Miscellaneous Cardiac Diseases

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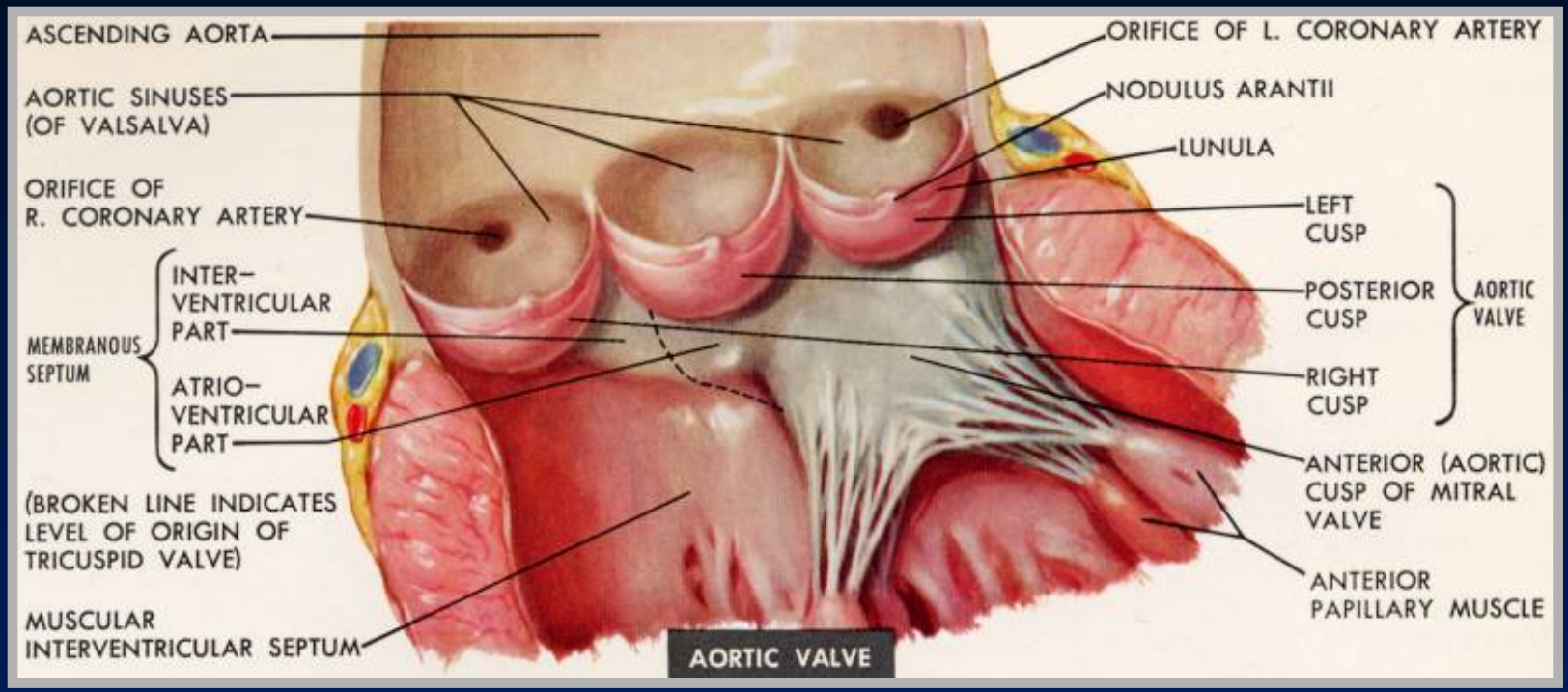
Sinus of Valsalva Aneurysm



Types

- Congenital
- Inherited
- Acquired

Sinus of Valsalva Aneurysm
Congenital

- Usual type
- Involves a single cusp
- Most often arise from R coronary sinus

Sinus of Valsalva Aneurysm
Inherited

- Associated with Marfan's Disease
- All cusps involved
- Produce aortic regurgitation

Sinus of Valsalva Aneurysm
Acquired

- Usually 2° to endocarditis of aortic valve
- Other causes
  - Syphilis
  - Atherosclerosis
  - Dissecting aneurysm
  - Marfan's

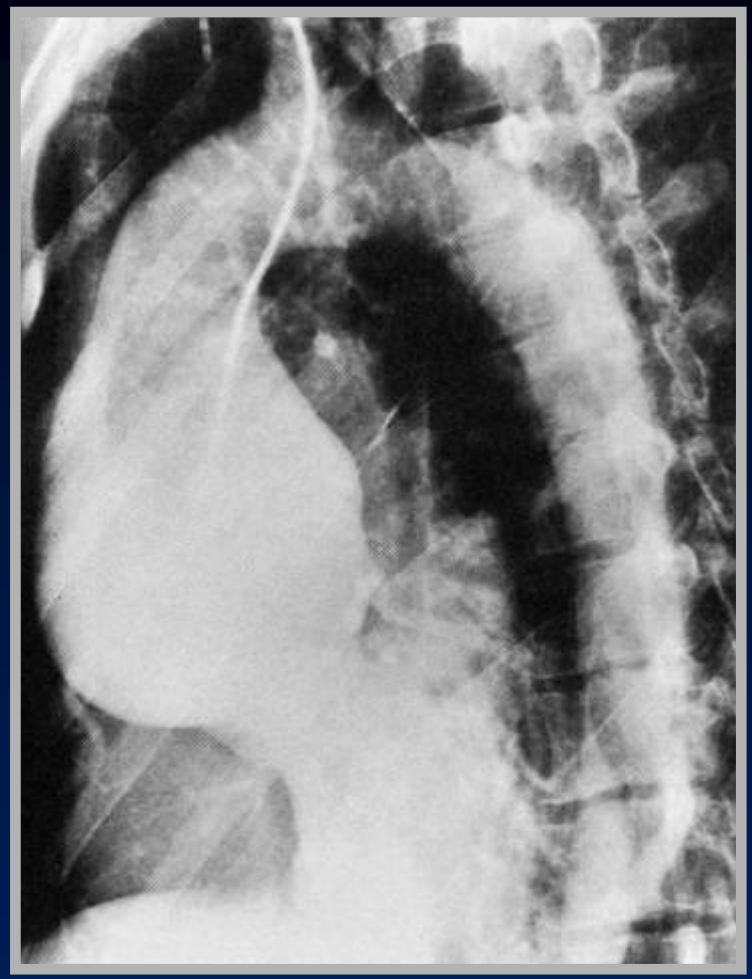
Sinus of Valsalva Aneurysm
X-ray Findings

- Since aortic root is intracardiac, usual aneurysm is not visible
- Rarely, a large aneurysm of L aortic sinus → bulge L upper heart border in region of LA appendage

Sinus of Valsalva Aneurysm

Sinus of Valsalva Aneurysm
Other X-ray Findings

- Rarely, a large aneurysm of the R aortic sinus → bulge on R heart border
- Usually the aneurysm dilates the aortic ring → AI



Aneurysm of Sinus of Valsalva and Proximal Ascending Aorta = annuloaortic ectasia

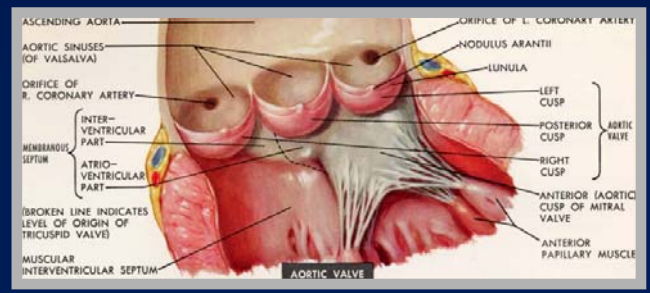
Ruptured Sinus of Valsalva Aneurysm

Ruptured Sinus of Valsalva Aneurysm Congenital vs. Acquired

- Congenital forms (usually R sided) always produce an intracardiac fistula
  - Most congenital aneurysms rupture during third or fourth decade of life
- Acquired forms can produce either intra- or extracardiac fistulae

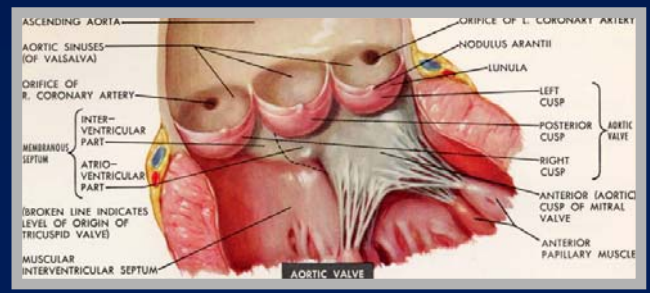
Ruptured Sinus of Valsalva Aneurysm
General

- May rupture → aortic-cardiac fistula
  - L → R shunt usually
- Most ruptures involve R coronary sinus
  - Into R ventricle
- Posterior (non-coronary) aortic sinus ruptures occasionally
  - Into R atrium



Ruptured Sinus of Valsalva Aneurysm
General

- Rupture of aneurysms of L sinus are very rare
  - May rupture into the pericardial space

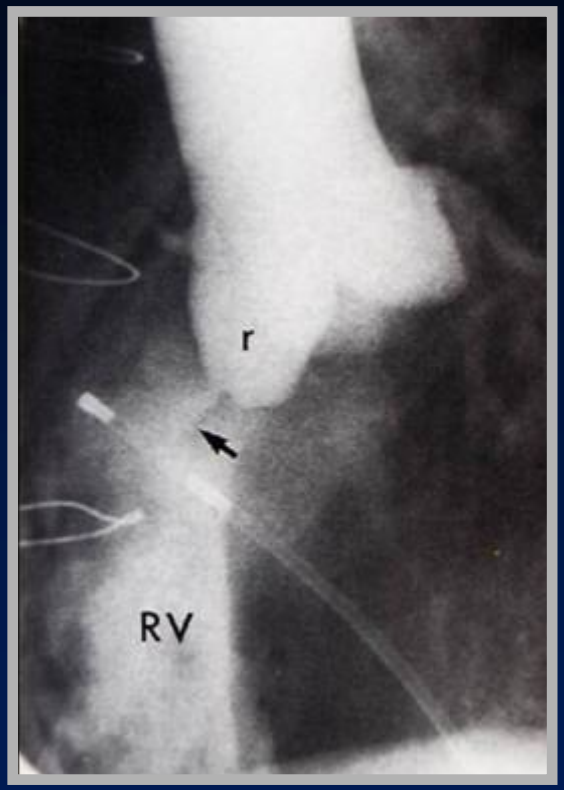


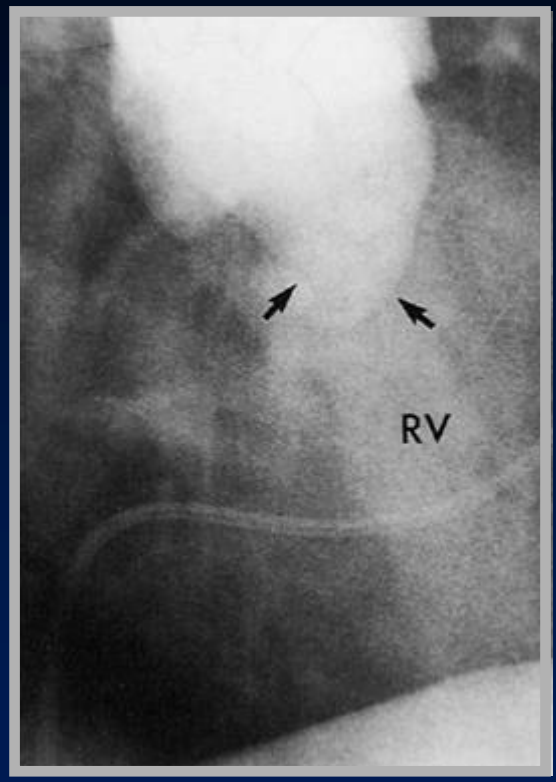
Ruptured Sinus of Valsalva Aneurysm Clinical

- Symptoms due to sudden onset of massive aortic regurg or L → R shunt
  - Acute onset of SOB
  - Chest pain
  - Acute onset of murmur
  - CHF
  - Death

Ruptured Sinus of Valsalva Aneurysm X-ray Findings

- Acute CHF
- Followed by L → R shunt
- With rupture of L sinus, LA may suddenly enlarge





Two cases of rupture of right coronary sinus into RV

Congenital Defect in the Pericardium

Congenital Pericardial Defect
Embryogenesis

- Premature atrophy of left duct of Cuvier (cardinal vein)
- Failure of nourishment of left pleuro-pericardial membrane ➜ failure of pericardium to develop

Congenital Pericardial Defect
General

- Male:female ratio of 3:1
- May be detected at any age
  - Most common in low 20's

Congenital Pericardial Defect
Location

- Foraminal defect on left side 35%
- Complete absence of left side gives levoposition of heart 35%
- Diaphragmatic surface 17%
- Total bilateral absence 9%
- Right sided 4%

Congenital Pericardial Defect
Associations

- Bronchogenic cysts
- VSD, PDA, mitral stenosis
- Diaphragmatic hernia
- Sequestration

Congenital Pericardial Defect
Clinical

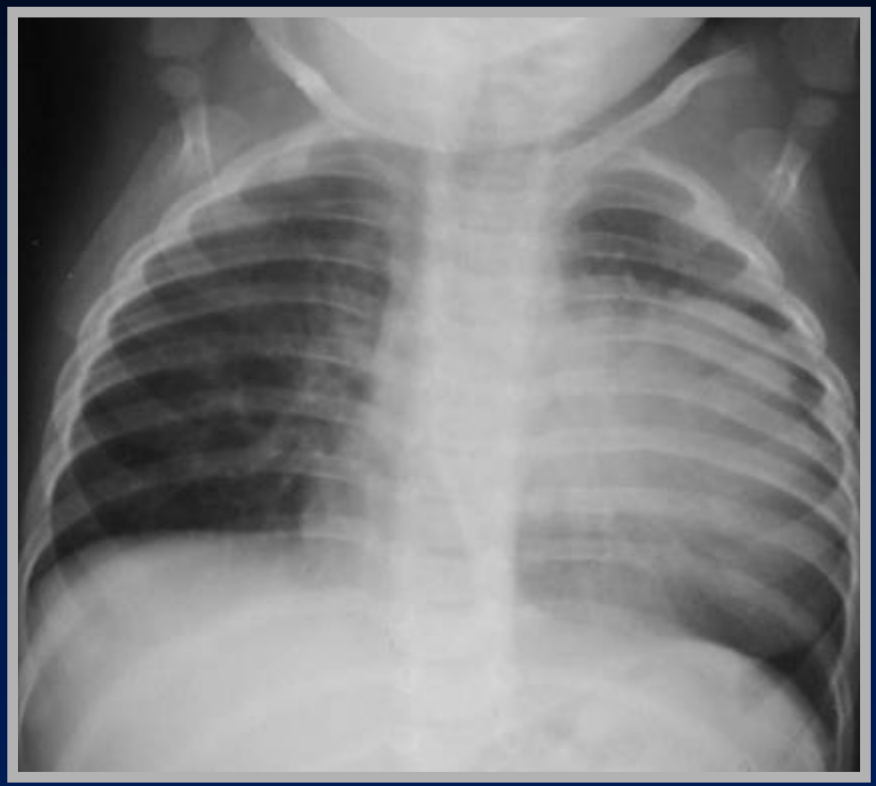
- Mostly asymptomatic
- May have:
  - Tachycardia
  - Palpitations
  - Right bundle block
  - Positional discomfort lying on left side
  - Chest pain

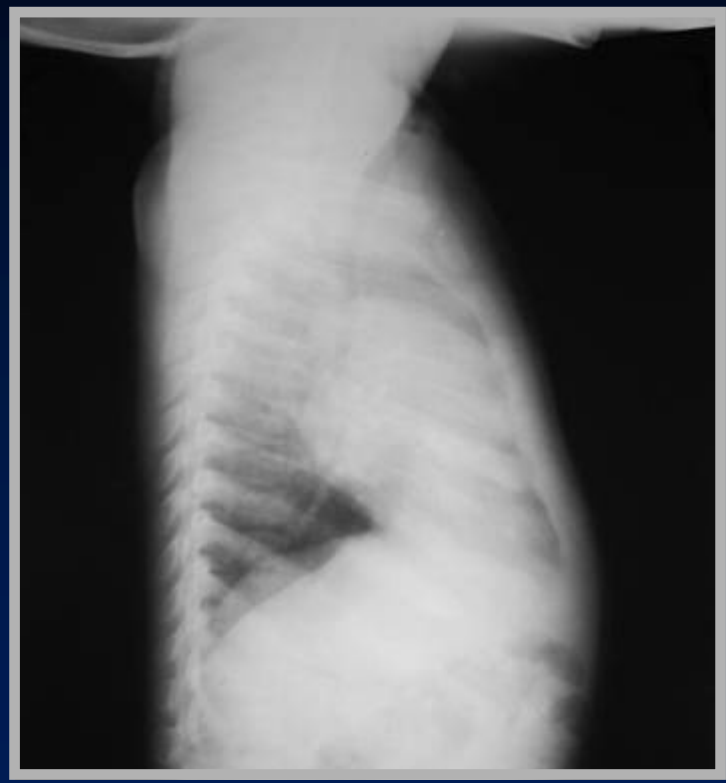
Congenital Pericardial Defect
X-ray Findings

- Focal bulge in area of main pulmonary artery
- Sharply marginated
- Lung may interpose between heart-left hemidiaphragm
- Increased distance between sternum and heart 2° absence of sternopericardial ligament

Congenital Pericardial Defect
X-ray Findings-Continued

- Levoposition of heart
- Pneumopericardium following pneumothorax





Congenital Defect in the Pericardium

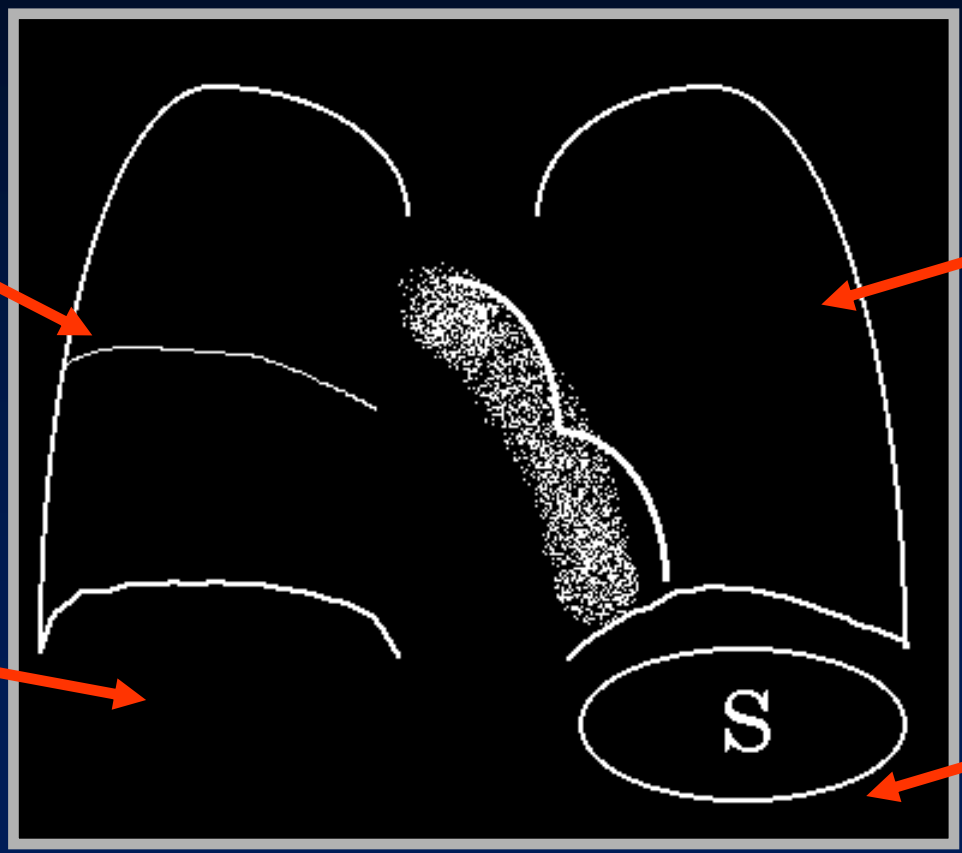
Congenital Pericardial Defect
Treatment

- Since herniation and strangulation of left atrial appendage or herniation of LA/LV may occur
- Foraminal defect requires surgery

Cardiac Malpositions

Heterotaxy Syndromes

Trilobed and Bilobed Lungs



Naming Rules

- Since anatomic side (i.e. “left” or “right”) in complex lesions is frequently reversed or indeterminate
- Naming conventions for
  - Atria
  - AV valves
  - Ventricles
  - Ventricular outflow tracts

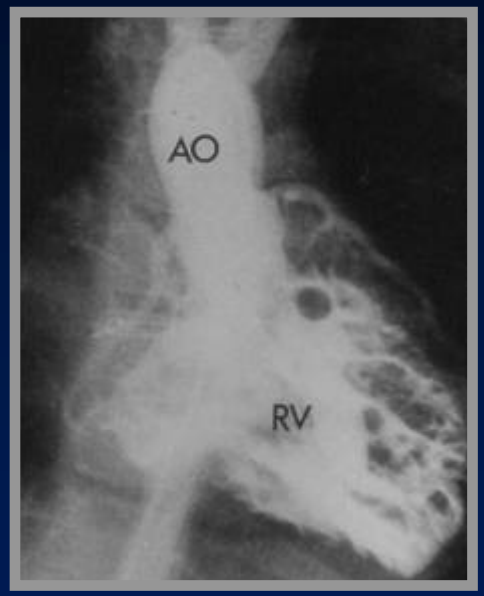
The Rules
How the atria are named

- Anatomic right atrium is on side of trilobed lung and liver
  - Shape of atrial appendage-broad and pyramidal
  - Same side as IVC
- Anatomic left atrium is on side of bilobed lung and spleen
  - Shape of atrial appendage-thin c narrow neck
  - Same side as aortic arch

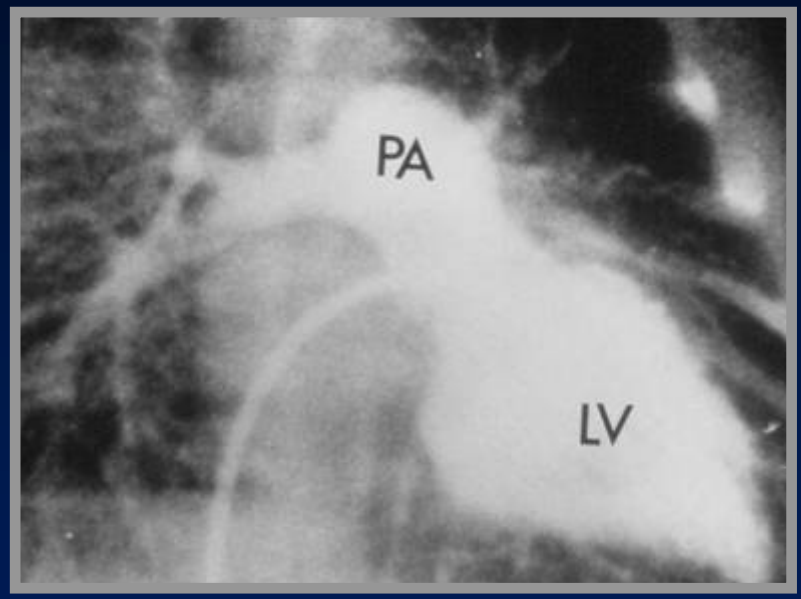
The Rules
How the ventricles are named

- Anatomic right ventricle is trabeculated ventricle
  - Coarse in both systole and diastole
  - Has tricuspid AV valve
- Anatomic left ventricle is smooth-walled ventricle
  - In diastole; fine trabeculations in systole
  - Has bicuspid AV valve

Anatomic Ventricles



Trabeculated ventricle-
Anatomic Right



Smooth ventricle-
Anatomic Left

The Rules
Mitral and tricuspid valves

- Tricuspid valve belongs to anatomic right ventricle
  - Not right atrium
- Mitral valve belongs to anatomic left ventricle
  - Not left atrium

AV Connections
Concordance

- Ventricles are concordant to the atria
  - When R atrium connects to R ventricle
  - L atrium connects to L ventricle
- Ventricles are discordant to the atria
  - When R atrium connects to L ventricle
  - When L atrium connects to R ventricle
- With atrial isomerism, AV connections are ambiguous

The Rules
Aortic and pulmonic valves

- Pulmonic valve is part of pulmonary artery
  - Not anatomic right ventricle
- Aortic valve is part of aorta
  - Not anatomic left ventricle
- Pulmonic infundibulum is part of anatomic right ventricle

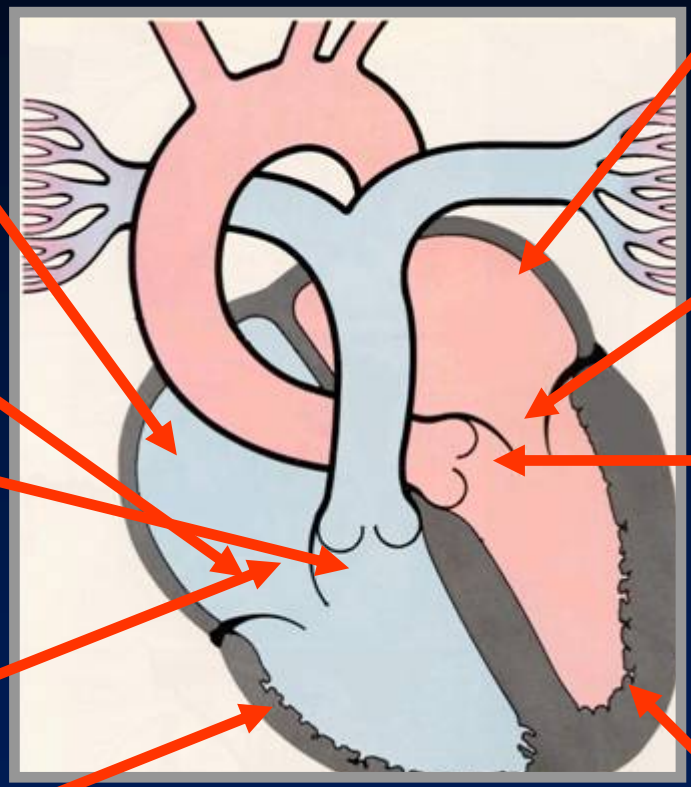
Anatomic R atrium is on side of trilobed lung-same side as IVC

Tricuspid valve belongs to anatomic RV

Pulmonic infundibulum belongs to anatomic RV

Pulmonic valve belongs to pulmonary artery

Anatomic R ventricle is trabeculated



Anatomic L atrium is on side of bilobed lung-same side as Ao arch

Mitral valve belongs to anatomic LV

Aortic valve belongs to aorta

Anatomic L ventricle is smooth

Situs
Definitions

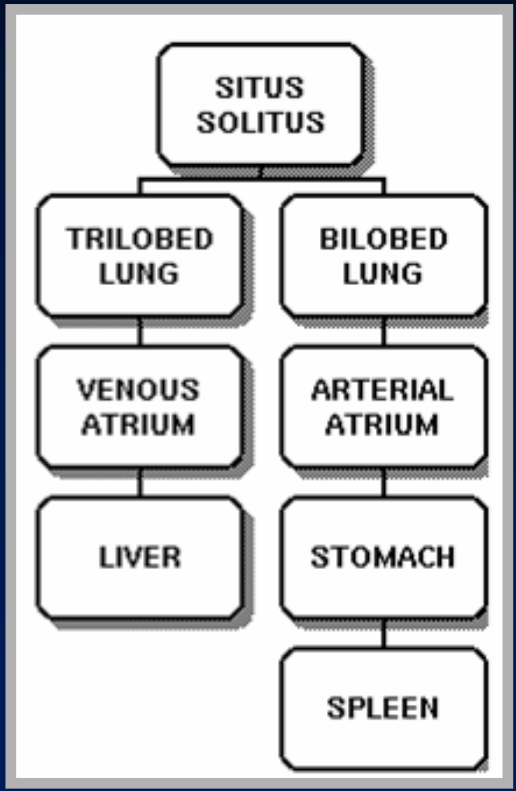
- Describes position of asymmetric organs in body
  - Lungs
  - Liver
  - Spleen
  - Stomach

Situs Solitus

- Normal anatomic relationships

Right side	Left side
Trilobed lung	Bilobed lung
Eparterial bronchus	Hyparterial bronchus
Anatomic right atrium	Anatomic left atrium
Liver	Spleen

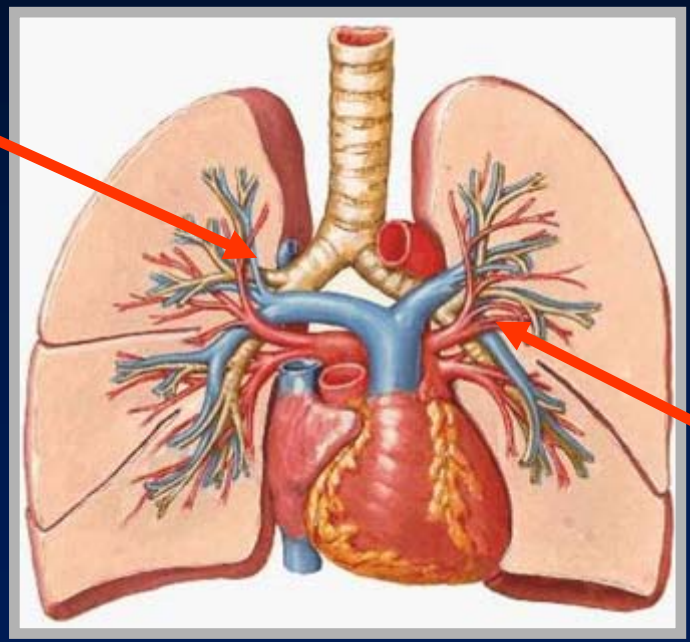
Situs Solitus



Situs Solitus
Hyparterial/Eparterial Bronchi

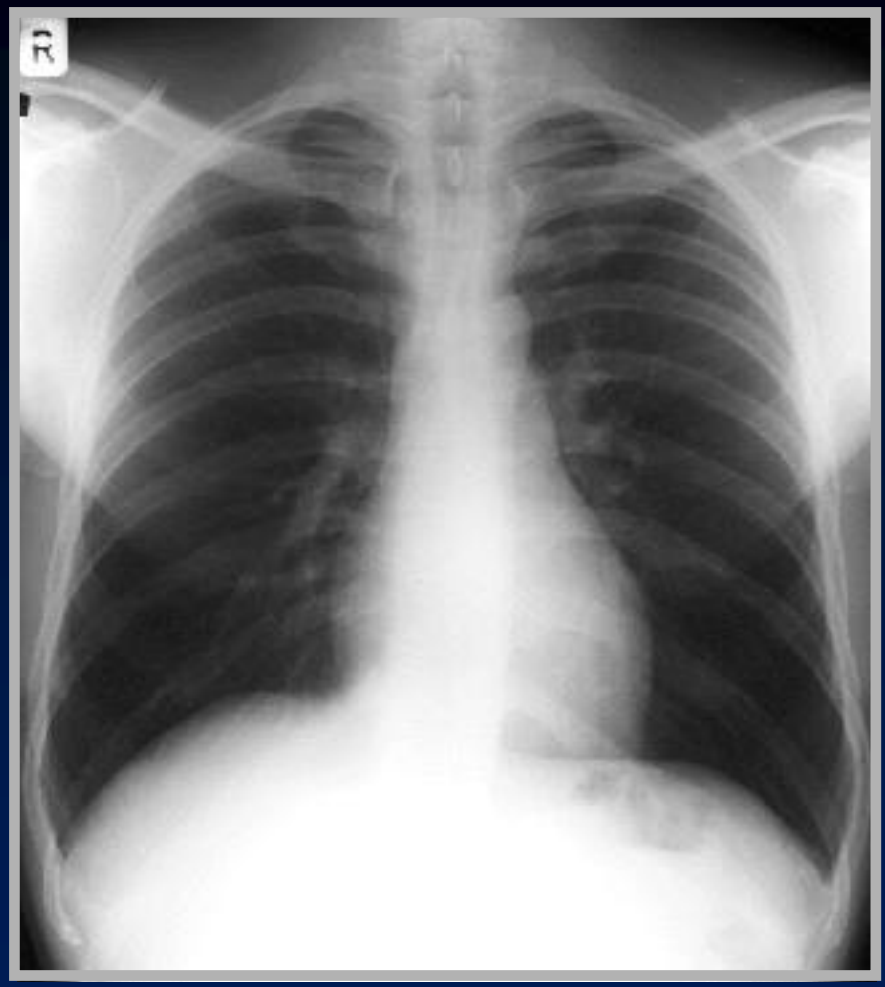
Eparterial bronchus-

First branch of Right mainstem bronchus is above pulmonary artery



Hyparterial bronchus-

First branch of Left mainstem bronchus is below pulmonary artery



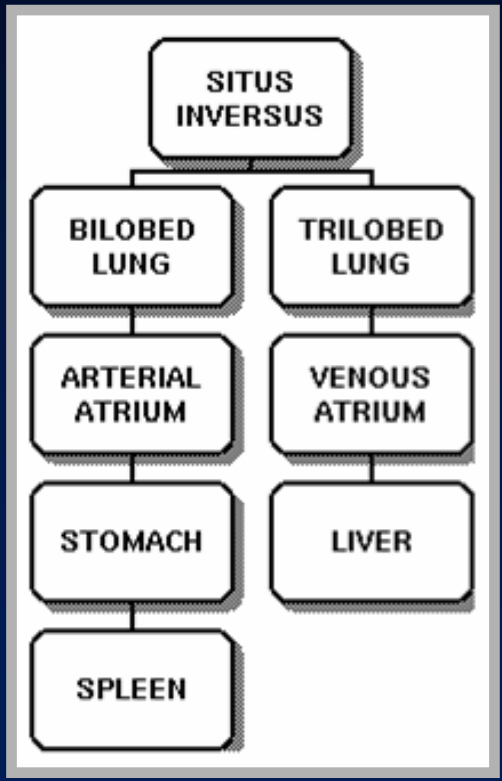
Situs Solitus
0.6 - 0.8% CHD

Situs Inversus

- Reversed anatomic relationships

Right side	Left side
Bilobed lung	Trilobed lung
Hyparterial bronchus	Eparterial bronchus
Anatomic left atrium	Anatomic right atrium
Spleen	Liver

Situs Inversus



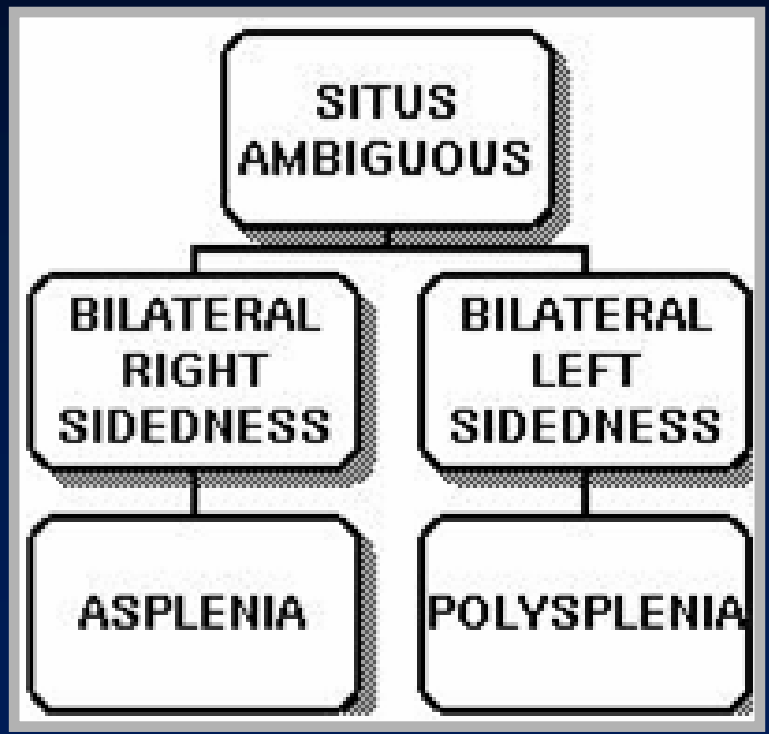
Situs Ambiguous

- Lungs and abdomen are symmetric so right and left sides can't be defined
  - Isomerism-both atria have the same features
    - Either right or left
  - Two kinds of situs ambiguous
    - Bilateral right-sidedness
    - Bilateral left-sidedness

Situs Ambiguous
Heterotaxy Syndrome

- Bilateral right-sidedness
  - Since, spleen is usually on left side
  - No spleen
    - Asplenia syndrome
- Bilateral left-sidedness
  - Since, spleen is usually on left side
  - Many spleens
    - Polysplenia syndrome

Situs Ambiguous



Cardiac Positions

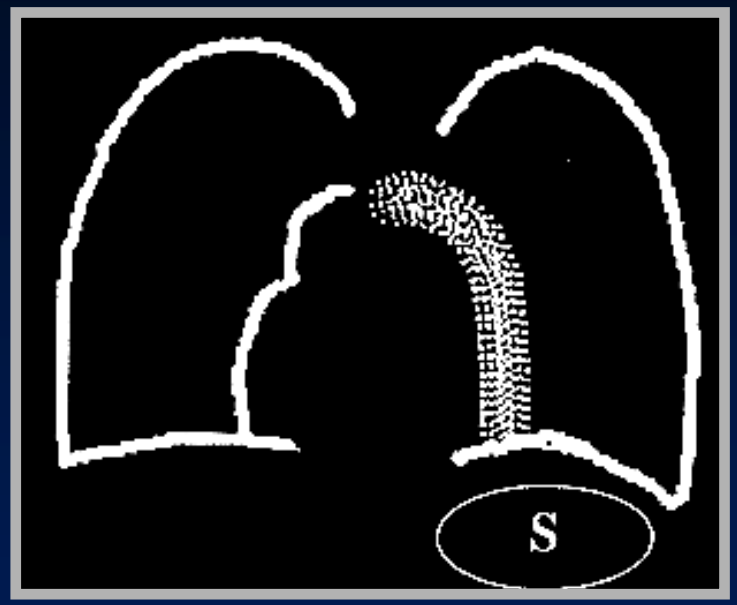
- Position of cardiac apex
  - Levocardia on the left
  - Dextrocardia on the right
  - Mesocardia in the midline
- Cardiac malposition
  - Anything other than situs solitus with levocardia

Cardiac Malpositions
Types

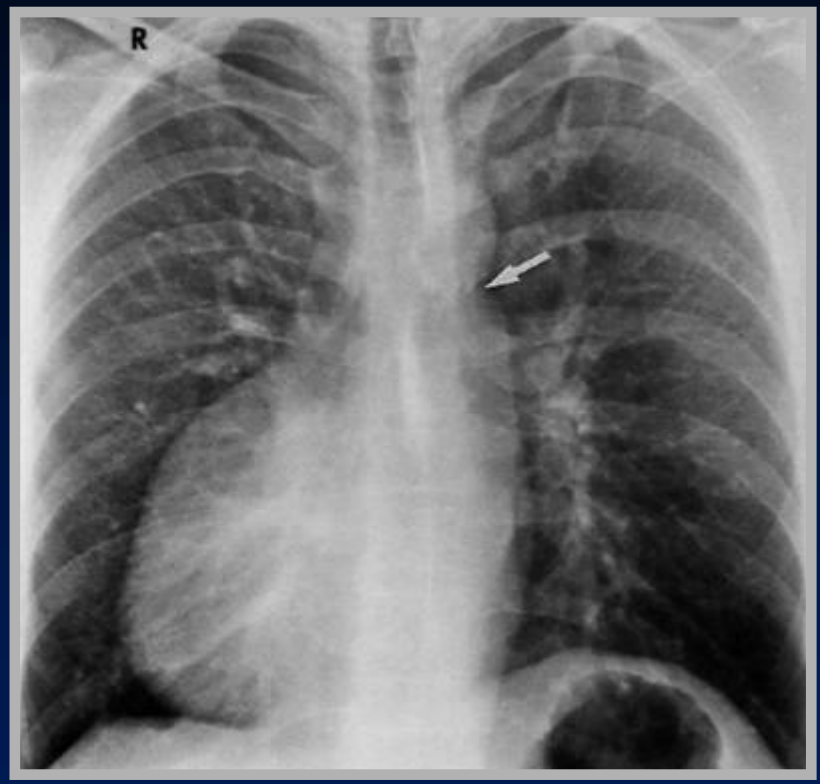
- Situs solitus with dextrocardia
- Situs inversus with levocardia
- Situs inversus with dextrocardia

Situs Solitus with Dextrocardia

Test Alert

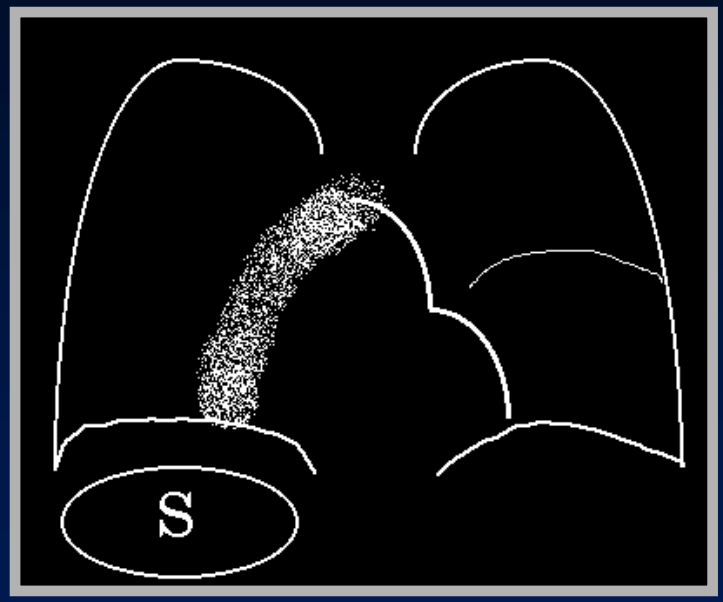


95% chance of CHD of which 80% corrected transposition
If cyanotic with ↑ flow, then tricuspid atresia.
If cyanotic with ↓ flow, then corrected transposition.
If asplenia, then 100% have common ventricle.
Interrupted IVC common.

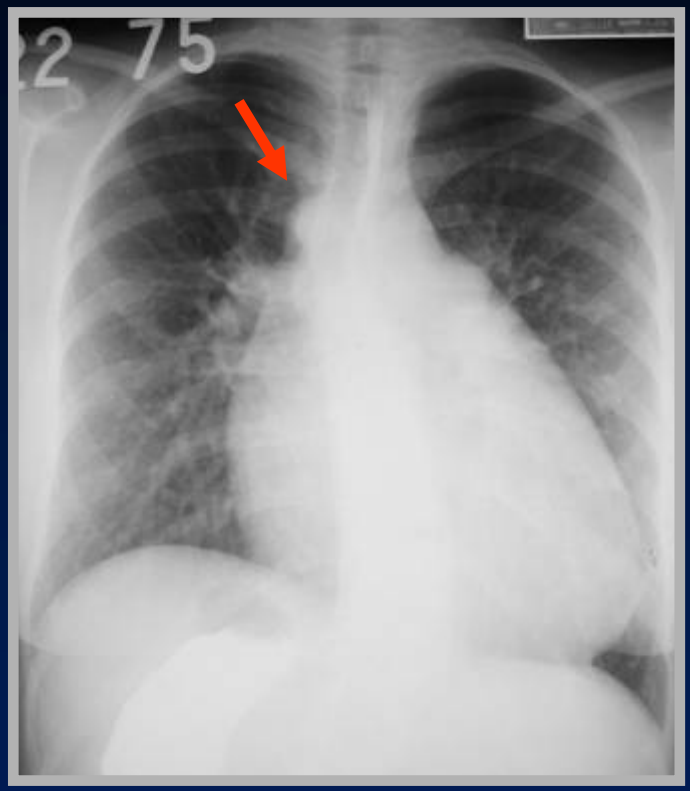


Situs Solitus with Dextrocardia
Coarctation of Aorta

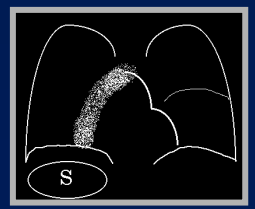
Situs Inversus with Levocardia

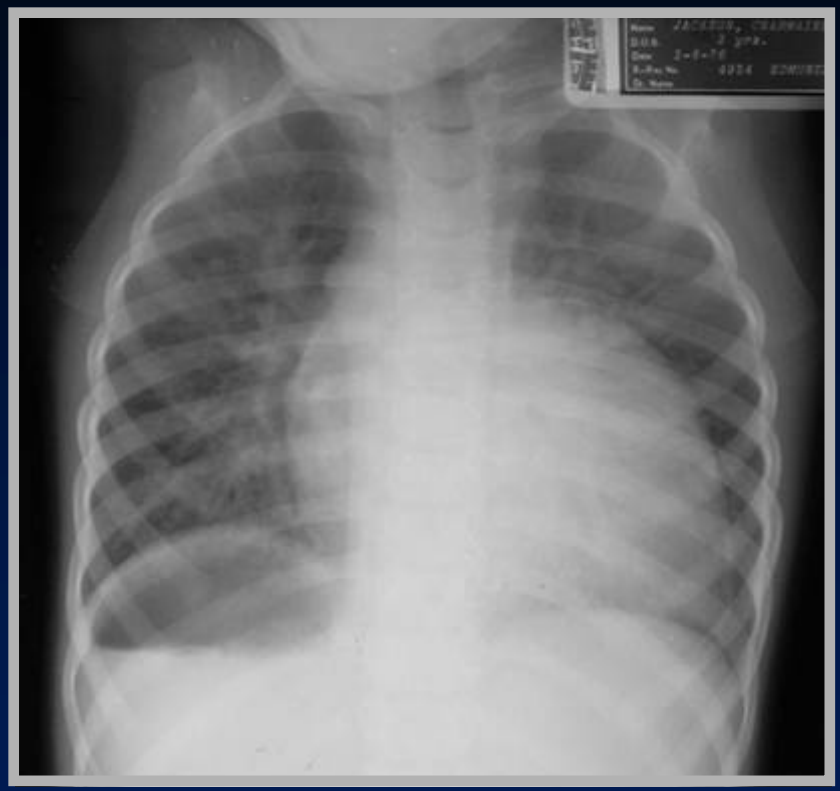


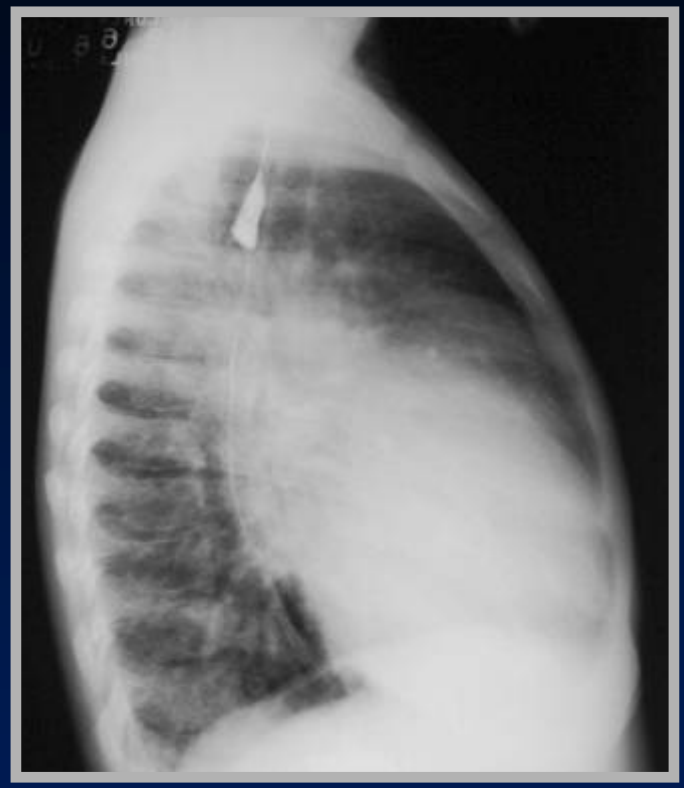
Rare, but 100% CHD
If asplenia, 100% have common ventricle
Interruption of IVC common



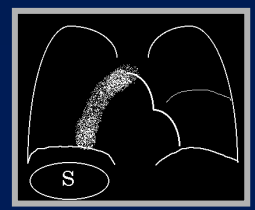
Situs Inversus with Levocardia



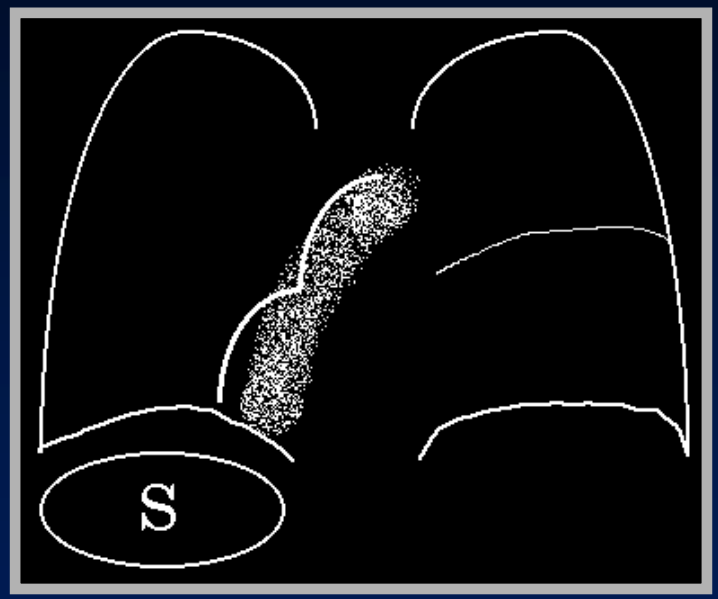




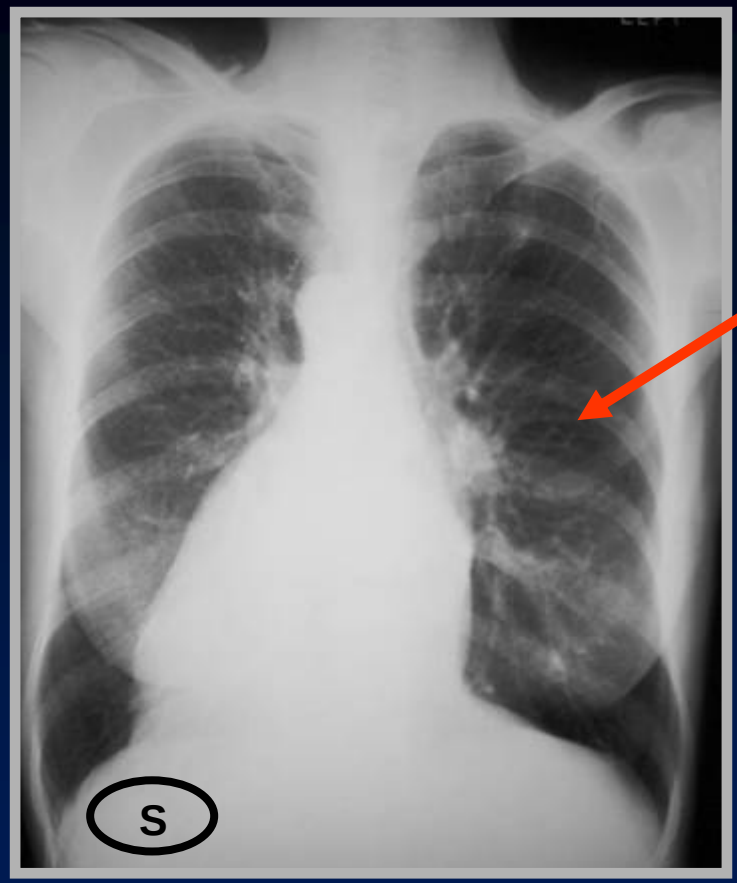
Situs Inversus with Levocardia
Transposition



Situs Inversus with Dextrocardia

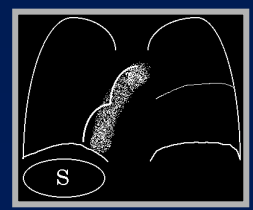


3-5% CHD
Most common is Corrected Transposition
Kartgener's

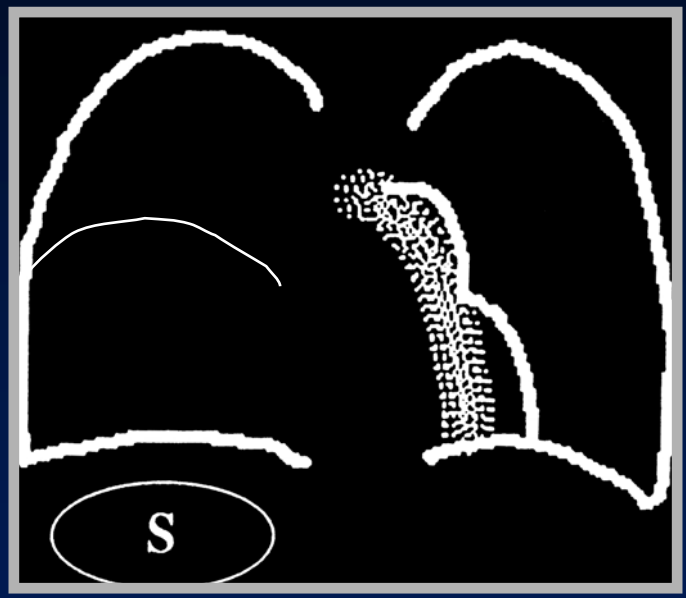


Trilobed lung

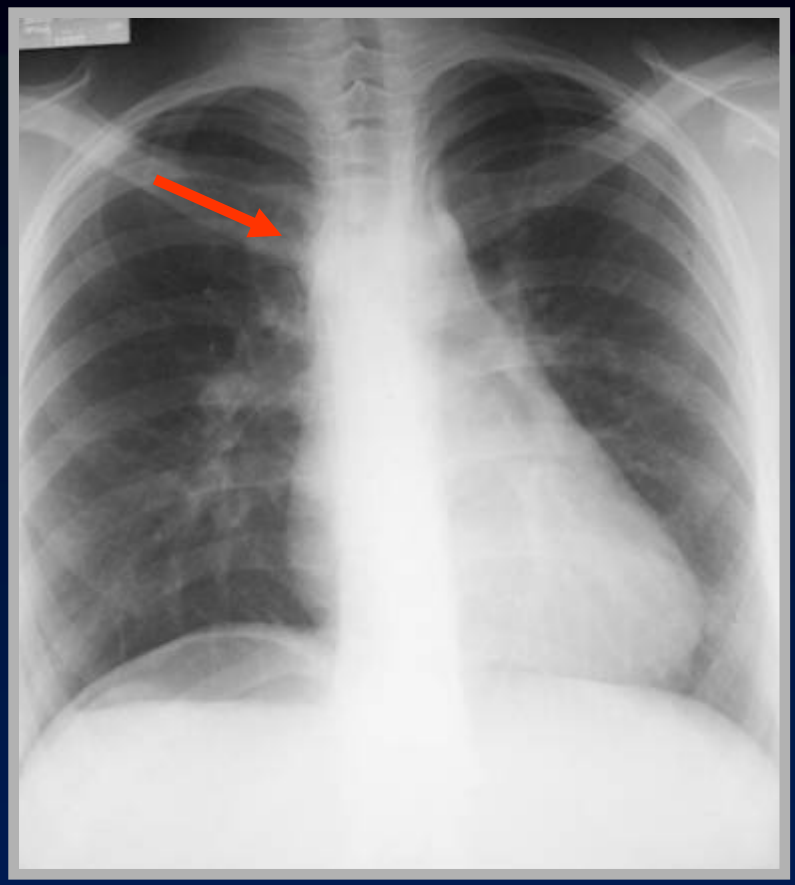
Situs Inversus with Dextrocardia

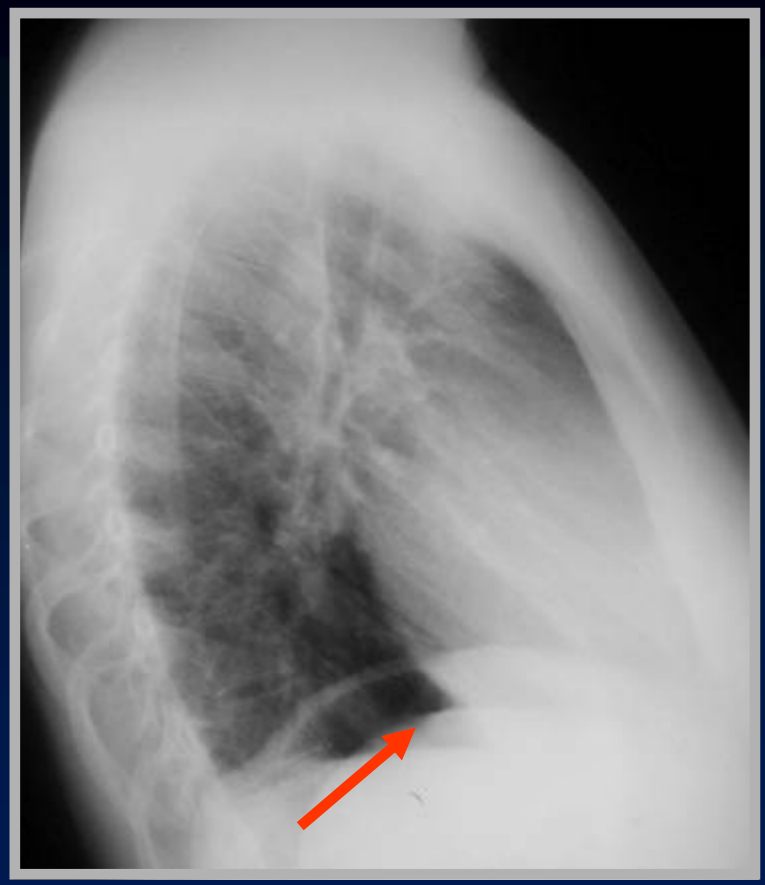


Situs Solitus
Malposition of the Stomach

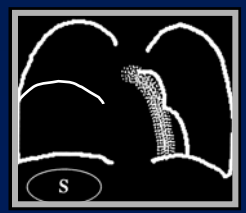


R/O asplenia
Most have CHD (L → R shunt)
Most with polysplenia have azygous continuation of IVC

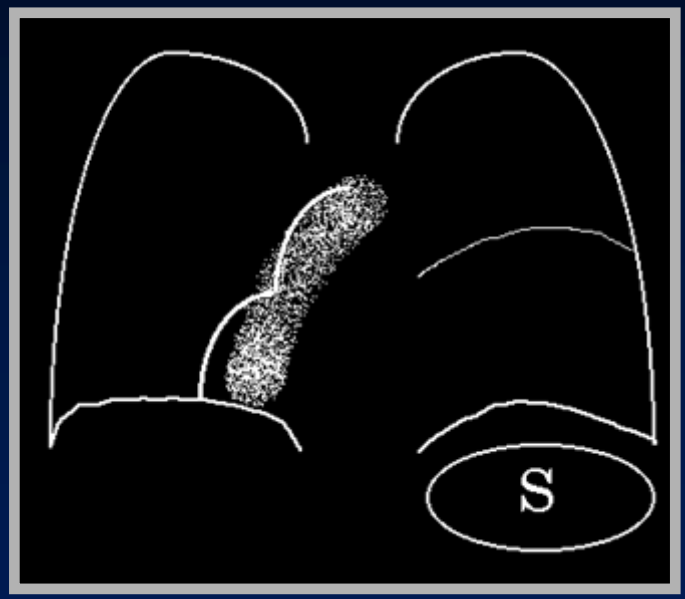




Situs Solitus with Malposition of the Stomach



Situs Inversus
Malposition of the Stomach



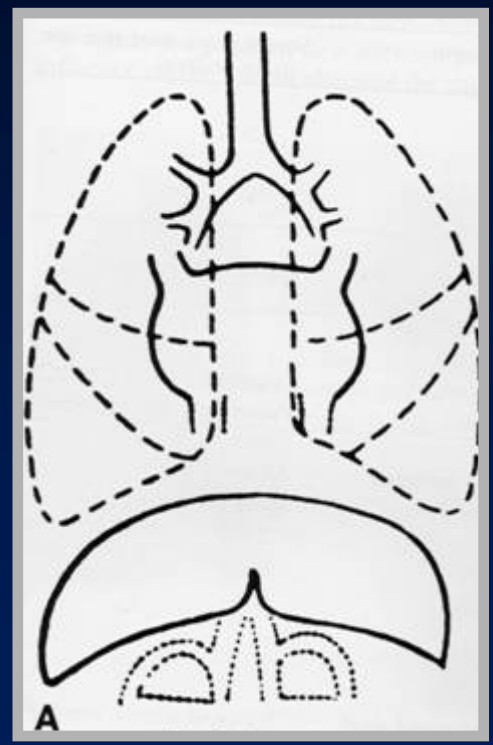
Situs Inversus with Malposition of the Stomach
95% CHD of which 80% are Corrected Transposition

Situs Ambiguous
Heterotaxy Syndrome

- Bilateral right-sidedness
  - Since, liver is usually on right side
  - No spleen
    - Asplenia syndrome
- Bilateral left-sidedness
  - Since, spleen is usually on left side
  - Multiple spleens
    - Polysplenia syndrome

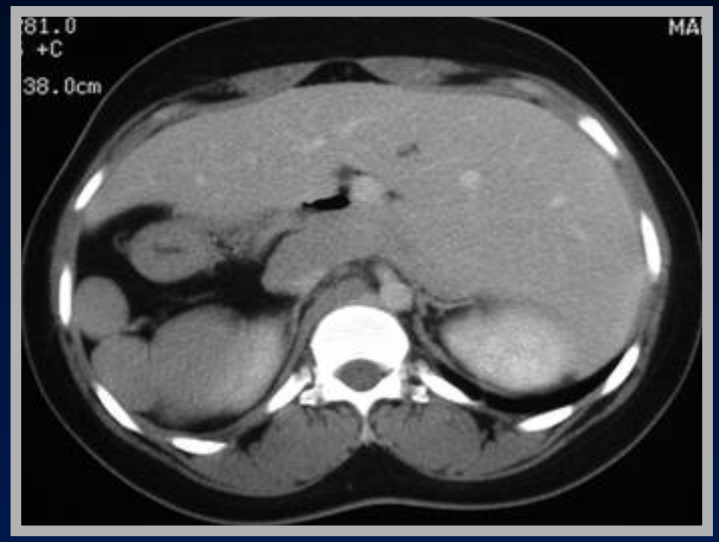
Asplenia
Bilateral Right-sidedness

- Male
- Cyanotic
- High risk of infection
- Severe cardiac abnormalities
  - Transposition
  - TAPVR

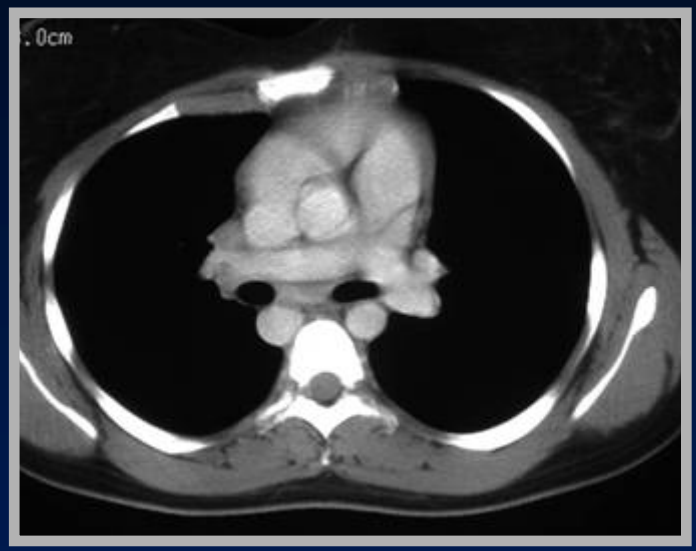


Polysplenia
Bilateral left-sidedness

- Female
- Abnormalities are more benign
  - Azygous continuation of IVC
  - Bilateral superior vena cava
  - PAPVR
  - ASD



Situs Ambiguous-polysplenia

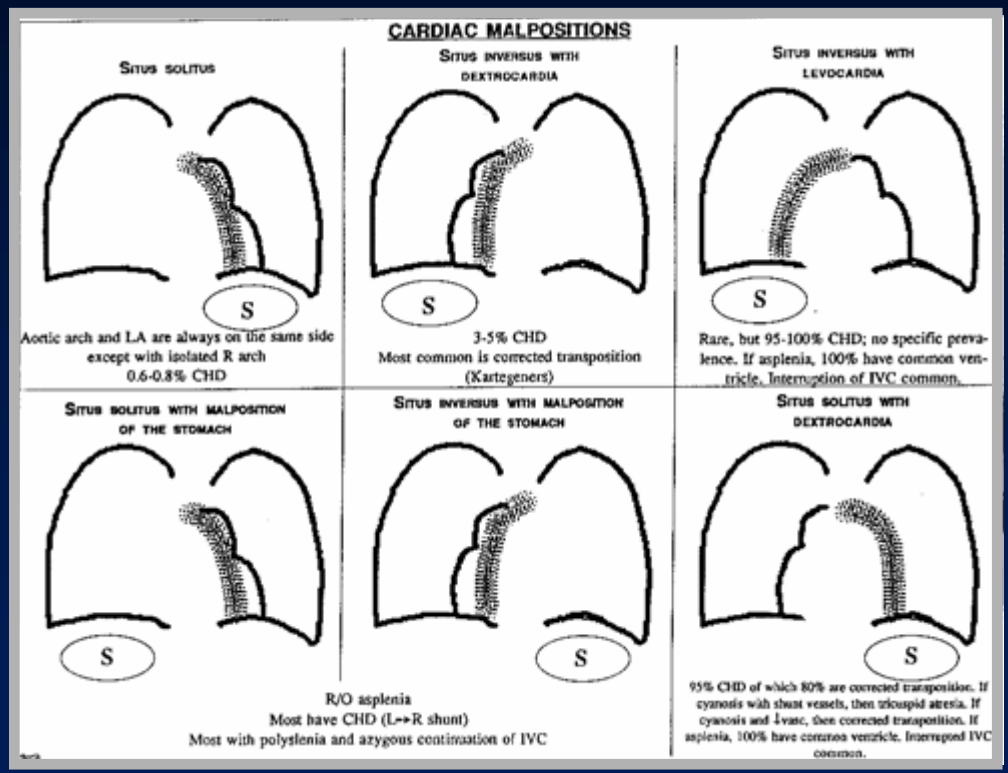


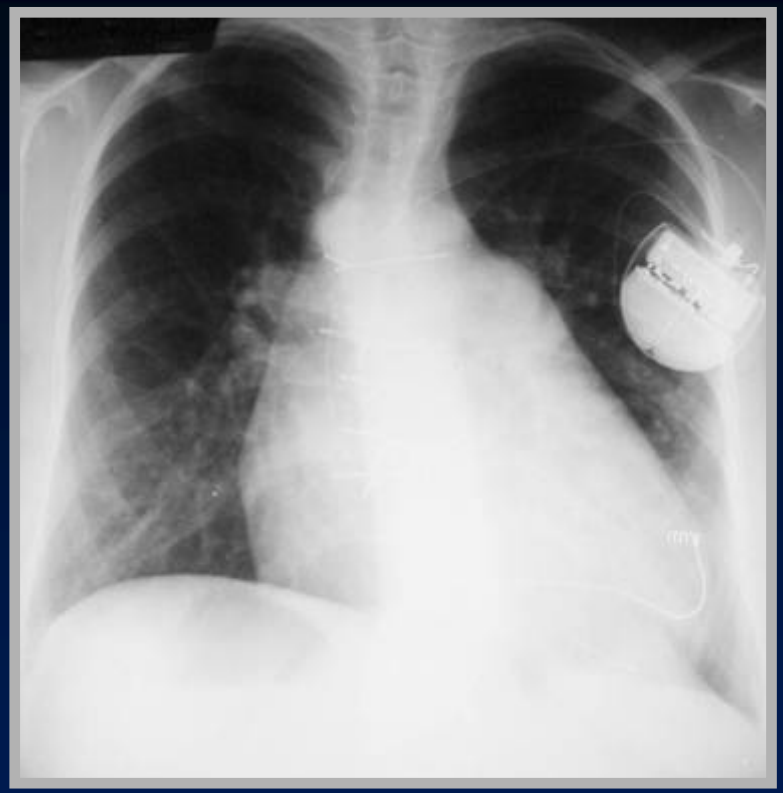
Situs Ambiguous-polysplenia

Approach to Cardiac Malpositions

- Which side is heart on
- Which side is trilobed lung on
- Which side is arch on
- Which side is stomach bubble on
- Check for asplenia
  - Midline liver
  - Minor fissures in both lungs

Another Approach





Situs Inversus with Levocardia

Rare, but 100% CHD
If asplenia, 100% have common ventricle
Interruption of IVC common

Situs ambiguous
Asplenia with dextrocardia

Complex CHD

Nine Lesions Which Produce 75% of All Severe Congenital Heart Lesions In the Neonate

- **Decreased flow**
  1. Tetralogy of Fallot
  2. Tricuspid Atresia
  3. Severe Pulmonic Stenosis
  4. Ebstein's
- **Increased Flow**
  5. Transposition
  6. VSD

Nine Lesions Which Produce 75% of All Severe Congenital Heart Lesions In the Neonate

- **Pulmonary venous hypertension**
  7. Hypoplastic left heart
  8. Coarctation of the aorta
  9. TAPVR with infradiaphragmatic obstruction
- **What's left**
  - Left-to-right shunts
    - ASD
    - PDA
  - Truncus arteriosus

The End