



Fetal Cardiology

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Fetal Cardiology

- Cardiac anomalies are the most frequently overlooked group of abnormalities
- Congenital heart disease = 0.8% of all pregnancies
 - 4% one sibling affected; 10% two siblings affected
 - 9% father affected
 - 12% mother affected
- Causes > 50% deaths from congenital disease

Fetal Cardiology

- Risk Factors for congenital heart disease:
 - Family history
 - Recurrence risk (hypoplastic left heart as high as 13.5%)
 - Nongestational DM
 - Maternal infection (rubella)
 - Lupus
 - Drugs (anticonvulsants, etoh, amphetamines, ocp, vit A, steroids, etc.)

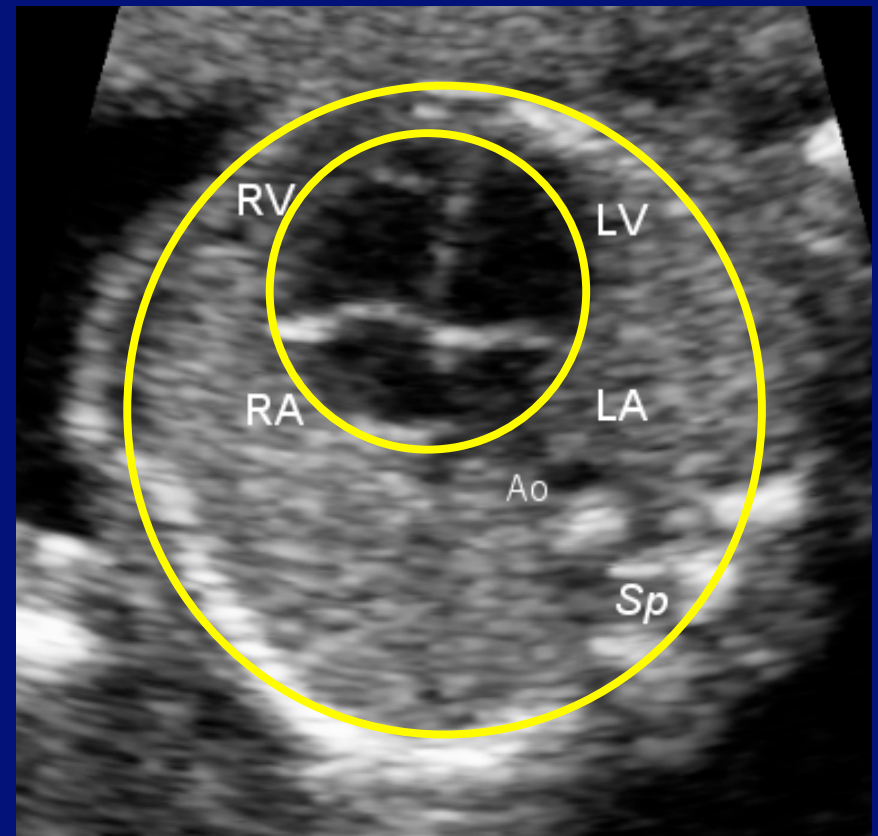
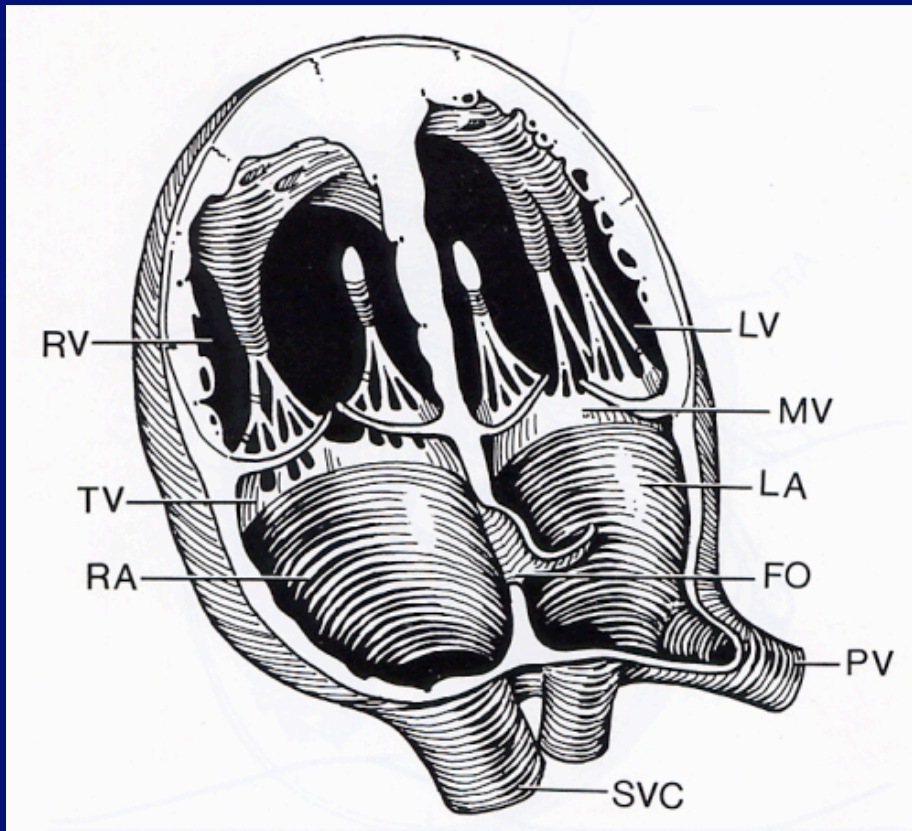
Fetal Cardiology

- AIUM / ACR standards in the 2nd and 3rd trimesters include:
 - Four chamber view
 - Position of fetal heart in the thorax
- LVOT and RVOT not yet part of standards
- 4 chamber view alone: 33-63% sensitive
- With outflow tracts: 83-85% sensitive [2]

GOALS

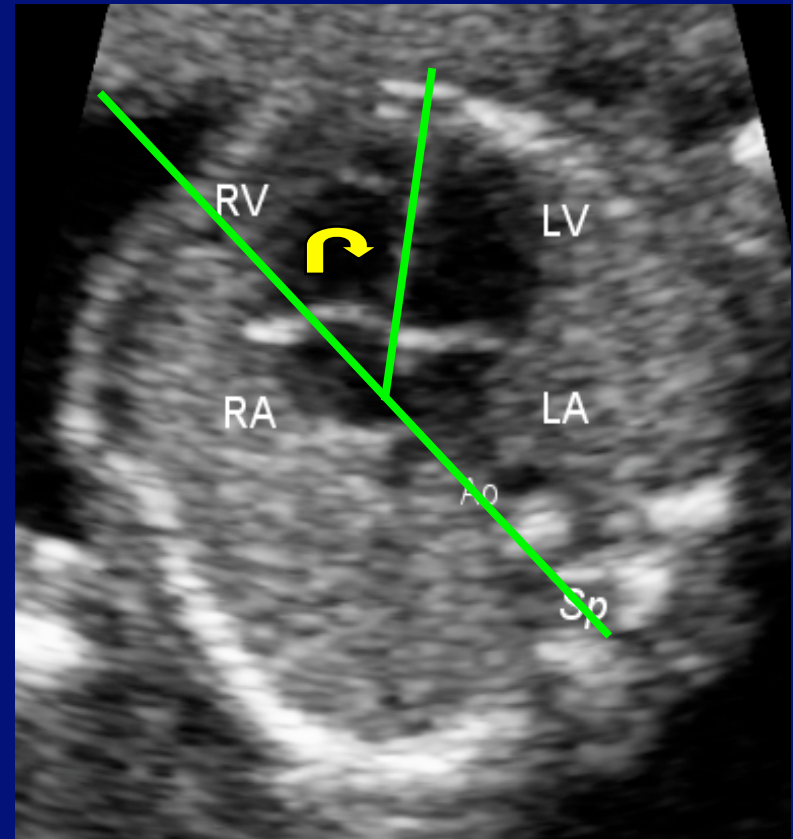
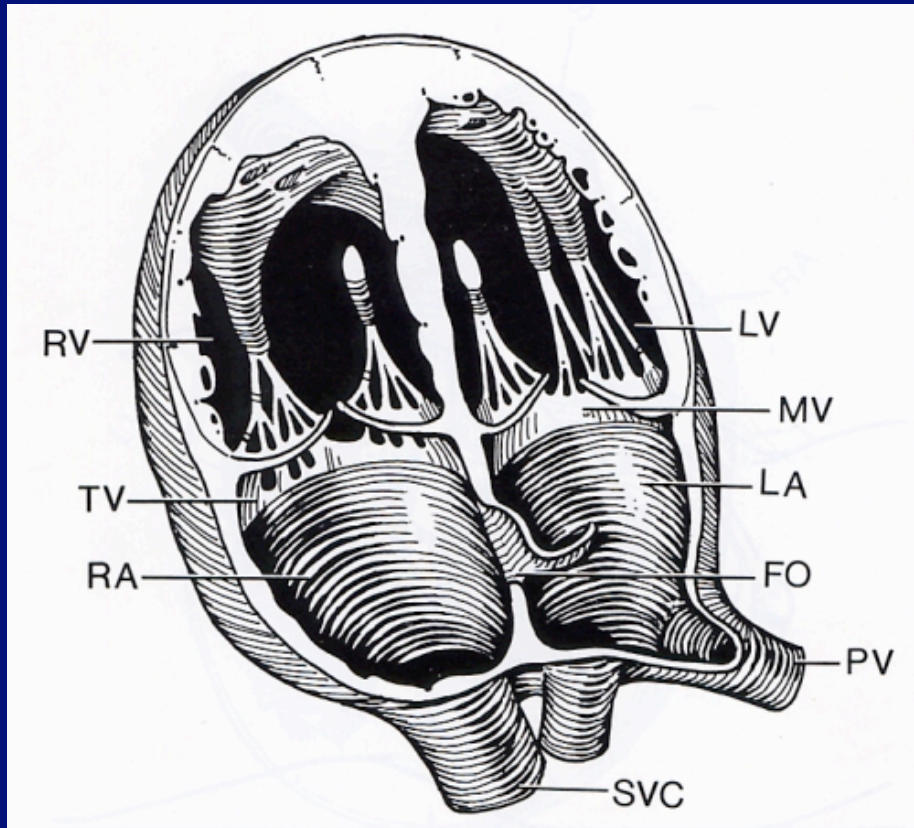
- Review normal cardiac anatomy and its sonographic appearance (four chamber, LVOT, RVOT)
- Explore diagnostic pitfalls
- Review the appearance of more common structural cardiac defects

The Four Chamber View



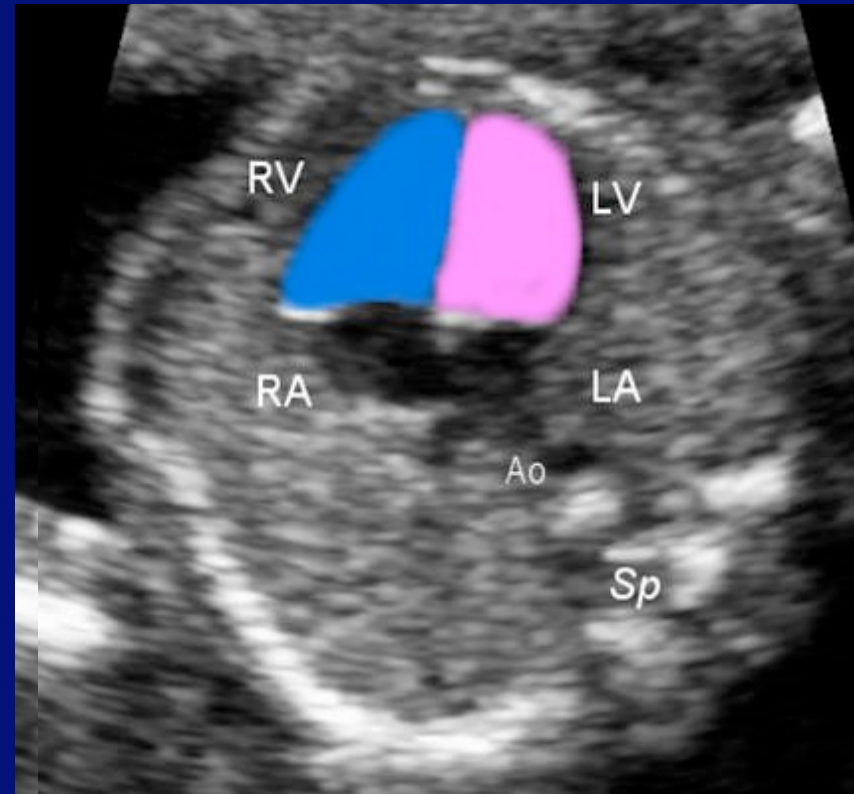
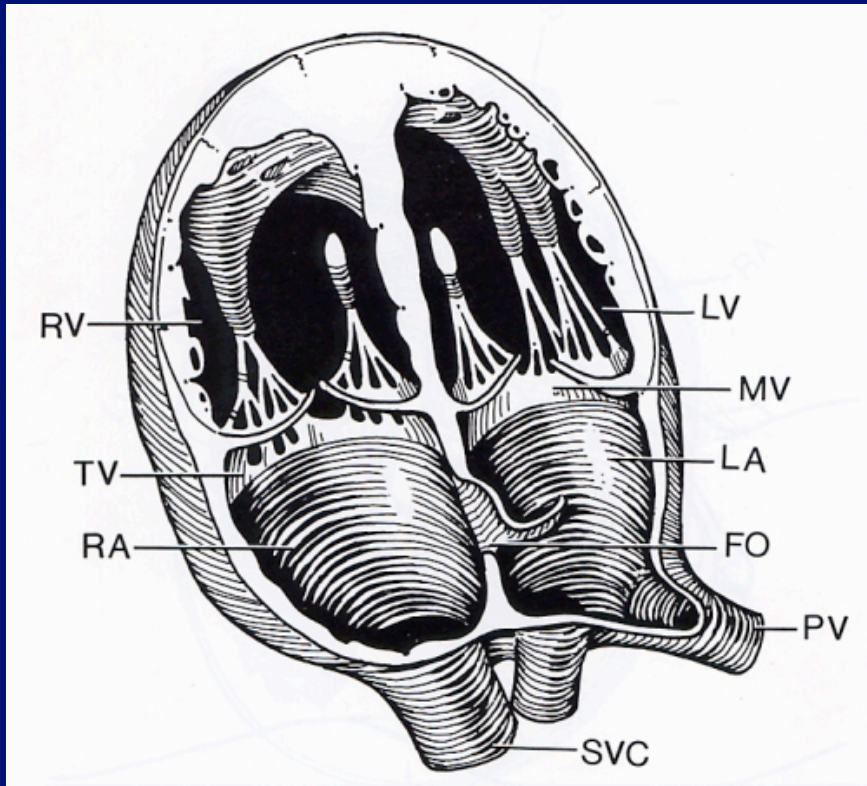
1. Heart fills one third of the chest

The Four Chamber View



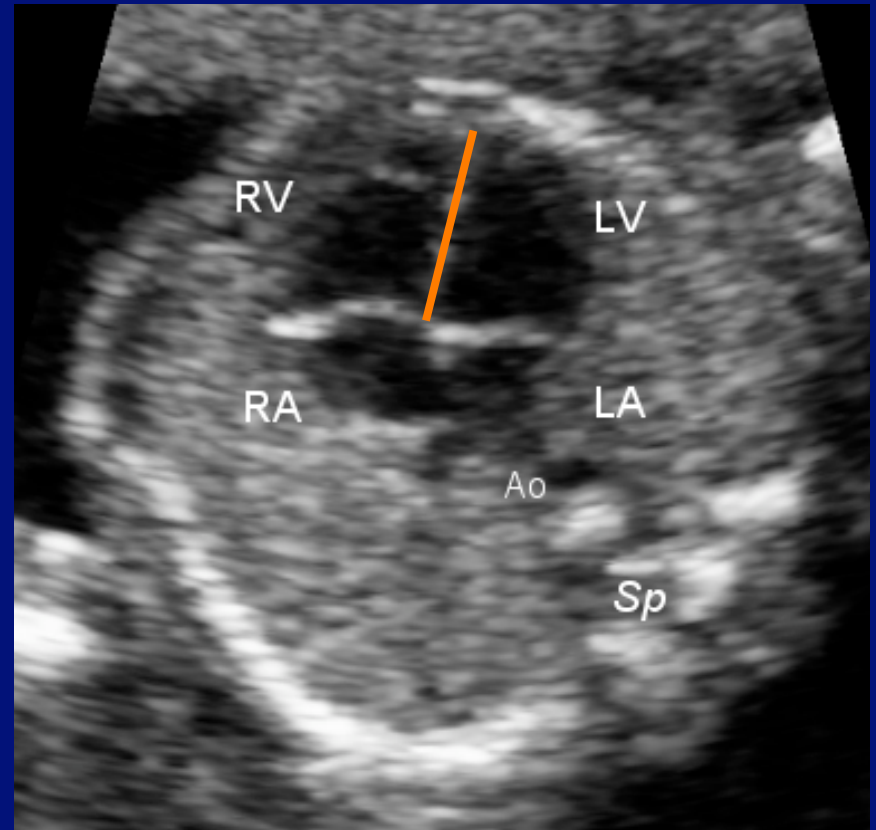
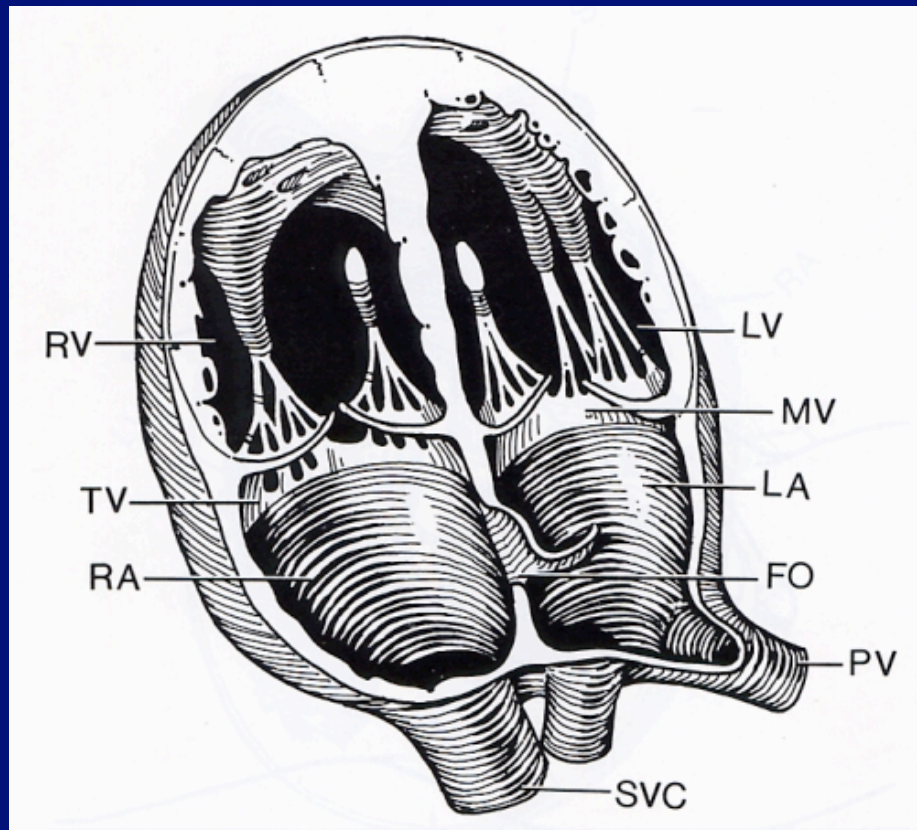
2. Apex points to the left (45 degree angle)

The Four Chamber View



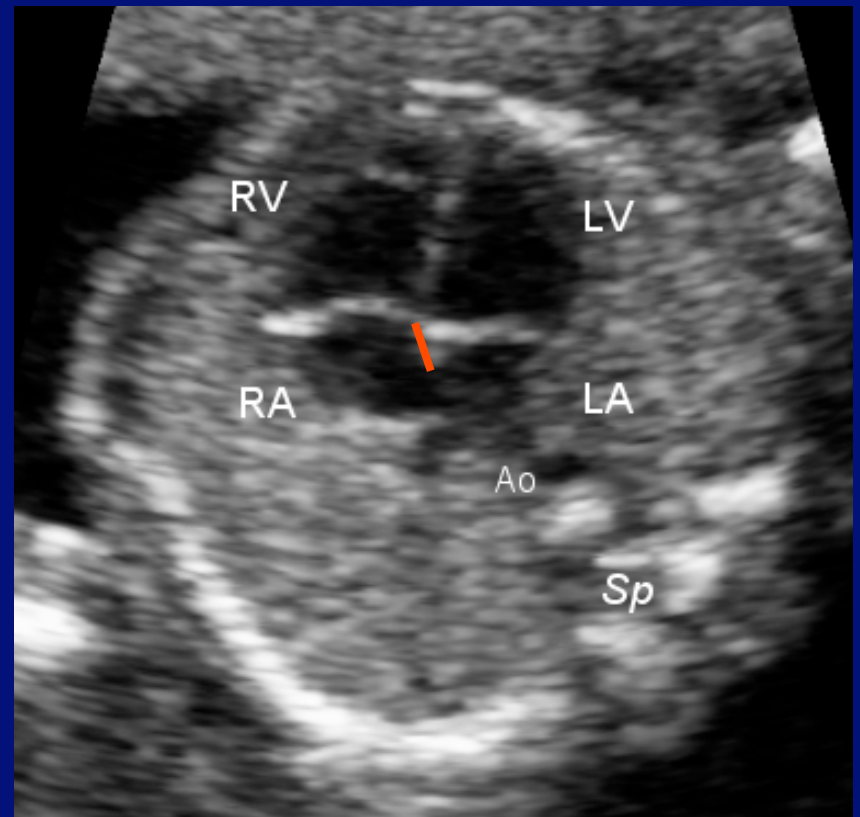
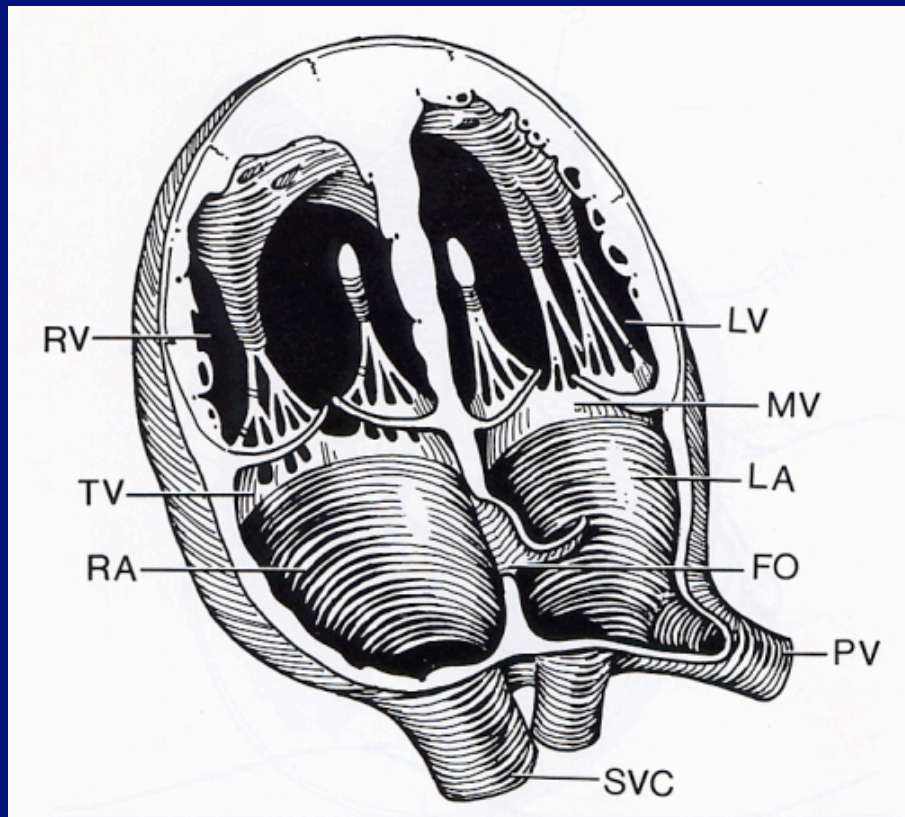
3. Size of right chambers approximates left chambers

The Four Chamber View



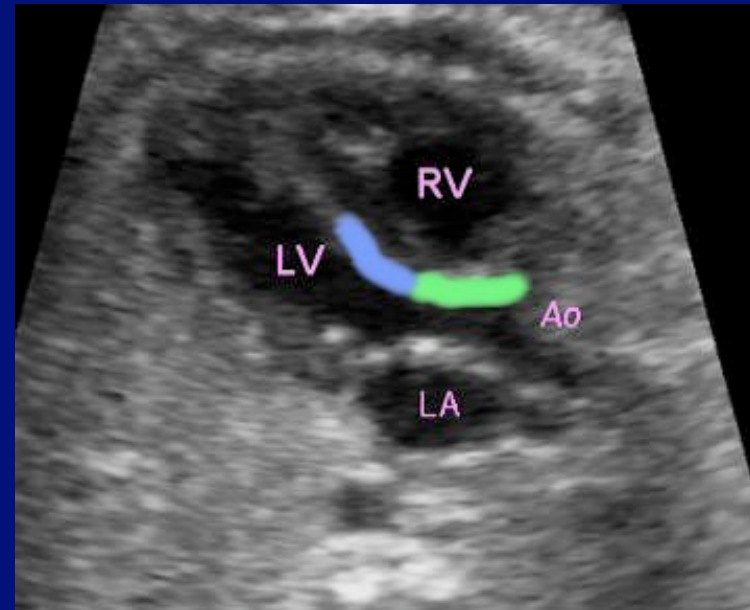
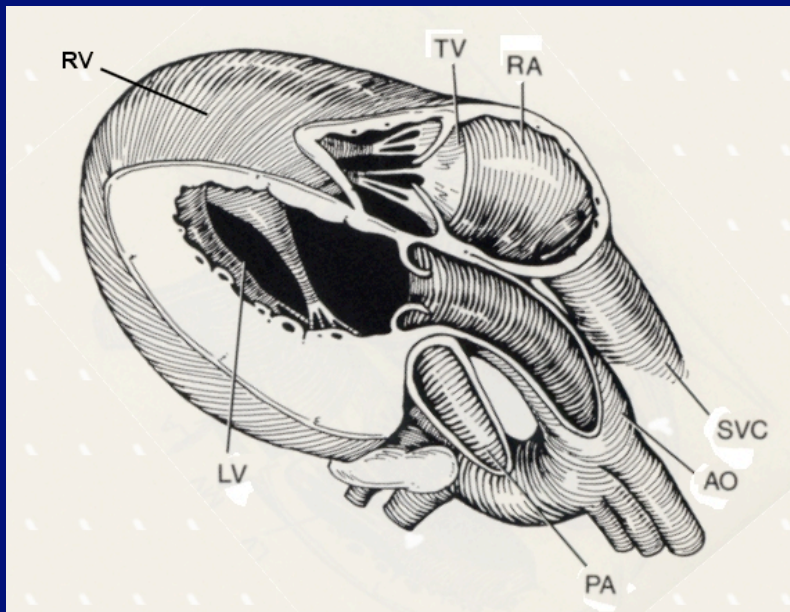
1. MV and TV move on real time imaging
5. Ventricular septum symmetric

The Four Chamber View



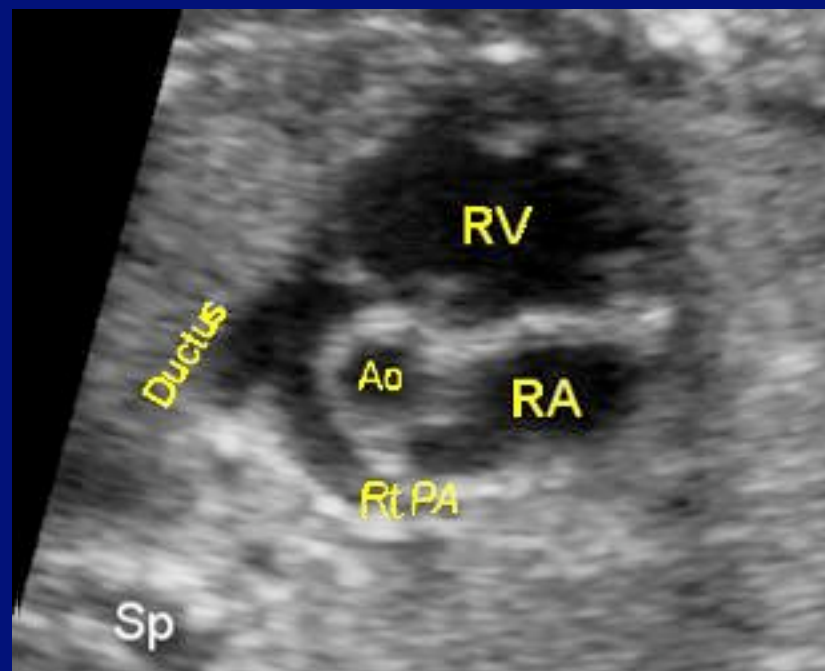
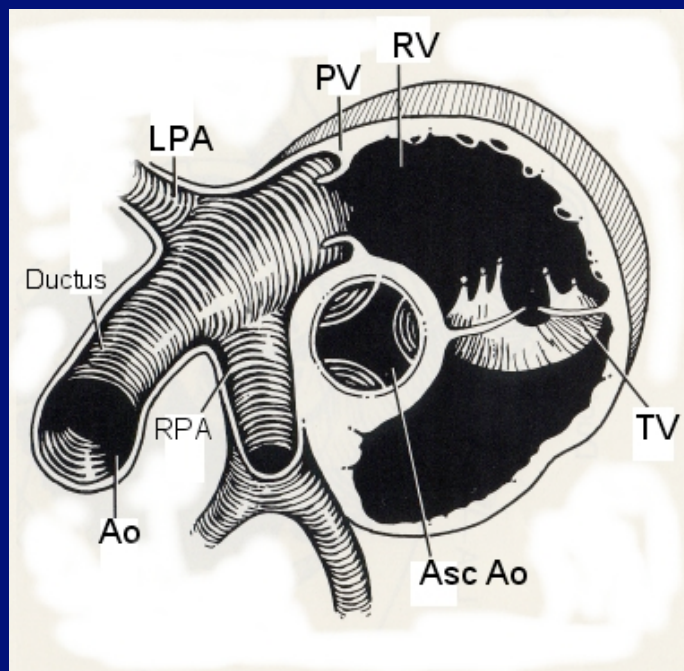
6. Portion of the atrial septum present (crus)

Left Ventricular Outflow Tract

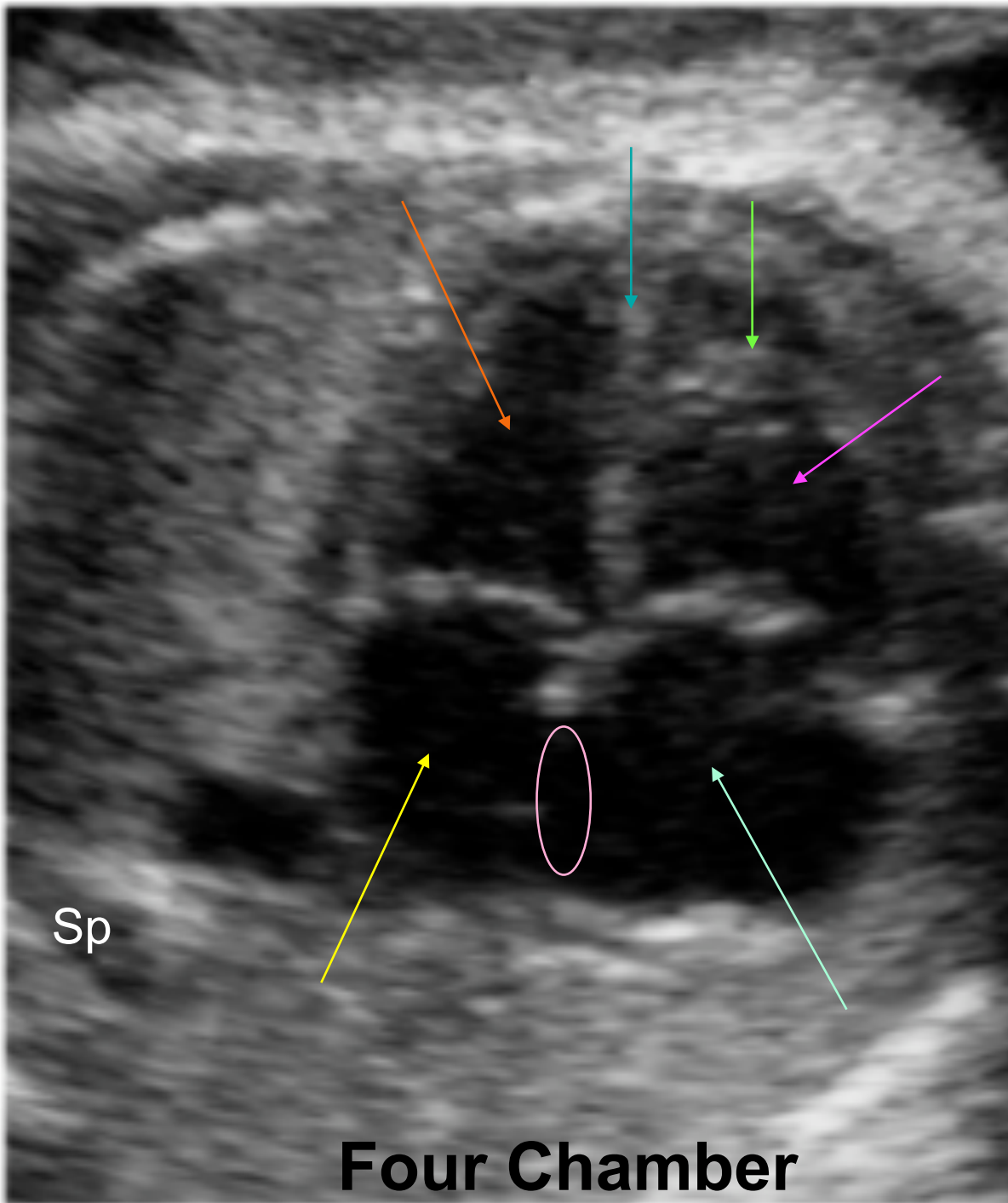


- Identify: LV, RV, IV septum, aorta (normal caliber), +/- LA, +/- RA
- Medial wall of the ascending aorta merges with the top of the IV septum (most frequent location for VSD)
- Pathology: VSD, tetralogy of Fallot, transposition, truncus arteriosus

Right Ventricular Outflow Tract



- Identify: branching of the main PA into right PA and ductus arteriosus (to desc Aorta), asc aorta in cross section, desc aorta to left of spine; verify PA crosses anterior to asc aorta
- Pathology: transposition, truncus arteriosus



Spine

Left Atrium

Right Atrium

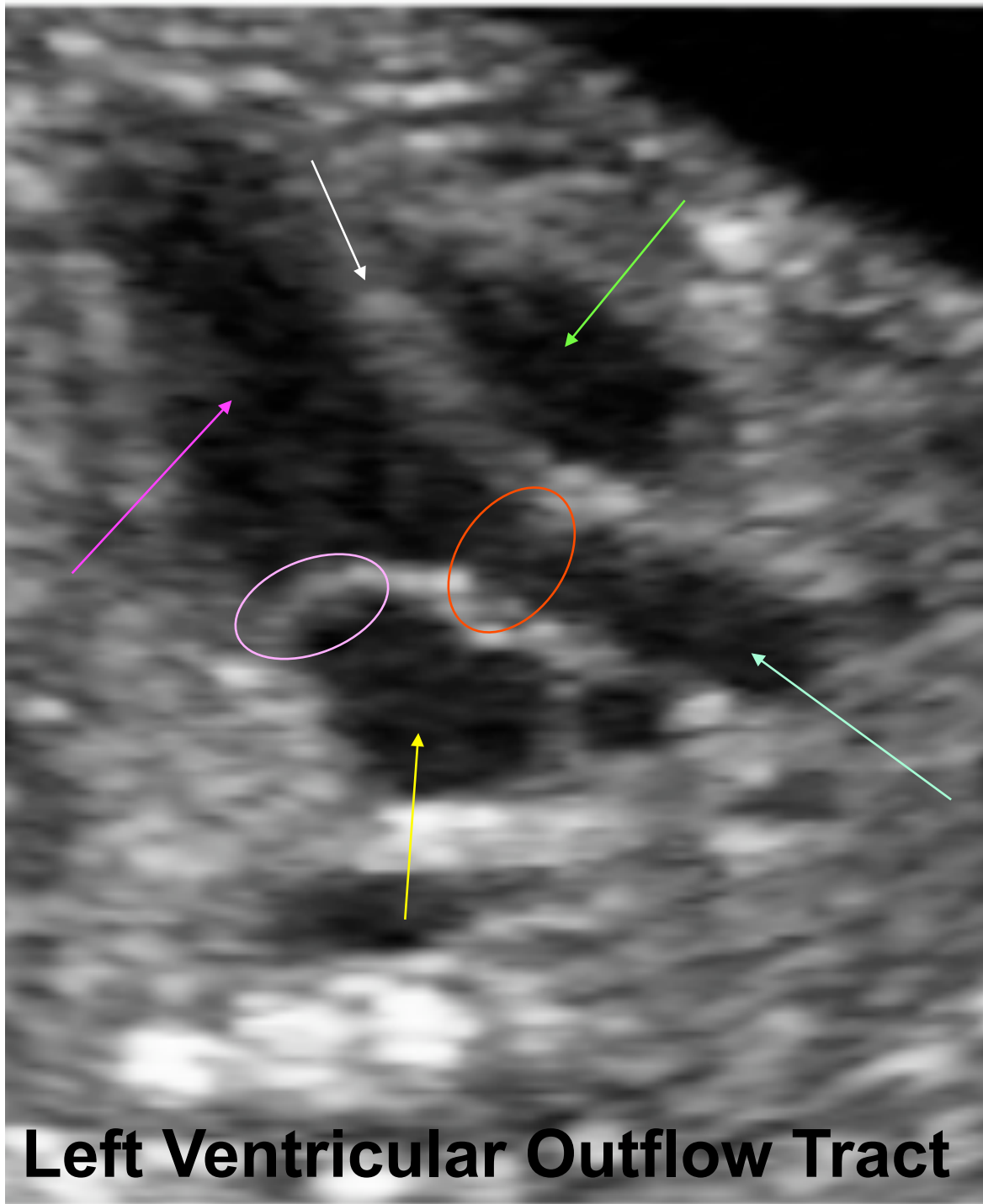
Left Ventricle

Right Ventricle

Moderator Band

Foramen Ovale

Interventricular Septum



Left Atrium

Left Ventricle

Right Ventricle

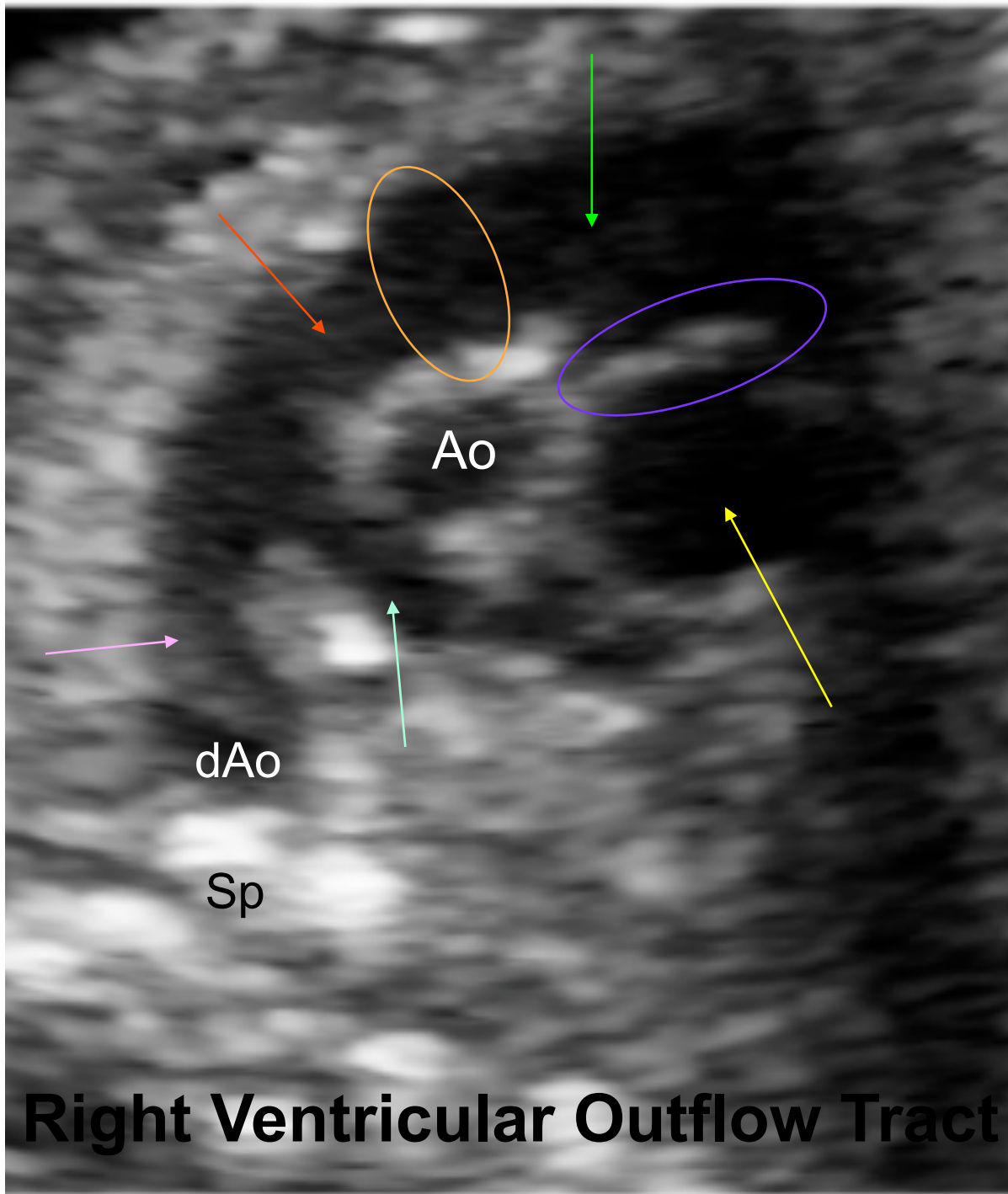
Aorta

Mitral Valve (closed)

Aortic Valve (open)

Interventricular Septum

Left Ventricular Outflow Tract



Right Atrium

Right Ventricle

Main Pulmonary Artery

Right Pulmonary Artery

Ductus Arteriosus

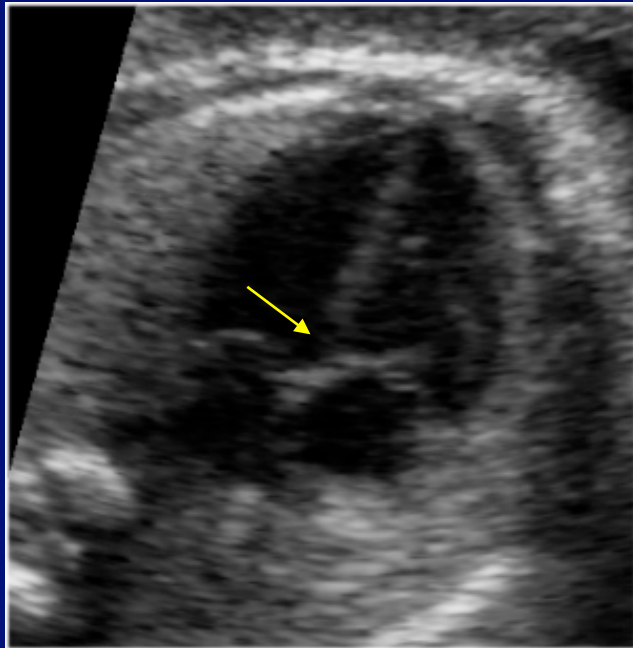
Ascending Aorta

Tricuspid Valve

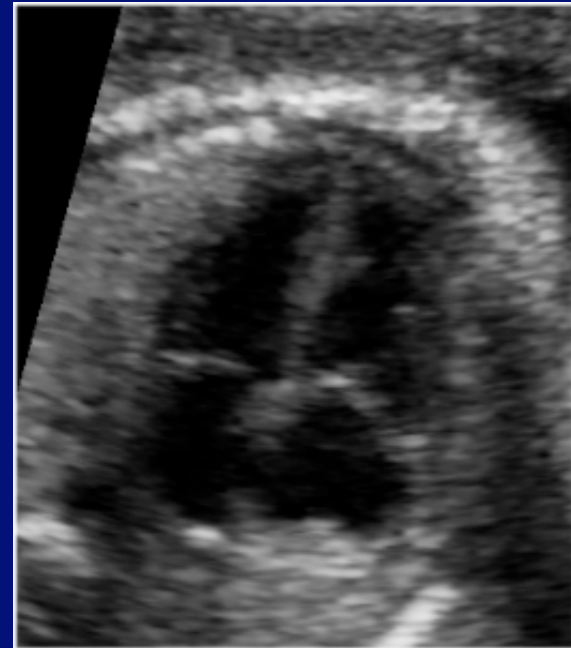
Pulmonic Valve (open)

Descending Aorta

Pitfalls: PseudoVSD



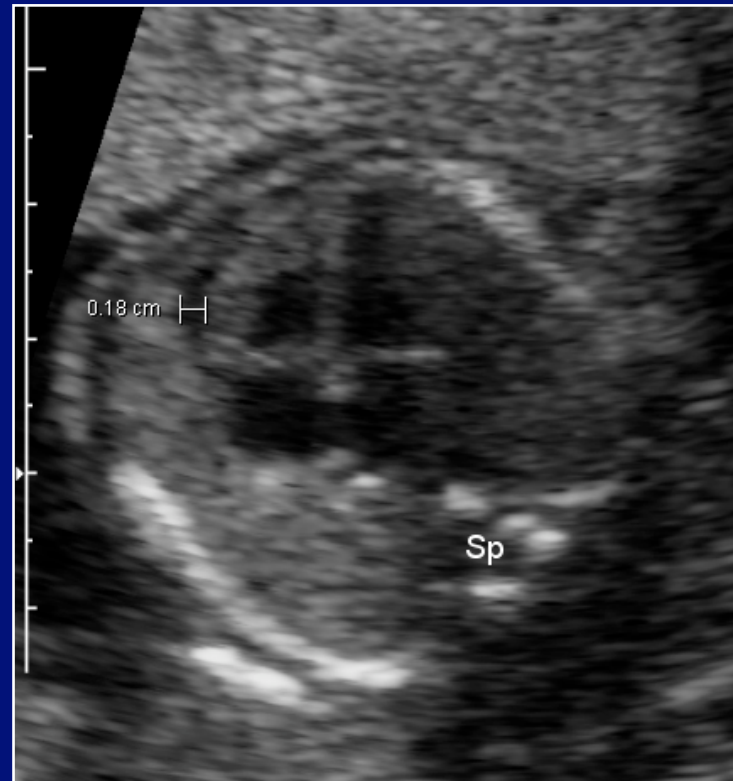
IV septum parallels US beam
(echo dropout)



Changing Angle

- When the IV septum parallels the US beam, resultant echo drop out looks like VSD
- IV septum normally tapers near the AV valves
- Corrected by changing the angle of view

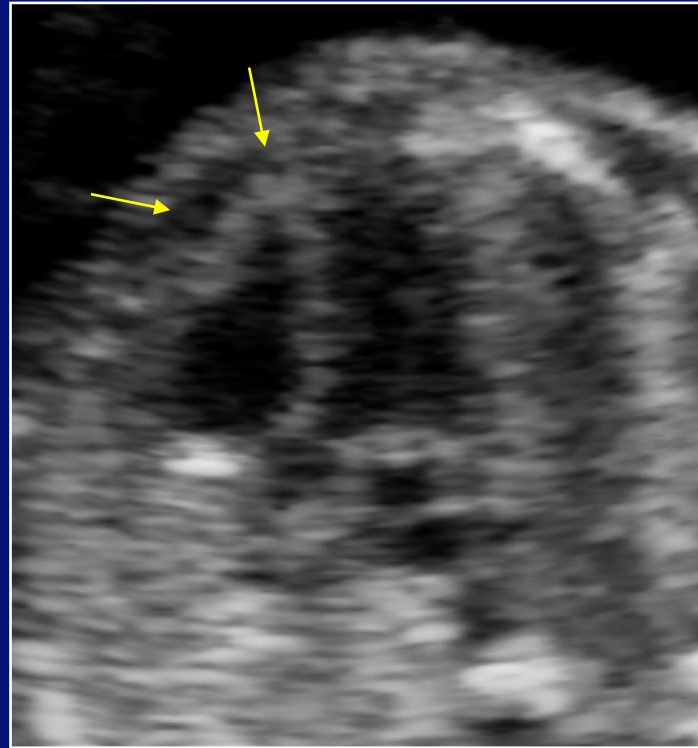
Pericardial effusion ???



NO!!!

Normal amount fluid (< 2mm)

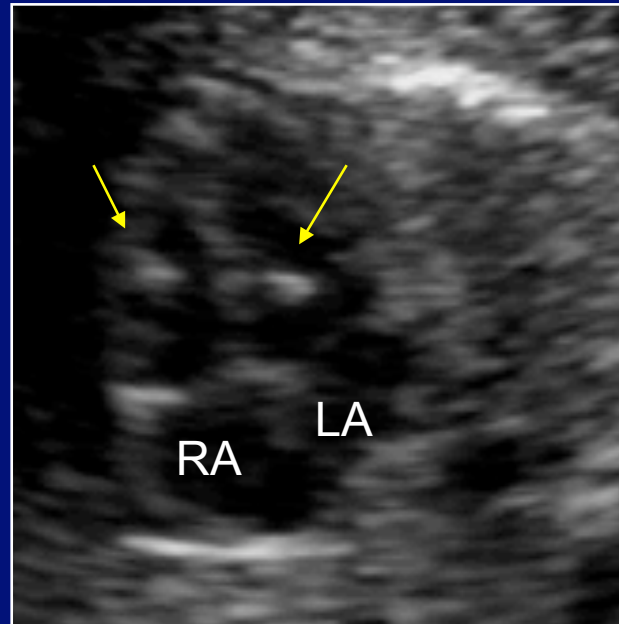
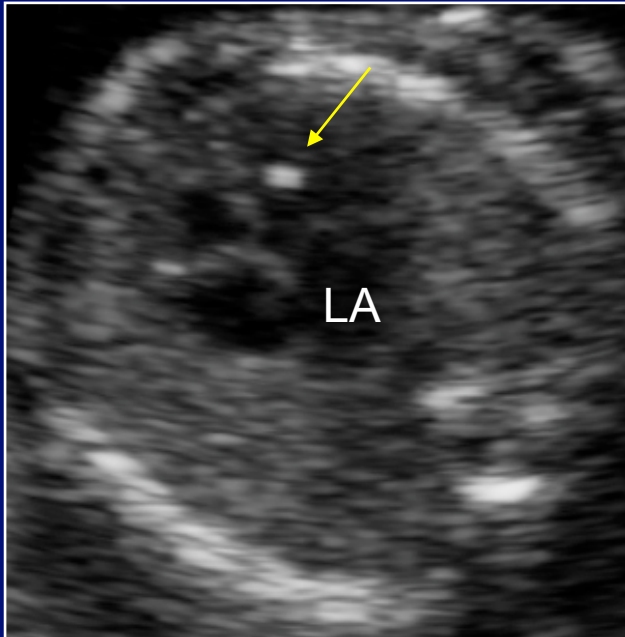
Pericardial Effusion???



NO !!!

The hypoechoic myocardium extends into the interventricular septum (arrow), allowing differentiation from a pericardial effusion

Echogenic foci: EFLV, EFRV



- Weak association with cardiac abnormalities
- Doubles the risk for Trisomy 21
- Can be confused with portions of the chordae tendinae (differentiate on real time imaging)

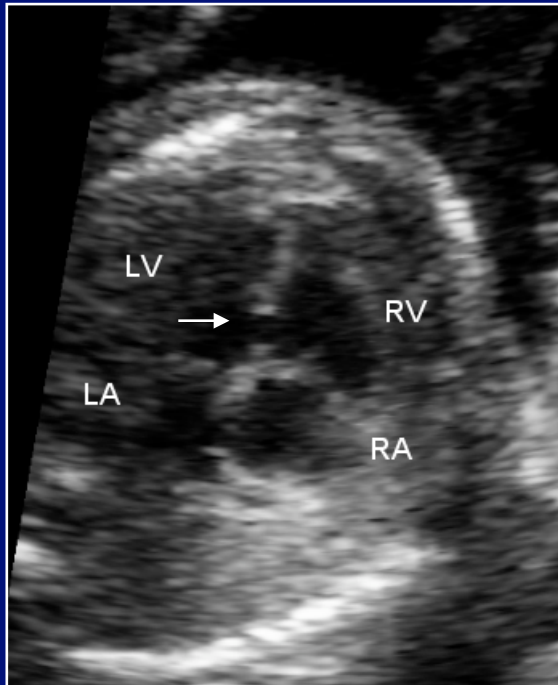
Pathology

- VSD
- Tetralogy of fallot
- Endocardial cushion defect
- Transposition of the great vessels
- Hypoplastic left heart
- Hypoplastic right heart
- Double outlet right ventricle
- Truncus arteriosus
- Ebstein anomaly
- Coarctation of the aorta

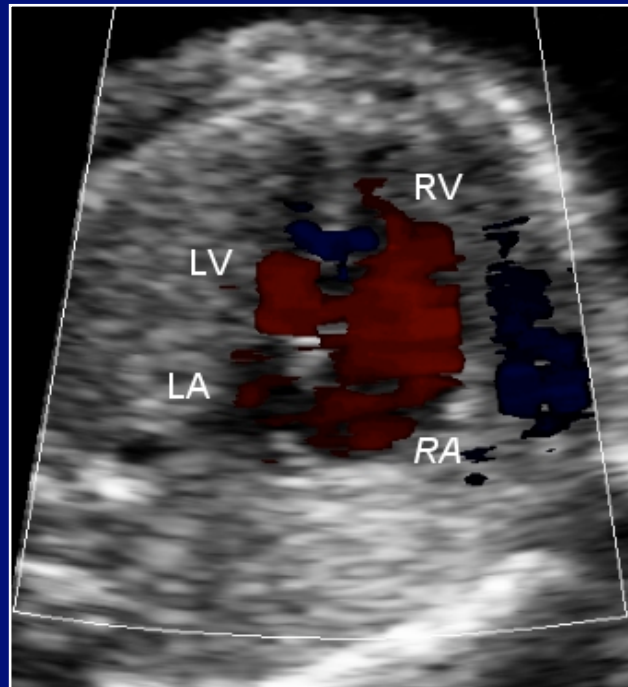
Ventricular Septal Defect (VSD)

- Most frequent cardiac defect (20-30%) [4]
- IV septum: inlet, trabecular, infundibular and membranous portions (muscular and membranous)
- Maldevelopment of muscular septum or endocardial cushion; improper resorption of the muscular ridge
- Left to right shunt

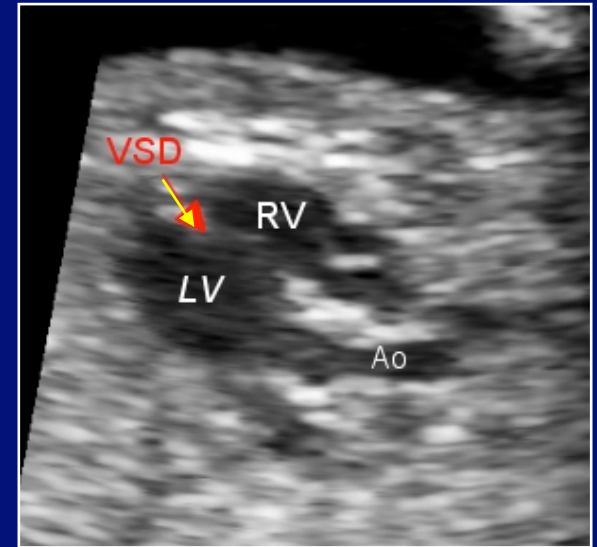
VSD



4 Chamber View

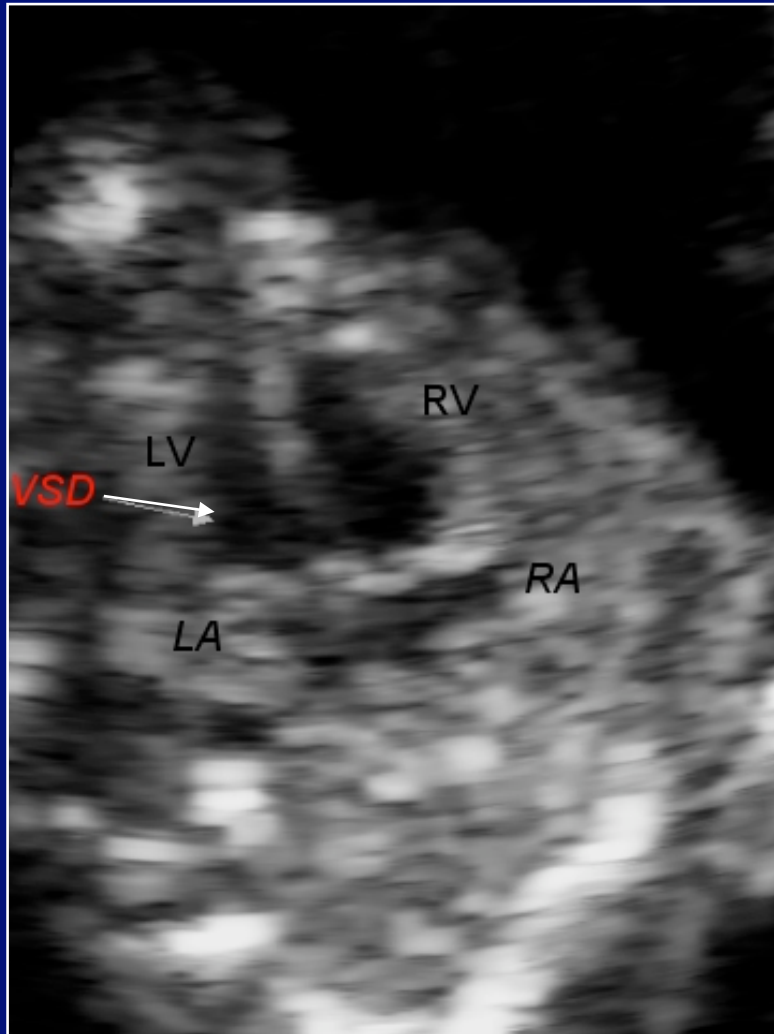


4 Chamber View

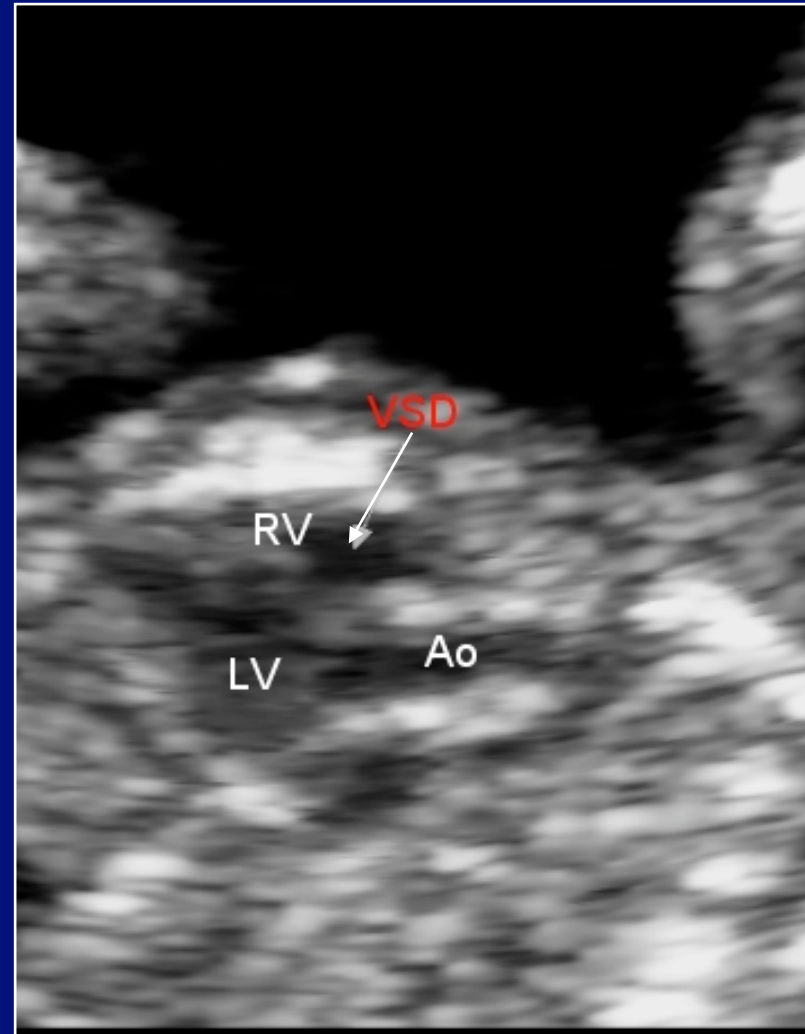


LVOT

VSD

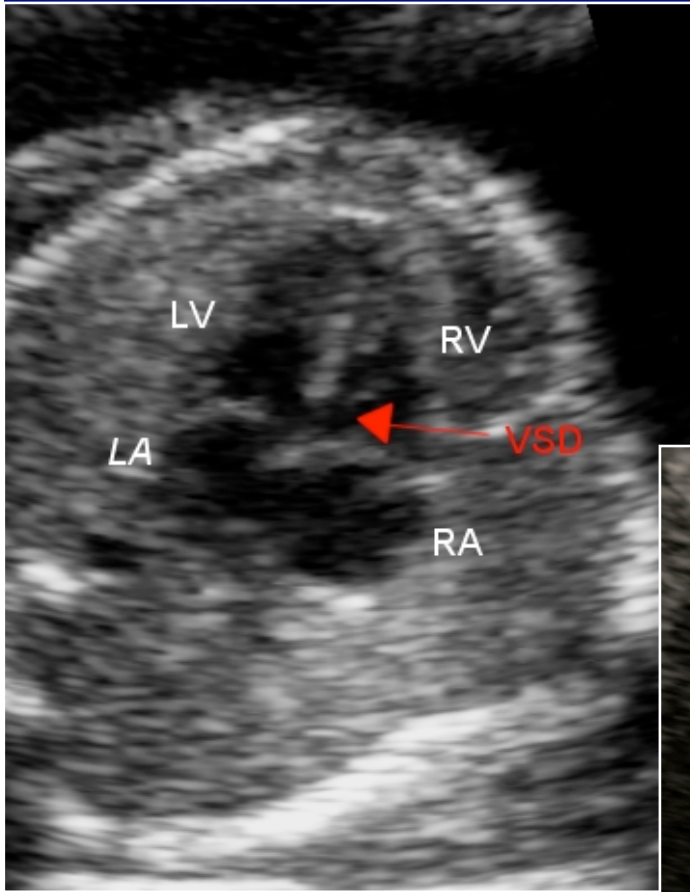


4 Chamber

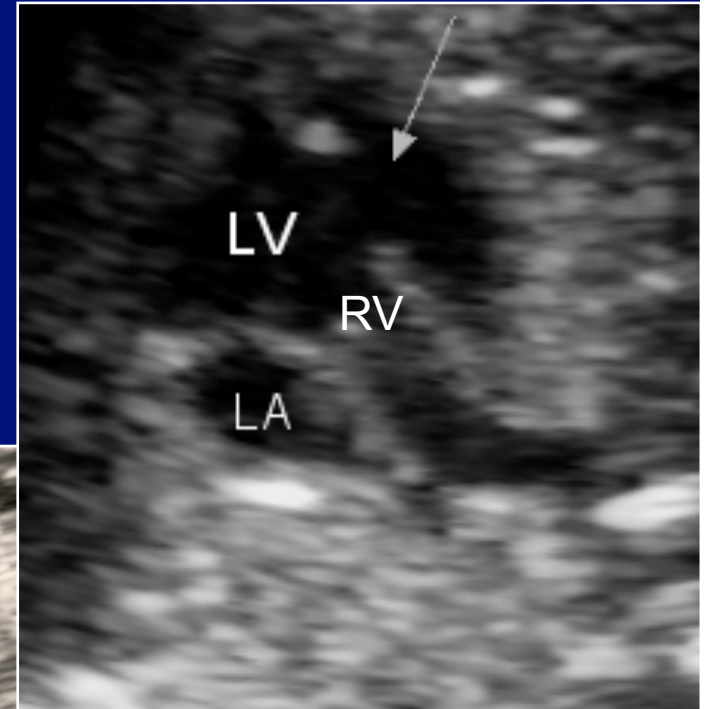


LVOT

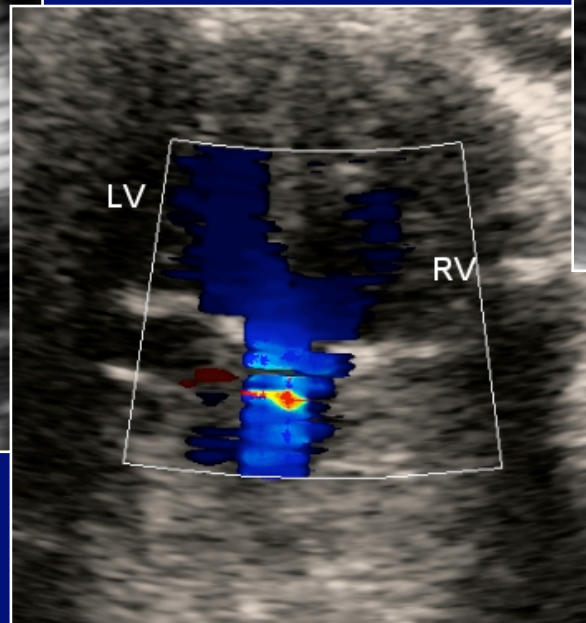
VSD



4 Chamber

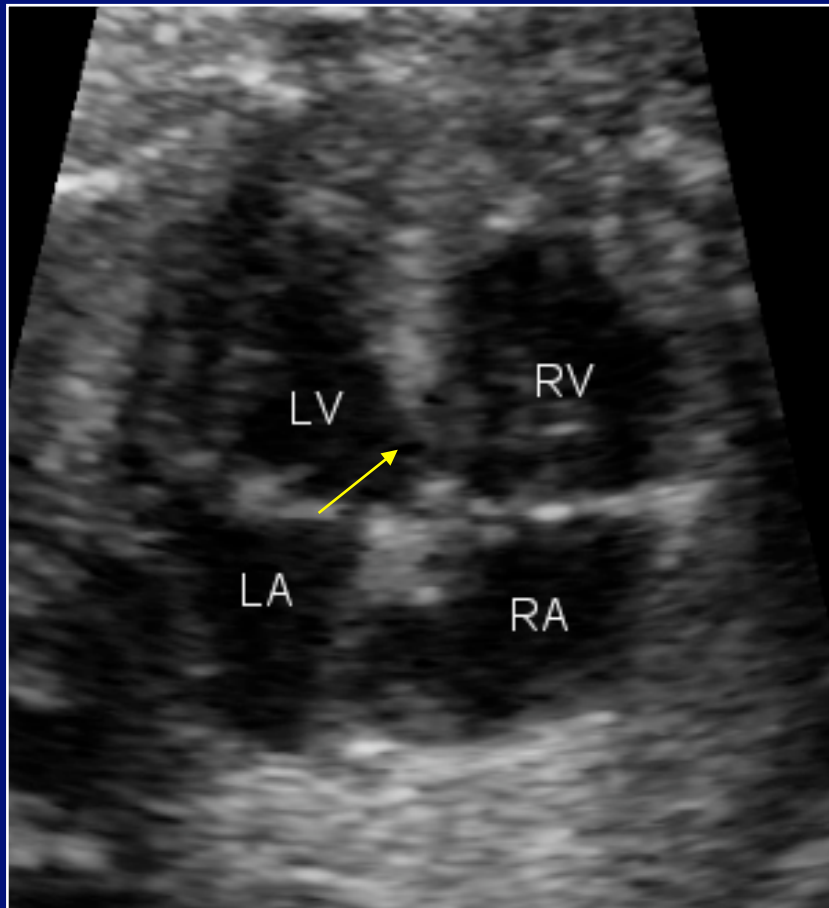


LVOT

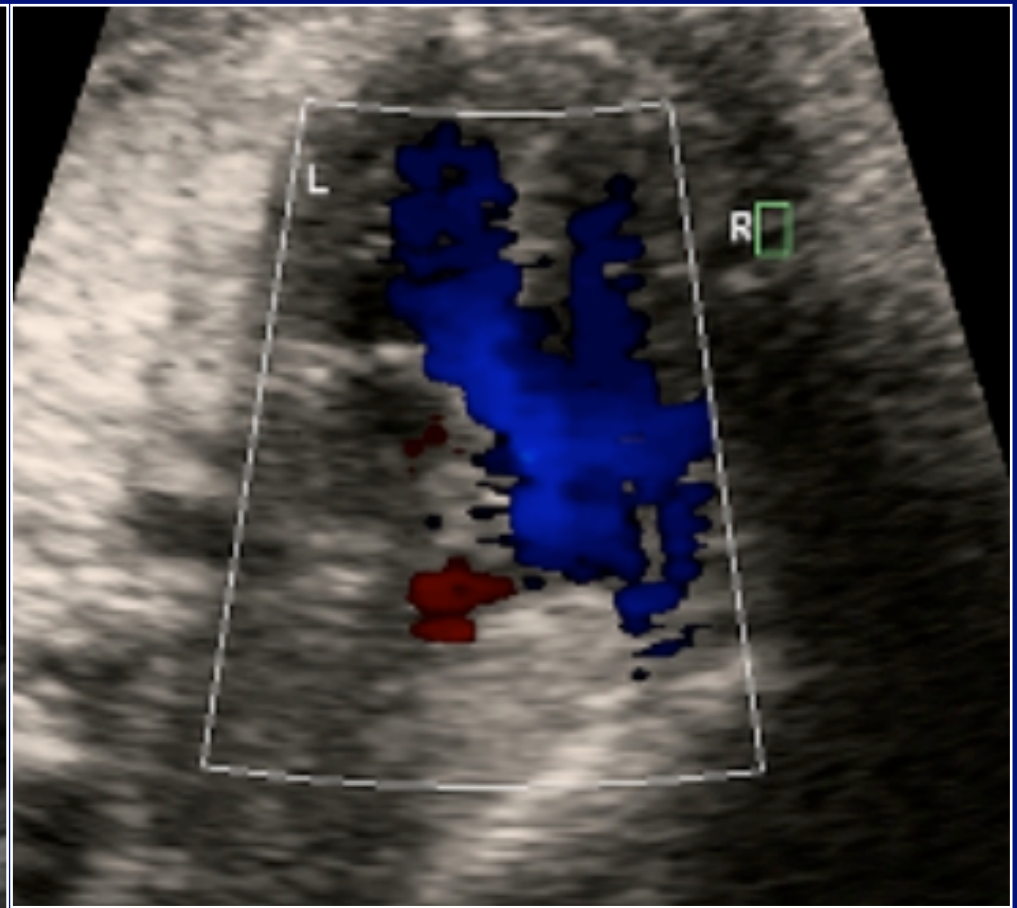


4 Chamber

VSD



4 Chamber



Color
(left to right shunt)

VSD video clips



click image to play video

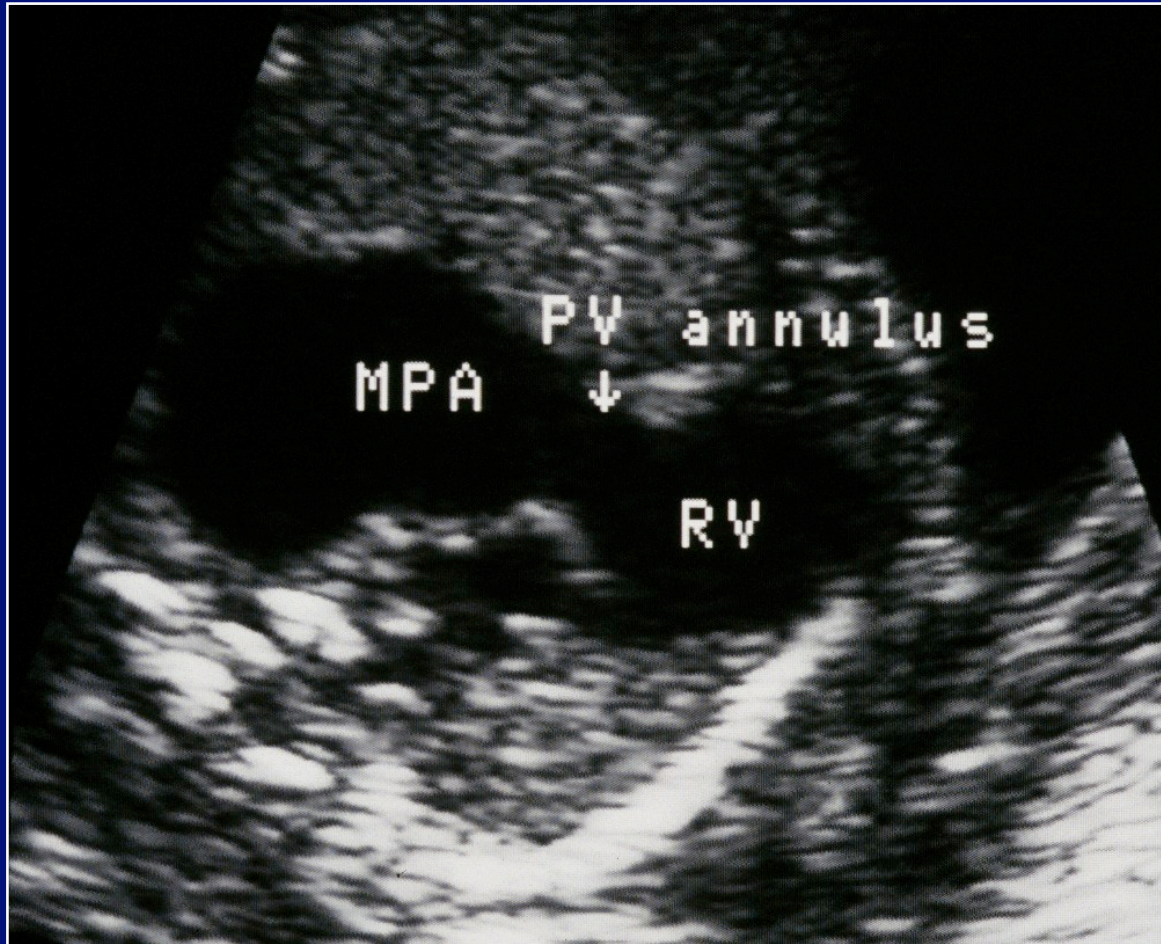
Tetralogy of Fallot

- VSD, infundibular pulmonic stenosis, over-riding aorta, RV hypertrophy
- 5-10% of CHD ^[1]
- Frequently not visualized on four chamber view (aside from VSD)

Tetralogy of Fallot

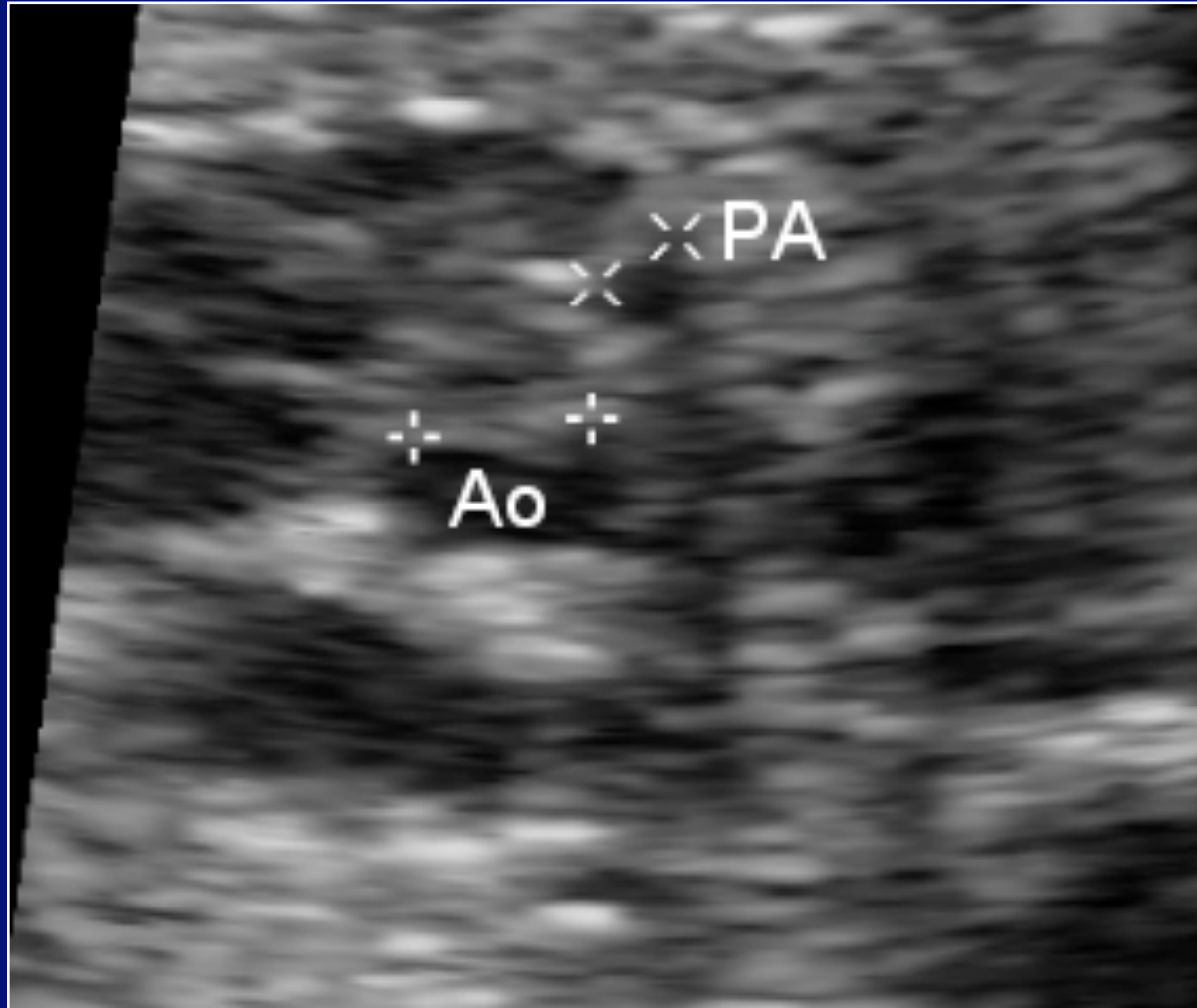
- Associated with chromosome anomalies (12-50%) [1]
- Recurrence risk = 2.5% (1 sib), 8% (2 sibs) [4]
- Cause = unequal division of the conus into smaller RV portion and larger LV portion [4]

Tetralogy of Fallot



Courtesy of Dr Mark Skalansky

Tetralogy of Fallot



Caliber of aorta > PA

Tetralogy of Fallot video



click image to play video

Courtesy of Dr Alfred Albahamed

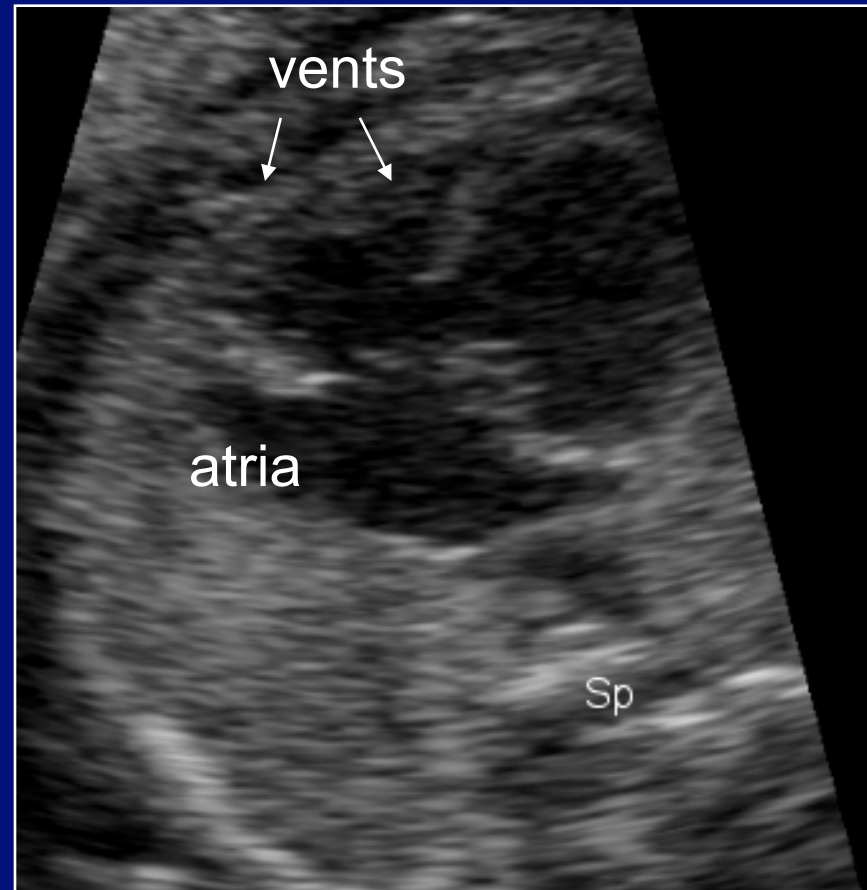
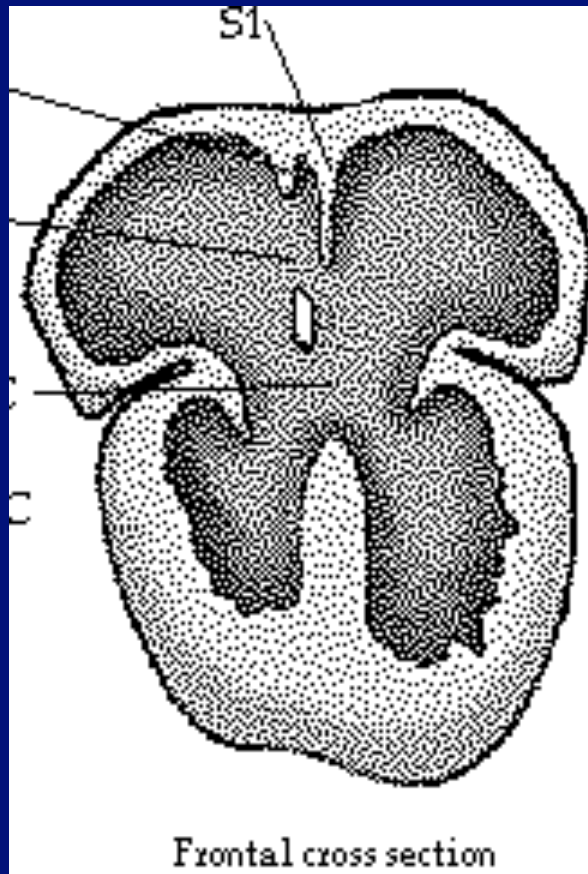
Endocardial Cushion Defect

- 5% of CHD
- Recur risk = 3% (1 sib), 10% (2 sibs), 1% (dad), 14% (mom)
- Freq assoc with other anomalies; strong assoc with Trisomy 21
- Large defect at the crus of heart on four chamber view

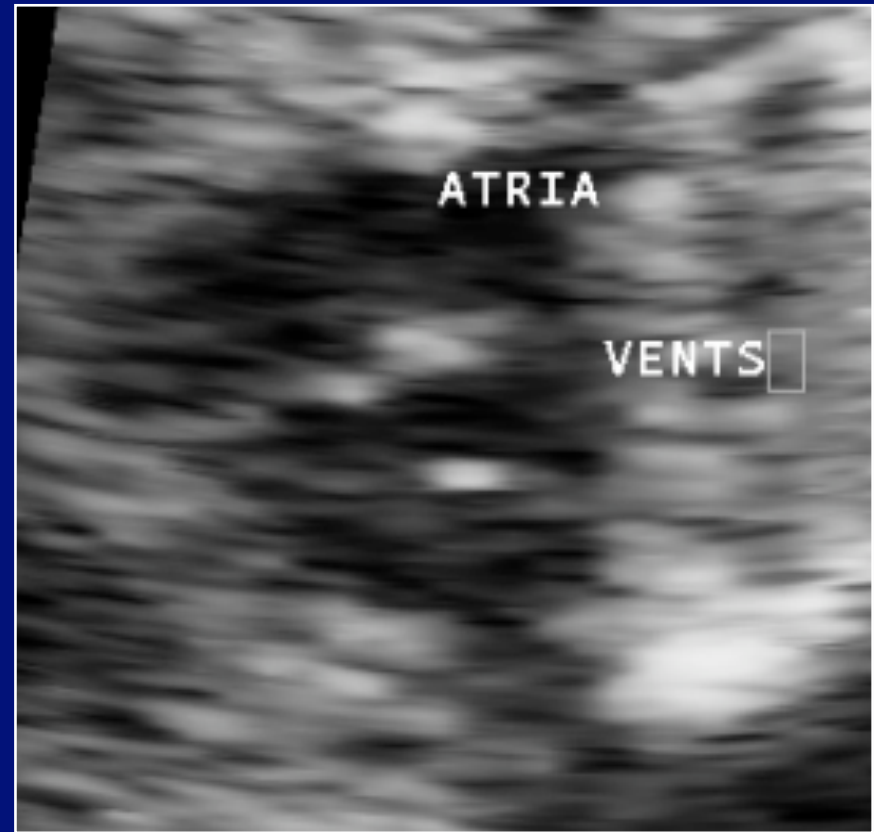
Endocardial Cushion Defect

- Several types depend on how AV valves attach. Most common is type III, complete AV canal and common AV valve
- Endocardial cushions fail to fuse; cause defect in both the atrial and ventricular septae (AV canal)

Endocardial Cushion Defect

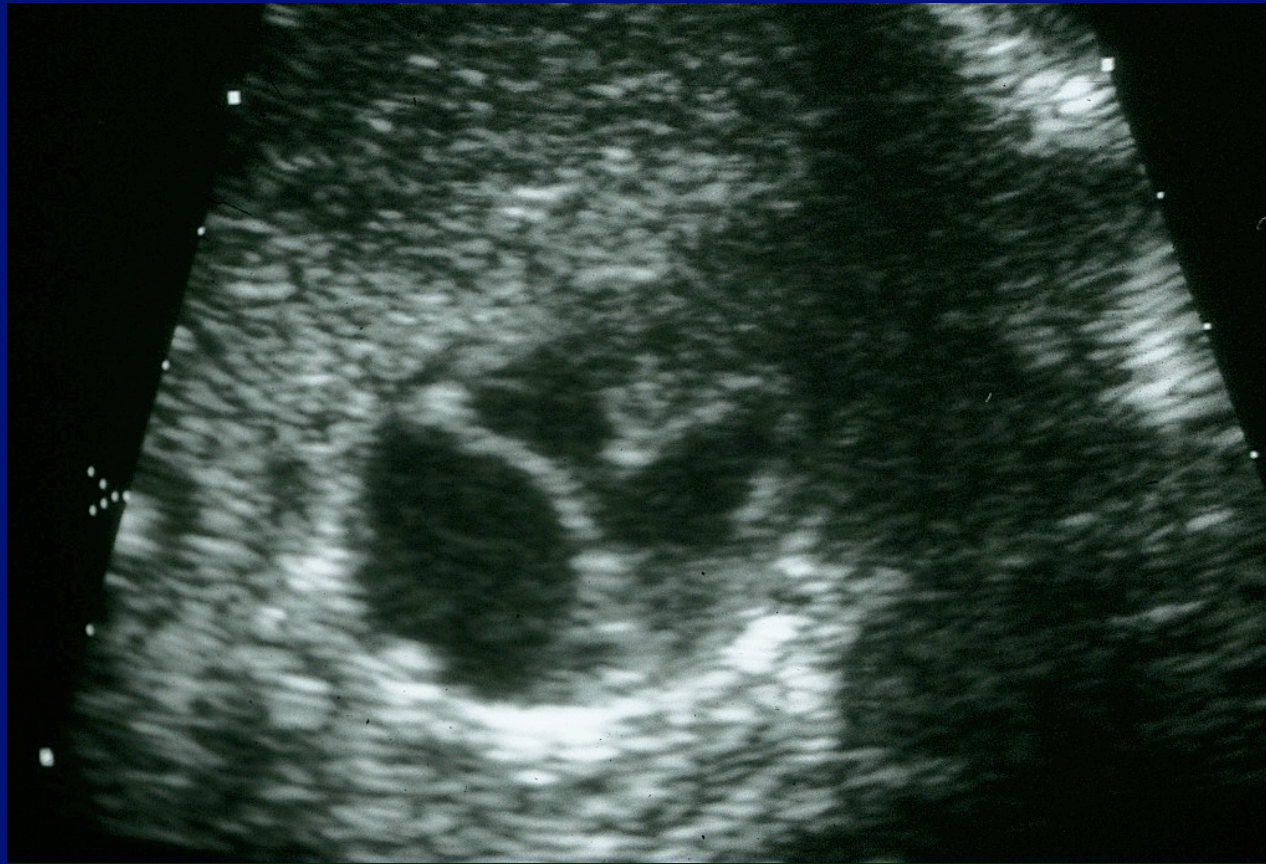


Endocardial Cushion Defect



4 Chamber View

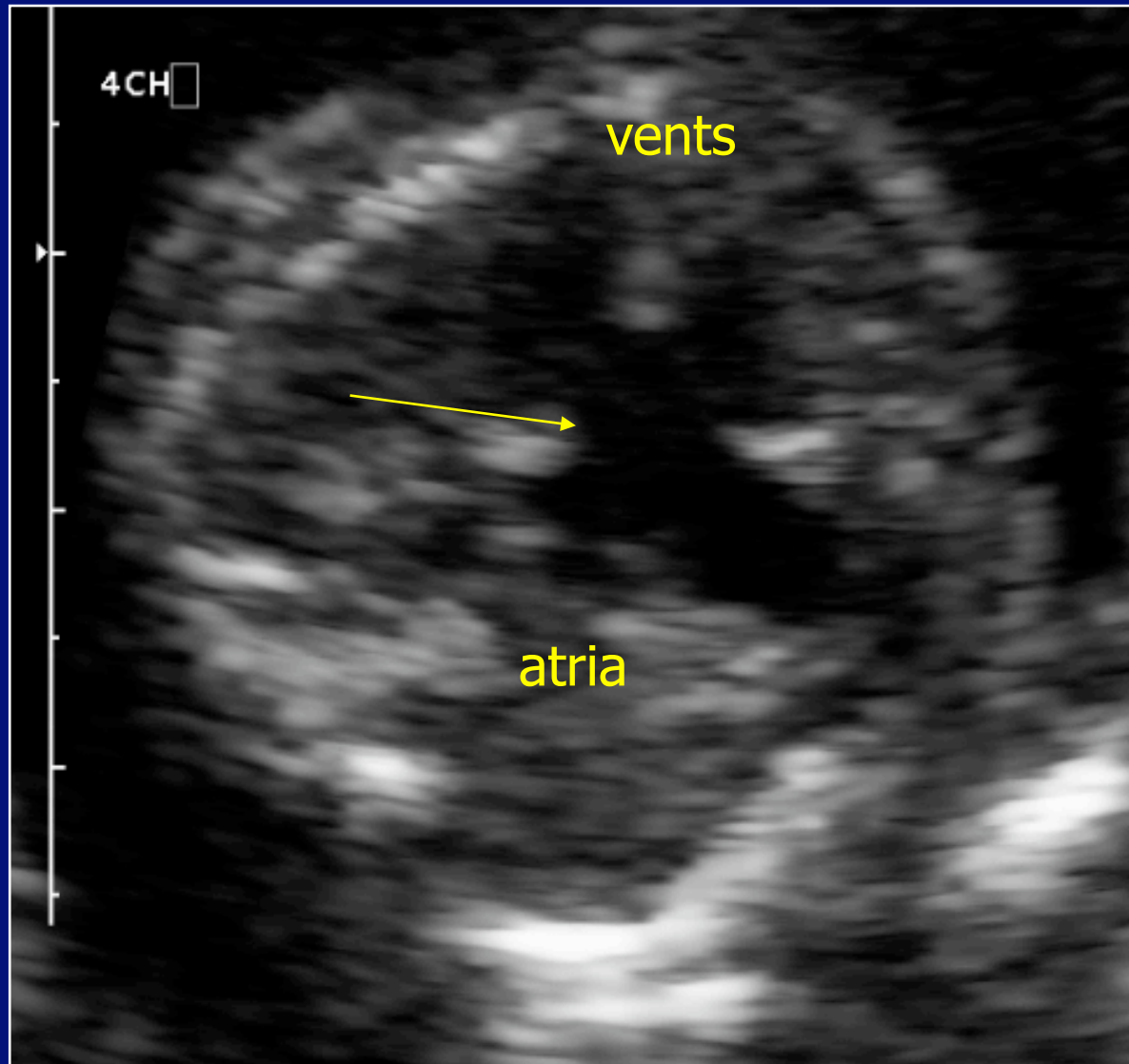
Endocardial Cushion Defect



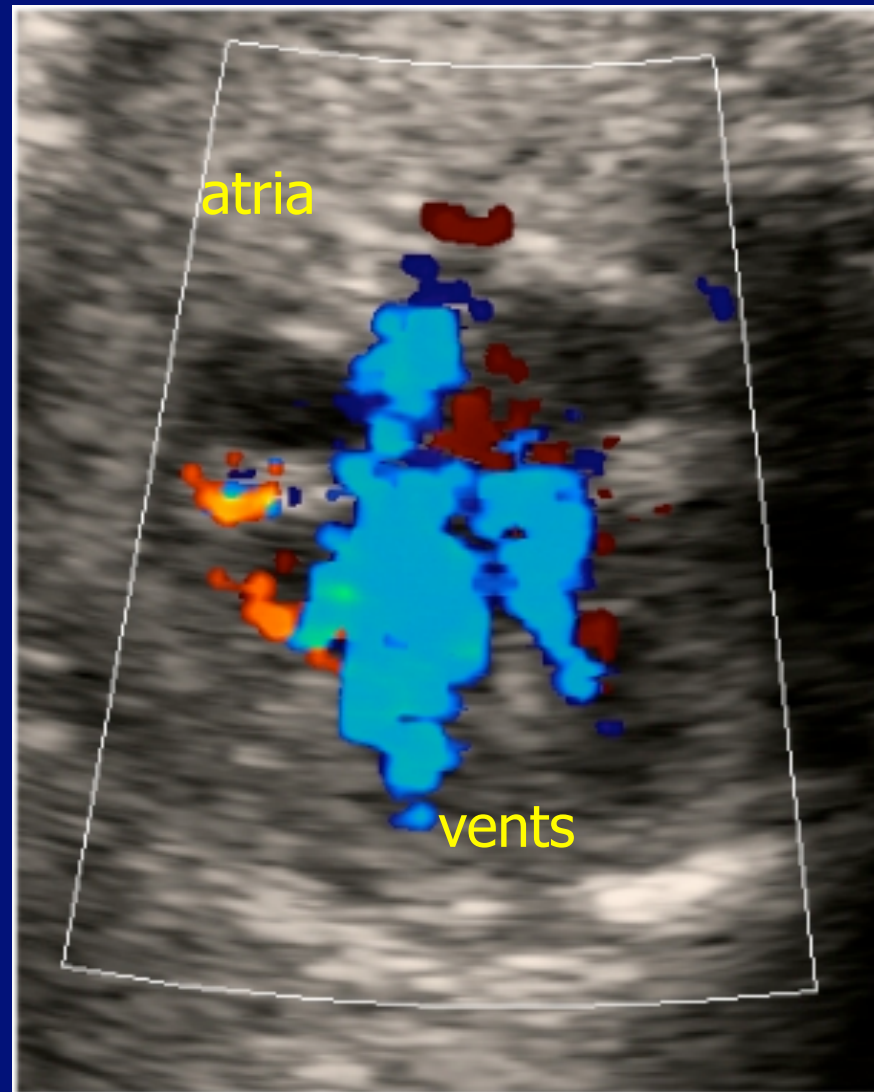
4 Chamber View

Courtesy of Dr Mark Skalansky

Endocardial Cushion Defect



Endocardial Cushion Defect

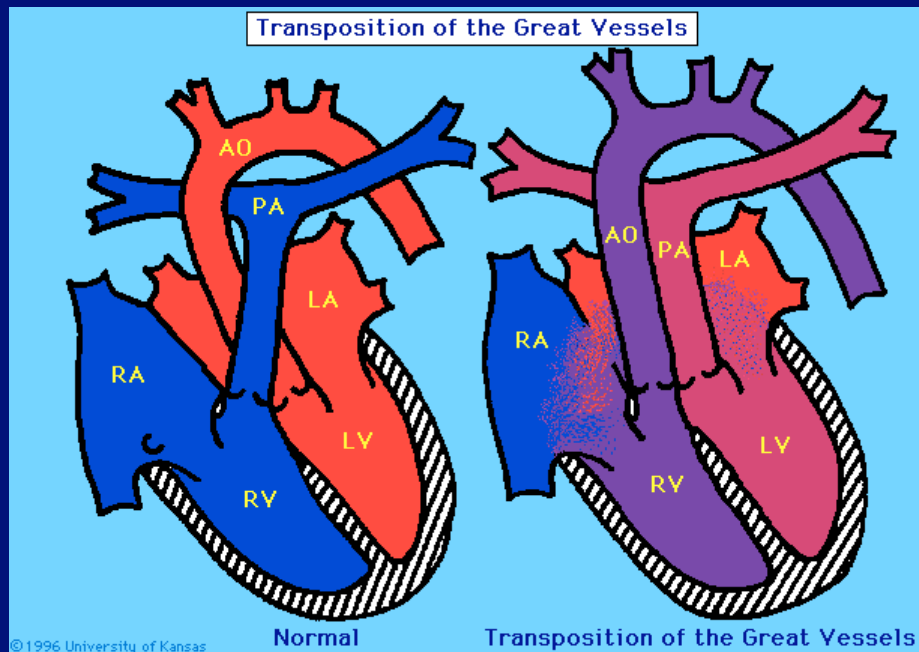


Endocardial Cushion Defect video



click image to play video

Transposition of Great Vessels



- D-type (“complete”) = heart tube loops to the right... normal orientation of ventricles; but vessels malform (cyanosis)
- L-type (“congenitally corrected”) = heart tube loops to the left... ventricular inversion, but normal vessel orientation (no cyanosis)

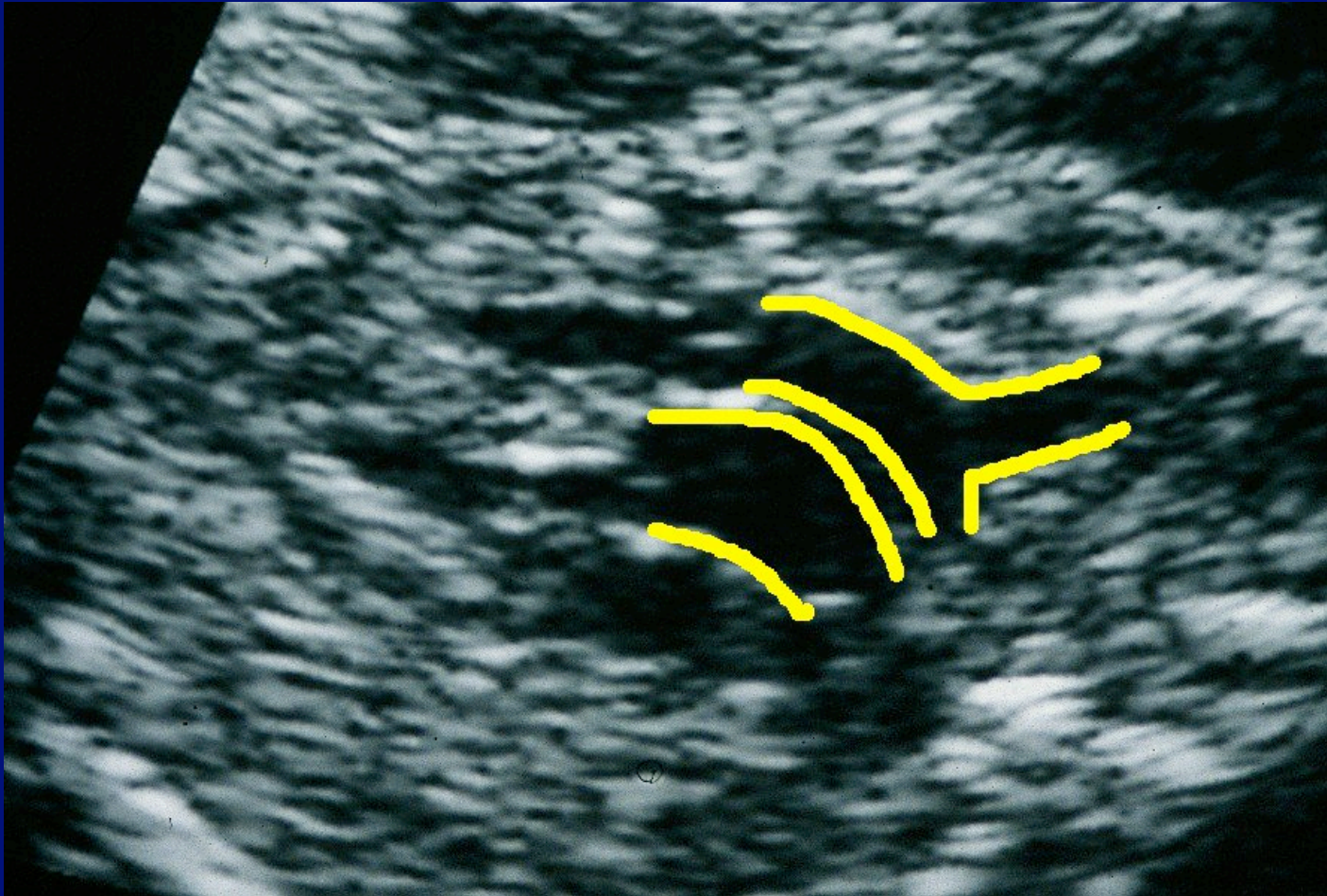
Transposition of the Great Vessels

- 4-6% of CHD
- Recurrence risk = 1.5% (1 sib), 5% (2 sibs)
- Association with excessive vitamin A
- Rarely associated with chromosome abnormalities [4]

Transposition Great Vessels

- VSD (30-50%)
- Aorta arises anterior and to the right of PA; great vessels parallel each other
- 5 year survival (surgery) 90%

Transposition Great Vessels



Courtesy of Dr Mark Skalansky

Transposition Great Vessels video



click image to play video (video starts slowly)

Courtesy of Dr Alred Albufamed

Hypoplastic Left Heart

- 2-4% of CHD [3]
- Hypoplasia of the LV; MV / AV stenosis or atresia
- Assoc with coarctation of aorta (80%) when AV atretic
- Hypothesis: low blood flow to LV causes hypoplasia

Hypoplastic Left Heart

- Causes hypoperfusion of coronary arts; CHF from ischemia
- Cyanosis at birth if LA to RA shunt not adequate
- Associated with chromosomal anomalies = 16% (one half from Trisomy 18 alone) ^[4]

Hypoplastic Left Heart

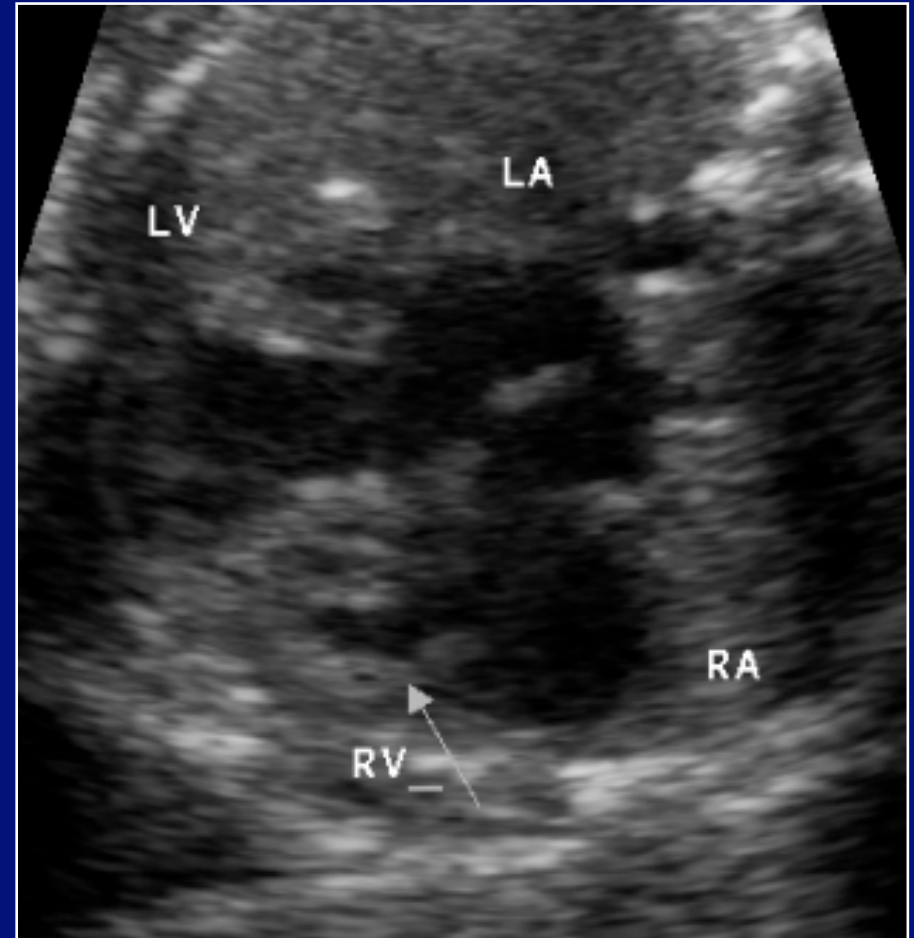
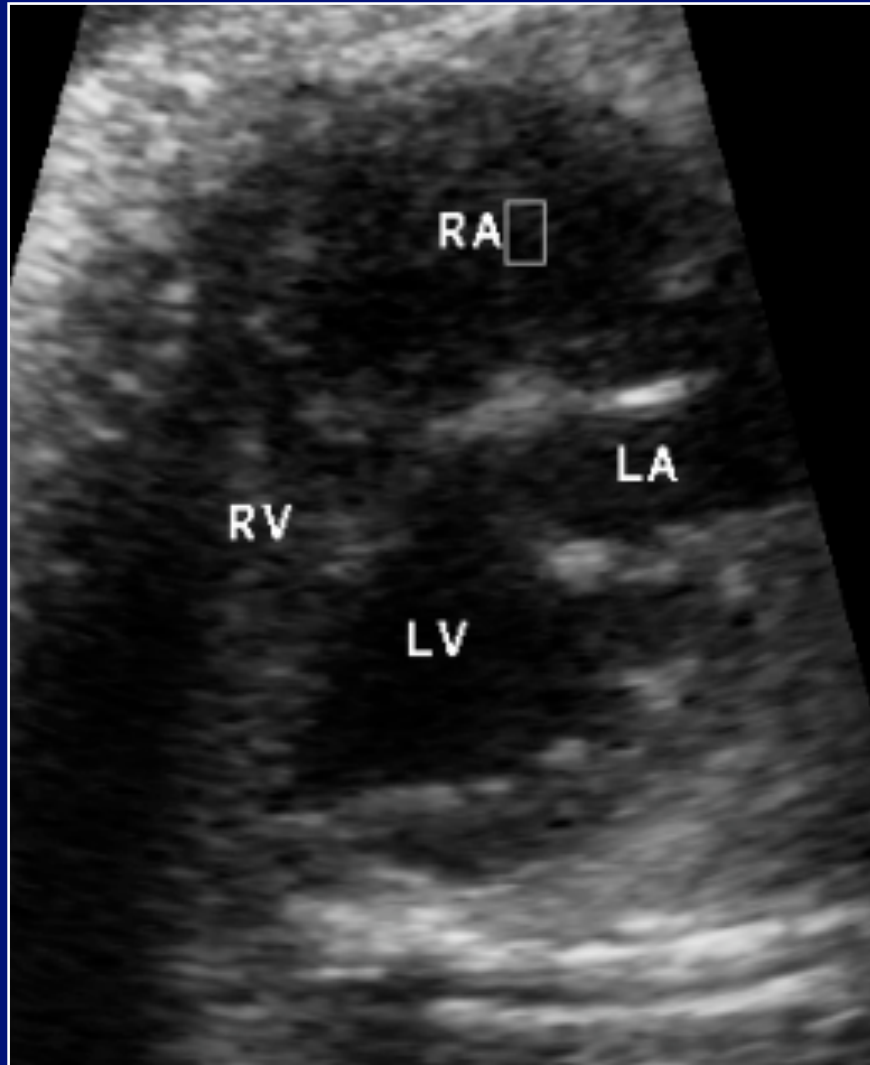


4 Chamber View

Hypoplastic Right Heart

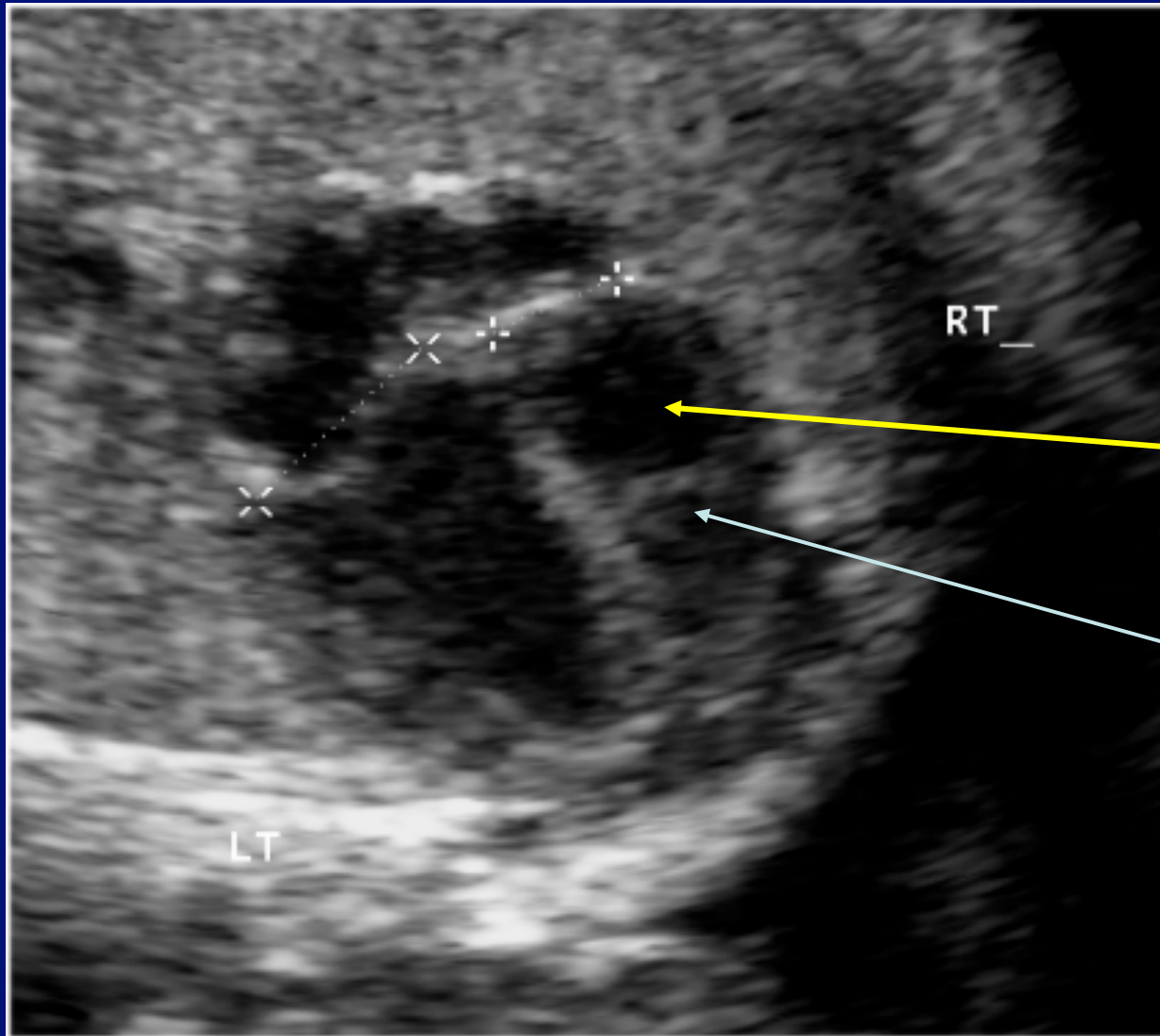
- Underdevelopment of right sided heart structures
- Hypoplasia of the RV and PA; RV wall thick; hypoplastic or incompetent TV; PV atresia
- Relies on patent ductus arteriosus for blood supply to lungs (PGE1)
- Tx = surgical systemic to pulmonary shunt (Blalock Taussig Shunt)

Hypoplastic Right Heart



Small RV, thick wall

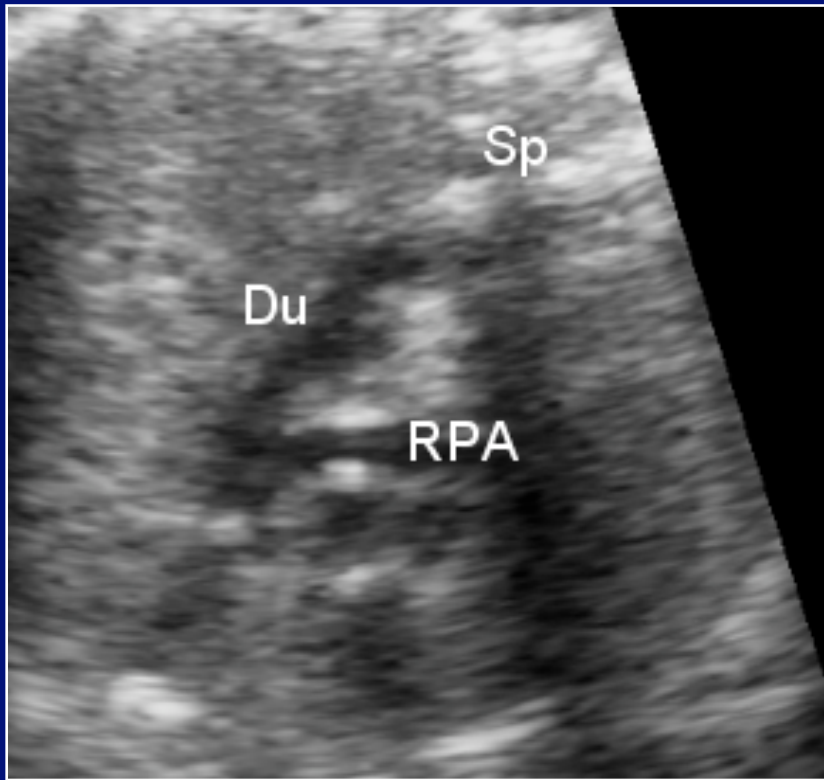
Hypoplastic Right Heart



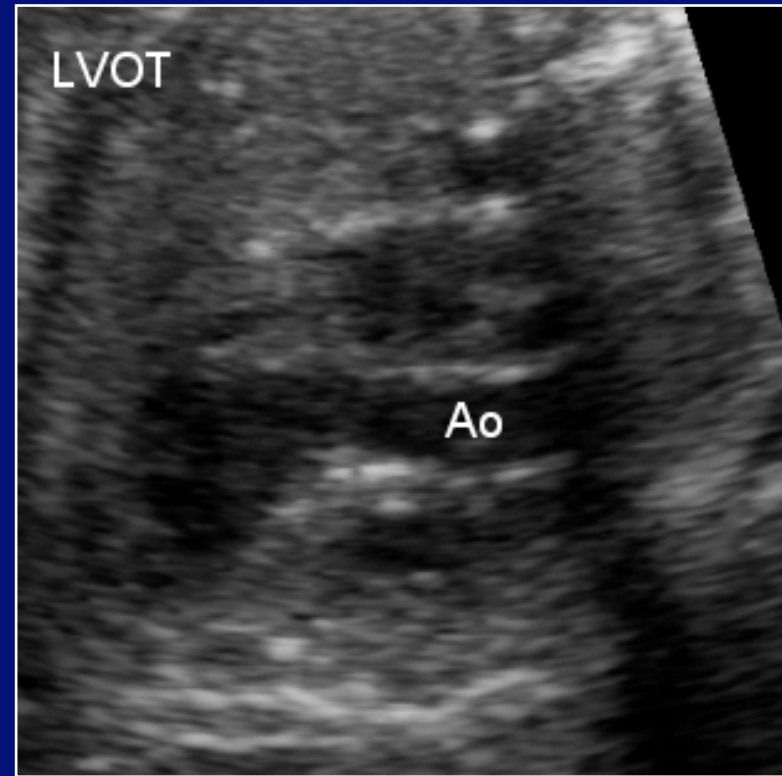
Small RV

Notice the
moderator band

Hypoplastic Right Heart



RVOT



LVOT

Caliber of PA < aorta

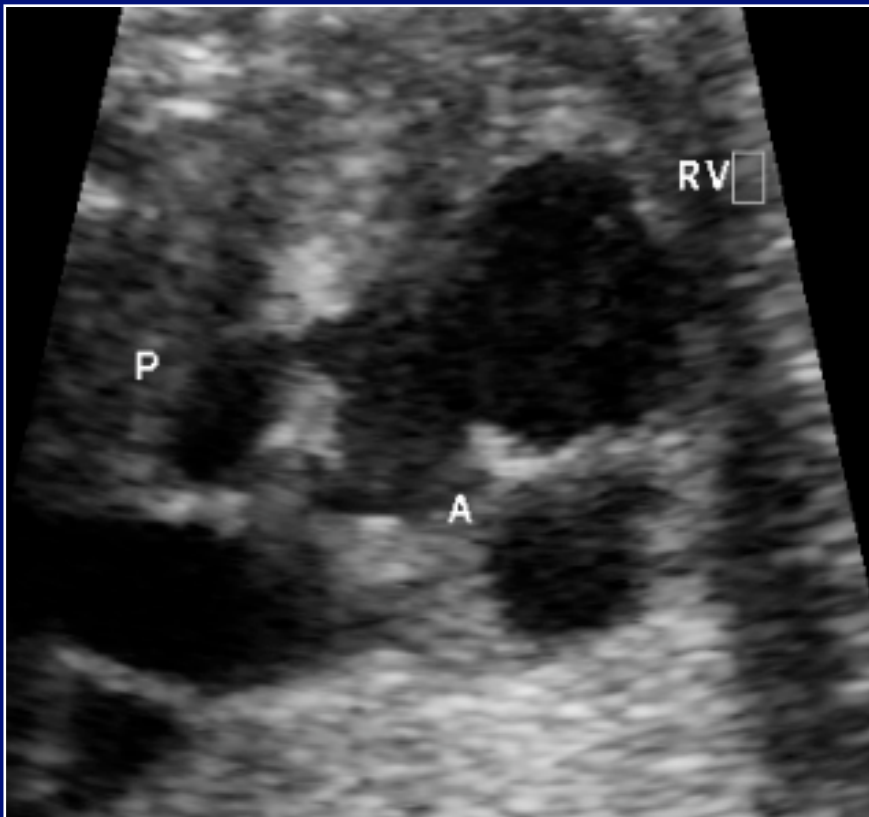
Double Outlet Right Ventricle

- 1% of CHD
- Recurrence risks not defined
- Both PA and at least half of the aorta originate from RV
- Karyotype abnormalities = 5%; including Trisomy 13,18 ^[4]

Double Outlet Right Ventricle

- Associated with VSD, ascending aortic stenosis, pulmonic stenosis, AV abnormalities
- Complex embryologic changes involving bulbus cordis (RV), conus cordis (septum), and truncus arteriosus (great vessels)

Double Outlet Right Ventricle



Both the PA and aorta (more than 50%) arise from the RV

Double Outlet Right Ventricle video



click image to play video

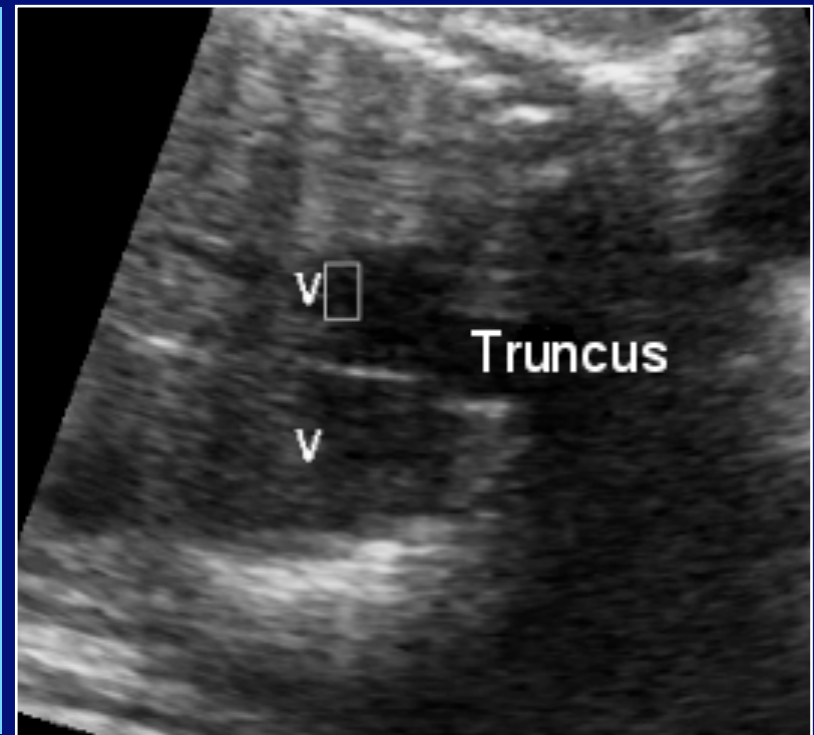
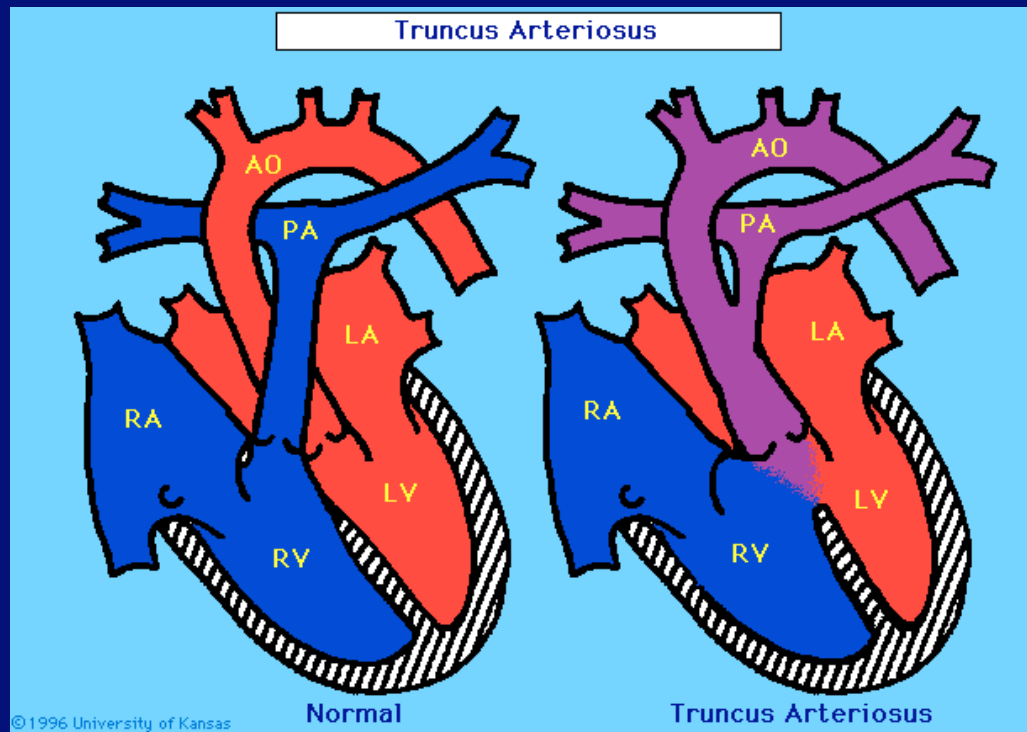
Truncus Arteriosus

- <1% CHD
- Single arterial vessel “truncus” arises from the heart and supplies the coronary arteries, pulmonary and systemic circulations
- Recurrence risk = 1% (1 sib), 3% (2 sibs) [4]

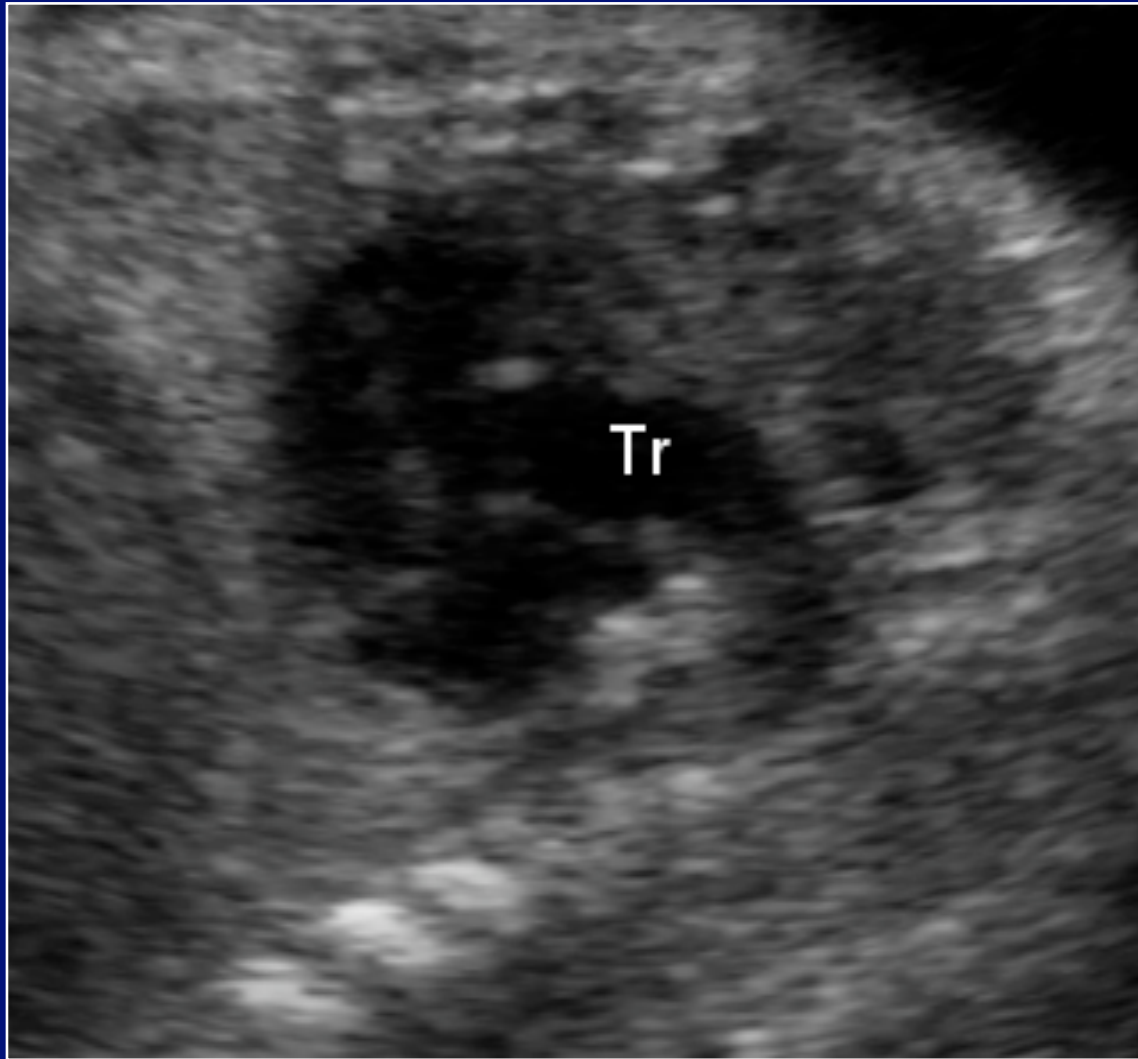
Truncus Arteriosus

- Other associated pathology: VSD, abnormal tricuspid valve, ASD...
- 20% overall mortality (surg < 6mo to avoid pulmonary HTN)
- Differentiate from tetralogy of Fallot = no RVOT; look for origin of PAs from truncus

Truncus Arteriosus



Truncus Arteriosus



Truncus Arteriosus

Four Types (Van Praagh Classification)

1. Main PA arises from truncal root and divides
2. Both PAs arise from the truncal root separately
3. Left PA supplied by collaterals from aortic arch
4. Aortic arch interrupted; desc aorta supplied by ductus (10-15%)

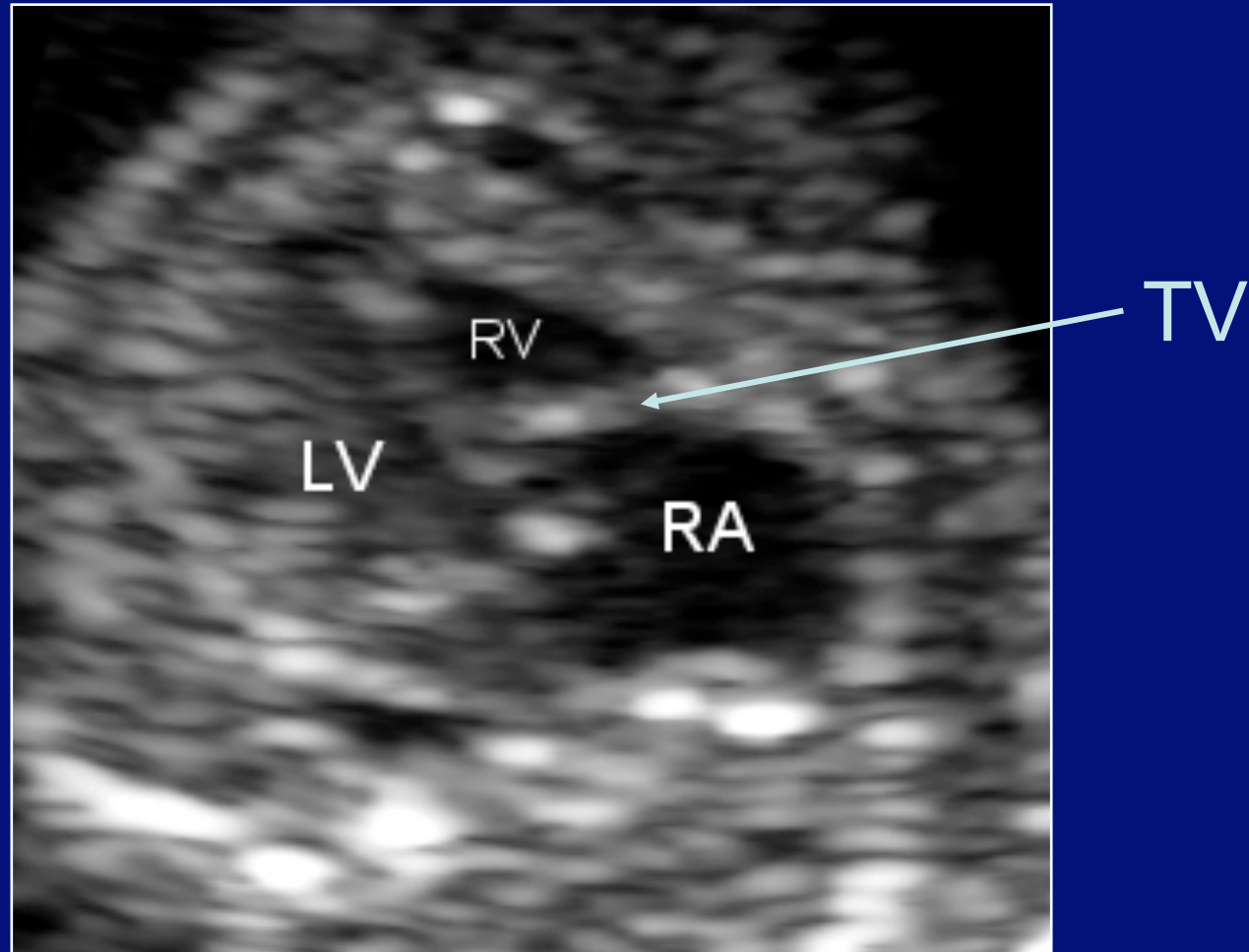
Ebstein Anomaly

- <1% CHD
- Recurrence risk = 1% (1 sib), 3% (2 sibs)
- Apical displacement of TV; small RV; RA grossly dilated because TV incompetent

Ebstein Anomaly

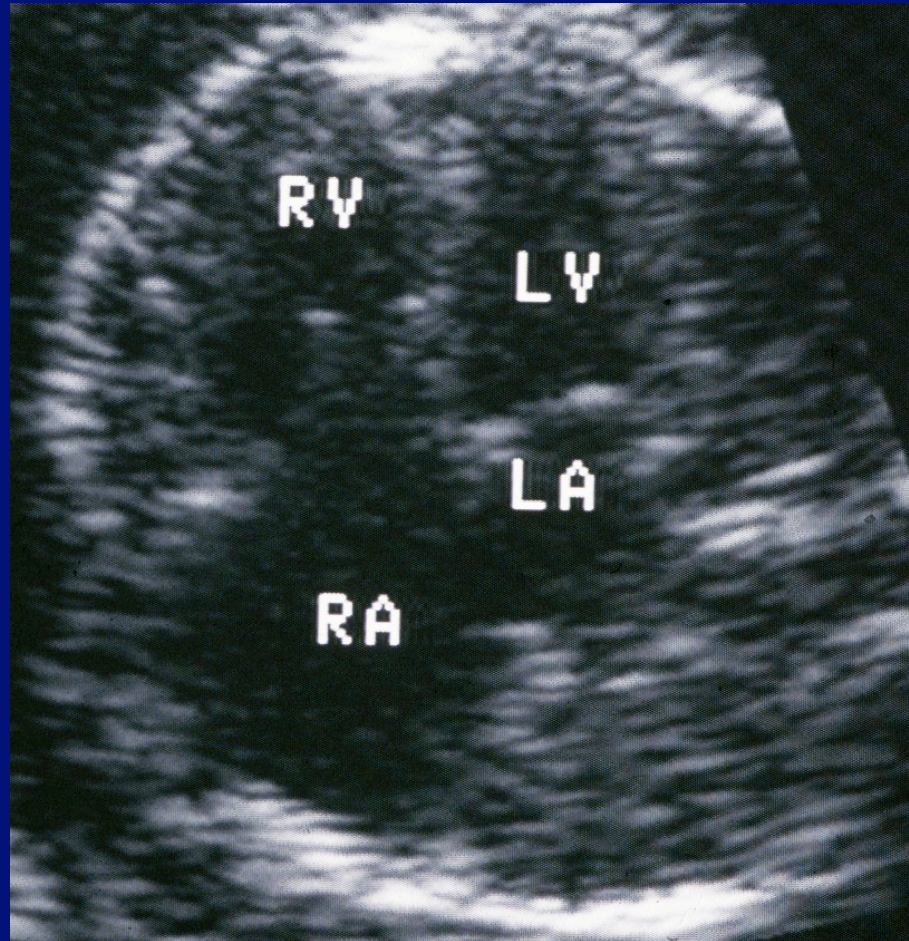
- Assoc with ASD, WPW syndrome (30%), RV outflow obstruction (PS); CHF in utero from TR ^[1]
- Surgery to replace TV ^[1] ; arrhythmia is frequent after correction (dilated RA)

Ebstein Anomaly



4 Chamber View

Ebstein Anomaly



Courtesy of Dr Mark Skalansky

Ebstein Anomaly



click image to play video

Courtesy of Dr Alfred Albafamed

Coarctation of Aorta

- <1% CHD
- Recurrence risk = 2% (1 sib), 6% (2 sibs)
- When ductus closes, tissue at insertion on proximal desc aorta constricts (post ductal coarctation)
- 6th arch forms ductus arteriosus; 4th arch forms transverse aorta

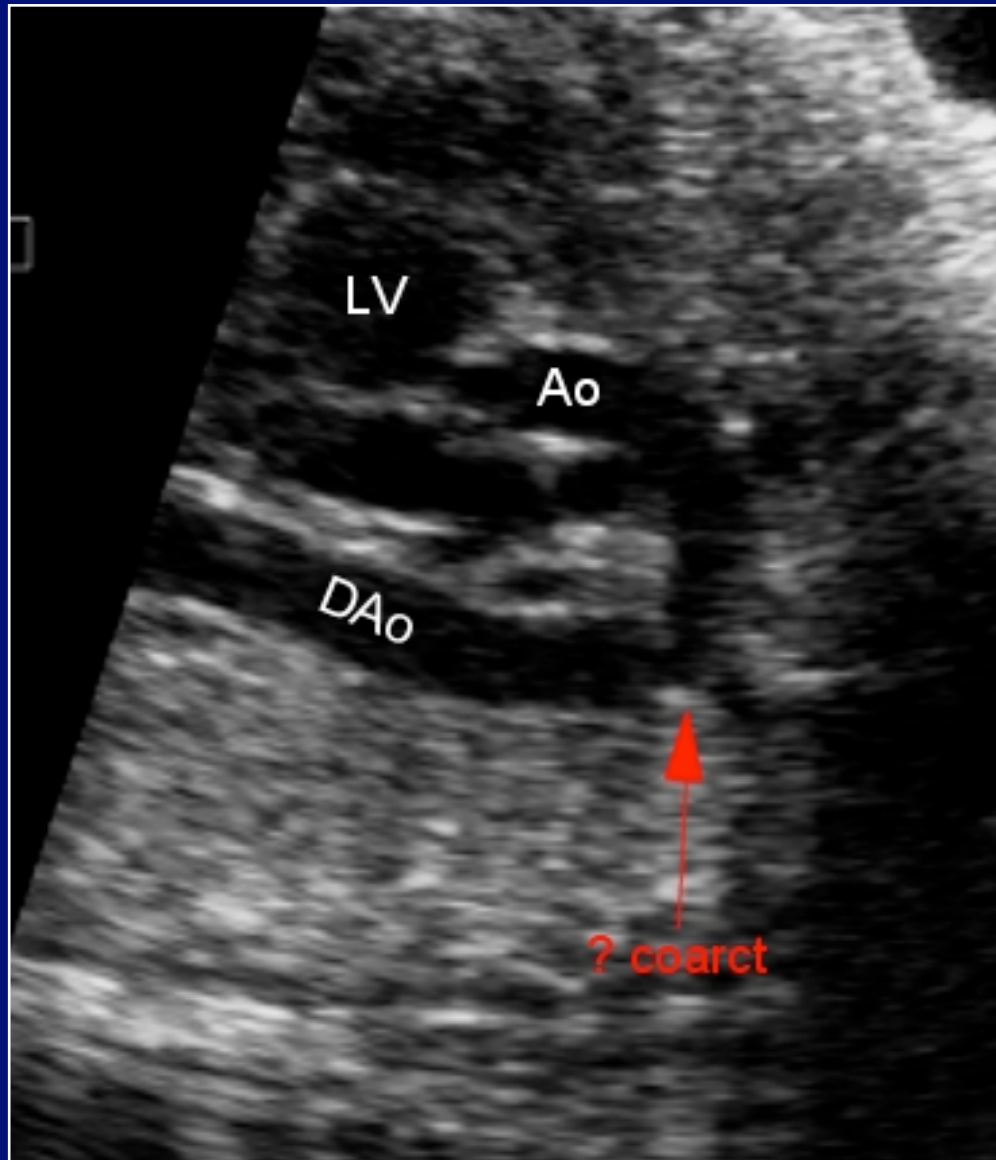
Coarctation of Aorta

- Narrowing just distal to takeoff of left subclavian artery
- Hypoplasia of 4th arch
- Visualization of arch is insensitive
- Large RV and PA relative to LV and Aorta

Coarctation of Aorta

- Flow across TV / MV $> 2 / 1$
(normal $< 1.8 / 1$)
- Assoc w/ other cardiac defects =
bicuspid aortic valve, VSD, ASD...
- 10% association with chromosomal
anomalies (especially Turner's)

Coarctation of Aorta

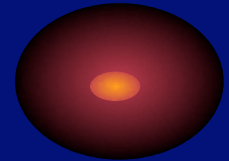
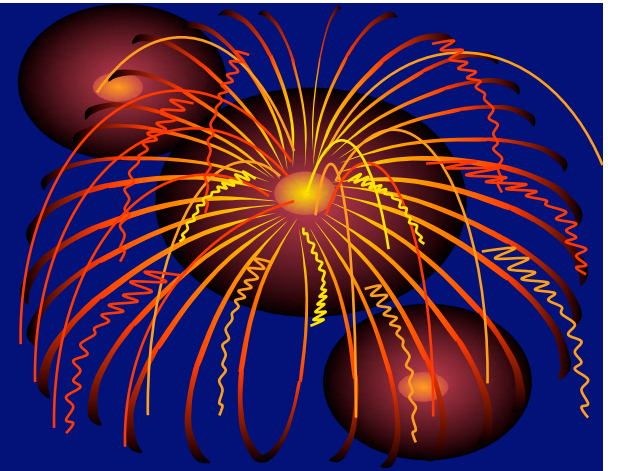


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THANK YOU !

