Università di Pisa
Scuola di Specializzazione in Cardiologia

Seminario

Classificazione delle cardiopatie congenite

Sandra Giusti

Fondazione Gabriele Monasterio
Cardiologia Pediatrica
Ospedale del Cuore G.Pasquinucci, Massa

Peacock TB

On malformation of the human heart

London: John Churchill 1858

Abbot M.E.

Atlas of congenital cardiac disease

New York: American Heart Association, 1936

Shinebourne E.A., MaCartney F.J., Anderson R.H.

Sequential chamber localization: the logical approach to the diagnosis in congenital heart disease

Br Heart J, 1976; 38: 327-340

Sequential segmantal analysis

ATRIA

- **SITUS AND VENOUS CONNECTION**

VENTRICLES

- **A-V CONNECTION**

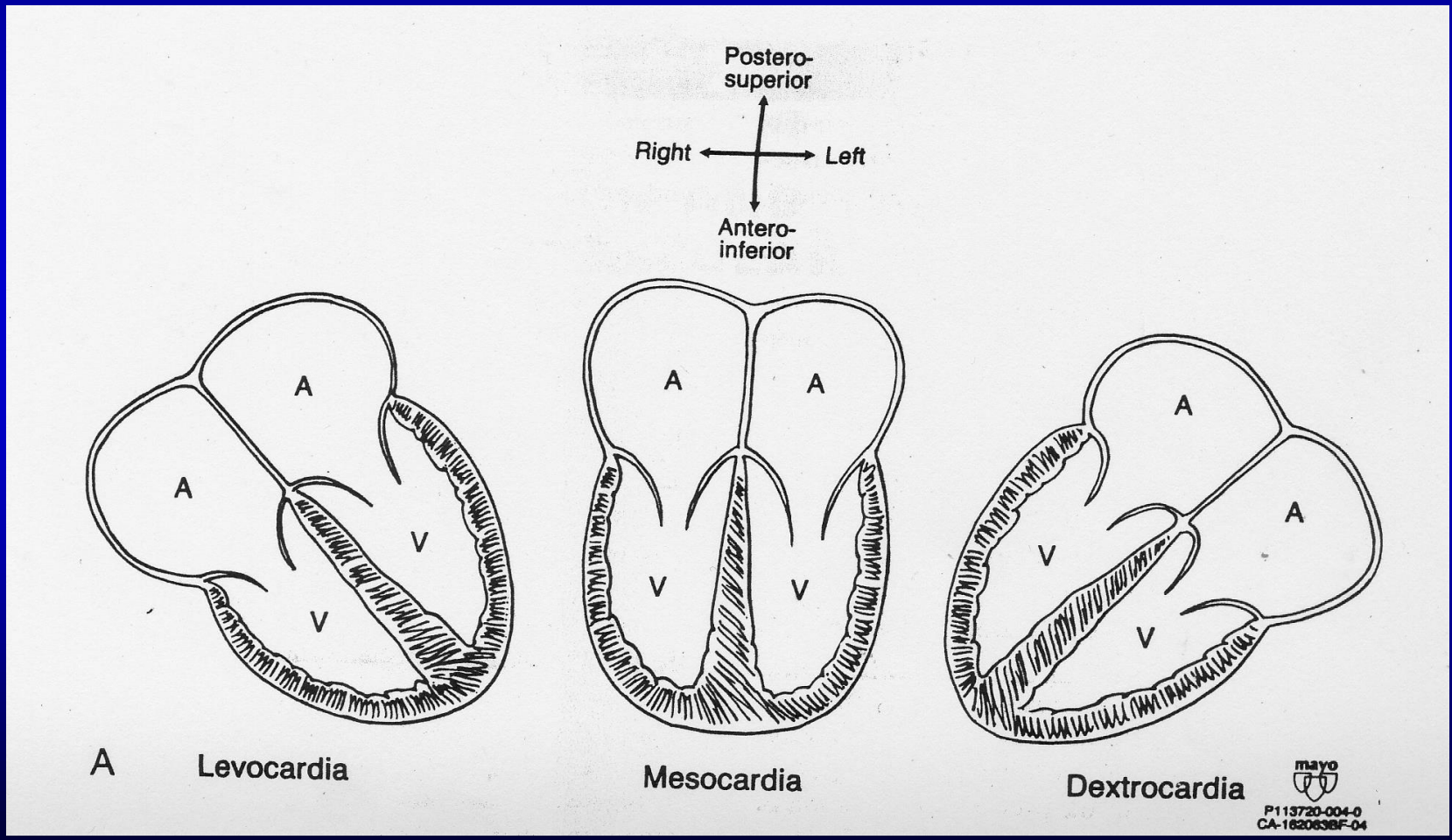
GREAT ARTERIES

- **V-A CONNECTION**

Cardiac position

- Left-sided
- Right-sided
- Midline
- Ectopia cordis (cervical, thoracic, abdominal)

Orientation in the chest



Visceral sidedness (situs)

- Cardiovascular
- Respiratory
- Digestive

situs solitus

situs inversus

situs ambiguus ***(isomeric or indeterminate)***

Isomerism

- Right isomerism = bilateral right sidedness

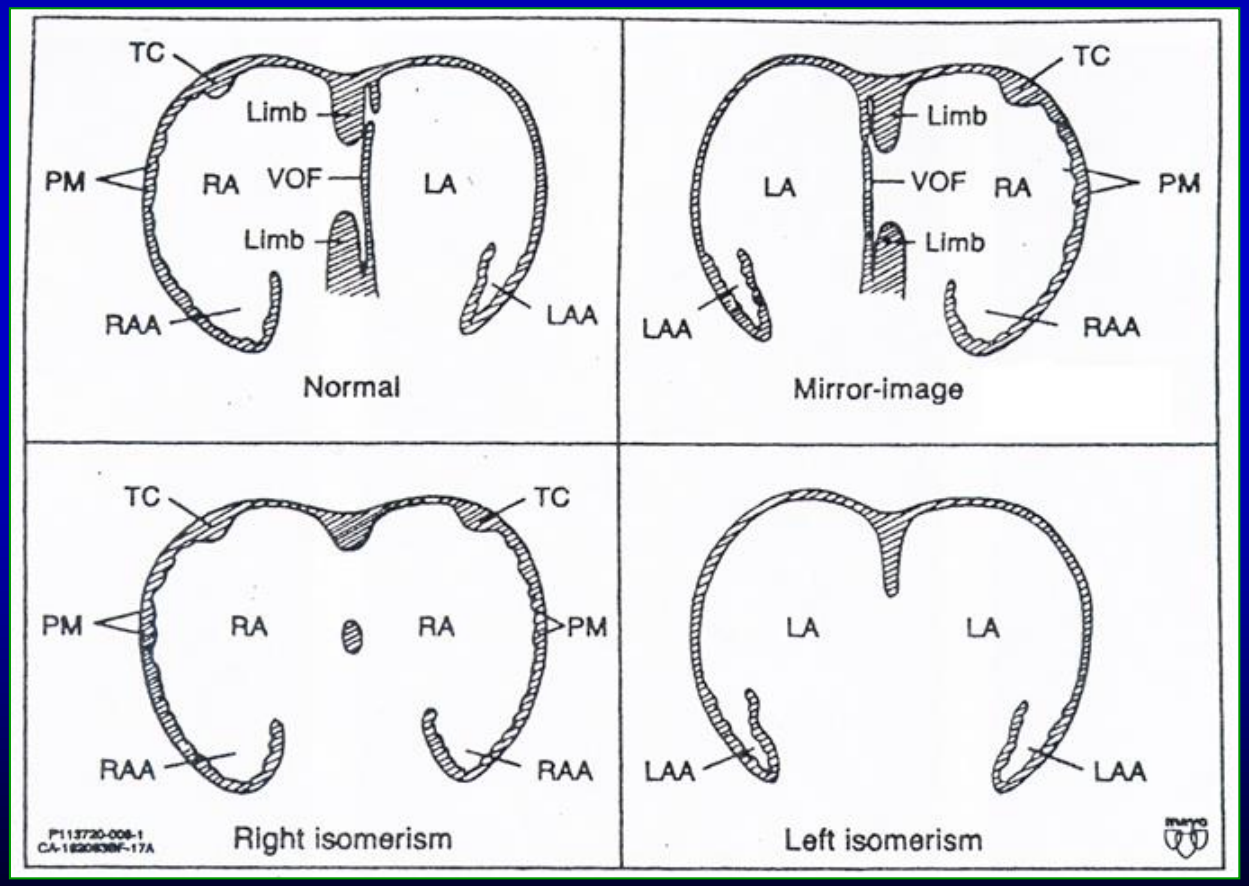
  asplenia

- Left isomerism = bilateral left sidedness

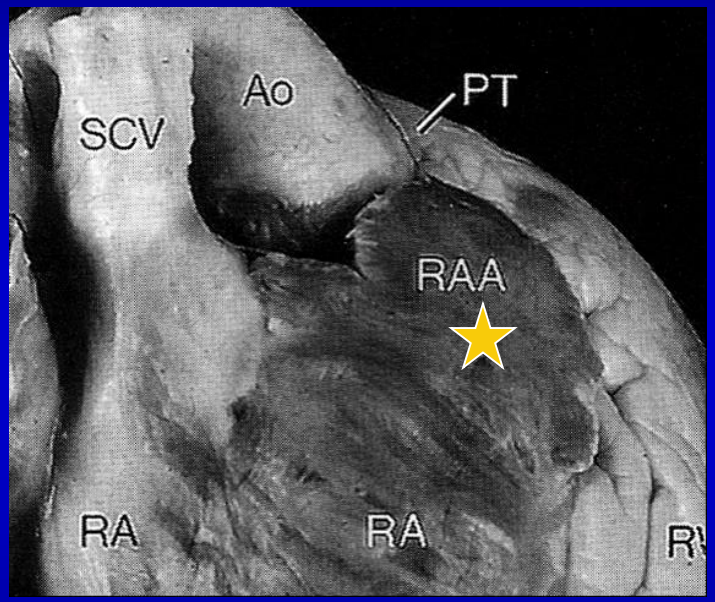
  polysplenia

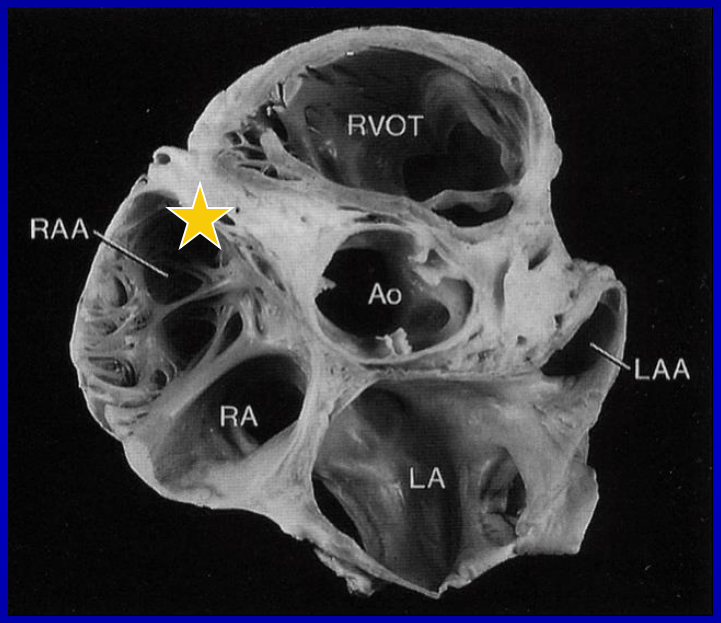
Cardiac sidedness

Position of the morphologic right atrium



Morphology of the right atrium



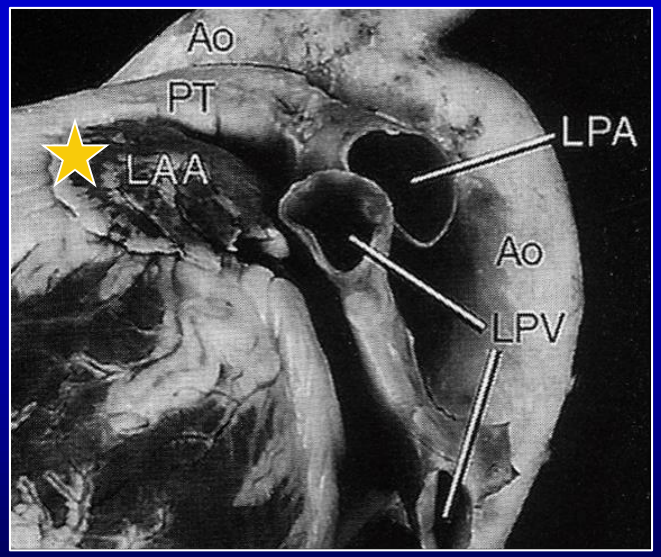


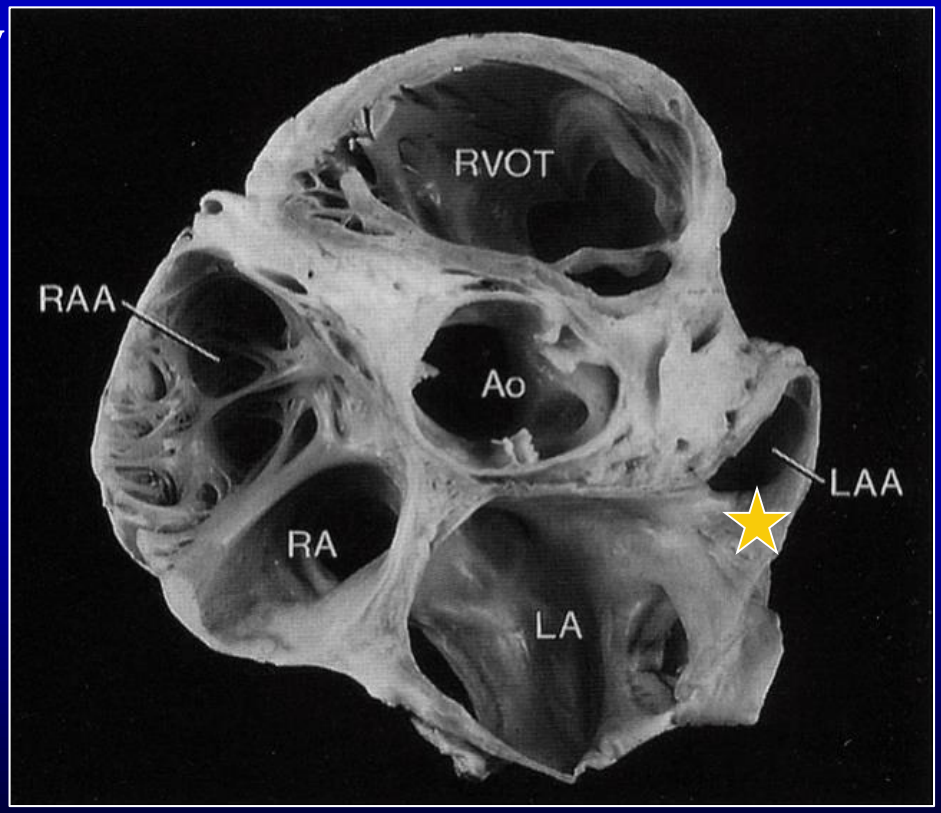
- triangular shape
- large communication with atrial cavity

Morphology of the left atrium

finger-like

narrow communication with atrial cavity

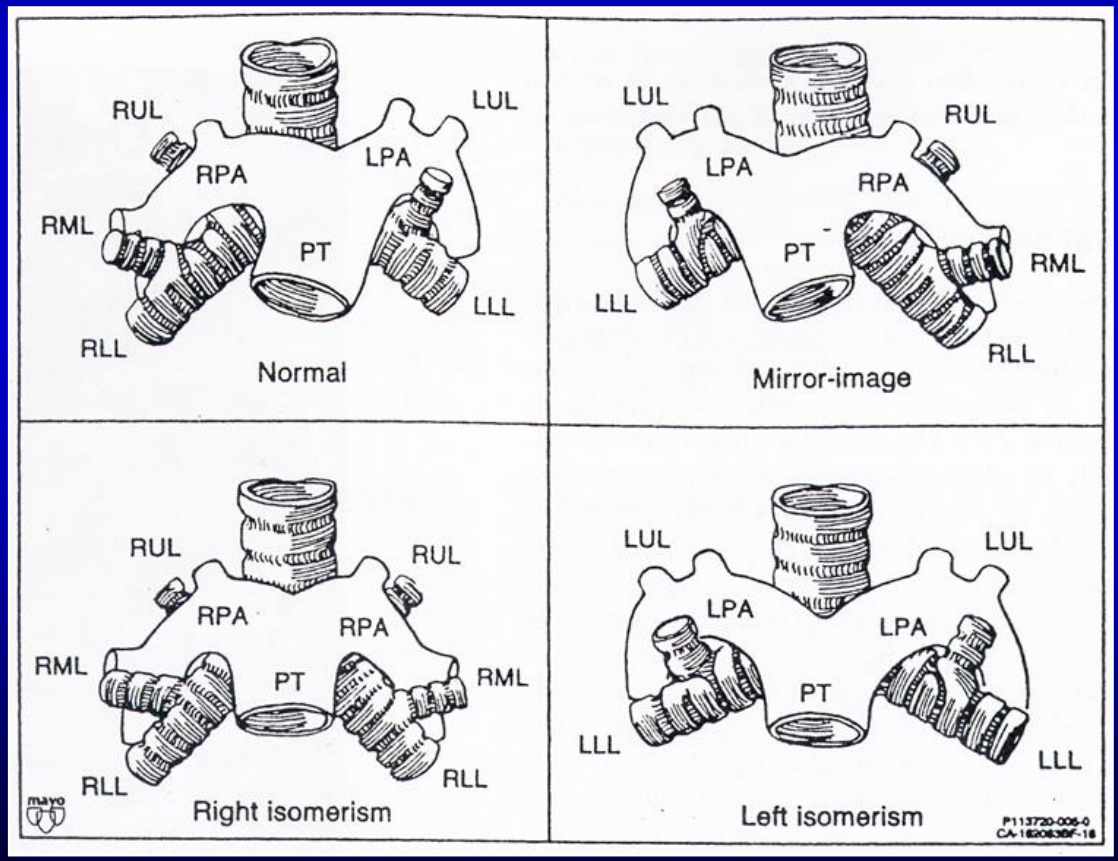




Isomerism

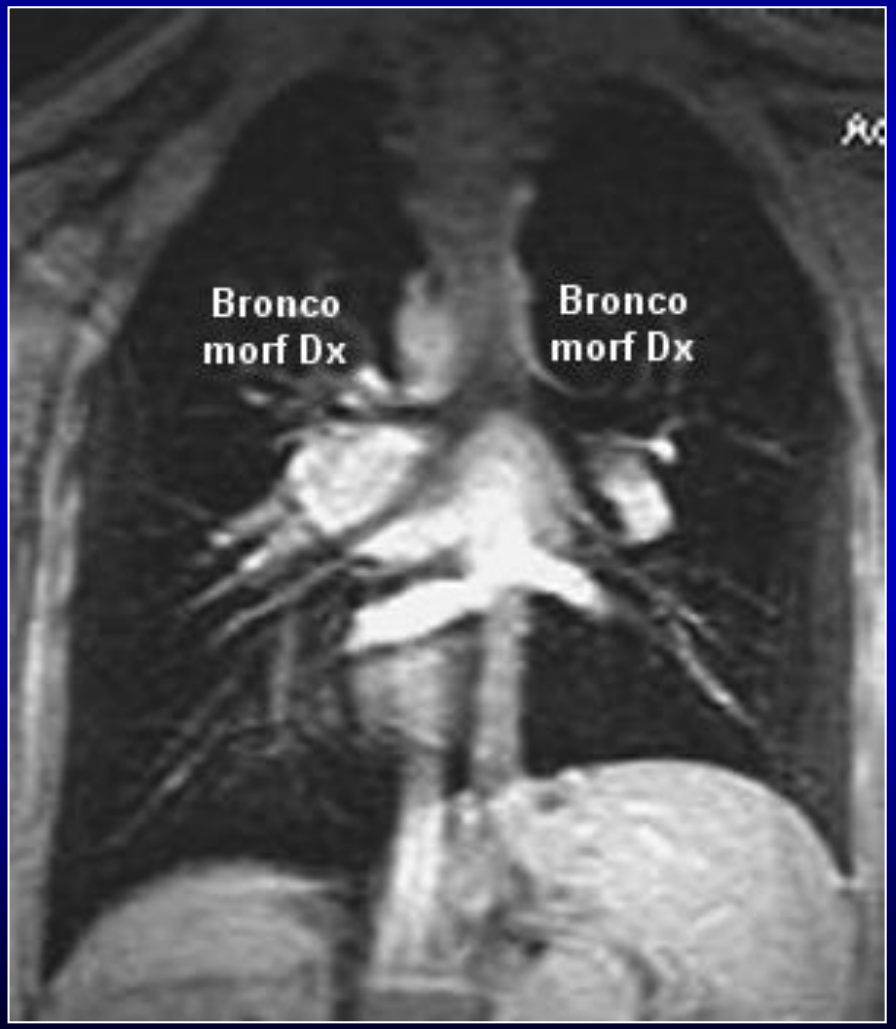
Pulmonary sidedness

Positions of the morphologic right and left lungs



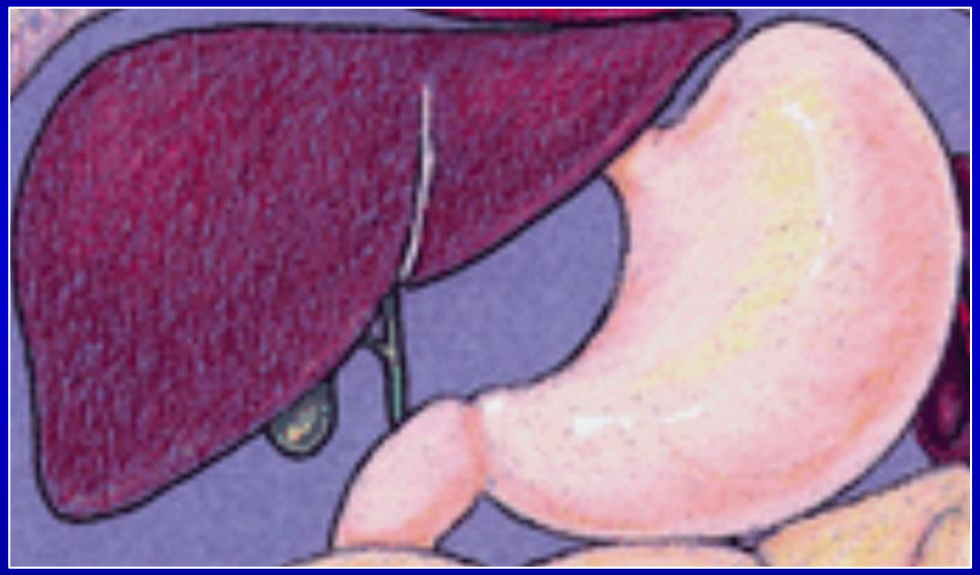
Isomerism

Isomerism



Abdominal sidedness

location of the liver and the stomach



Spleen (if present) and pancreas generally on the same side of the vertebral column as the stomach

Situs assessment

Atriovisceral situs

DETERMINED

SOLITUS

INVERSUS

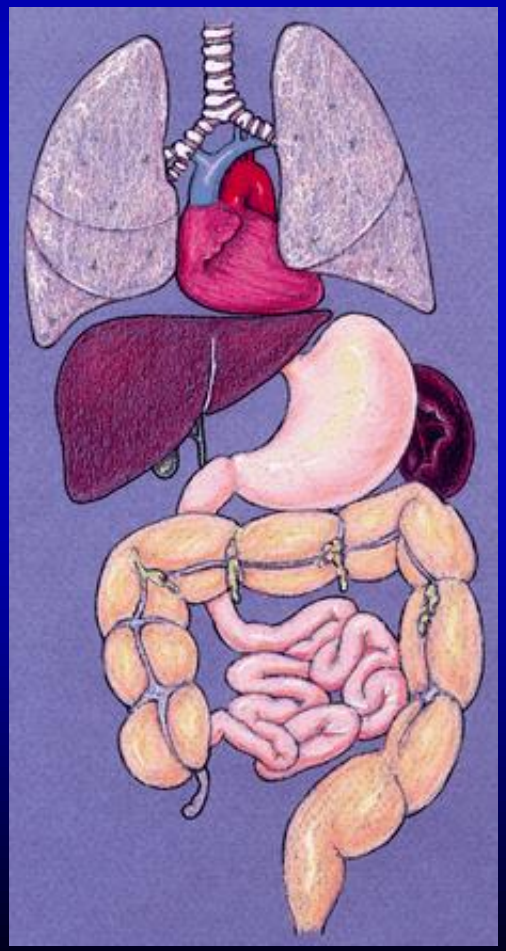
(mirror image)

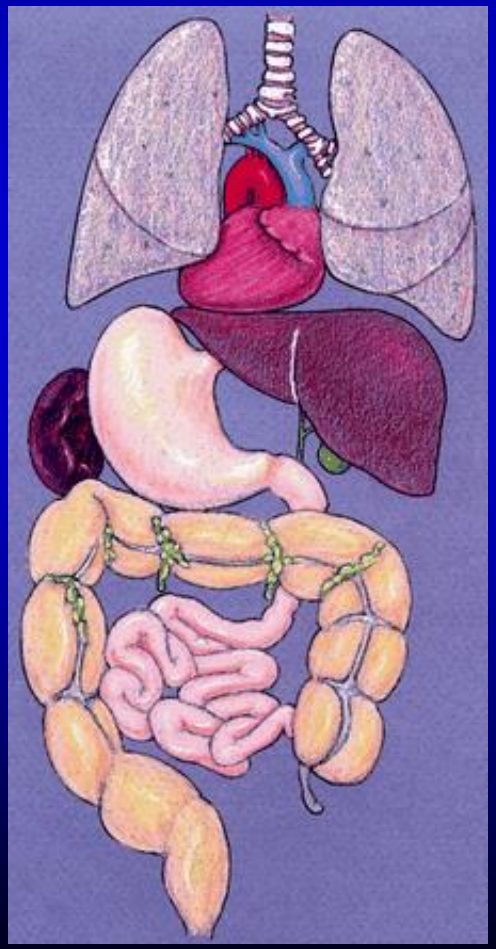
UNDETERMINED

(HETEROTAXY / SITUS AMBIGUUS)

Determined Atriovisceral Situs

SITUS SOLITUS	SITUS INVERSUS



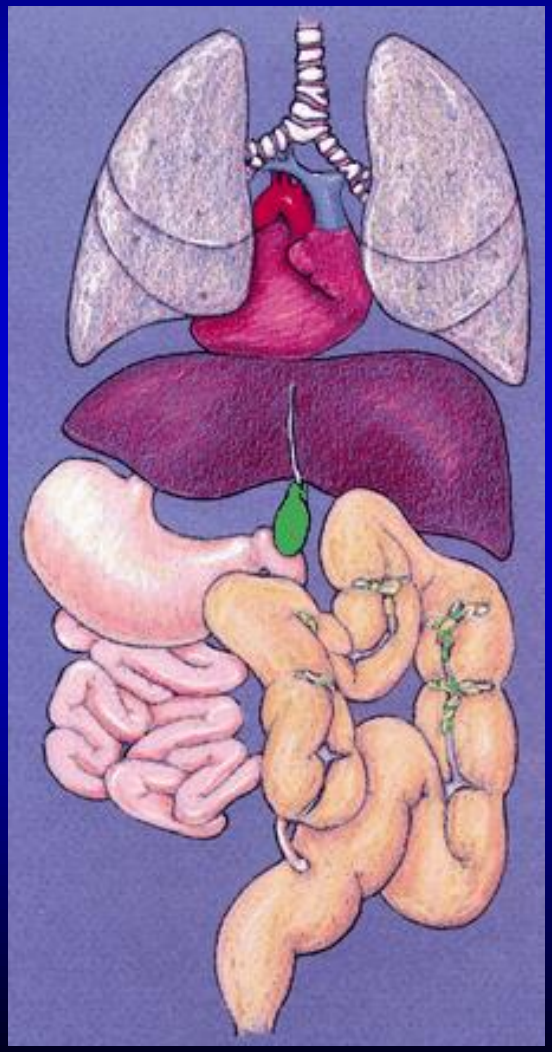


Right isomerism

2 R bronchi

2 R atria

Midline liver



Absent spleen

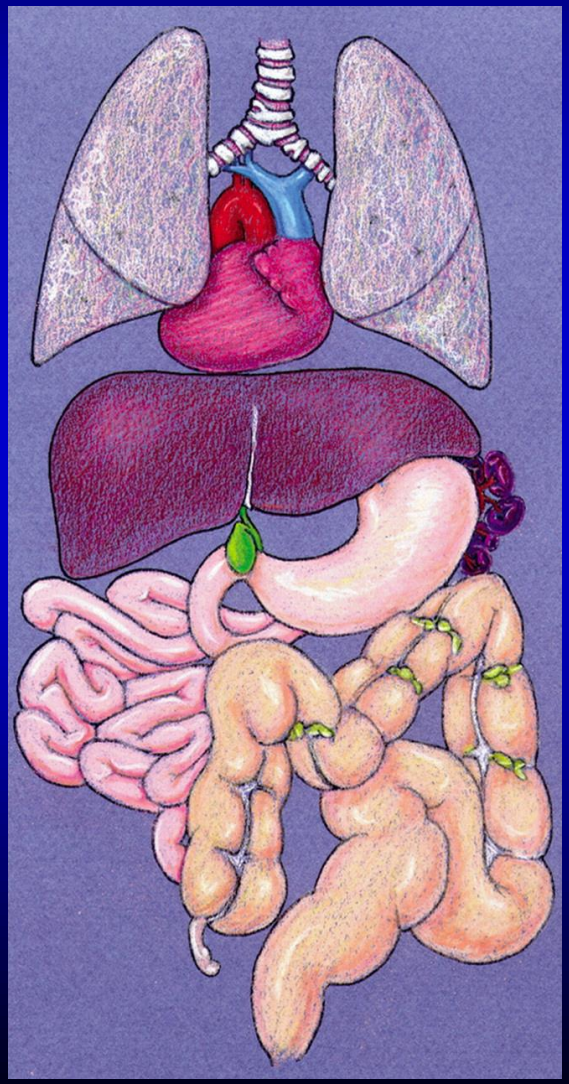
asplenia syndrome

Left isomerism

2 L bronchi

2 L atria

Sidedness of abdominal viscera indeterminate, mirror-image or normal



Supranumerary spleens

→ ***polysplenia syndrome***

Venous connections

SYSTEMIC VENOUS connection

Superior vena cava (SVC)

↓

MORPH. R ATRIUM

↑

Inferior vena cava (IVC)

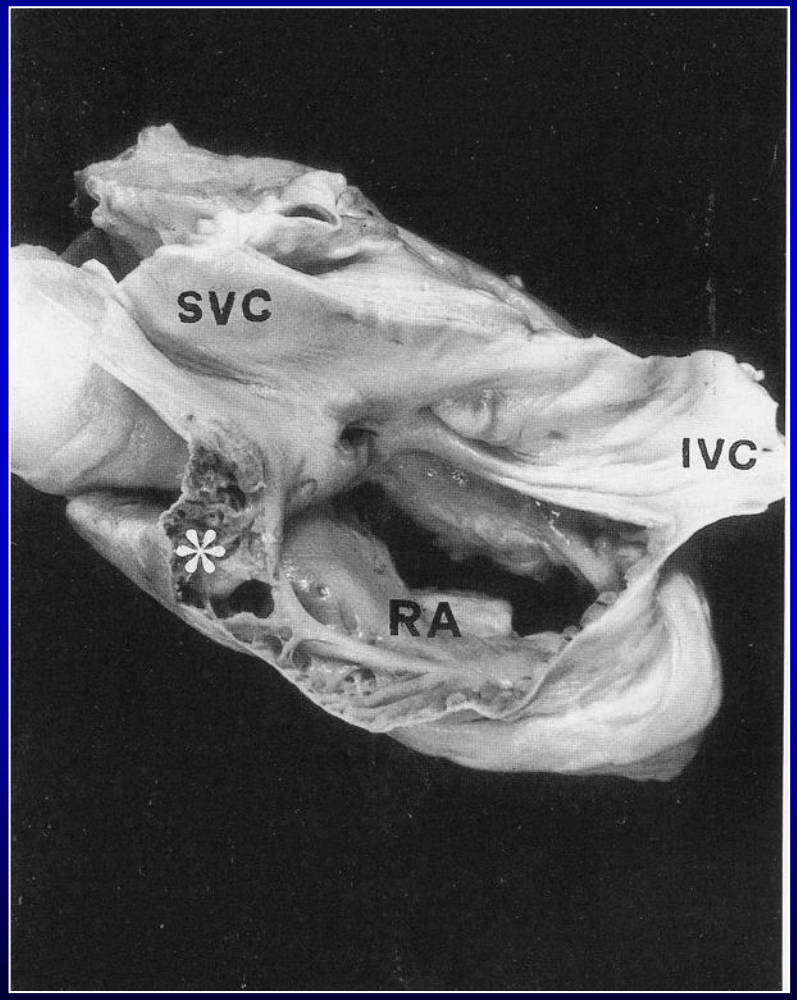
PULMONARY VENOUS connection

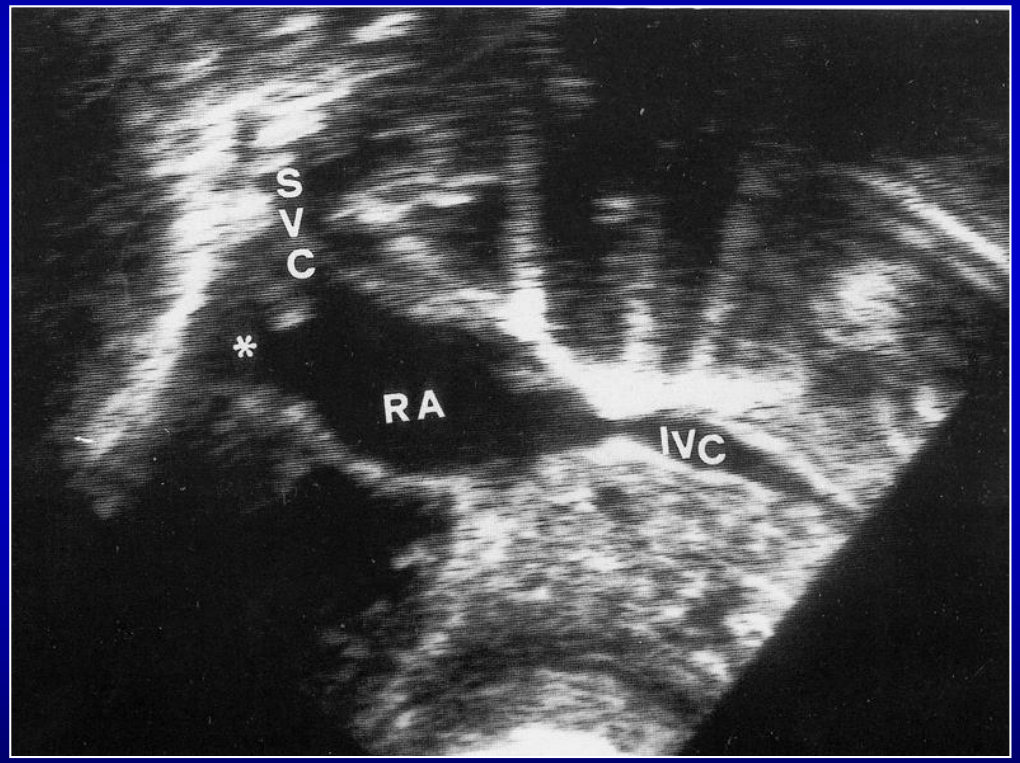
4 pulmonary veins

↓

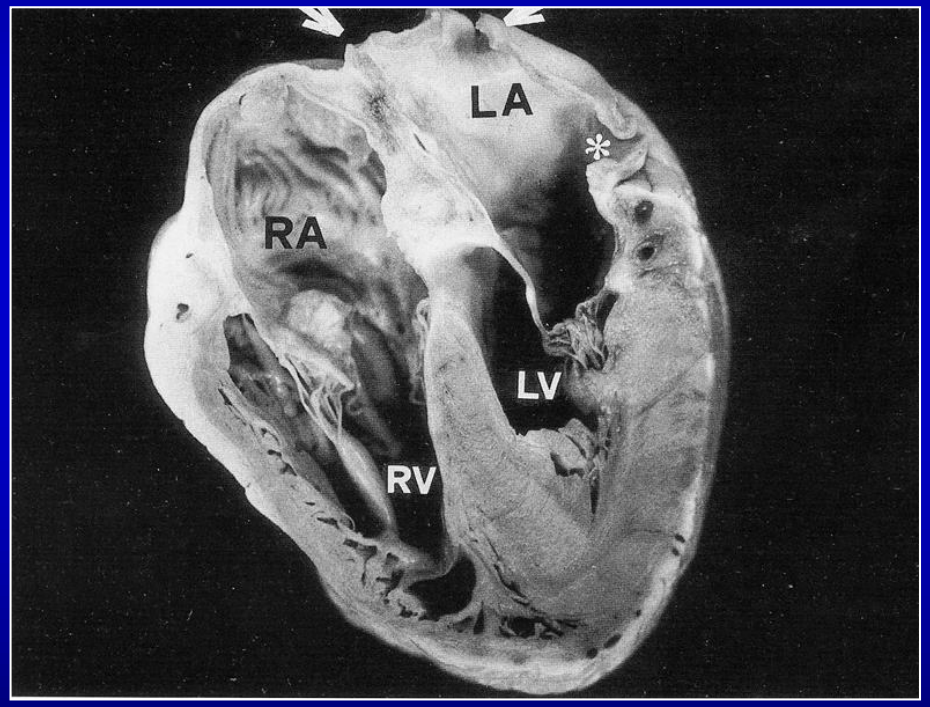
MORPH. L ATRIUM

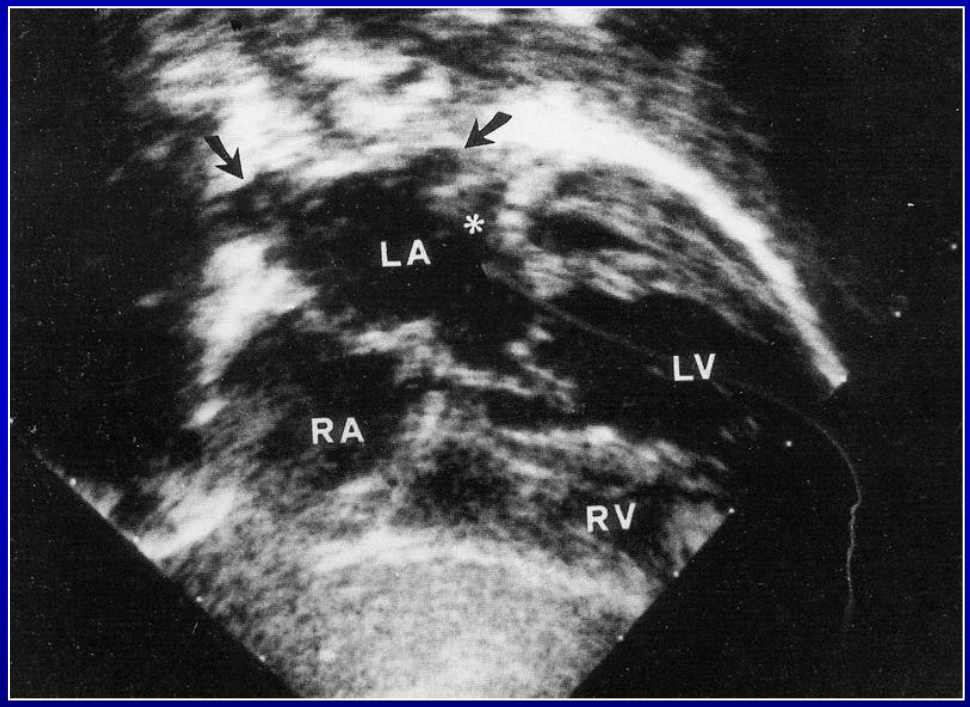
Venous connections





Venous connections



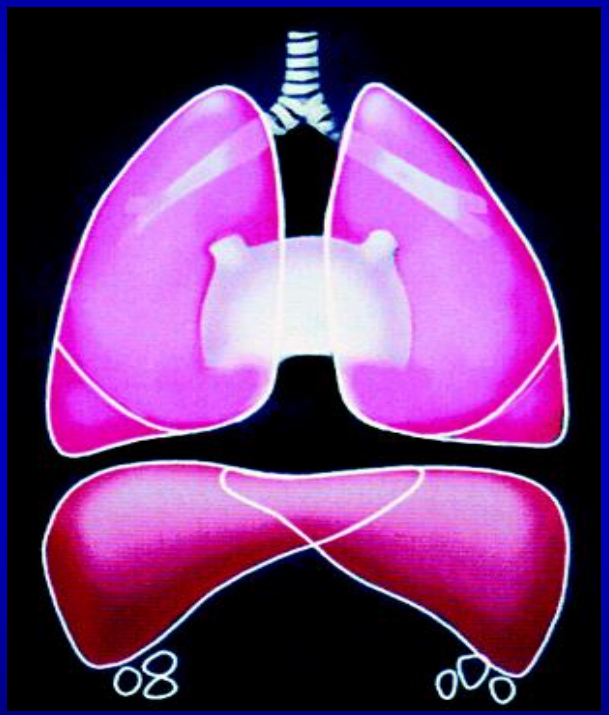


Venous connections

RIGHT ISOMERISM

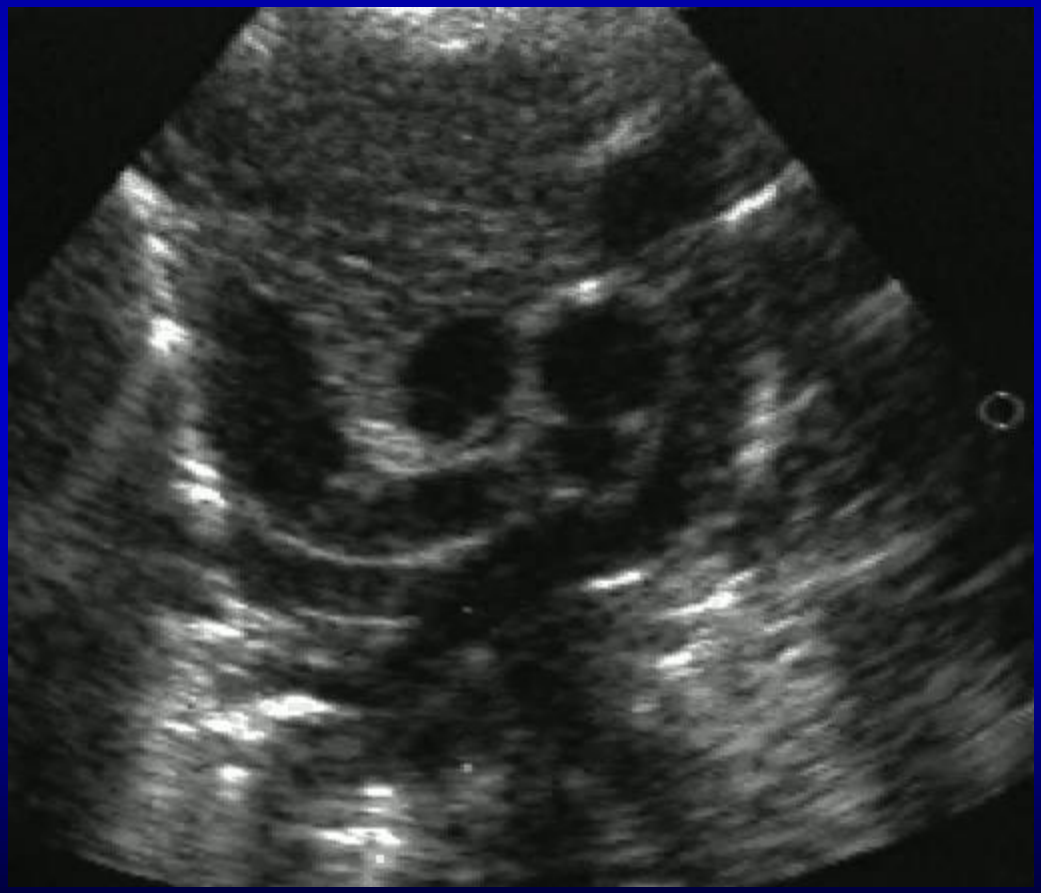
- IVC and hepatic veins drain separately
- 2 superior caval veins
- TAPVD

LEFT ISOMERISM



- Interruption of suprahepatic tract of IVC and azygos continuation
- Pulmonary venous drainage into both atria

Anomalous venous connection

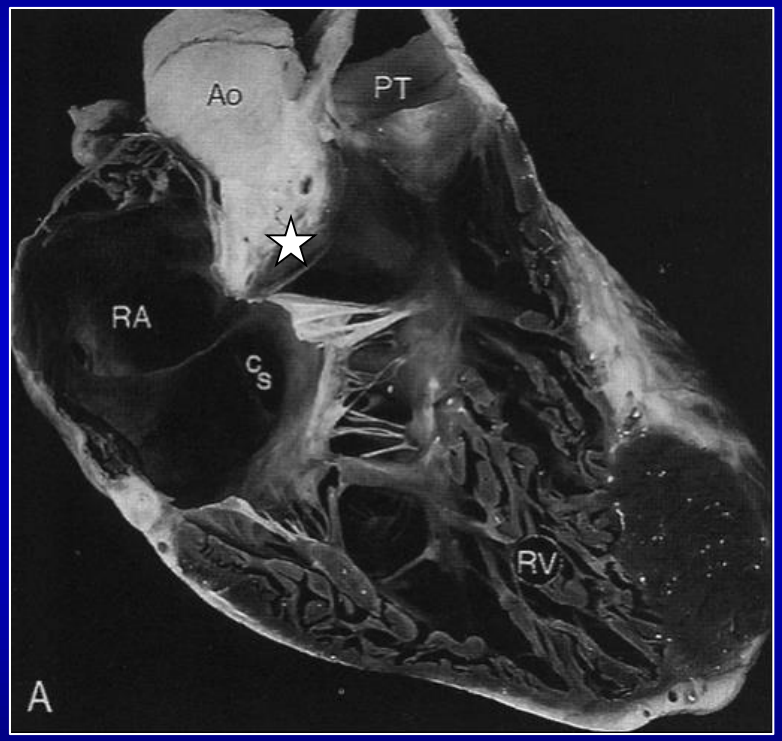


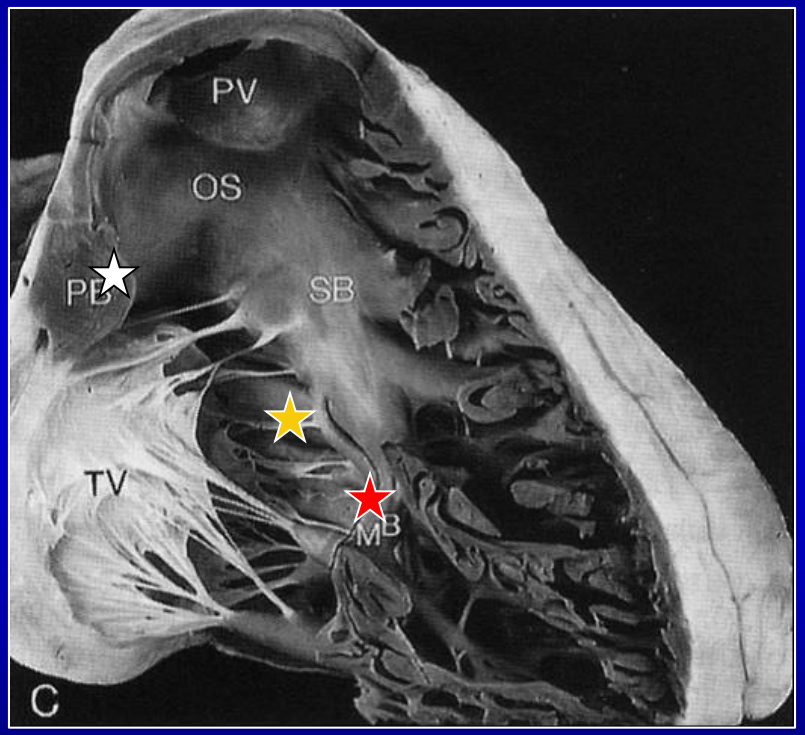
Anomalous venous connection

Atrioventricular connections

- Biventricular :
  - Concordant
  - Discordant
  - Ambiguous
- Univentricular:
  - Double inlet
  - Single inlet
  - Common inlet

Morphology of the right ventricle





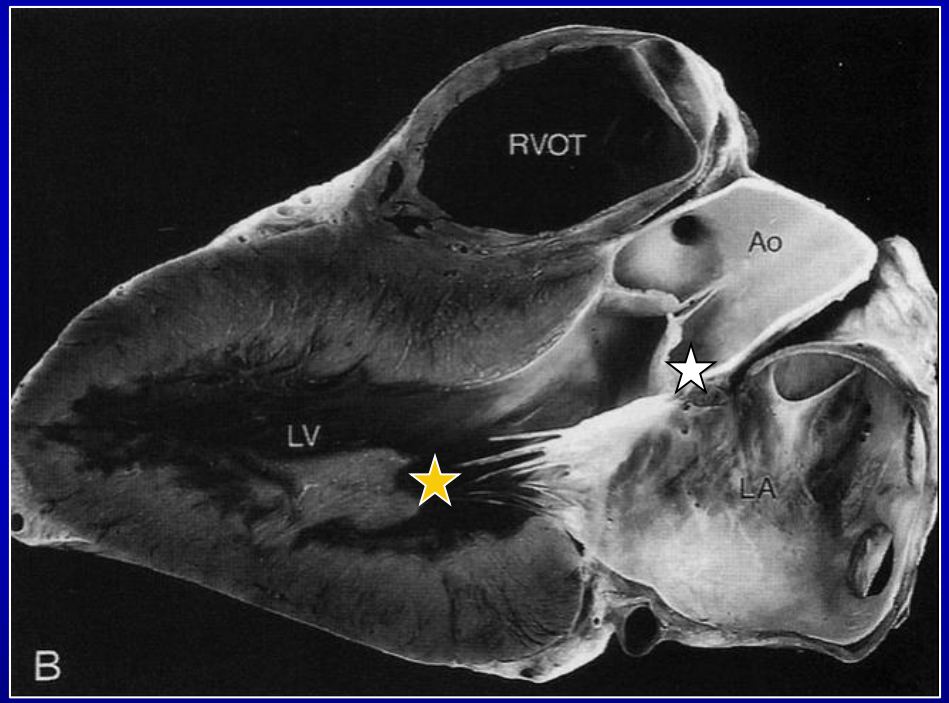
heavy trabeculations

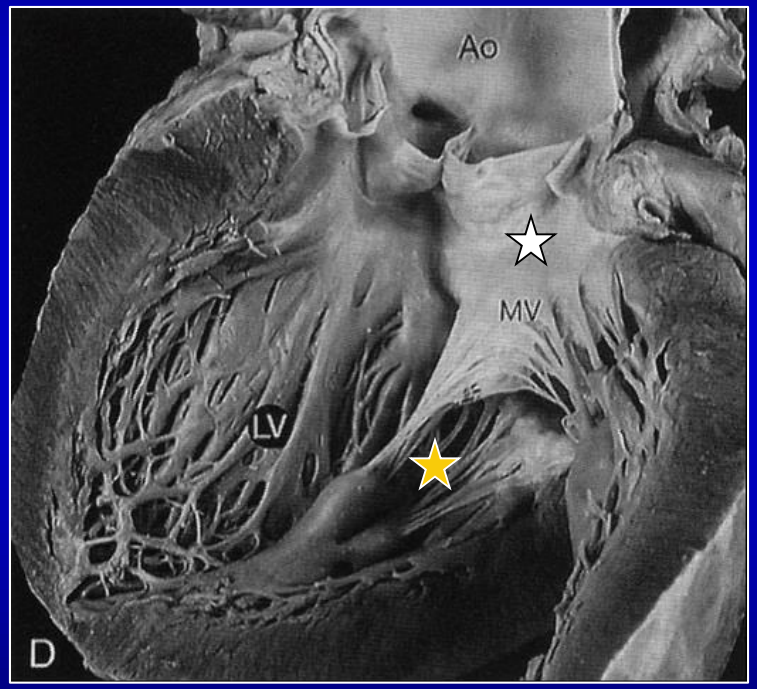
moderator band

ventriculo-infundibular fold

morphology of tricuspid valve

Morphology of the left ventricle





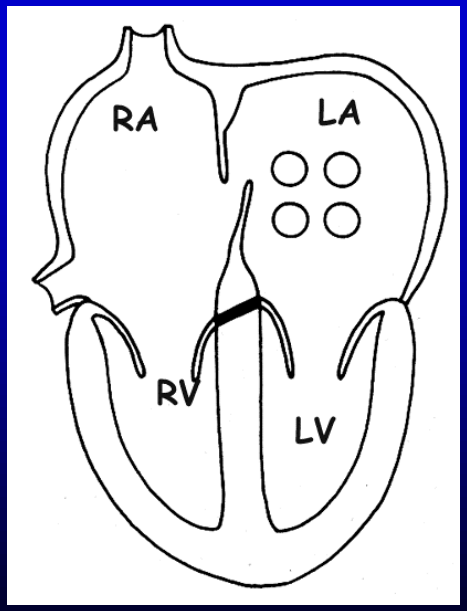
smooth endocardial surface, fine apical trabeculations
mitro-aortic fibrous continuity
morphology of mitral valve

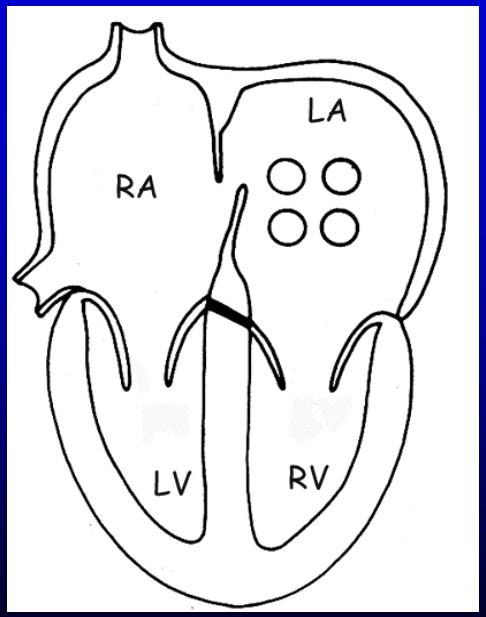
Atrioventricular Connection

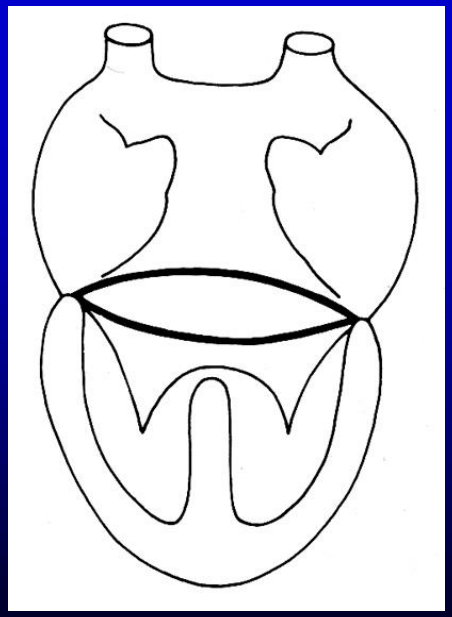
Biventricular Connection

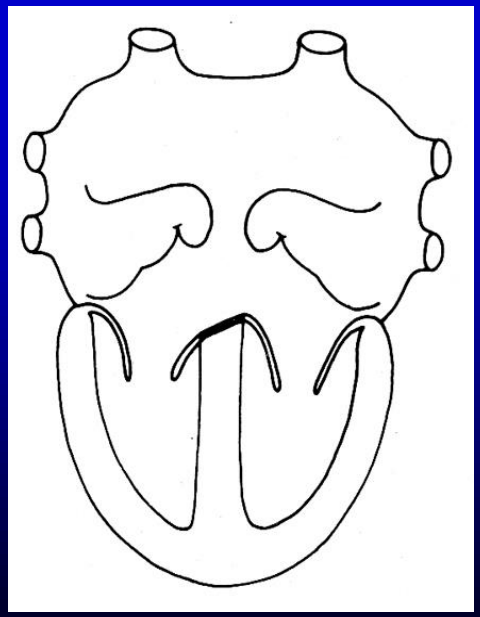
atria are connected with two distinct ventricles

concordant **discordant** **ambiguous**

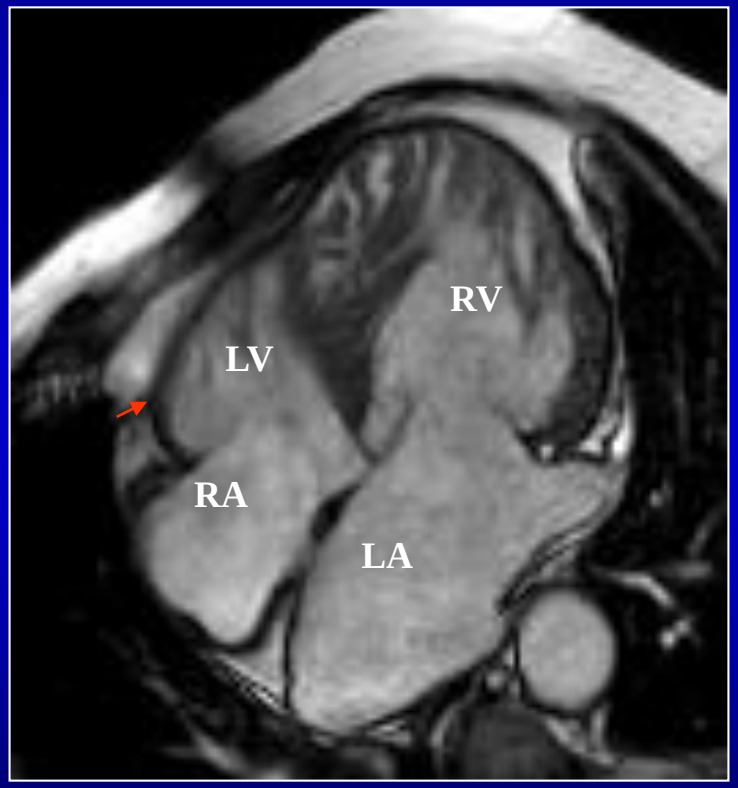








cTGA

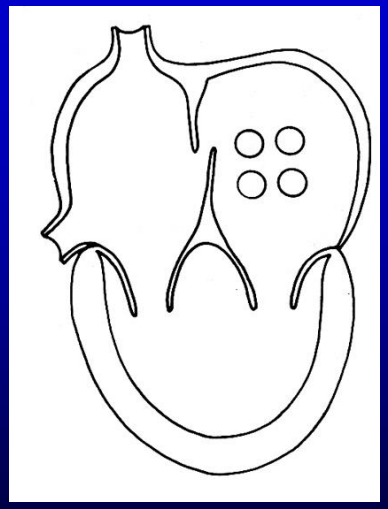


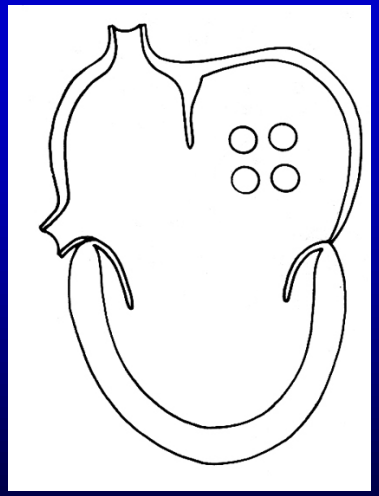
Atrioventricular Connection

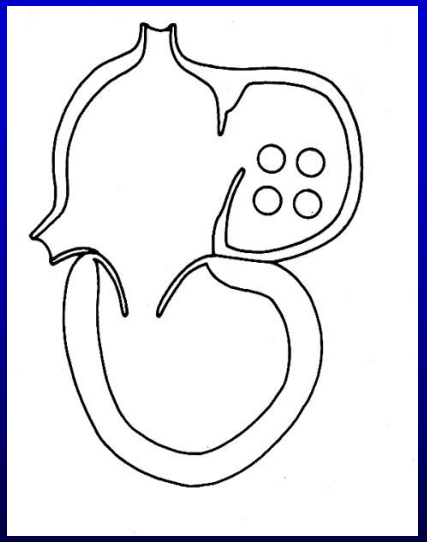
Univentricular Connection

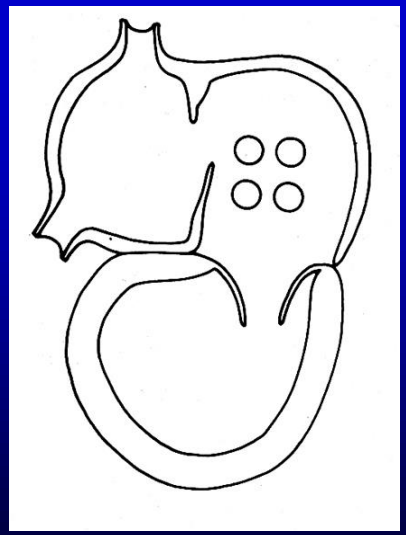
atria are connected with a single ventricle

Double inlet ***Common inlet*** ***Single inlet***

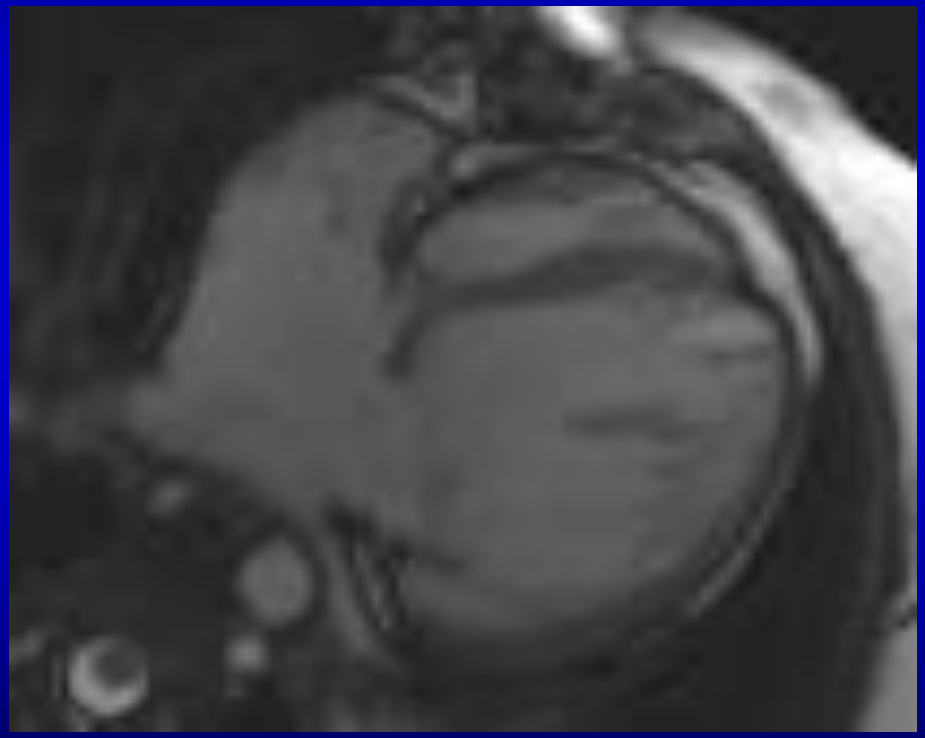




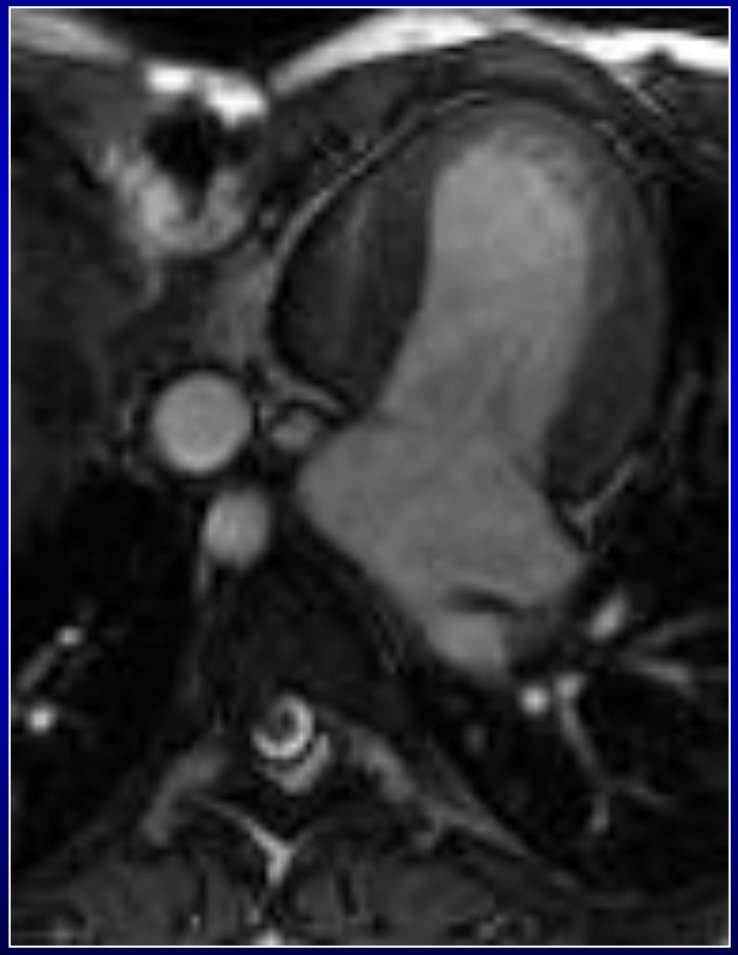




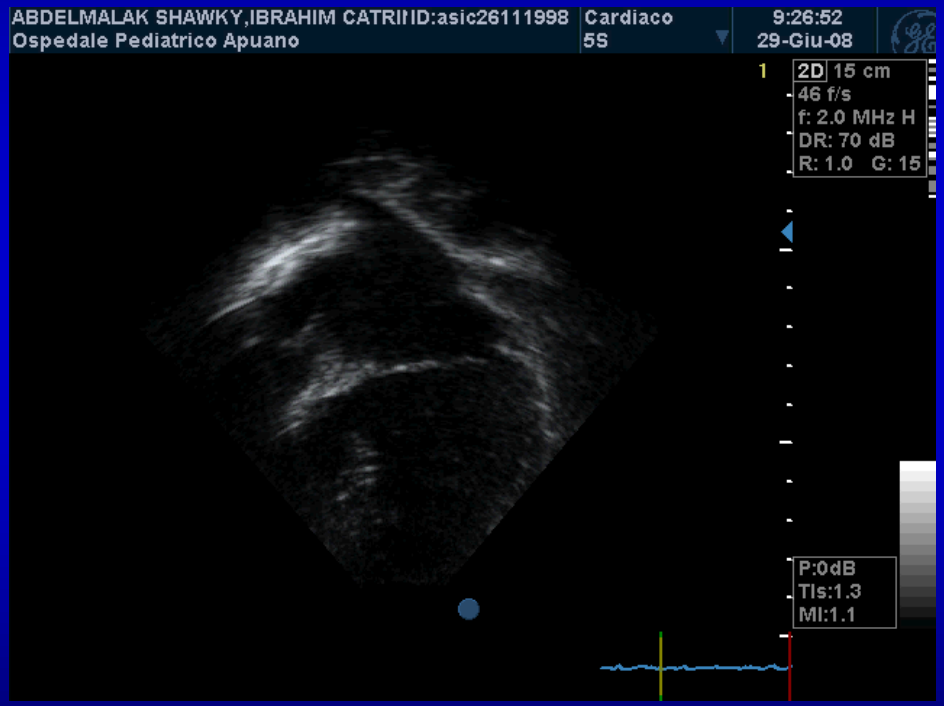
Tricuspid atresia

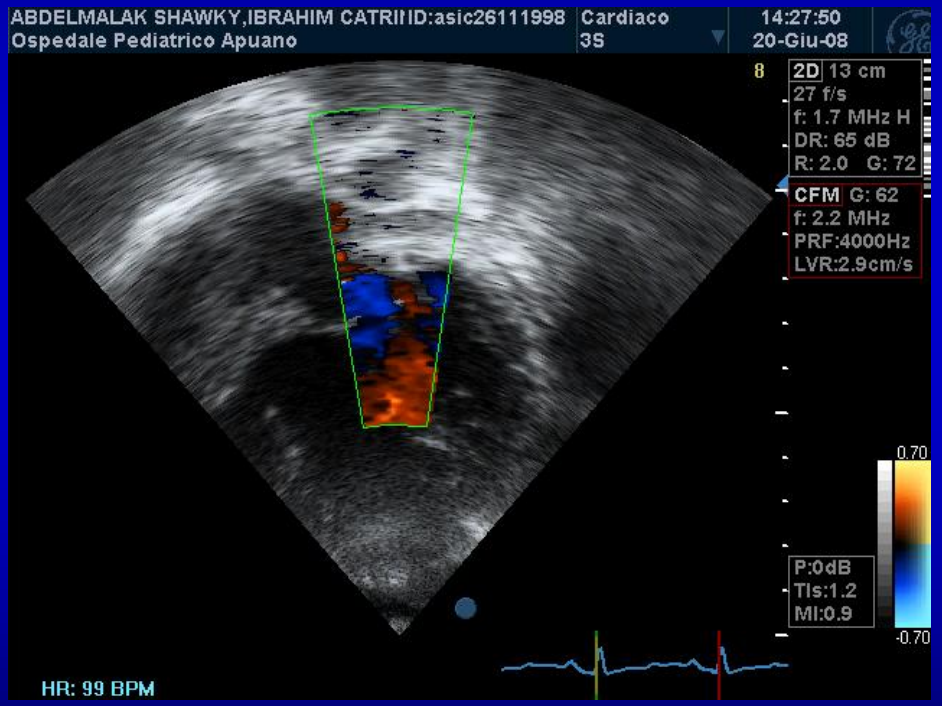


Tricuspid atresia



Tricuspid atresia

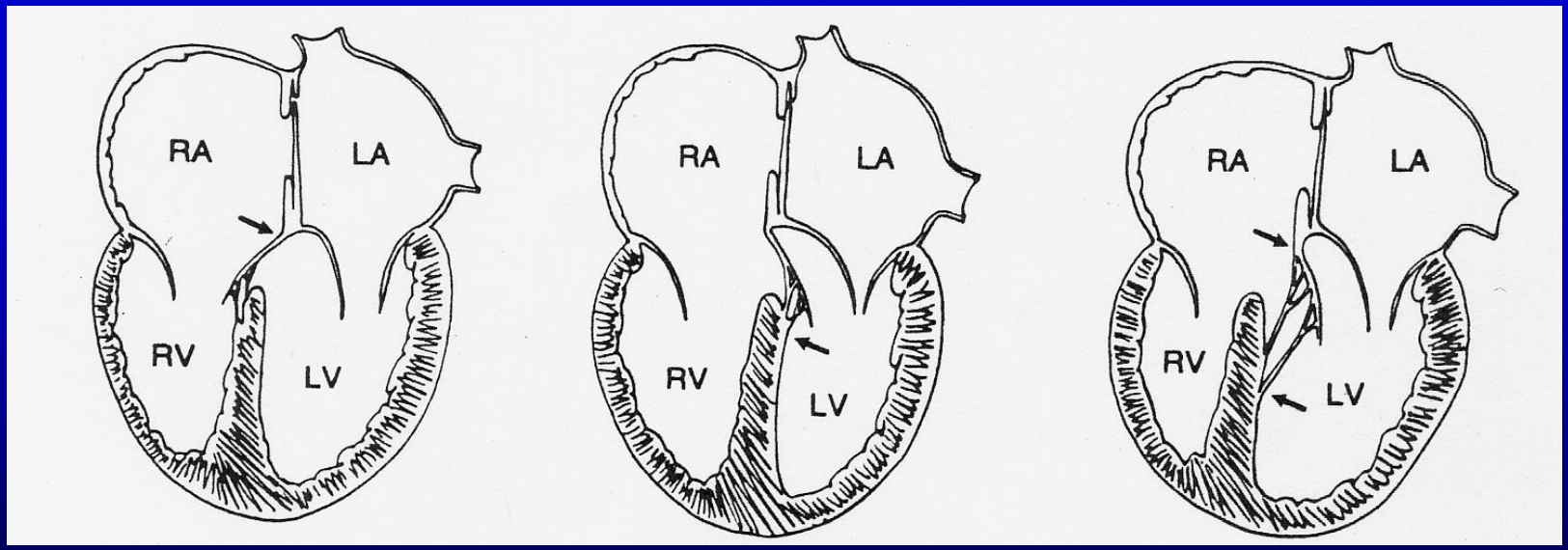




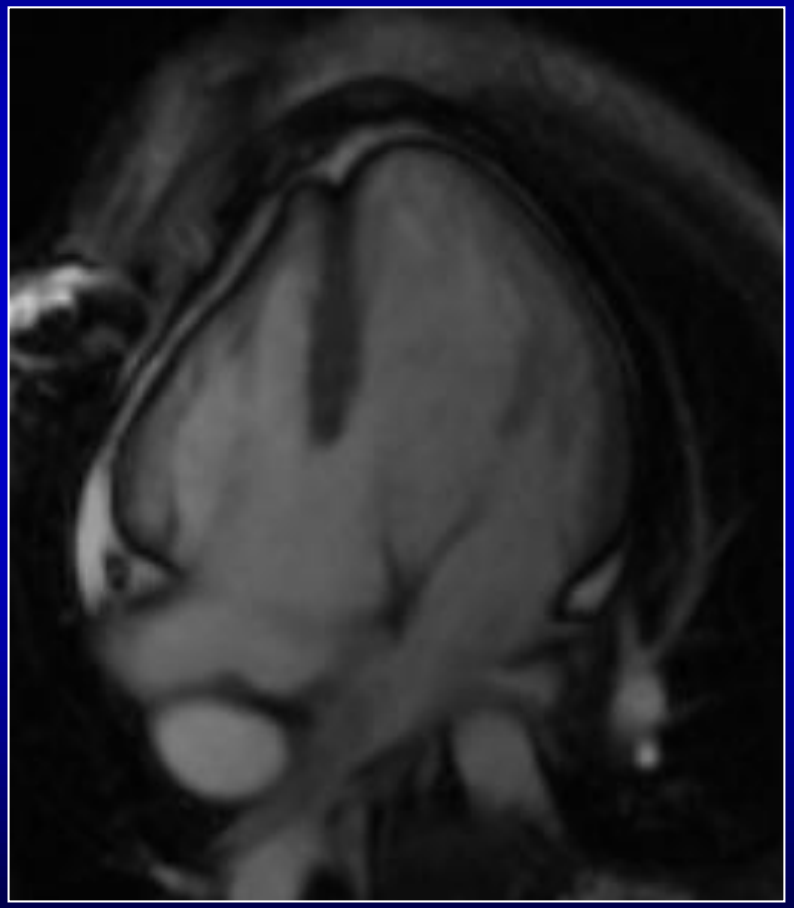
Atrioventricular Connection

Straddling/Overriding

OVERRIDING **STRADDLING** **STRADDLING WITH OVERRIDING**



Straddling atrioventricular valve

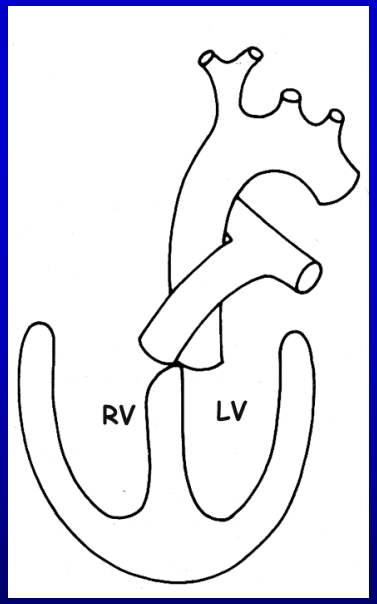


Atrioventricular connections

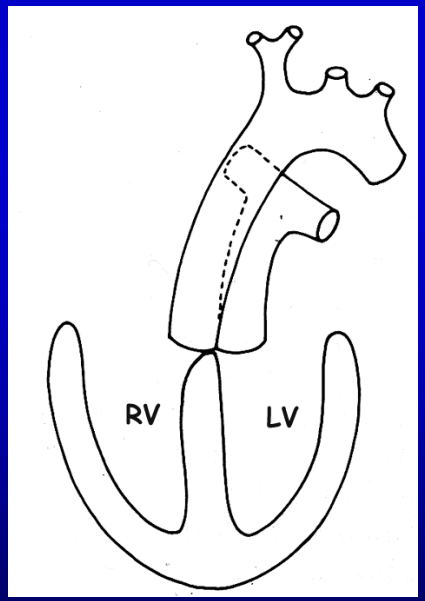
- Concordance
- Discordance
- Double outlet
- Single outlet
- Common outlet

Atrioventricular connections

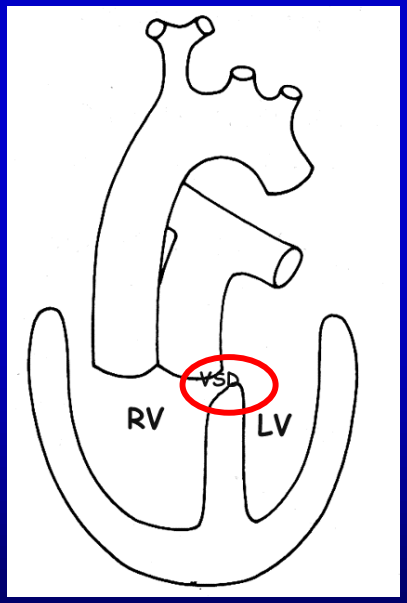
CONCORDANT

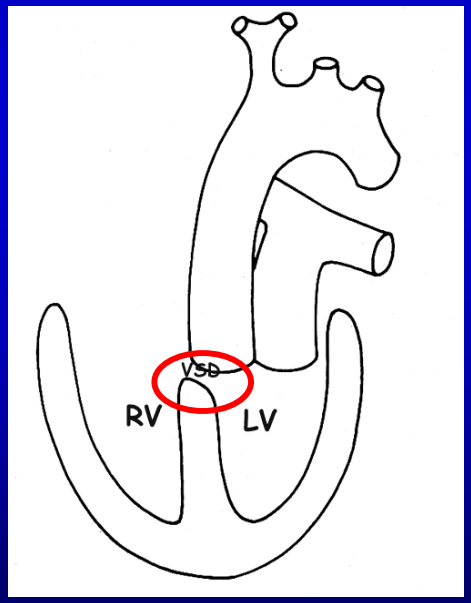


DISCORDANT



DOUBLE OUTLET

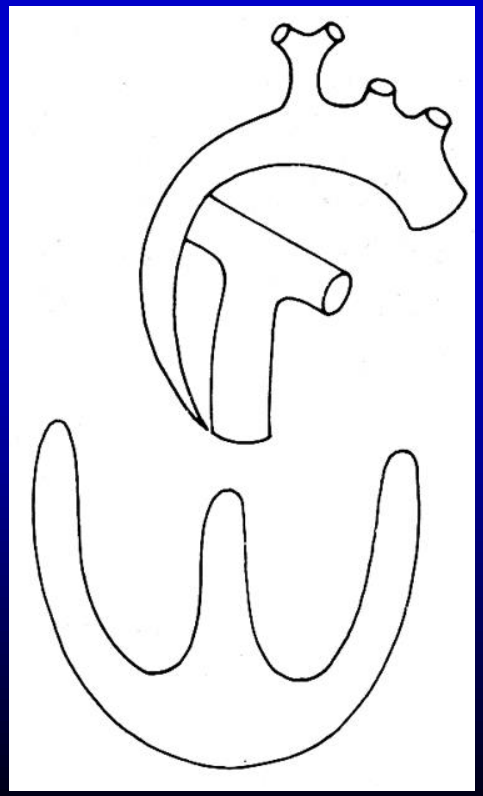




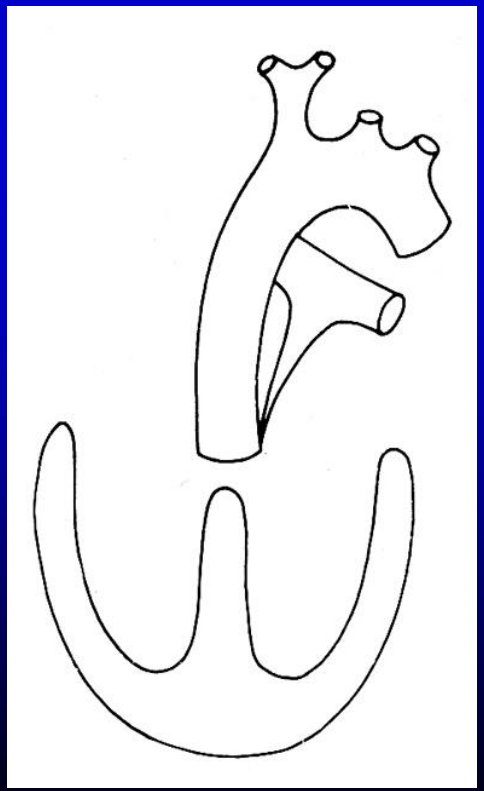
Atrioventricular connections

Single Outlet

Ao Atresia

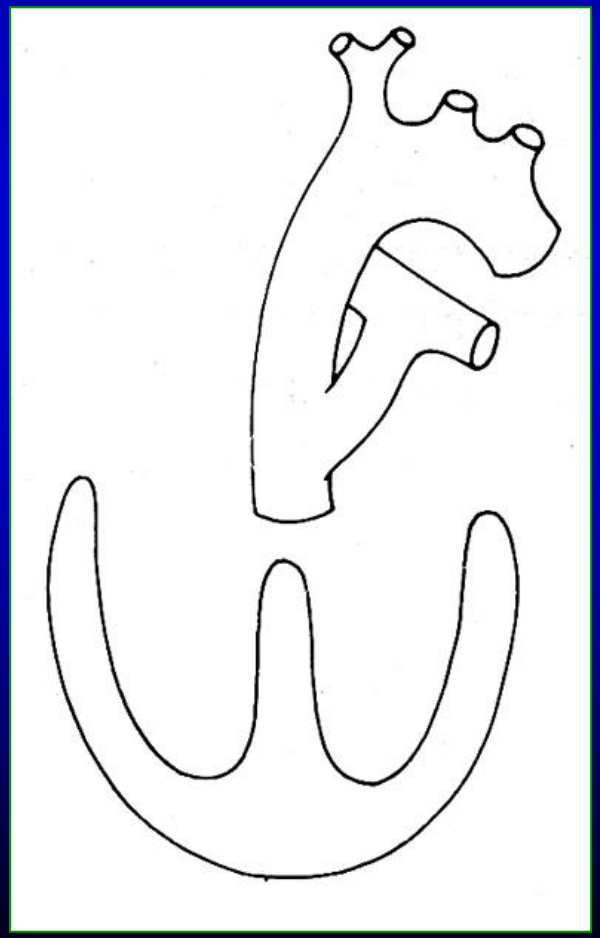


Pul. Atresia

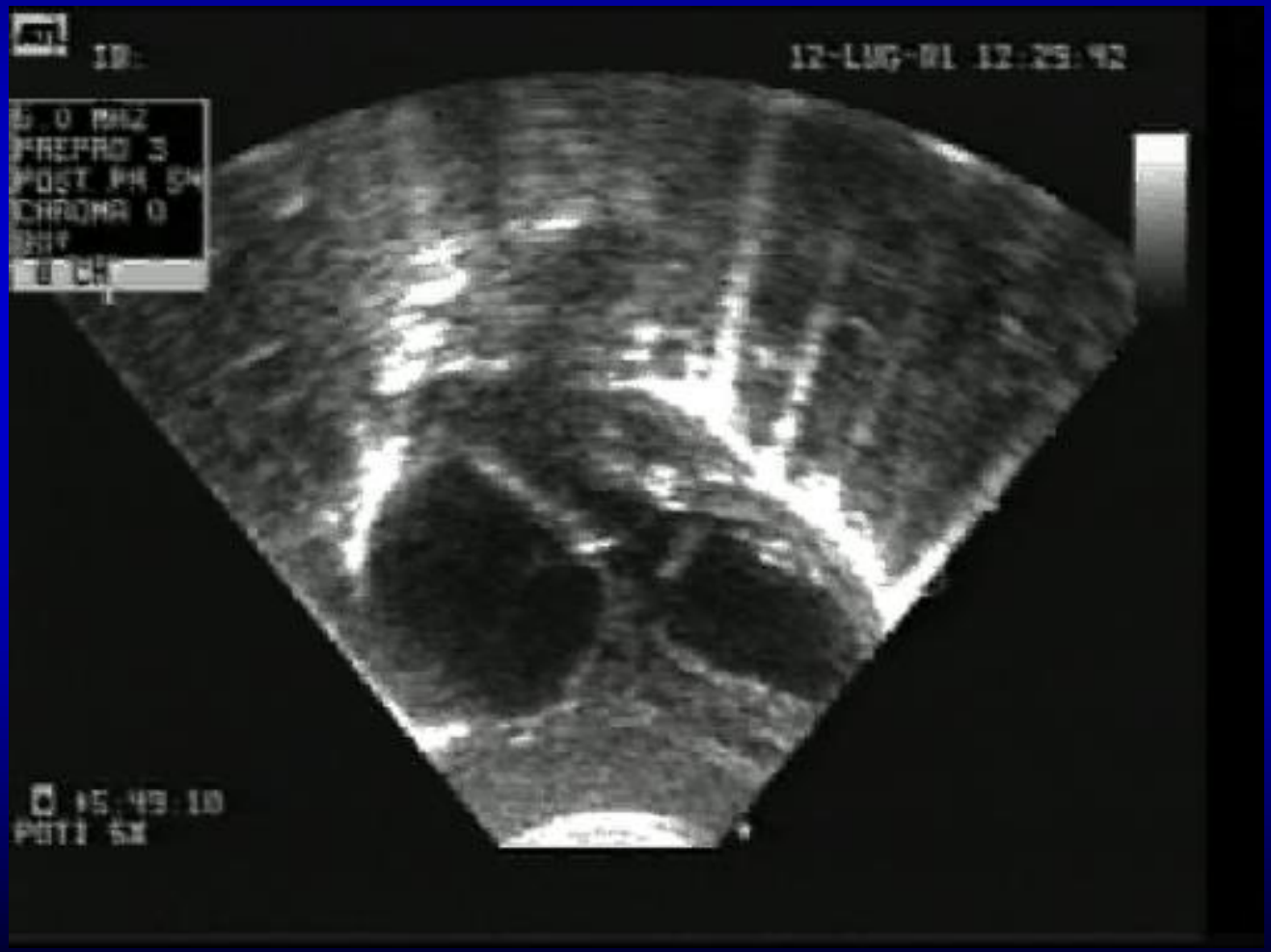


Atrioventricular connections

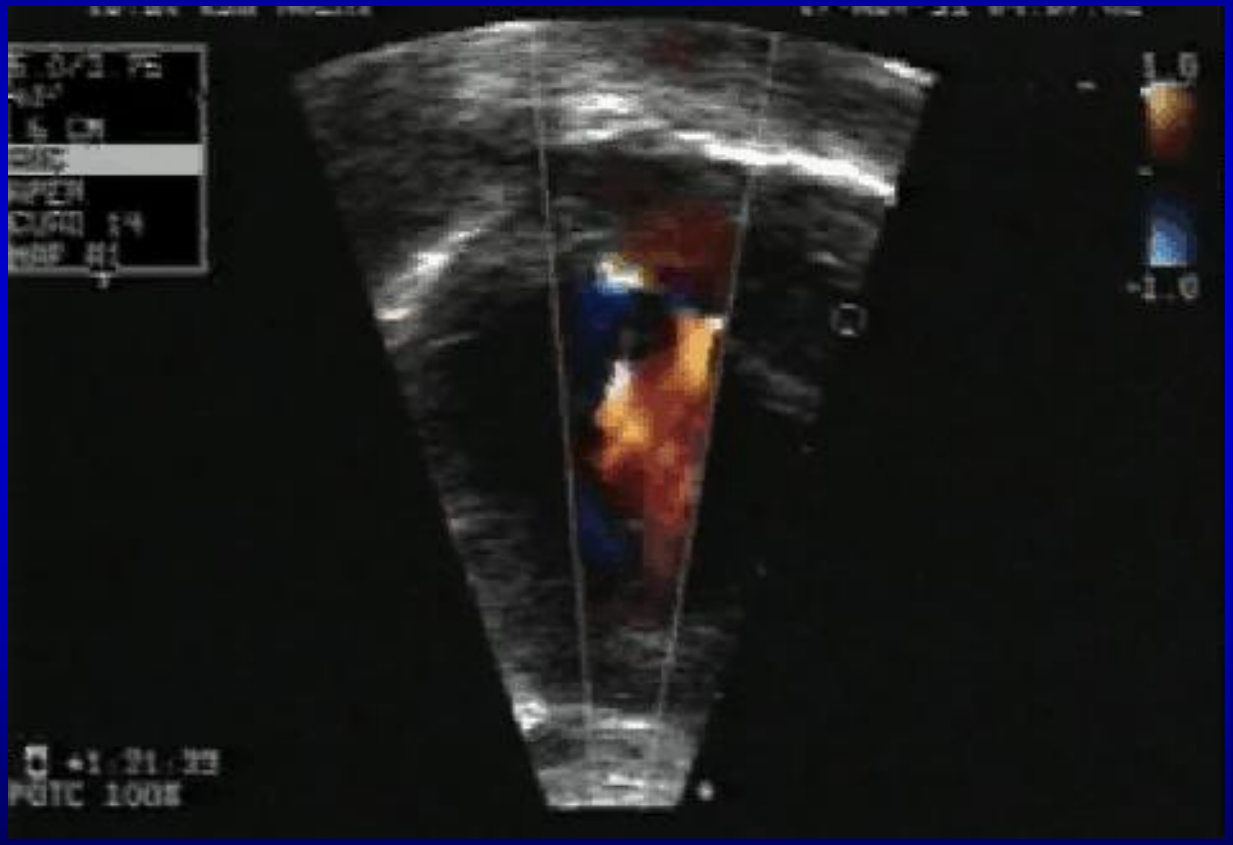
Common outlet



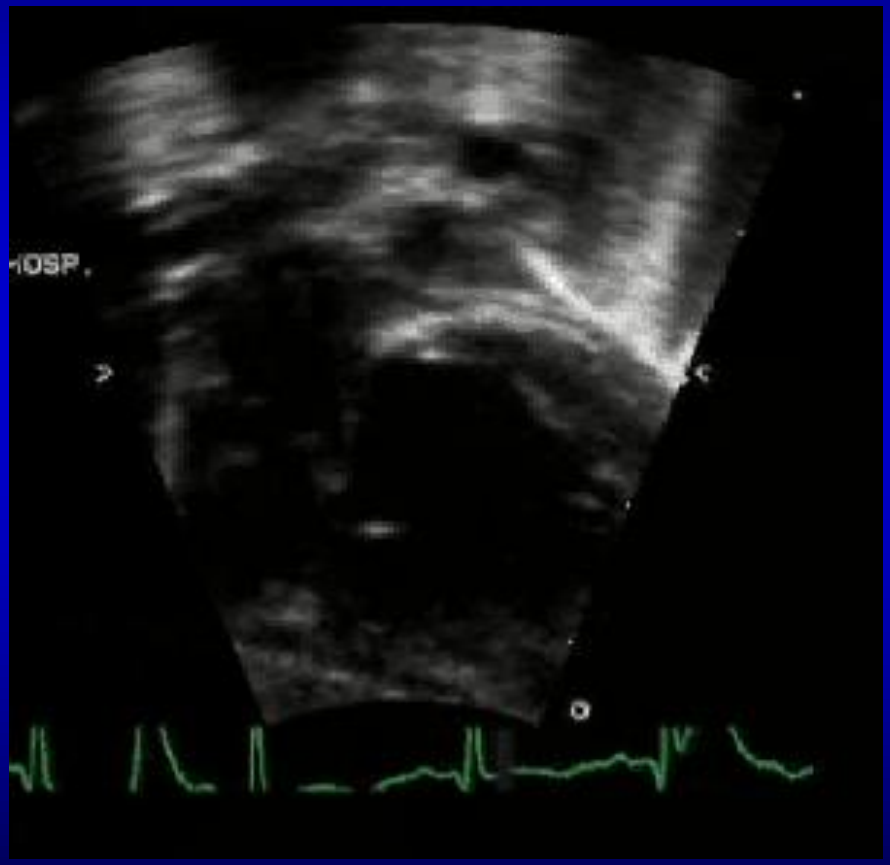
Transposition of the Great Arteries



Aortic atresia



Truncus



Diagnostic tools

- **Chest X-Ray**
  - bronchial morphology
  - position of heart and cardiac apex
  - position of liver and stomach
- **Abdominal Echo**
  - position of liver and stomach
  - presence (and number) of spleen(s)
  - position of aorta and vena cavaà
- **Echocardiography**
- **Cardiovascular MRI**
- **Cardiac Catheterization**

Università di Pisa
Scuola di Specializzazione in Cardiologia
Direttore Prof. Alberto Balbarini

Seminario

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Ospedale del Cuore G.Pasquinucci, Massa