

# Obstructive Lesions

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or click left mouse button**

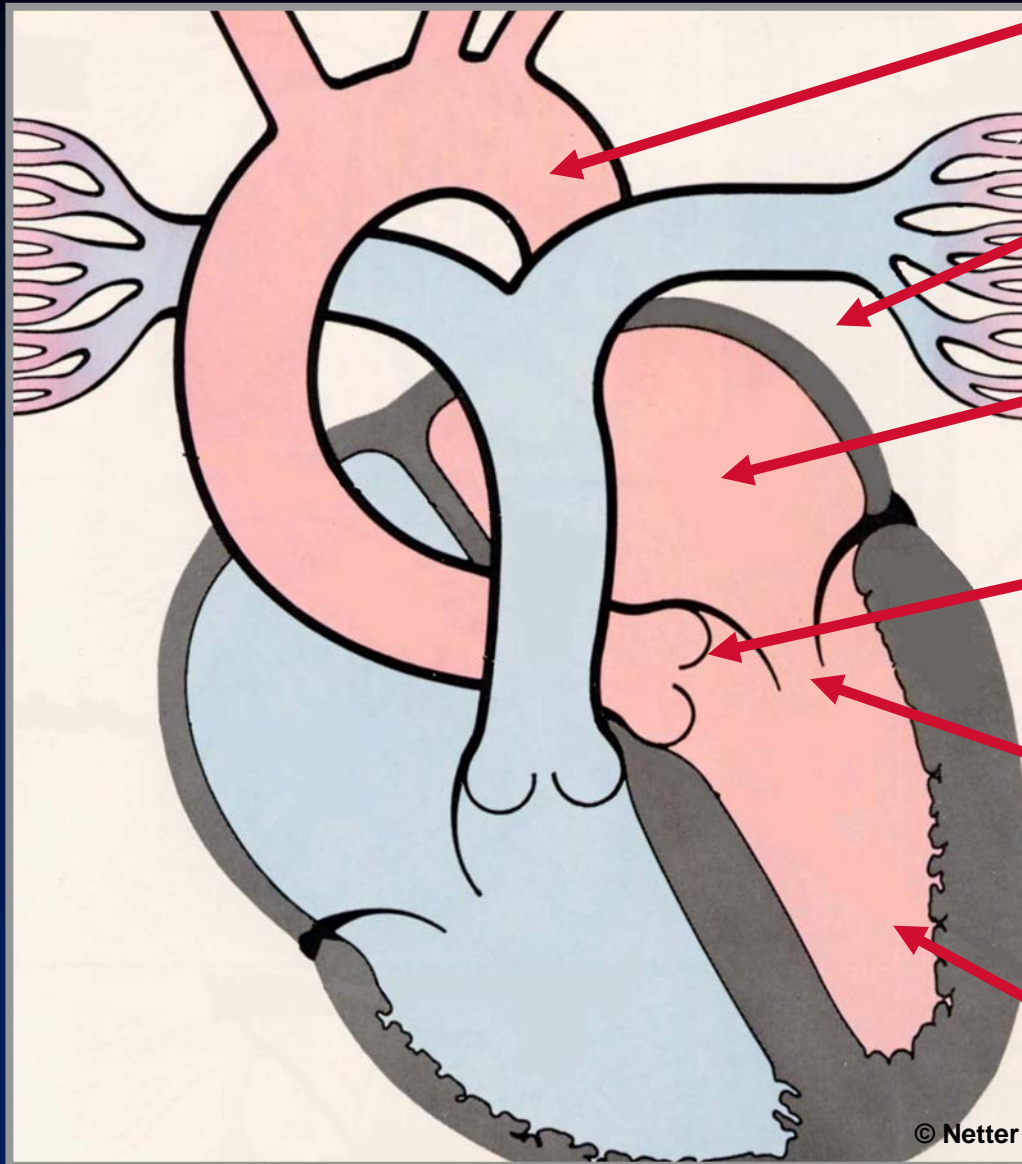
# **Lesions That Cause CHF**

# **CHF In Newborn**

## **Impede Return of Flow to Left Heart**

- **Infantile coarctation**
- **Congenital aortic stenosis**
- **Hypoplastic left heart syndrome**
- **Congenital mitral stenosis**
- **Cor triatriatum**
- **Obstruction to venous return from lungs**
  - **TAPVR from below diaphragm**

# Causes of CHF in the Newborn



**Coarctation of the Aorta**

**Obstruction to venous return from lungs**

**Cor Triatriatum**

**Congenital Aortic Stenosis**

**Congenital Mitral Stenosis**

**Hypoplastic Left Heart**

# Diagnosing CHF in a Newborn

- Usually have cardiomegaly
- Ill-defined bronchovascular bundles
- Flattening of diaphragm
  - Air hunger
- Rare
  - Kerley B lines
  - Pleural effusions



# **CHF In Chronologic Sequence**

# Commonest Cause of CHF

## In Chronologic Sequence

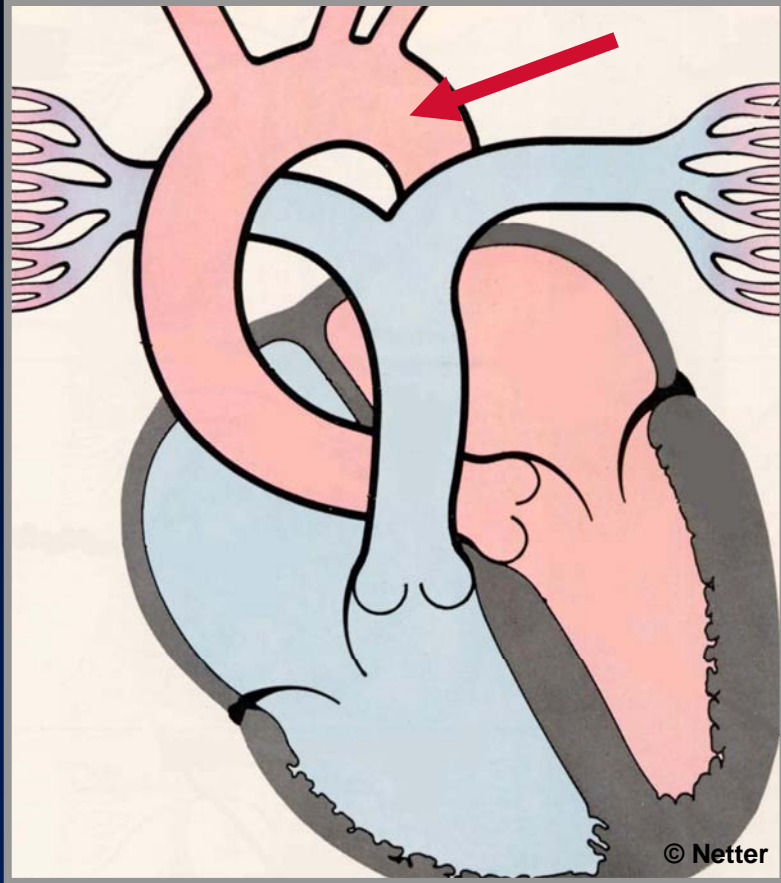
**< 24 hrs.....**Intrauterine arrhythmia

**First week.....** Hypoplastic Left Heart Syndrome

**2-6 weeks.....** Infantile coarctation

**1-4 months.....** Large L → R shunts

*VSD, ASD, PDA, AV Canal*



# Coarctation Of the Aorta



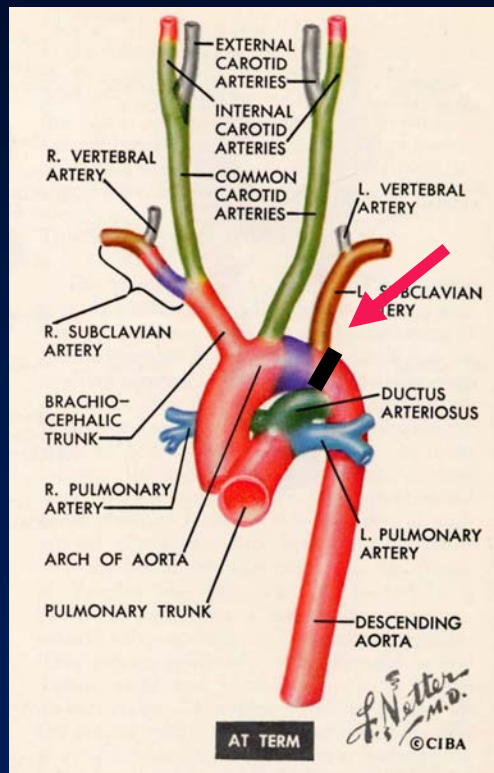
# **Coarctation of the Aorta**

## **General**

- **2X more common in males**
- **Common classification**
  - **Infantile or preductal form**
  - **Adult or juxtaductal form**
- **Relationship of ductus to coarct affects clinical picture**

# Coarctation of the Aorta

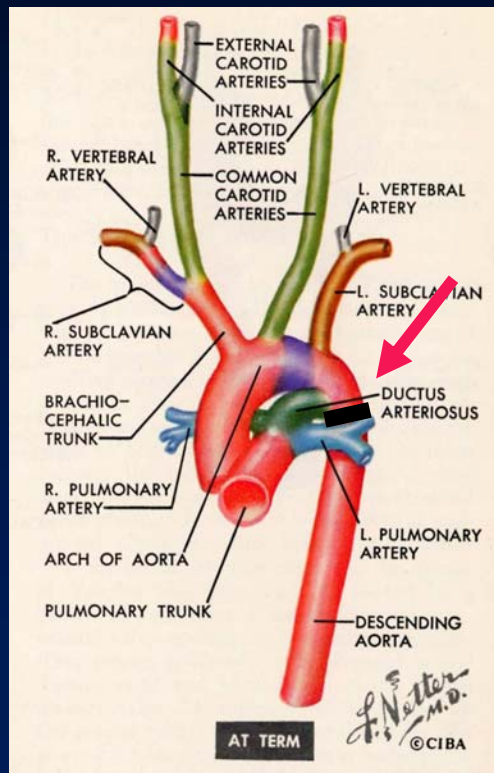
## Coarctation Proximal to Ductus



- Flow is frequently from PA to Ao through Ductus
- Cyanosis in lower half of body as
  - Unoxygenated blood from PA feeds lower extremities
- Oxygenated blood from LV goes to major vessels of head and neck
  - Not cyanotic

# Coarctation of the Aorta

## Coarctation Distal to Ductus



- Flow is initially from Ao to PA (L → R shunt)
- If there is Eisenmenger's physiology, the flow reverses and goes from PA → Ao (R → L shunt)
- Cyanosis
- More common form

# **Coarctation of the Aorta**

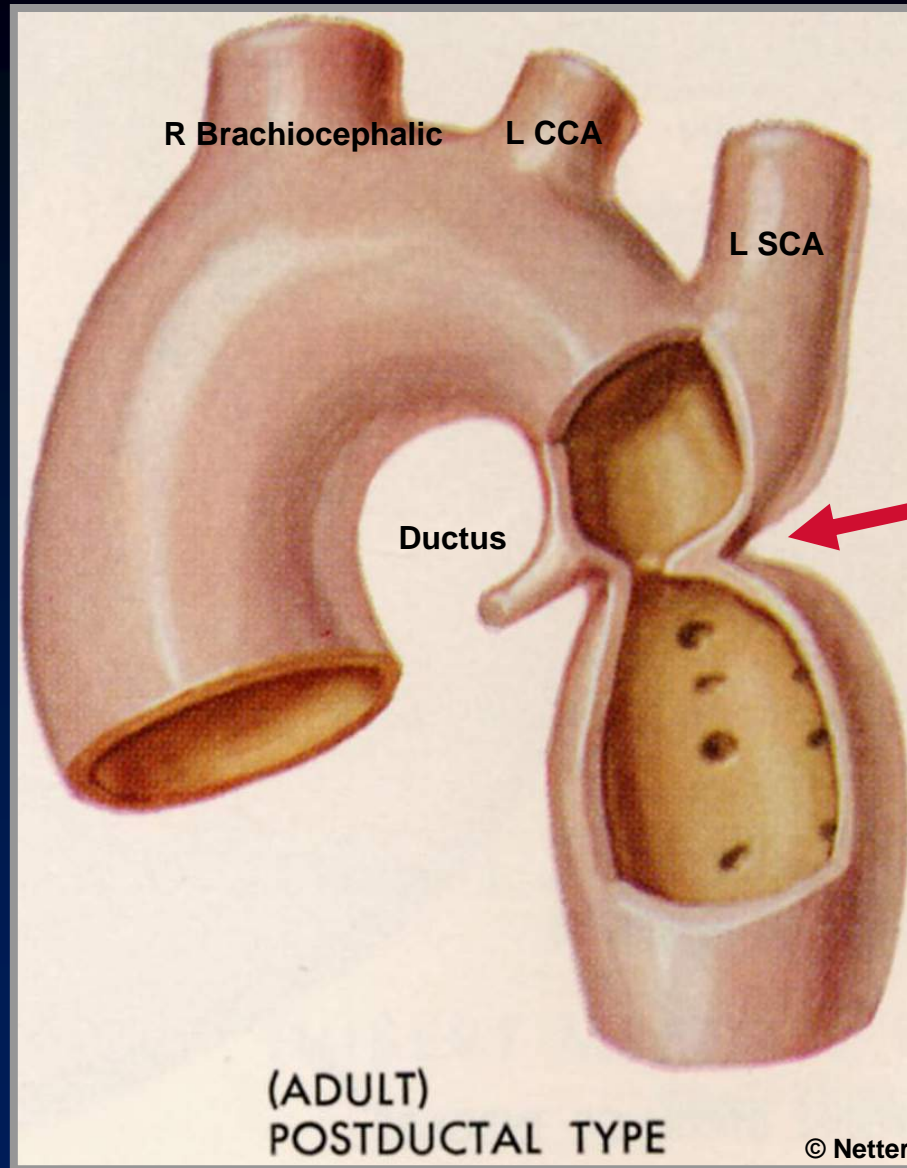
## **Other Classifications**

- **More complicated classifications take following into account**
  - **Location and length of coarct**
  - **Patency of ductus arteriosus**
  - **Relationship of coarct to ductus**

# **Coarctation of the Aorta**

## **Adult Form**

- **Adult or juxtaductal (postductal) form is more common than infantile**
- **Usually localized**
- **Area of coarctation just beyond origin of LSCA at level of ductus**





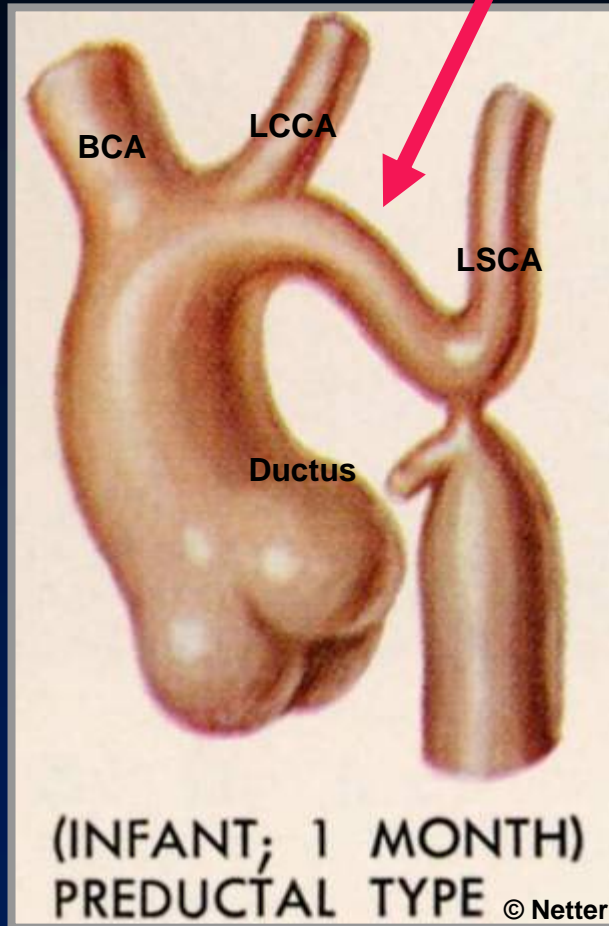
**Coarctation of the Aorta**

# **Coarctation of the Aorta**

## **Infantile Form**

- **Infantile, preductal form = diffuse type**
- **Long, tubular segment of narrowed aorta**
  - **From just distal to brachiocephalic artery to level of ductus**
- **Intracardiac defects (VSD, ASD, deformed mitral valve) present in 50% of diffuse type**
  - **Also patent ductus arteriosus**





# **Coarctation of the Aorta**

## **Associated Defects**

- **Bicuspid aortic valve (most common associated defect seen in 50%)**
- **VSD**
- **ASD**
- **Transposition**
- **25% of patients with Turner's Syndrome have coarctation of aorta**

# **Coarctation of the Aorta**

## **Shone Syndrome**

- **Coarctation of aorta**
- **Aortic stenosis**
- **Parachute mitral valve**
- **Supravalvular mitral ring**

# **X-Ray Findings**

## **Rib Notching**

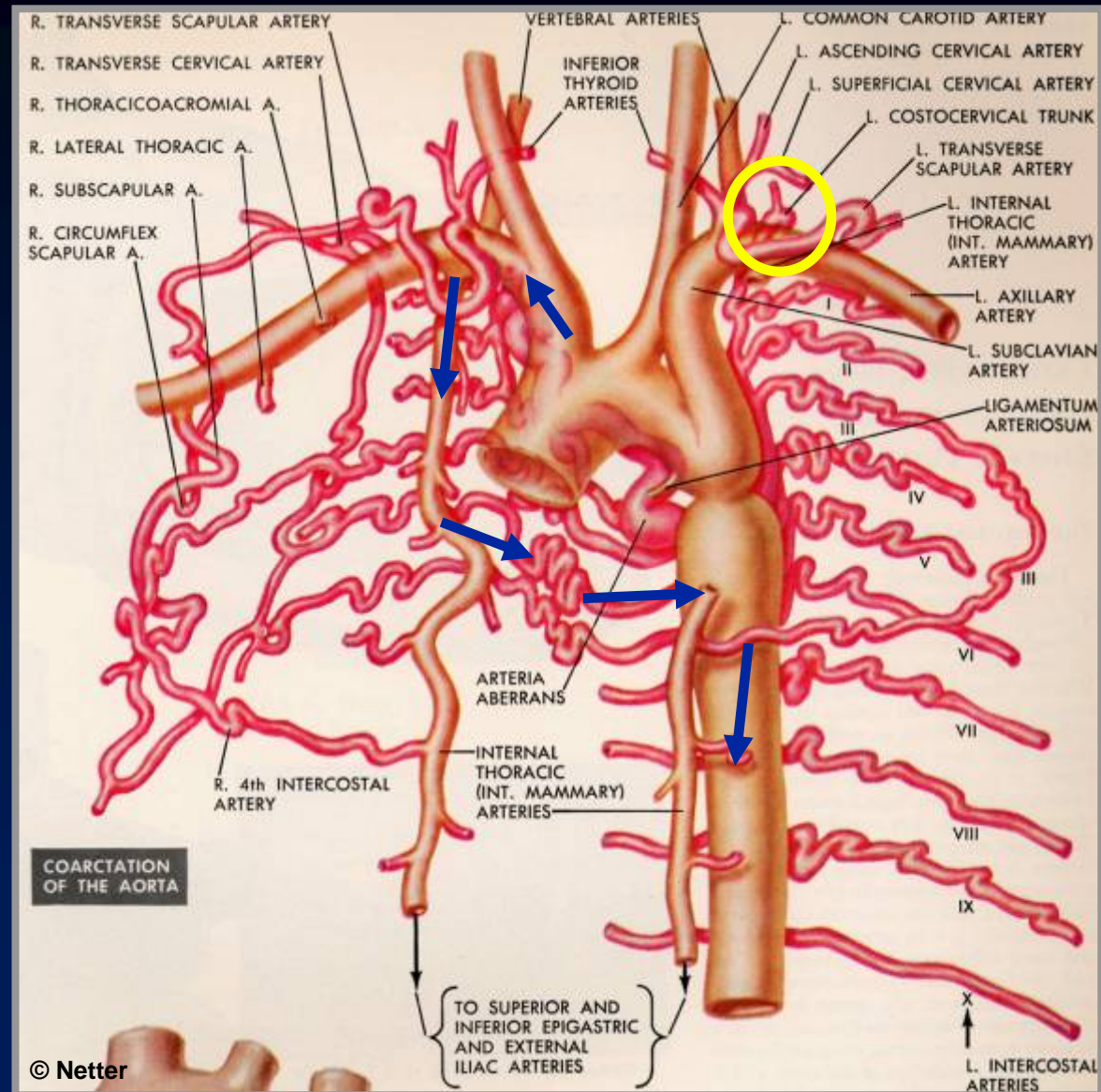
- **Single best sign**
- **Older the person, more likely to have rib notching (uncommon <6 yrs)**
- **Majority with coarcts display it >20 years of age**
- **Rib notching occurs in high pressure circuit**

# Coarctation of the Aorta

To supply aorta  
distal to ductus,  
flow in the 3<sup>rd</sup>-8<sup>th</sup>  
intercostals  
reverses

Blood flows from  
subclavian →  
internal mammary  
→ intercostals →  
aorta

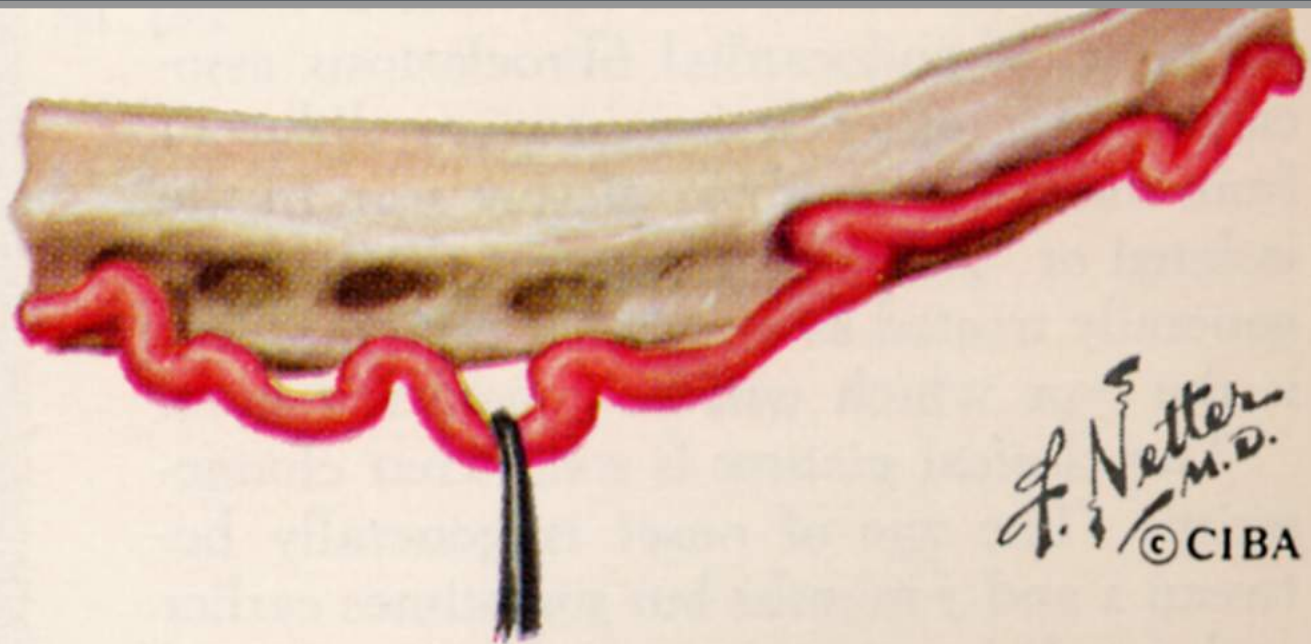
First two  
intercostals arise  
from costocervical  
trunk and do not  
serve aorta



# **X-Ray Findings**

## **Rib Notching**

- **Most often involves 4th-8th rib**
  - Sometimes may involve 3rd and 9th
- **Does not involve 1st and 2nd ribs**
  - Intercostals come off costocervical trunk and do not supply collateral flow to descending aorta
    - ▲ 4th-8th do anastomose with internal mammary to form collaterals for descending aorta

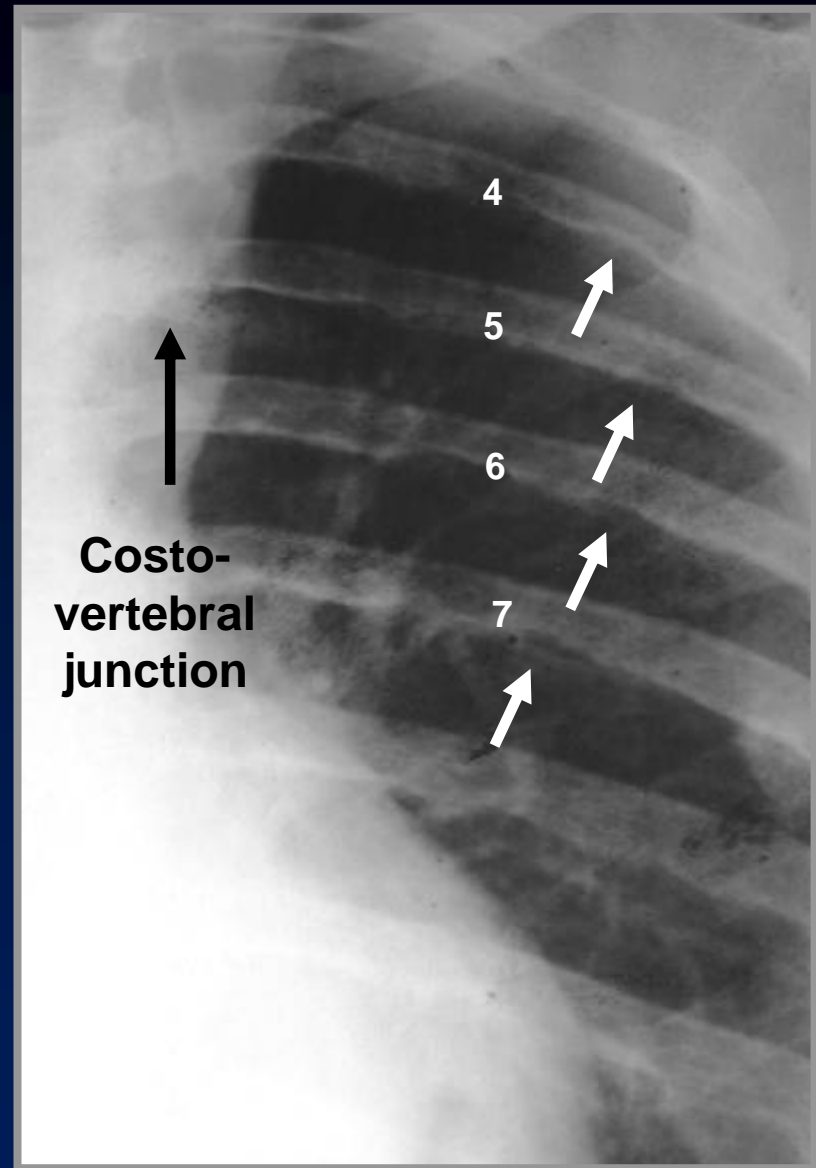
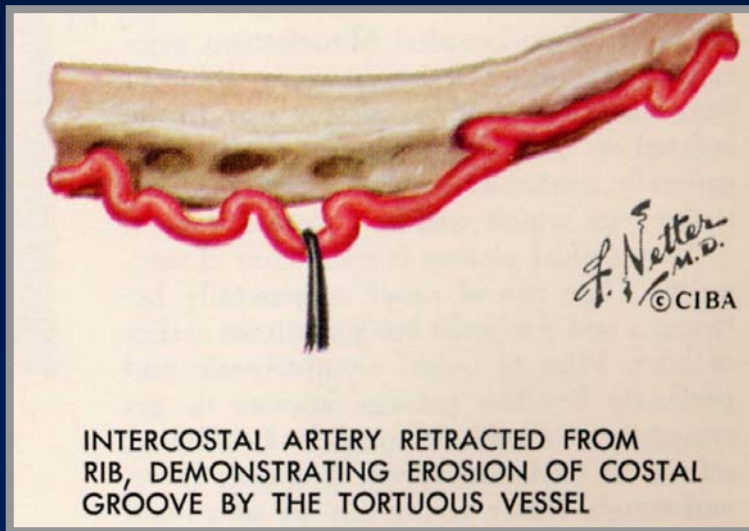


INTERCOSTAL ARTERY RETRACTED FROM  
RIB, DEMONSTRATING EROSION OF COSTAL  
GROOVE BY THE TORTUOUS VESSEL



# Rib Notching in Coarctation

Regresses after coarct is repaired





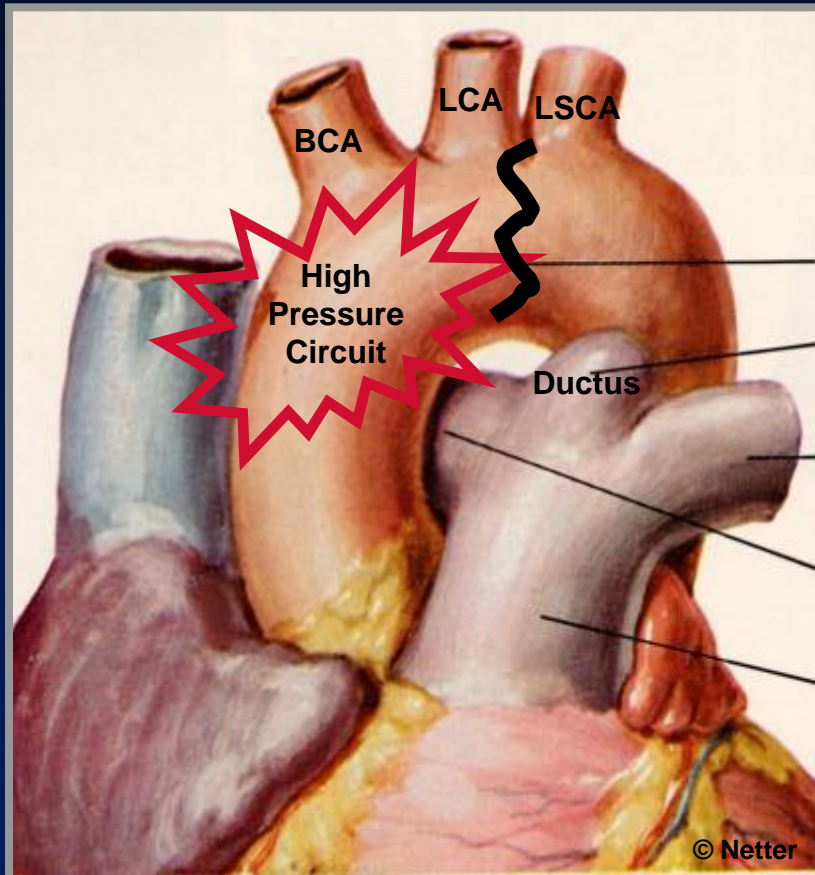
# **X-Ray Findings**

## **Rib Notching–Unilateral**

**Rib notching occurs in the high  
pressure circuit**

# X-Ray Findings

## Unilateral Right Rib Notching



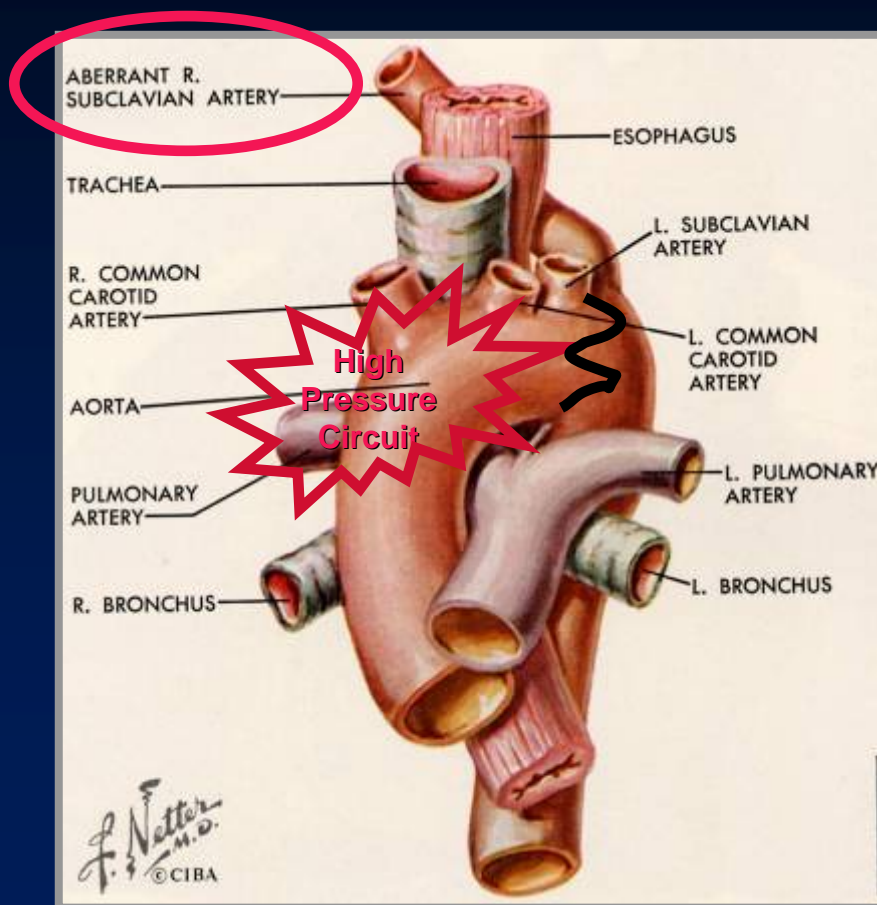
**Notching occurs in the high pressure circuit**

**Isolated right-sided rib notching**

**Coarct originates between the LCCA and the LSCA**

# X-Ray Findings

## Unilateral Left Rib Notching



**Notching occurs in the high pressure circuit**

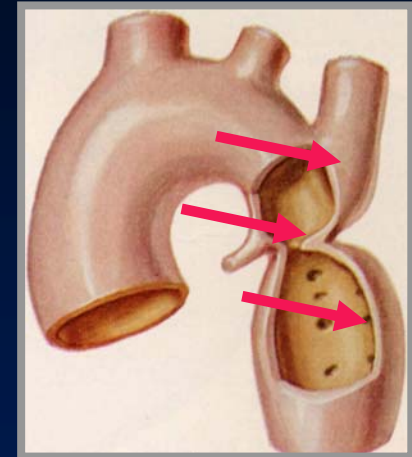
**Isolated left- sided rib notching**

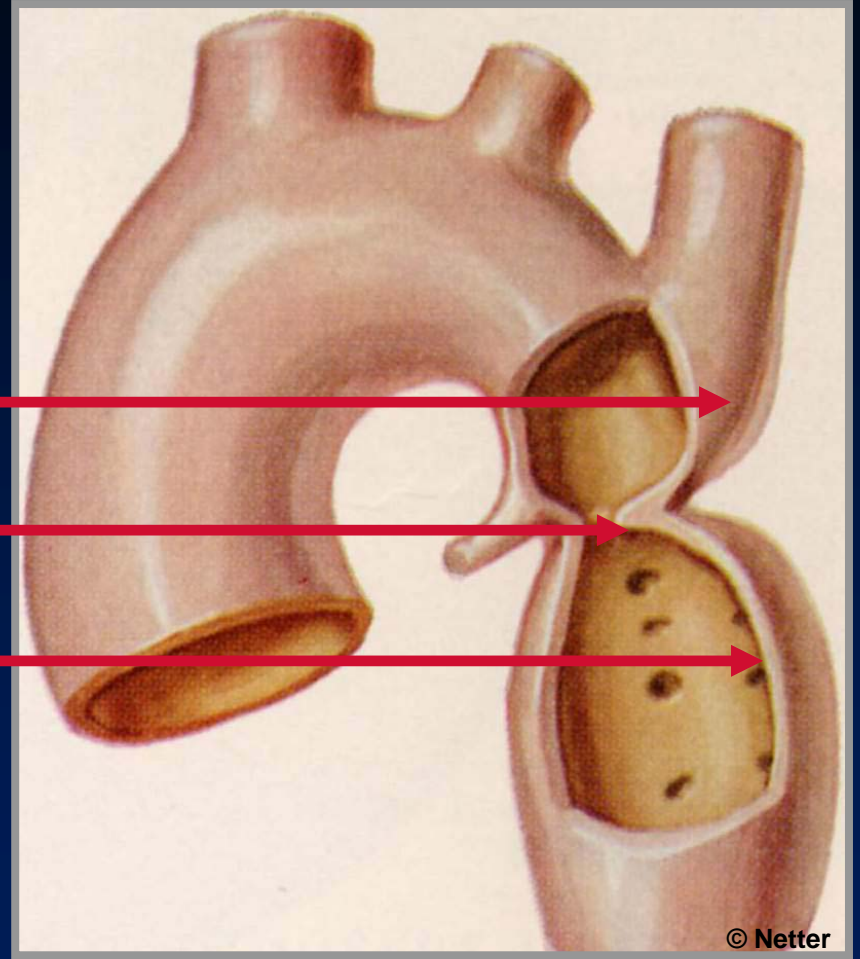
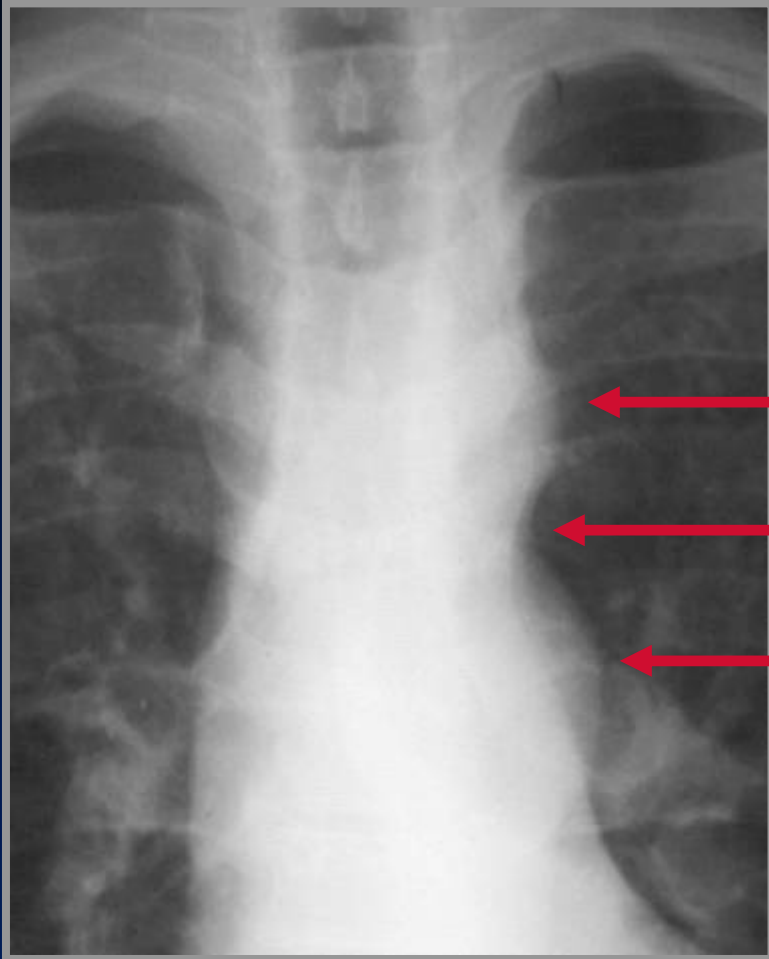
**Anomalous RSCA originates distal to site of coarct**

# X-Ray Findings

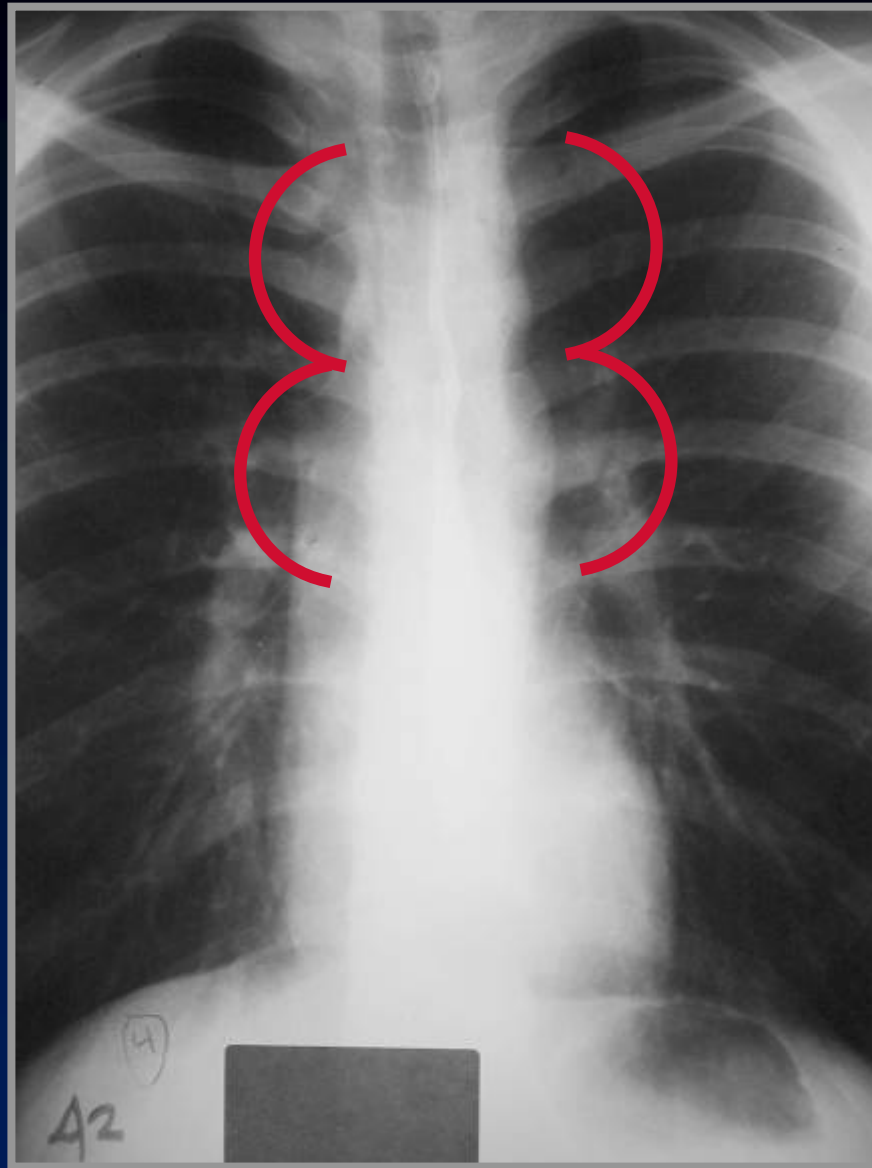
## Figure 3 Sign

- Caused by (in order)
  - Dilated LSCA or aortic knob
  - “Tuck” of coarct itself
  - Poststenotic dilatation
- Occurs in 1/3–1/2 of patients with coarct
  - Not in children
- Matched by “reverse 3” or “E” on barium-filled esophagus





**Reverse 3 sign  
on barium filled  
esophagus**



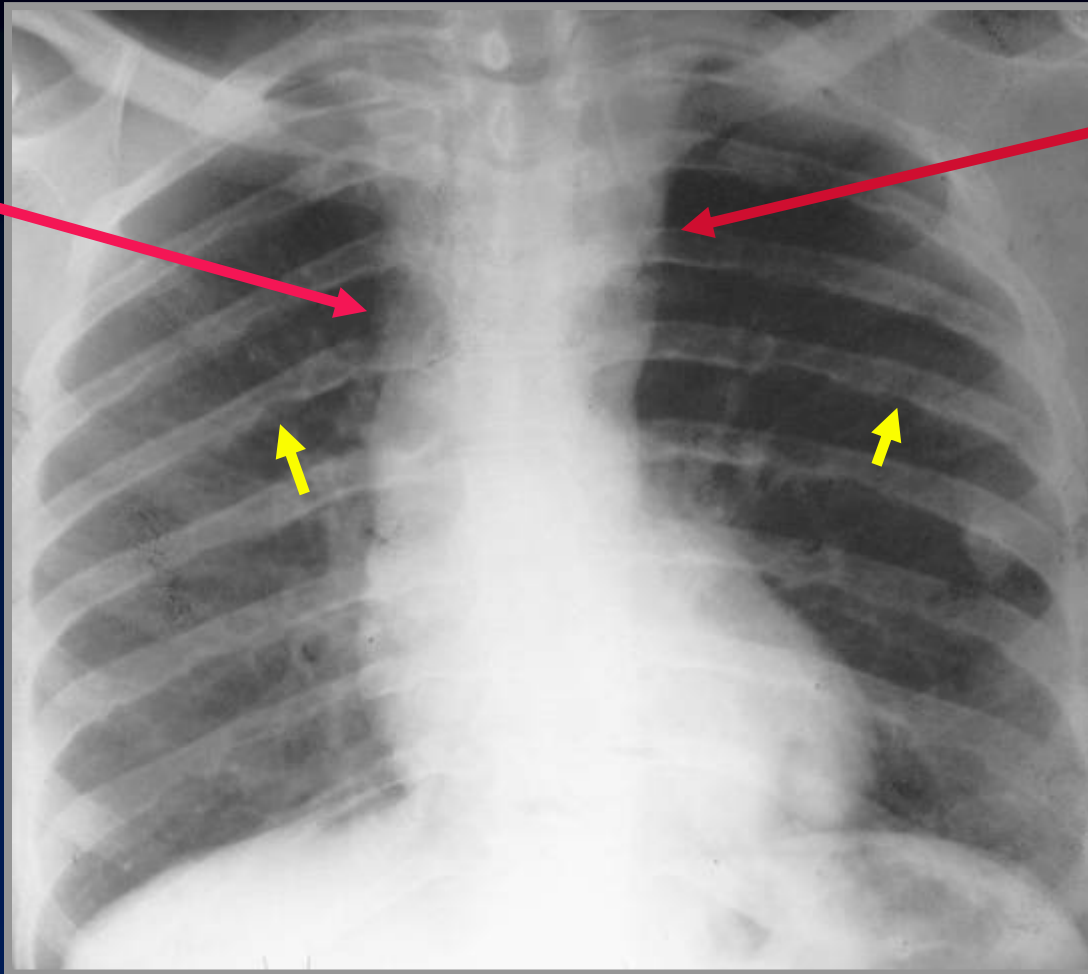
**“Figure 3 sign”  
caused by  
coarctation**

**Coarctation of the Aorta**

# **X-Ray Findings Continued**

- **Convexity of left side of mediastinum just above aortic knob 2° to**
  - **Dilated aorta proximal to coarct, or**
  - **Dilated LSCA**
    - ▲ **May be congenital or may be 2° to ↑ pressure**
- **Convexity of ascending aorta in 1/3**
  - **May be normal or small in others**

**Ascending  
Ao may be  
dilated,  
normal or  
small**



**Convexity  
above aortic  
knob due to  
dilated LSCA  
or Aorta  
proximal to  
coarct**

## **Coarctation of the Aorta**



# **Coarctation of the Aorta**

## **Clinical Findings–Infancy**

- **Severe CHF most common from 2nd to 6th week of life**
- **Weak or absent leg pulses**
- **Lower BP in the legs than in the arms**
- **EKG: RV hypertrophy because RV assumes most of the cardiac output during fetal life in these patients**

# **Coarctation of the Aorta**

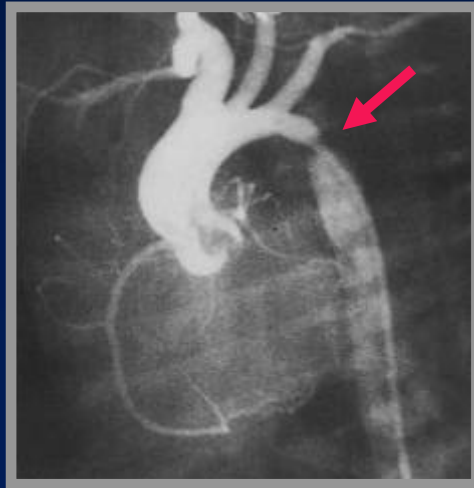
## **Echocardiographic Findings**

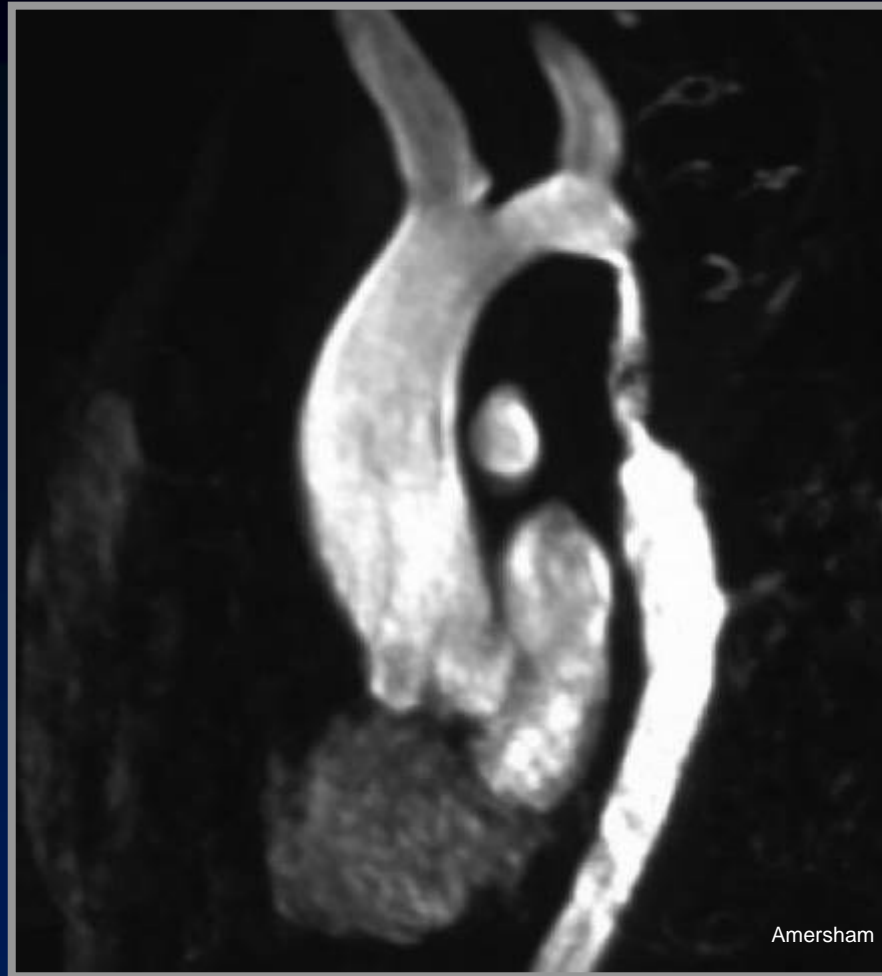
- **In infants, 2D echo can demonstrate coarcts from suprasternal notch**
- **Echo helpful in excluding associated hypoplastic left heart syndrome**

# Coarctation of the Aorta

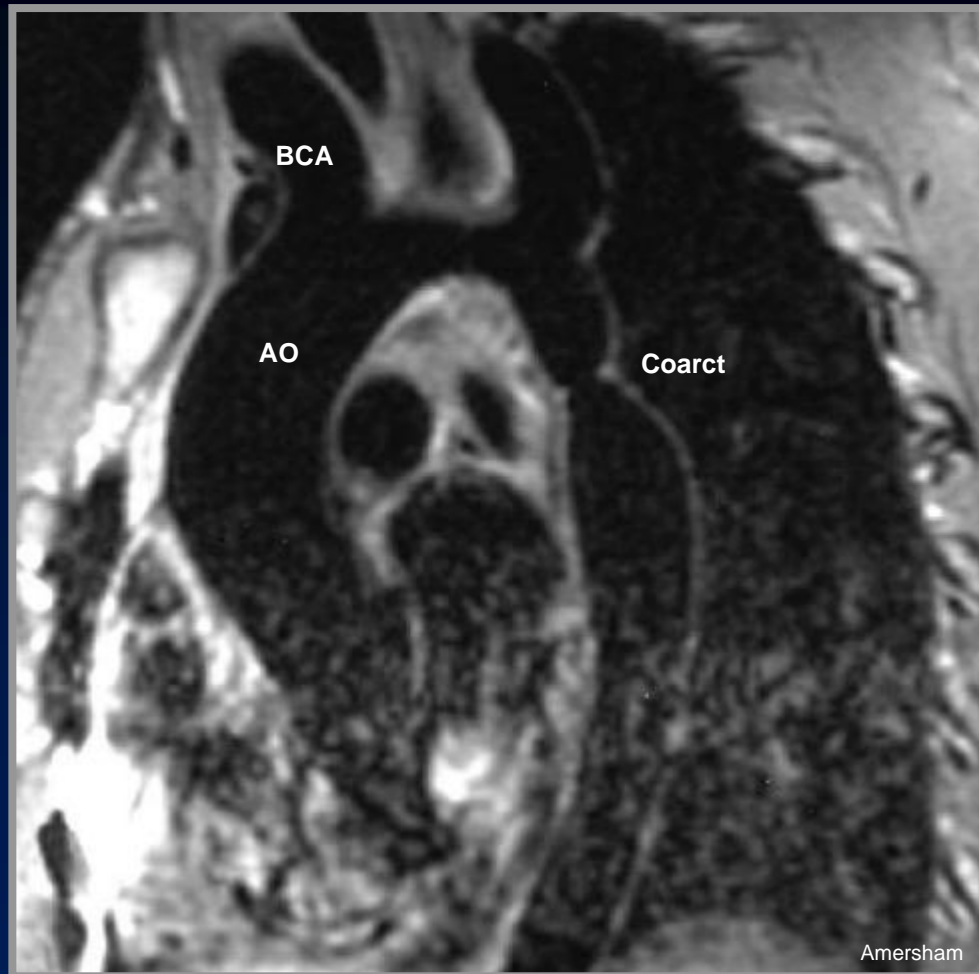
## MRI and Angiography

- MRI preferred study in children/adults
- Aortography offers greatest resolution

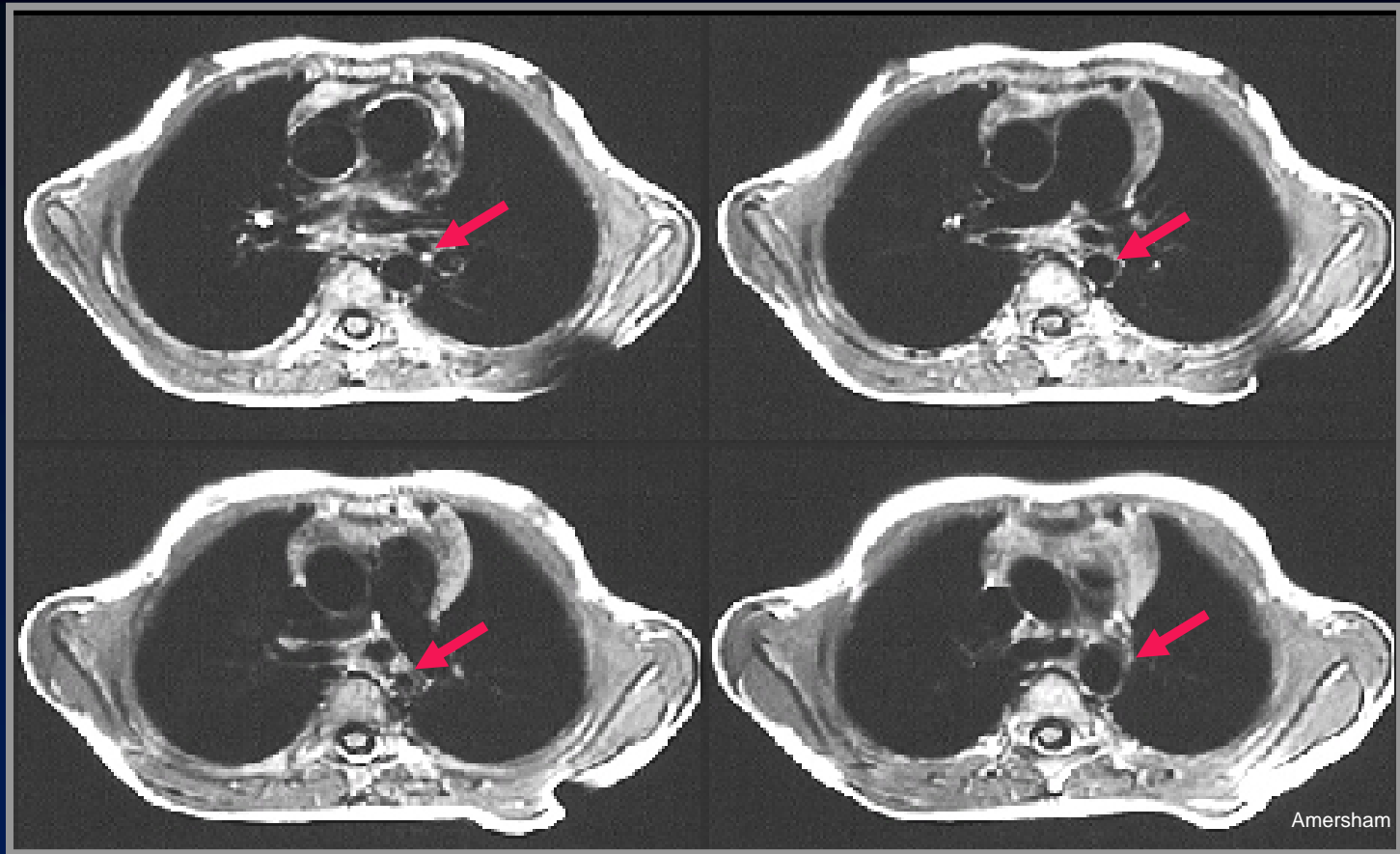




**Contrast enhanced MRA shows long segment coarctation of the aorta**



**Oblique sagittal spin-echo-Coarctation of the Aorta**



**Axial spin-echo MRI-Coarctation of the Aorta**

# **Coarctation of the Aorta**

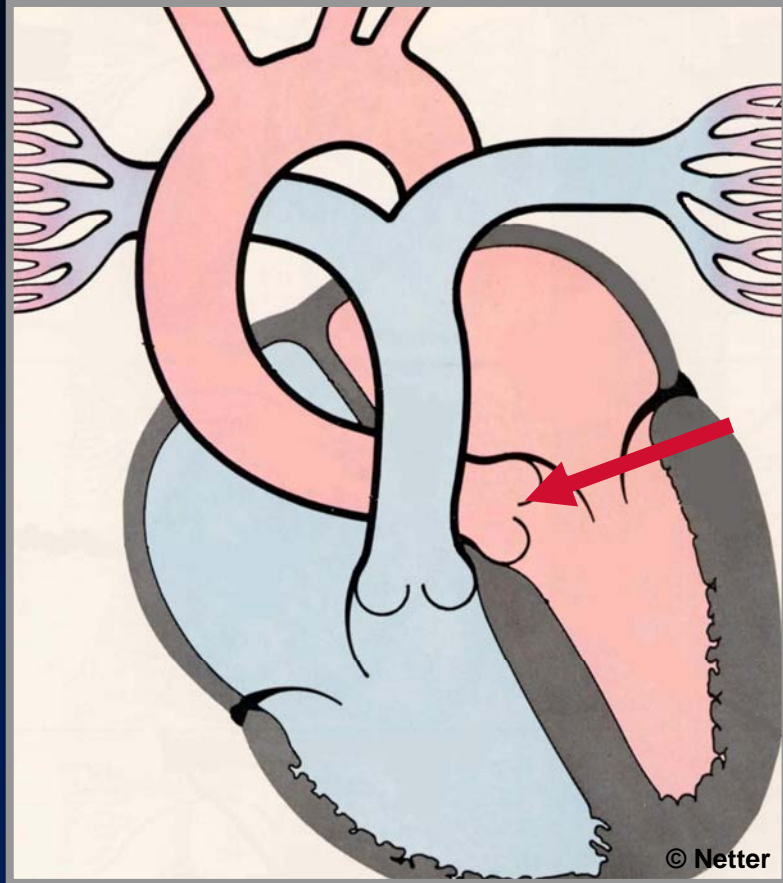
## **Complications**

- **Heart failure in neonate**
- **Subarachnoid bleeds 2° ruptured Berry aneurysms**
- **Dissection of aorta**
- **Bacterial endocarditis**
- **Mycotic aneurysm**

# Pseudocoarctation

- Buckling of aorta resembles true coarctation
- No pressure gradient ( $<30\text{mmHg}$ )
- Figure 3 sign present
- No rib notching





# **Congenital Aortic Stenosis**

# **Congenital Aortic Stenosis**

## **Valvular-General**

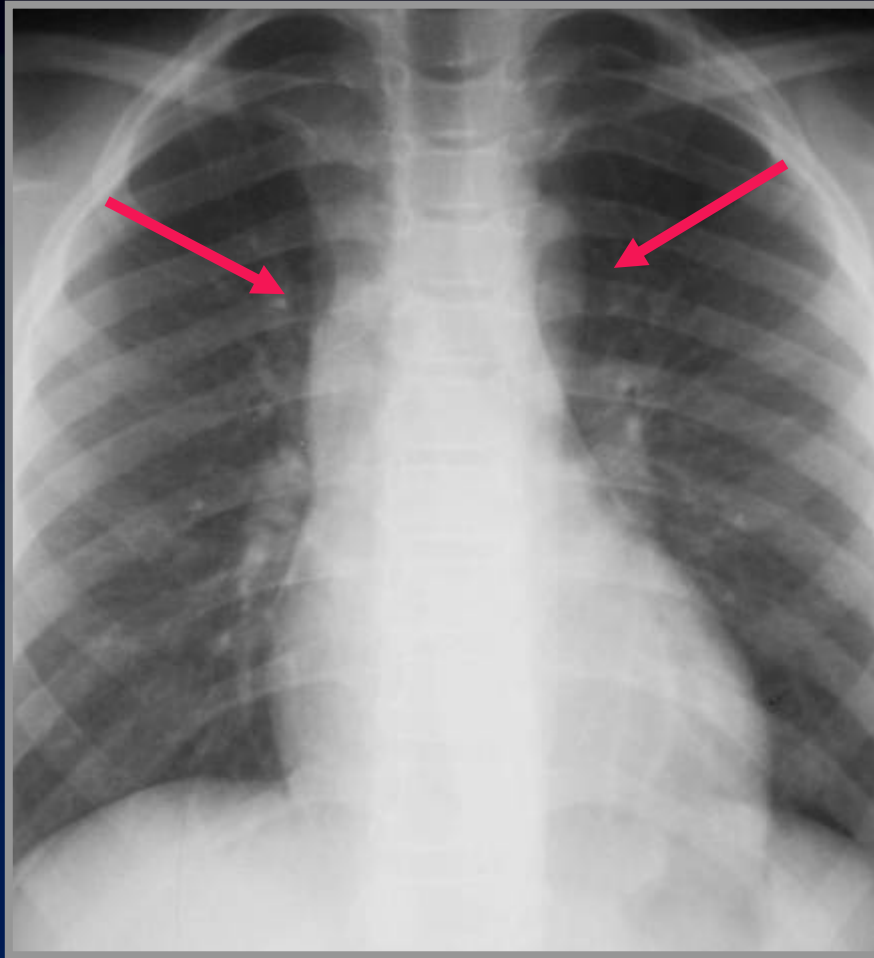
- **Bicuspid aortic valve is most common congenital cardiac anomaly (2%)**
- **Usually not stenotic in infancy**
- **Becomes stenotic when fibrosis and calcification occur**
- **About half of those with coarctation have bicuspid Ao valve**

# Congenital Aortic Stenosis

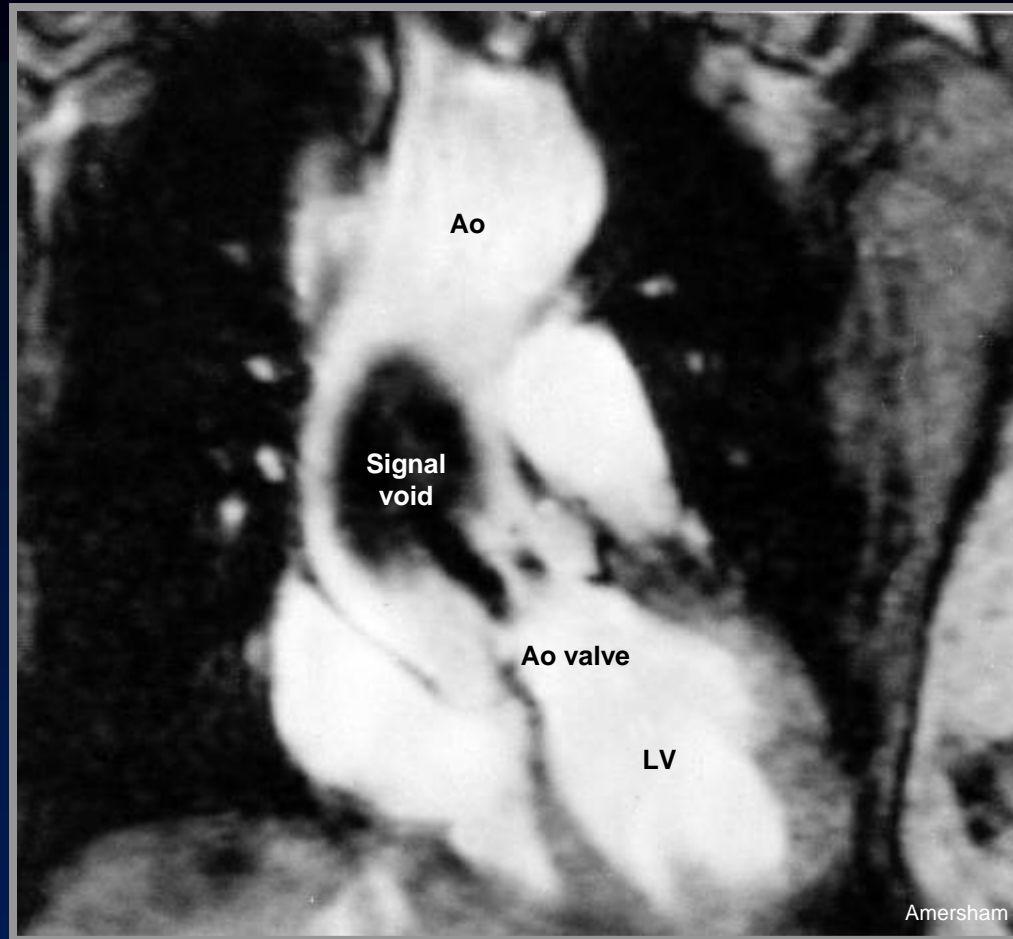
## Angiography

- Domed and thickened leaflets in systole
- Two leaflets and two sinuses of Valsalva



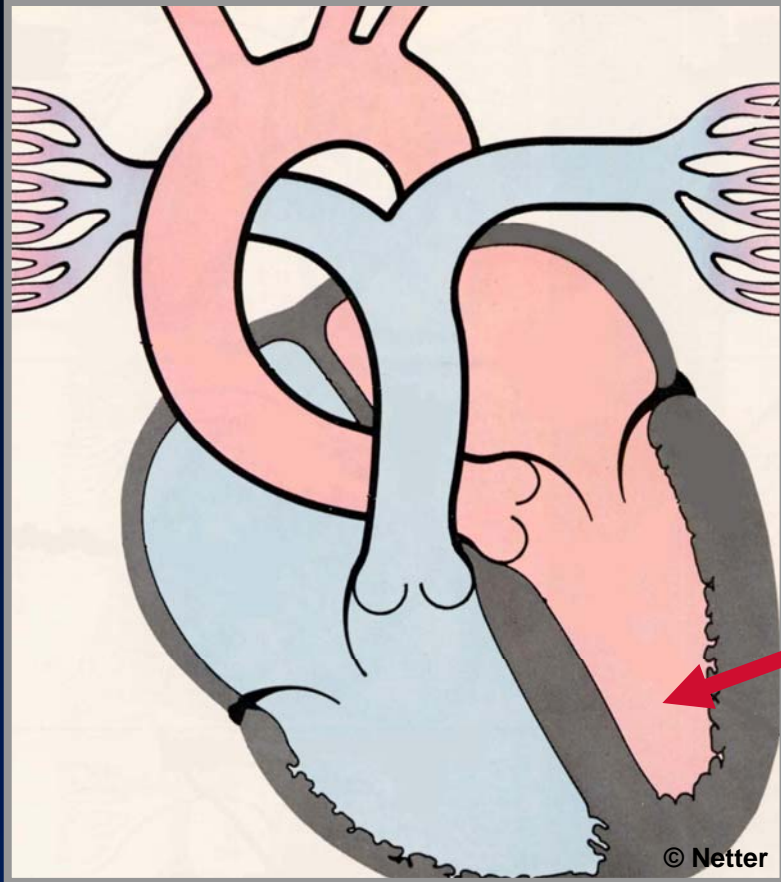


**Congenital Aortic Stenosis (10 yo)**



### **Aortic Stenosis**

**Coronal cine MRI image demonstrates a systolic signal void originating at the stenotic aortic valve. Ascending aorta is dilated**



# Hypoplastic Left Heart Syndrome

**Aortic Atresia**

# **Hypoplastic Left Heart Syndrome**

## **General**

- **Most common cause of death from cardiac cause during first week of life**
- **Common clinical expression of this lesion is CHF in first week of life**
  - **Usually cyanotic**
- **Heart is enlarged in most**

# **Hypoplastic Left Heart Syndrome**

## **General**

- **Small ascending aorta**
  - **Common to all forms**
  - **Sometimes infantile coarctation**
- **Often associated mitral stenosis or atresia or aortic stenosis or atresia**
- **In 90%, size of LA and LV small**
- **A large PDA is essential**
  - **VSD, ASD also present**

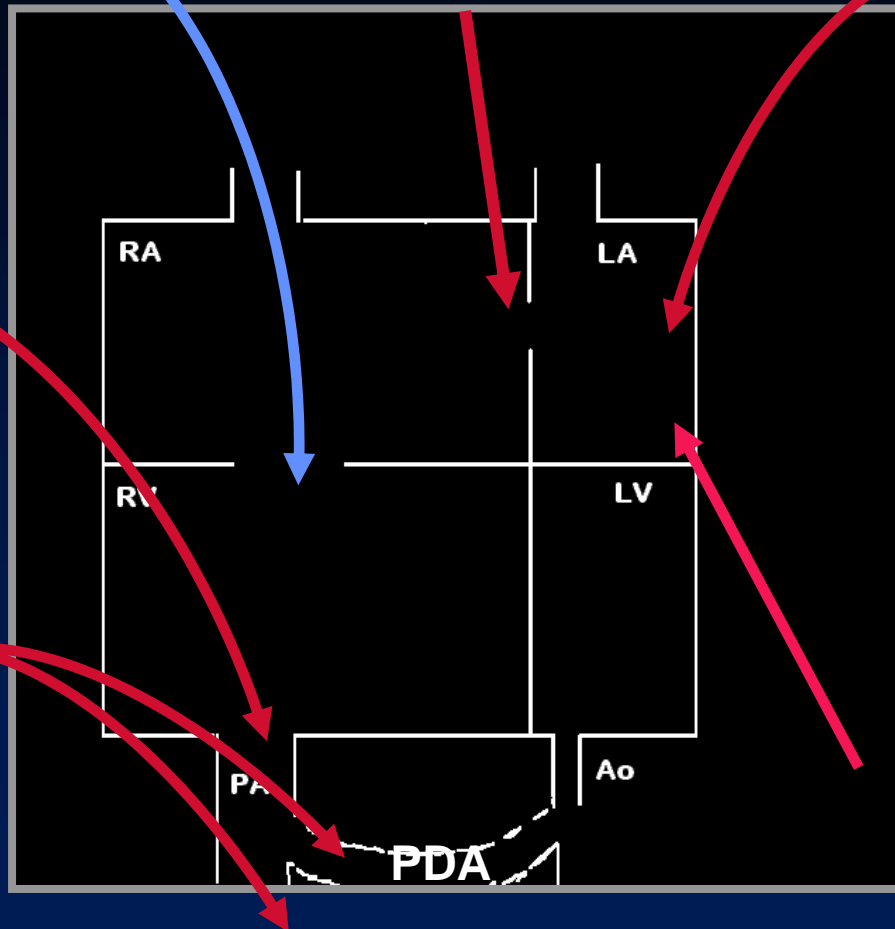


RA, RV and  
PA are  
enlarged

Oxygenated  
and  
deoxygenated  
blood enter PA

Blood passes  
through PA  
→ lungs and  
into large  
PDA → aorta  
→ to body

Passes to RA via  
ASD (L → R shunt)



Oxygenated  
blood  
returning  
from lungs  
can not  
enter LV

Obstruction  
to return of  
blood from  
lungs → CHF

**Hypoplastic Left Heart Syndrome**

**Cyanotic**

# **Hypoplastic Left Heart Syndrome Pathophysiology**

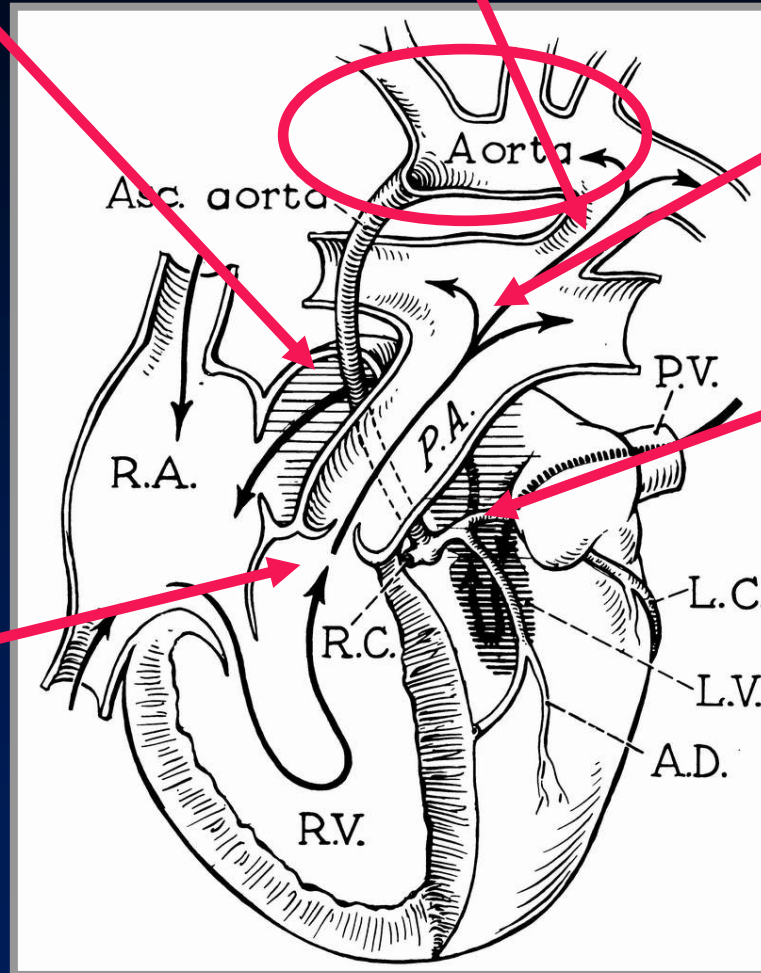
- **Since outflow tract from L heart is small, aerated blood always shunted**
- **Large PDA needed to get aerated blood to body**
- **Blood to head, arms and coronaries flows through PDA, then backwards through arch**

**Need L → R  
shunt  
through ASD  
to get blood  
out of LA**

**Some blood passes  
through large PDA to  
aorta and out to body**

**Some  
deoxygenated  
blood goes to  
lungs**

**Blood  
returning from  
body mixes  
with  
oxygenated  
blood from LA;  
passes into PA**

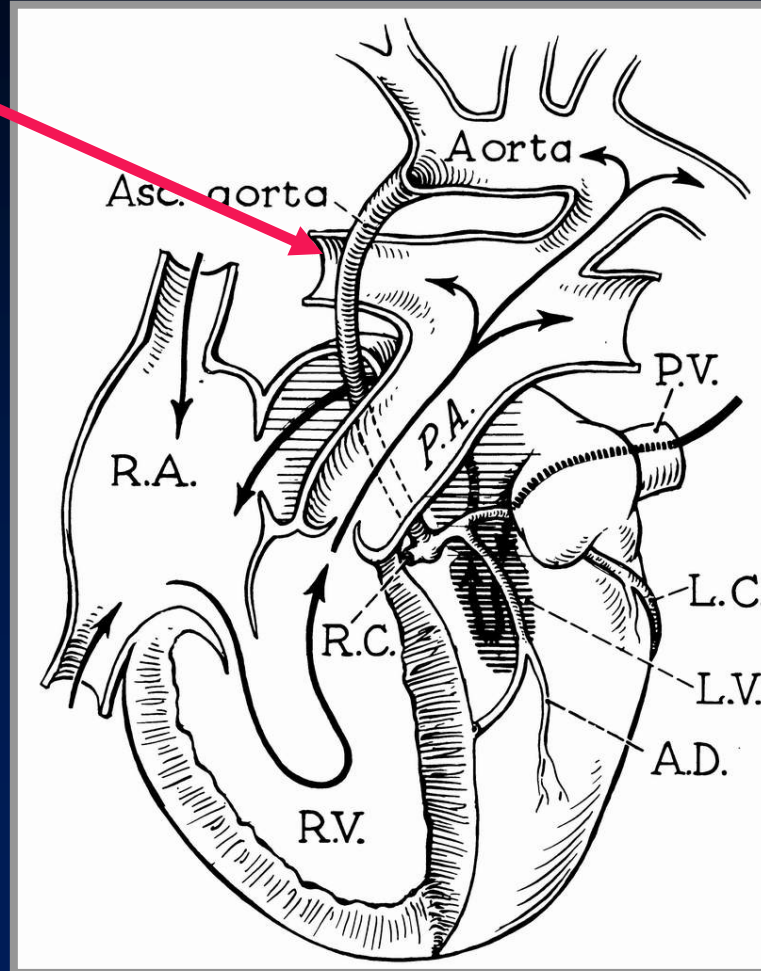
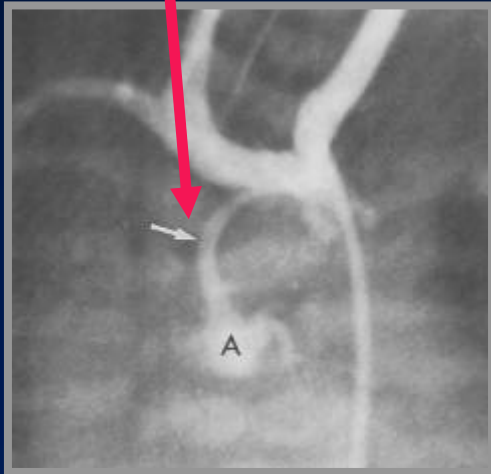


**Blood  
returning  
from lungs  
can not exit  
LA to LV  
because of  
atretic mitral  
valve**

**Hypoplastic Left Heart Syndrome**

# Hypoplastic Left Heart Syndrome

Atretic aorta



# **Hypoplastic Left Heart Syndrome Associated Anomalies**

- **Coarctation of the aorta**
- **Interruption of the aortic arch**
- **AV communis**
- **Anomalies of the R subclavian artery**
- **Bicuspid aortic valve**

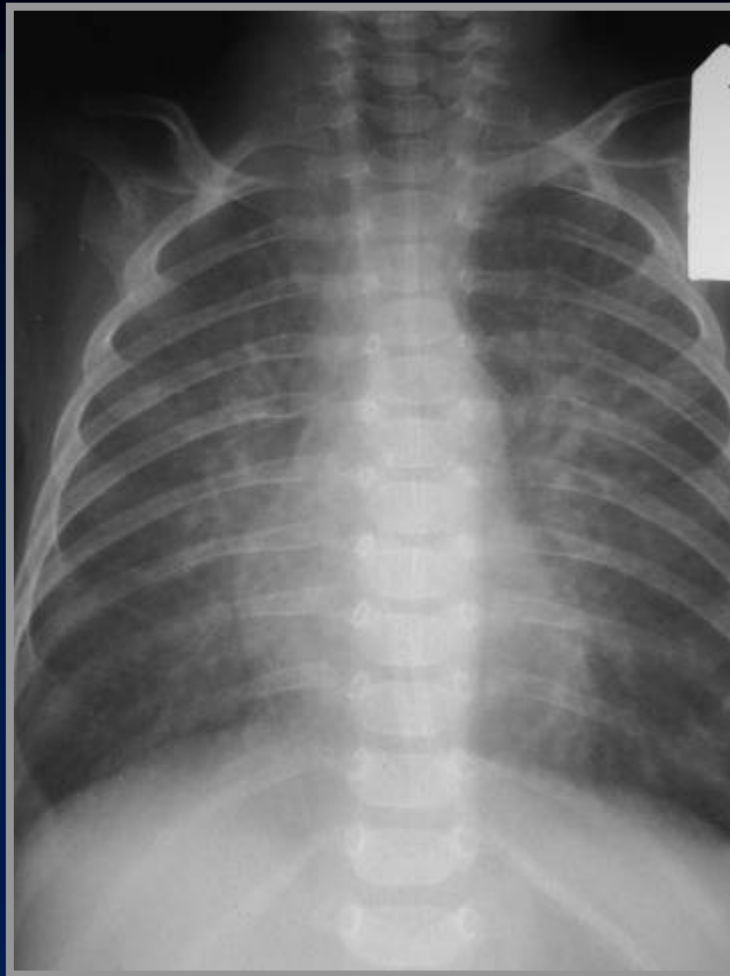
# **Hypoplastic Left Heart Syndrome**

## **X-ray Findings**

- **Increased load on RV → marked cardiomegaly at birth**
- **Obstruction to return of blood from lungs → CHF at birth**
  - **Most common cause of CHF in first two weeks of life**

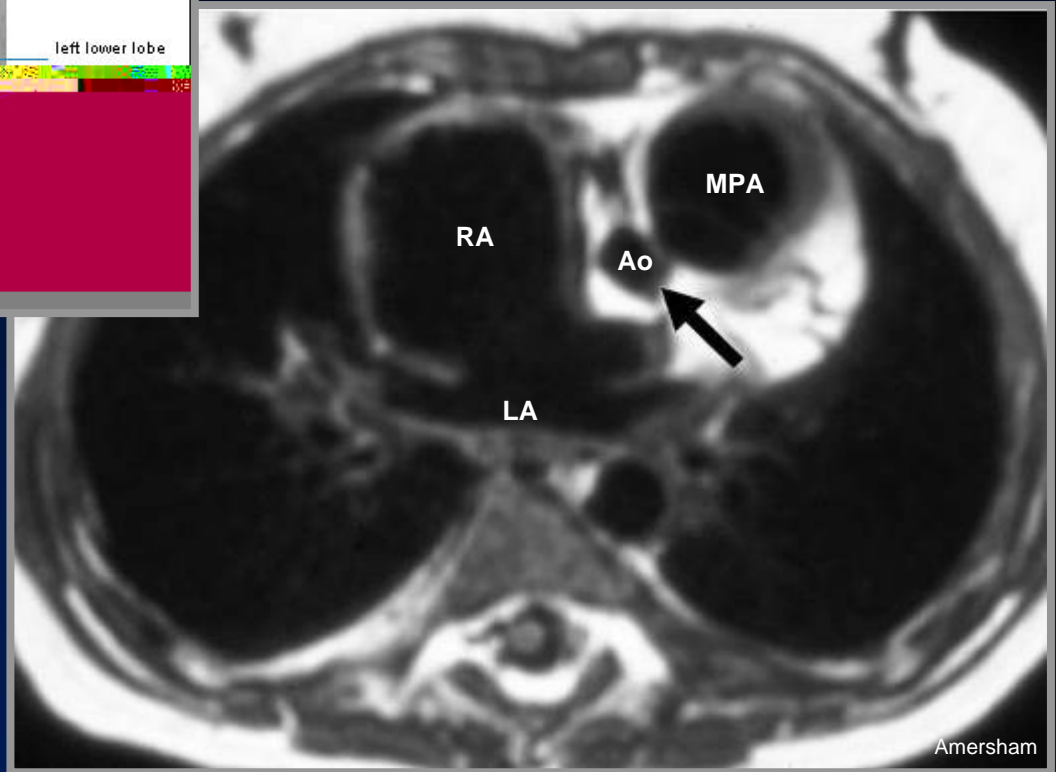
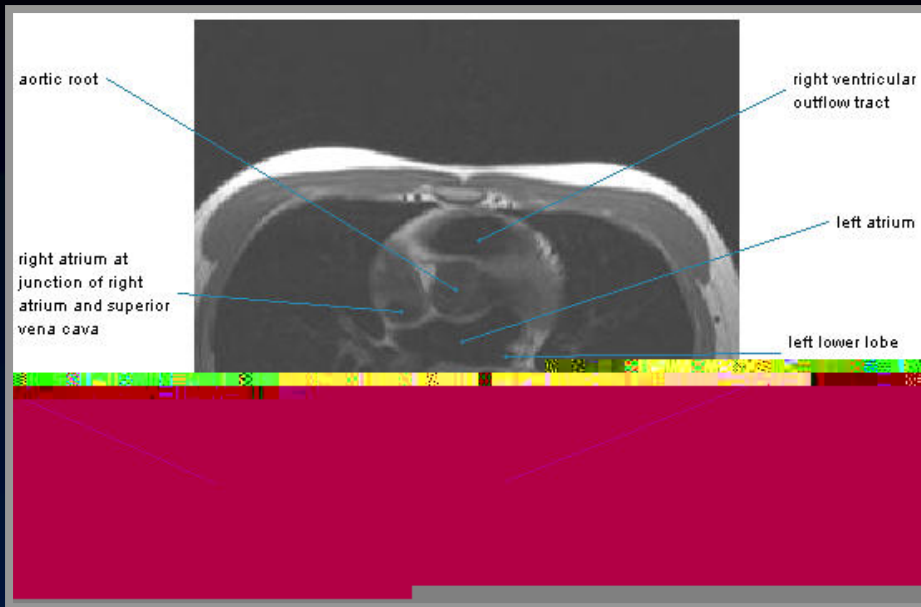


**Hypoplastic Left Heart Syndrome**



**Hypoplastic Left Heart Syndrome**





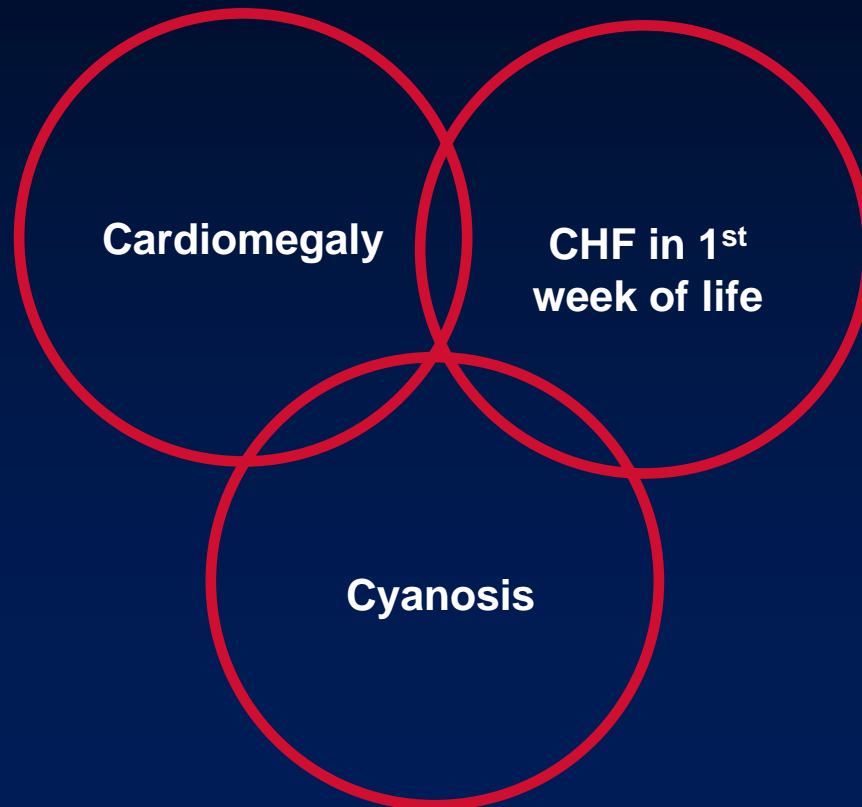
**Hypoplastic Left heart Syndrome**  
**Gated spin echo at base of heart shows hypoplastic aorta**  
**(arrow) posterior and right of main pulmonary artery**

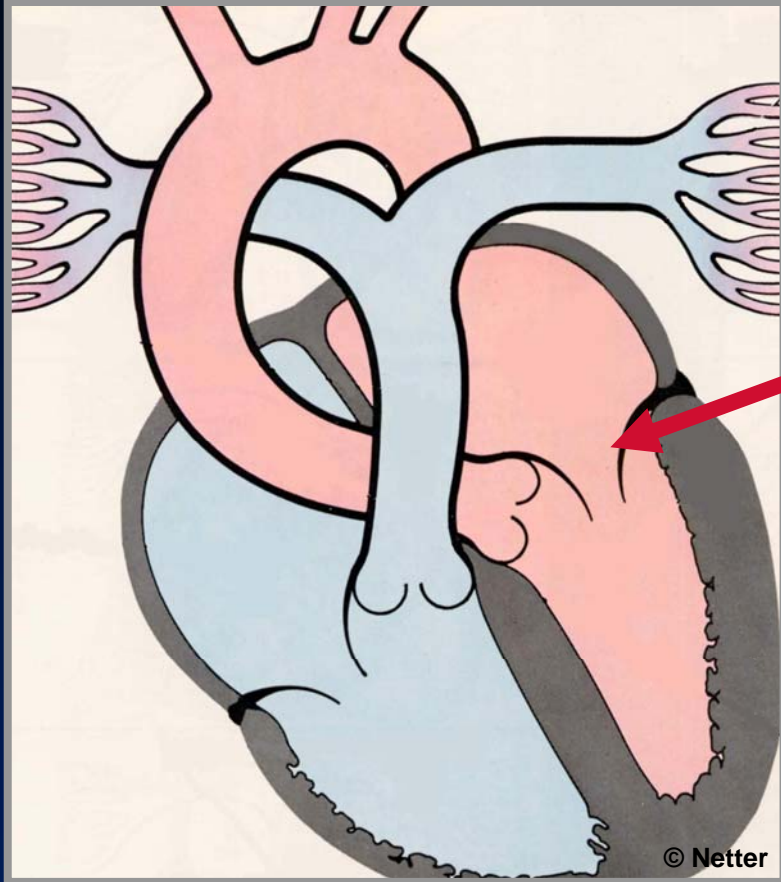
# **Hypoplastic Left Heart Syndrome**

## **Diagnosis**

- **Diagnosis can be made by echo**
- **Catheterization may be hazardous**
  - **Spasm of PDA during cath can → death**

# Hypoplastic Left Heart Syndrome Triad





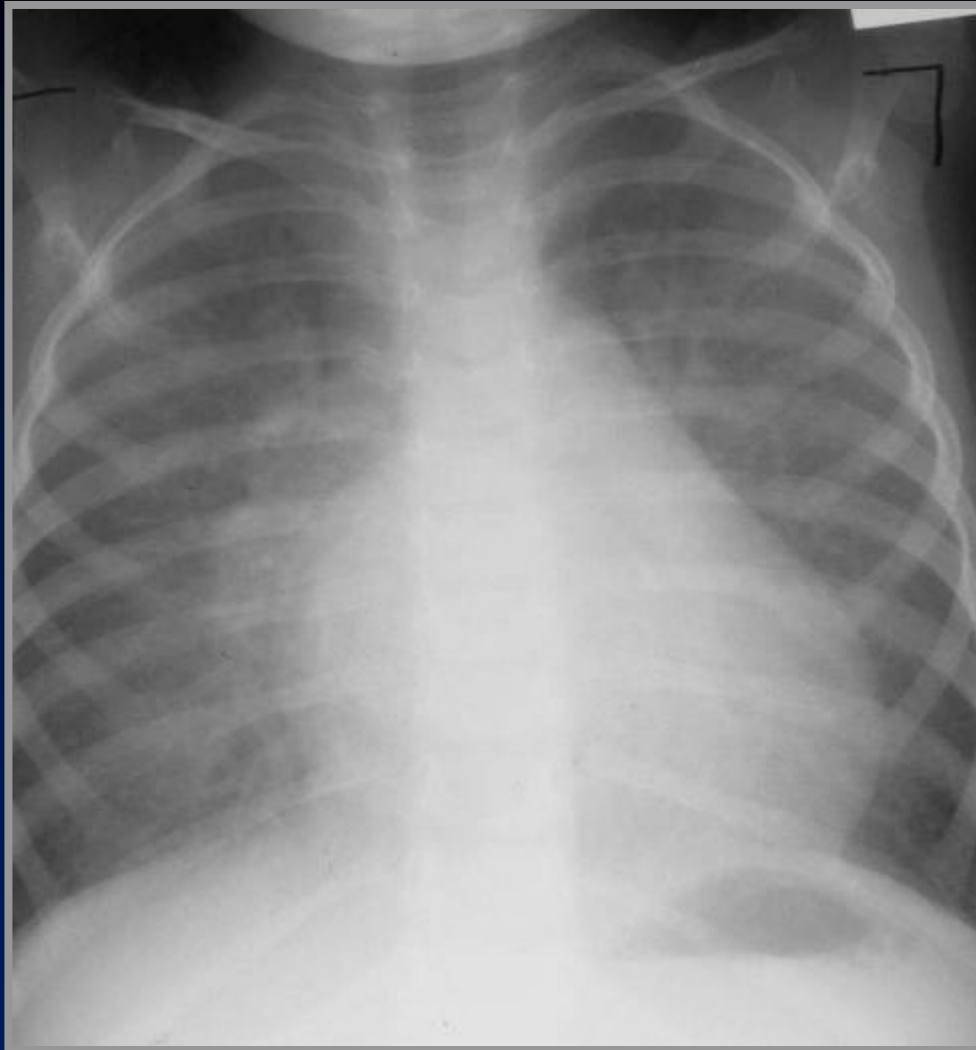
# Congenital Mitral Stenosis

# **Congenital Mitral Stenosis**

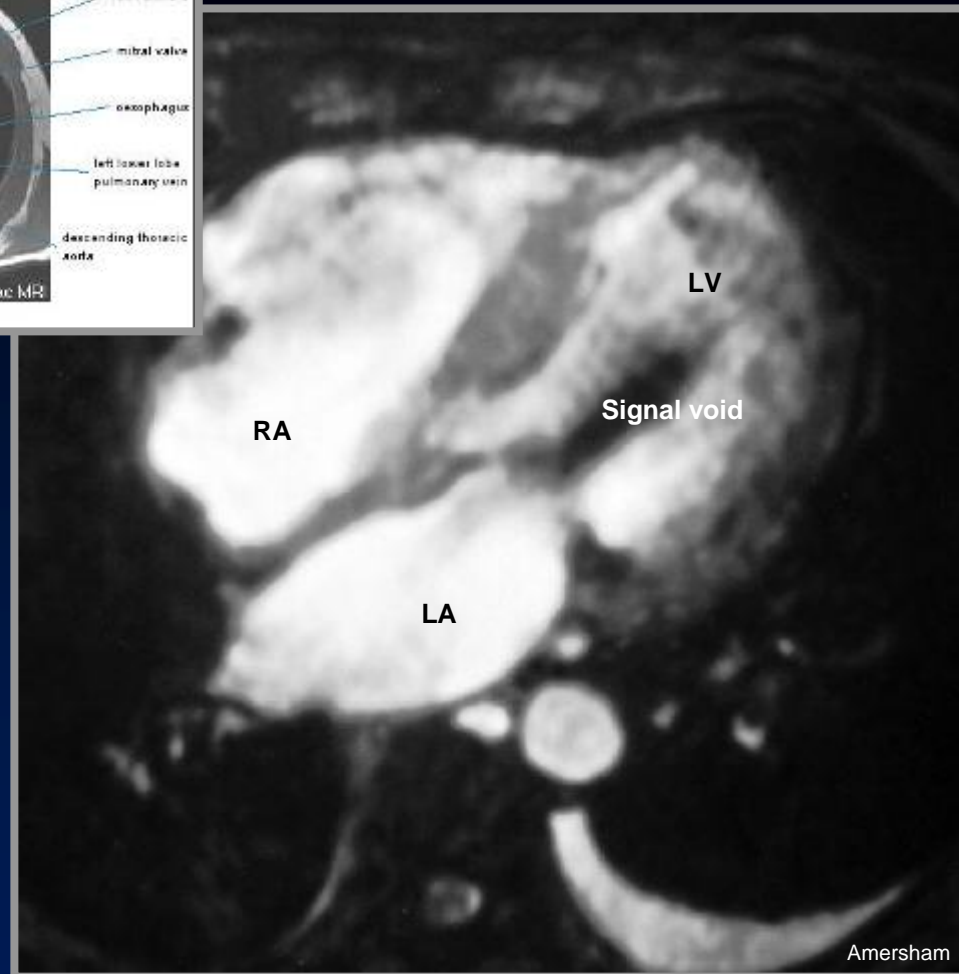
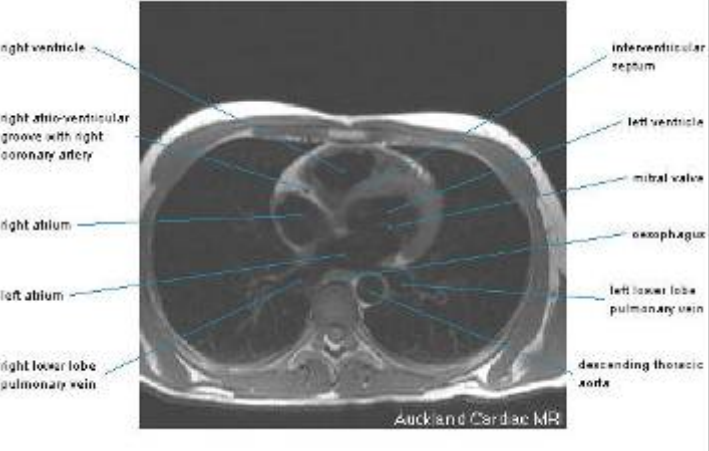
- **Exists as isolated abnormality 25% of time**
- **Coexists with VSD 30% of time**
- **Coexists with another form of left ventricular outflow obstruction 40% of time—SHONE'S Syndrome**

# Shone's Syndrome

- Parachute mitral valve
- Supravalvular mitral ring
- Subaortic stenosis
- Coarctation of aorta

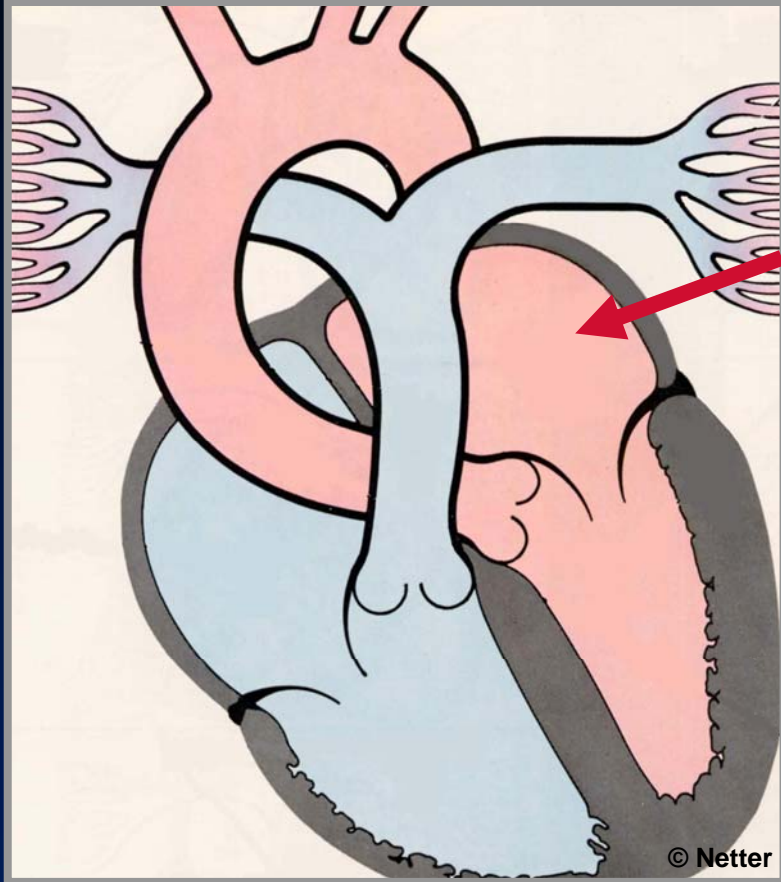


**Congenital Mitral Stenosis**



**Cine MR image in axial plane demonstrates a diastolic signal void emanating from the mitral valve indicative of mitral stenosis**





# Cor Triatriatum

# Cor Triatriatum

## General

- Rare congenital anomaly
- Fibromuscular septum with single, large, opening separates embryonic common pulmonary vein from left atrium



# Cor Triatriatum

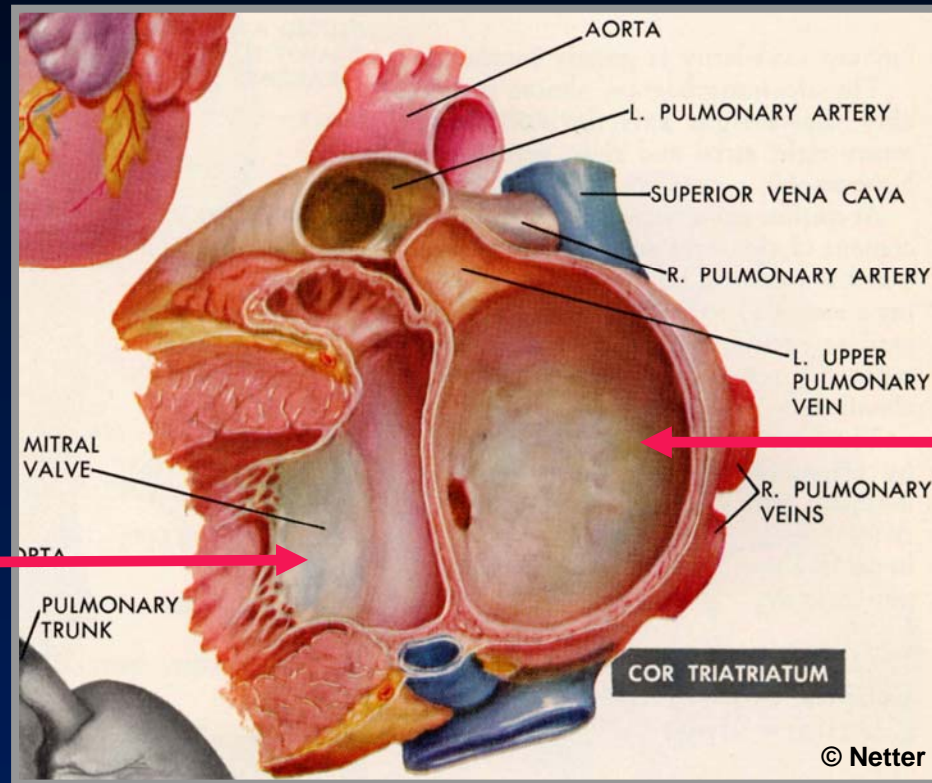
## Anatomy

- Proximal, accessory chamber lies posteriorly and receives pulmonic veins
- Distal, true atrial chamber lies anteriorly, emptying into left ventricle through mitral valve



# Cor Triatriatum

**Distal, true atrial chamber lies anteriorly and contains mitral valve**



**Proximal accessory chamber lies posterior and receives pulmonary veins**

# **Cor Triatriatum**

## **Associations**

- **ASD**
- **PDA**
- **Anomalous pulmonary venous drainage**
- **Left SVC**
- **VSD**
- **Tetralogy of Fallot**

# **Cor Triatriatum**

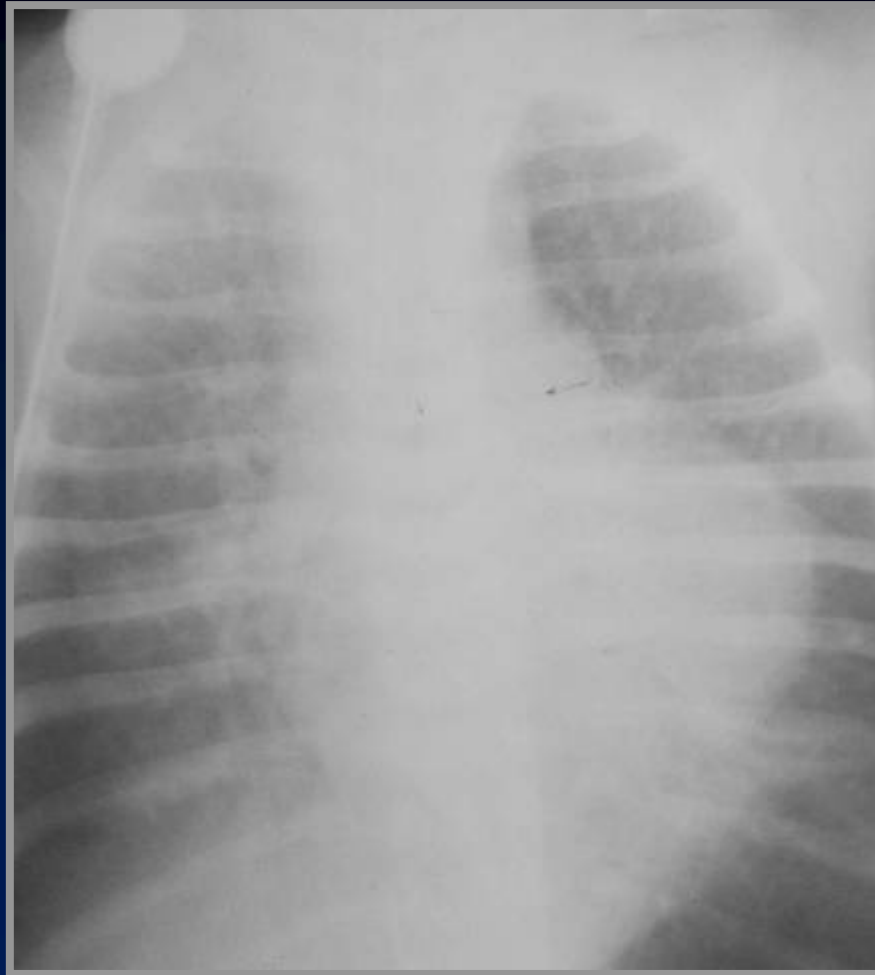
## **Clinical**

- **Clinically similar to mitral stenosis**
- **Dyspnea**
- **Heart failure**
- **Failure to thrive**

# **Cor Triatriatum**

## **X-ray Findings**

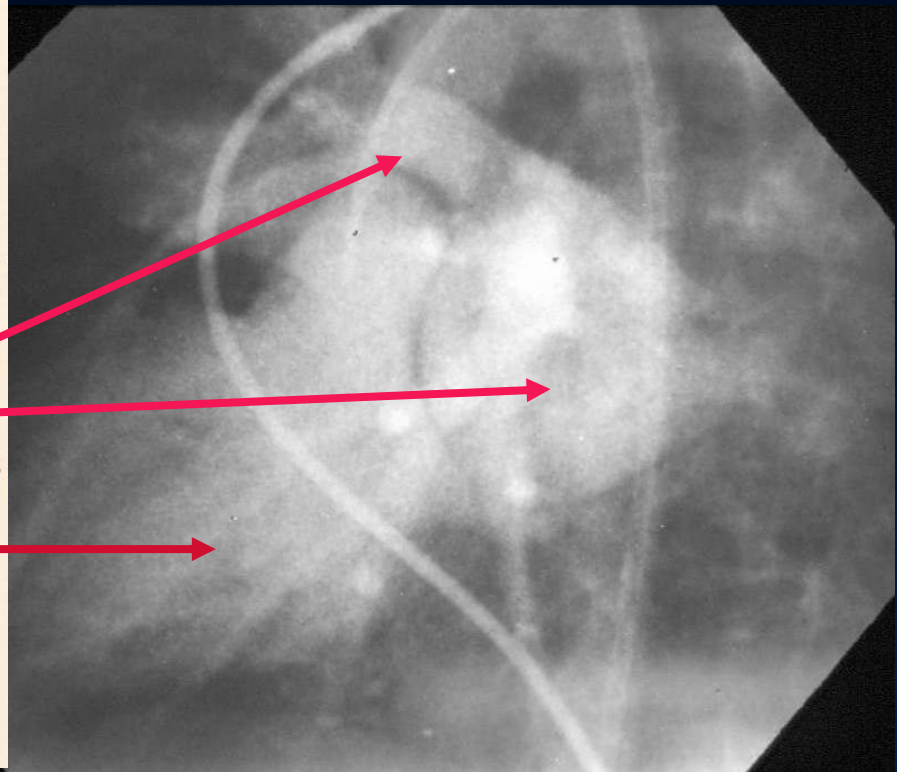
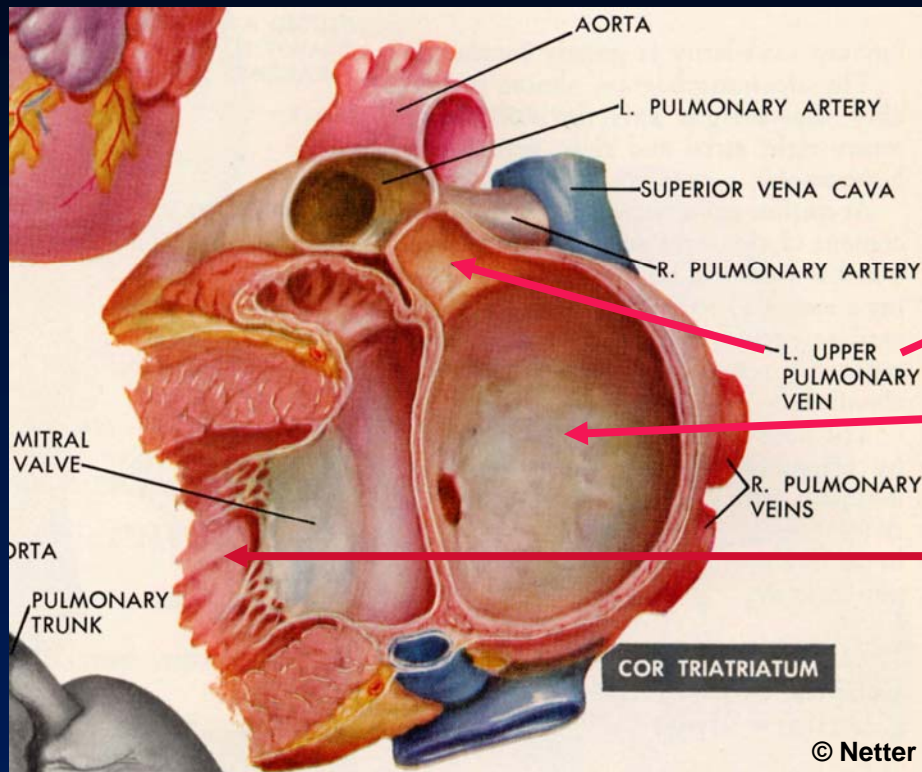
- **Pulmonary edema**
- **Enlarged LA**

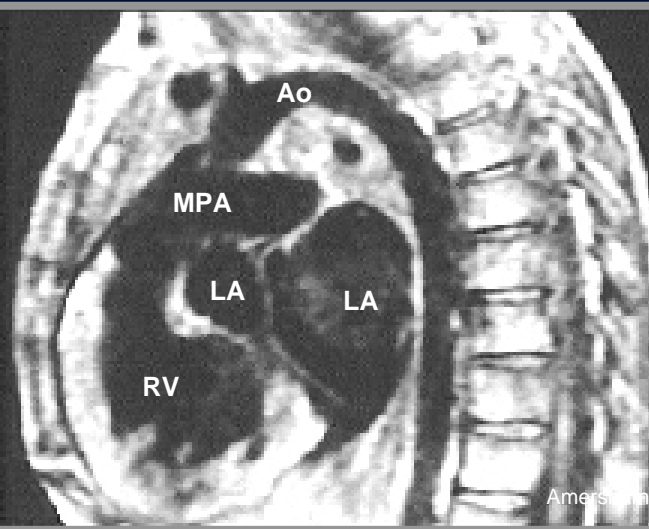
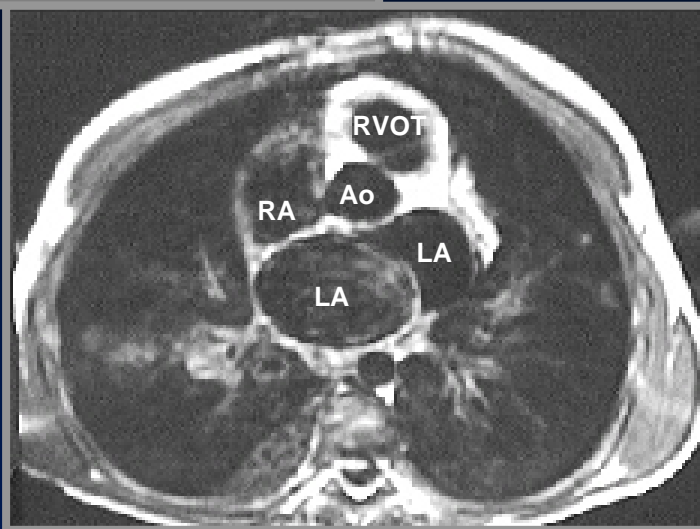
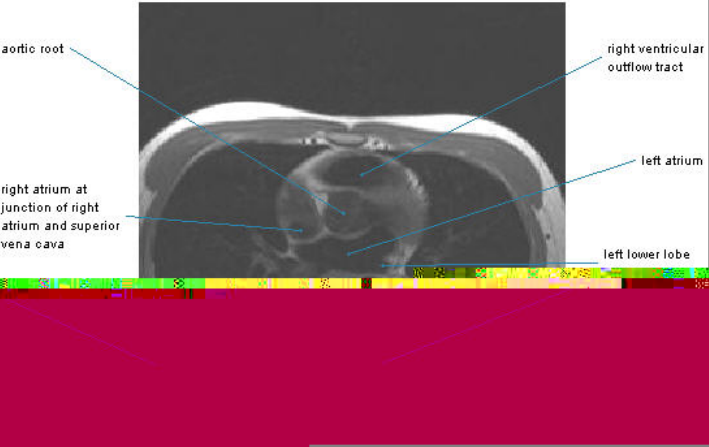


**Cor Triatriatum**



# Cor Triatriatum - angiography





## Cor Triatriatum



# **Cor Triatriatum**

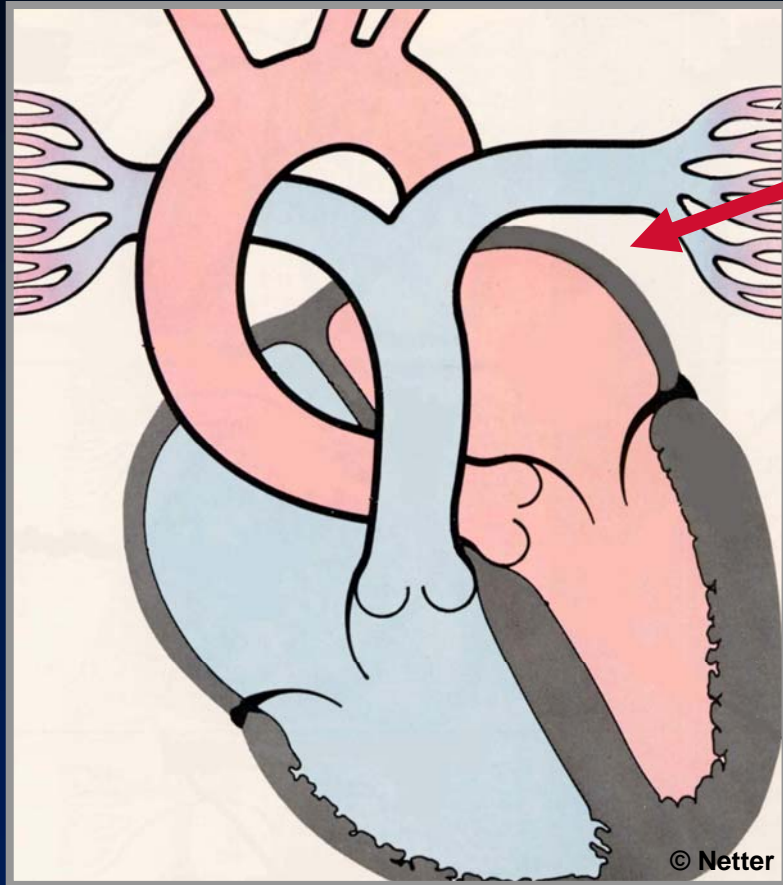
## **Treatment**

- **Surgical excision of obstructing membrane**

# Cor Triatriatum

## Prognosis

- Usually fatal in first 2 years of life
  - Associated abnormalities



# Obstruction Of the Venous Return from the Lungs

TAPVR from below  
Diaphragm

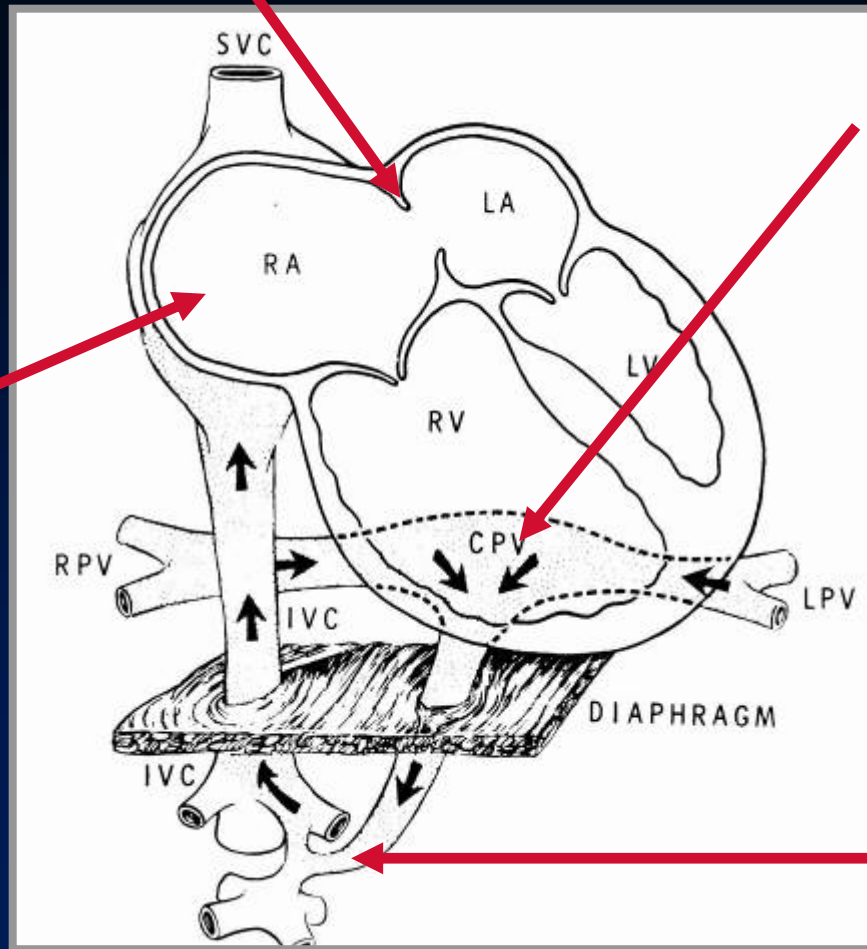
# **TAPVR**

## **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**

Obligatory R → L shunt to  
carry oxygenated blood to  
body

Oxygenated  
blood  
returns to  
RA



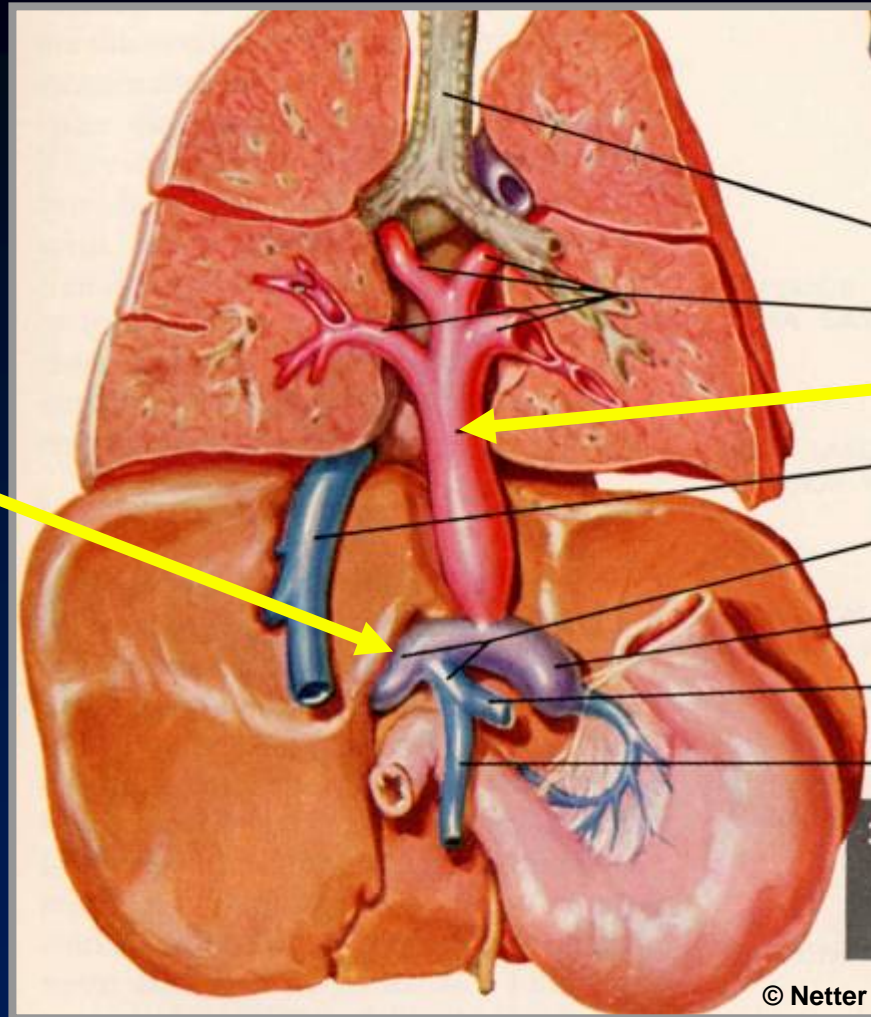
Blood returning  
from lungs →  
pulmonary veins  
which are  
constricted by  
diaphragm → CHF

Pulmonary veins  
empty into portal  
vein or IVC

**TAPVR-Type III-Infradiaphragmatic**



**Portal vein**

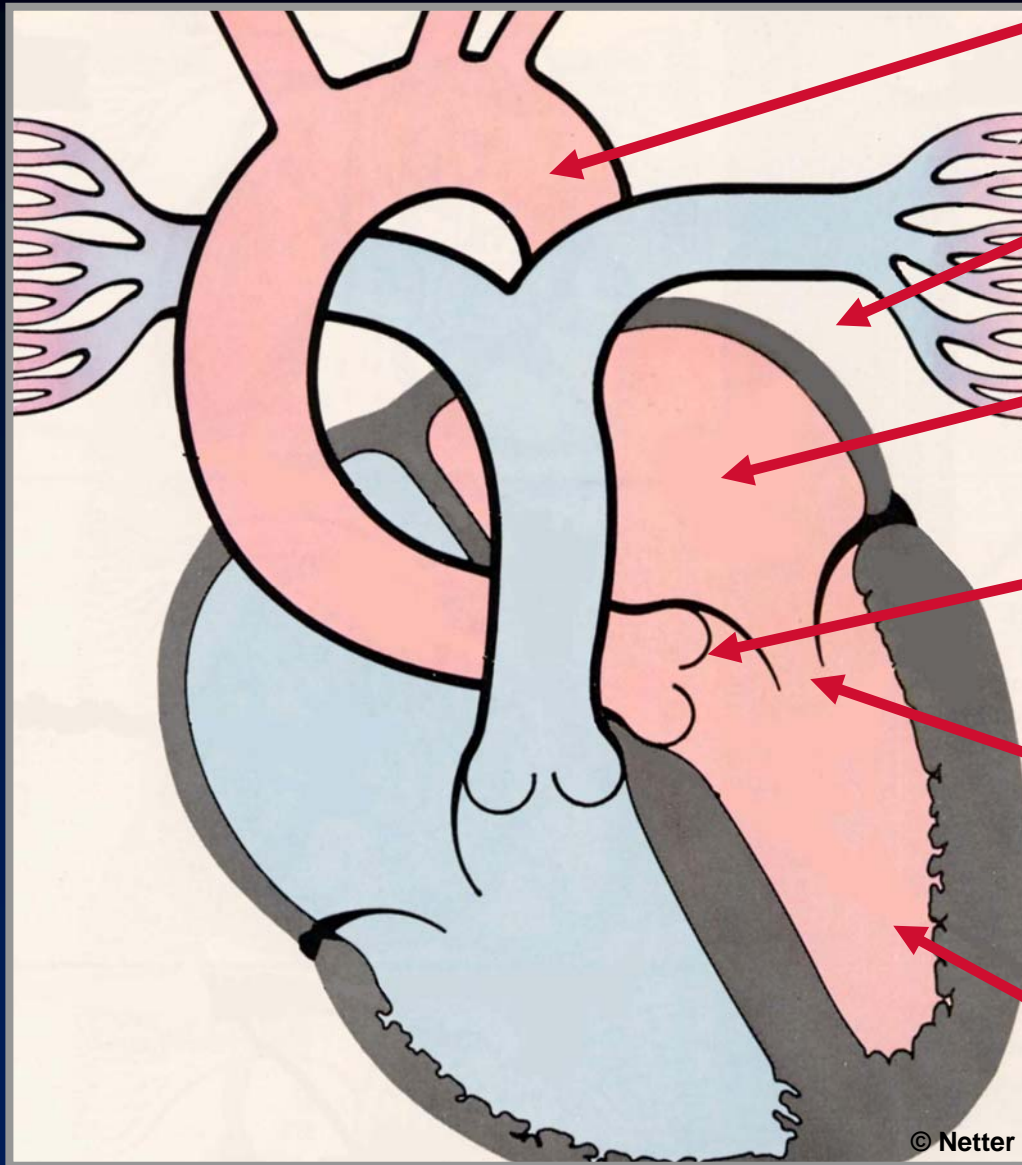


**Pulmonary  
veins**

**TAPVR-Type III-  
Infradiaphragmatic**



# Causes of CHF in the Newborn



**Coarctation of the Aorta**

**Obstruction to venous return from lungs**

**Cor Triatriatum**

**Congenital Aortic Stenosis**

**Congenital Mitral Stenosis**

**Hypoplastic Left Heart**

**The End**