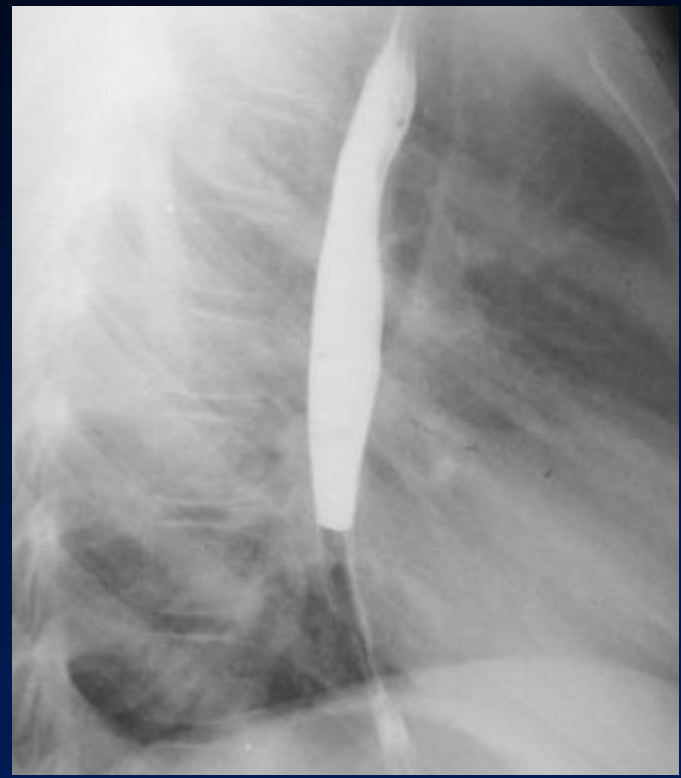
Left to Right Shunts

In Slide Show mode, to advance slides, press spacebar or click left mouse button



7 yo acyanotic female

Atrial Septal Defect

Atrial Septal Defect
Four Major Types

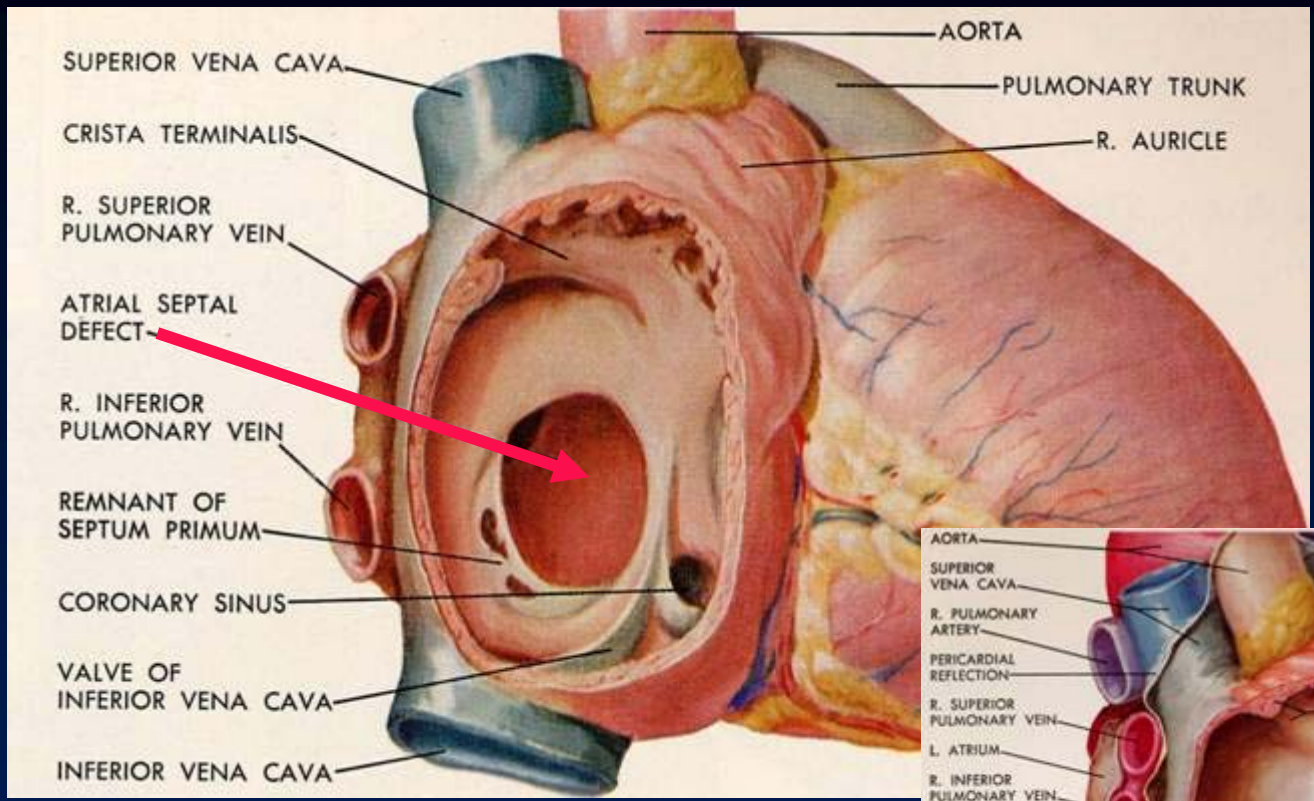
- Ostium secundum
- Ostium primum
- Sinus venosus
- Posteroinferior

Atrial Septal Defect
General

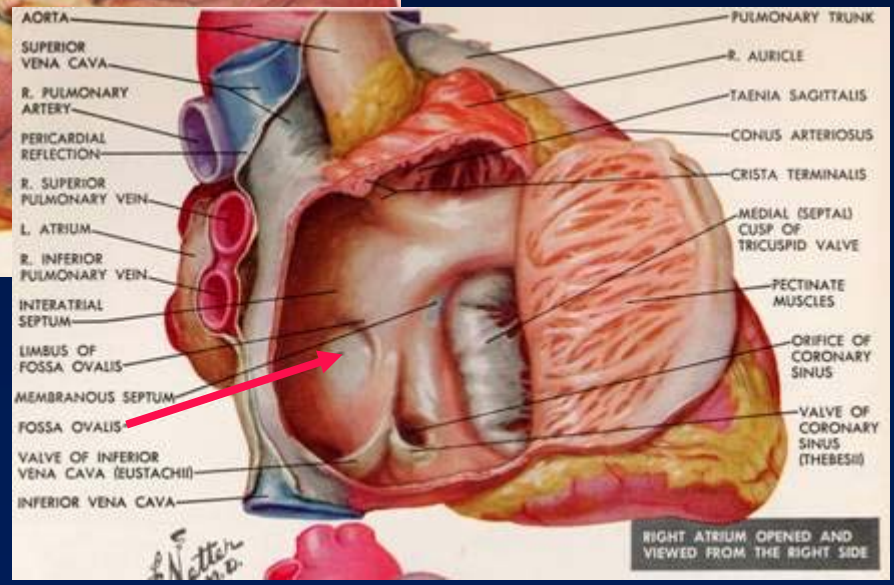
- 4:1 ratio of females to males
- Most frequent congenital heart lesion initially diagnosed in adult
- Frequently associated with Ellis-van Creveld and Holt-Oram syndromes
- Associated with prolapsing mitral valve

Atrial Septal Defect
Ostium Secundum Type

- Most common is ostium secundum (60%) located at fossa ovalis
- High association with prolapse of mitral valve



Normal



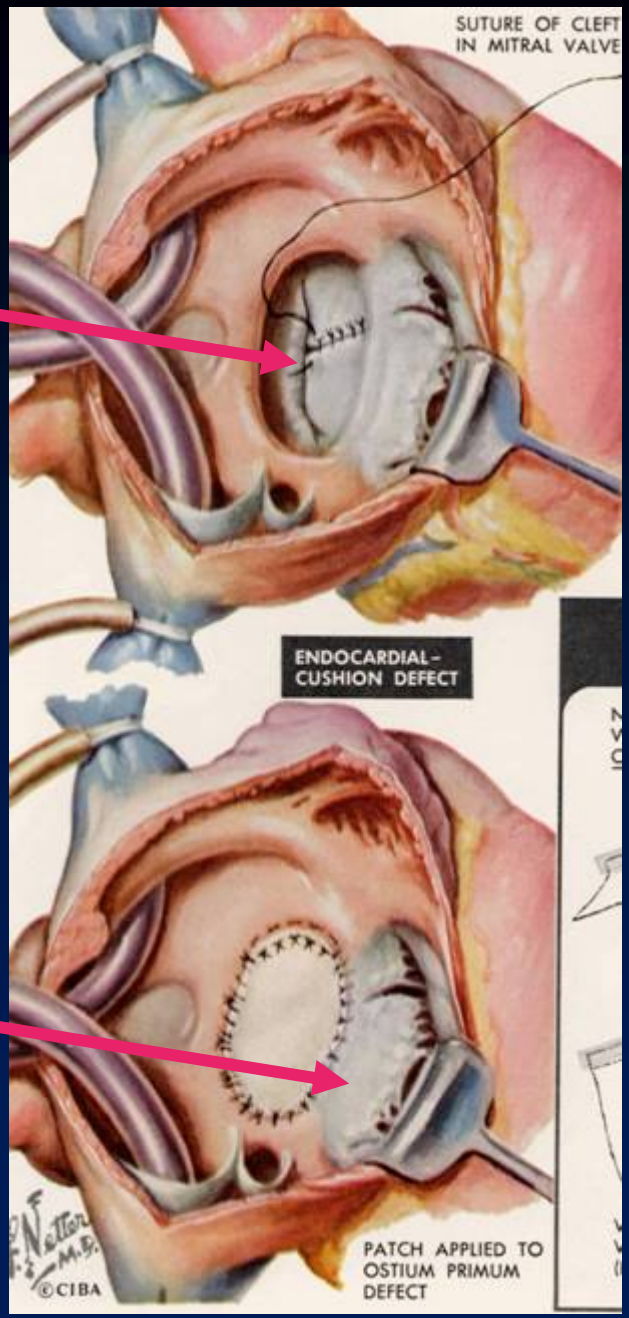
Right atrium open looking into left atrium through ASD

Atrial Septal Defect
Ostium Primum Type

- Ostium primum type usually part of endocardial cushion defect
- Frequently associated with cleft mitral and tricuspid valves
- Tends to act like VSD physiologically

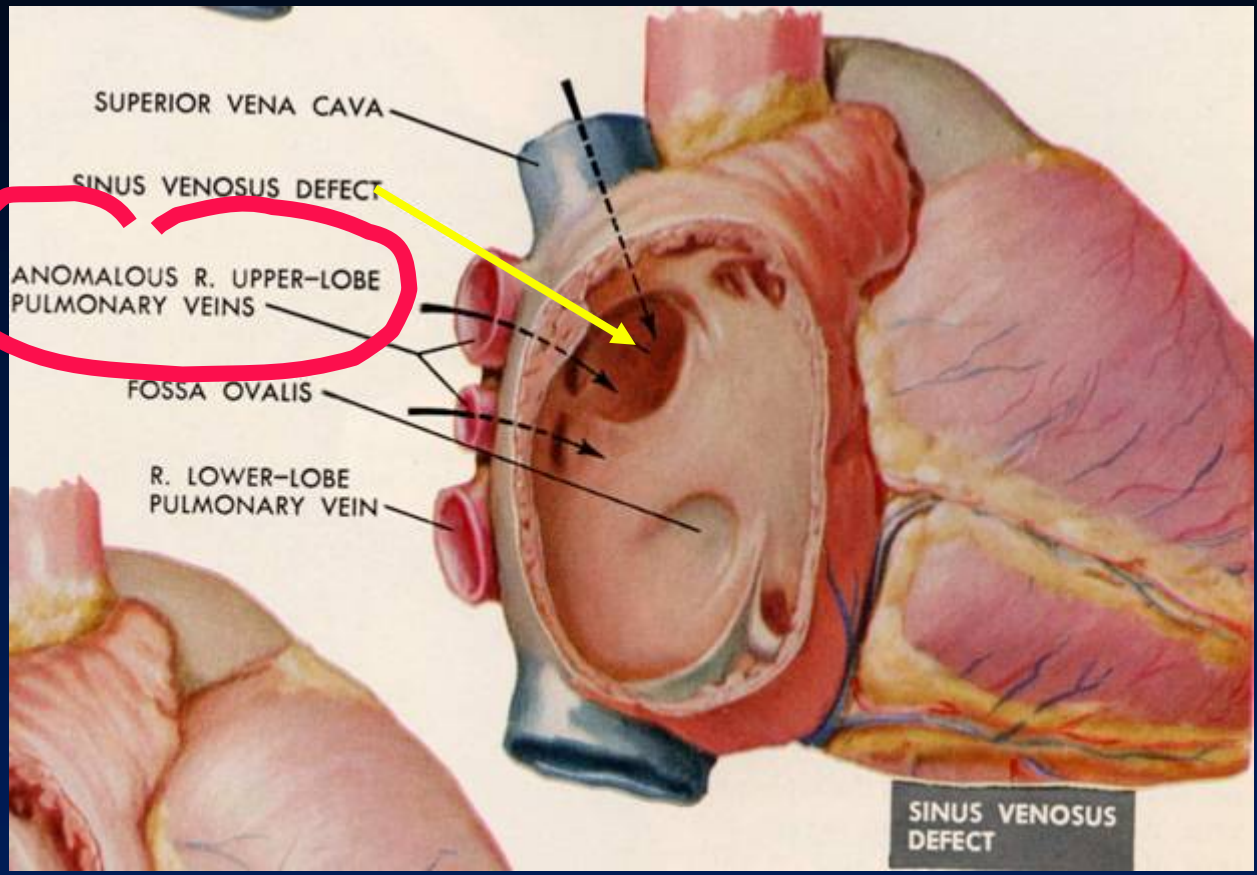
Looking through ostium primum defect at cleft mitral valve

Proximity of ostium primum defect to tricuspid valve



Atrial Septal Defect
Sinus Venosus Type

- Sinus venosus type located high in inter-atrial septum
- 90% association of anomalous drainage of R upper pulmonary vein with SVC or right atrium
  - Partial anomalous pulmonary venous return



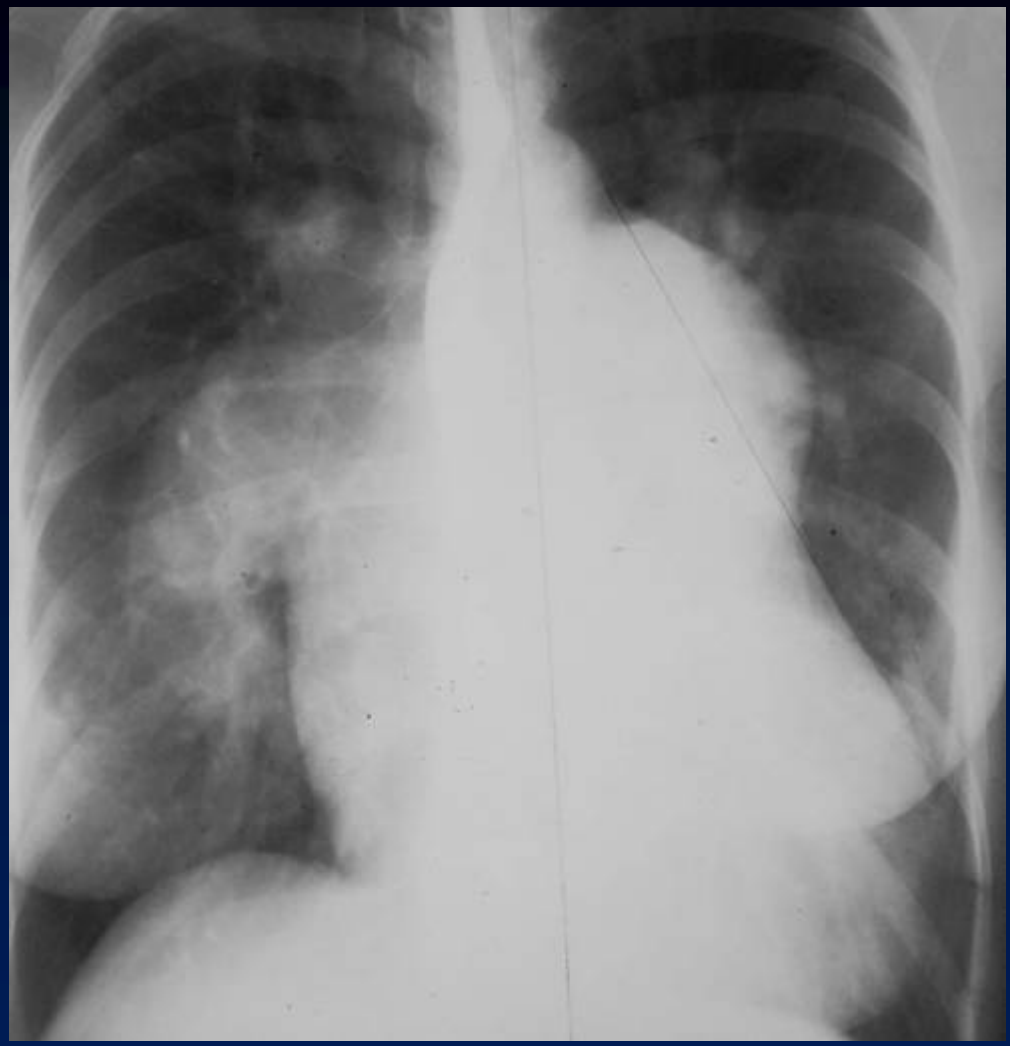
Right atrium open looking into left atrium through ASD

Atrial Septal Defect
Posteroinferior Type

- Most rare type
- Associated with absence of coronary sinus and left SVC emptying into LA

Atrial Septal Defect
Pulmonary Hypertension

- Rare in ostium secundum variety (<6%)
  - Low pressure shunt from LA ➜ RA
- More common in ostium primum variety
  - Behaves physiologically like VSD



37 yo female with severe PAH 2°
ostium primum type of ASD

Atrial Septal Defect
X-Ray Findings

- Enlarged pulmonary vessels
- Normal-sized left atrium
- Normal to small aorta

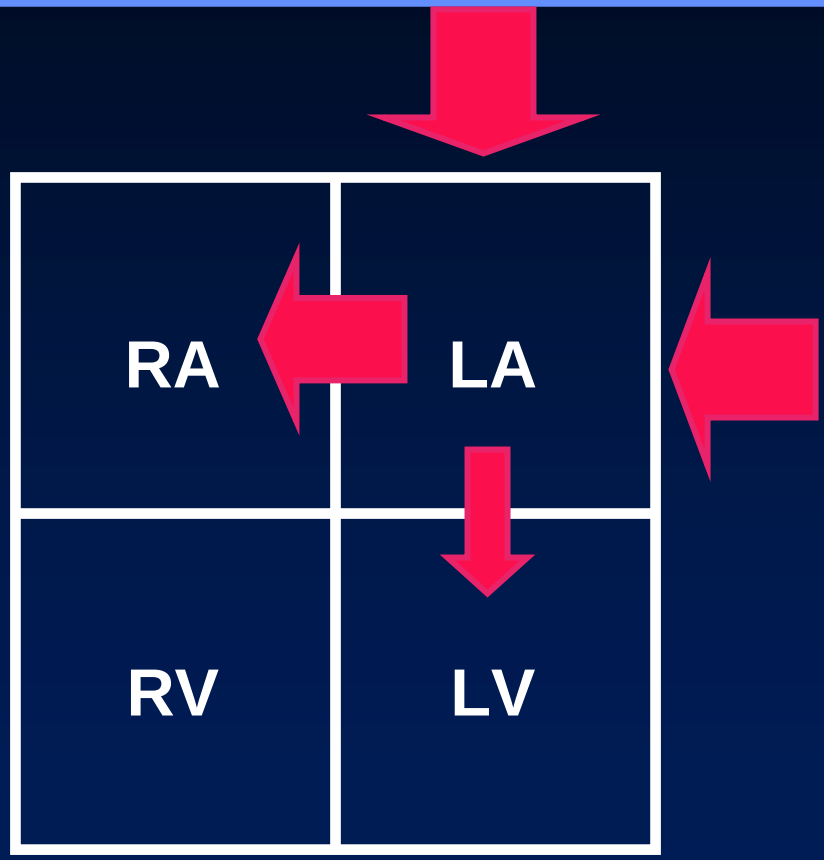
Atrial Septal Defect
Complications

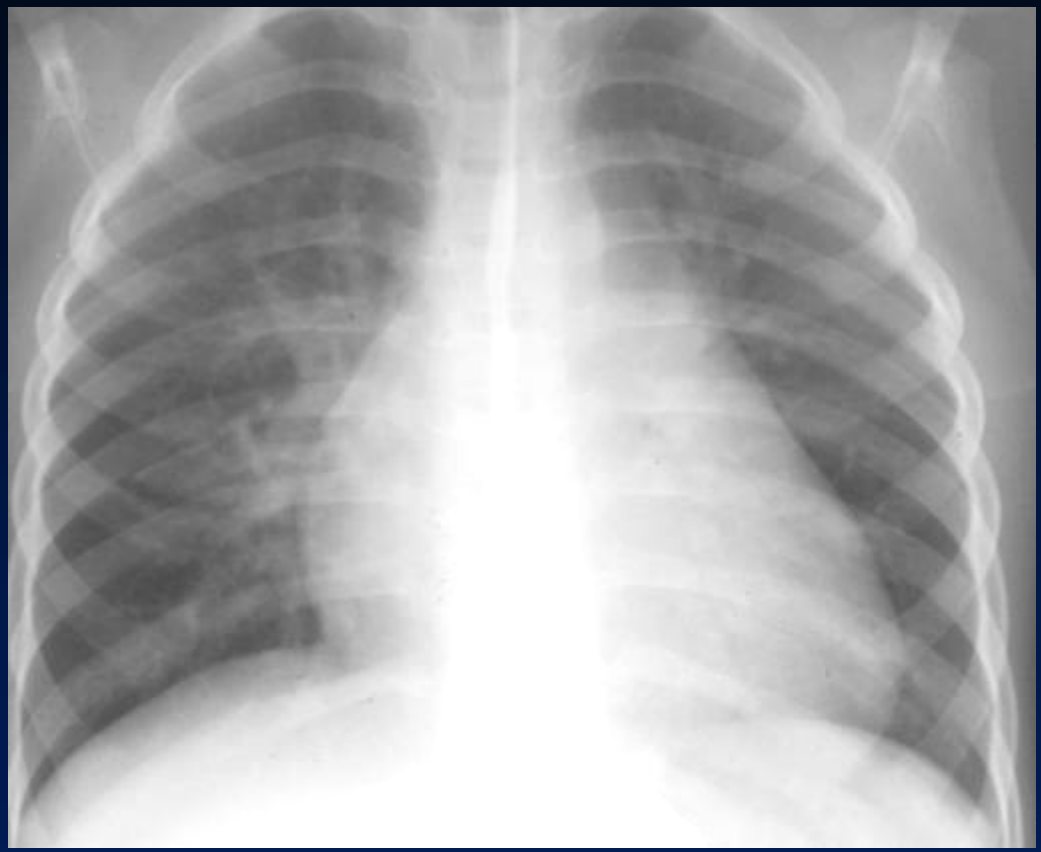
- Large shunts associated with
  - Pulmonary infections and cardiac arrythmias
- Higher incidence of pericardial disease with ASD than any other CHD
- Bacterial endocarditis is rare

Differentiating ASD, PDA and VSD

⇩	**LA**	Ao
ASD	⇔	⇩
PDA	⇧	⇧
VSD	⇧	⇔

Atrial Septal Defect
Why the Left Atrium Isn't Enlarged





1 yo acyanotic female

Ventricular Septal Defect

Ventricular Septal Defect
General

- Most common L → R shunt
- Shunt is actually from left ventricle into pulmonary artery more than into right ventricle

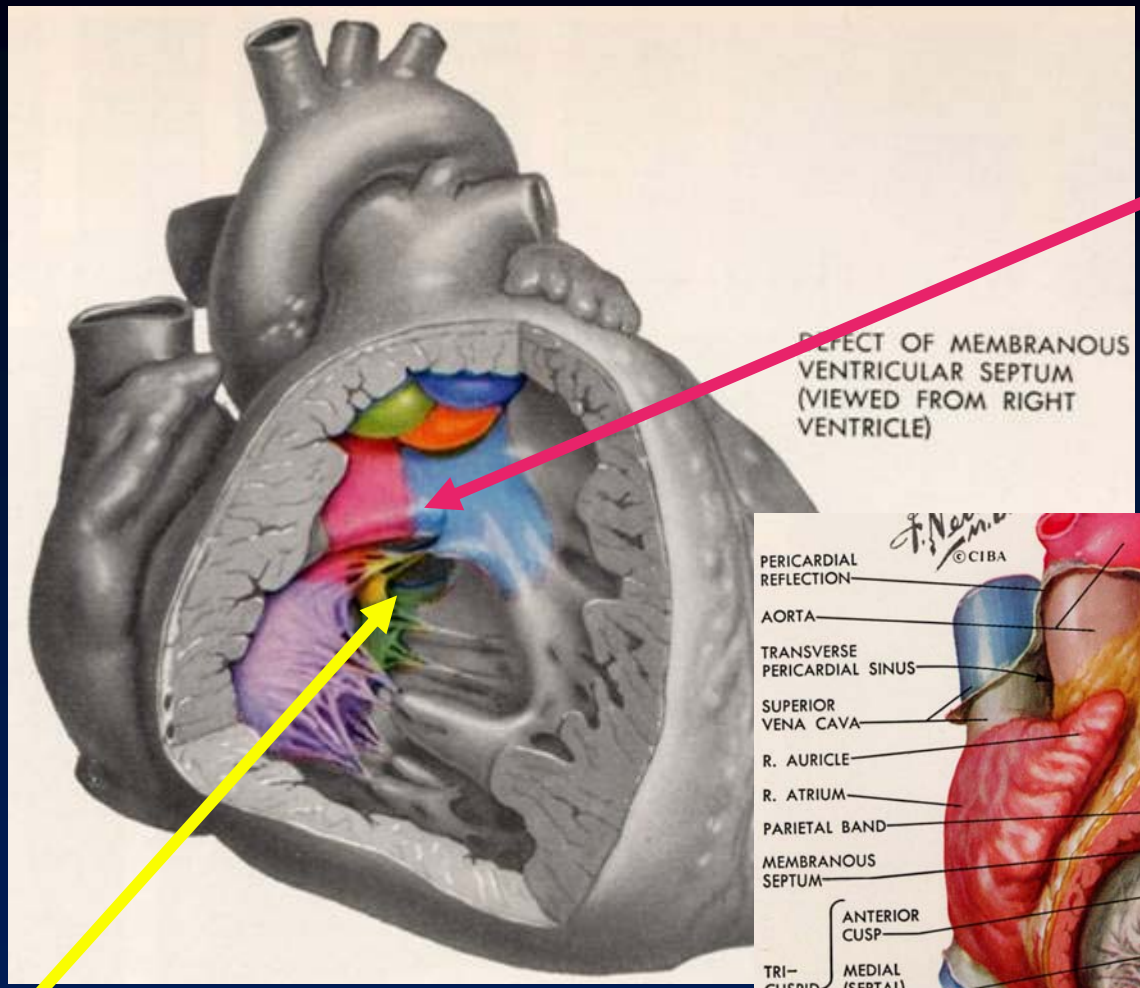
Ventricular Septal Defect
Types

- Membranous
- Supracristal
- Muscular
- AV canal

Ventricular Septal Defect
Membranous

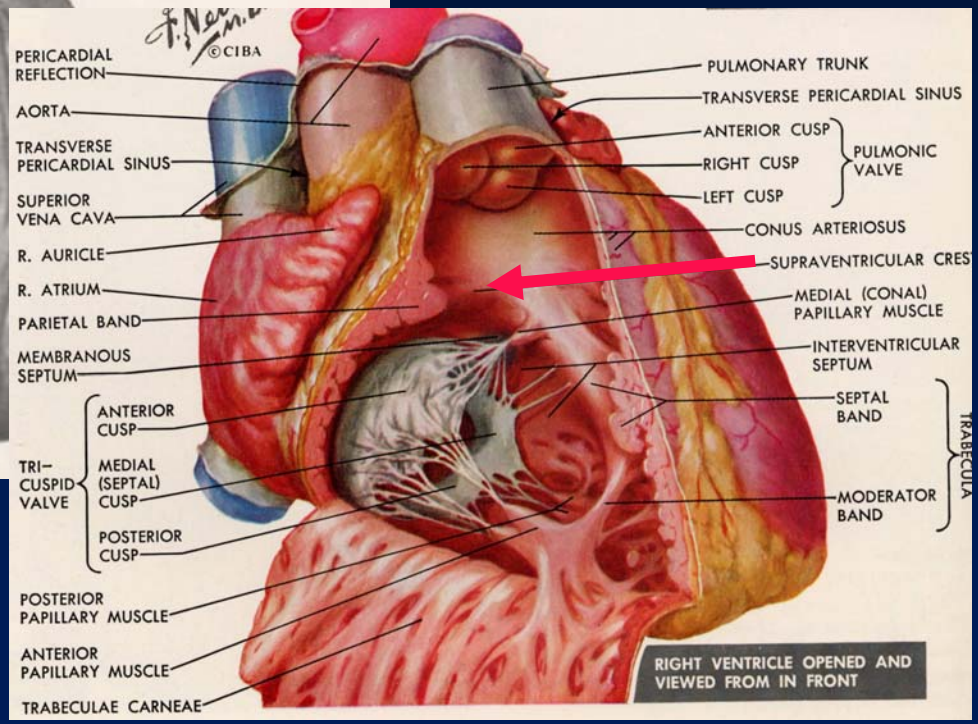
- Membranous = perimembranous VSD (75-80%–most common)
- Location: Posterior and inferior to crista supraventricularis near right and posterior (=non-coronary) aortic valve cusps
- Associated with: small aneurysms of membranous septum

Right ventricle opened

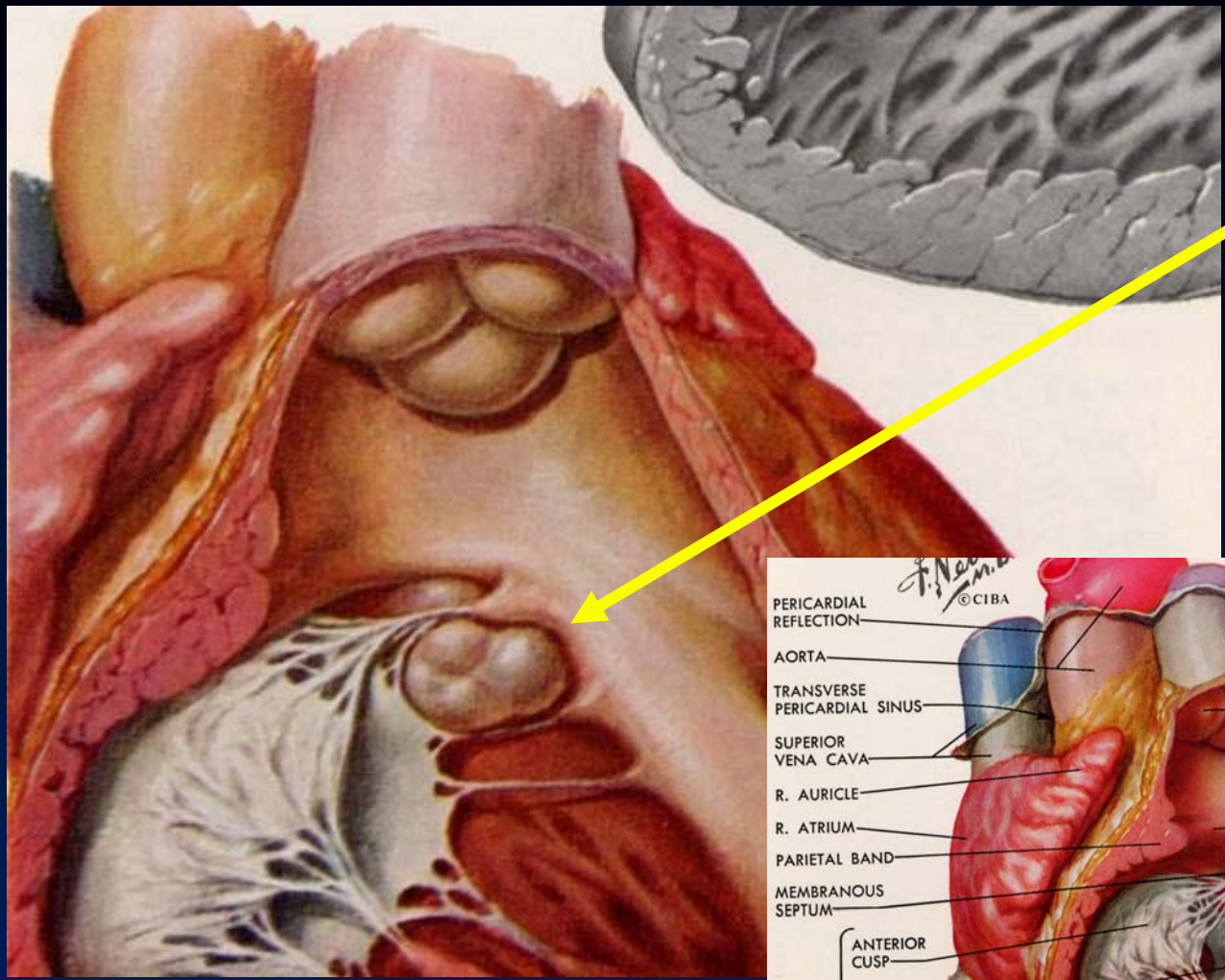


Crista supraventrularis

Normal

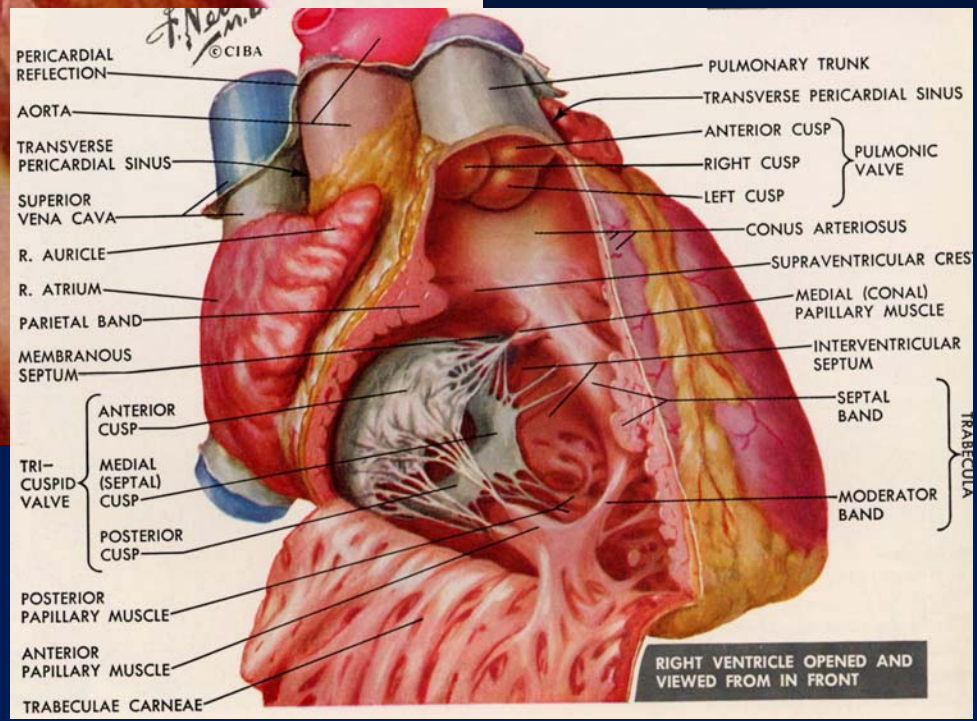


Membranous VSD



Aneurysm of membranous septum

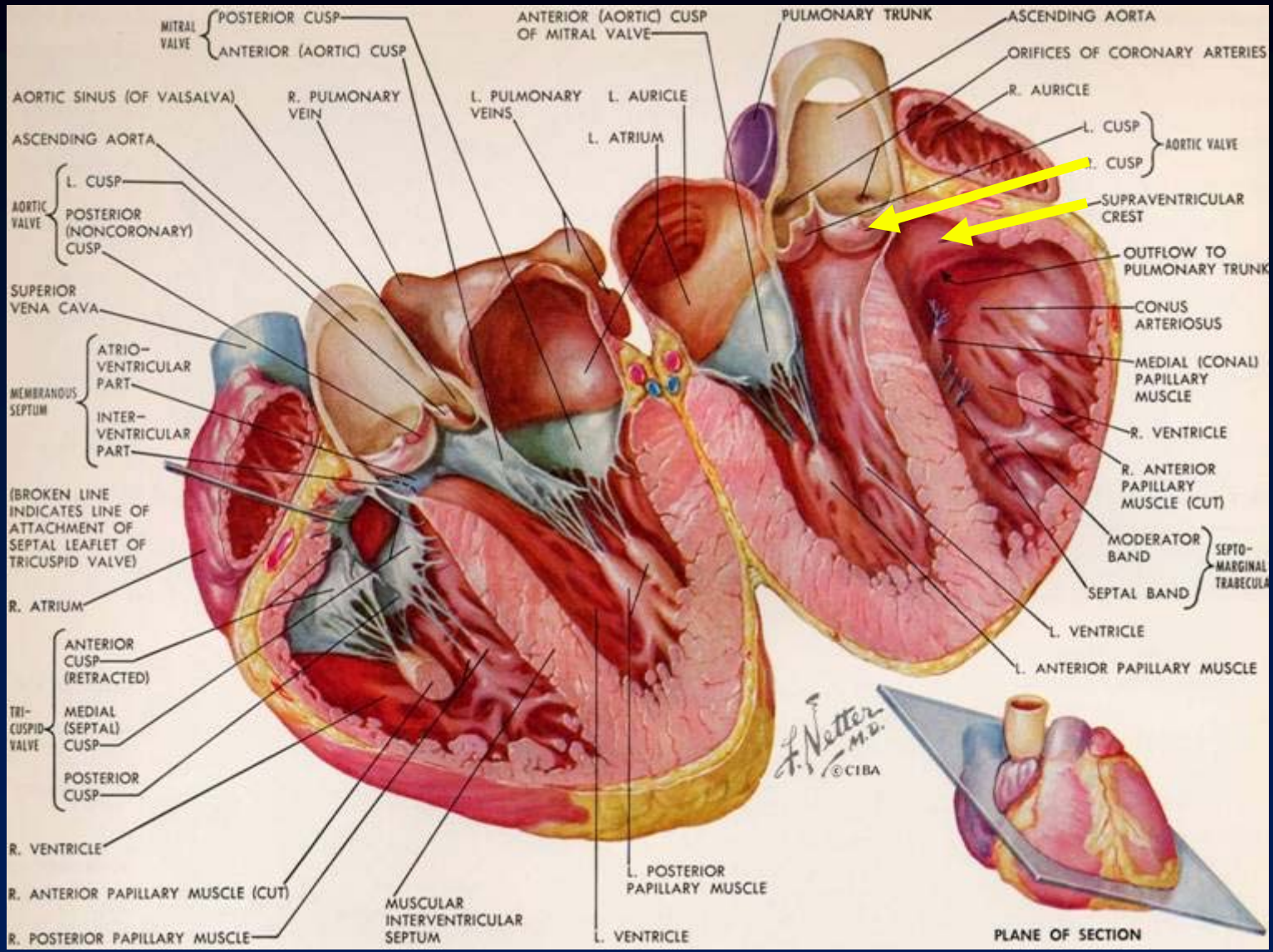
Normal



Ventricular Septal Defect
Supracristal

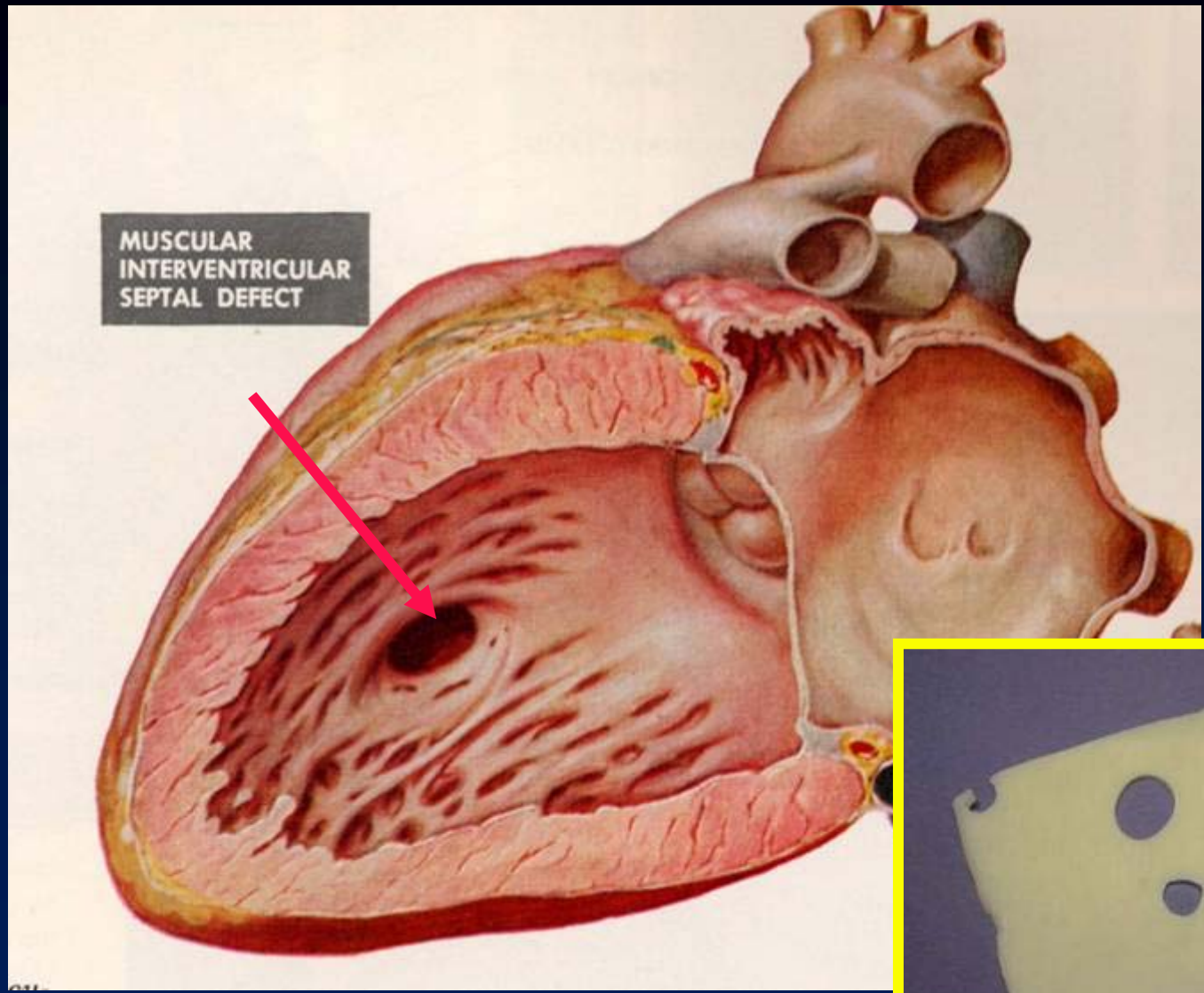
- Supracristal = conal VSD (5%–least common)
- Crista supraventricularis= inverted U-shaped muscular ridge posterior and inferior to the pulmonic valve high in interventricular septum
- On CXR: right aortic valve cusp may herniate → aortic insufficiency

LV open **RV open**

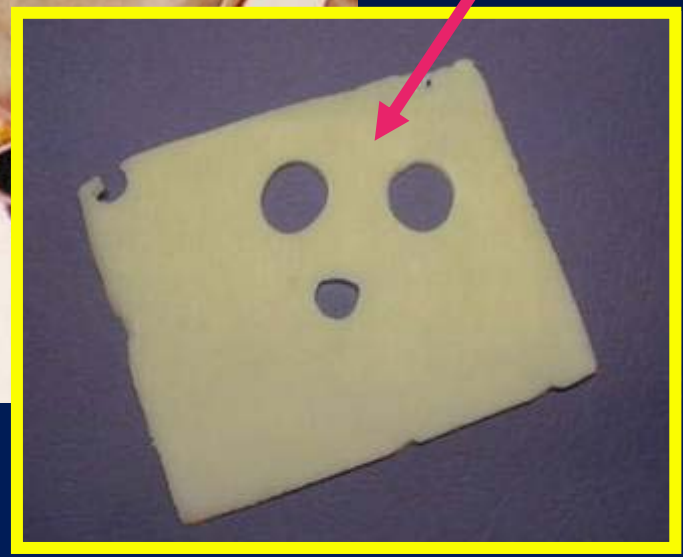


Ventricular Septal Defect
Muscular

- Muscular VSD (5–10%)
- Low and anterior within trabeculations of muscular septum
- May consist of multiple VSDs = “swiss-cheese septum”

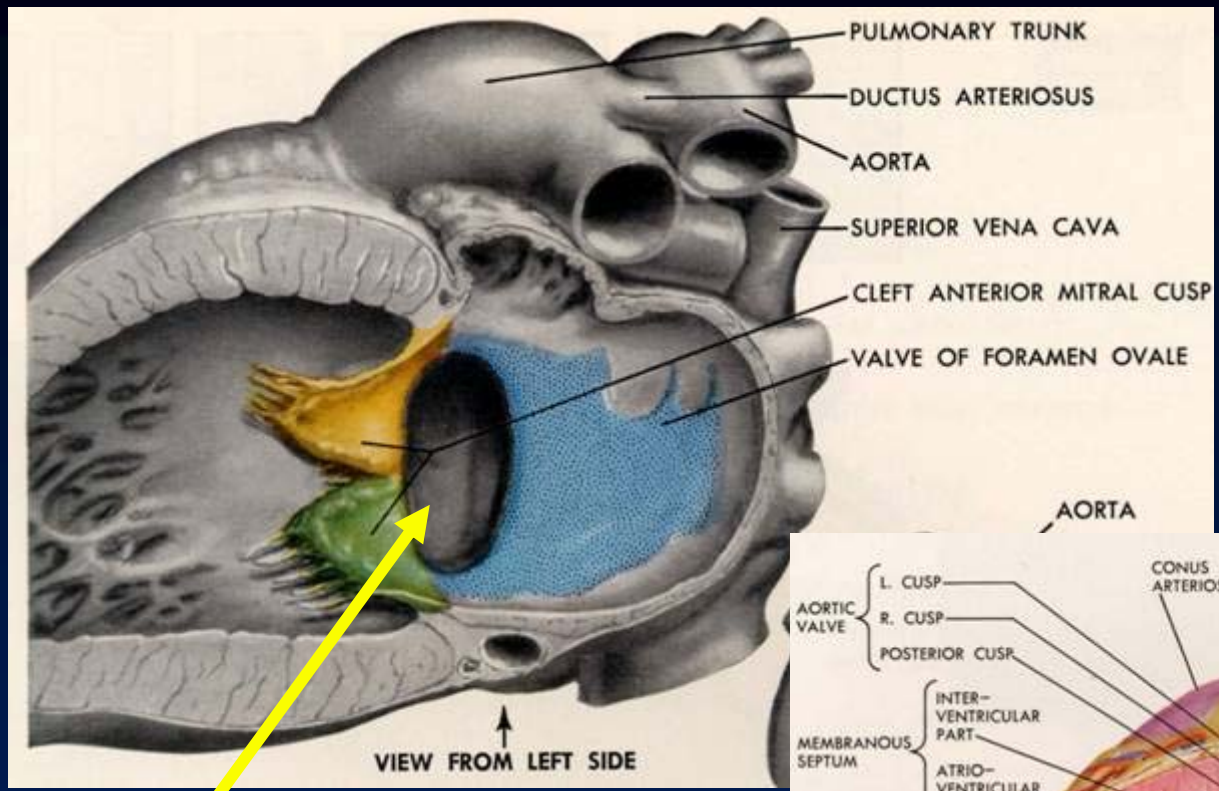


Swiss cheese

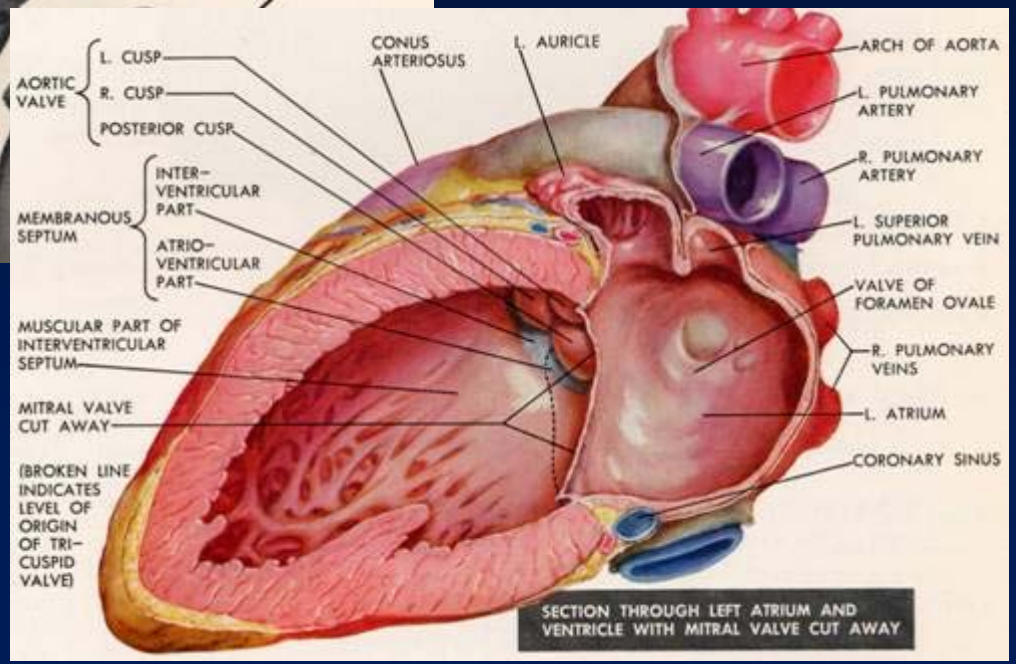


Ventricular Septal Defect
AV Canal

- Atrioventricular canal = endocardial cushion type = posterior VSD (5–10%)
- Location: adjacent to septal and anterior leaflet of mitral valve
- Large VSD → pulmonary hypertension, eventually shunt reversal
  - Eisenmenger's physiology
- Very large VSD → CHF soon after birth



Large posterior VSD (AV canal)



Ventricular Septal Defect
Natural History

- Natural history of VSD is affected by two factors:
  - Location of defect
    - Muscular and perimembranous have high incidence of spontaneous closure
    - Endocardial cushion defects have low rate of closure

Ventricular Septal Defect
Natural History

- Size of the defect
  - Larger the defect, more likely to → CHF
  - Smaller the defect, more likely to be asymptomatic

Ventricular Septal Defect
Eisenmenger Physiology

- Progressive increase in pulmonary vascular resistance
  - Intimal and medial hyperplasia ➜
  - Reversal of L ➜ R shunt to R ➜ L shunt
  - Cyanosis

Ventricular Septal Defect
Clinical Course

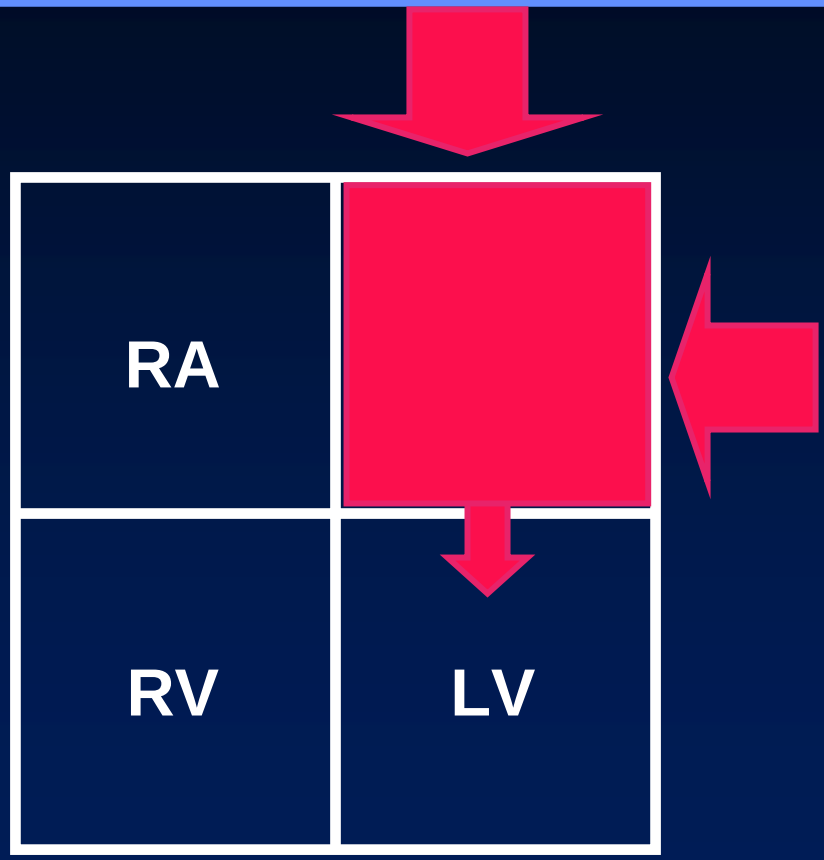
- Neonates usually asymptomatic because of high pulmonary vascular resistance from birth to 6 weeks
- Common cause of CHF in infancy
- Bacterial endocarditis may develop
- Severe pulmonary hypertension → Eisenmenger's physiology/cyanosis

Ventricular Septal Defect
X-ray Findings

- Prominent main pulmonary artery
  - Adult
- Shunt vasculature (increased flow to the lungs)
- LA enlargement (80%)
- Aorta normal in size

5 yo acyanotic male

Ventricular Septal Defect
Why Left Atrium Is Enlarged



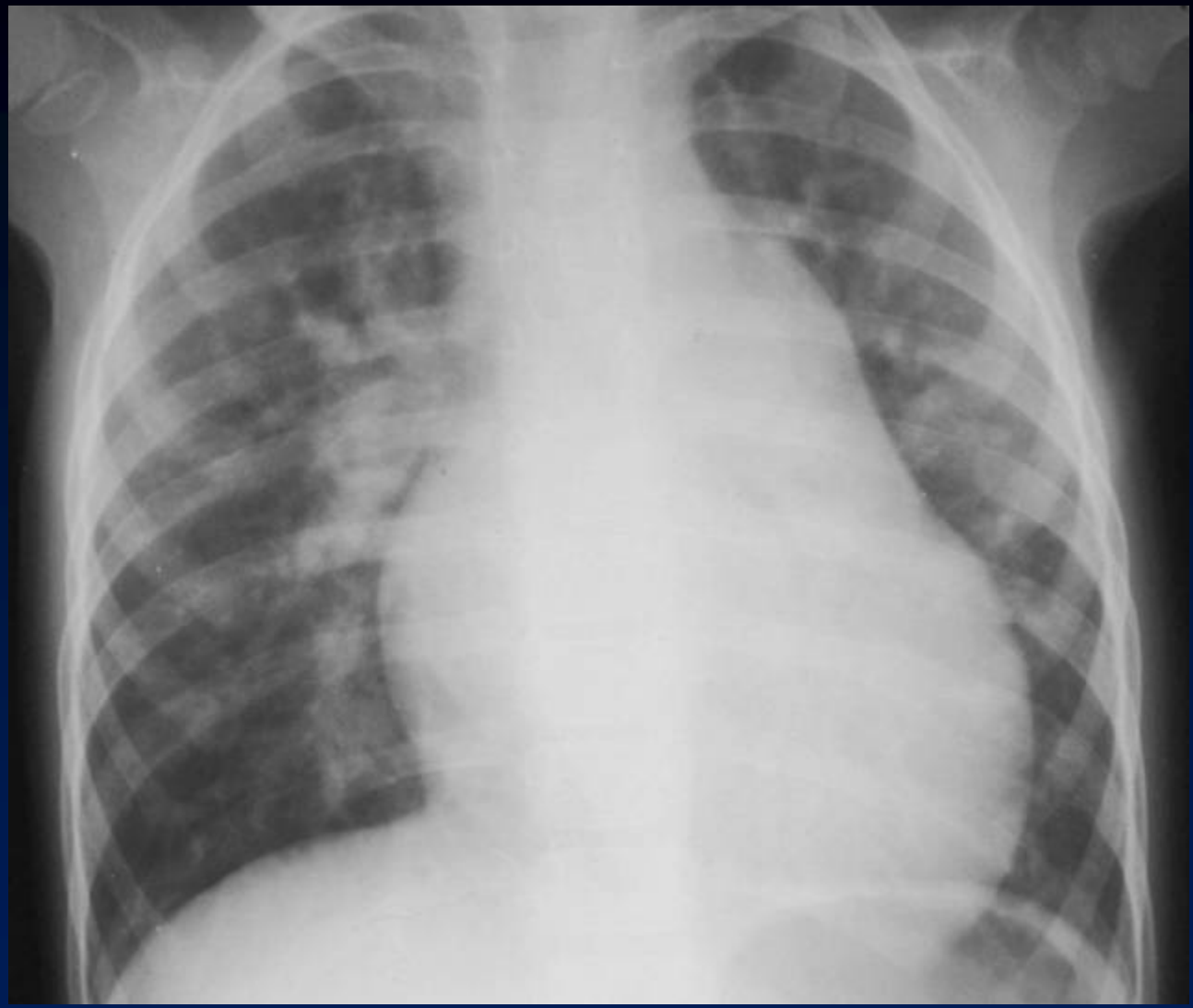
4 mos old acyanotic female

Ventricular Septal Defect
Prognosis

- Spontaneous closure occurs in
  40% during first 2 years of life
- 60% by 5 years

Ventricular Septal Defect
Indications For Surgery

- Greater than 2:1 shunt, surgery required before pulmonary arterial hypertension develops
- CHF unresponsive to medical management
- Failure to grow
- Supracristal defects because of their high incidence of AI



8 mos old acyanotic female

Patent Ductus Arteriosus

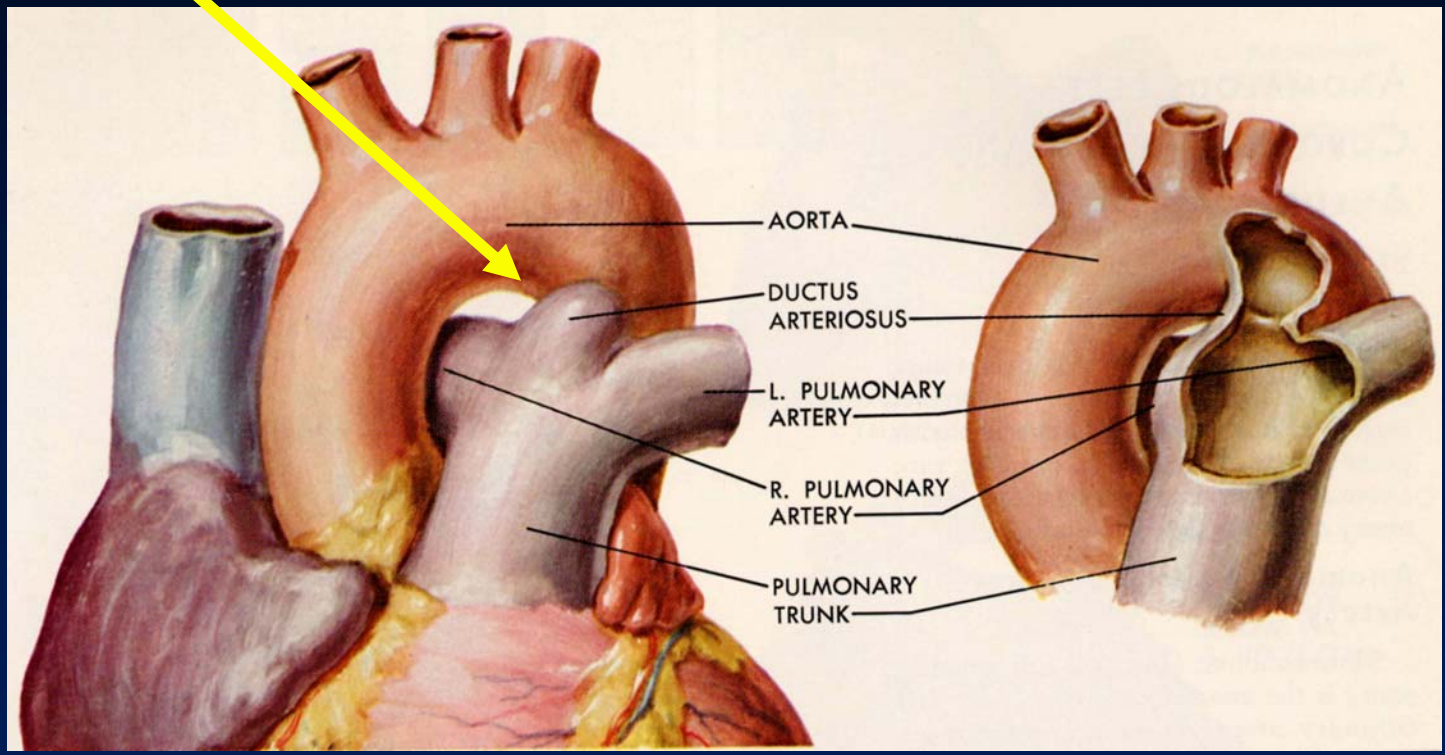
Patent Ductus Arteriosus
General

- Higher incidence in
  - Trisomy 21
  - Trisomy 18
  - Rubella
  - Preemies
- Predominance in females 4:1

Patent Ductus Arteriosus
Anatomy

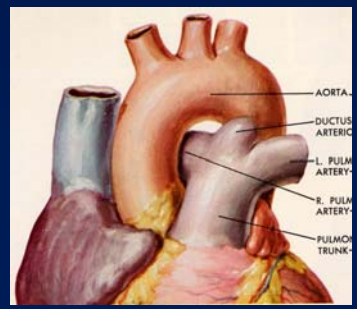
- Ductus connects pulmonary artery to descending aorta just distal to left subclavian artery

Ductus Arteriosus



Ductus Arteriosus
Physiology

- In fetal life, shunts blood from pulmonary artery to aorta
- At birth, increase in arterial oxygen concentration ↑ constriction of ductus

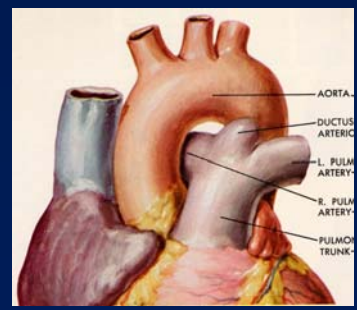


Ductus Arteriosus
Normal Closure

- Functional closure
  - By 24 hrs of life
- Normal anatomic closure
  - Complete by 2 months in 90%
- Closure at 1 year in 99%

Patent Ductus Arteriosus
Pathophysiology

- Ductus may persist
  - Because of defect in muscular wall of ductus, or
  - Chemical defect in response to oxygen
- Anatomic persistence of ductus beyond 4 months is abnormal
- Blood is shunted from aorta to pulmonary arteries



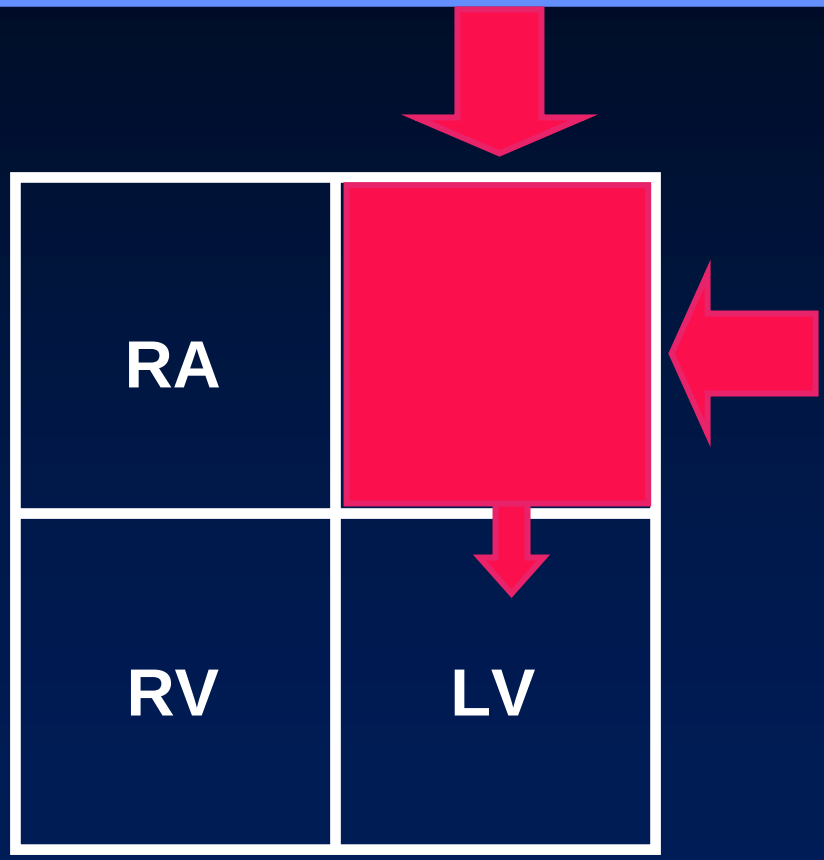
Patent Ductus Arteriosus
Clinical

- Common cause of CHF in premature infants
  - Usually at age 1 week (after HMD subsides and pulmonary arterial pressure falls)
- Wide pulse pressure
- Continuous murmur

Patent Ductus Arteriosus
X-ray Findings

- Cardiomegaly
- Enlarged left atrium
- Prominent main pulmonary artery (adult)
- Prominent peripheral pulmonary vasculature
- Prominence of ascending aorta

Patent Ductus Arteriosus
Why Left Atrium Is Enlarged



Patent Ductus Arteriosus
Calcifications

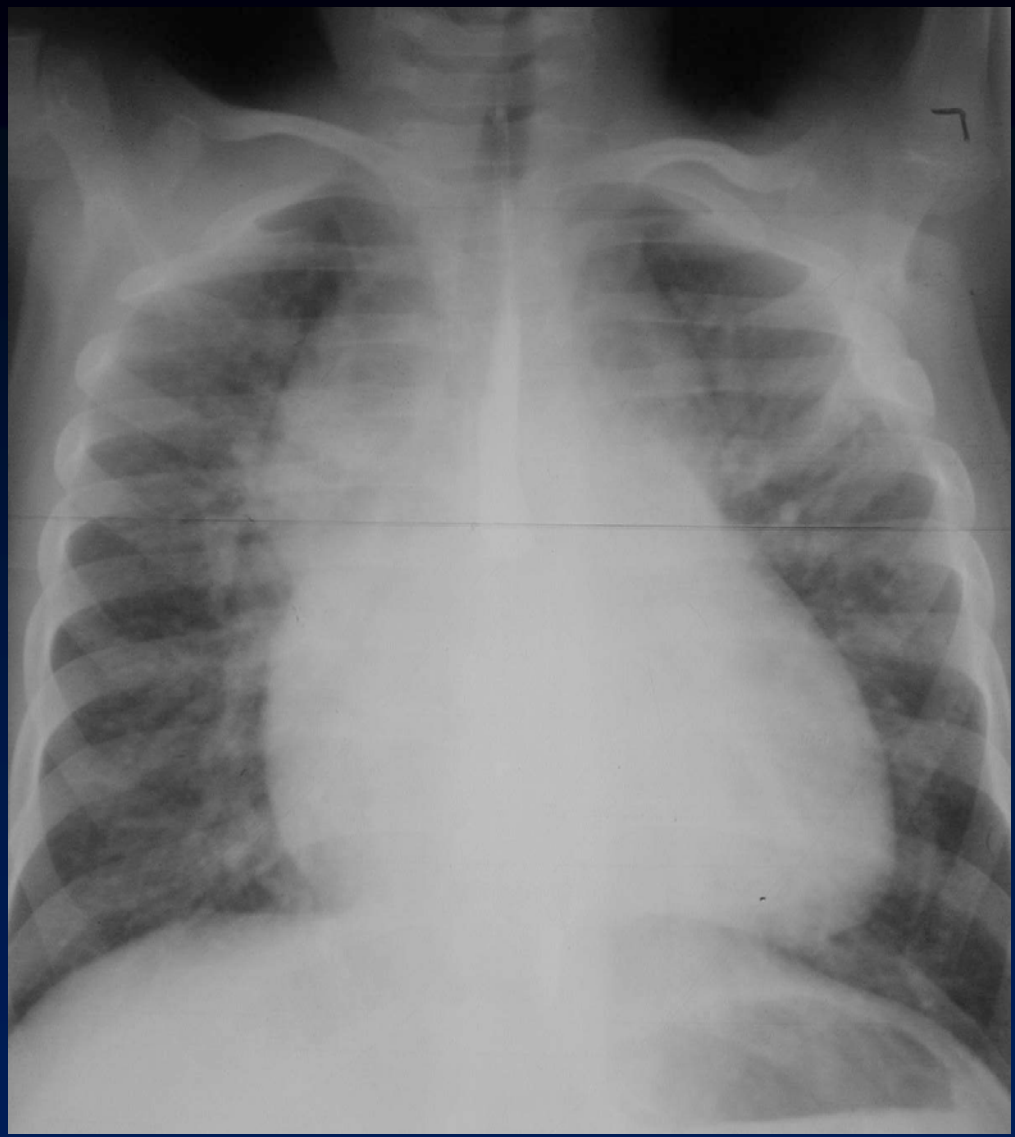
- Punctate calcification at site of closed ductus is normal finding
- Linear or railroad track calcification at site of ductus may be seen in adults with PDA

Patent Ductus Arteriosus
Prognosis

- Spontaneous closure may occur

Patent Ductus Arteriosus
Complications

- CHF
- Failure to grow
- Pulmonary infections
- Bacterial endocarditis
- Eisenmenger's physiology with advanced lesions



2 yo old cyanotic female

Partial or Total Anomalous Pulmonary Venous Return

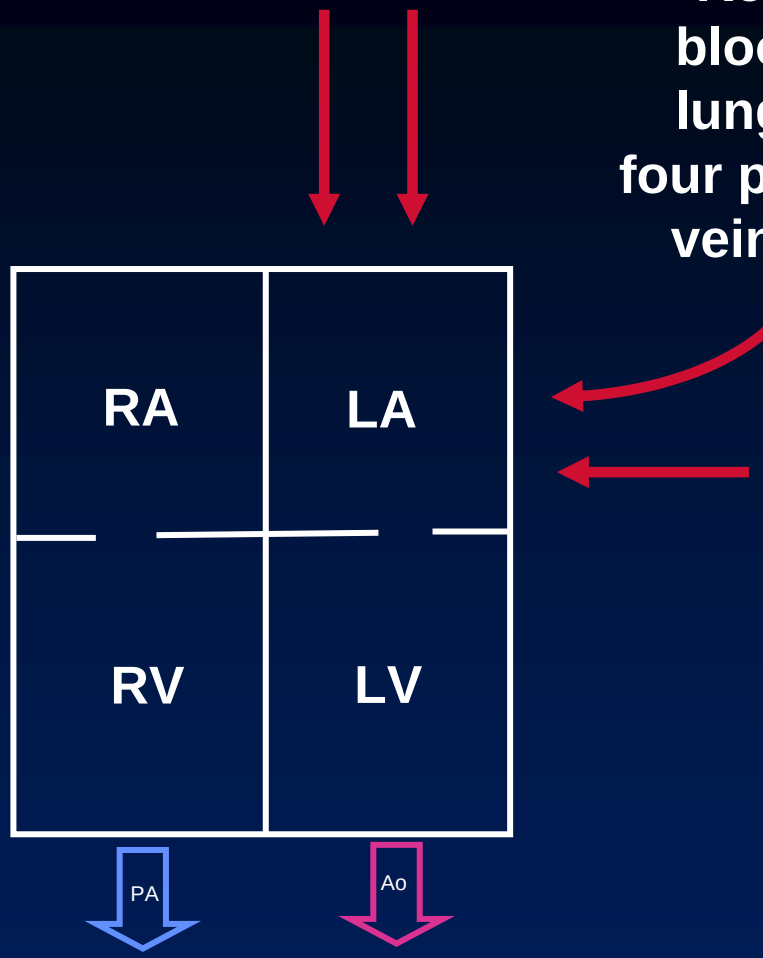
Cyanosis With Increased Vascularity

- Truncus types I, II, III
- TAPVR
- Tricuspid atresia*
- Transposition*
- Single ventricle

* Also appears on DDx of Cyanosis with Inc Vascularity

Two Types

- Partial (PAPVR)
  - Mild physiologic abnormality
  - Usually asymptomatic
- Total (TAPVR)
  - Serious physiologic abnormalities



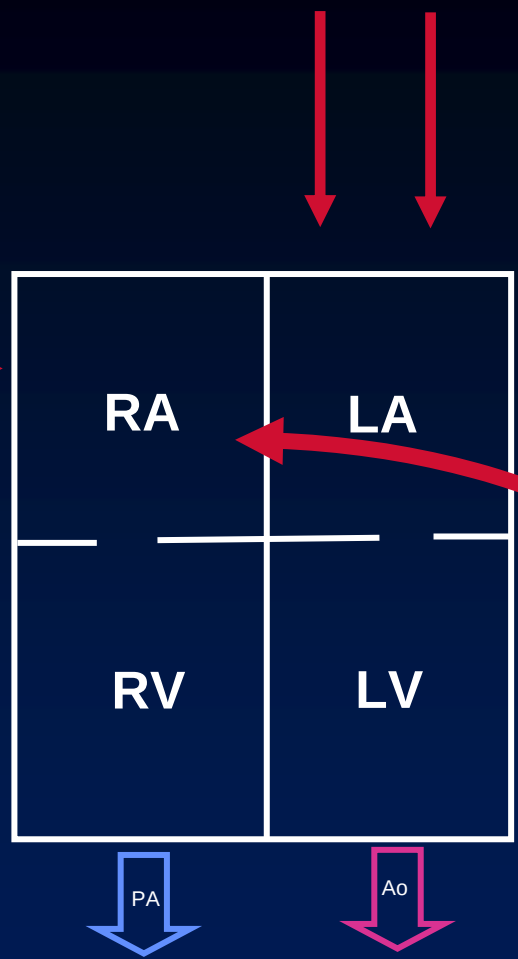
Normal heart

PAPVR
General

- One of the four pulmonary veins may drain into right atrium
- Mild or no physiologic consequence
- Associated with ASD
  - Sinus venosus or ostium secundum types

Return of blood from lungs is mostly to LA

One vein abnormally connected to right heart



Frequently associated with sinus venosus or secundum ASD

Partial Anomalous Pulmonary Return

TAPVR
General

- All have shunt through lungs to Ü R side of heart
- All must also have R → L shunt for survival
  - Obligatory ASD to return blood to the systemic side
- All are cyanotic
- Identical oxygenation in all four chambers

TAPVR
Types

- Supracardiac
- Cardiac
- Infracardiac
- Mixed

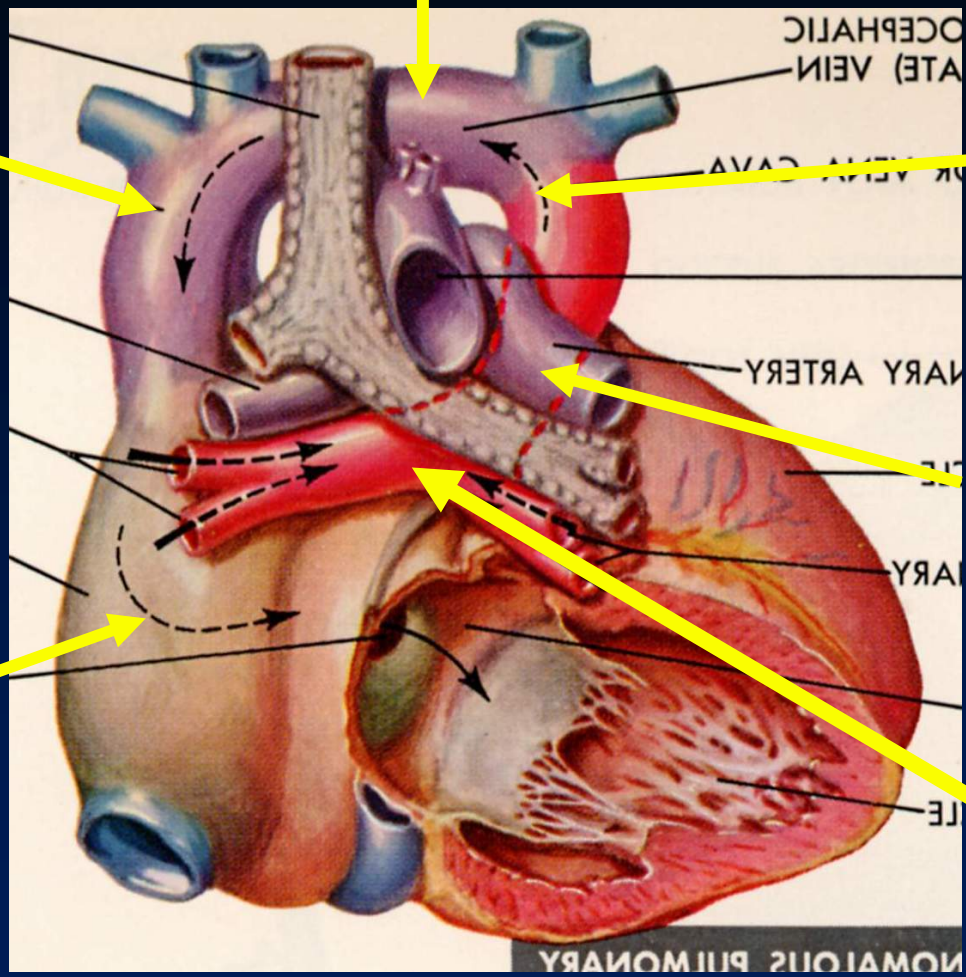
TAPVR
Supracardiac Type—Type I

- Most common (52%)
- Pulmonary veins drain into vertical vein (behind left pulmonary artery) → left brachiocephalic vein → SVC
- DDx: VSD with large thymus

Left Brachiocephalic vein

Right superior vena cava

Left superior vena cava

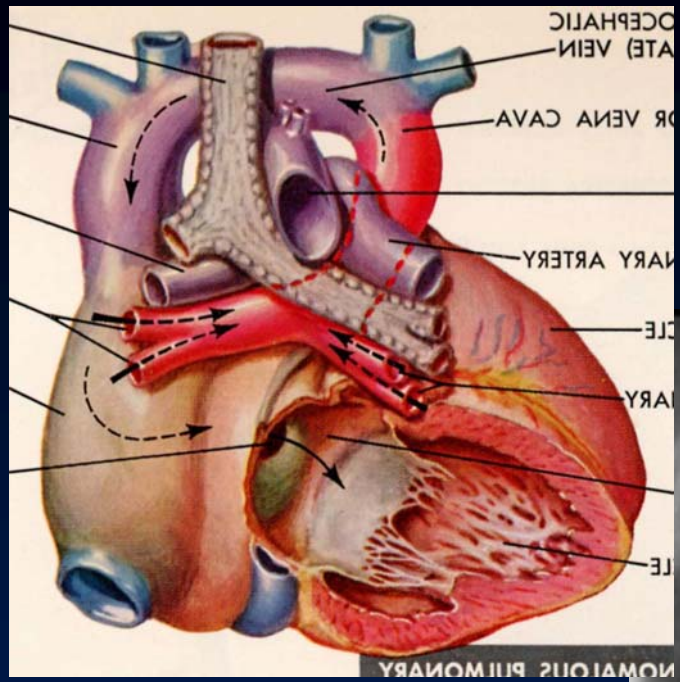


Vertical vein

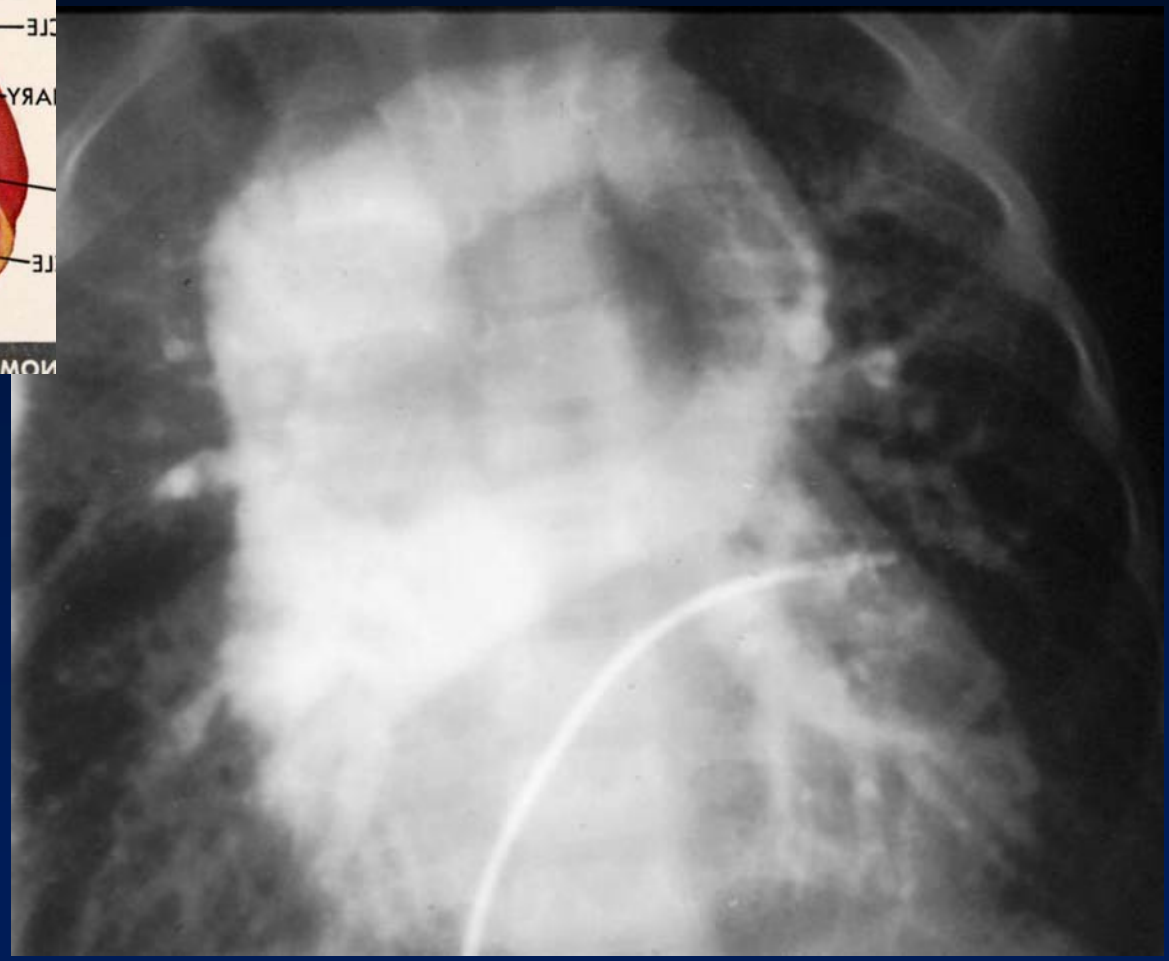
Right atrium

Pulmonary veins

TAPVR-Supracardiac Type 1

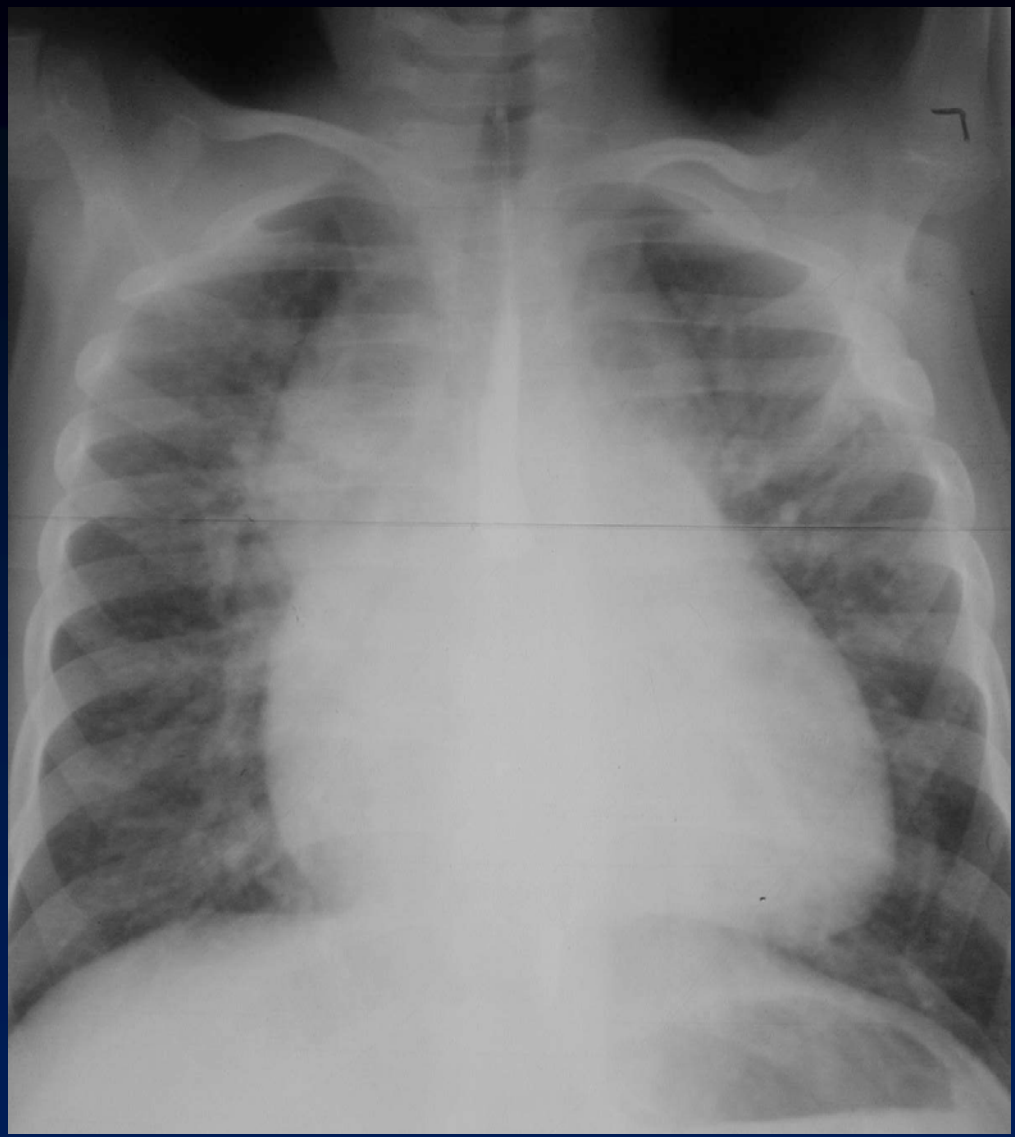


TAPVR-Supracardiac Type 1

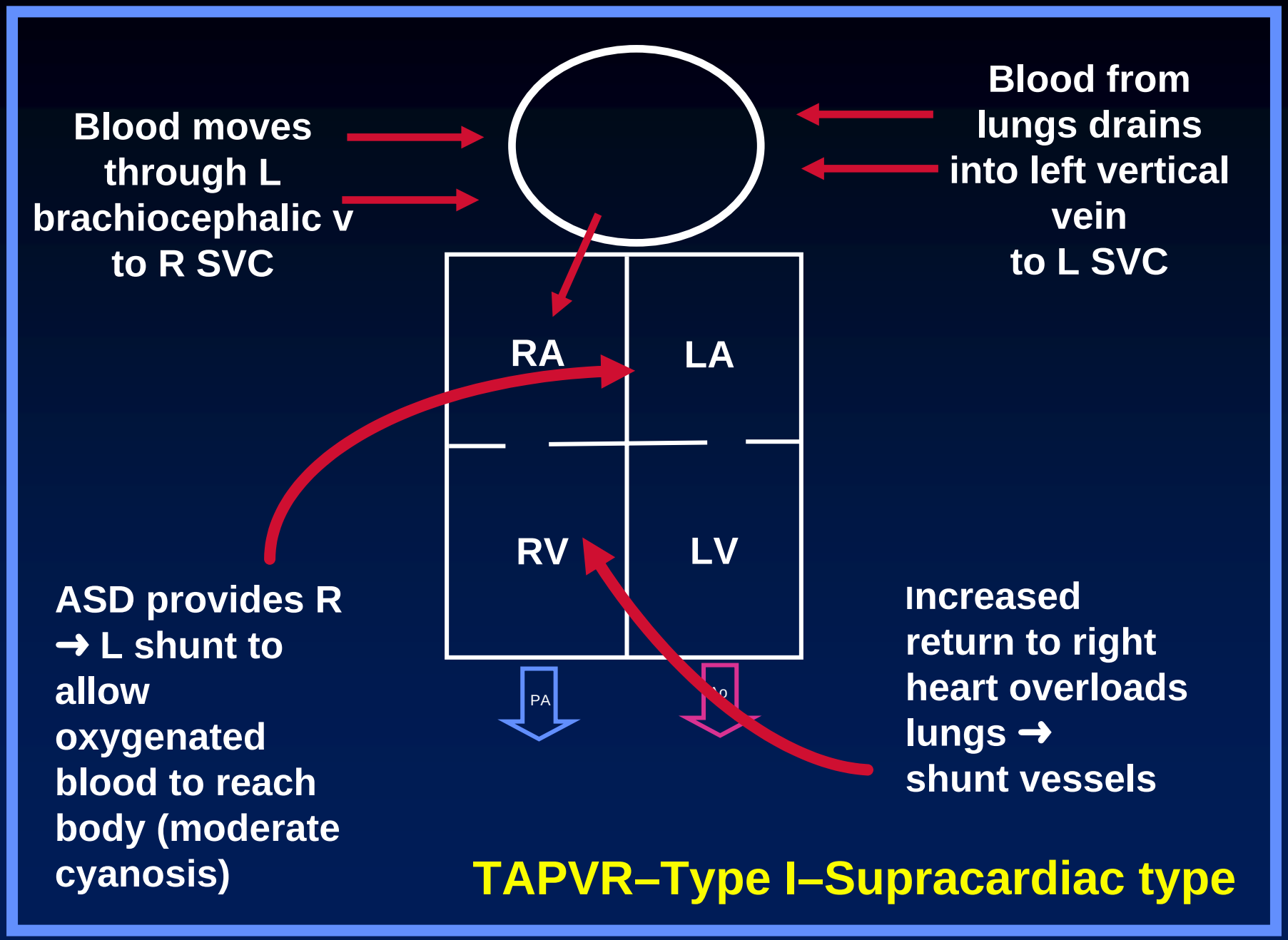


TAPVR
Supracardiac Type 1—X-ray Findings

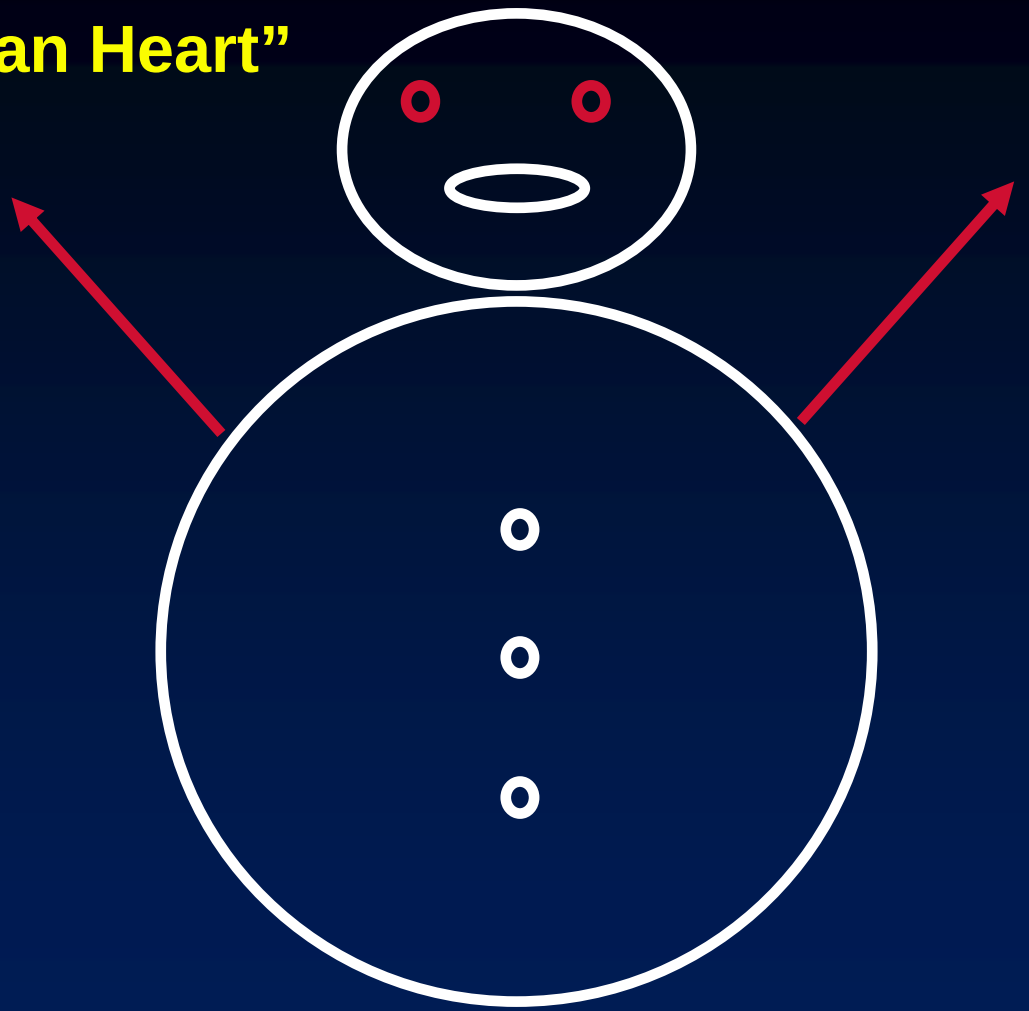
- Snowman heart = dilated SVC+ left vertical vein
- Shunt vasculature 2° increased return to right heart
- Enlargement of right heart 2° volume overload



TAPVR-Supracardiac Type 1


Blood moves through L brachiocephalic v to R SVC
Blood from lungs drains into left vertical vein to L SVC
RA
LA
RV
LV
PA
ASD provides R → L shunt to allow oxygenated blood to reach body (moderate cyanosis)
Increased return to right heart overloads lungs → shunt vessels
TAPVR–Type I–Supracardiac type

“Snowman Heart”

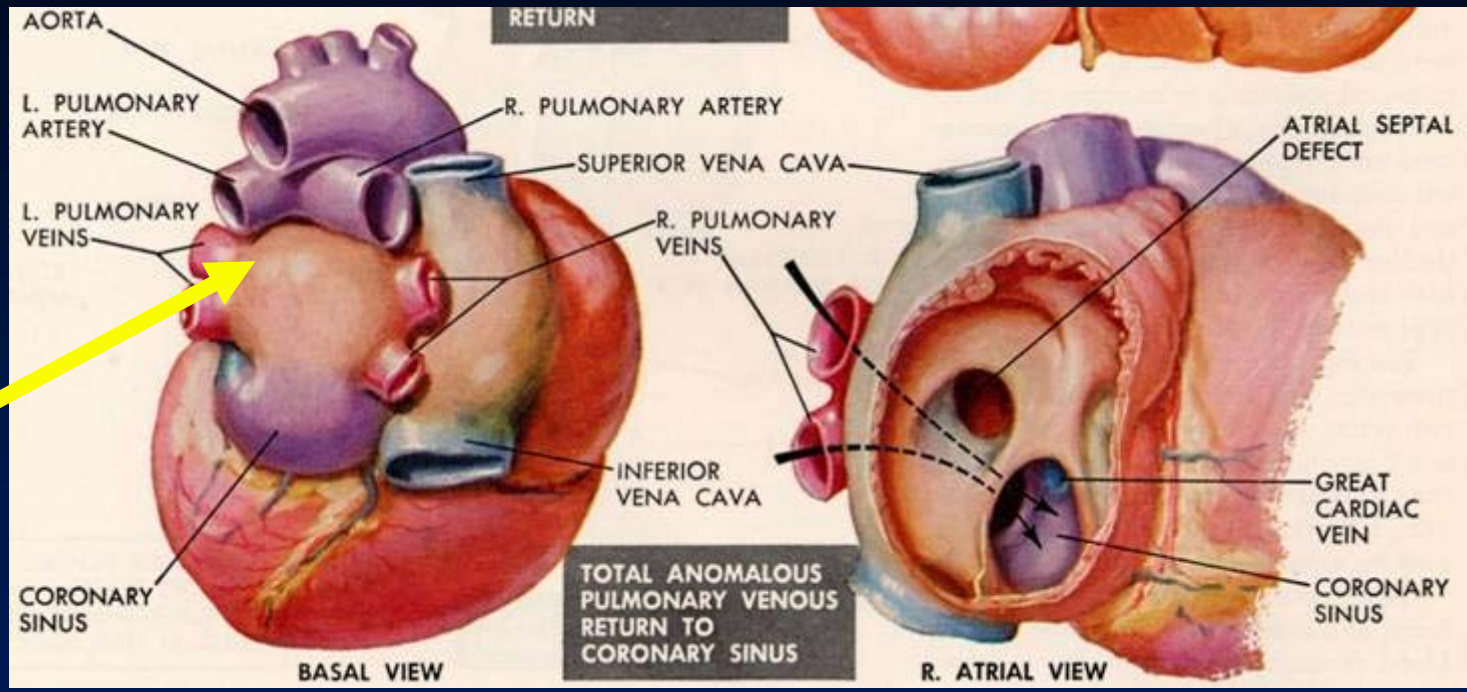


TAPVR–Type I–Supracardiac type

TAPVR
Cardiac Type—Type II

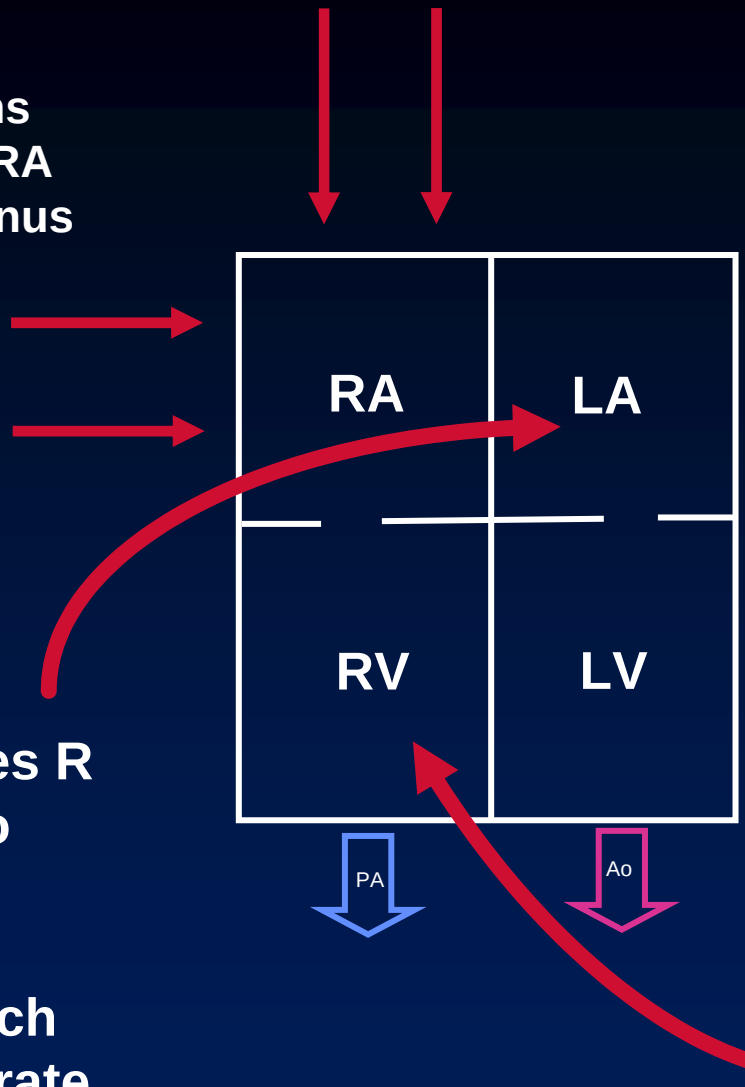
- Second most common: 30%
- Drains into coronary sinus or RA
  - Coronary sinus more common
- Increased pulmonary vasculature
- Overload of RV → CHF after birth
- 20% of I's and II's survive to adulthood
  - Remainder expire in first year

Coronary sinus



TAPVR-Coronary Sinus-Type II

Blood returns from lung to RA or coronary sinus



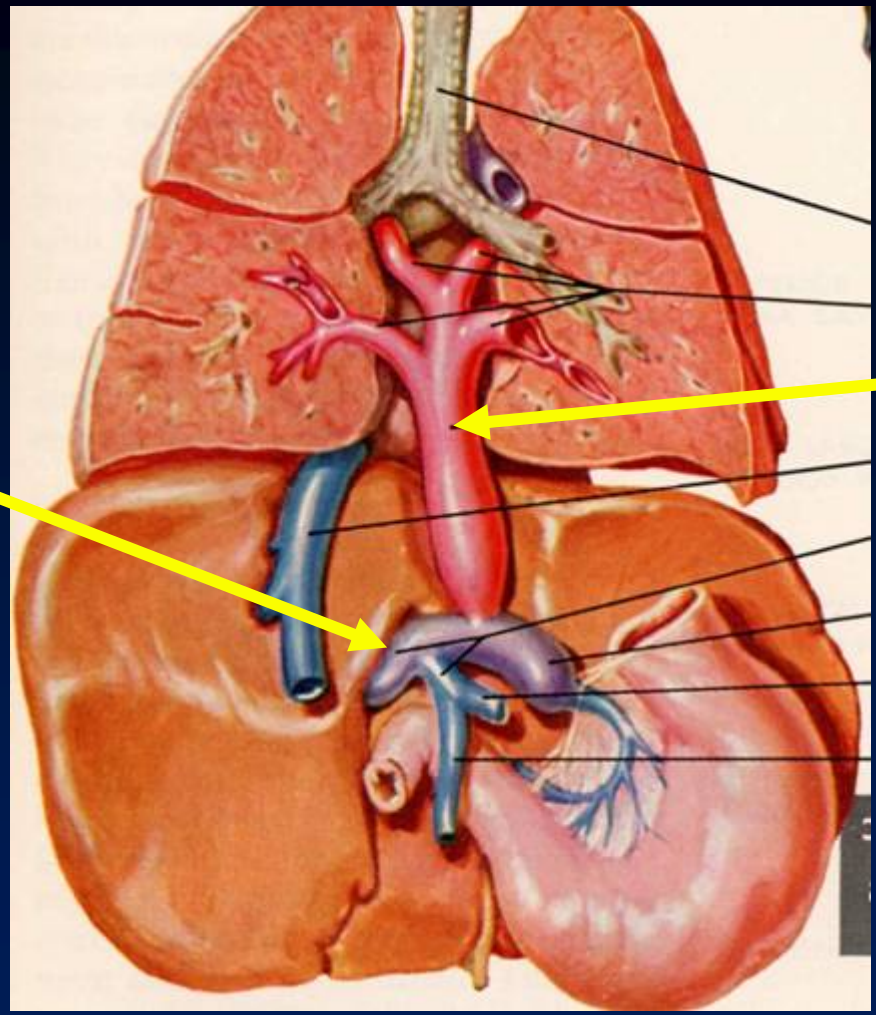
ASD provides R → L shunt to allow oxygenated blood to reach body (moderate cyanosis)

Increased return to right heart overloads lungs → shunt vessels

TAPVR–Type II–Cardiac Type

TAPVR
Infracardiac Type—Type III

- Percent of total: 12%
- Long pulmonary veins course down along esophagus
- Empty into IVC or portal vein (more common)
- Vein constricted by diaphragm as it passes through esophageal hiatus



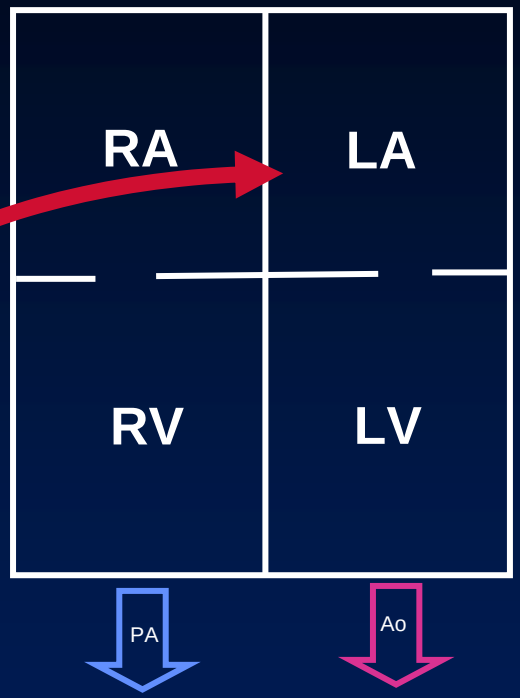
TAPVR-Type III-Infradiaphragmatic

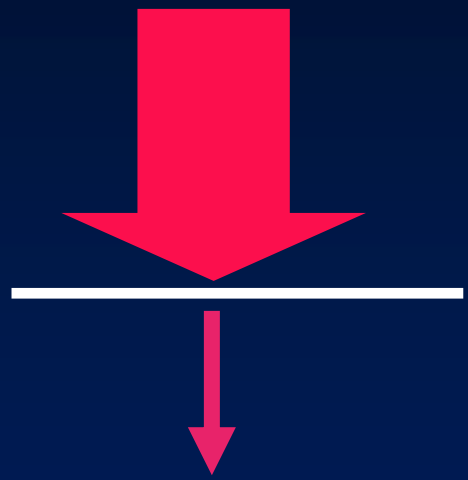
TAPVR
Infracardiac Type—Continued

- Severe CHF (90%) 2° obstruction to venous return
- Cyanotic 2° right Ü left shunt through ASD
- Associated with asplenia (80%), or polysplenia
- Prognosis=death within a few days

CHF vasculature

Blood returning from lungs → pulmonary veins which are constricted by diaphragm → CHF





ASD provides R → L shunt to allow oxygenated blood to reach body (cyanotic)

To portal v → IVC → RA

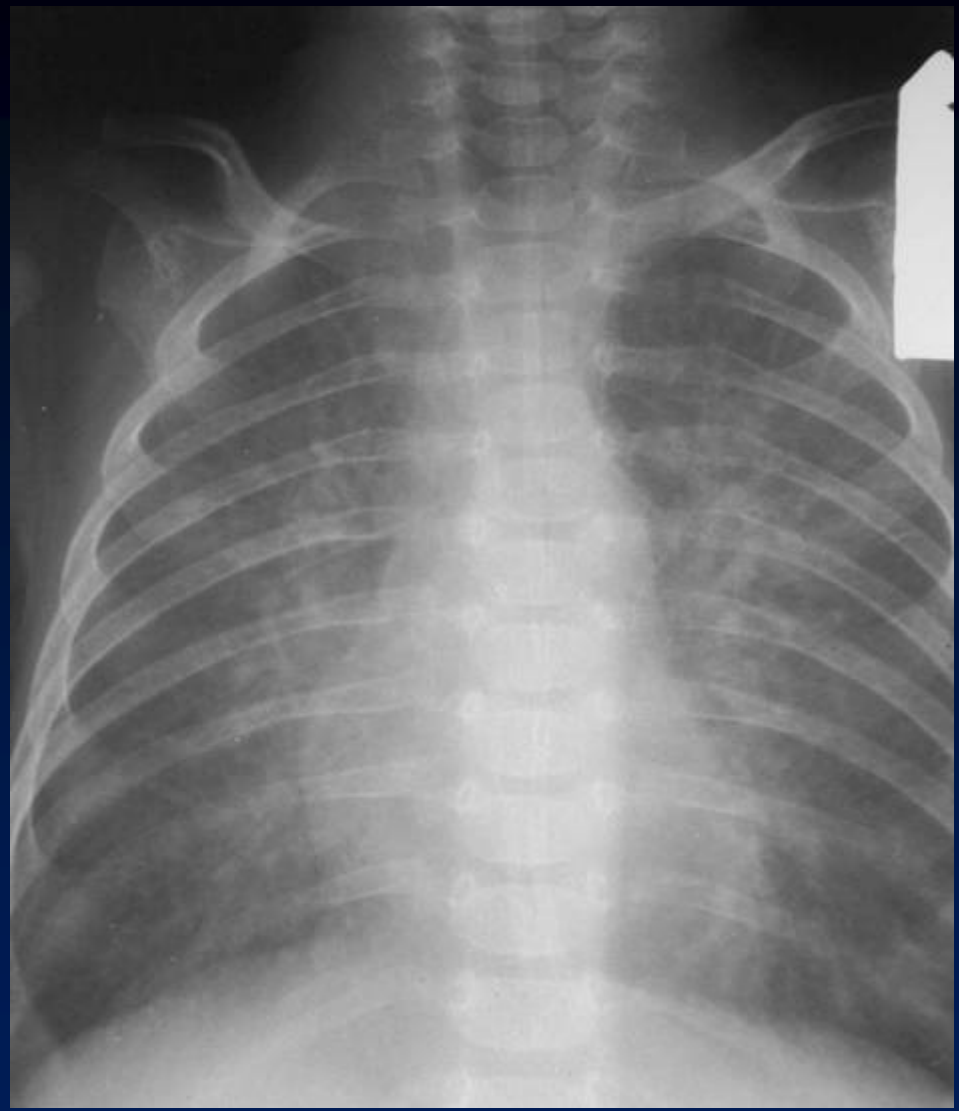
TAPVR–Type III–Infracardiac type

TAPVR
Mixed Type—Type IV

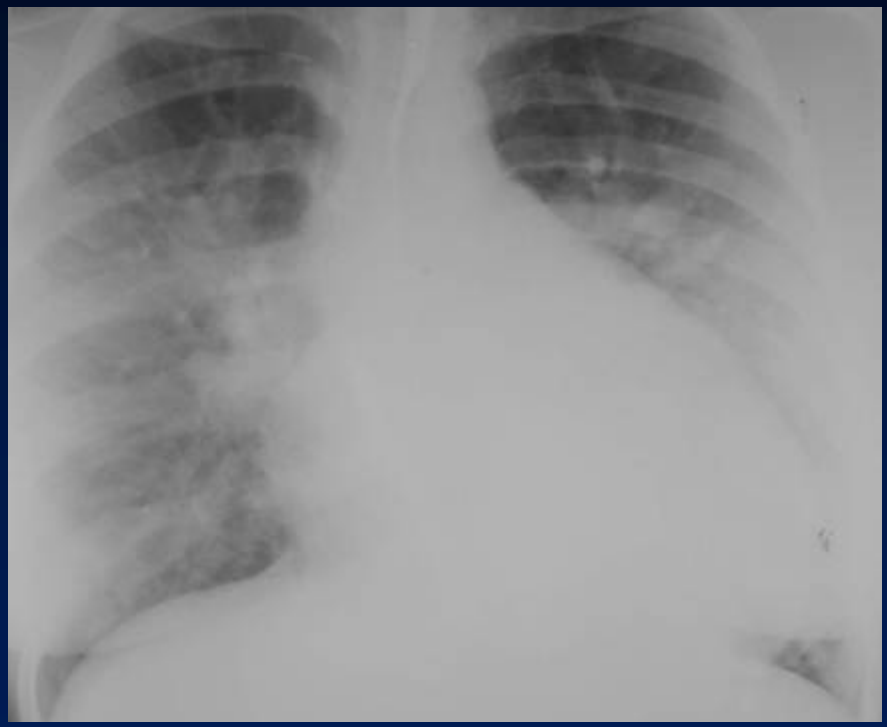
- Percent of total: 6%
- Mixtures of types I – III

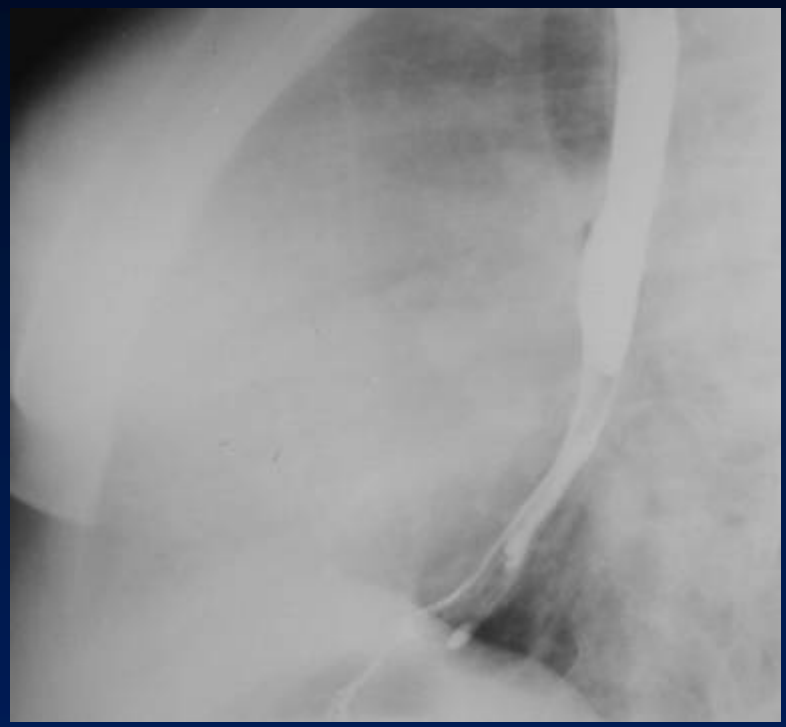
Unknowns

ASD (primum) with PAH



TAPVR from below diaphragm





VSD

ASD

The End