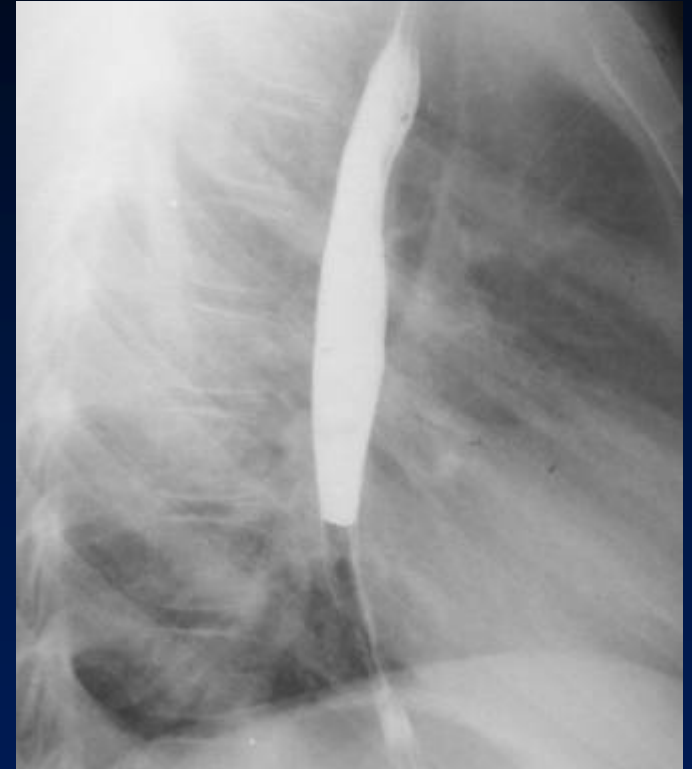


# Left to Right Shunts

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**

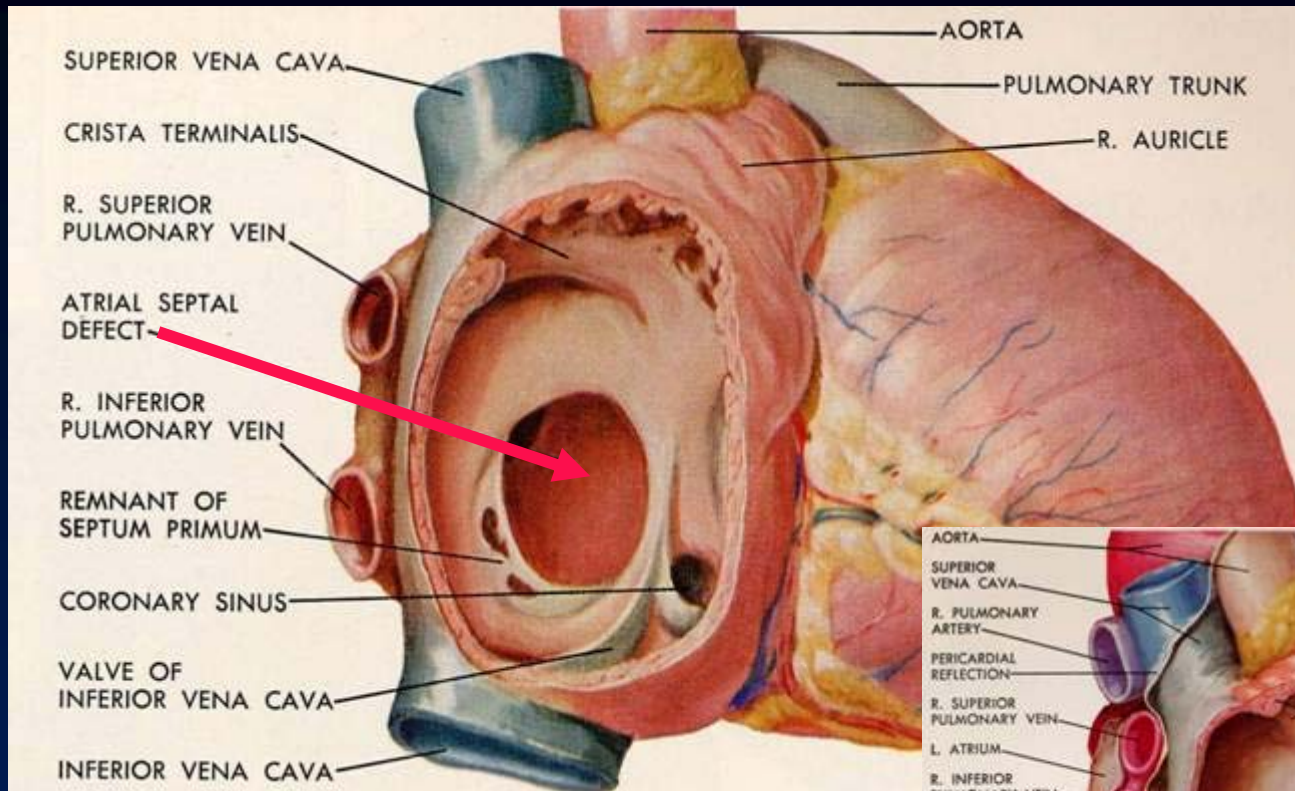
## **General**

- **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**

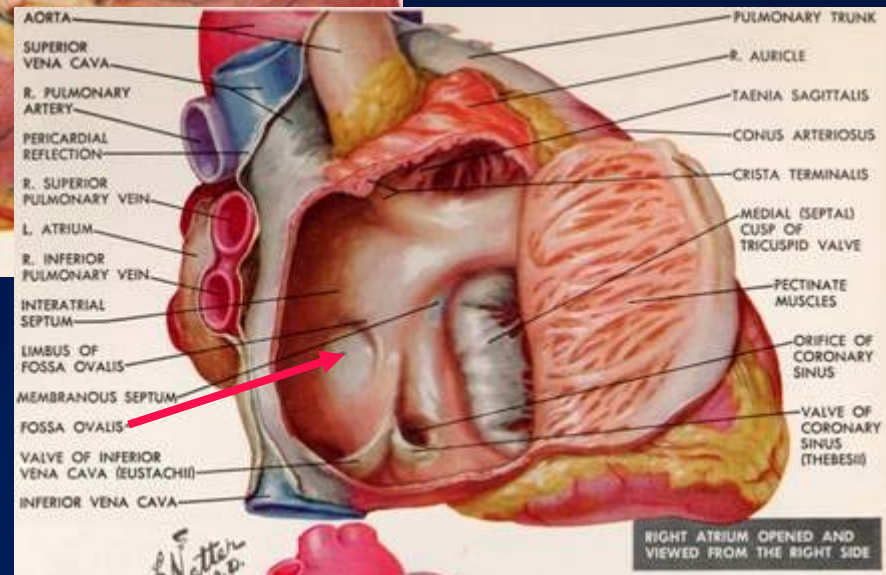
## **Ostium Secundum Type**

- **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**



RIGHT ATRIUM OPENED AND VIEWED FROM THE RIGHT SIDE

# **Atrial Septal Defect**

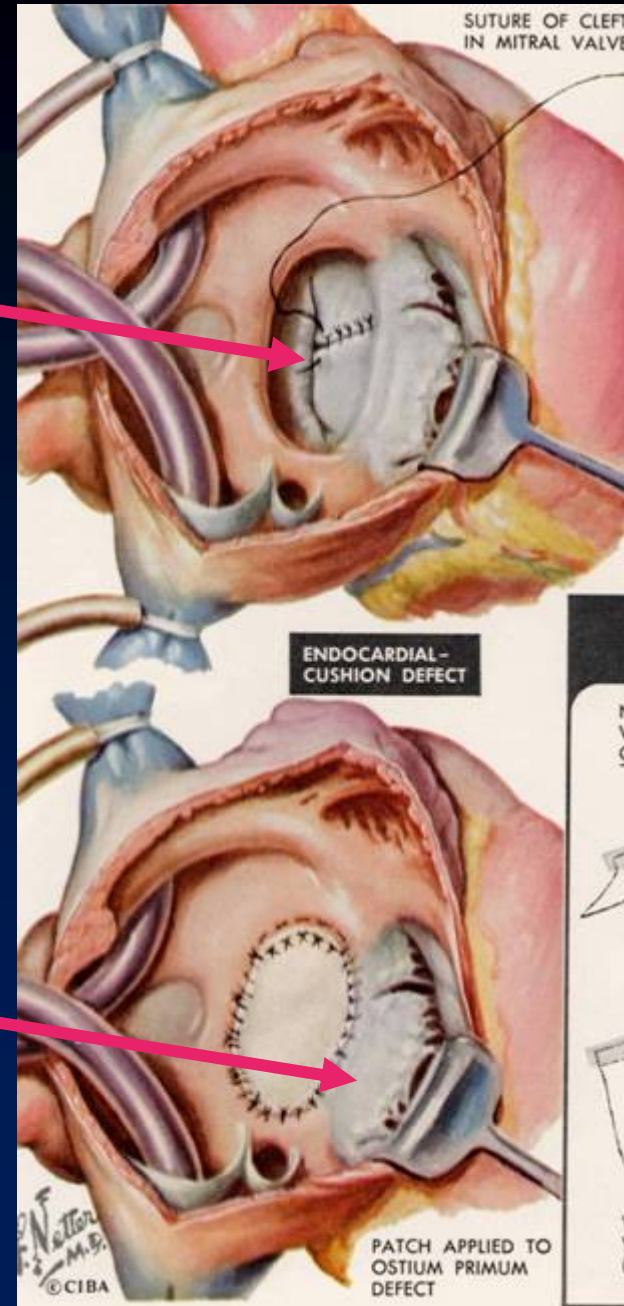
## **Ostium Primum Type**

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



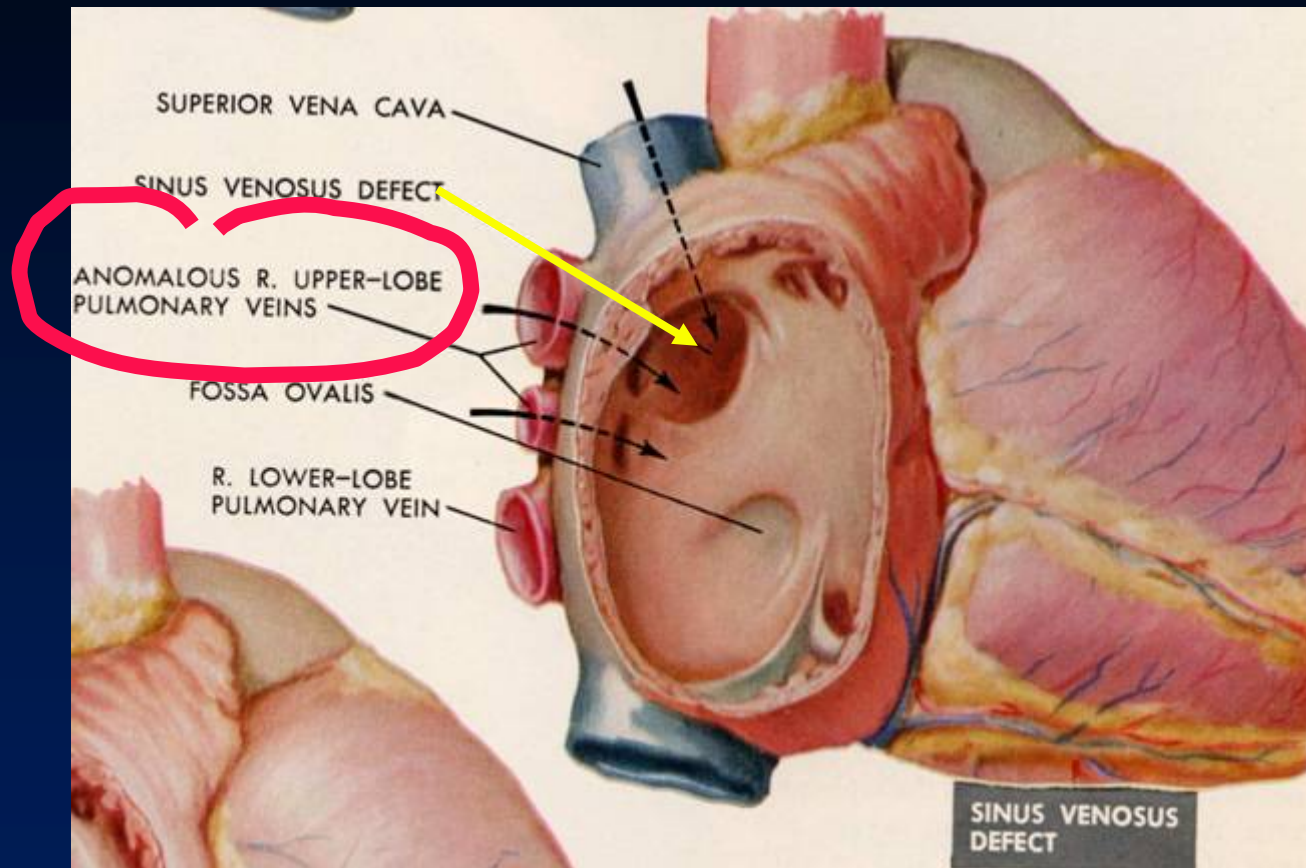
**Looking through  
ostium primum defect  
at cleft mitral valve**

**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**



© Frank Netter, MD Novartis®

**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**

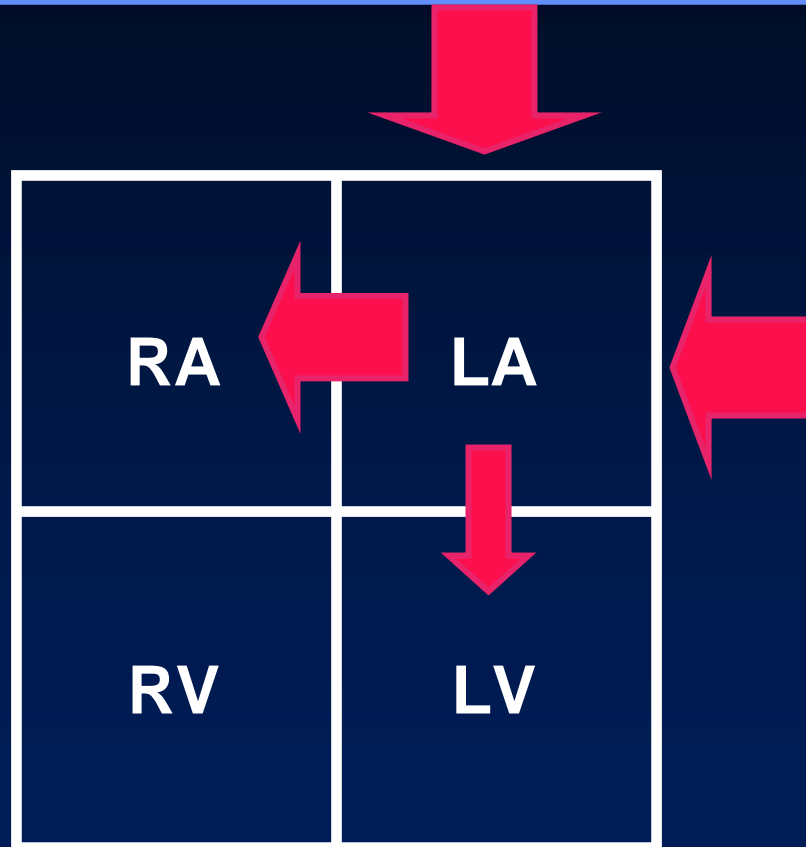
- **Large shunts associated with**
  - **Pulmonary infections and cardiac arrhythmias**
- **Higher incidence of pericardial disease with ASD than any other CHD**
- **Bacterial endocarditis is rare**

# 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**

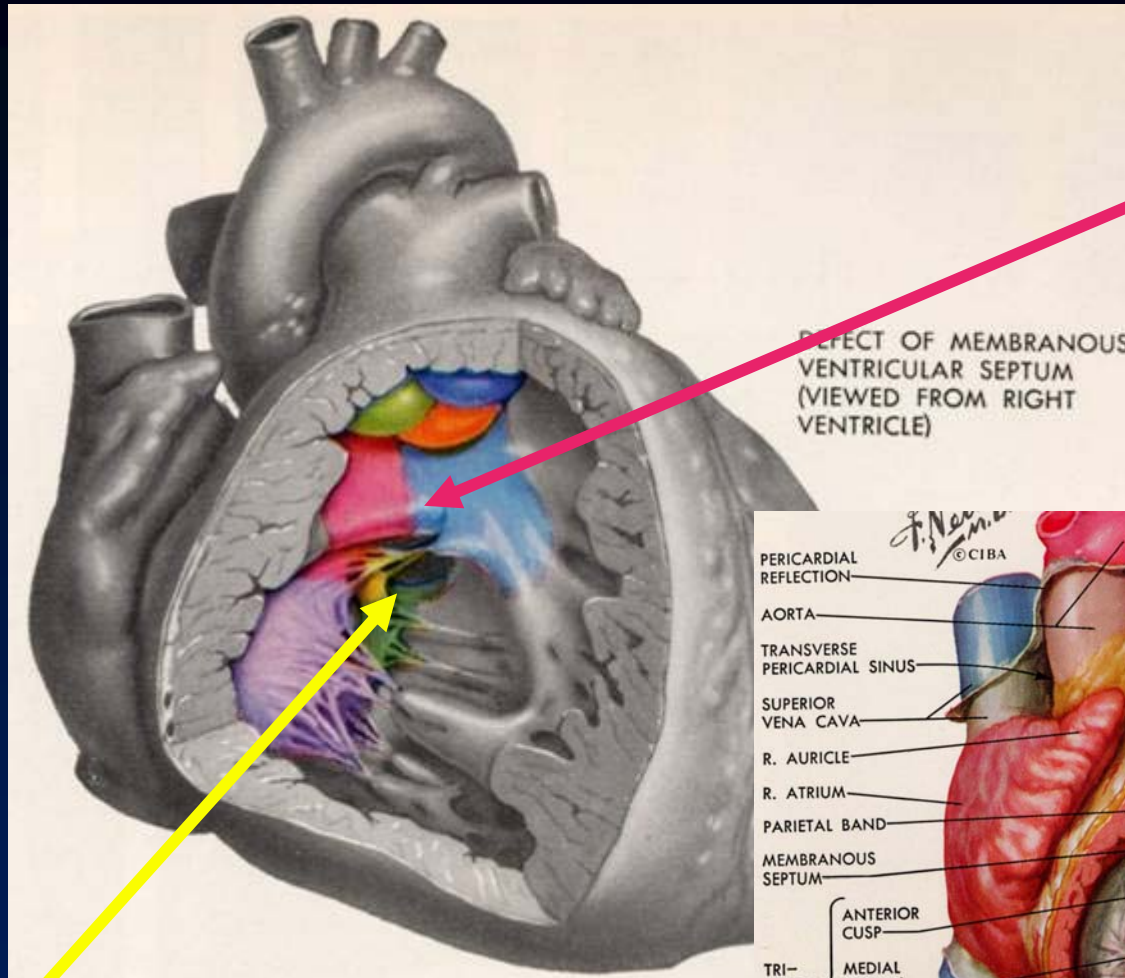
# **Ventricular Septal Defect**

## **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

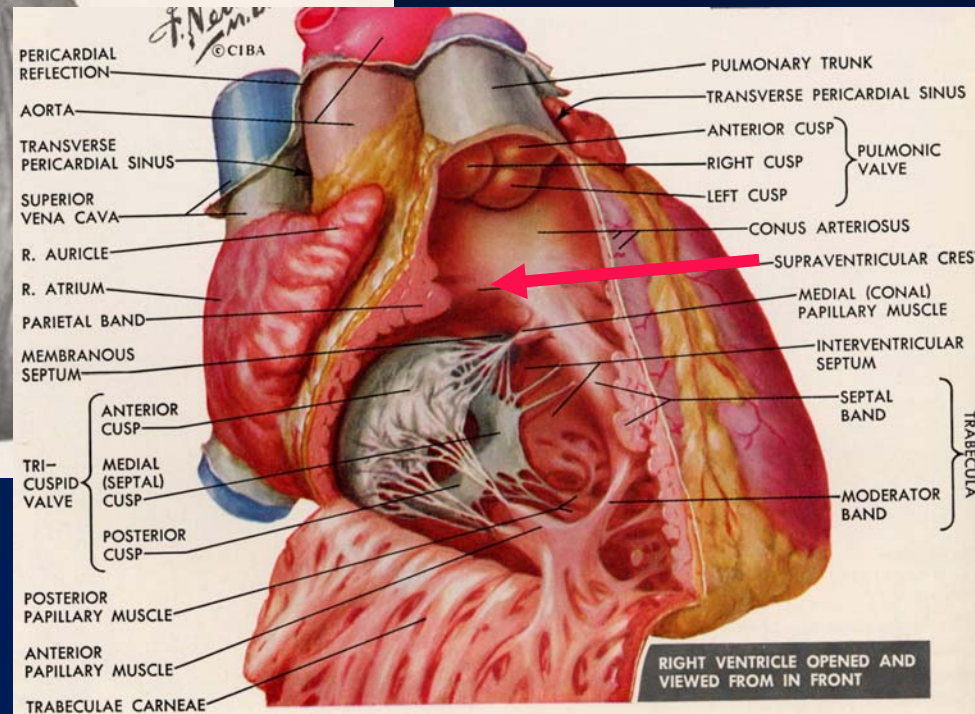


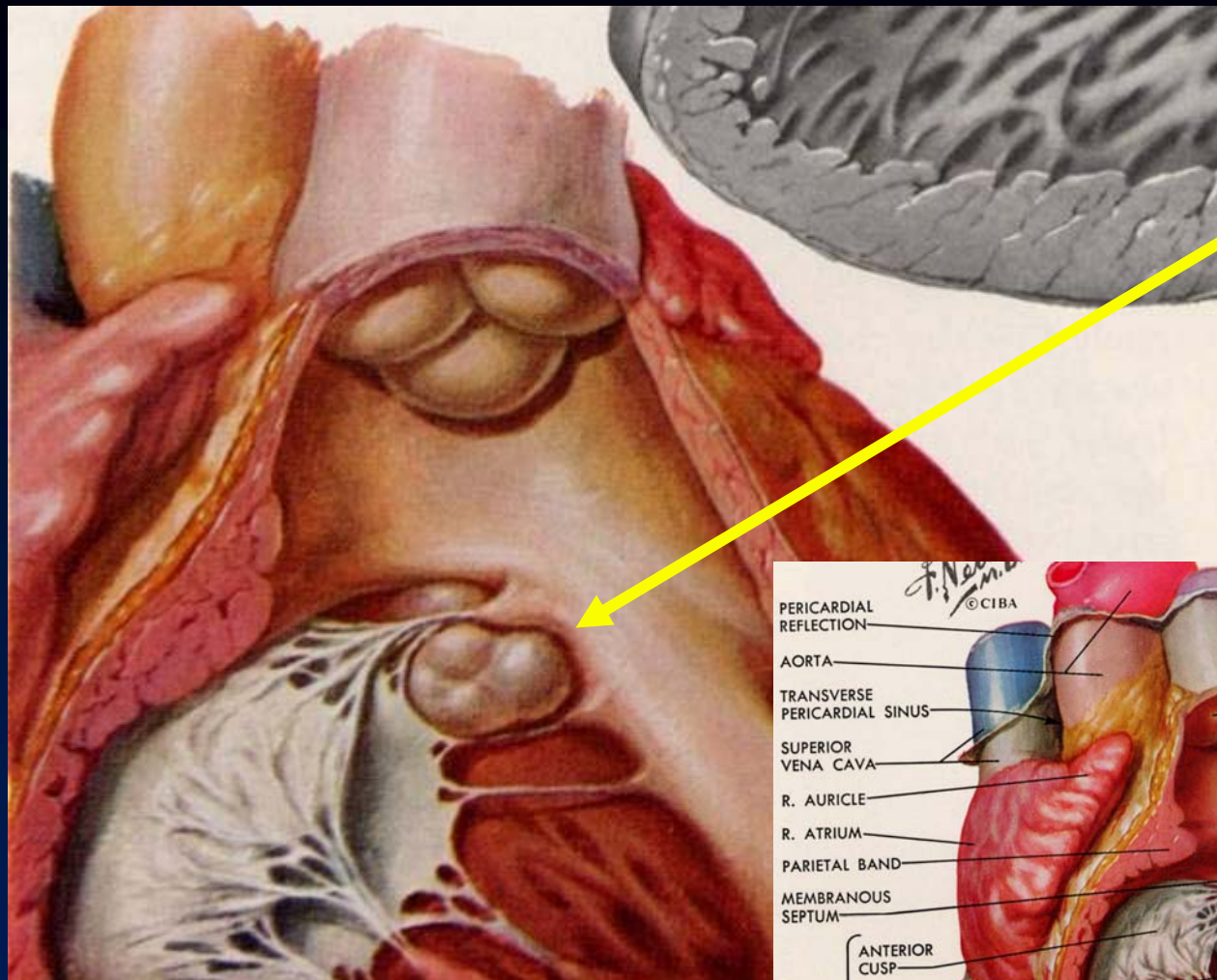
© Frank Netter, MD Novartis®

## Membranous VSD

Crista  
supraventricularis

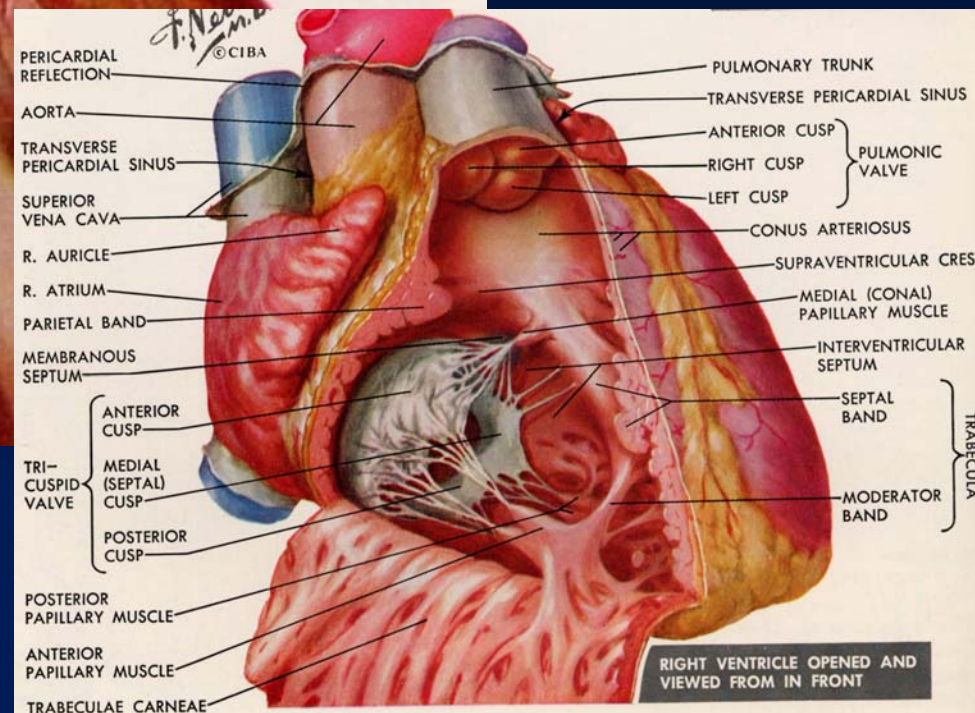
Normal





**Aneurysm of  
membranous  
septum**

**Normal**



© Frank Netter, MD Novartis®

© Frank Netter, MD Novartis®

# **Ventricular Septal Defect**

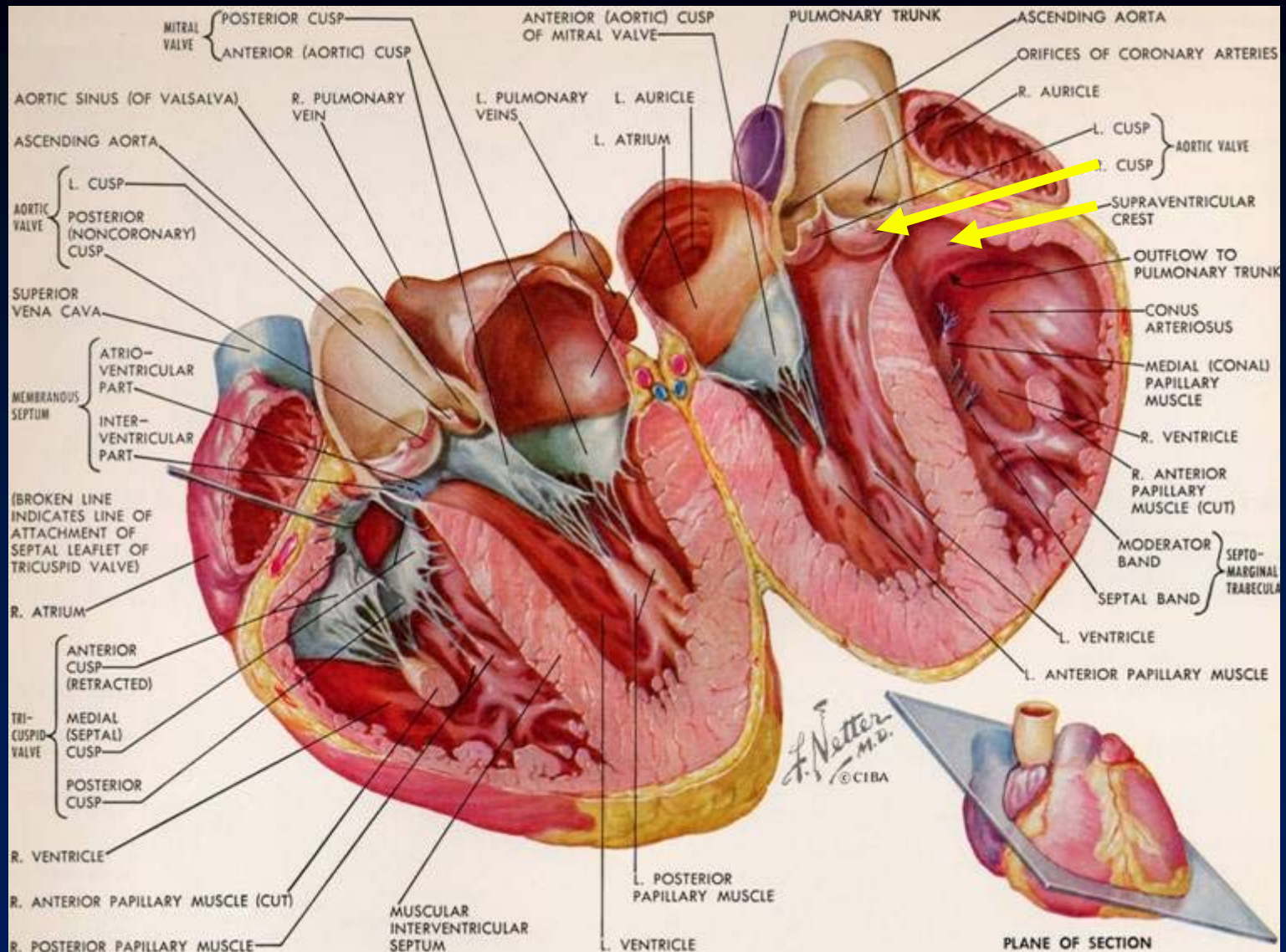
## **Supracristal**

- **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

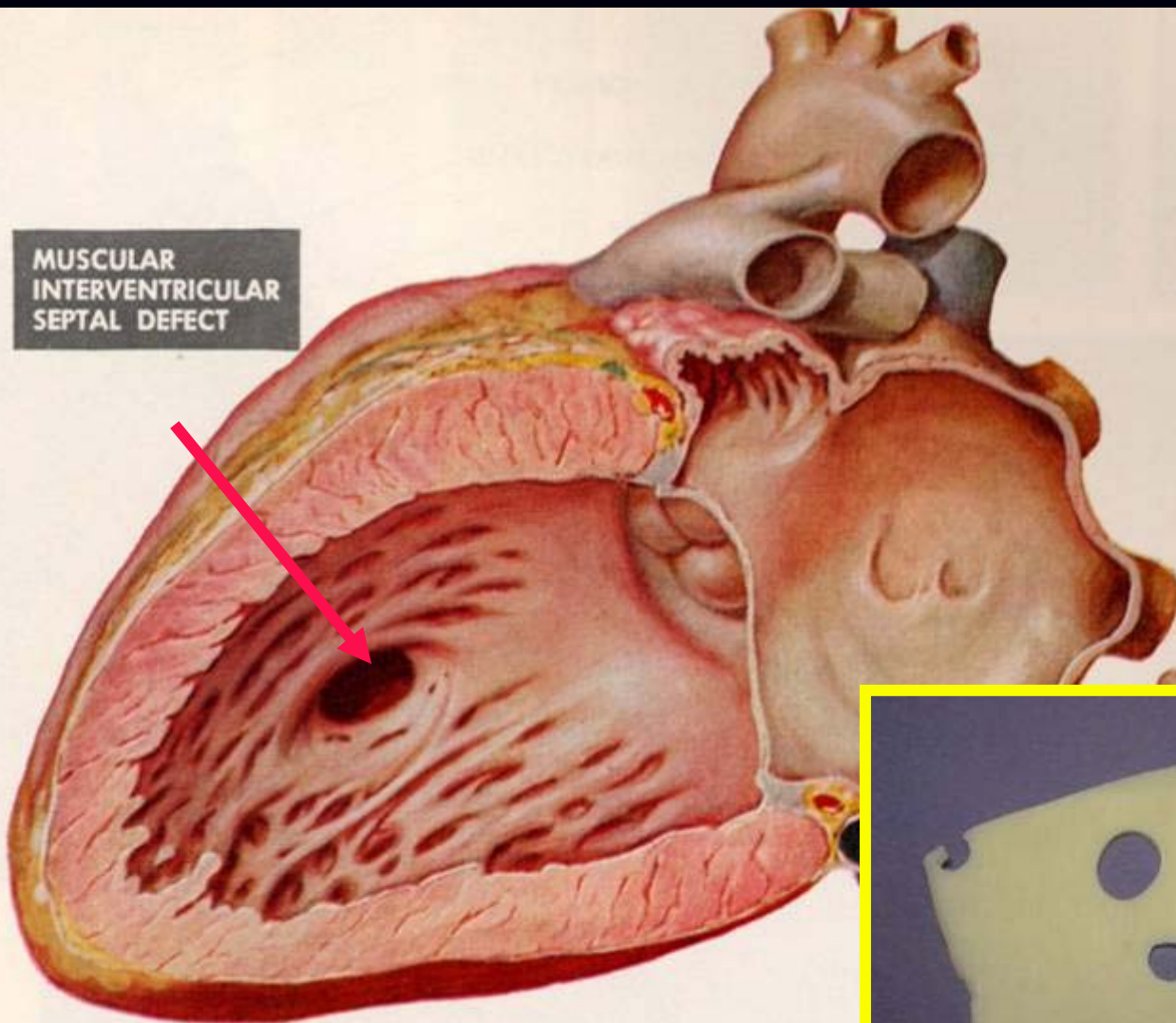


# Ventricular Septal Defect

## Muscular

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

MUSCULAR  
INTERVENTRICULAR  
SEPTAL DEFECT



Swiss  
cheese

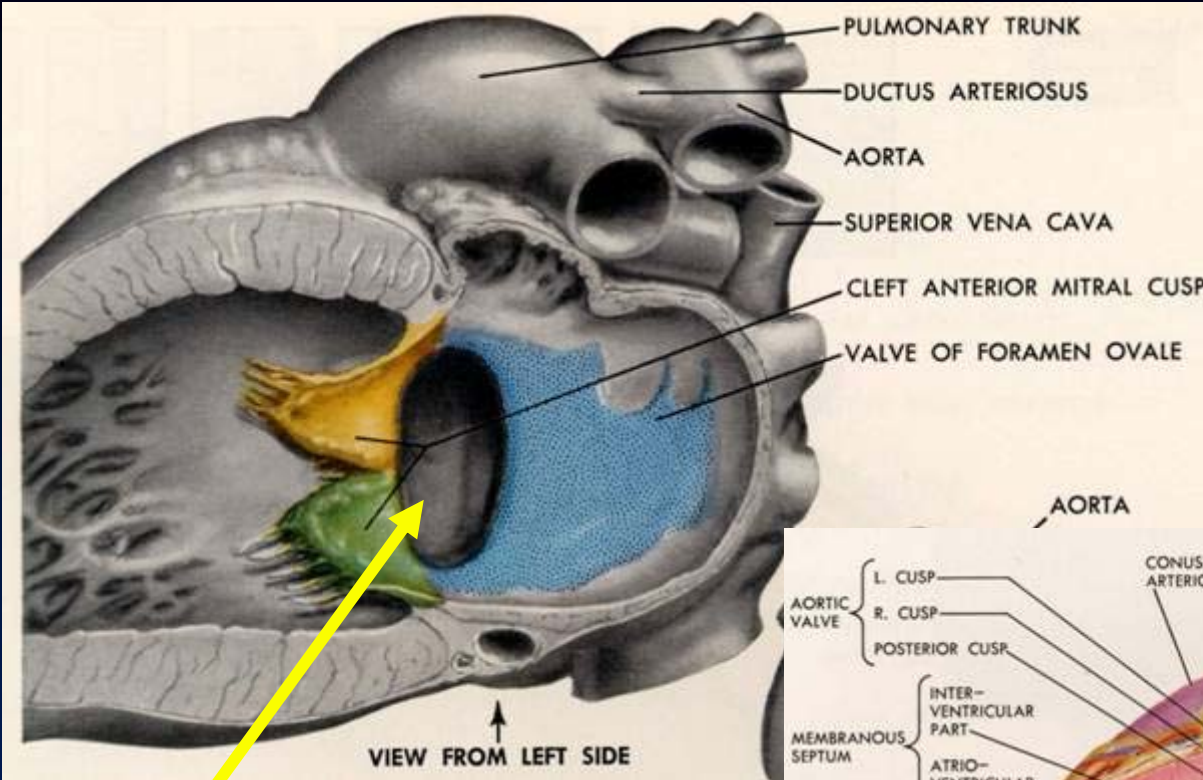


# **Ventricular Septal Defect**

## **AV Canal**

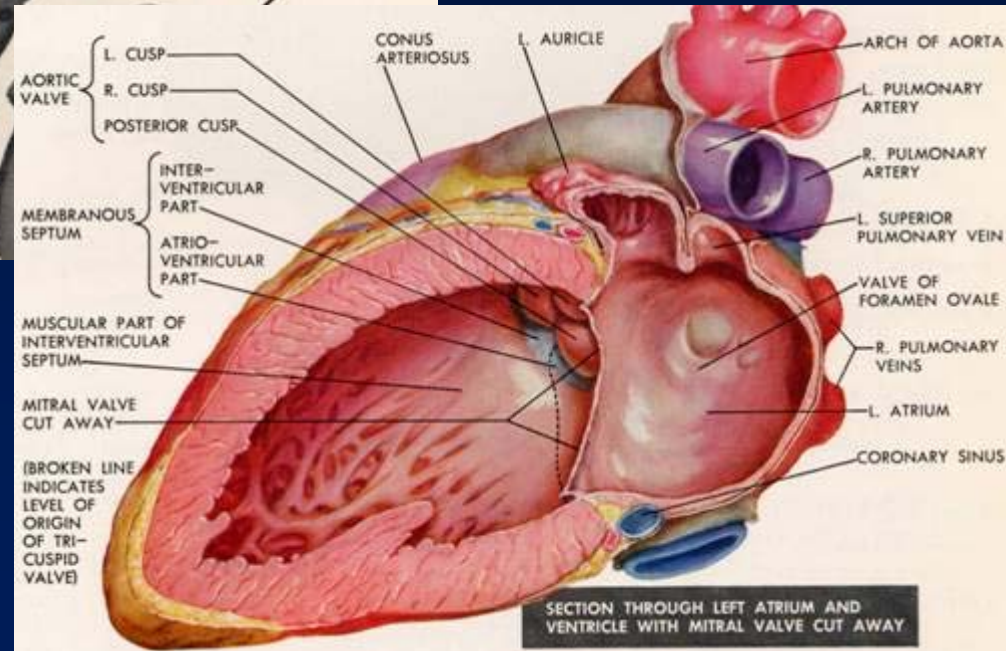
- **Atrioventricular canal = endocardial cushion type = posterior VSD (5–10%)**
- **Location: adjacent to septal and anterior leaflet of mitral valve**
- **Large VSD → pulmonary hypertension, eventually shunt reversal**
  - **Eisenmenger's physiology**
- **Very large VSD → CHF soon after birth**





© Frank Netter, MD Novartis®

**Large posterior VSD  
(AV canal)**



© Frank Netter, MD Novartis®



# **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**

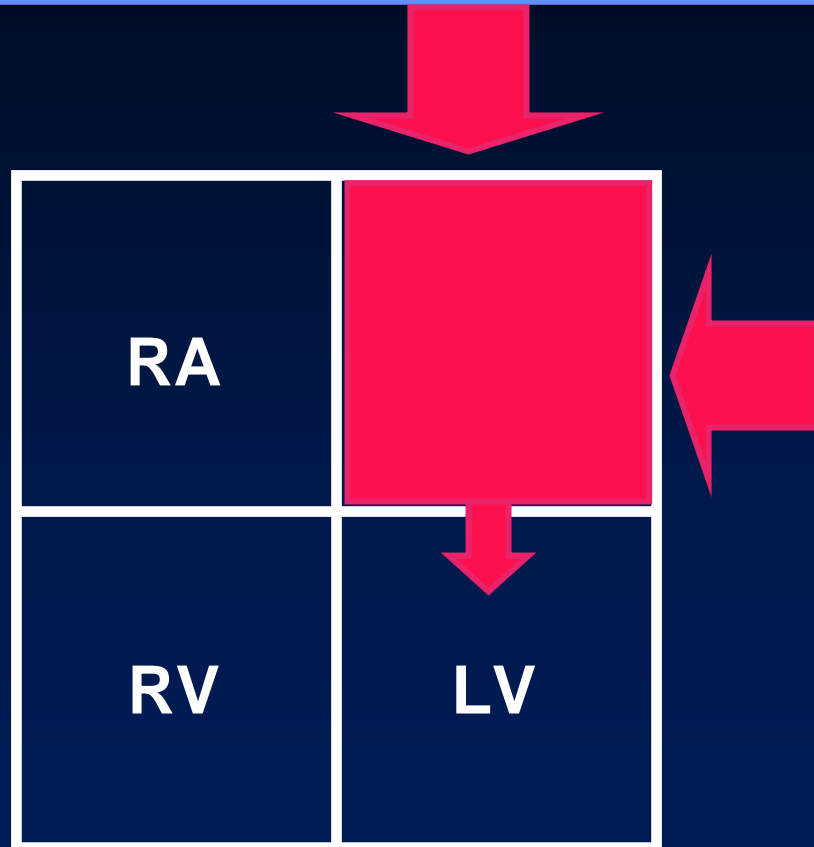
- **Prominent main pulmonary artery**
  - **Adult**
- **Shunt vasculature (increased flow to the lungs)**
- **LA enlargement (80%)**
- **Aorta normal in size**



**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
  - Premies
- Predominance in females 4:1

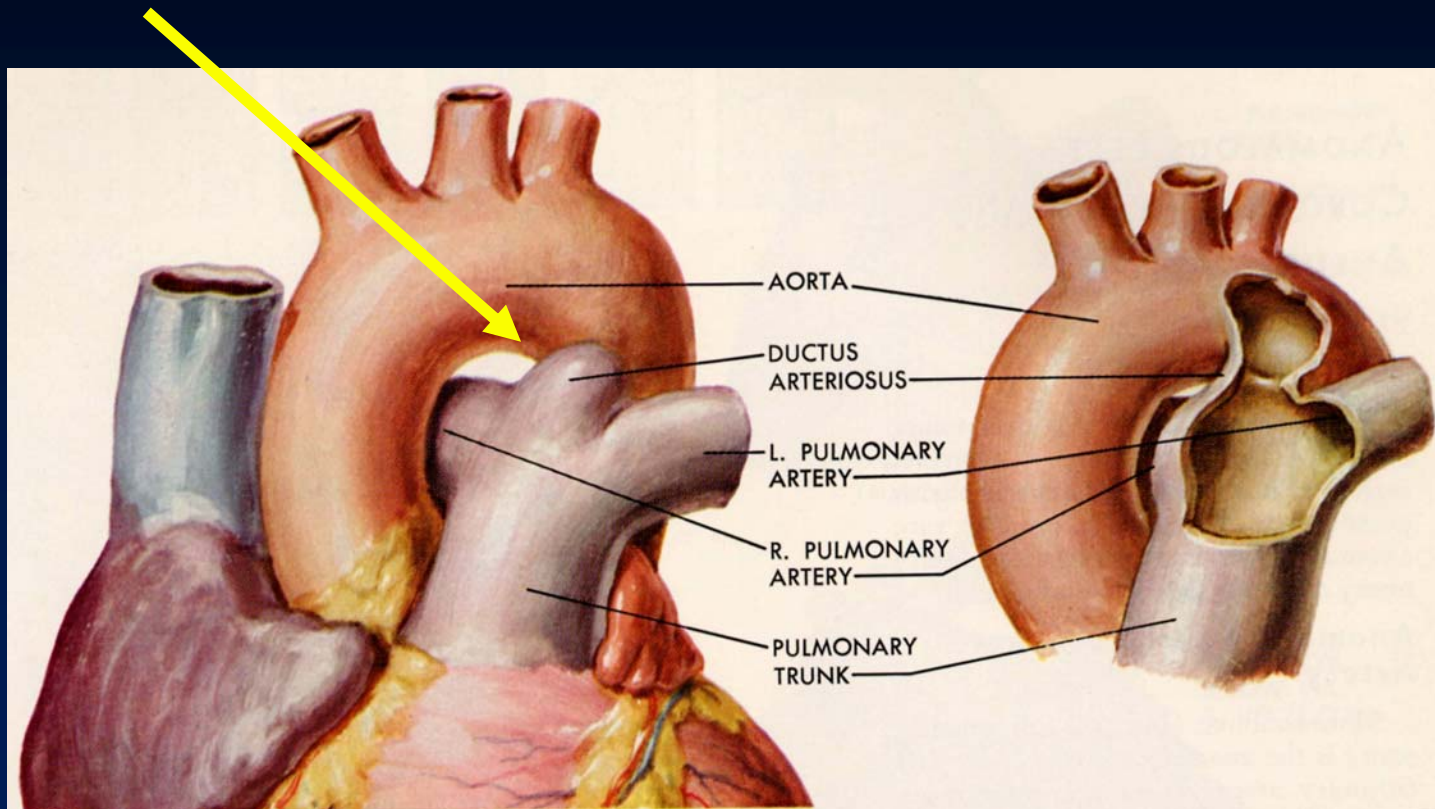
# **Patent Ductus Arteriosus**

## **Anatomy**

---

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

# Ductus Arteriosus

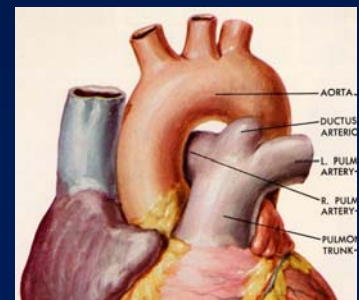


© Frank Netter, MD Novartis®

# Ductus Arteriosus

## Physiology

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





# Ductus Arteriosus

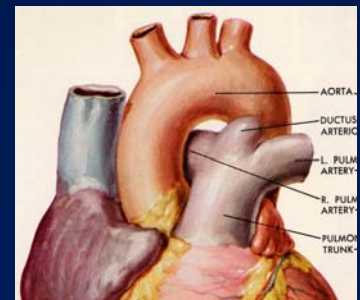
## Normal Closure

- **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

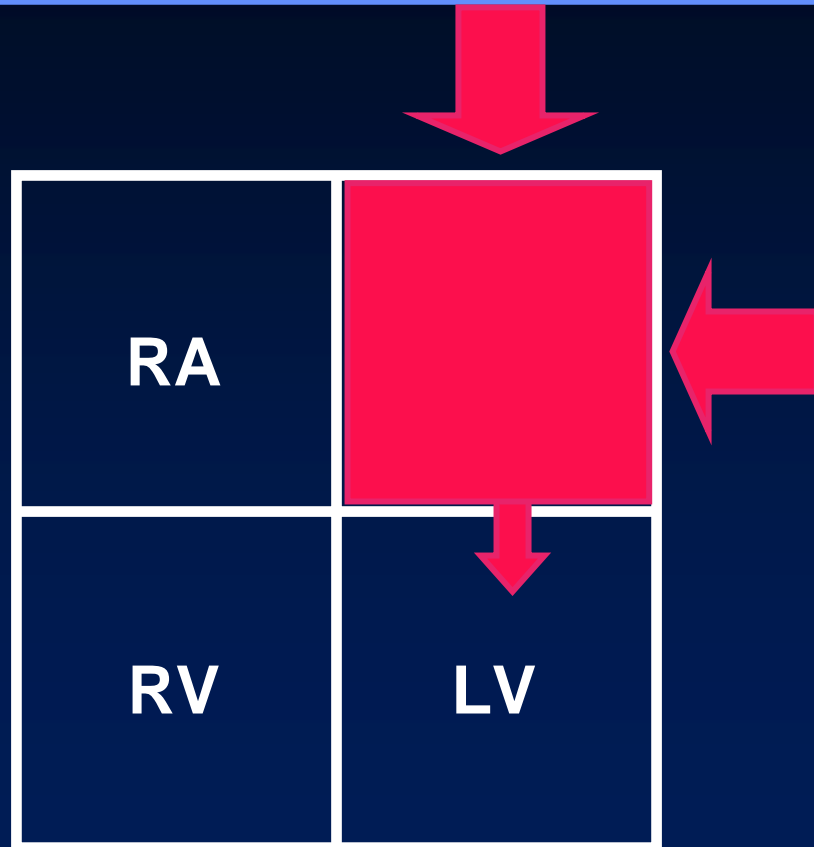
# **Patent Ductus Arteriosus**

## **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

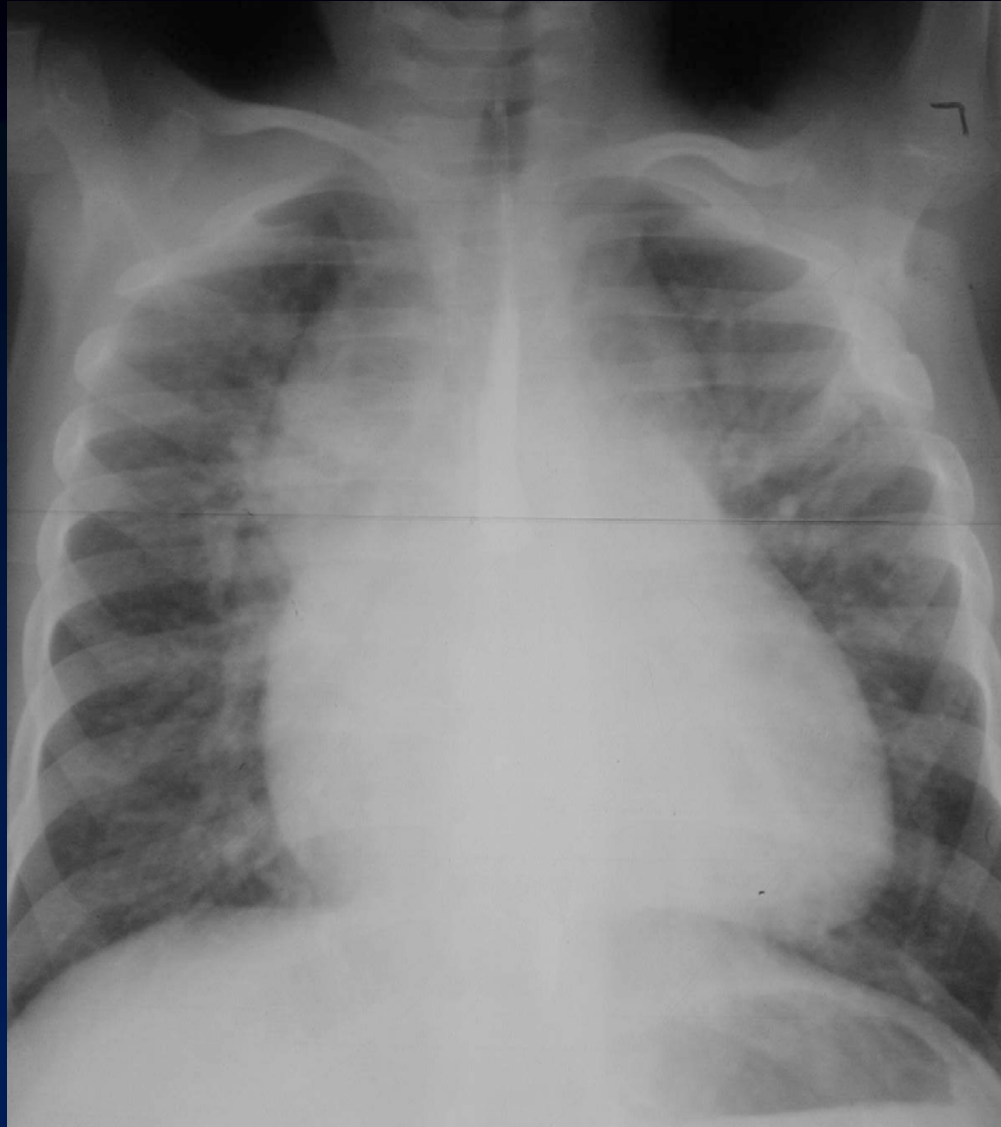
---

- 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**

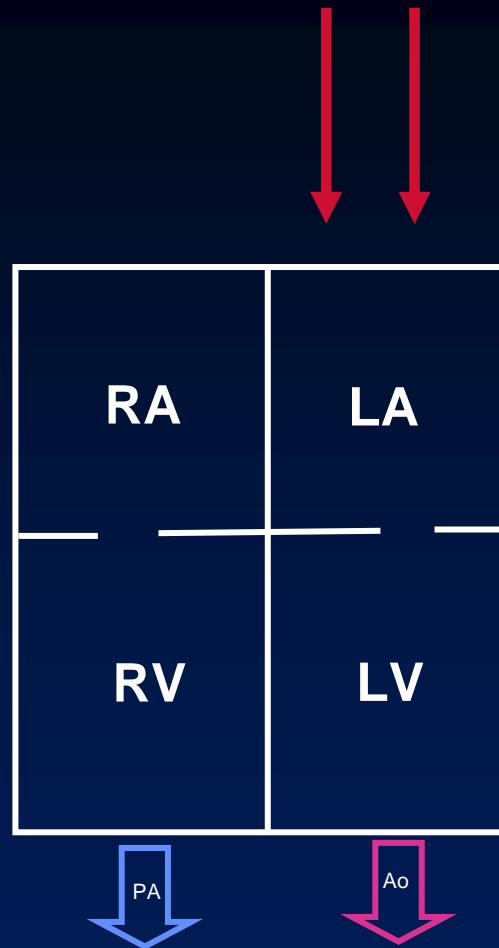
# Cyanosis With Increased Vascularity

- 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



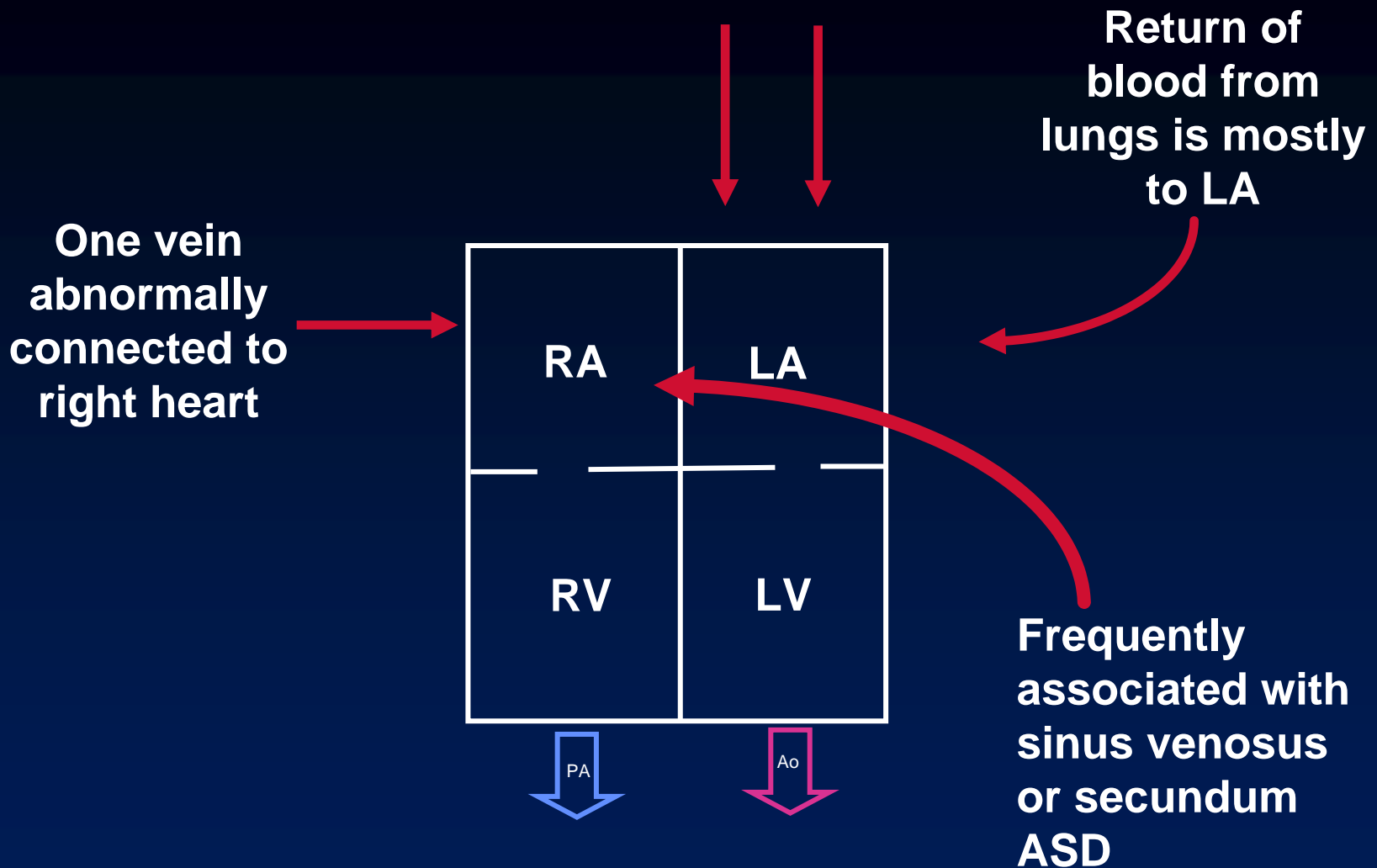
Return of  
blood from  
lungs is by  
four pulmonary  
veins to LA

**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**



## 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

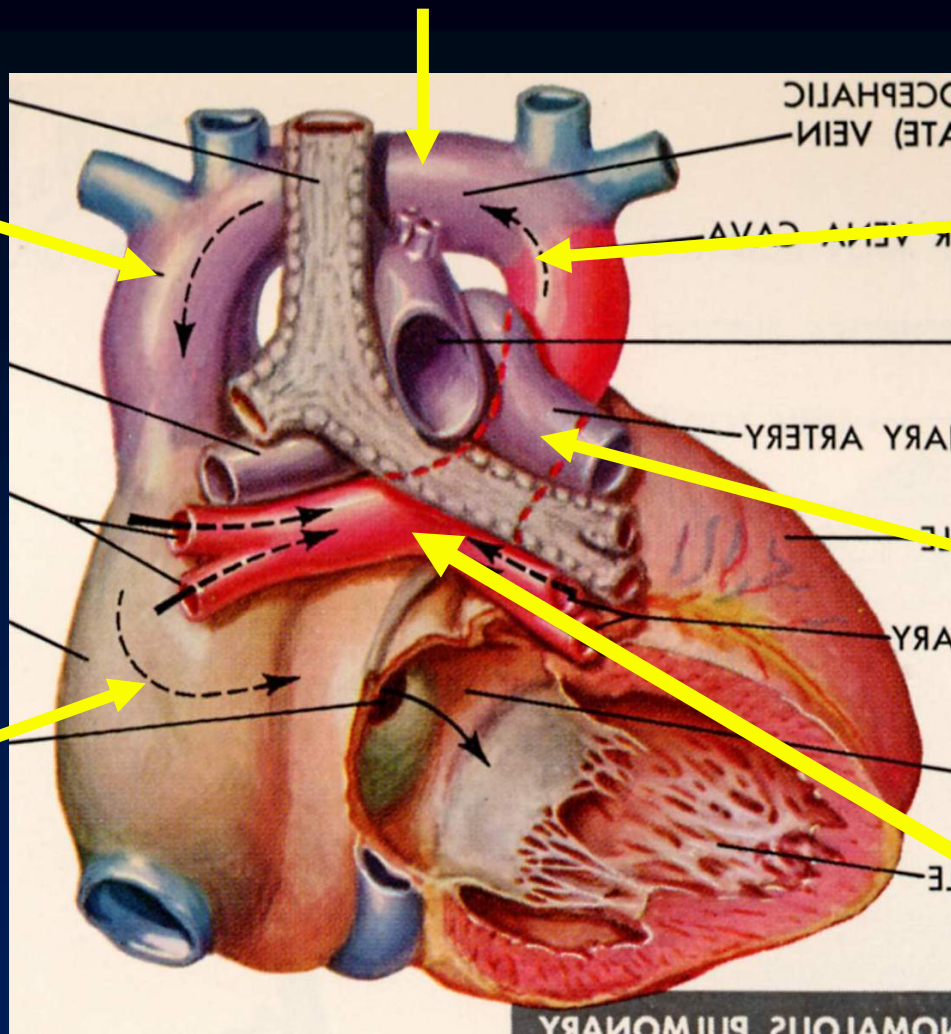
Right  
superior  
vena  
cava

Left  
superior  
vena cava

Vertical  
vein

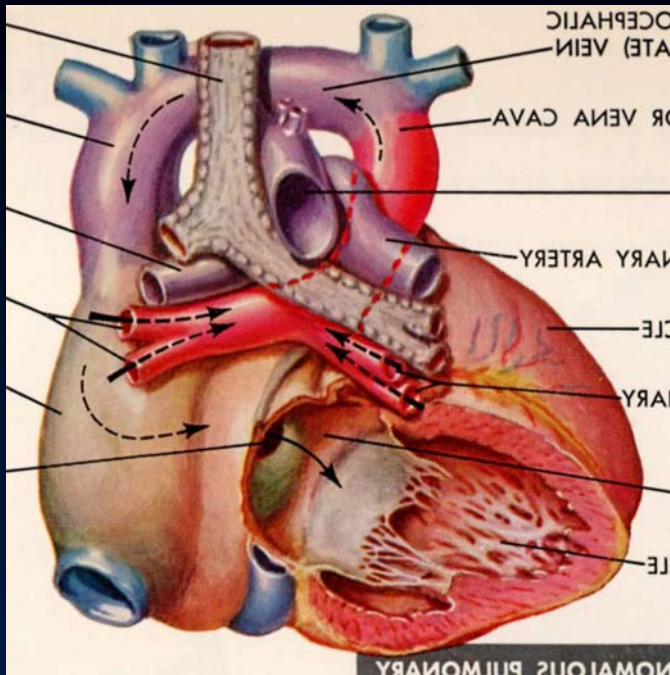
Right  
atrium

Pulmonary  
veins

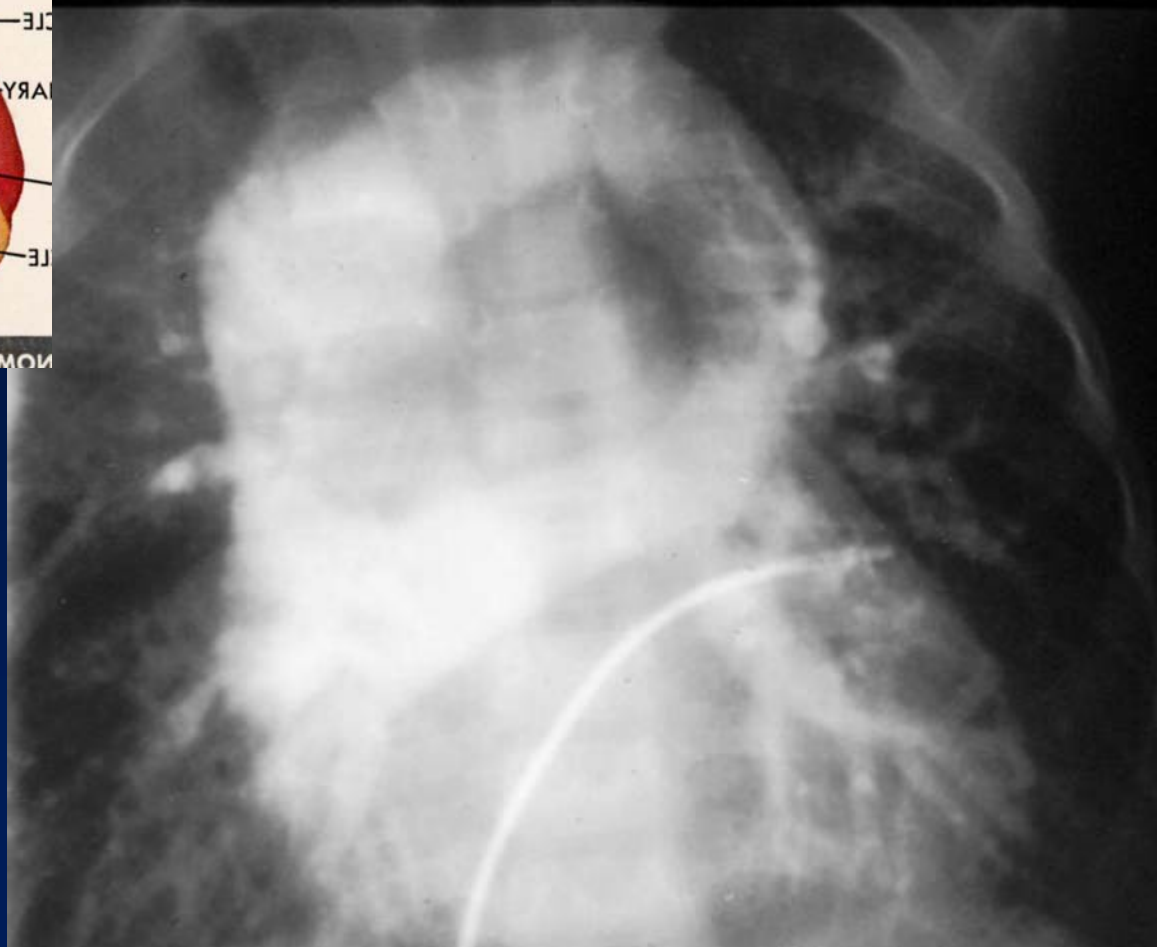


© Frank Netter, MD Novartis®

TAPVR-Supracardiac Type 1



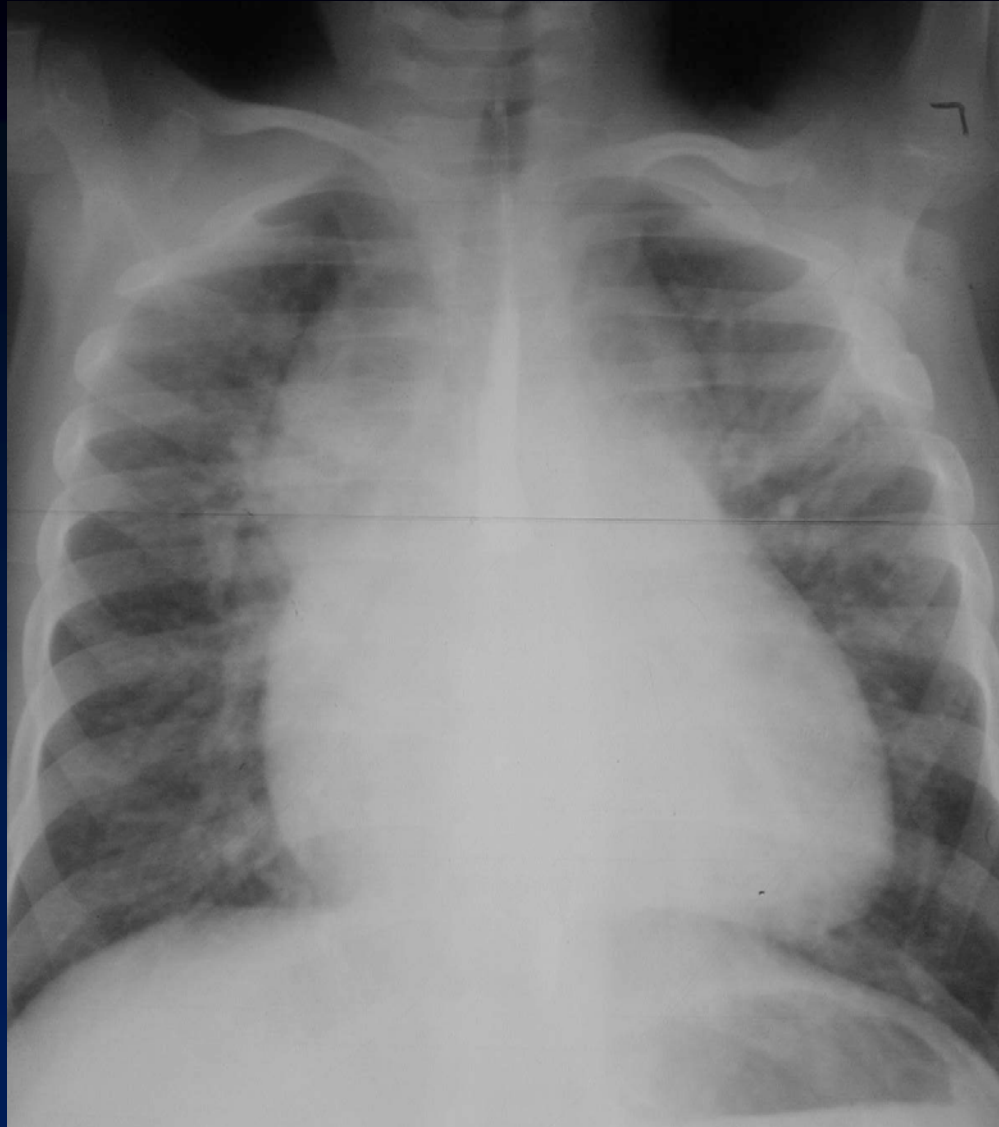
## TAPVR- Supracardiac Type 1



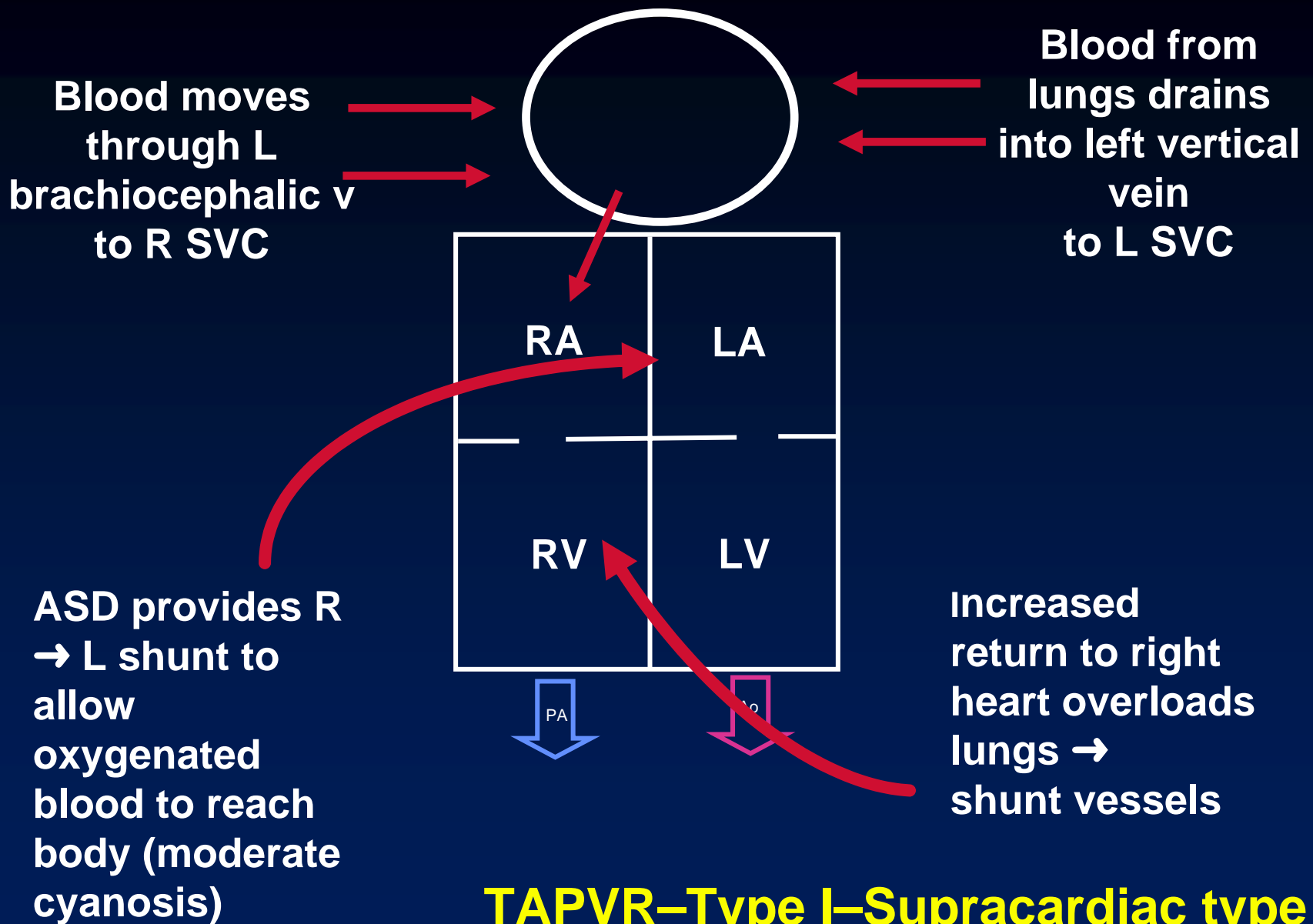
# **TAPVR**

## **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**

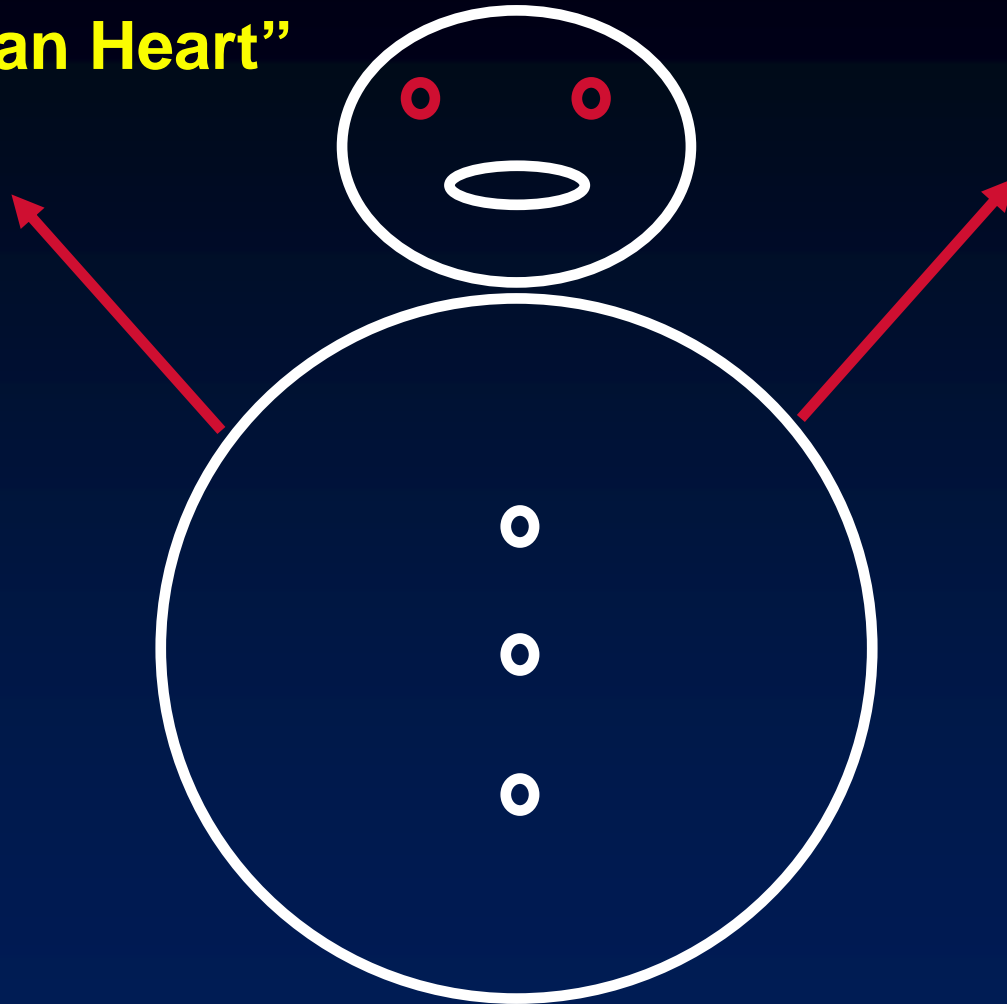


**TAPVR-Supracardiac Type 1**





**“Snowman Heart”**



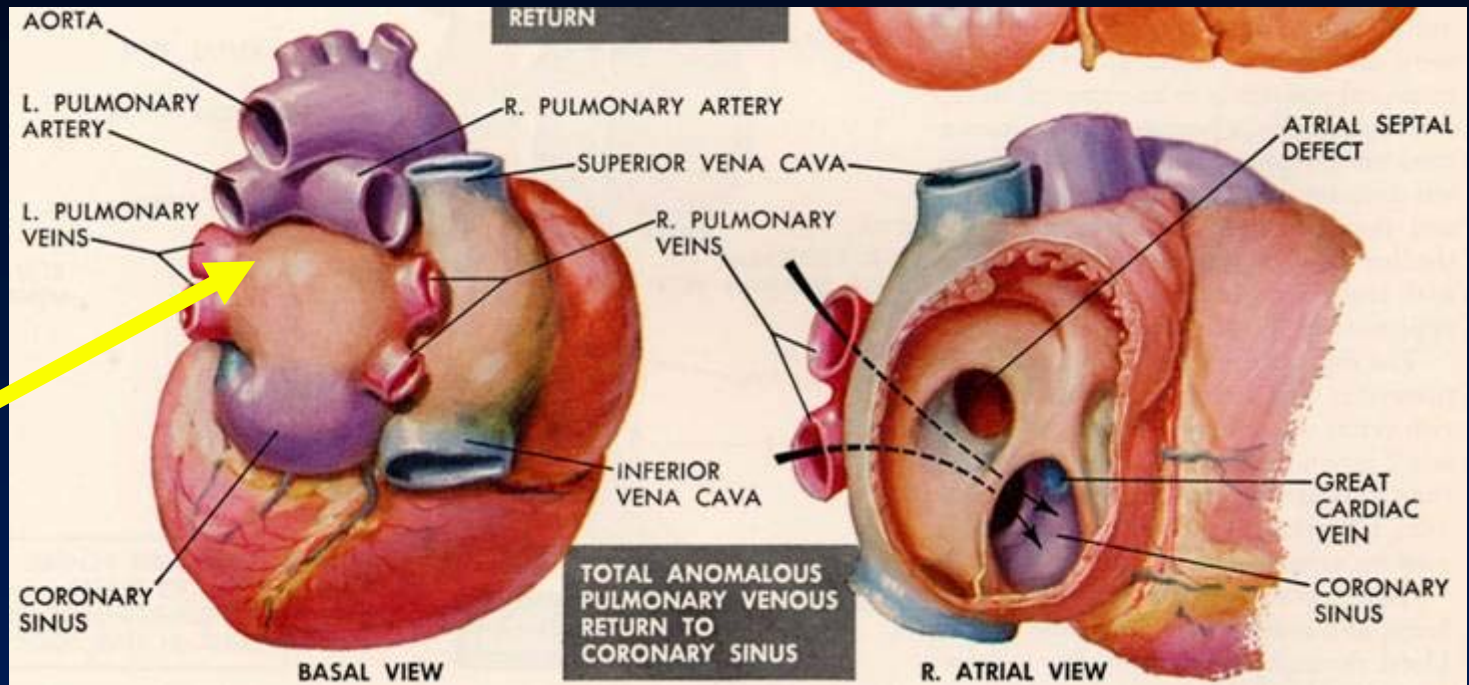
**TAPVR–Type I–Supracardiac type**

# **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**



© Frank Netter, MD Novartis®

## TAPVR-Coronary Sinus-Type II

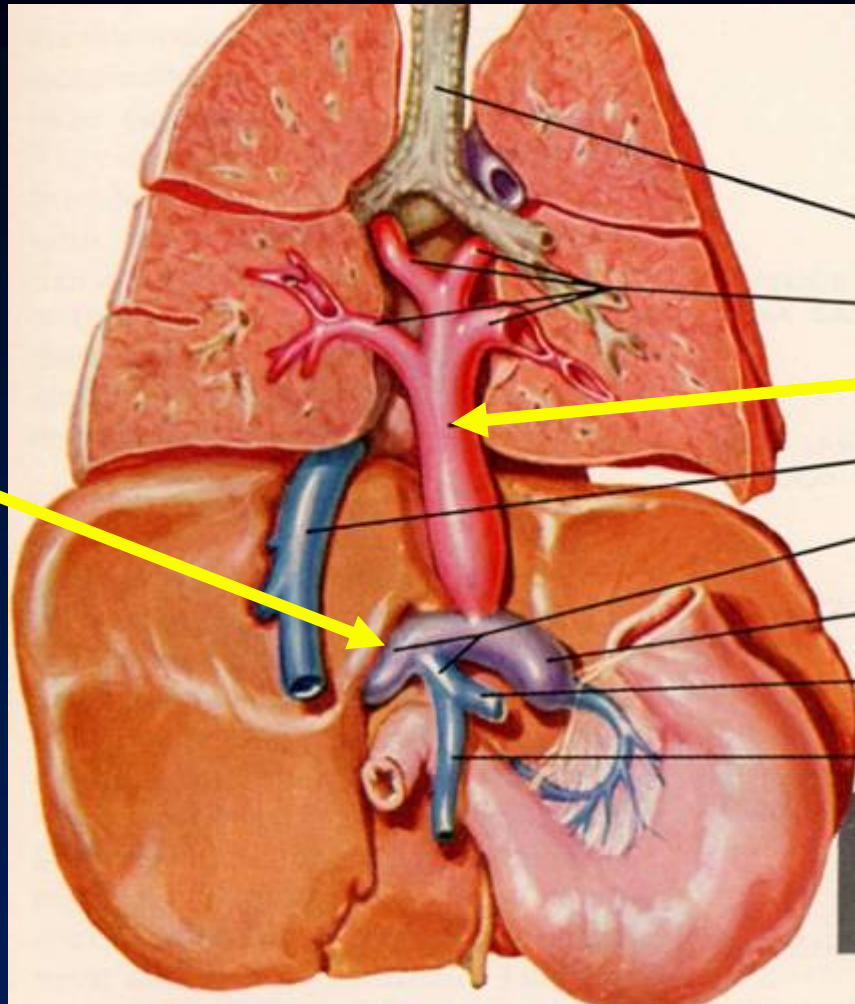


# **TAPVR**

## **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**

**Portal vein**



**Pulmonary  
veins**

© Frank Netter, MD Novartis®

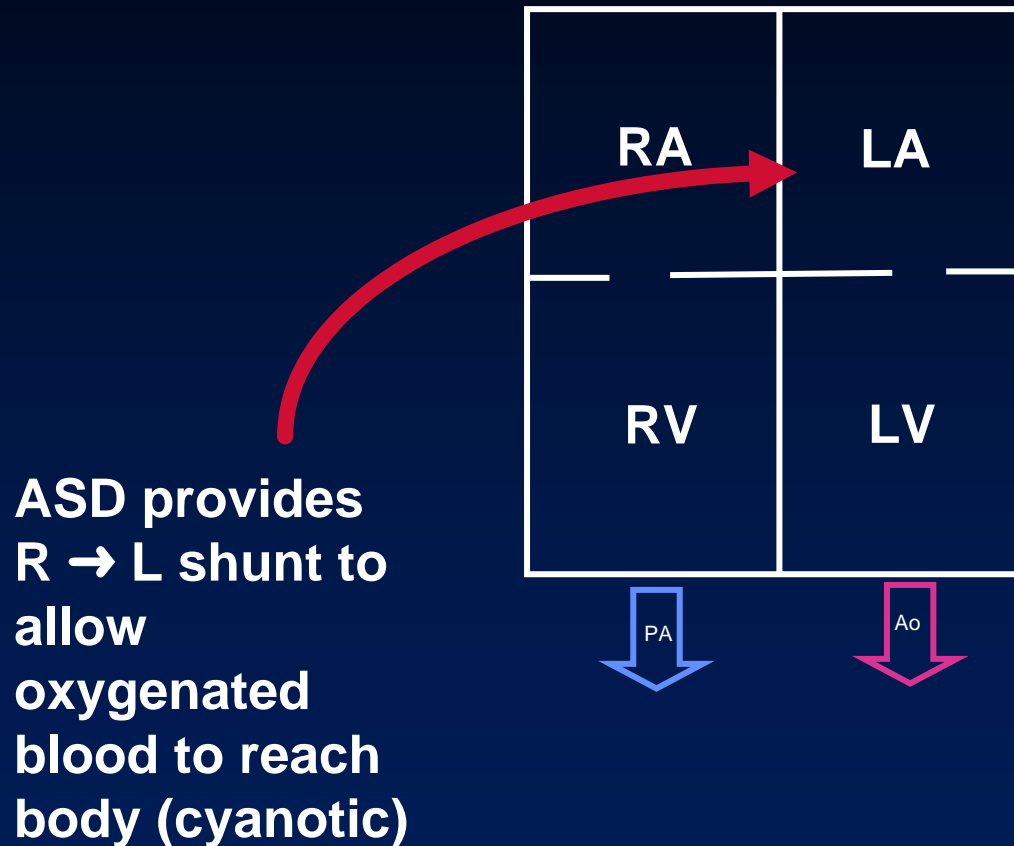
**TAPVR-Type III-  
Infradiaphragmatic**

# **TAPVR**

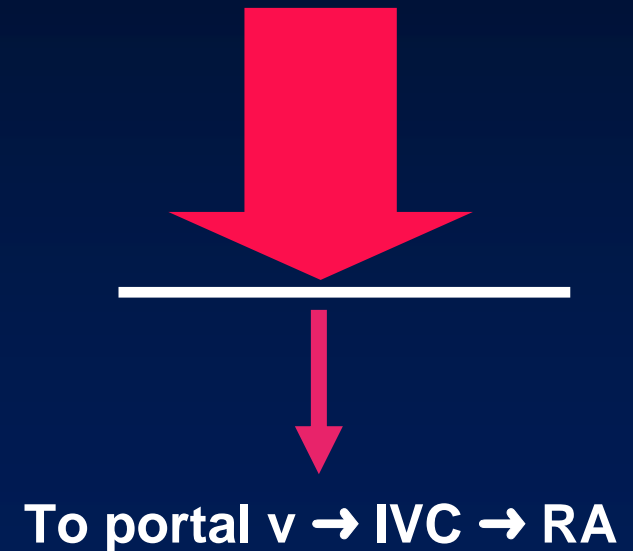
## **Infracardiac Type—Continued**

- **Severe CHF (90%) 2° obstruction to venous return**
- **Cyanotic 2° right Û 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



**TAPVR—Type III—Infracardiac type**



# **TAPVR**

## **Mixed Type—Type IV**

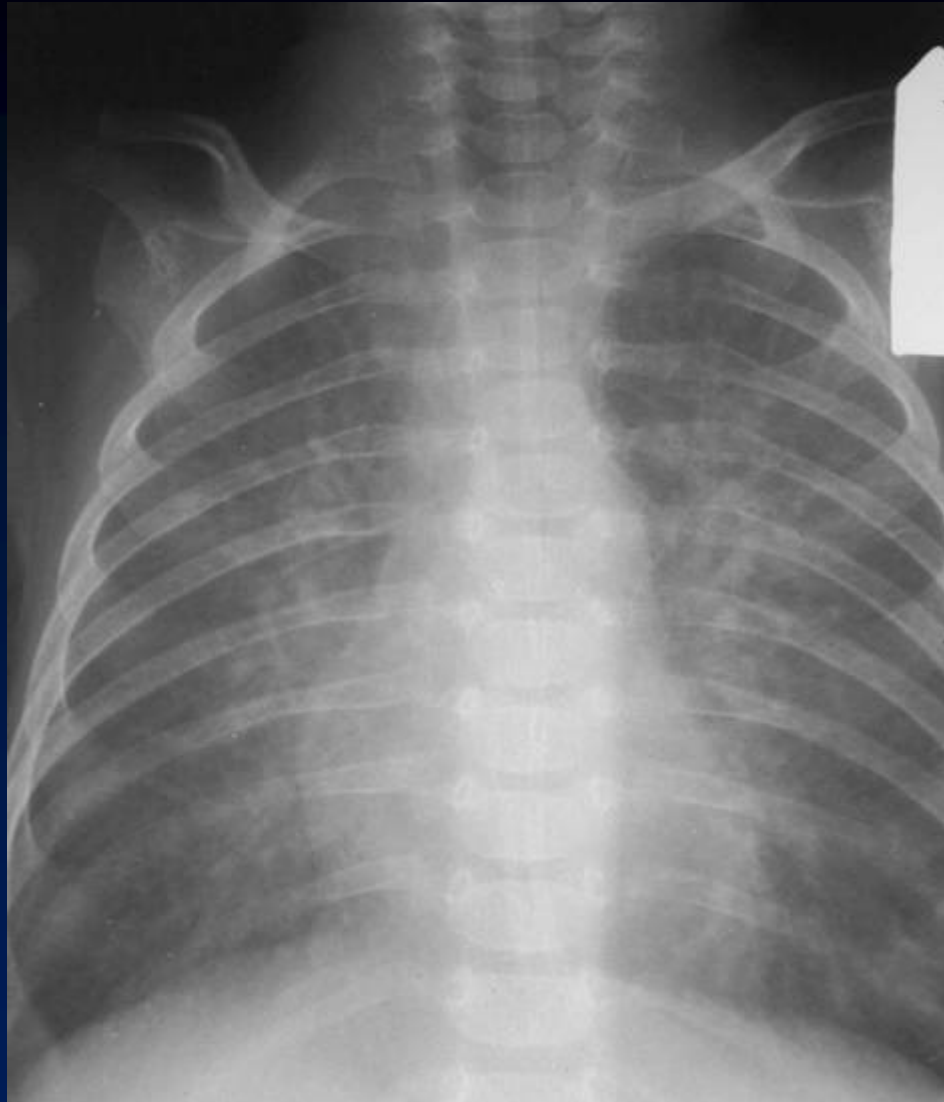
---

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

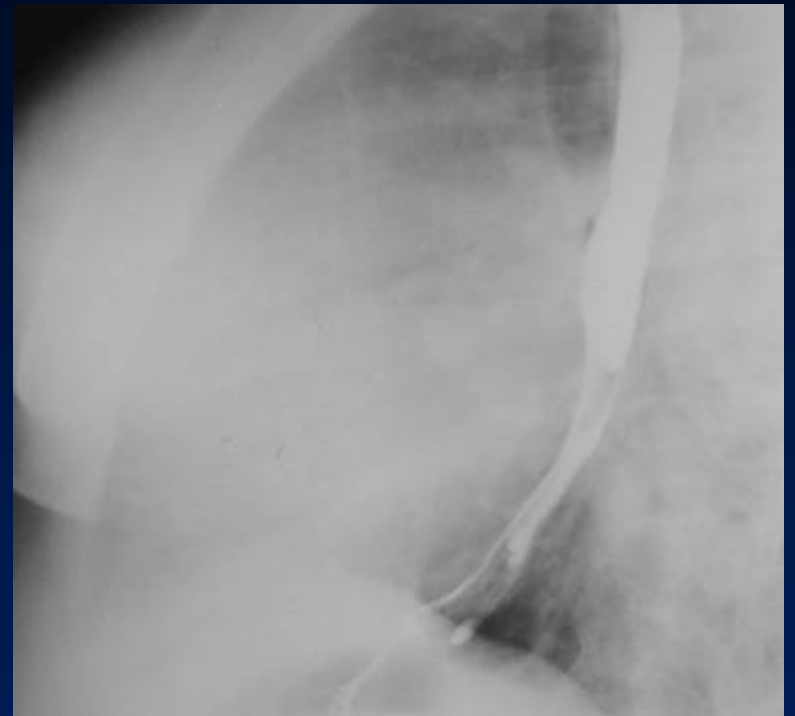
# Unknowns



**ASD (primum) with PAH**

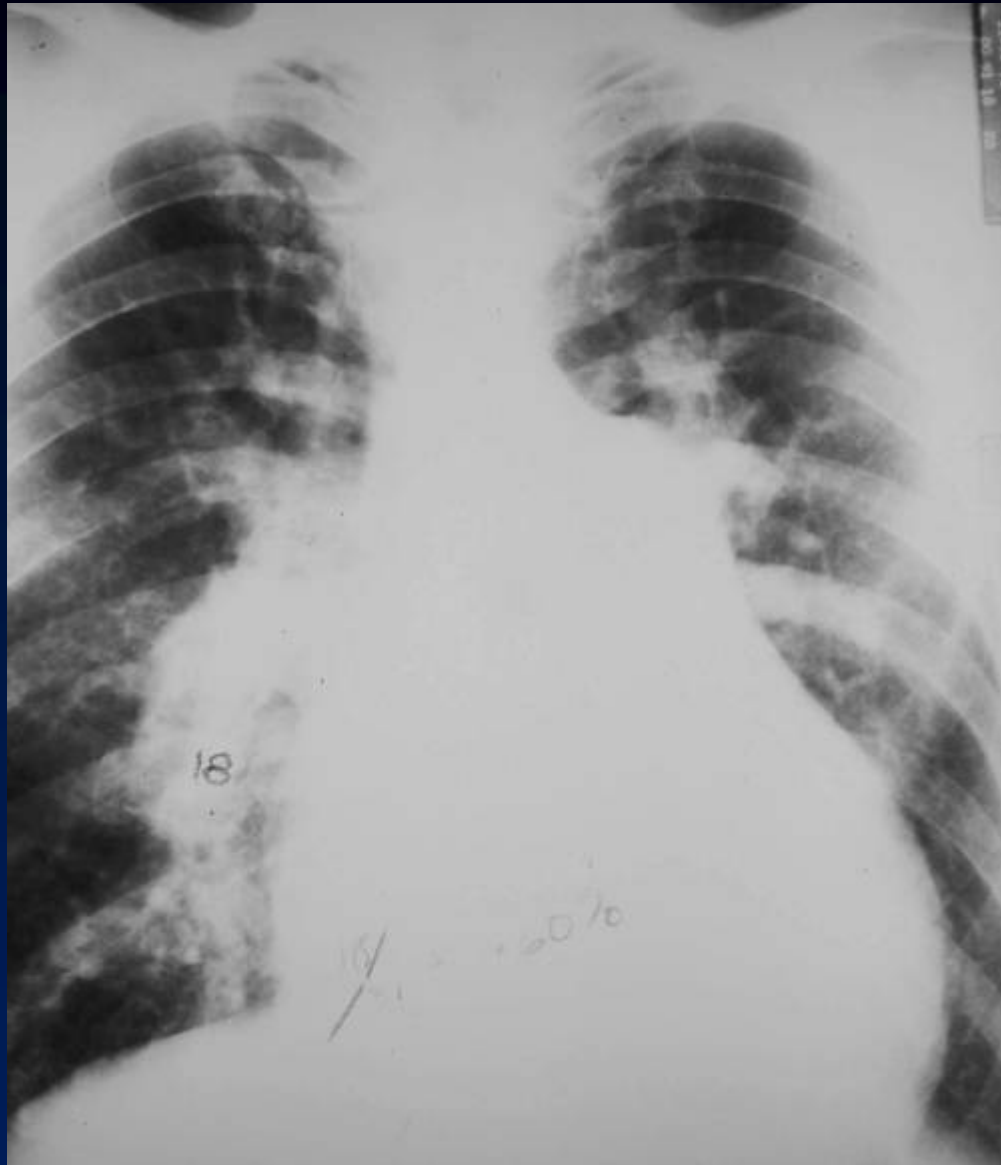


**TAPVR from below diaphragm**



**VSD**

ASD



**The End**