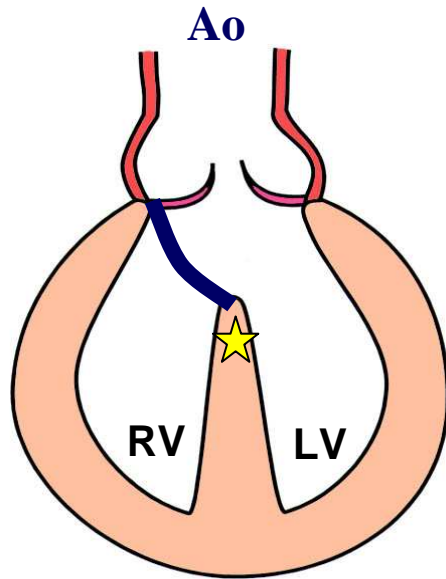
TOF



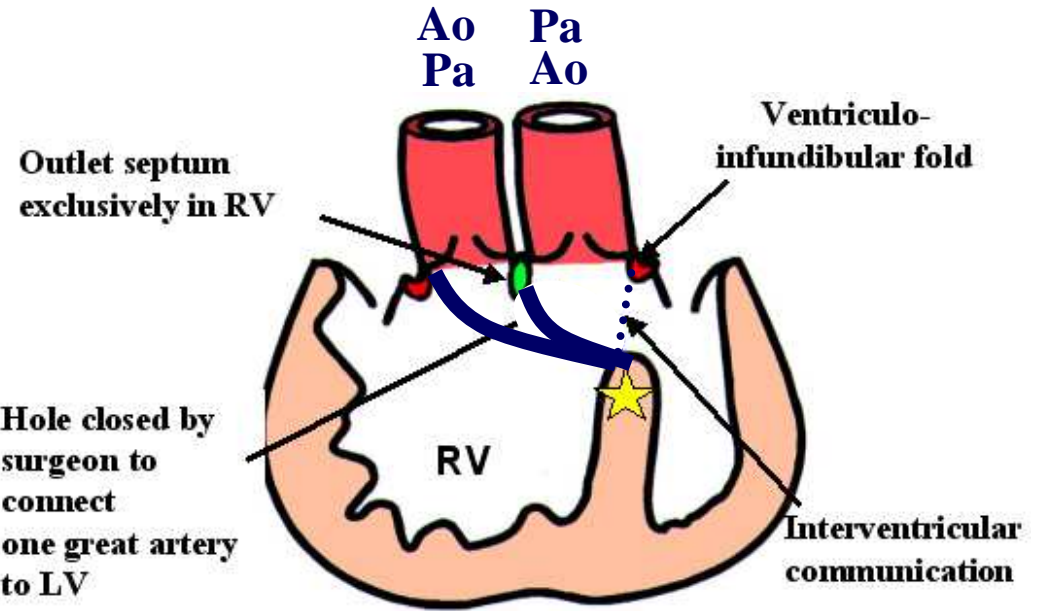
DORV



TOF vs Double outlet right ventricle

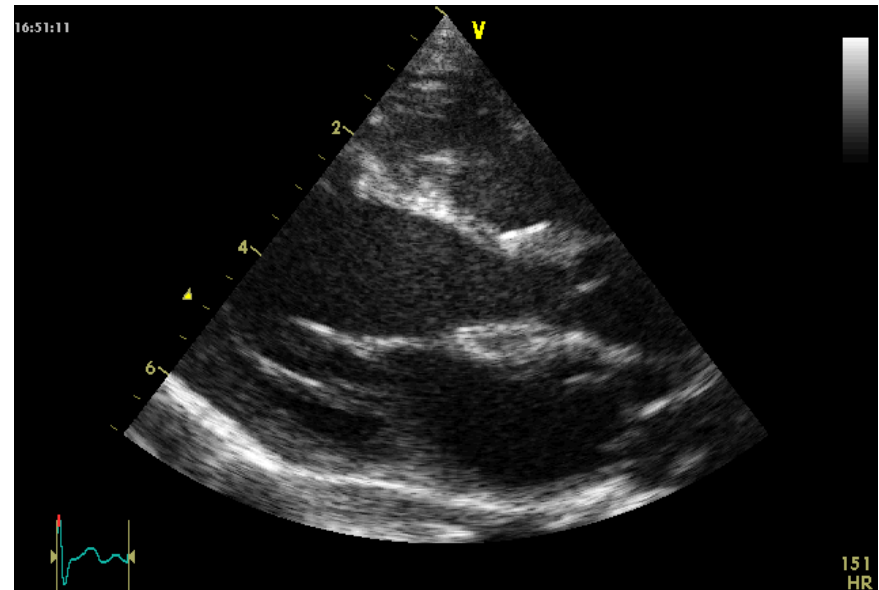
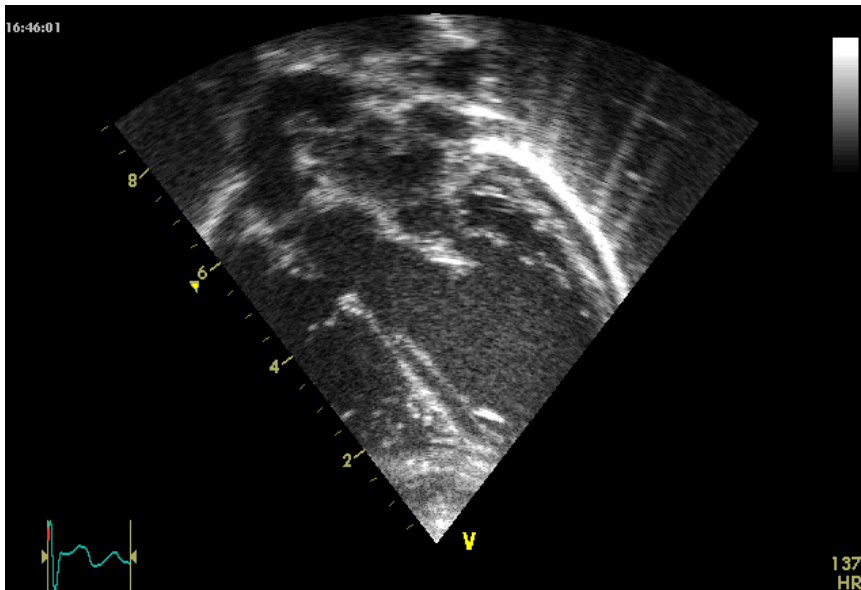


TOF



Double outlet right ventricle

Double conus = Double outlet right ventricle?



Double conus does not determine DORV!

How to diagnose

Correct definition of disease?

- Simple vs. Complex transposition?
- Transposition or Double outlet right ventricle?
- Transposition or Criss-cross heart
or Supero-inferior ventricles
- Transposition or Malposition (corrected or not, right or left?)

**AV concordance
&
VA discordance**

TGA

**Right/Left
ventricle
with
two outlets**

**DORV
DOLV**

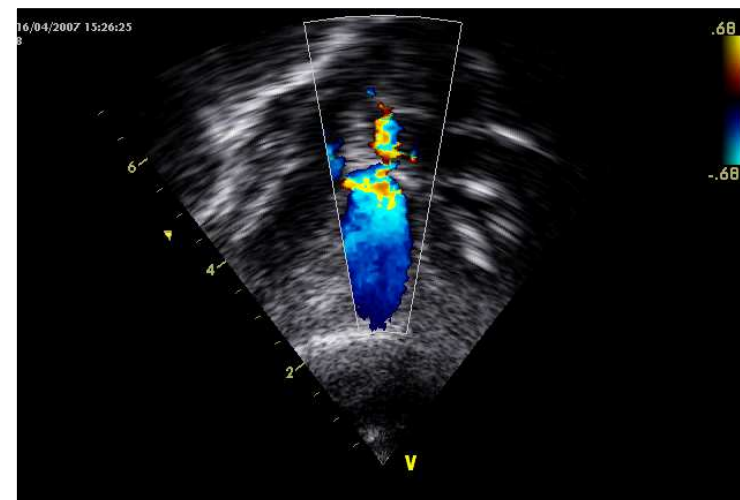
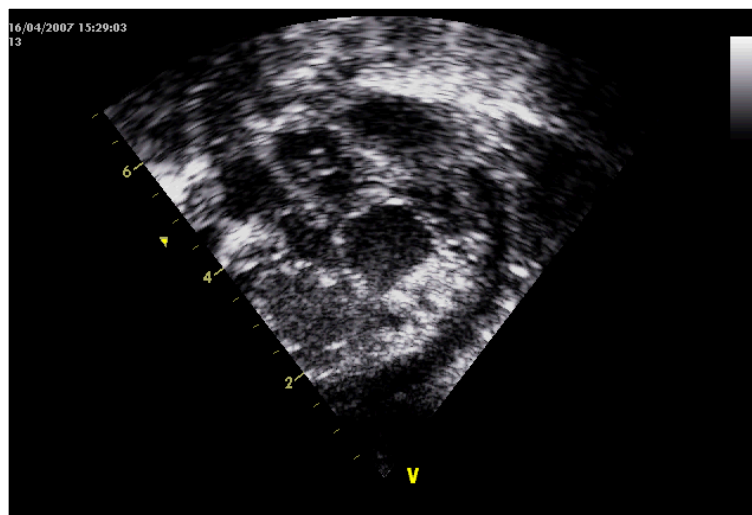
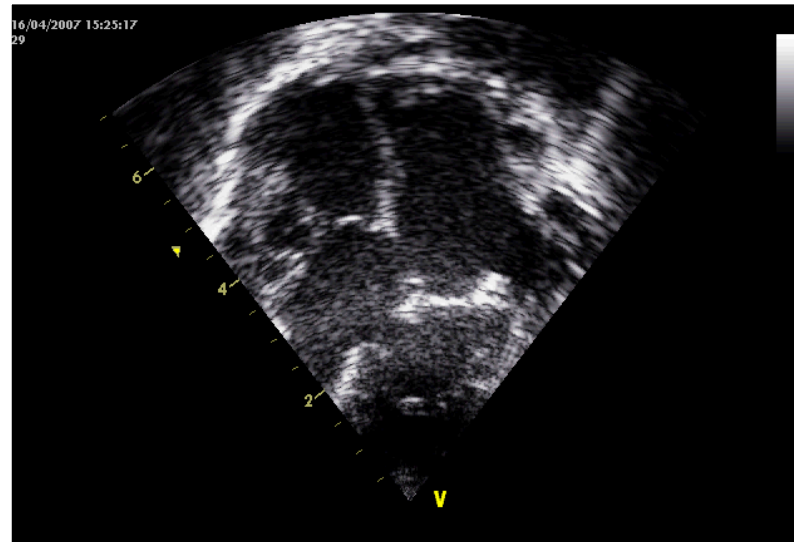
**AV discordance
&
VA discordance**

CCTGA

**AV discordance
&
VA concordance**

CCMGA

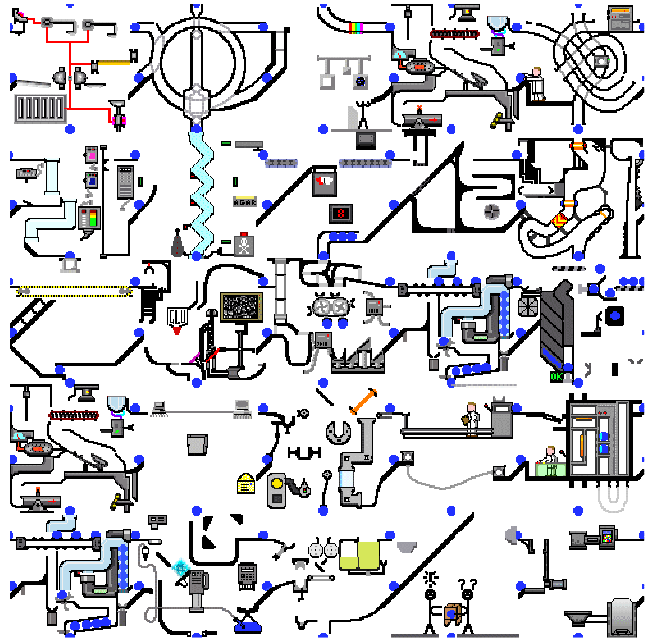
AV discordance & VA concordance (Congenitally corrected malposition)



Segmental approach

- Important to understand situs arrangement
- Provides complete coverage of lesion
- Minimises potential errors
- Easy for reading and interpretation

Sequential Segmental Approach



KISS

Keep
It
Simple,
Stupid

