Obstructive Lesions

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

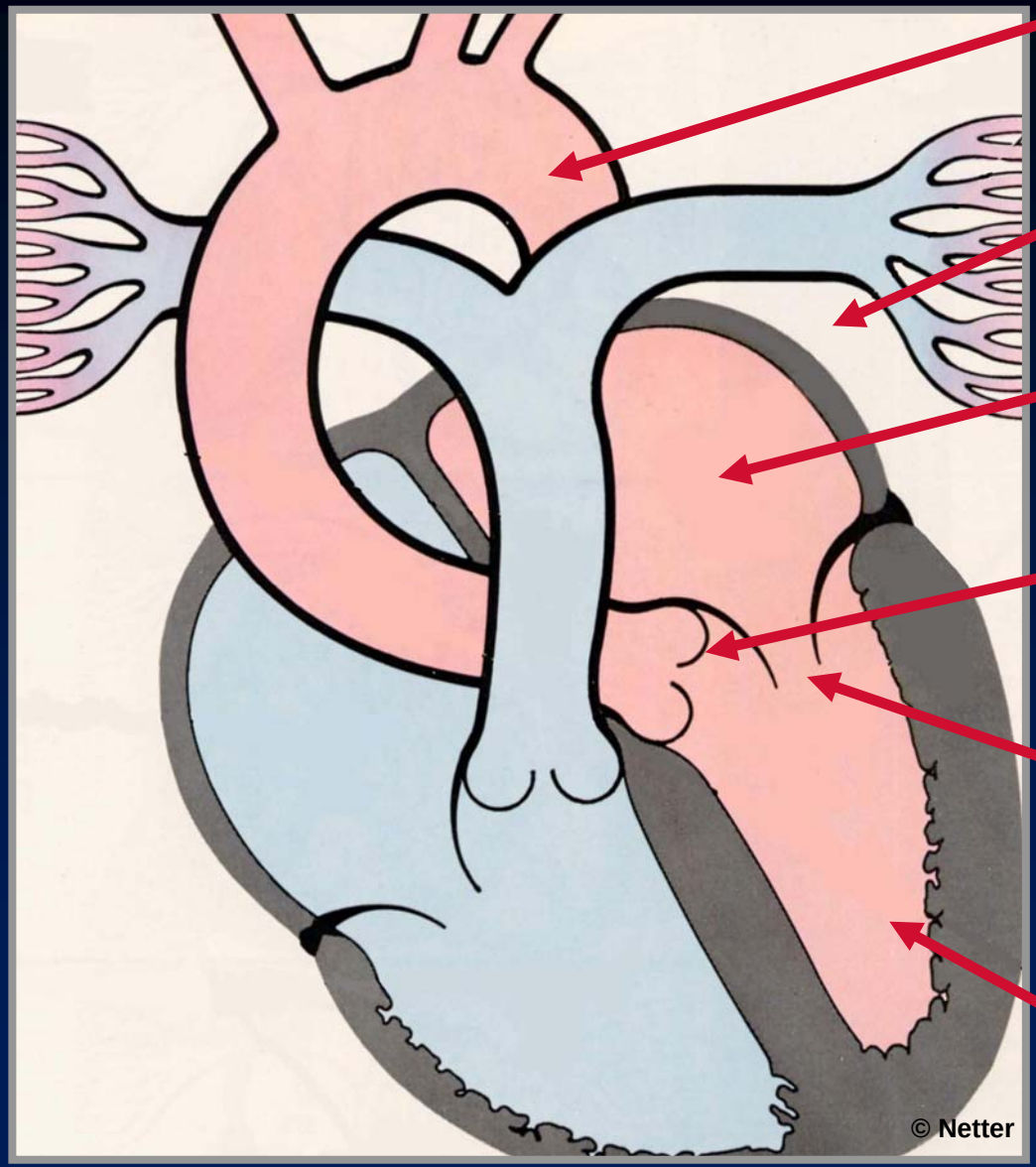
Lesions That Cause CHF

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

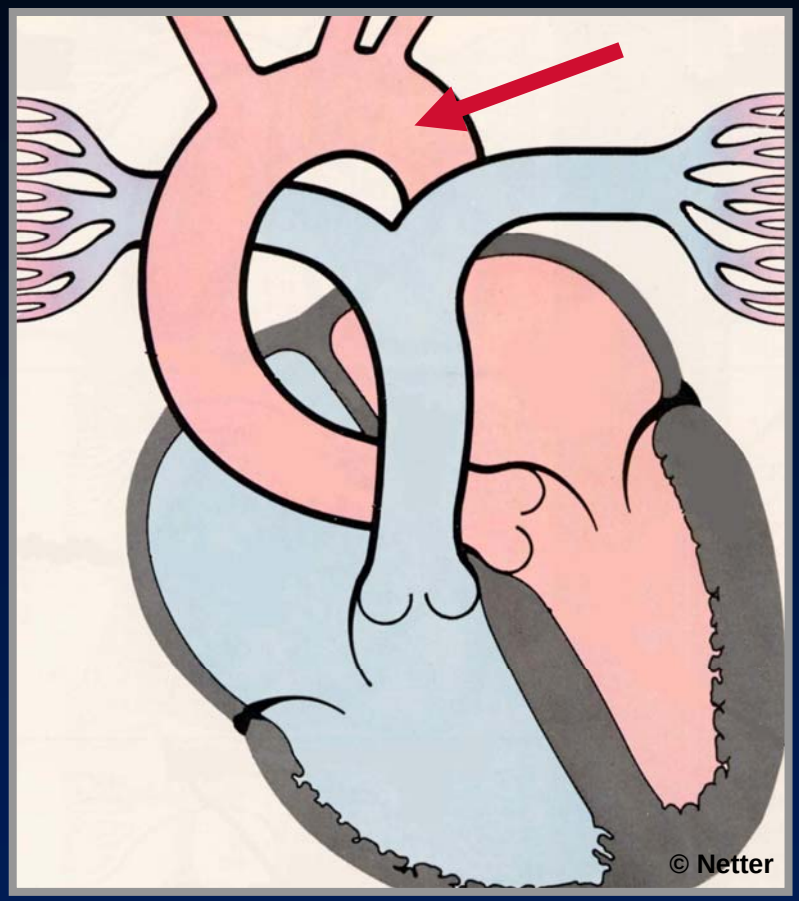
< 24 hrs……..**Intrauterine arrythmia**

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

2-6 weeks…….**Infantile coarctation**

1-4 months……**Large L ➞ R shunts**

VSD, ASD, PDA, AV Canal

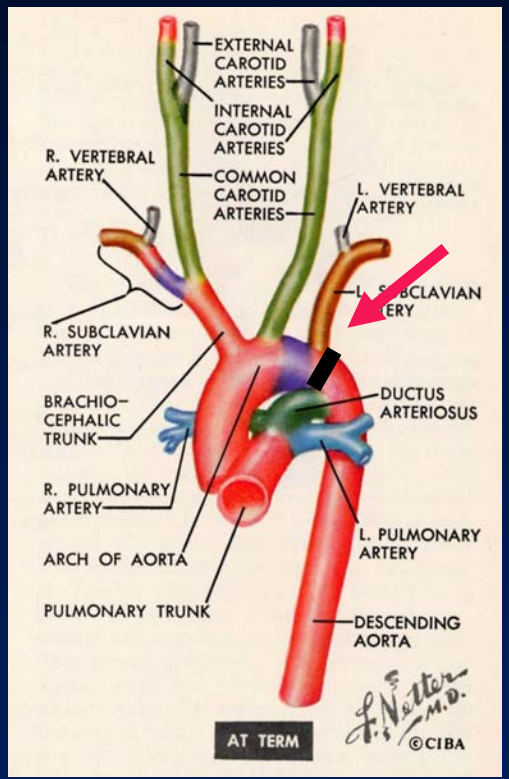


Coarctation Of the Aorta

Coarctation of the Aorta
General

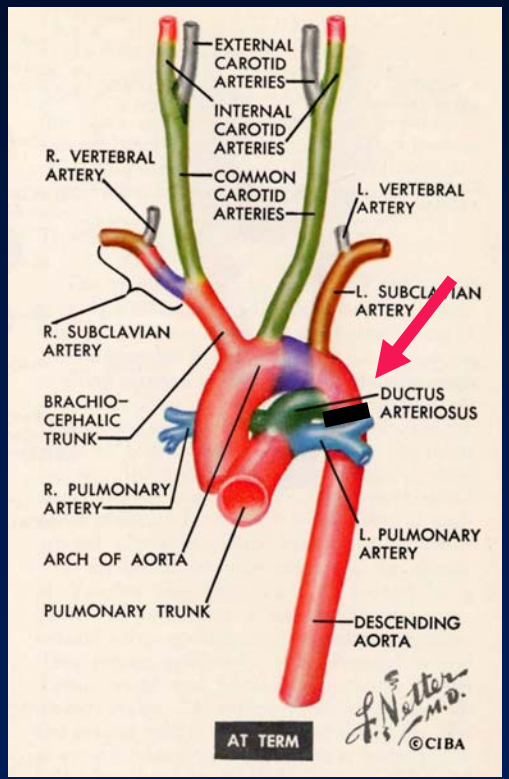
- 2X more common in males
- Common classification
  - Infantile or preductal form
  - Adult or juxtaductal form
- Relationship of ductus to coarct affects clinical picture

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



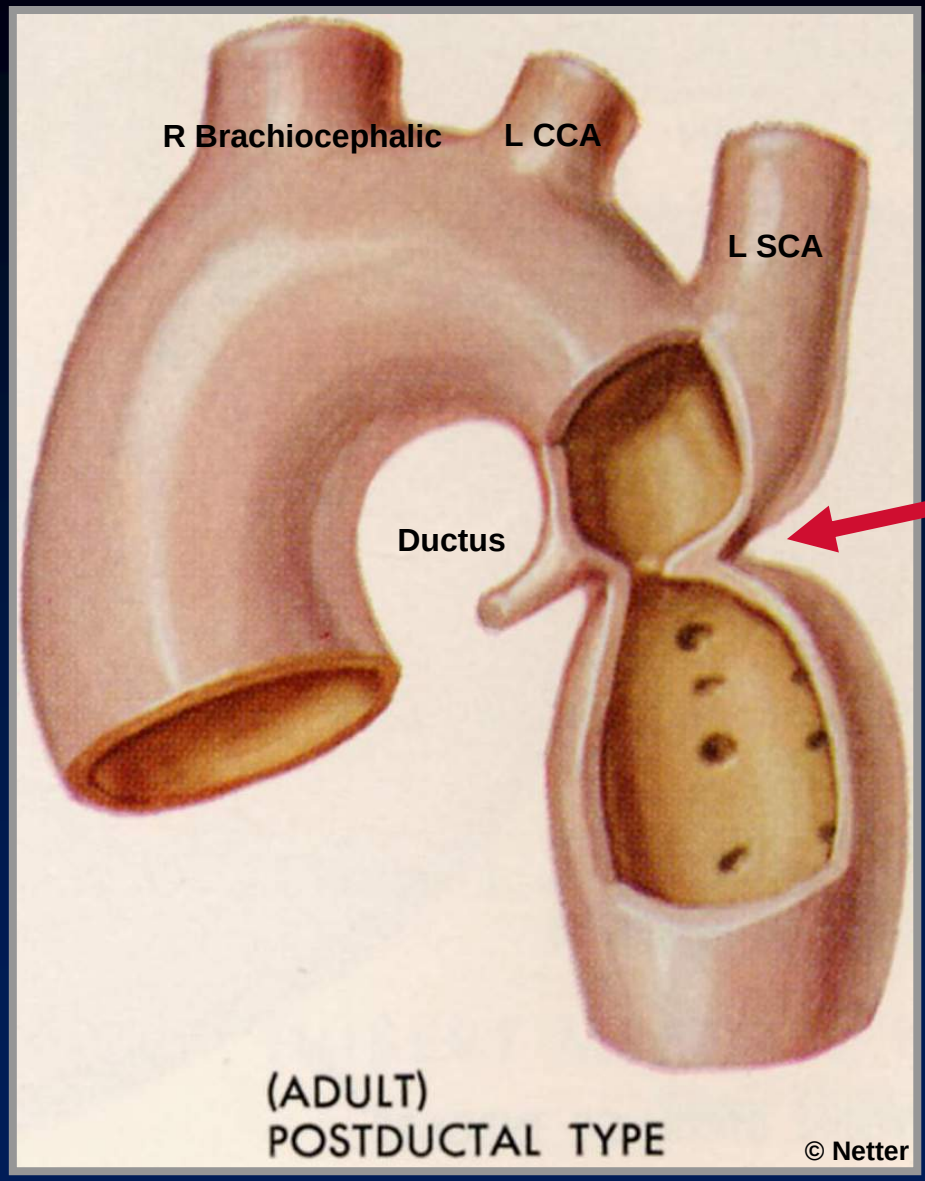
- Flow is initially from Ao to PA (L → R shunt)
- If there is Eisenmenger's physiology, the flow reverses and goes from PA → Ao (R → L shunt)
- Cyanosis
- More common form

Coarctation of the Aorta
Other Classifications

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

Coarctation of the Aorta
Adult Form

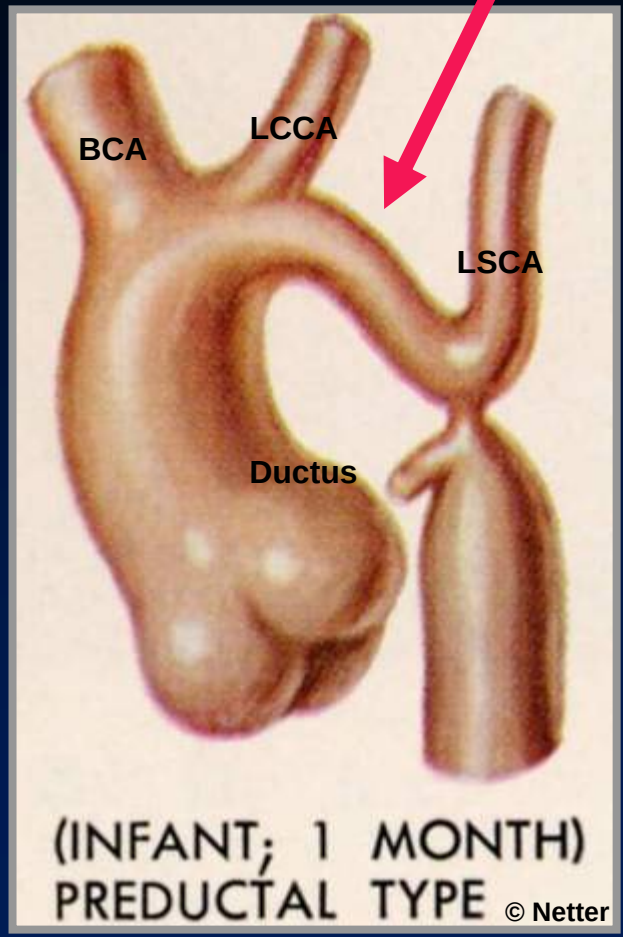
- Adult or juxtaductal (postductal) form is more common than infantile
- Usually localized
- Area of coarctation just beyond origin of LSCA at level of ductus


R Brachiocephalic
L CCA
L SCA
Ductus
(ADULT)
POSTDUCTAL TYPE
© Netter

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 arteriosis


BCA
LCCA
LSCA
Ductus
(INFANT; 1 MONTH)
PREDUCTAL TYPE
© Netter

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

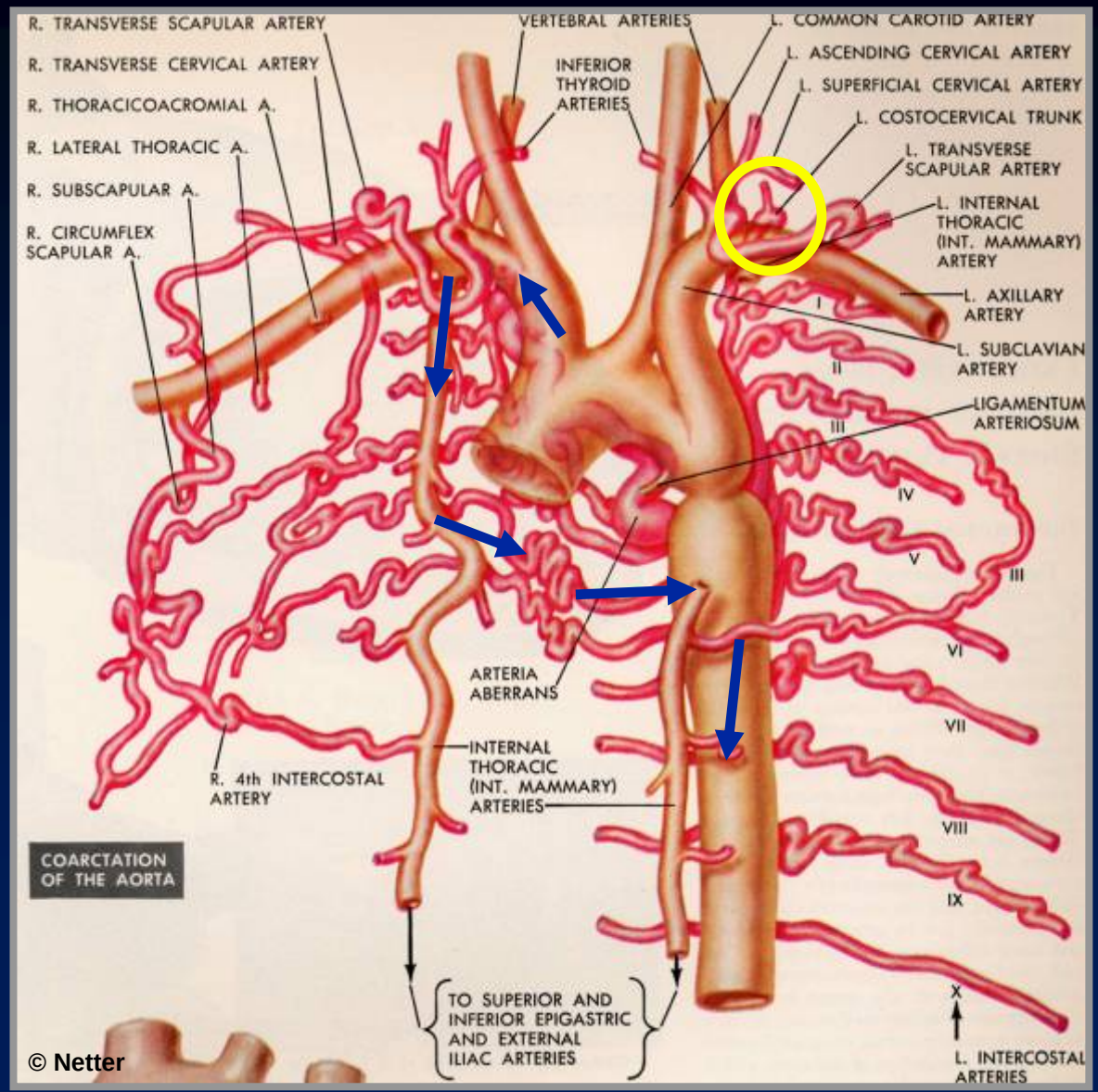
- Single best sign
- Older the person, more likely to have rib notching (uncommon <6 yrs)
- Majority with coarcts display it >20 years of age
- Rib notching occurs in high pressure circuit

Coarctation of the Aorta

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

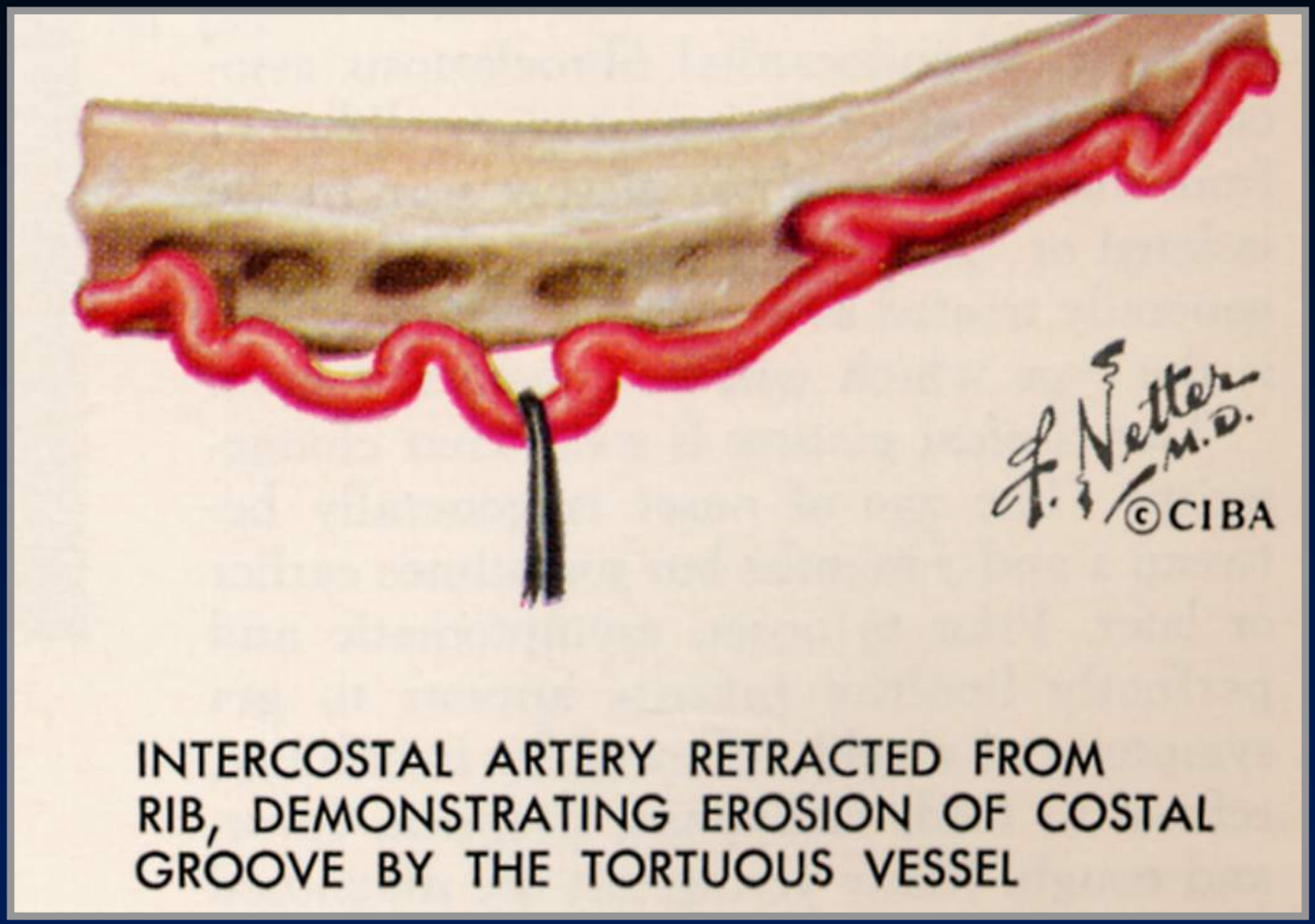
Blood flows from subclavian ➞ internal mammary ➞ intercostals ➞ aorta

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



X-Ray Findings
Rib Notching

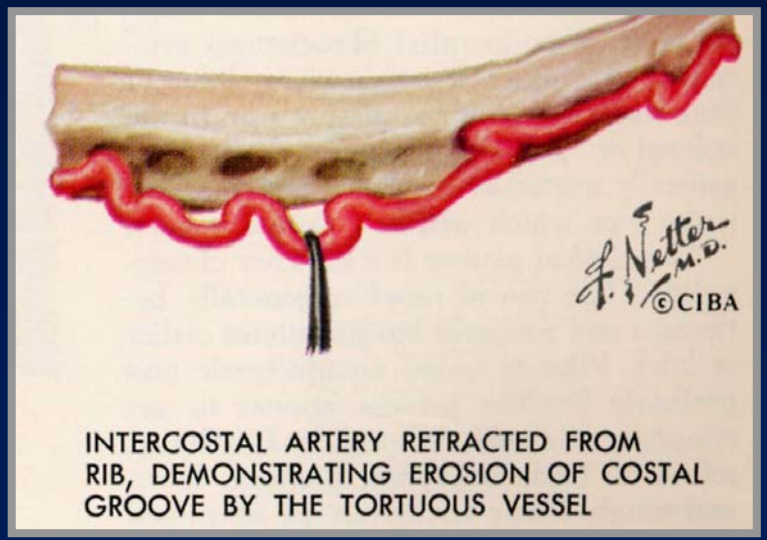
- Most often involves 4th-8th rib
  - Sometimes may involve 3rd and 9th
- Does not involve 1st and 2nd ribs
  - Intercostals come off costocervical trunk and do not supply collateral flow to descending aorta
    - 4th-8th do anastomose with internal mammary to form collaterals for descending aorta

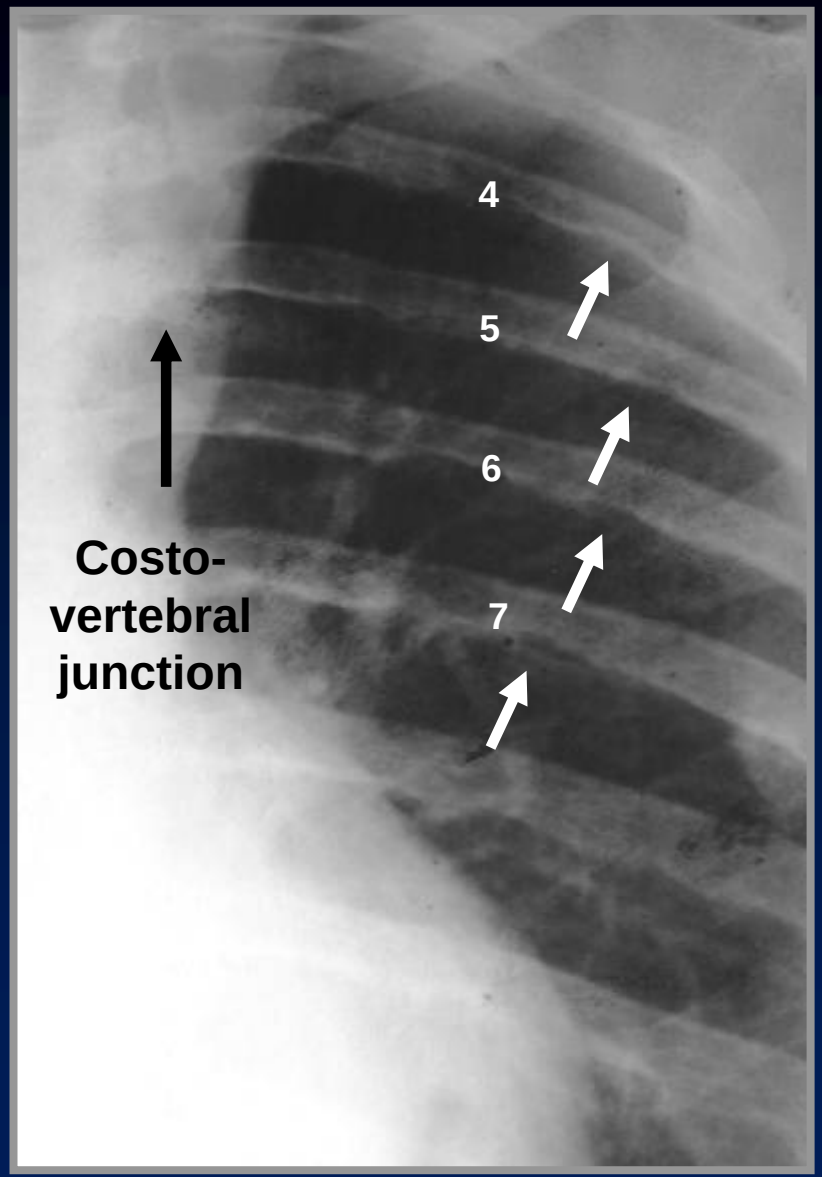


INTERCOSTAL ARTERY RETRACTED FROM RIB, DEMONSTRATING EROSION OF COSTAL GROOVE BY THE TORTUOUS VESSEL

Rib Notching in Coarctation

Regresses after coarct is repaired

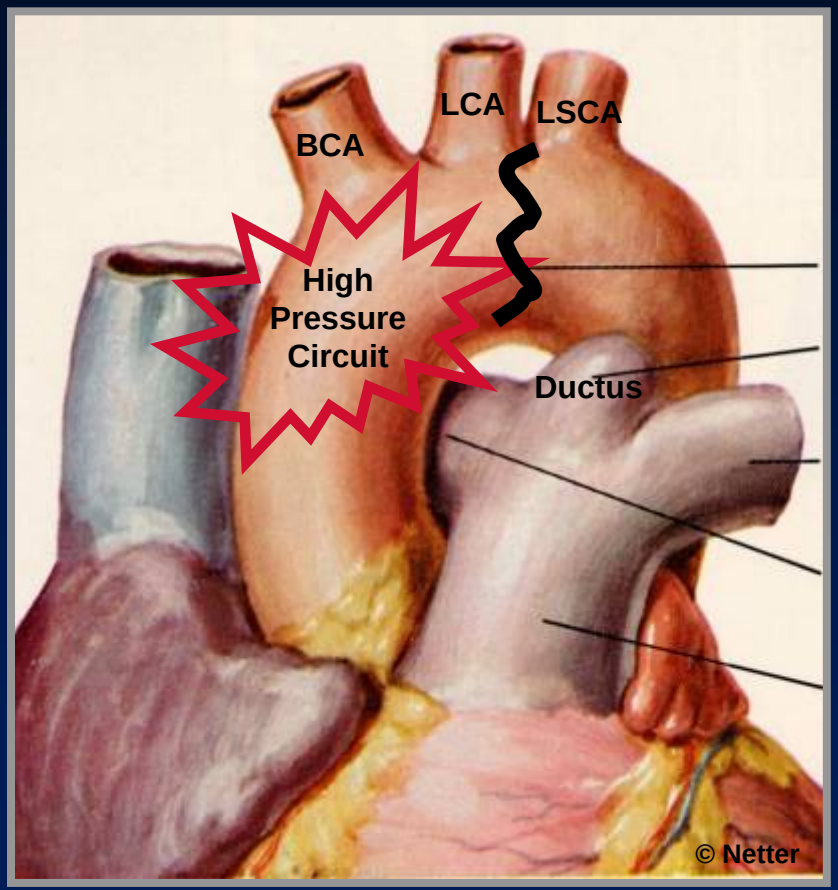




X-Ray Findings
Rib Notching–Unilateral

Rib notching occurs in the high pressure circuit

X-Ray Findings
Unilateral Right Rib Notching

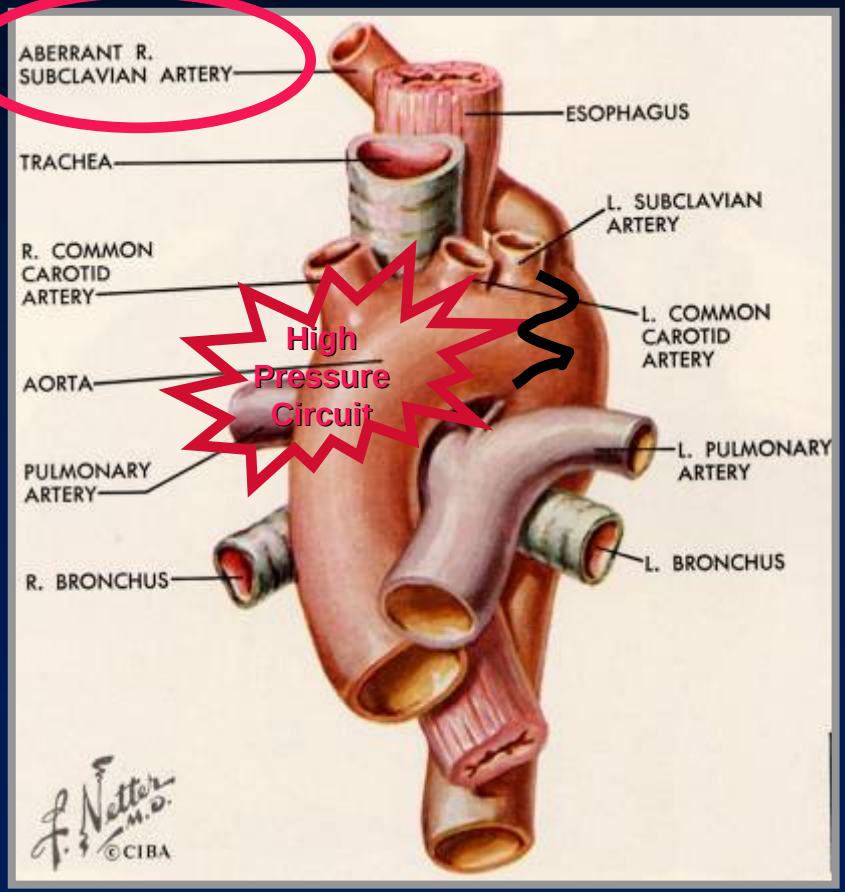


Notching occurs in the high pressure circuit

Isolated right-sided rib notching

Coarct originates between the LCCA and the LSCA

X-Ray Findings
Unilateral Left Rib Notching

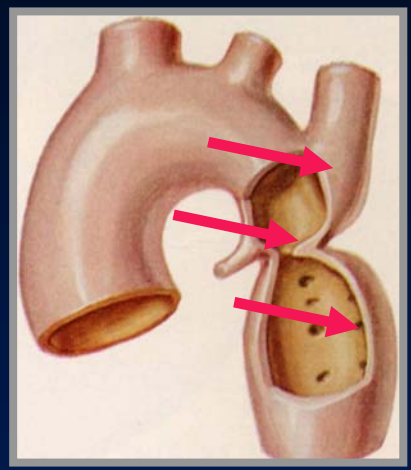


Notching occurs in the high pressure circuit

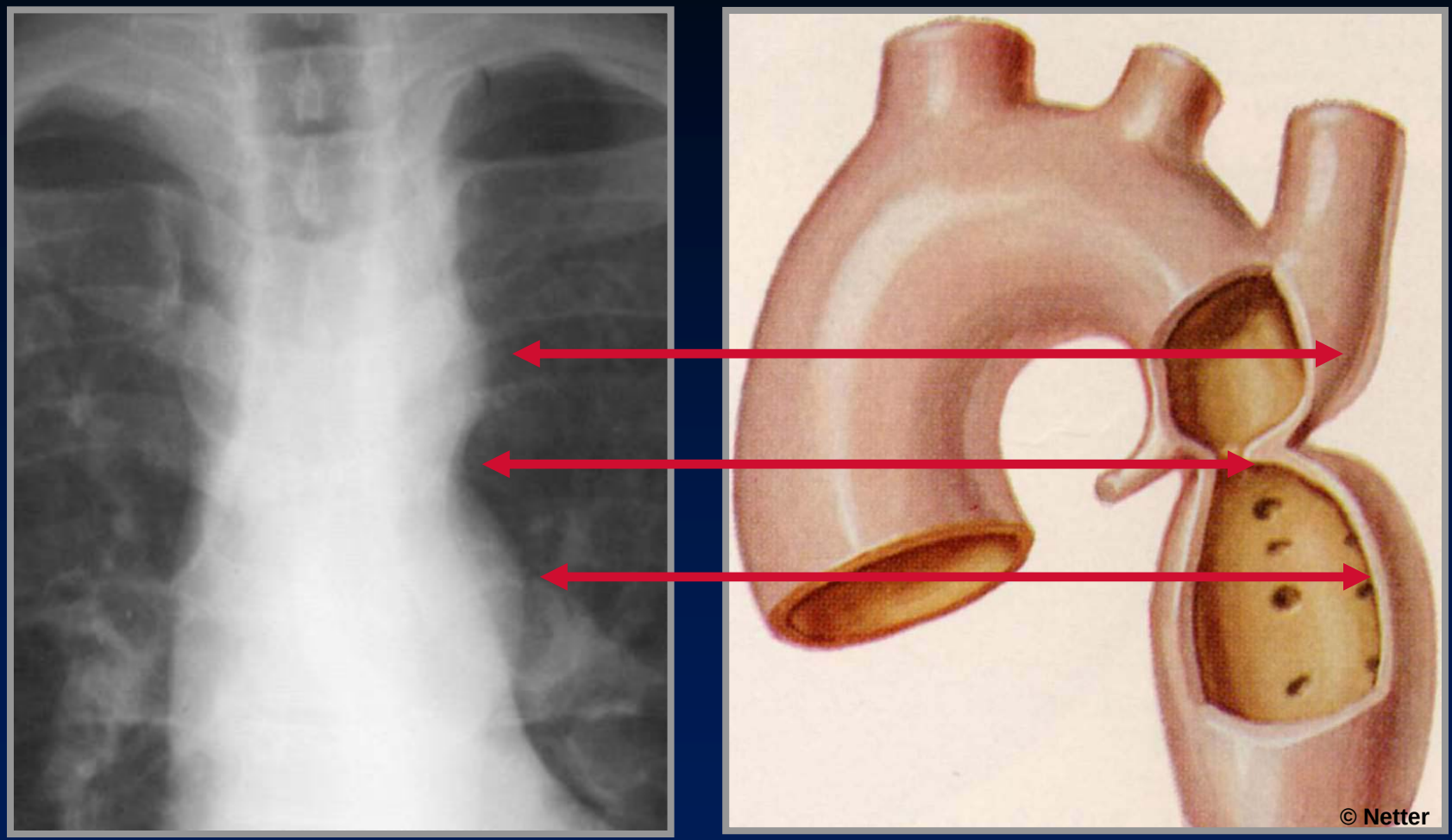
Isolated left- sided rib notching

Anomalous RSCA originates distal to site of coarct

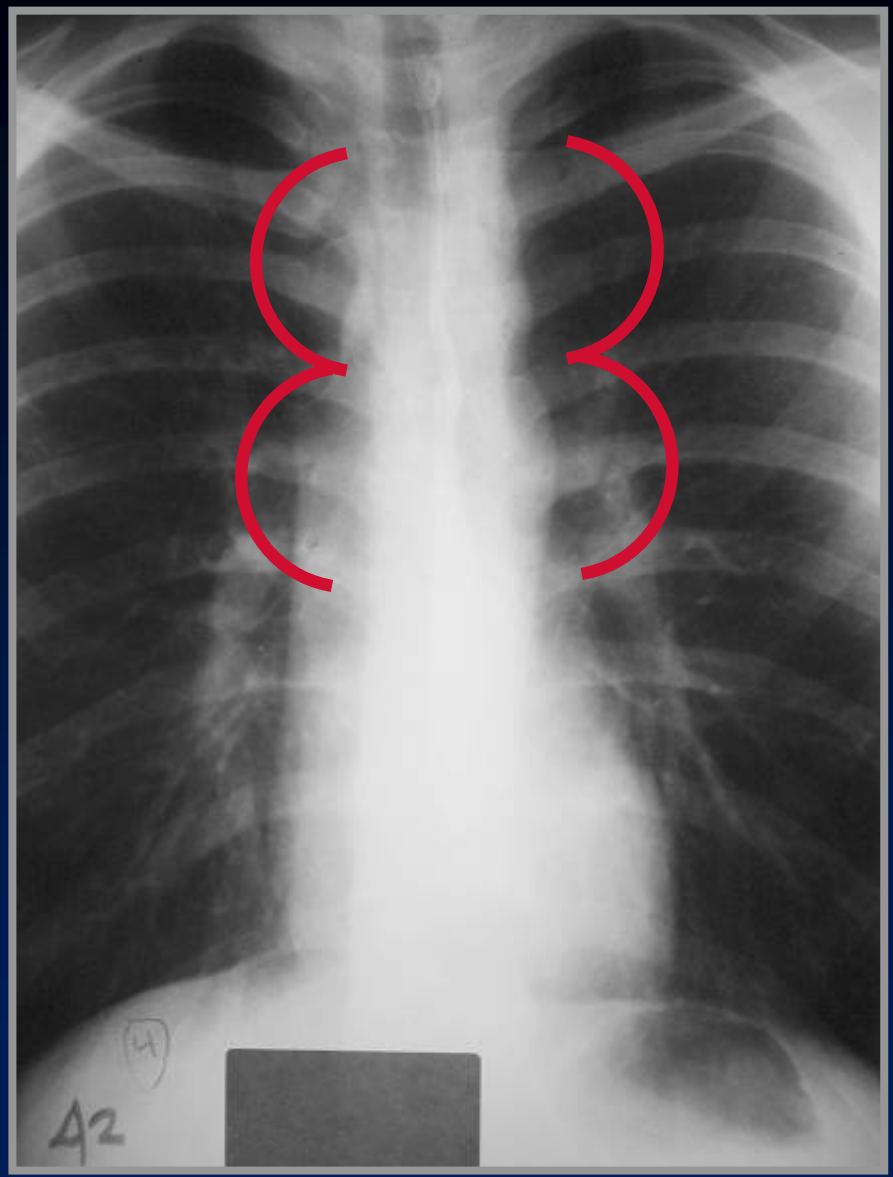
X-Ray Findings
Figure 3 Sign



- 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


© Netter

Reverse 3 sign on barium filled esophagus



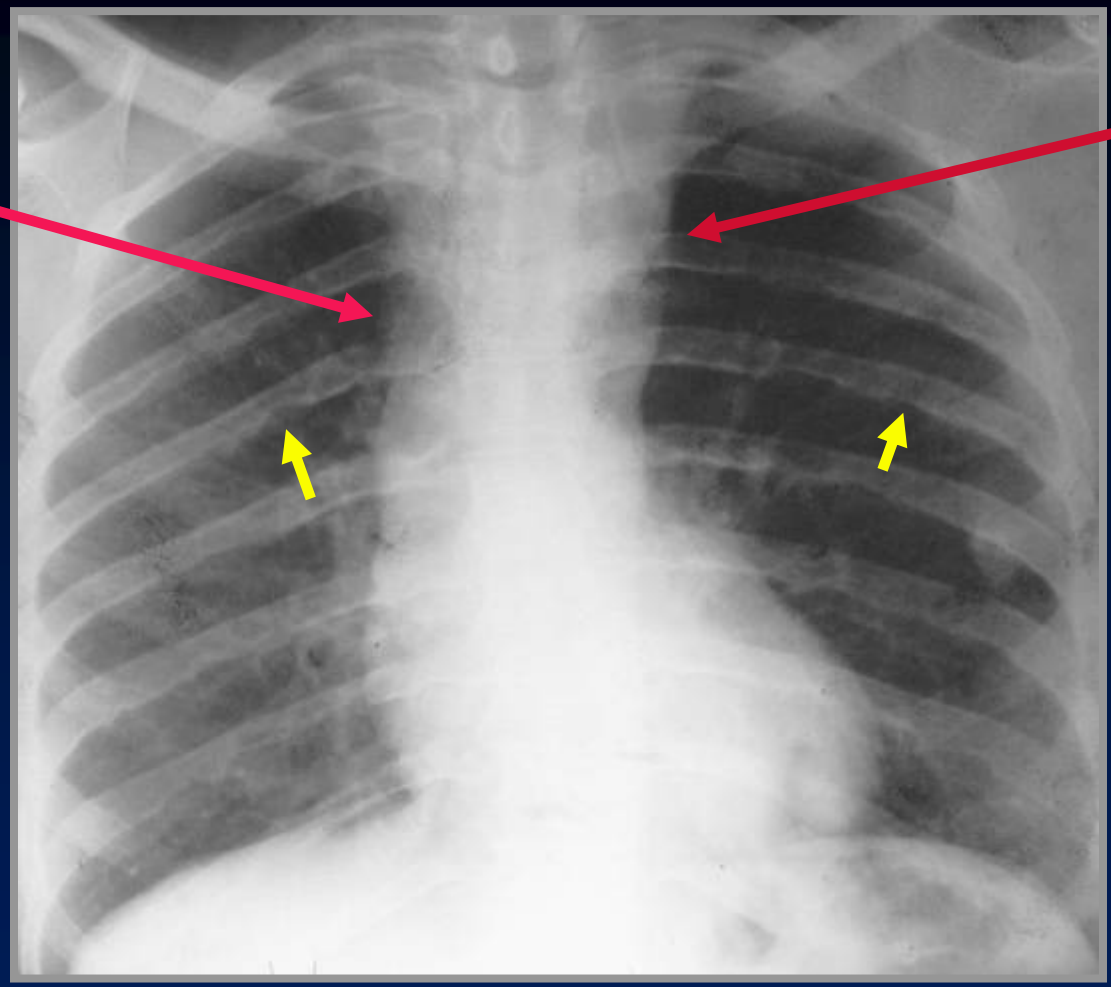
“Figure 3 sign” caused by coarctation

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

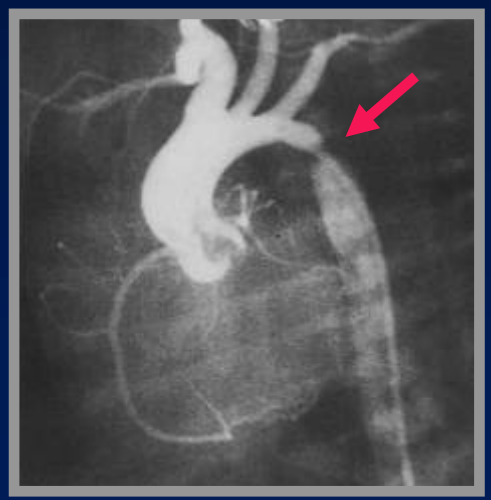
- Severe CHF most common from 2nd to 6th week of life
- Weak or absent leg pulses
- Lower BP in the legs than in the arms
- EKG: RV hypertrophy because RV assumes most of the cardiac output during fetal life in these patients

Coarctation of the Aorta
Echocardiographic Findings

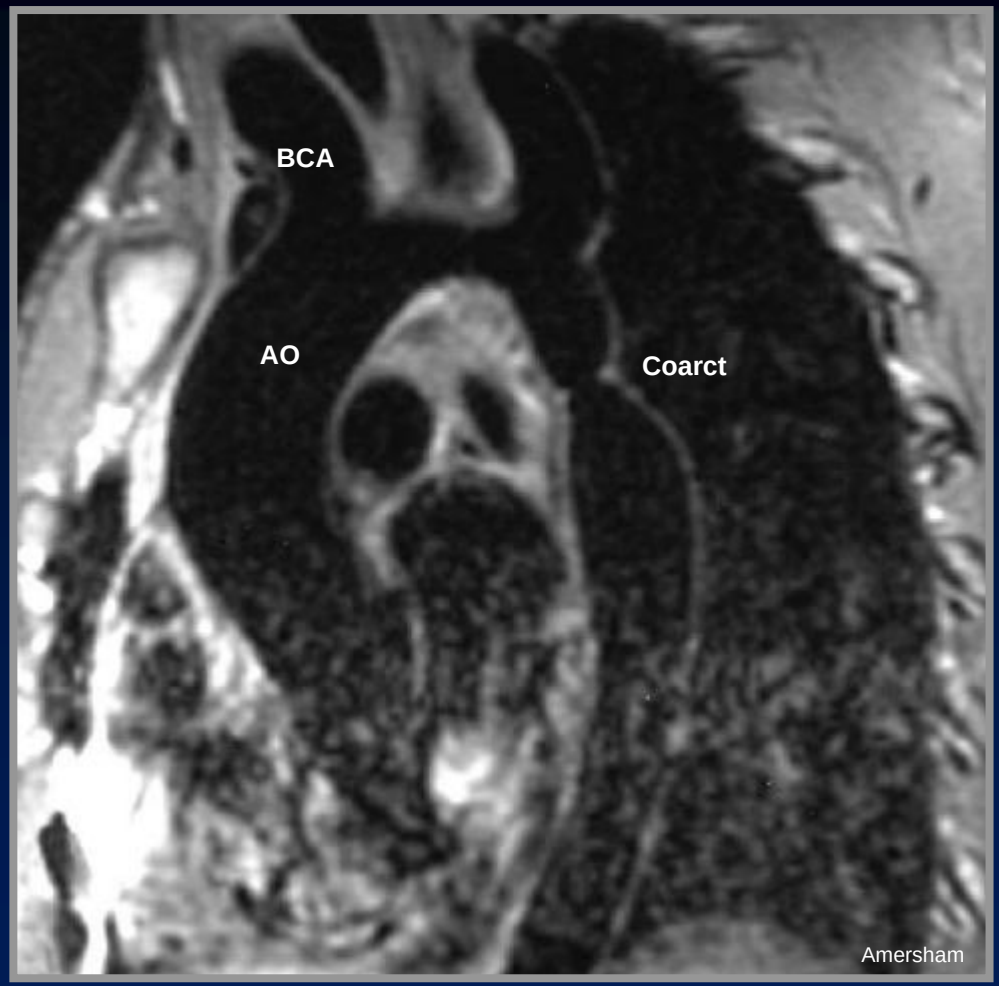
- In infants, 2D echo can demonstrate coarcts from suprasternal notch
- Echo helpful in excluding associated hypoplastic left heart syndrome

Coarctation of the Aorta
MRI and Angiography

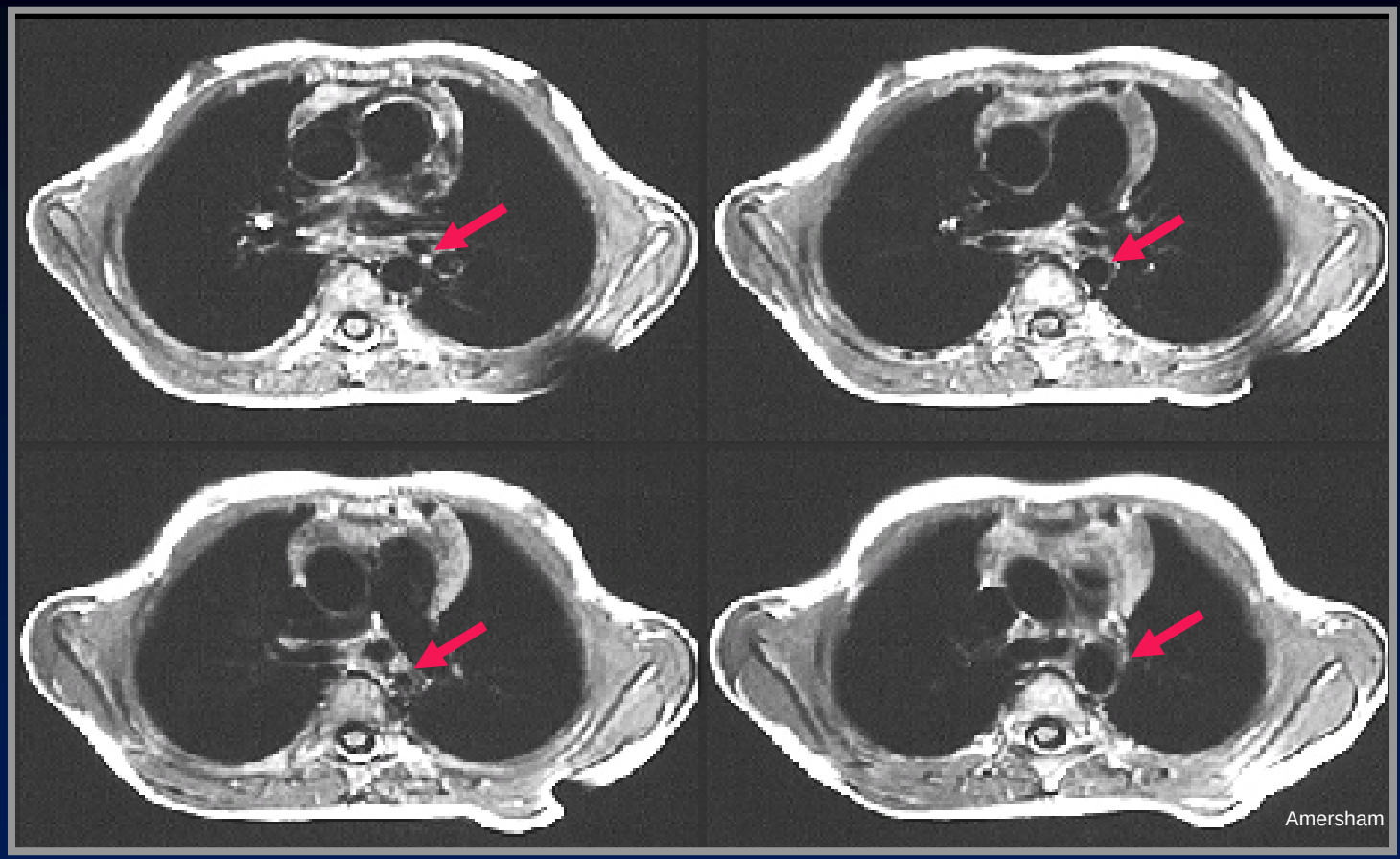
- MRI preferred study in children/adults
- Aortography offers greatest resolution



Contrast enhanced MRA shows long segment coarctation of the aorta



Oblique sagittal spin-echo-Coarctation of the Aorta



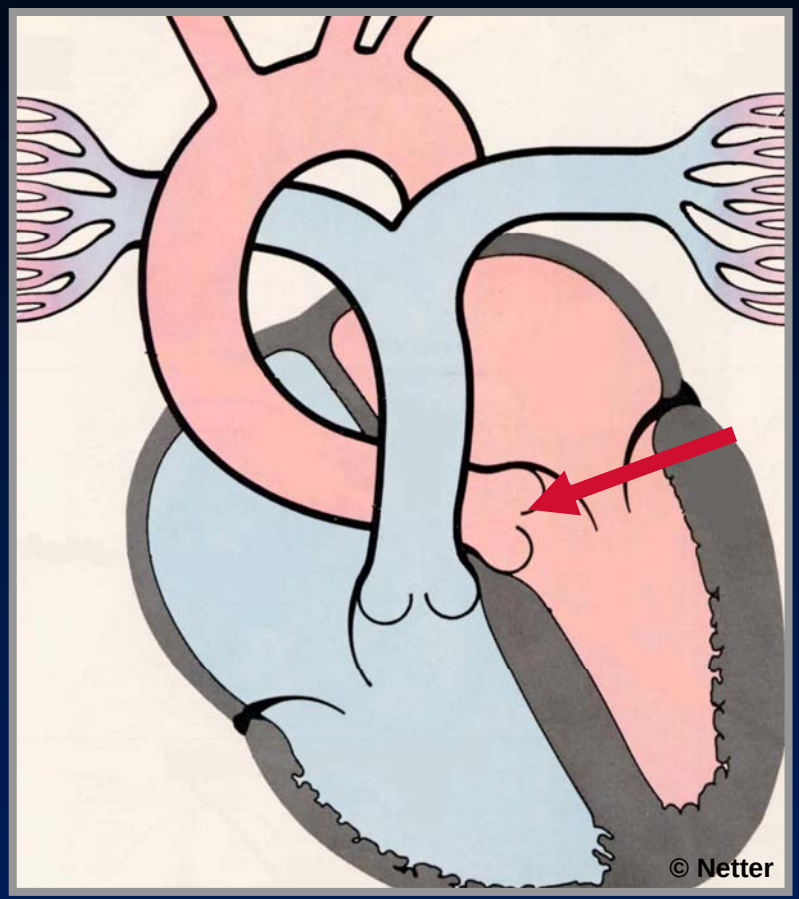
Axial spin-echo MRI-Coarctation of the Aorta

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 (<30mmHg)
- Figure 3 sign present
- No rib notching



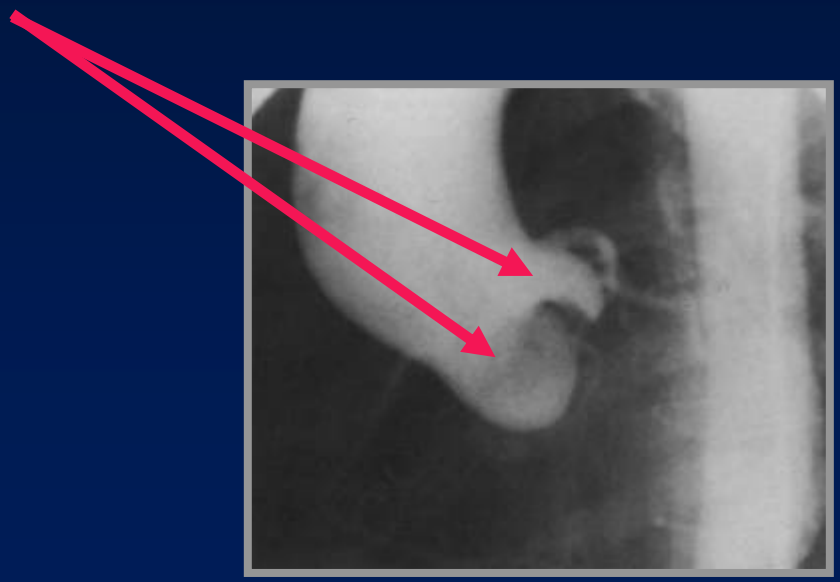
Congenital Aortic Stenosis

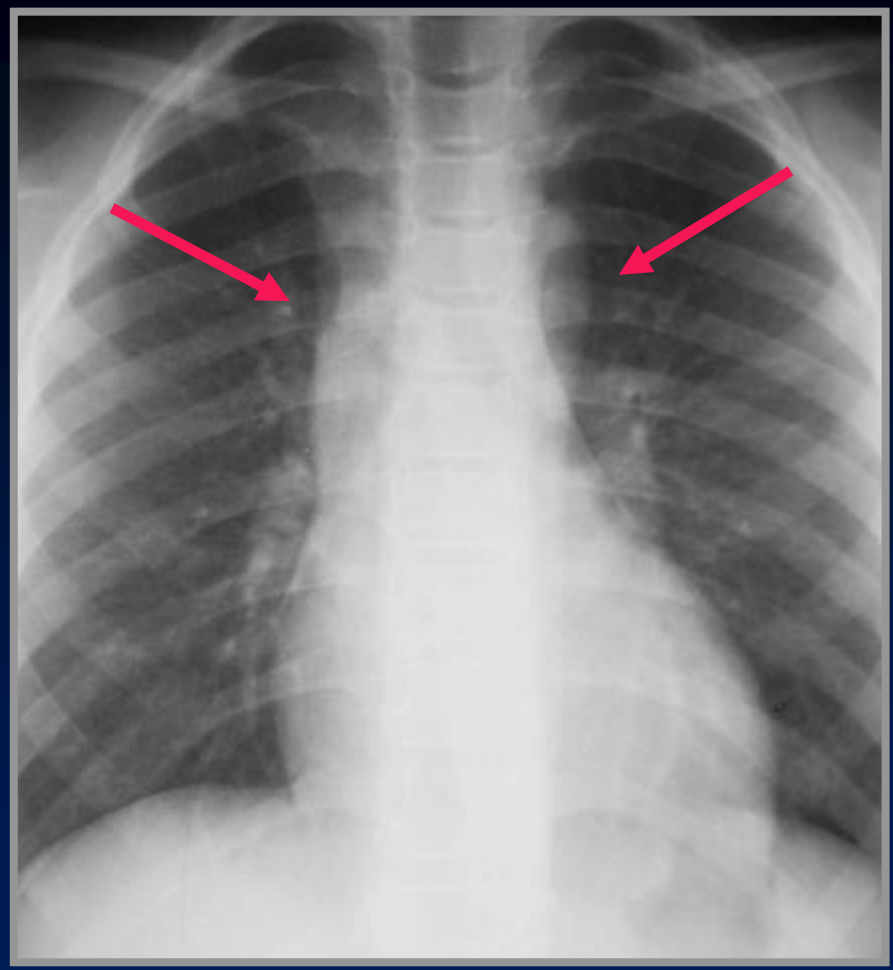
Congenital Aortic Stenosis
Valvular-General

- 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

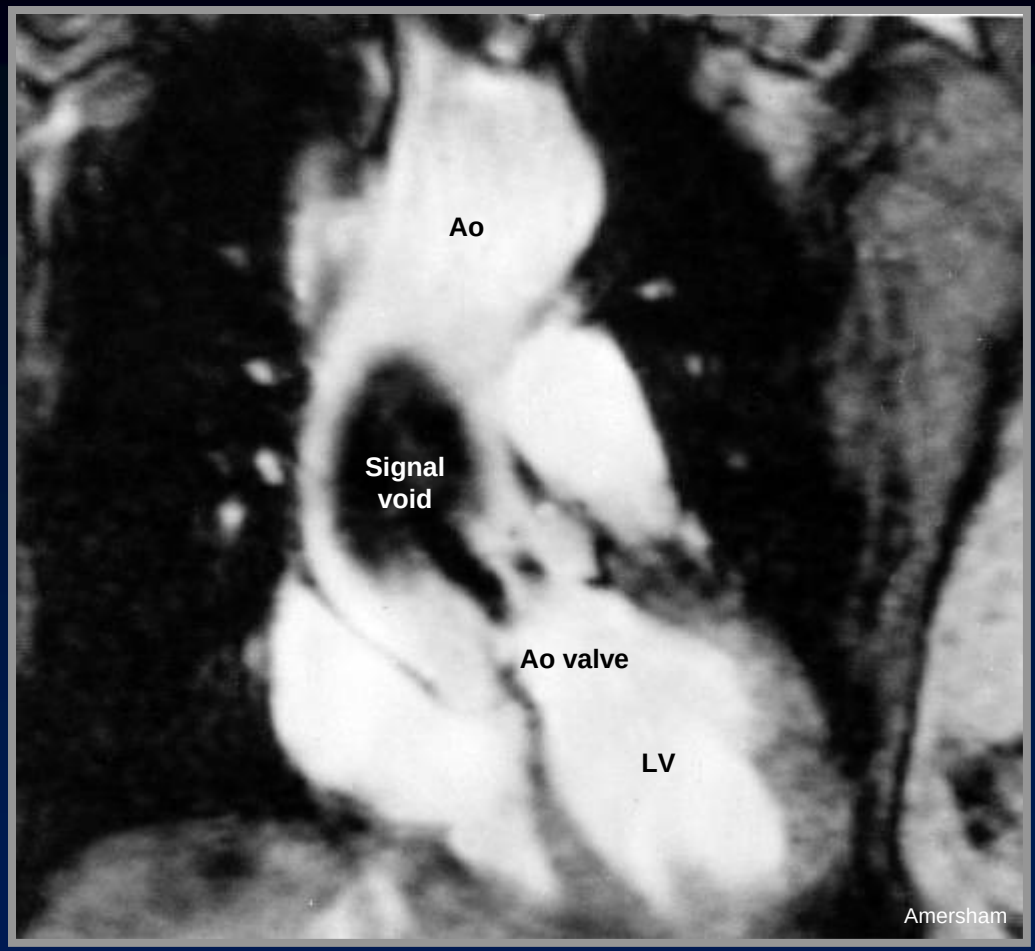
Congenital Aortic Stenosis
Angiography

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



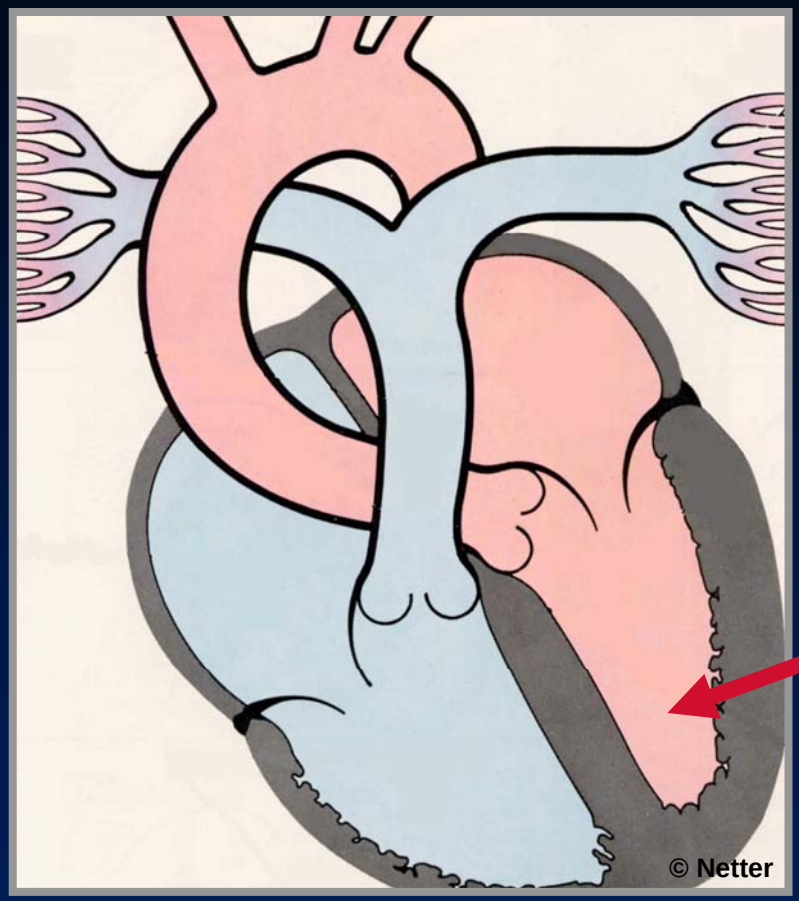


Congenital Aortic Stenosis (10 yo)



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

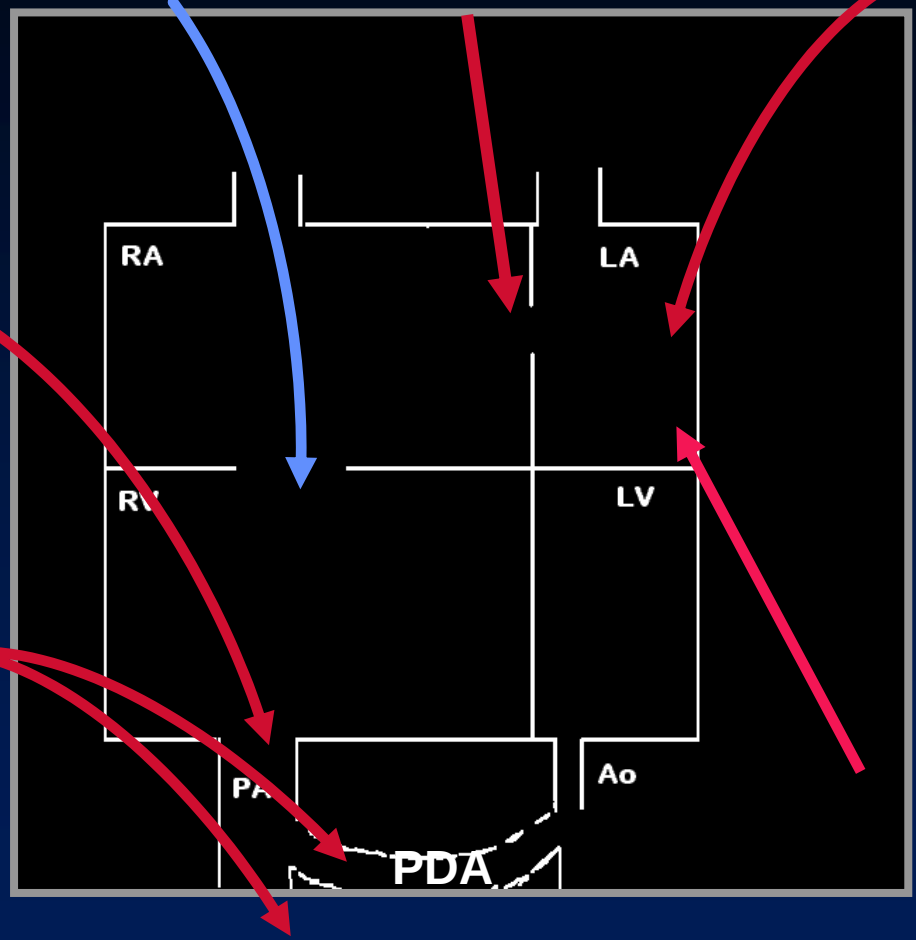
- Small ascending aorta
  - Common to all forms
  - Sometimes infantile coarctation
- Often associated mitral stenosis or atresia or aortic stenosis or atresia
- In 90%, size of LA and LV small
- A large PDA is essential
  - VSD, ASD also present

RA, RV and PA are enlarged

Passes to RA via ASD (L → R shunt)

Oxygenated blood returning from lungs can not enter LV

Oxygenated and deoxygenated blood enter PA



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

Obstruction to return of blood from lungs → CHF

Hypoplastic Left Heart Syndrome

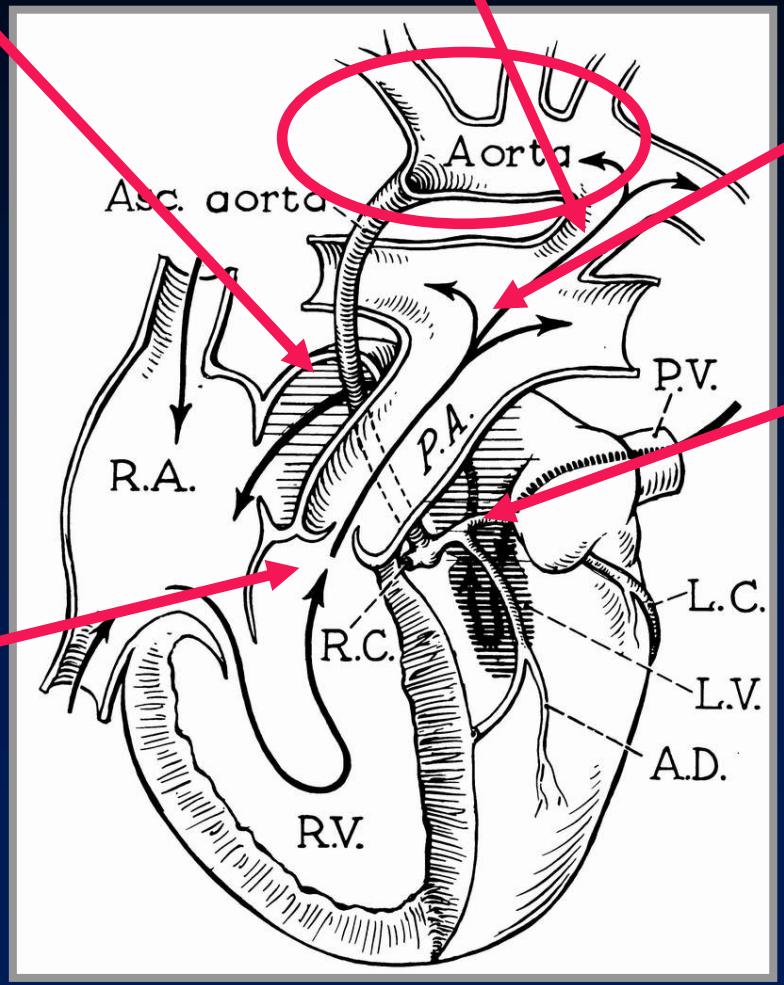
Cyanotic

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

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

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



Some deoxygenated blood goes to lungs

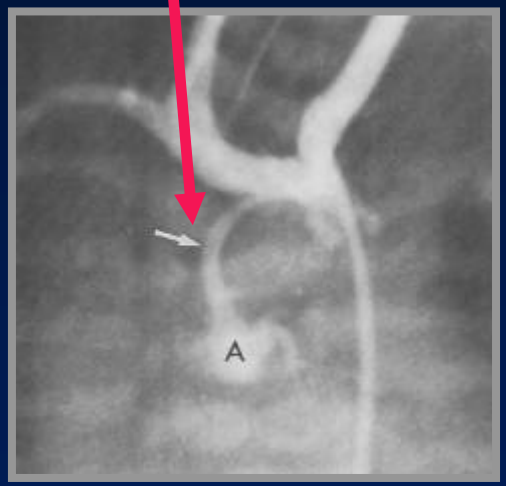
Blood returning from body mixes with oxygenated blood from LA; passes into PA

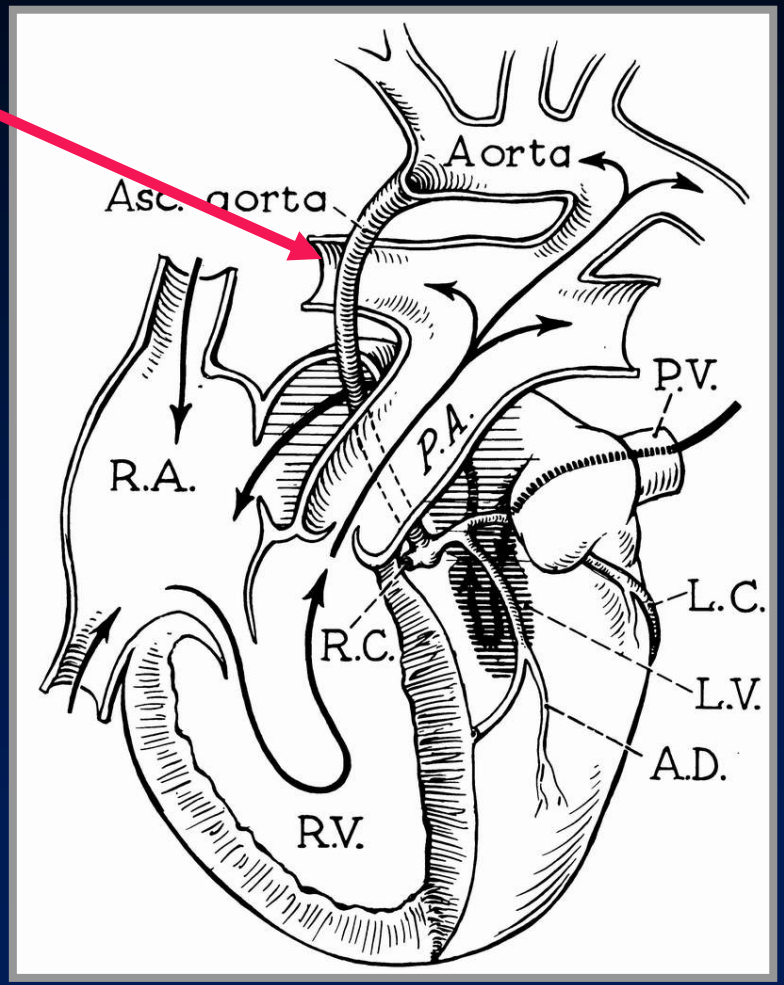
Blood returning from lungs can not exit LA to LV because of atretic mitral valve

Hypoplastic Left Heart Syndrome

Hypoplastic Left Heart Syndrome

Atretic aorta



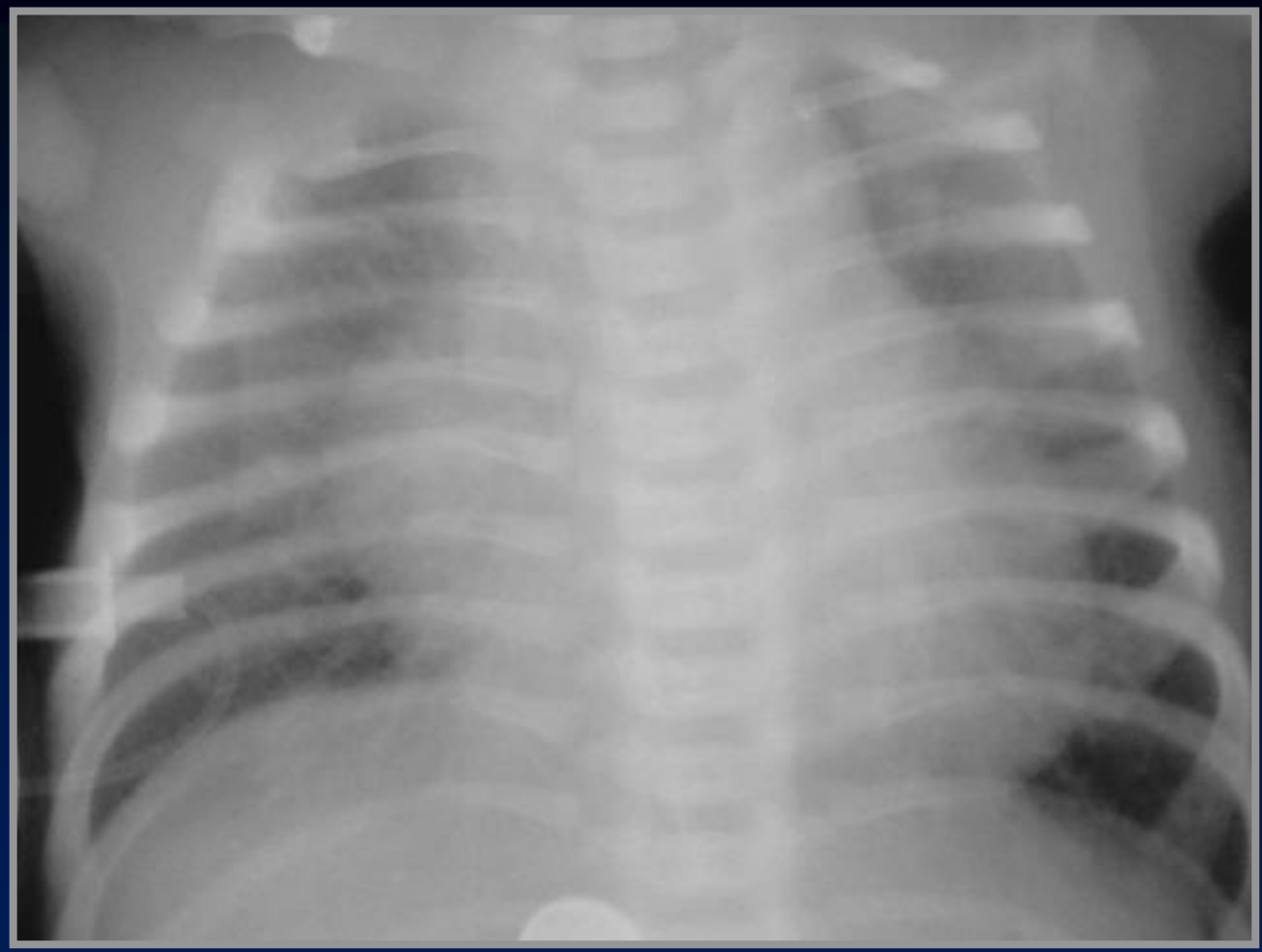


Hypoplastic Left Heart Syndrome
Associated Anomalies

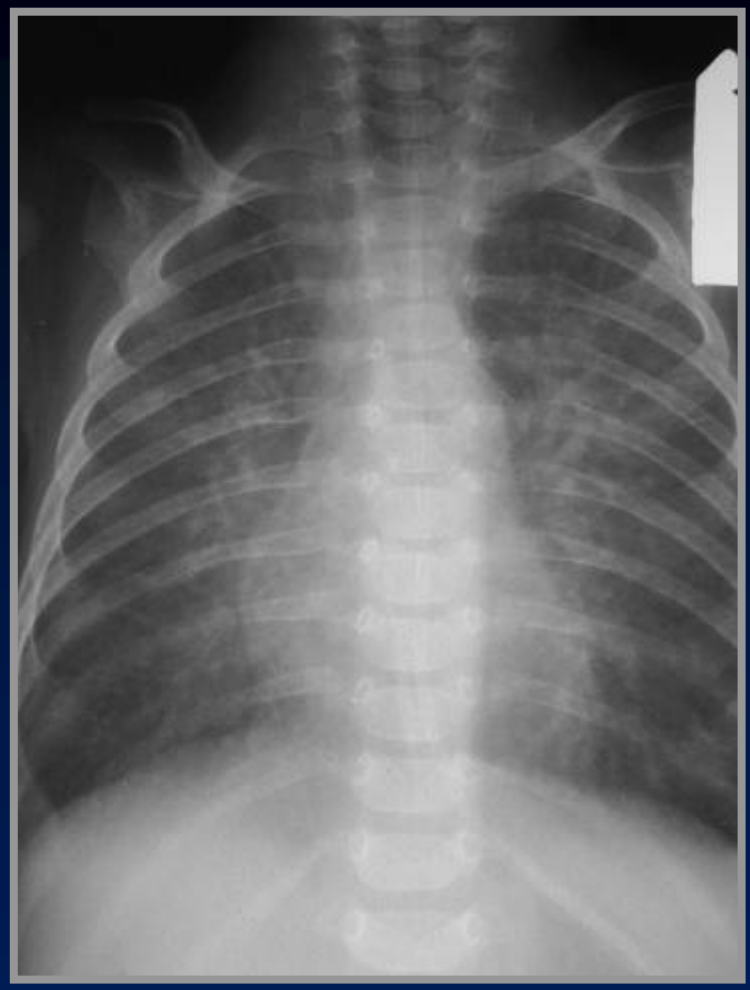
- Coarctation of the aorta
- Interruption of the aortic arch
- AV communis
- Anomalies of the R subclavian artery
- Bicuspid aortic valve

Hypoplastic Left Heart Syndrome
X-ray Findings

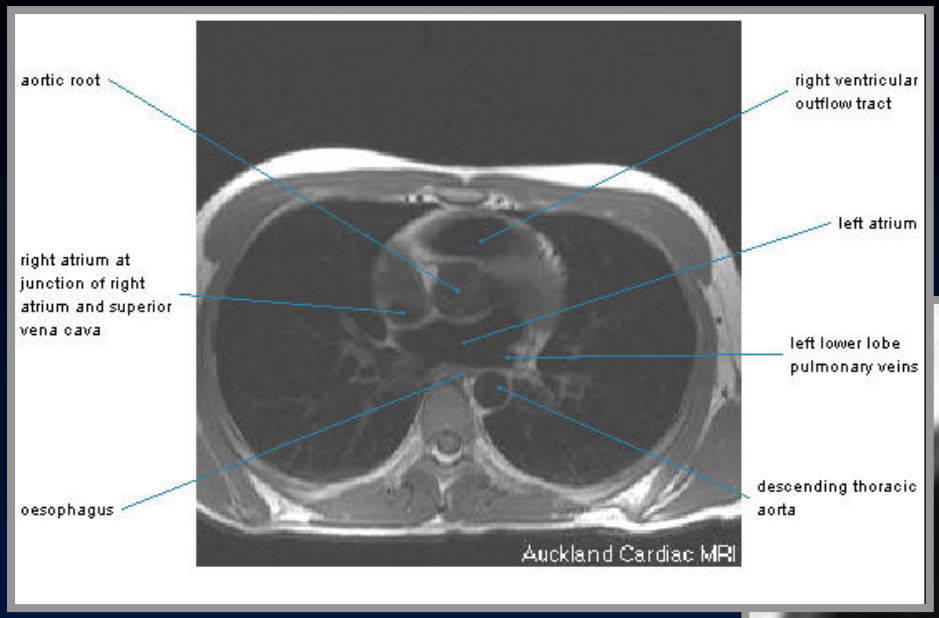
- Increased load on RV → marked cardiomegaly at birth
- Obstruction to return of blood from lungs → CHF at birth
  - Most common cause of CHF in first two weeks of life

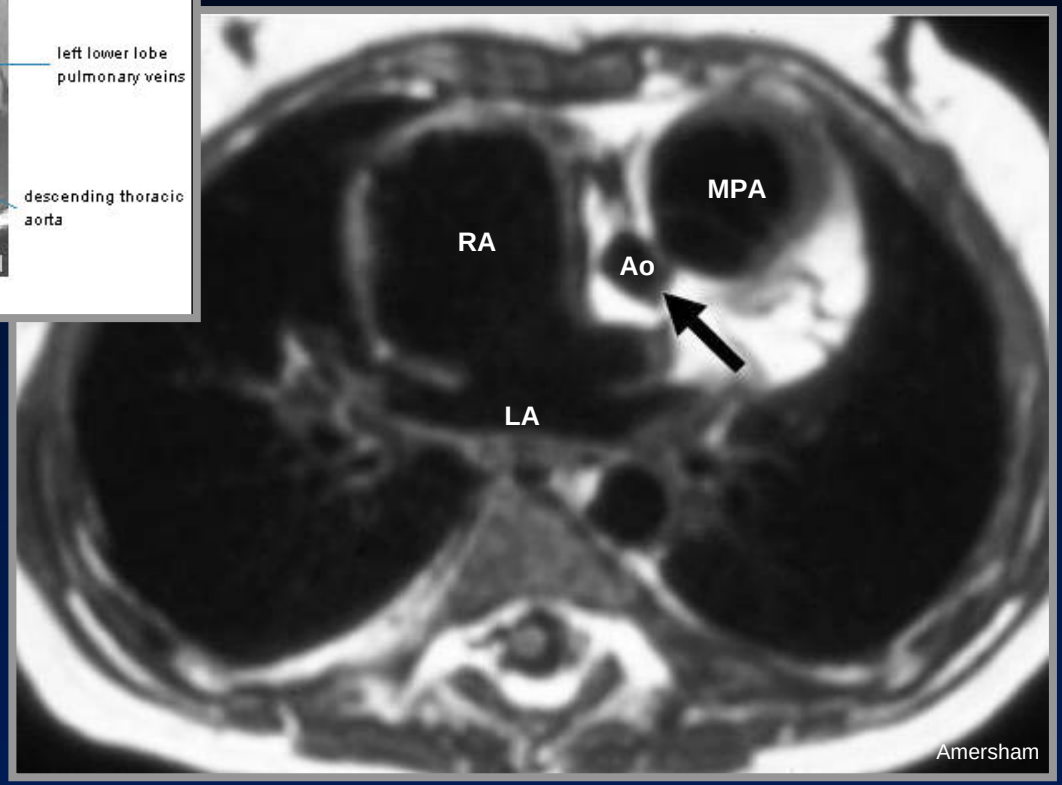


Hypoplastic Left Heart Syndrome



Hypoplastic Left Heart Syndrome





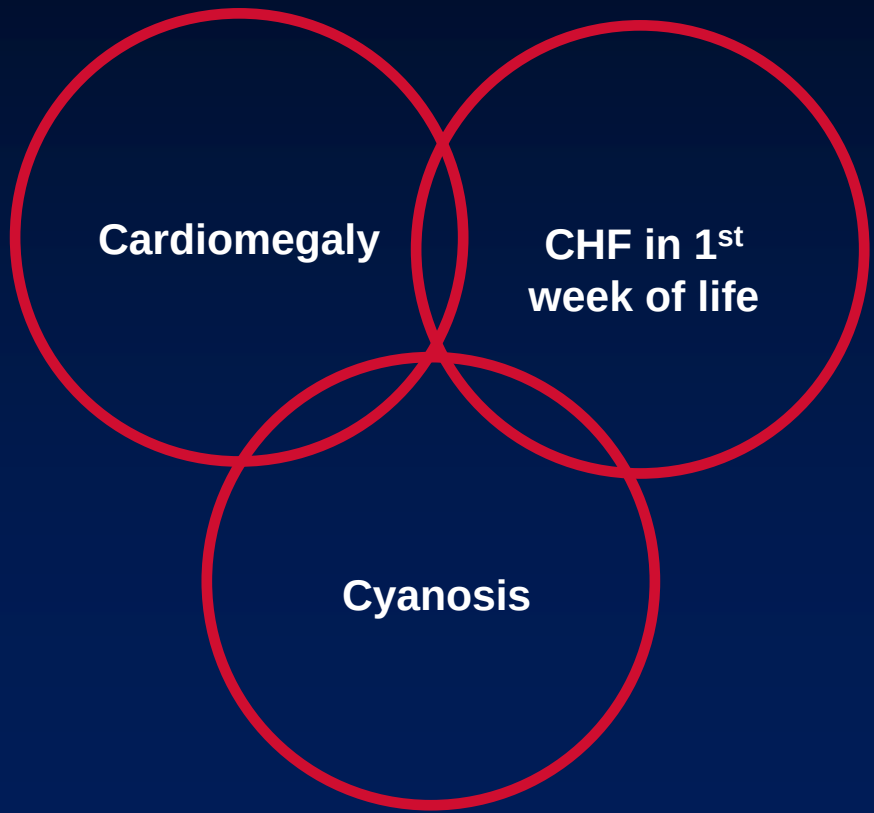
Hypoplastic Left heart Syndrome

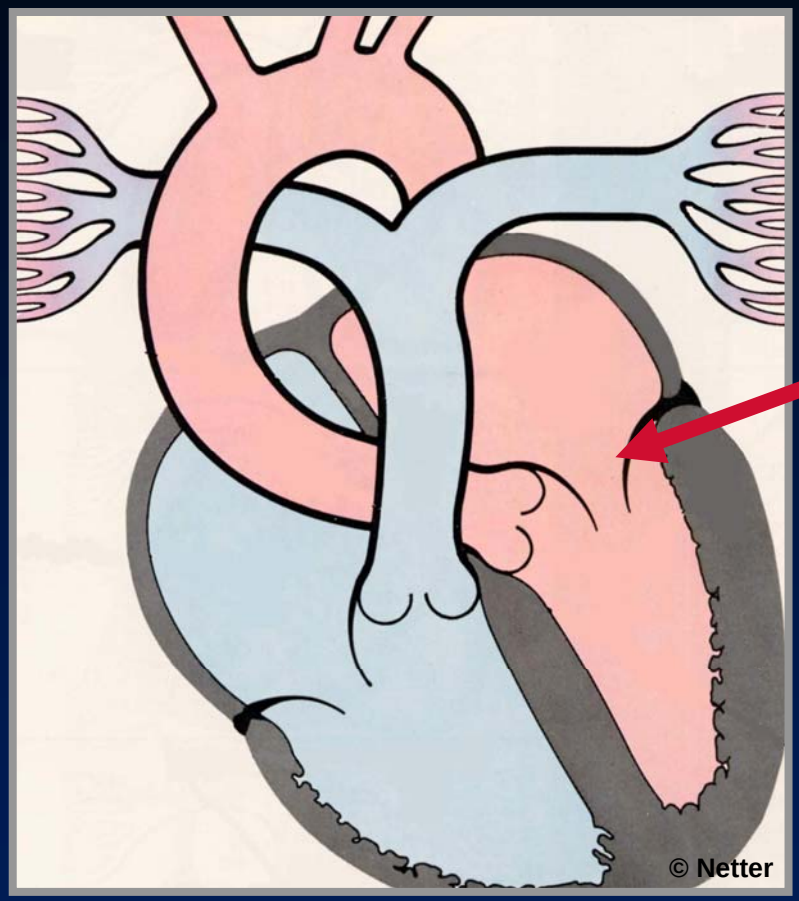
Gated spin echo at base of heart shows hypoplastic aorta (arrow) posterior and right of main pulmonary artery

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





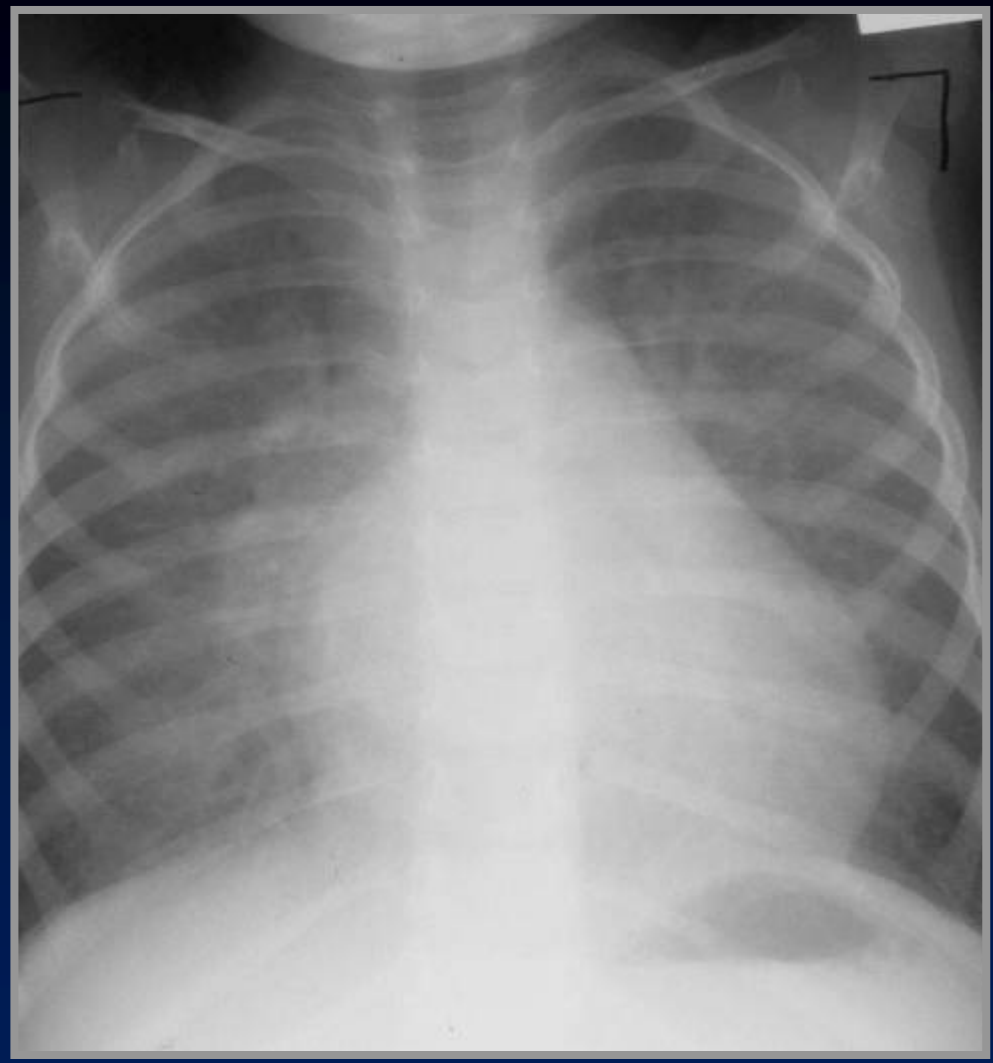
Congenital Mitral Stenosis

Congenital Mitral Stenosis

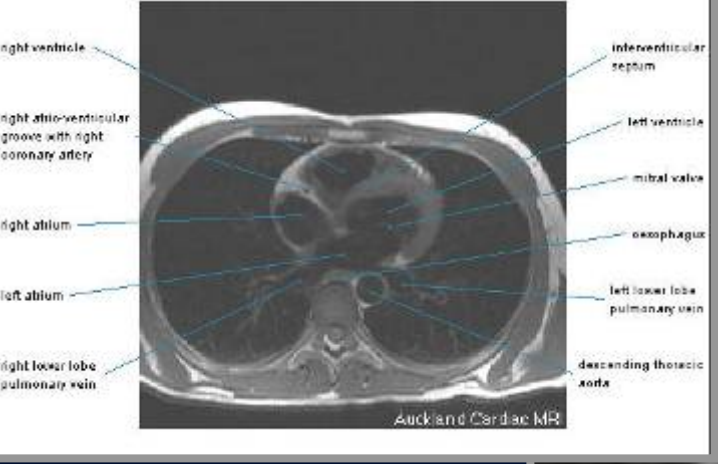
- Exists as isolated abnormality 25% of time
- Coexists with VSD 30% of time
- Coexists with another form of left ventricular outflow obstruction 40% of time—SHONE'S Syndrome

Shone's Syndrome

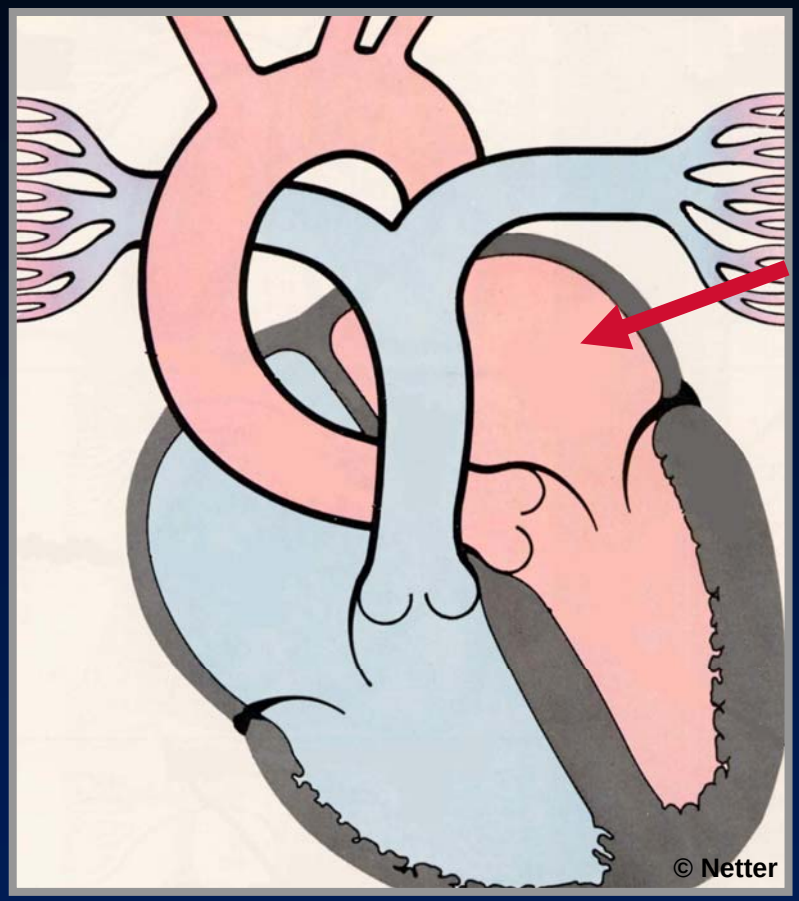
- Parachute mitral valve
- Supravalvular mitral ring
- Subaortic stenosis
- Coarctation of aorta



Congenital Mitral Stenosis



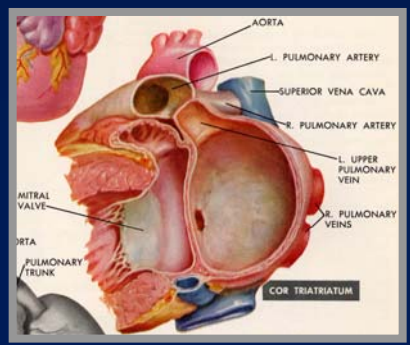
Cine MR image in axial plane demonstrates a diastolic signal void emanating from the mitral valve indicative of mitral stenosis



Cor Triatriatum

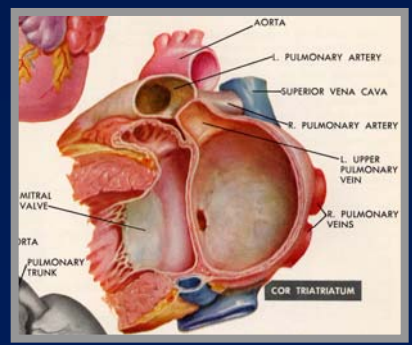
Cor Triatriatum
General

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



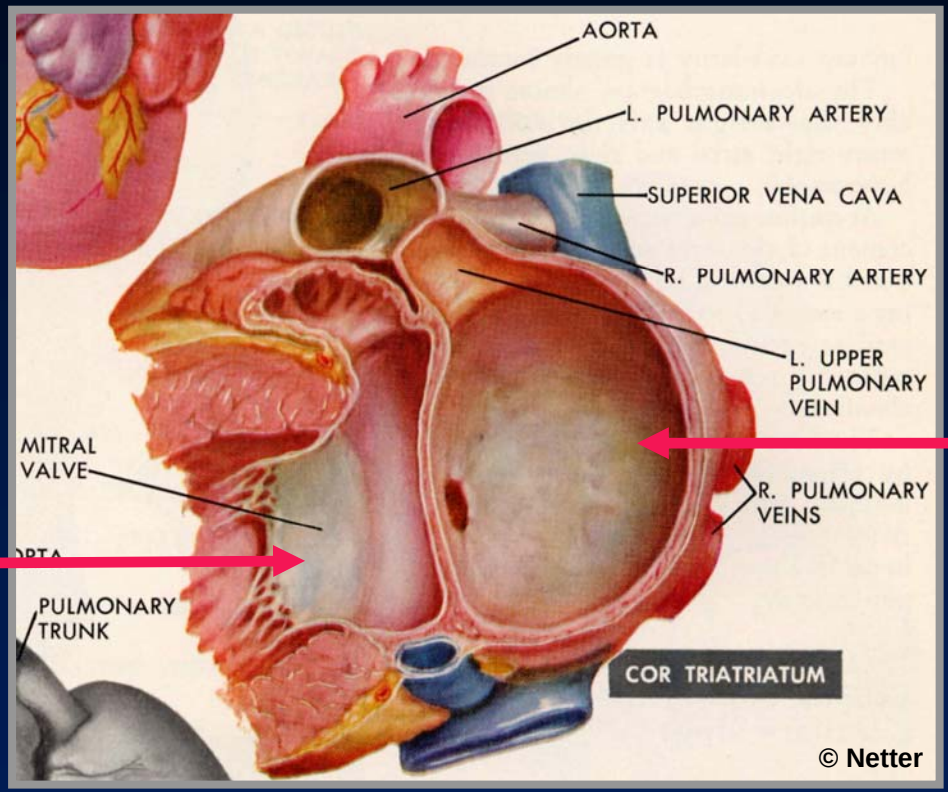
Cor Triatriatum
Anatomy

- 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

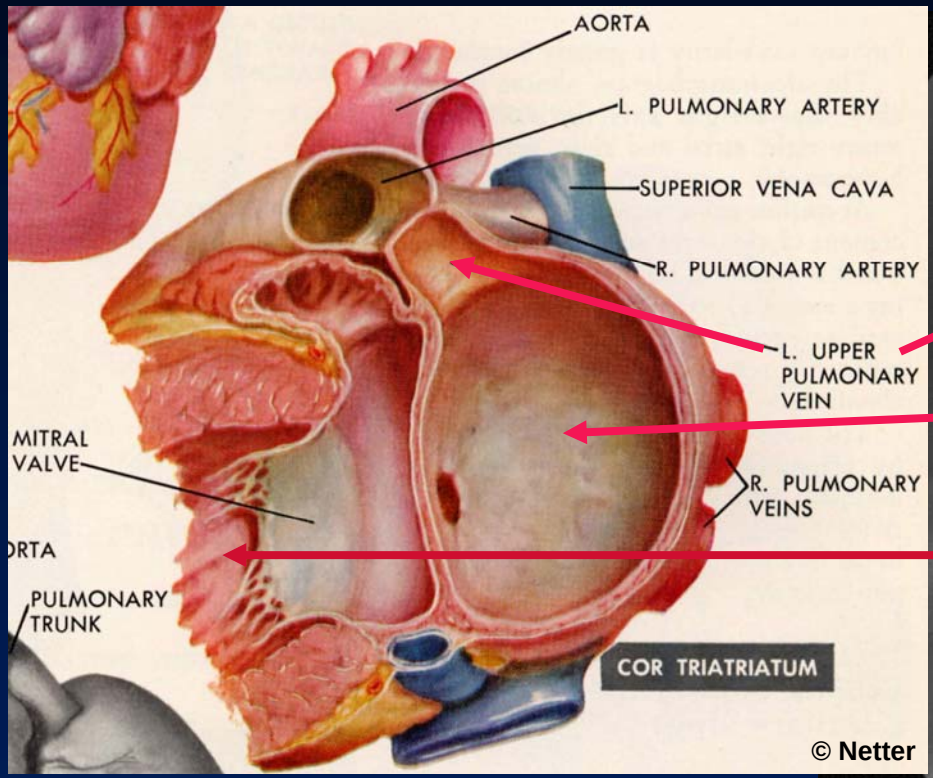
- Clinically similar to mitral stenosis
- Dyspnea
- Heart failure
- Failure to thrive

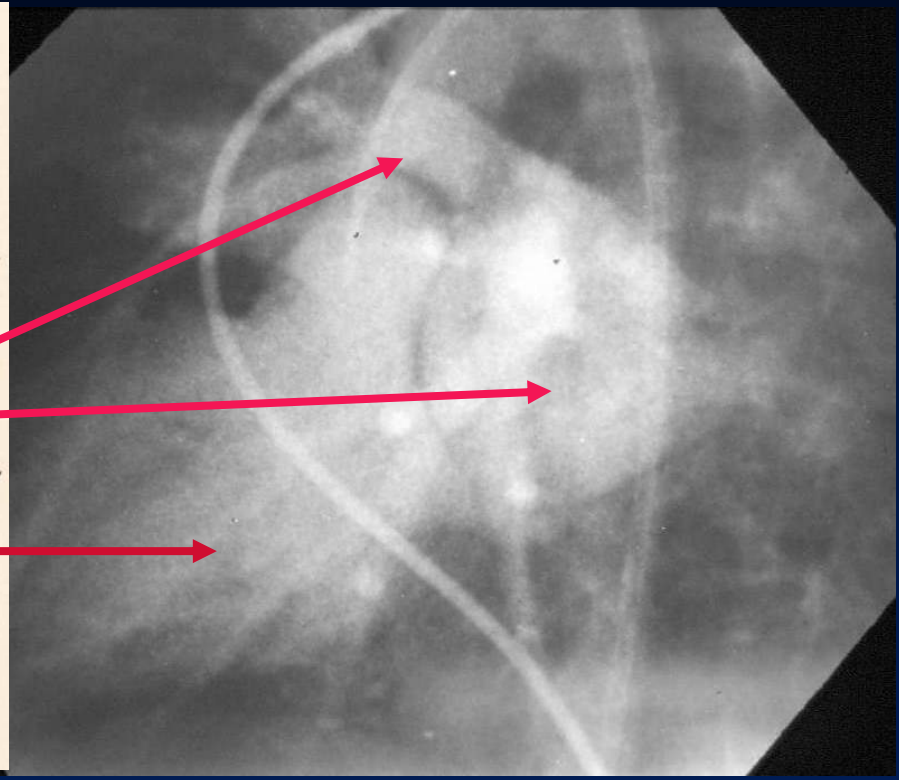
Cor Triatriatum
X-ray Findings

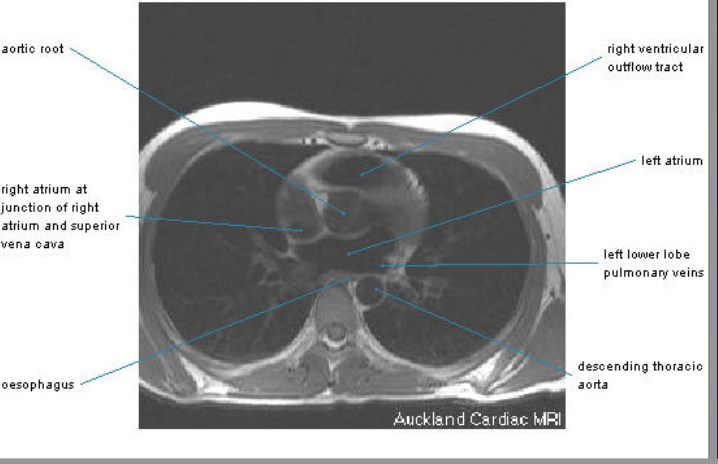
- Pulmonary edema
- Enlarged LA

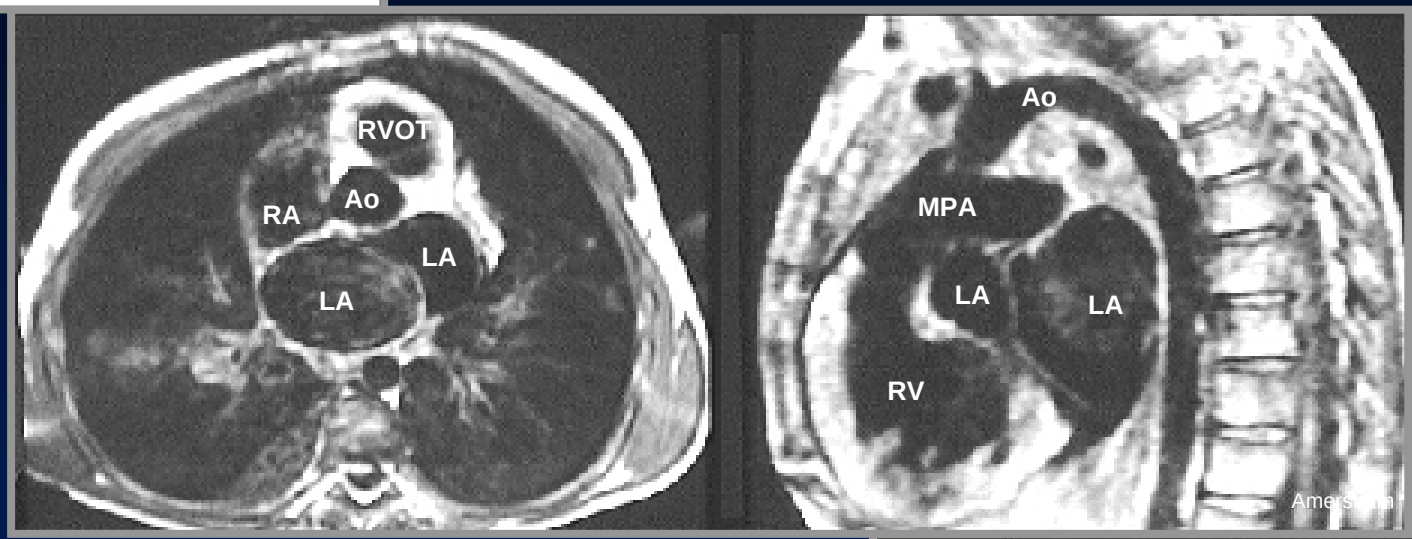
Cor Triatriatum

Cor Triatriatum - angiography









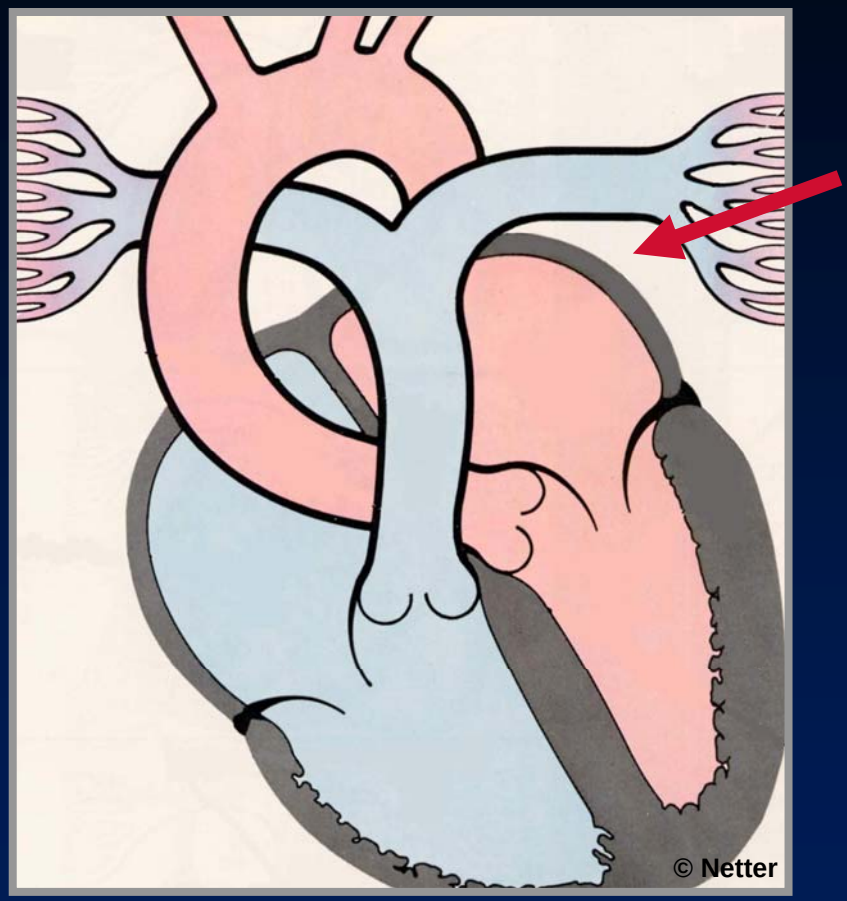
Cor Triatriatum

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

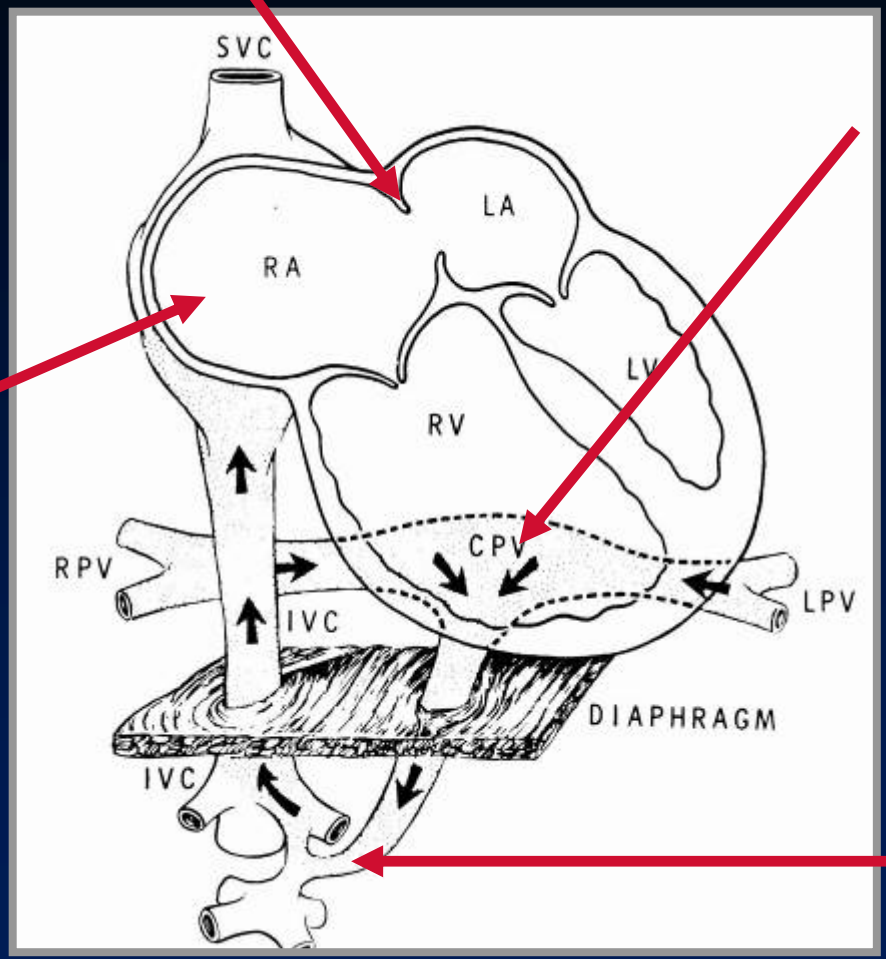
TAPVR
Infracardiac Type—Type III

- 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

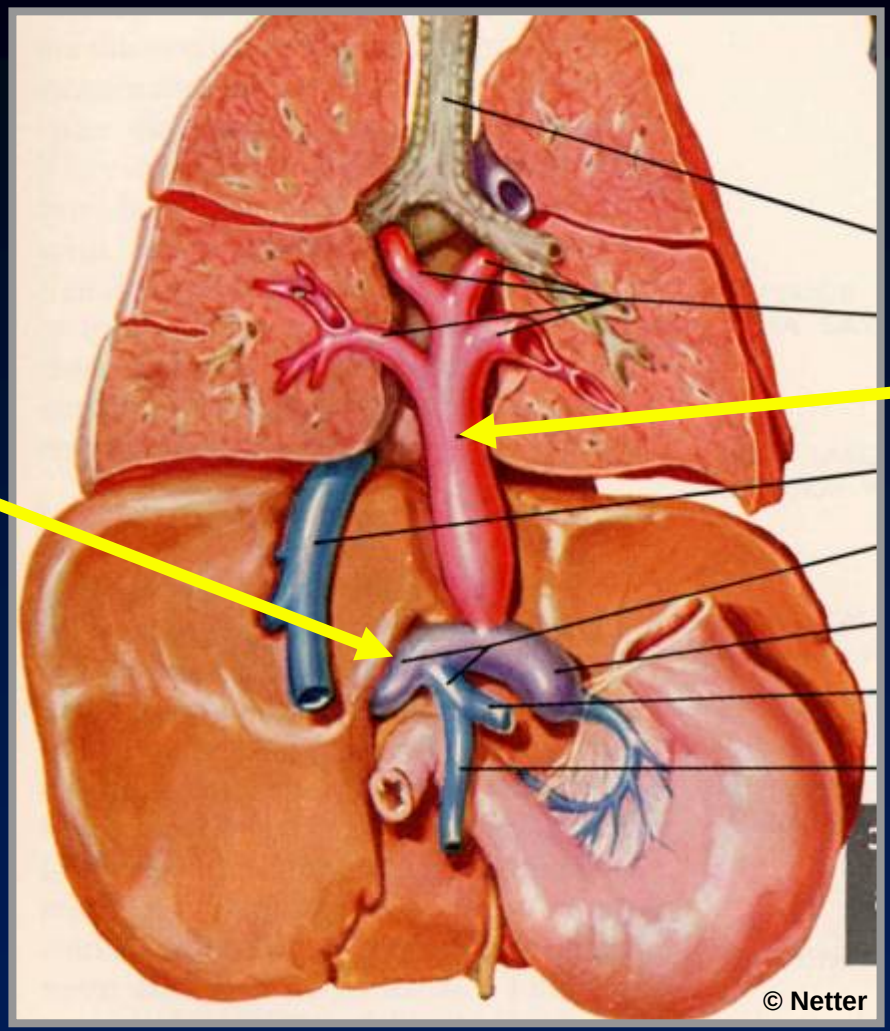
Blood returning from lungs → pulmonary veins which are constricted by diaphragm → CHF

Oxygenated blood returns to RA



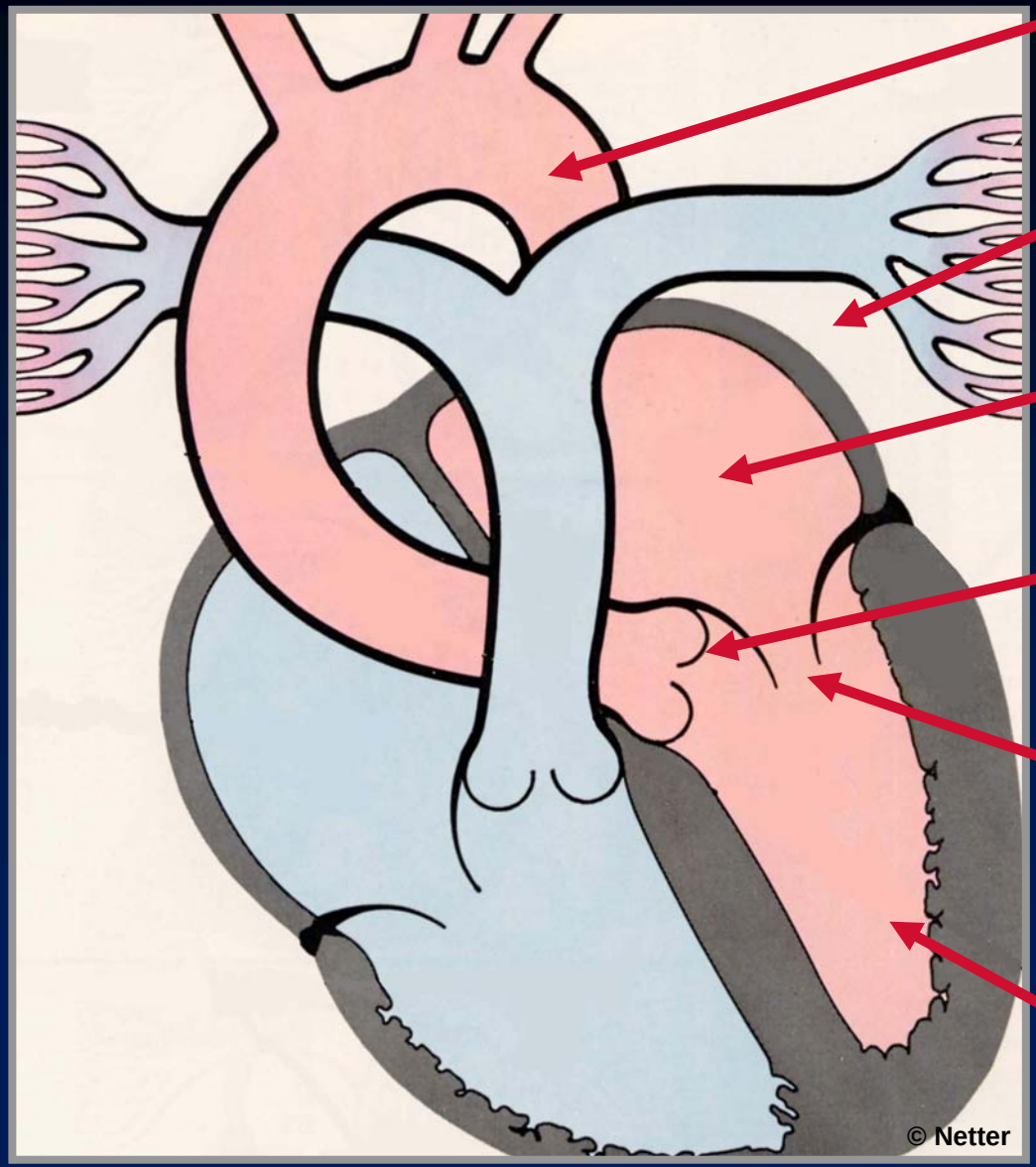
Pulmonary veins empty into portal vein or IVC

TAPVR-Type III-Infradiaphragmatic



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