

William Herring, M.D. © 2003

Diseases of the Great Vessels

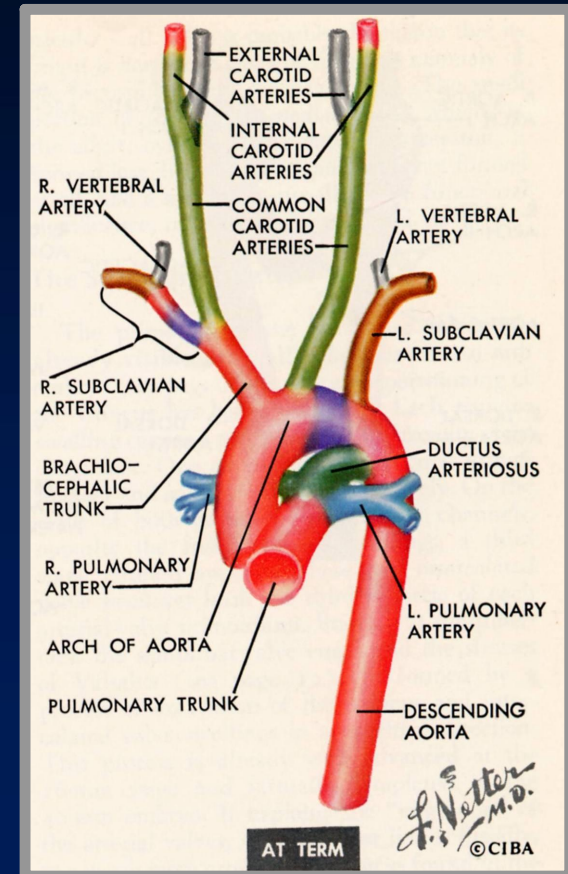
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Aortic Anomalies

General

- Most are asymptomatic
 - Unless they cause encircling vascular ring like pulmonary sling
- Can be complex lesions requiring multiple projections
 - MRI or CT



Aberrant Subclavian Arteries

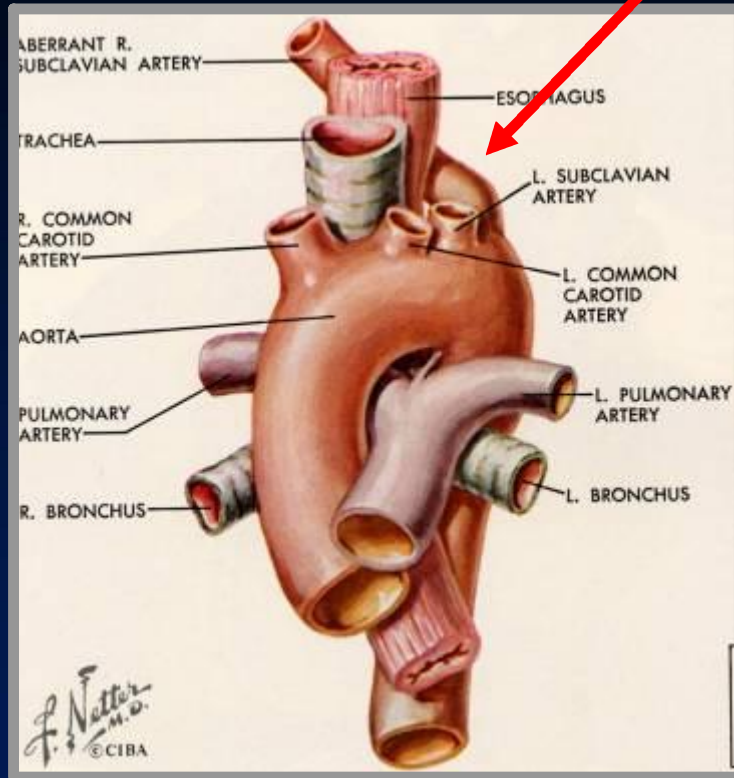
General

- **Left arch with aberrant right subclavian**
 - Usually passes posterior to esophagus
 - Dilated origin is “Diverticulum of Kommerell”
- **Right arch with aberrant left subclavian**
 - Most are asymptomatic
 - Passes behind esophagus
 - Low incidence of congenital heart dz

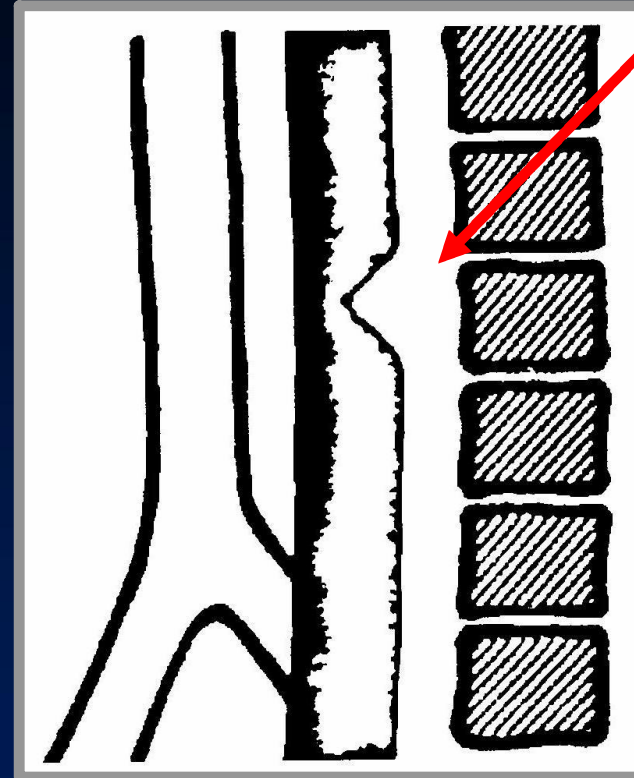
Left Aortic Arch With Anomalous RSCA

Left Arch with Anomalous RSCA

- Occurs in less than 1% of people
- Passes posterior to esophagus
 - Pushes trachea and esophagus forward
- Produces oblique shadow above aortic arch on frontal film
- Origin of RSCA may be dilated
 - Diverticulum of Kommerell

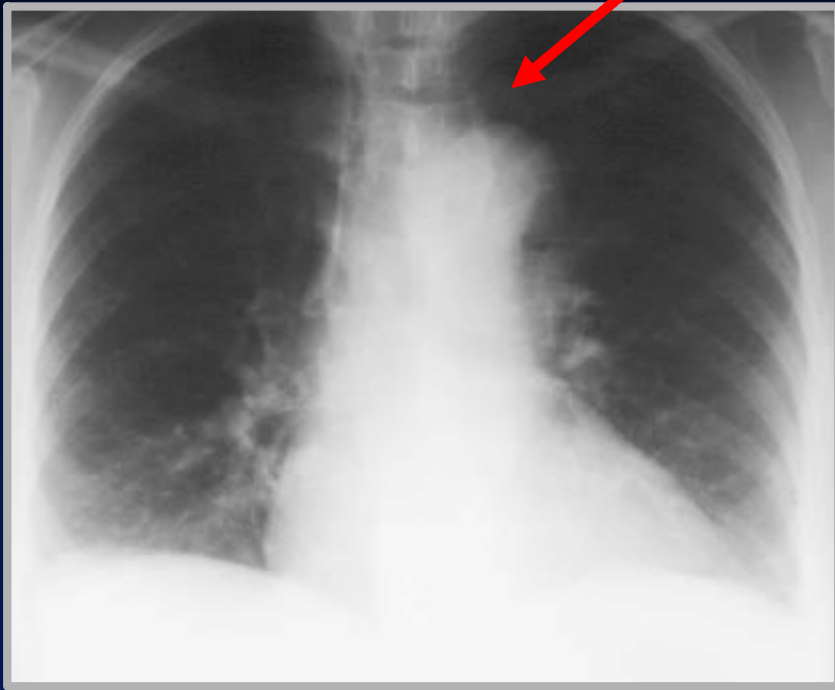


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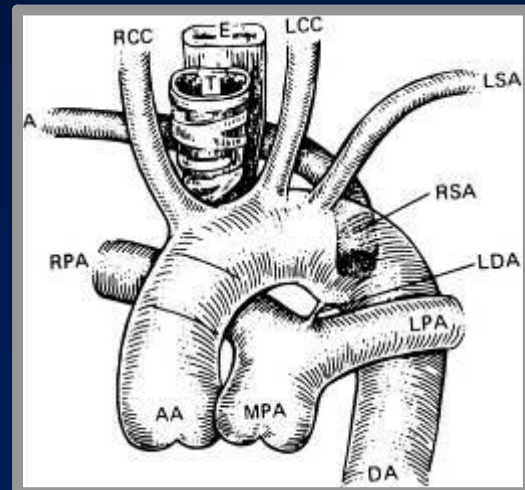
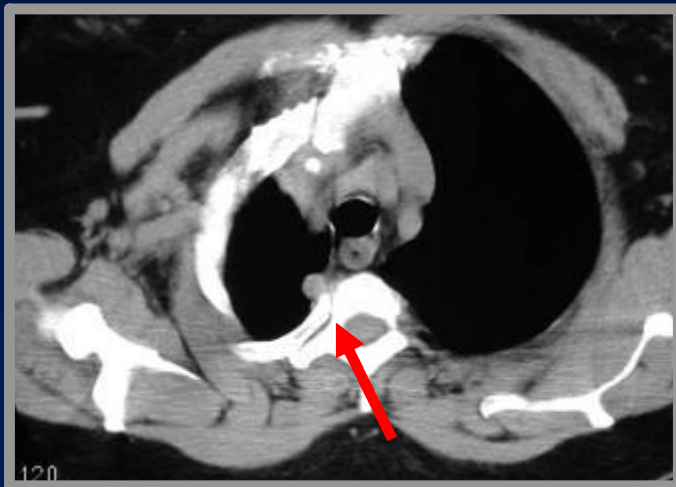
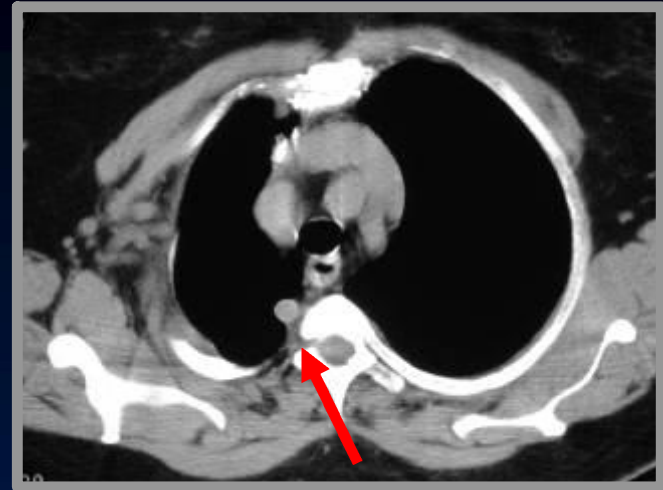
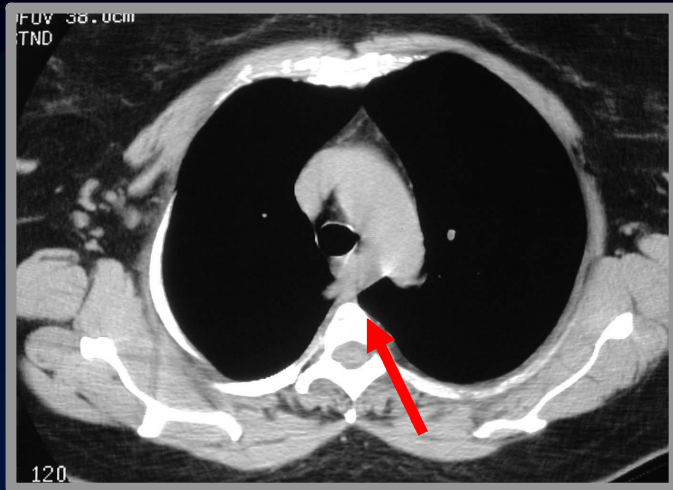


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Left Aortic Arch with Aberrant R SCA



Left Aortic Arch with Aberrant Right SCA



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Left Aortic Arch with Aberrant R SCA

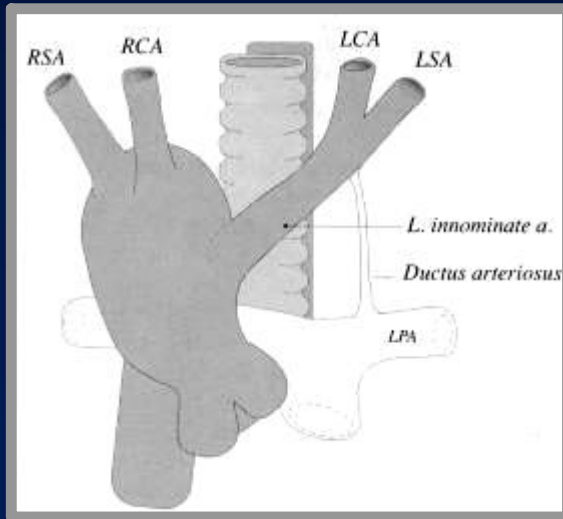
Right Aortic Arches

Right Aortic Arch Types

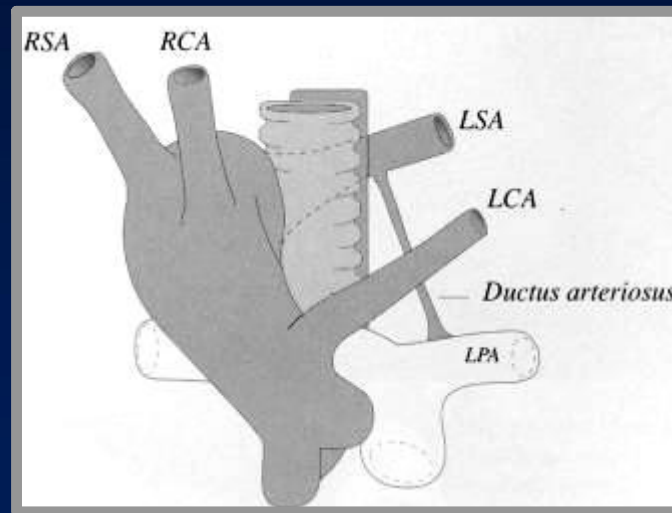
- At least five different types
- Only two of importance

Right Aortic Arch Types

- Mirror Image Type — Type I
- Aberrant left subclavian — Type II



Mirror Image



Aberrant LSCA

Right Aortic Arches

General

- **Recognized by leftward displacement**
 - **Of barium-filled esophagus**
 - **Of air-filled trachea**
- **Aortic knob is absent from left side**
- **Aorta descends on right**
- **Para-aortic stripe returns to left side of spine just above diaphragm**

Right Aortic Arches

General

- **Mirror-image type almost always has associated CHD**
 - Usually Tetralogy of Fallot
- **Aberrant Left Subclavian type rarely has associated CHD**
 - Most common variety of right arch

Right Aortic Arch

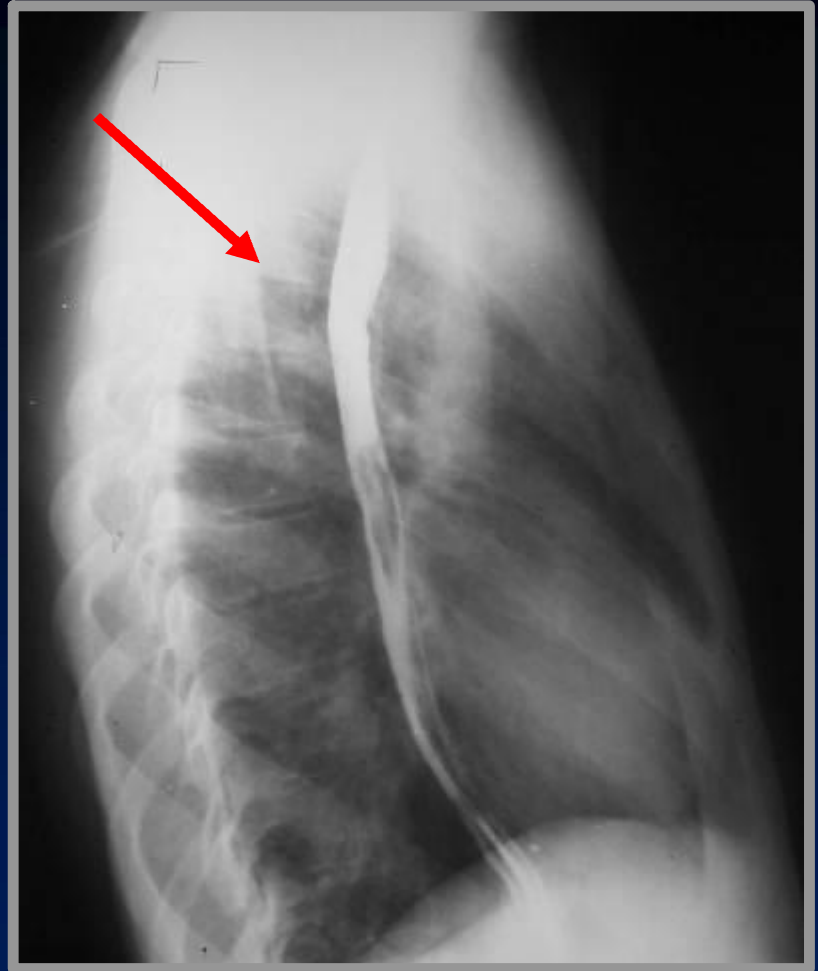
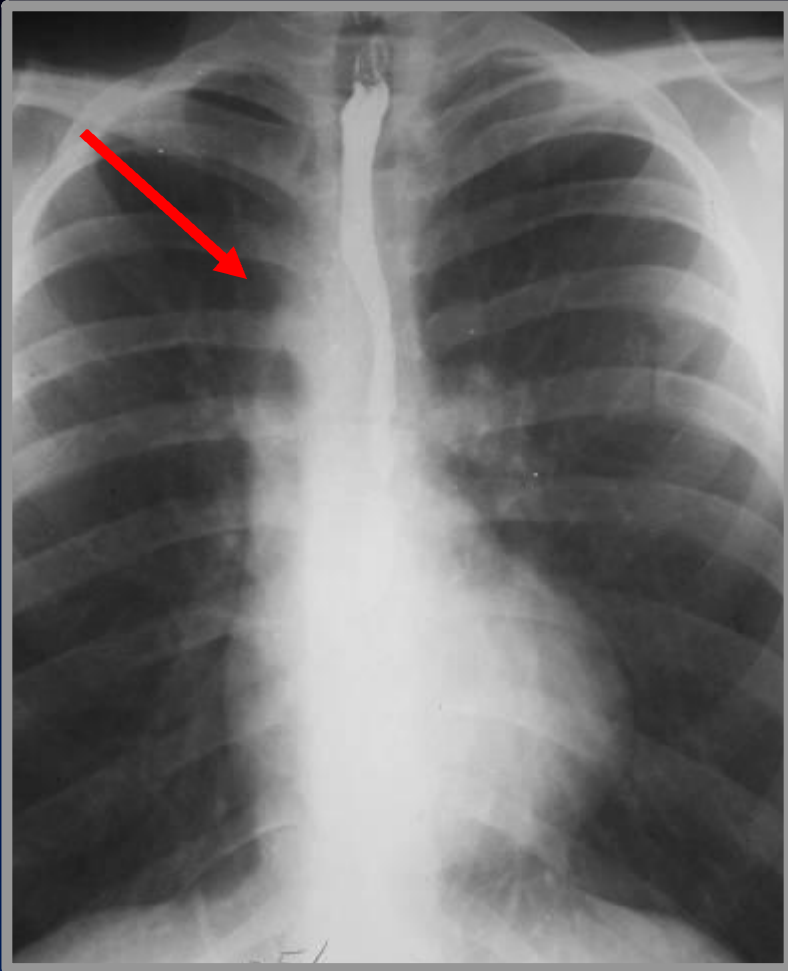
Type 1—Mirror Image Type

- **2° interruption of left arch just distal to ductus arteriosus**
- **Associated with congenital heart disease 98% of time**

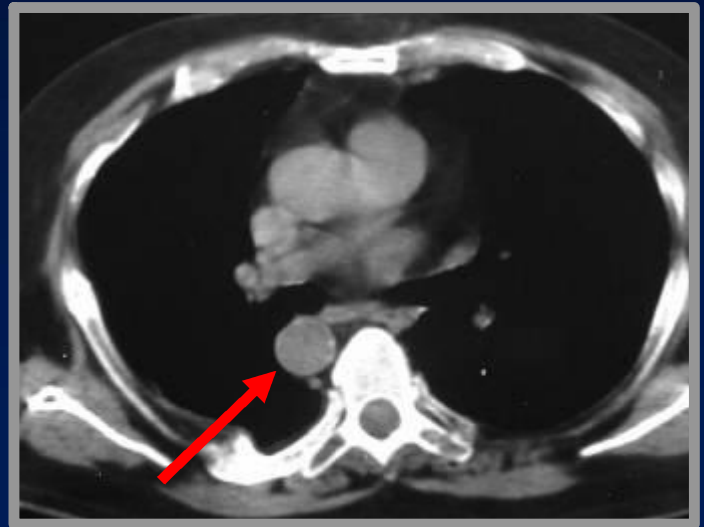
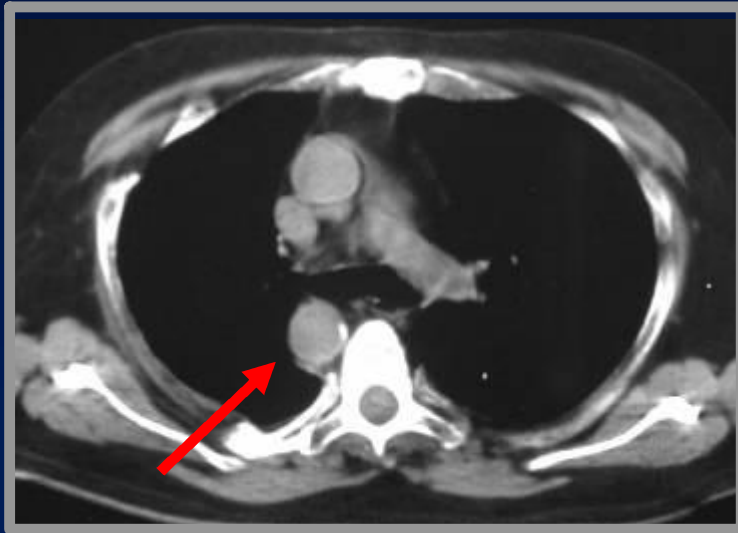
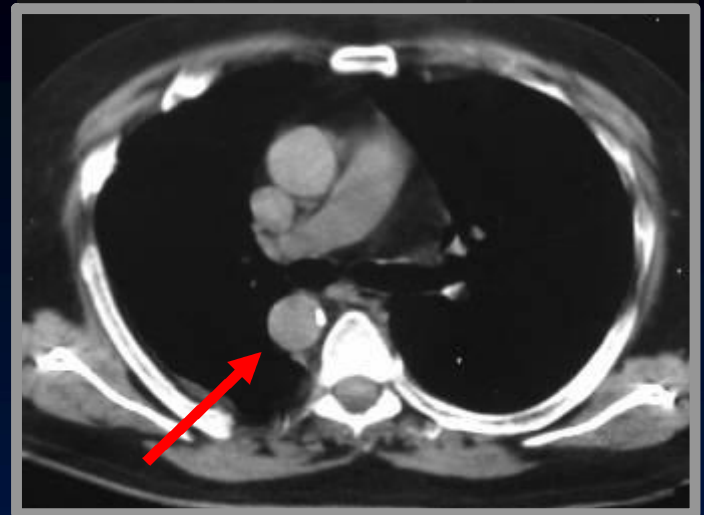
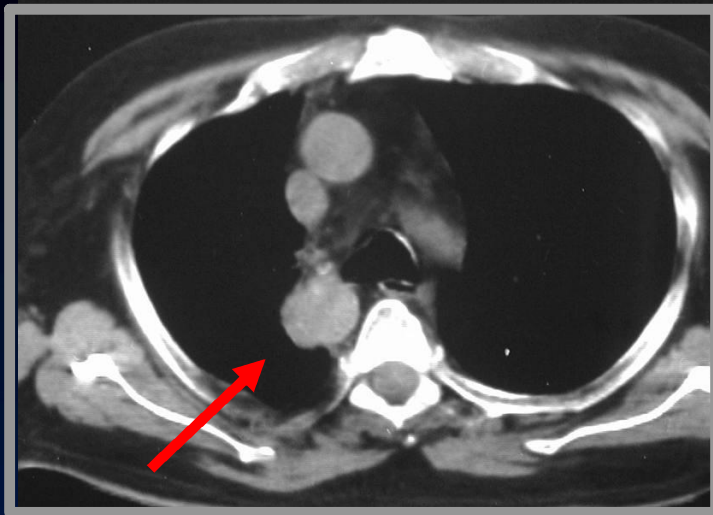
Right Aortic Arch

Type 1—Mirror Image Type—X-ray Findings

- **No posterior impression on trachea or barium-filled esophagus**
- **Heart is usually abnormal in size or shape**
- **Aorta descends on right**



Mirror Image Right Aortic Arch with TOF



Mirror Image Right Aortic Arch

Right Aortic Arch

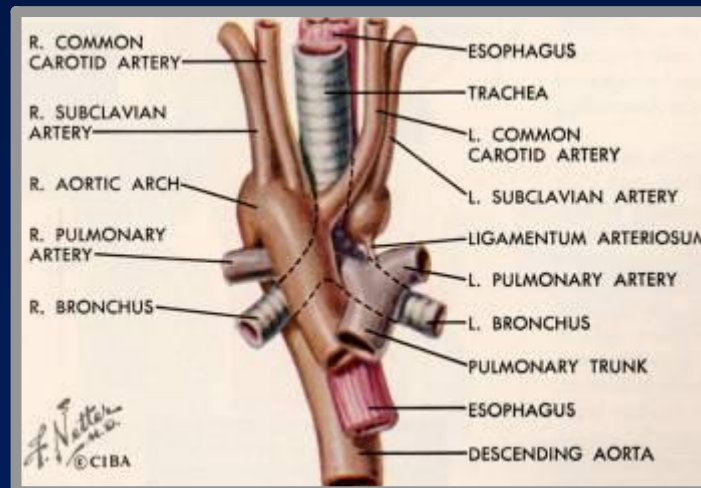
Type II—Aberrant Left Subclavian

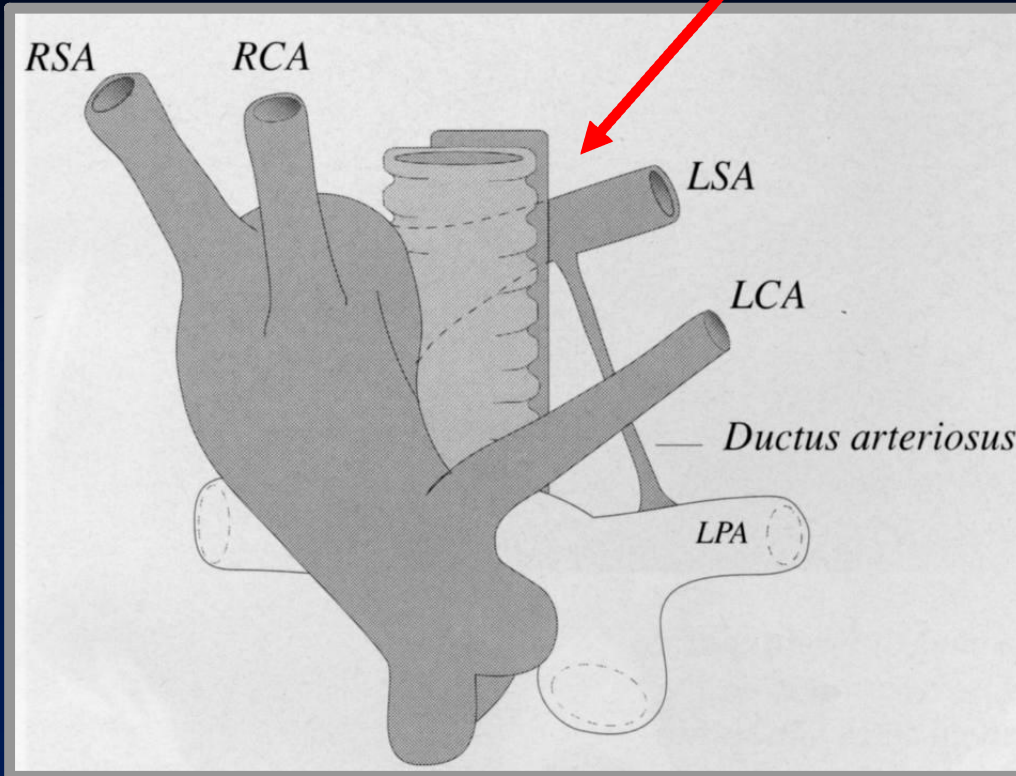
- **2° interruption of left aortic arch between LCC and LSC arteries**
- **Associated with cardiac defects 5-10% of the time**
 - **Tetralogy of Fallot most often (71%)**
 - **ASD or VSD next most often (21%)**
 - **Coarctation of aorta rarely (7%)**

Right Aortic Arch

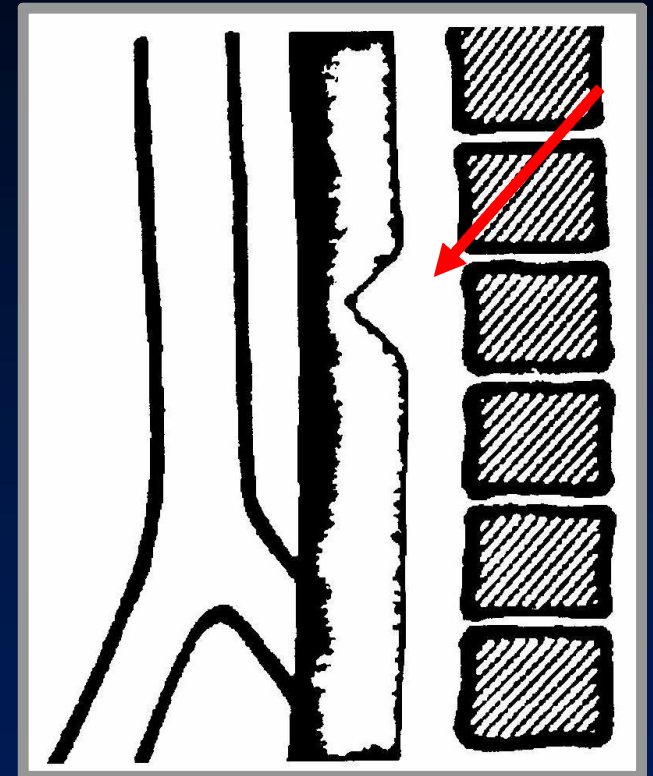
Type II—Aberrant Left Subclavian

- Anomalous left subclavian artery (retroesophageal and retrotracheal)
- Aorta descends on right





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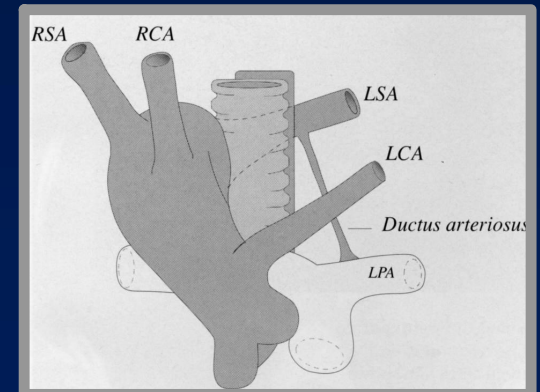
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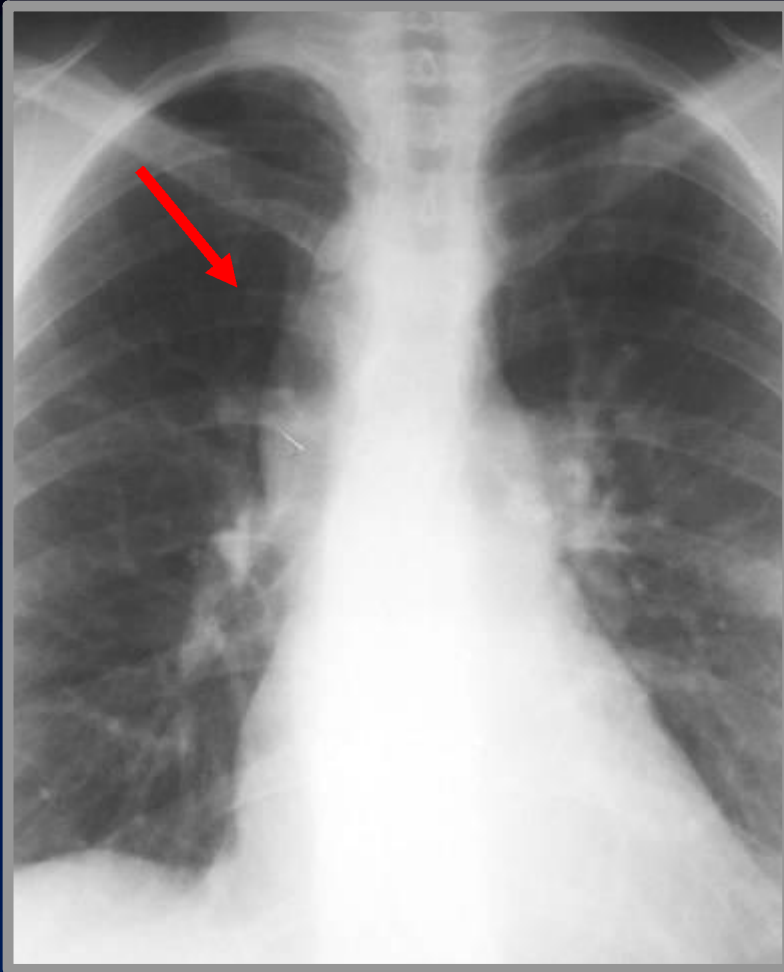
Right Arch with Aberrant LSCA

Right Aortic Arch

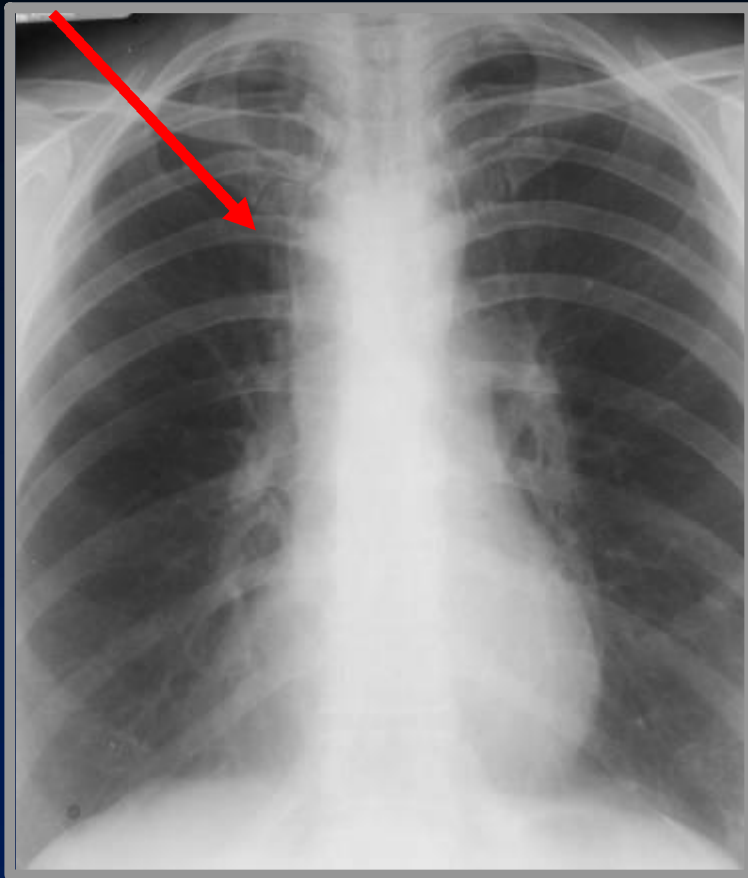
Aberrant Left Subclavian—X-ray Findings

- Posterior impression on trachea and barium-filled esophagus
- Heart is usually normal in size and shape
- Aorta descends on right

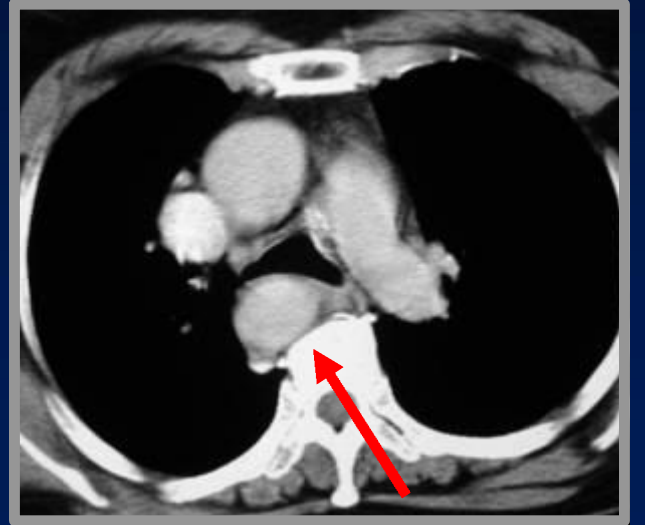
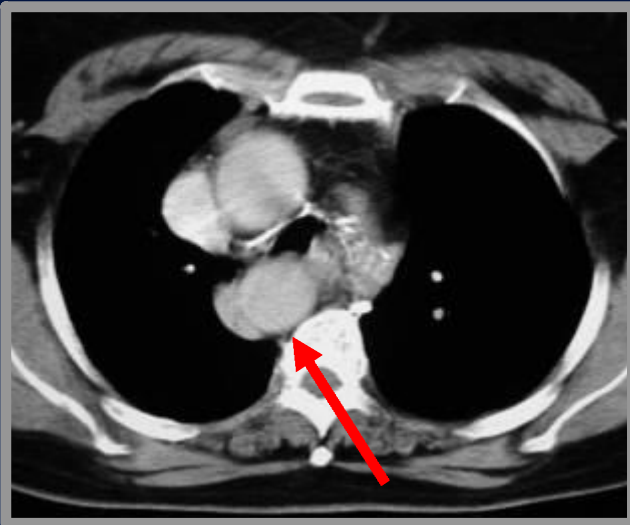
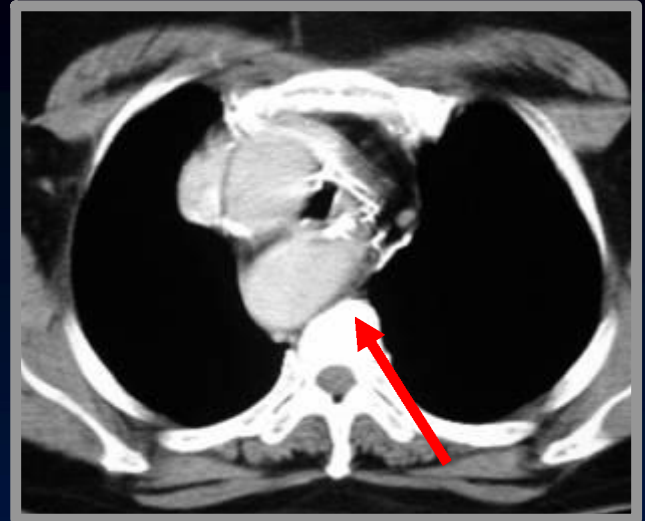
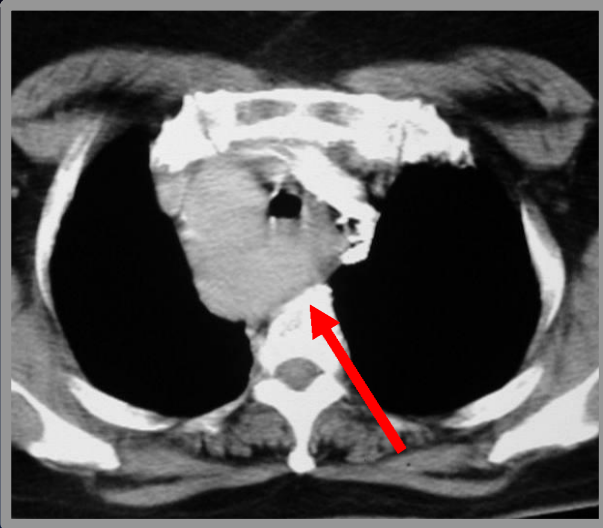




Right Aortic Arch with Aberrant Left Subclavian



Right Aortic Arch with Aberrant Left Subclavian



Right Aortic Arch with Aberrant Left Subclavian

**If the patient has
a Mirror Right arch,**

**Then it will be
associated**

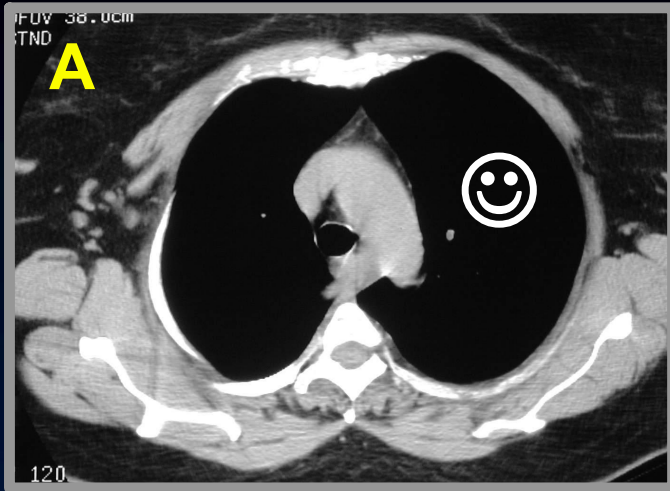
- **90% with Tetralogy of Fallot**
- **6% with Truncus Arteriosus**
- **5% with Tricuspid Atresia**

**If the patient has
this disease,**

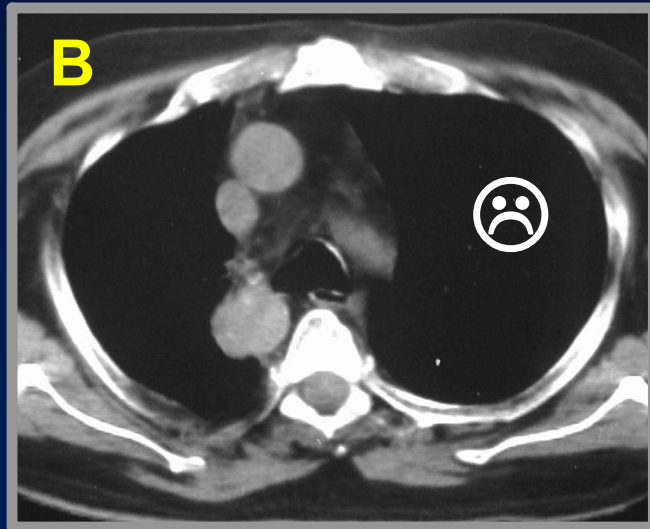
**This % will have a
Mirror Right arch**

- **Truncus arteriosus** **33%**
- **Tetralogy of Fallot** **25%**
- **Transposition** **10%**
- **Tricuspid atresia** **5%**
- **VSD** **2%**

**Apparent discrepancy due to much
higher ↑ incidence of TOF than Truncus**

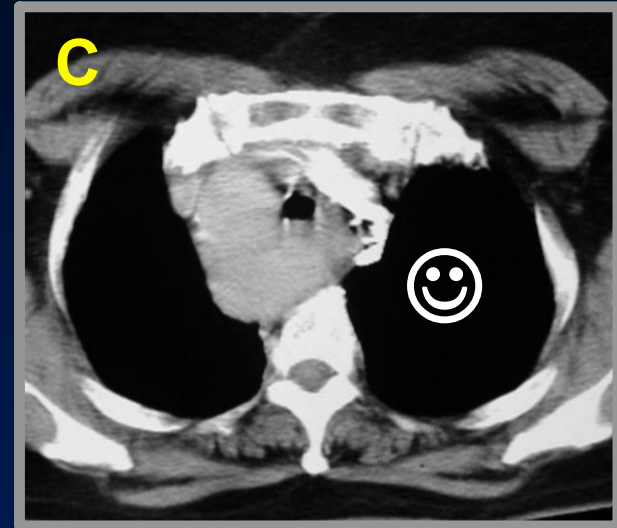


**Left Aortic Arch
with Aberrant R SCA**



**Mirror Image
Right Aortic Arch**

**Identify these three anomalies and
tell whether they are usually
associated with congenital heart
disease or not**



**Right Aortic Arch with
Aberrant Left Subclavian**

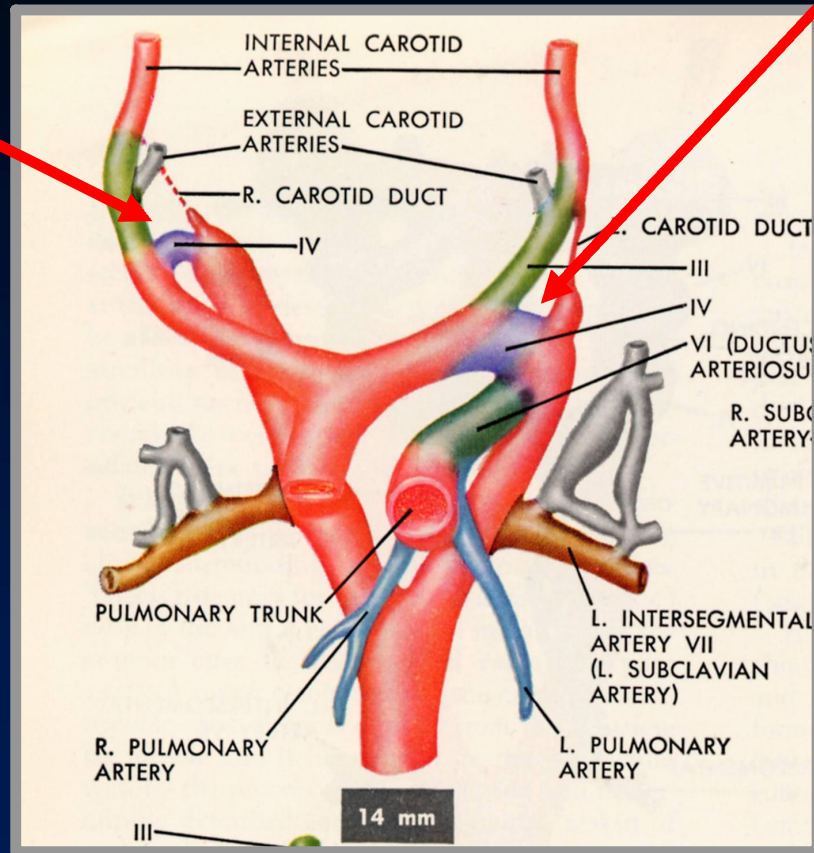
Double Aortic Arch

Double Aortic Arch

General

- **Most common vascular ring**
- **Rarely associated with congenital heart disease**
 - **But vascular ring → tracheal and/or esophageal compression**
- **Caused by persistence of R and L IV branchial arches**

R IV arch normally becomes most proximal segment of RSCA



L IV arch is part of normal aortic arch between LCC and LSCA

Persistence of both IV branchial arches forms a vascular ring or Double Aortic Arch

Double Aortic Arch

General

- **Passes on both sides of trachea**
 - **Joins posteriorly behind esophagus**
- **Right arch is larger and higher**
- **Left arch is smaller and lower**
 - **Ba swallow shows bilateral impressions on frontal view**
 - **Posterior impression on lateral view**
- **Angiogram is characteristic**

Double Aortic Arch

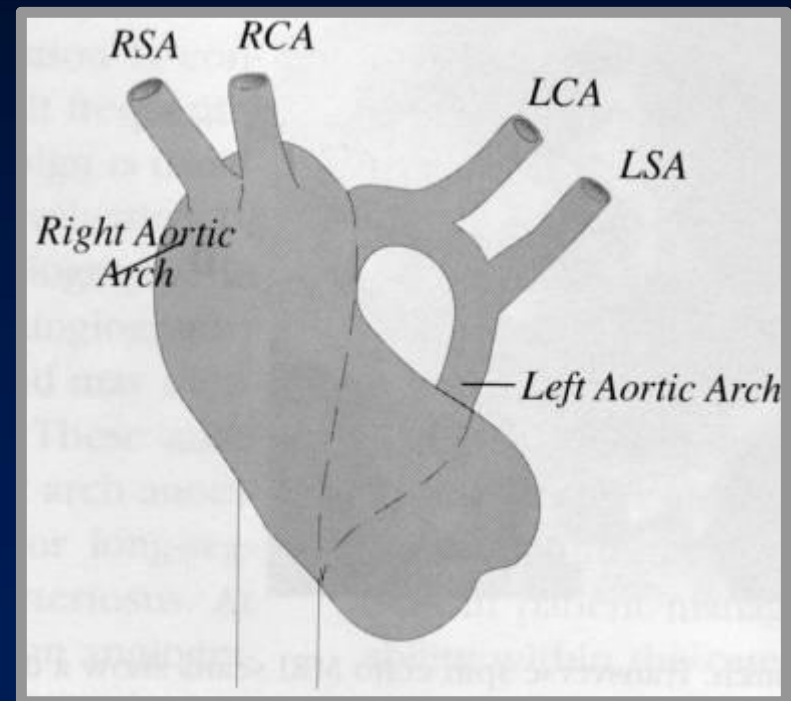
Clinical

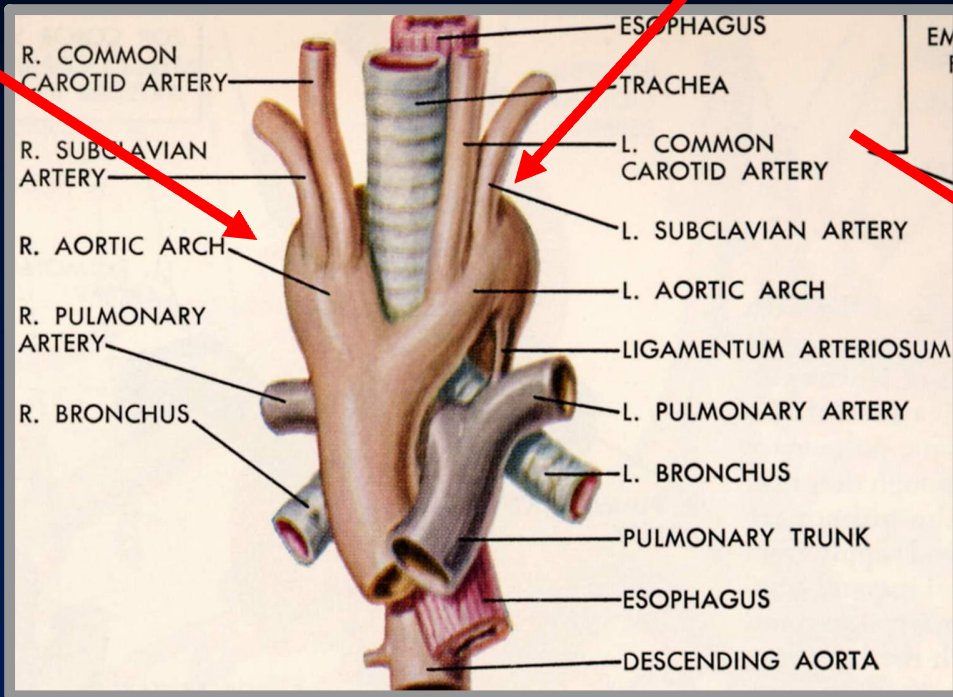
- **Symptoms may begin at birth**
- **Symptoms include**
 - **Tracheal compression, or**
 - **Difficulty swallowing**

Double Aortic Arch

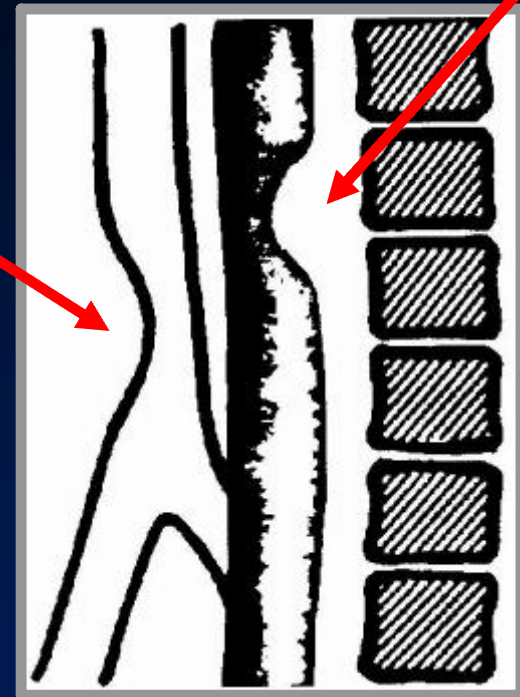
Anatomy

- Right arch supplies RSCA and RCC
- Left arch supplies LCC and LSCA





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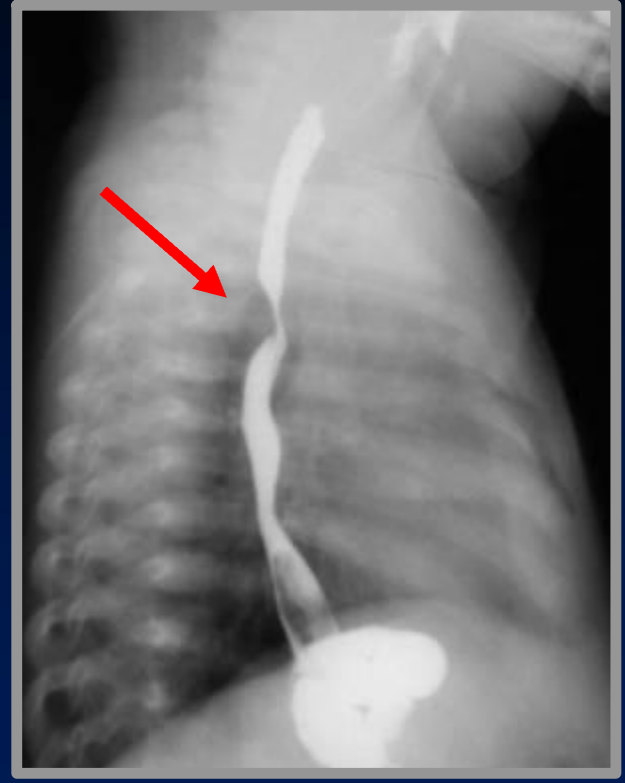
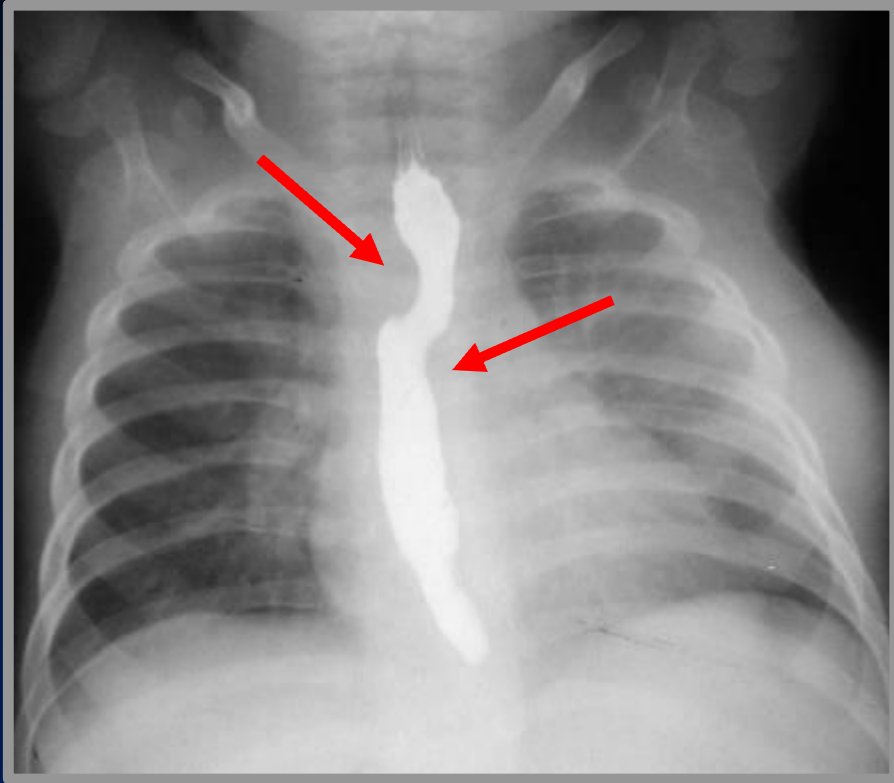
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Double Aortic Arch

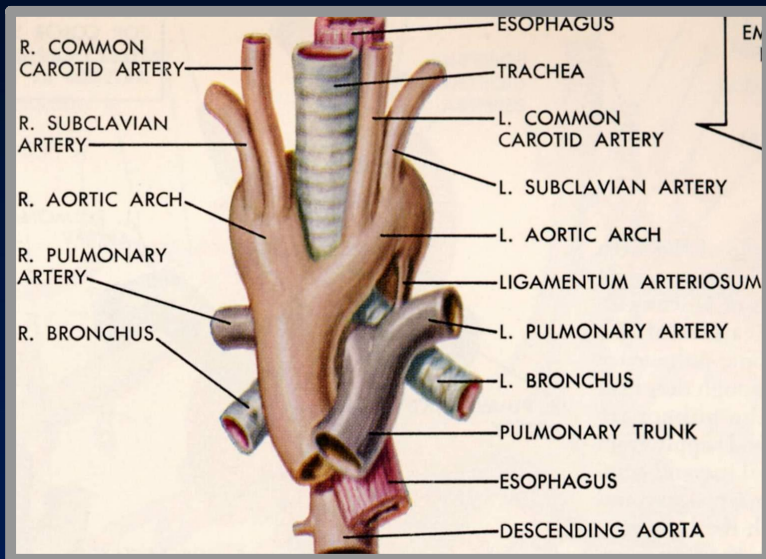
Double Aortic Arch

X-ray Findings

- Right arch is higher and larger
- Left arch is lower and smaller
 - Produces reverse S on esophagram on AP
- On lateral, arches are posterior to esophagus and anterior to trachea

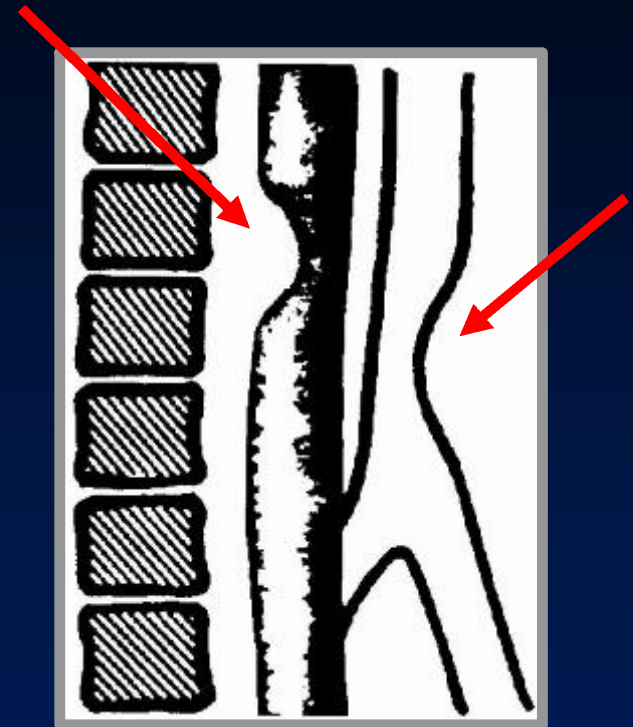
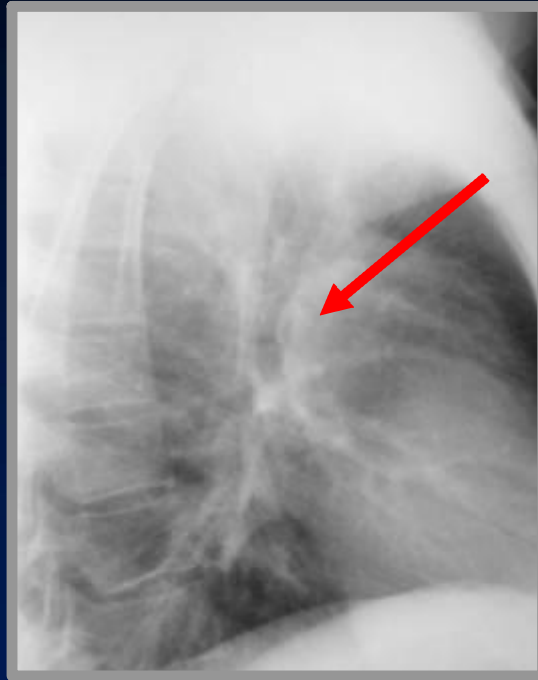
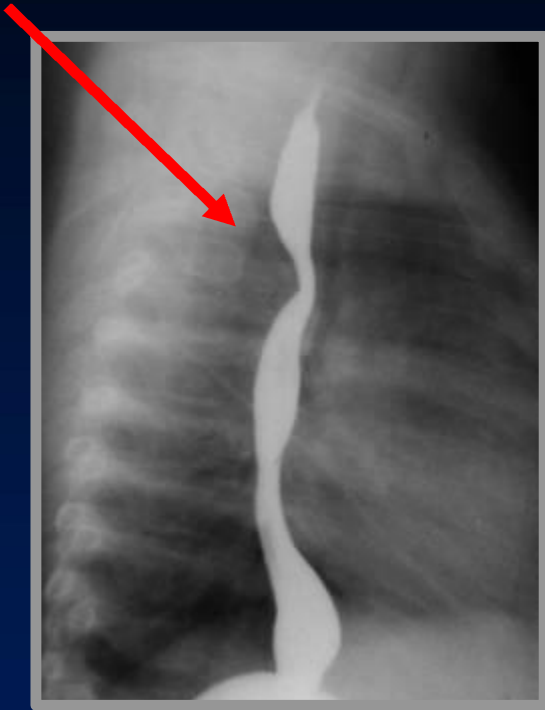


Double Aortic Arch



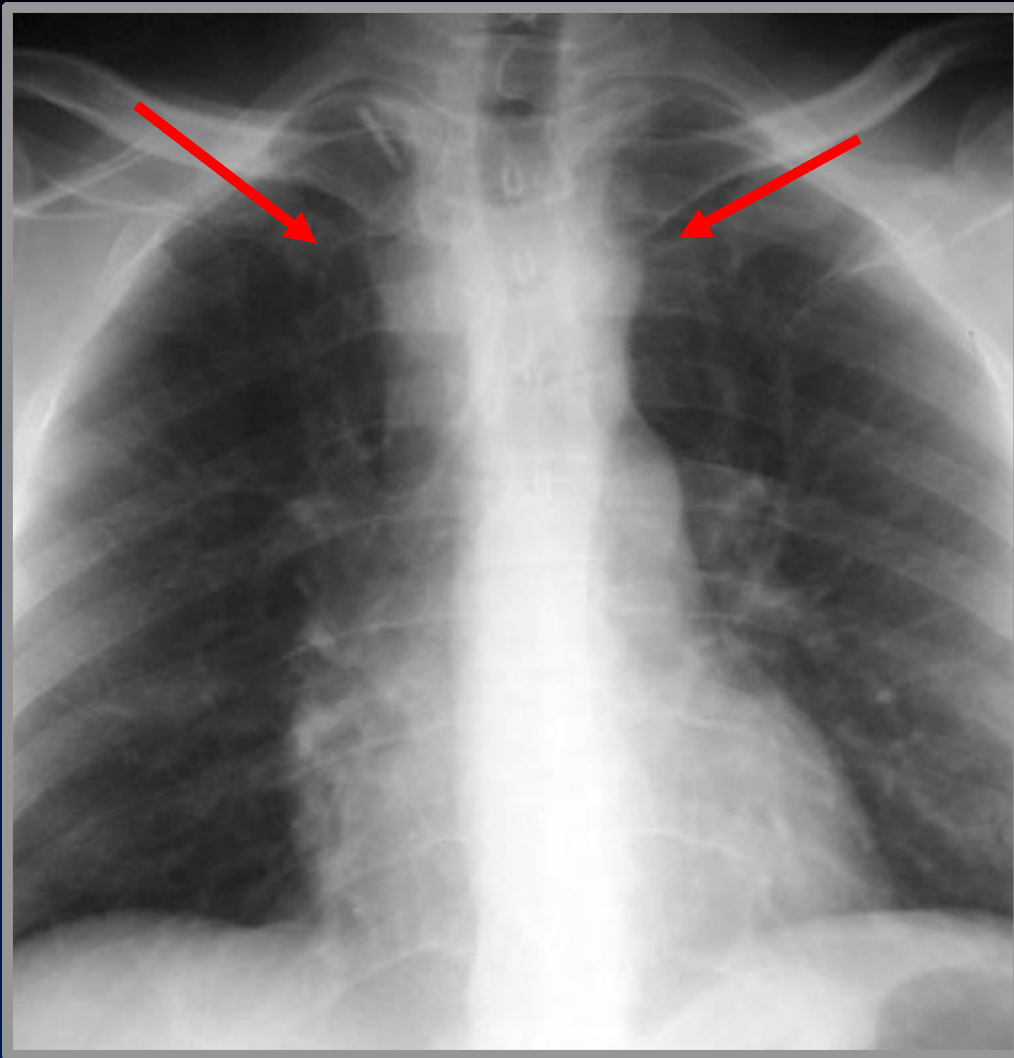
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Double Aortic Arch



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Double Aortic Arch Impressions on Trachea and Esophagus



Double Aortic Arch

Cervical Aortic Arch

Cervical Aortic Arch

General

- Rare
- Usually asymptomatic
 - May present as pulsating supraclavicular mass
 - May produce vascular ring and compress airway
- Embryogenesis uncertain
- Over 80% are *right-sided*

Cervical Aortic Arch

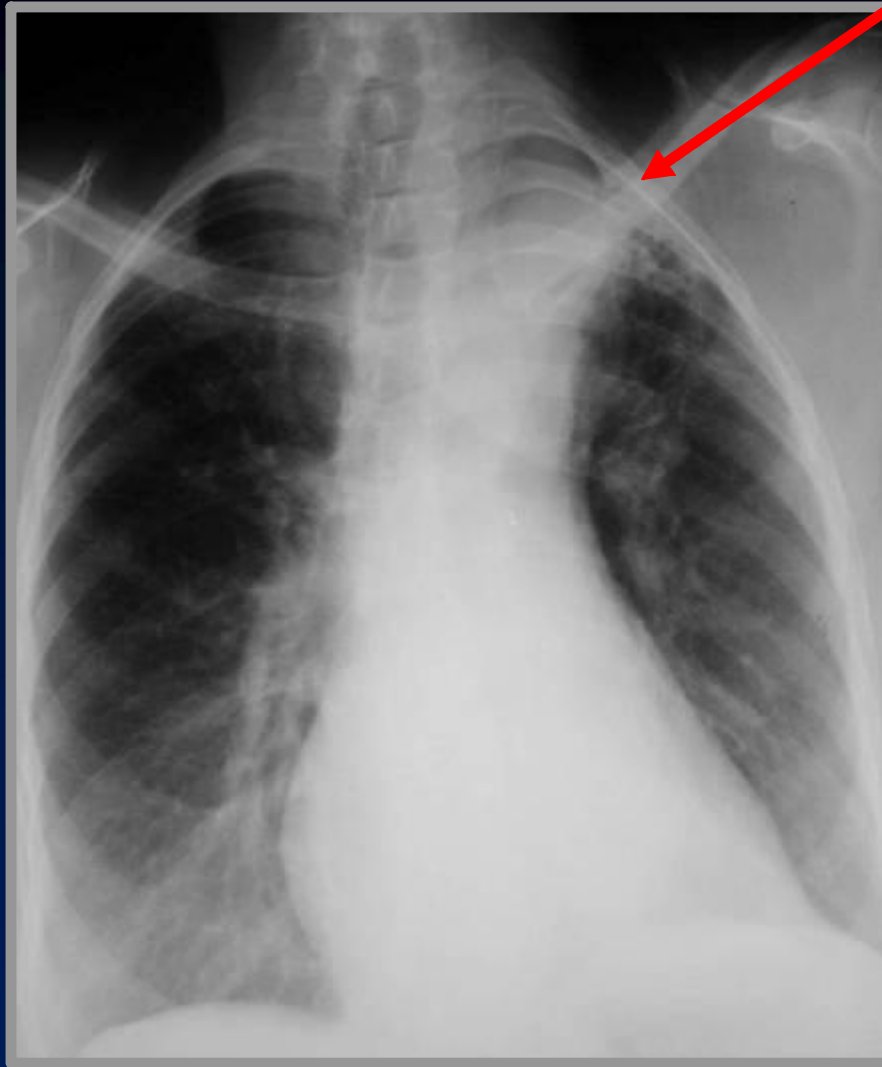
Imaging Findings—Right-sided lesions

- **Right-sided cervical aortic arches**
 - **Right apical mass-like density**
 - **Absence of aortic knob**
 - **Descend on the left**
 - **Displace the trachea and esophagus forward**
 - **Branching may be normal or mirror-image**

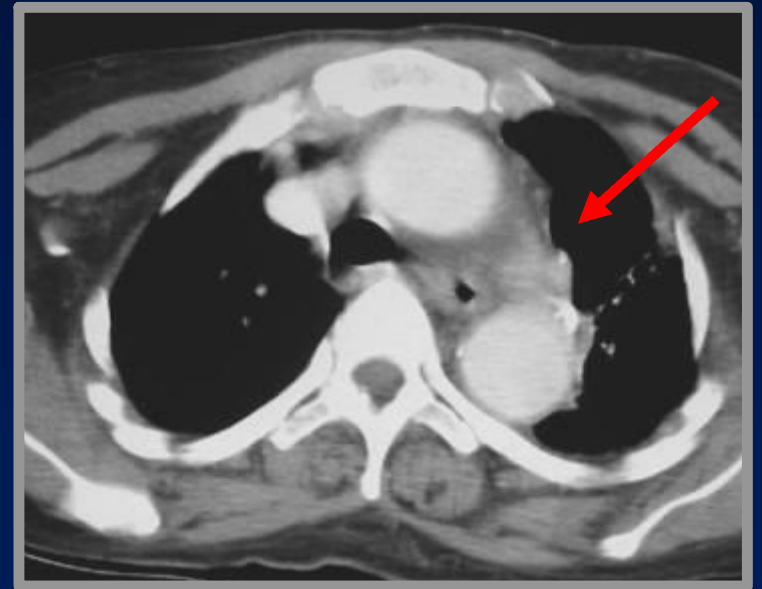
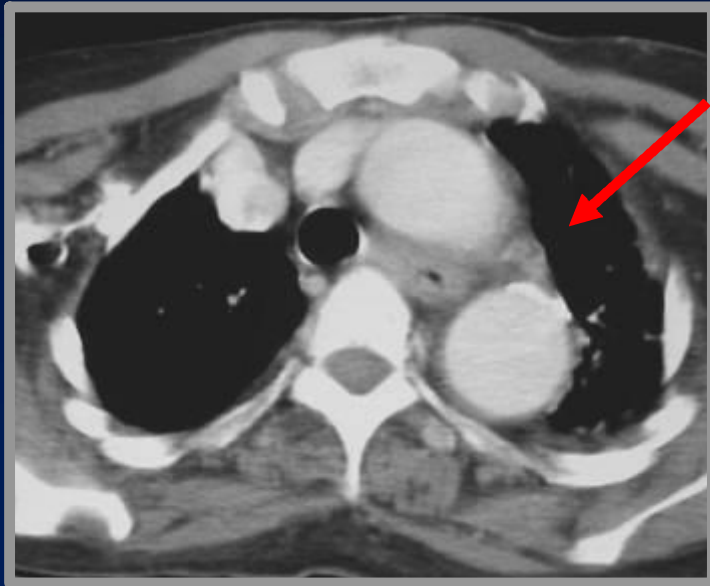
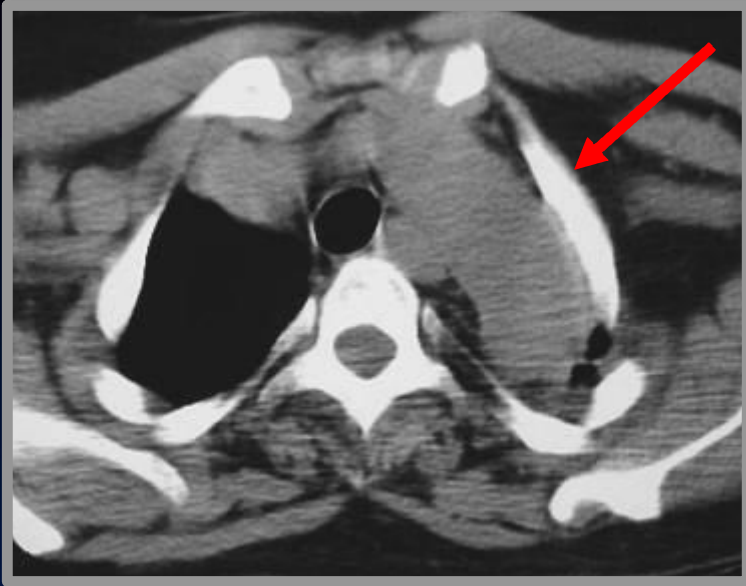
Cervical Aortic Arch

Imaging Findings—Left-sided lesions

- **Left-sided cervical aortic arches**
 - **Aortic knob at apex of lung**
 - **Descend on the left**
 - **Do not displace the trachea or esophagus forward**



Cervical Aortic Arch



Cervical Aortic Arch

Aortitis

Takayasu's Aortitis

Pulseless Disease

- **Chronic inflammatory arteritis**
- **Affects aorta, its main branches and pulmonary arteries**
- **15-40 years, 8:1 females, Oriental population**
- **LSCA, LCCA, brachiocephalic, renals, celiac commonly involved**

Takayasu's Aortitis

Type 3

- Most common is Type 3 (55%)
 - Stenoses of aortic arch, distal thoracic and abdominal aorta



Takayasu's Aortitis

Type 2

- Next most common is Type 2 (11%)
 - Segmental stenoses in descending thoracic and abdominal aorta

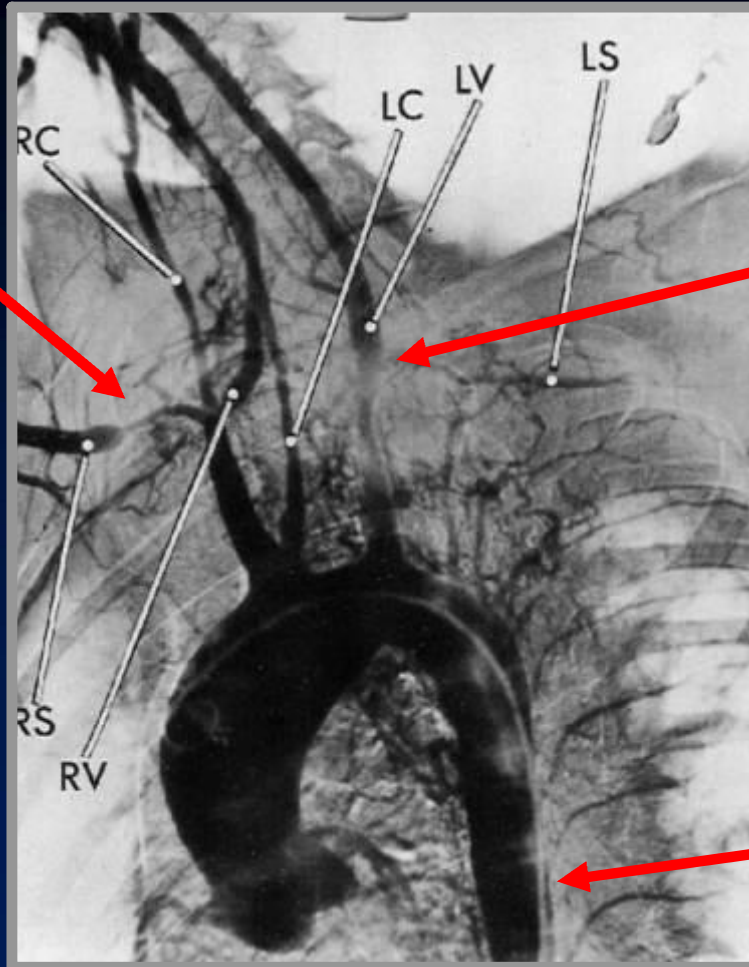


Takayasu's Aortitis

Type 1

- Next most common is Type 1 (8%)
 - Stenoses in arch, brachiocephalic, carotid and subclavian arteries





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Takayasu's Aortitis (Type 3)

Takayasu's Aortitis

Imaging Findings

- **On angiography, narrowing of aortic lumen**
- **On MRI, thickened aortic wall**
- **Associated aneurysms may be saccular or fusiform**

Other Forms of Aortitis

- Inflammation of intima and media
- Healing produces scarring - “tree-bark” appearance of luminal surface
- Aorta dilates
 - Ascending aorta more than arch
 - Abdominal aorta spared
 - Opposite of atherosclerosis

Other Forms of Aortitis

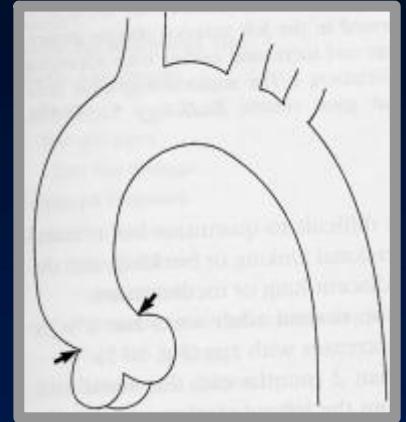
- Aortic wall becomes thickened on healing
- Usually results in aortic regurgitation
 - Diastolic murmur



Giant Cell Arteritis

Causes of Aortitis

- Rheumatic fever
- Reiter's syndrome
- Syphilis
 - Begins above sinotubular ridge
- Giant cell arteritis
- Ankylosing spondylitis
 - Crosses sinotubular ridge and dilates both root and ascending aorta



Sinotubular Ridge-
Jct of Sinuses of Valsalva
and tubular aorta

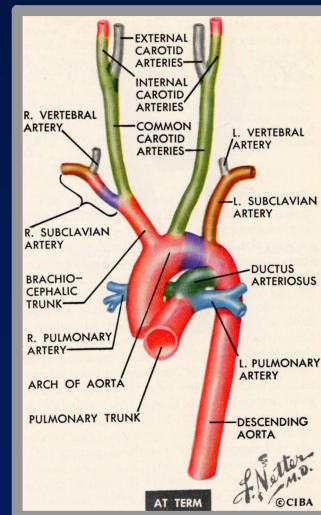
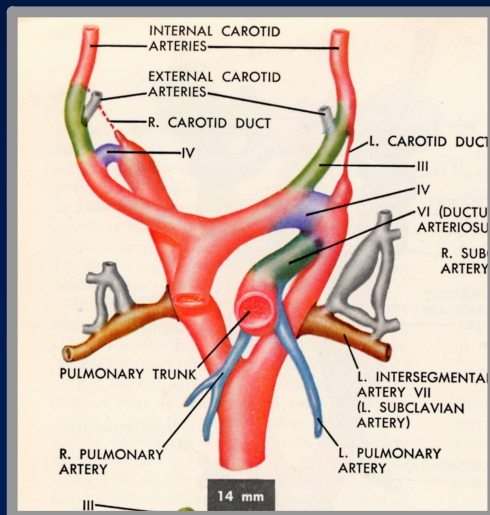


Syphilitic Aortitis

Pulmonary Sling

Pulmonary Sling Embryogenesis

- Failure of formation of left 6th aortic arch
→ absence of left pulmonary artery

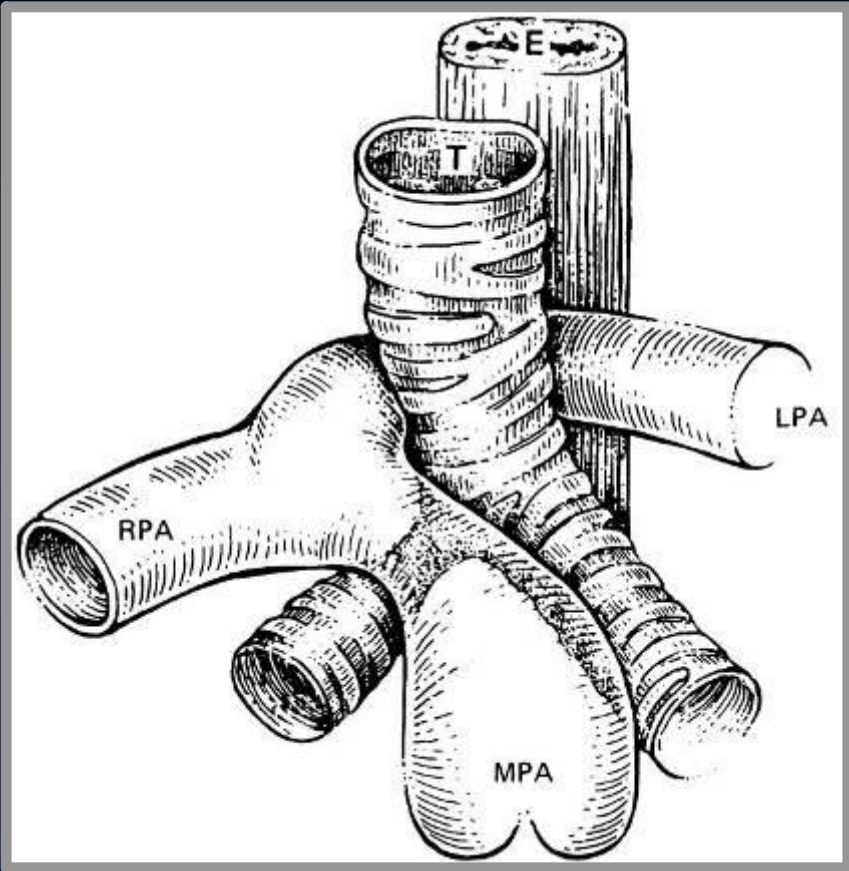


Proximal L VI arch normally becomes proximal segment of L PA; distal VI persists as ductus until birth

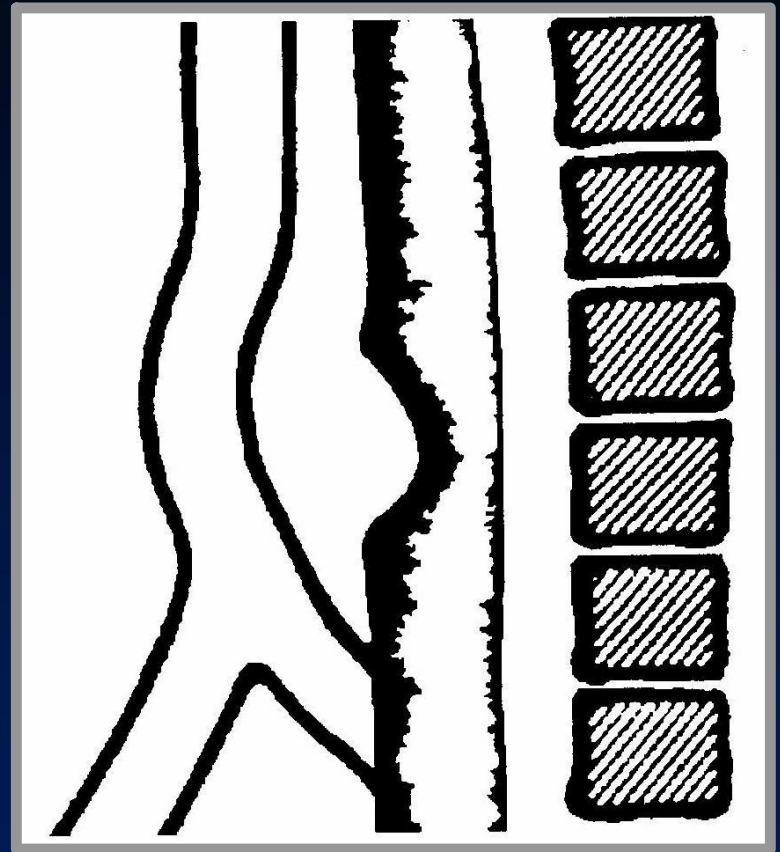
Pulmonary Sling

General

- **Aberrant origin of left pulmonary artery**
 - **From the right pulmonary artery**
- **Left pulmonary artery passes between trachea and esophagus**
- **Most have other anomalies**
 - **Stenosis of right mainstem bronchus**
 - **May lead to air-trapping, lobar emphysema and hyperlucent lung**



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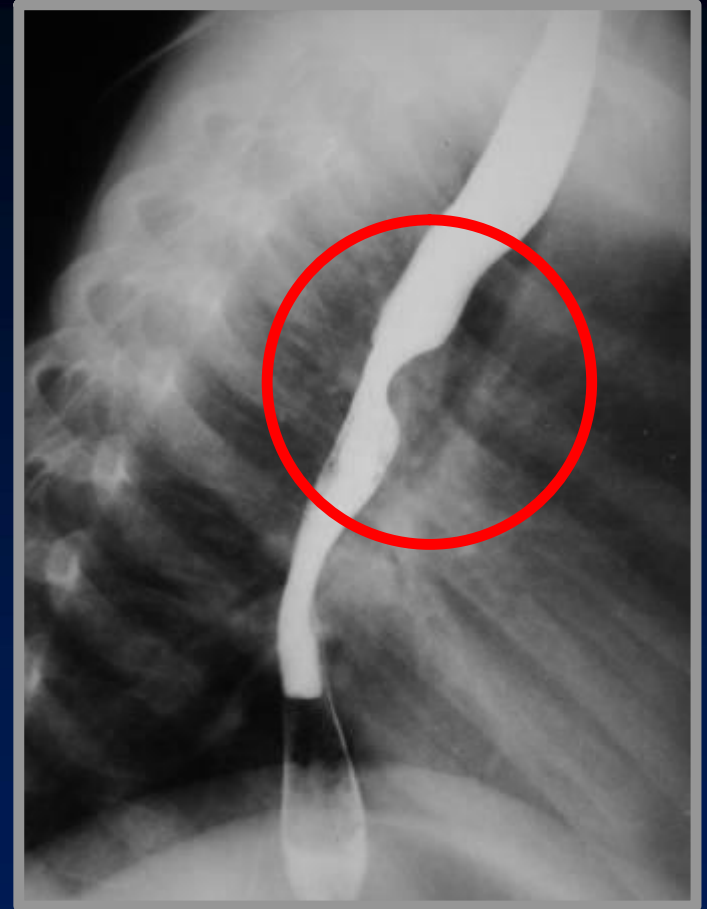
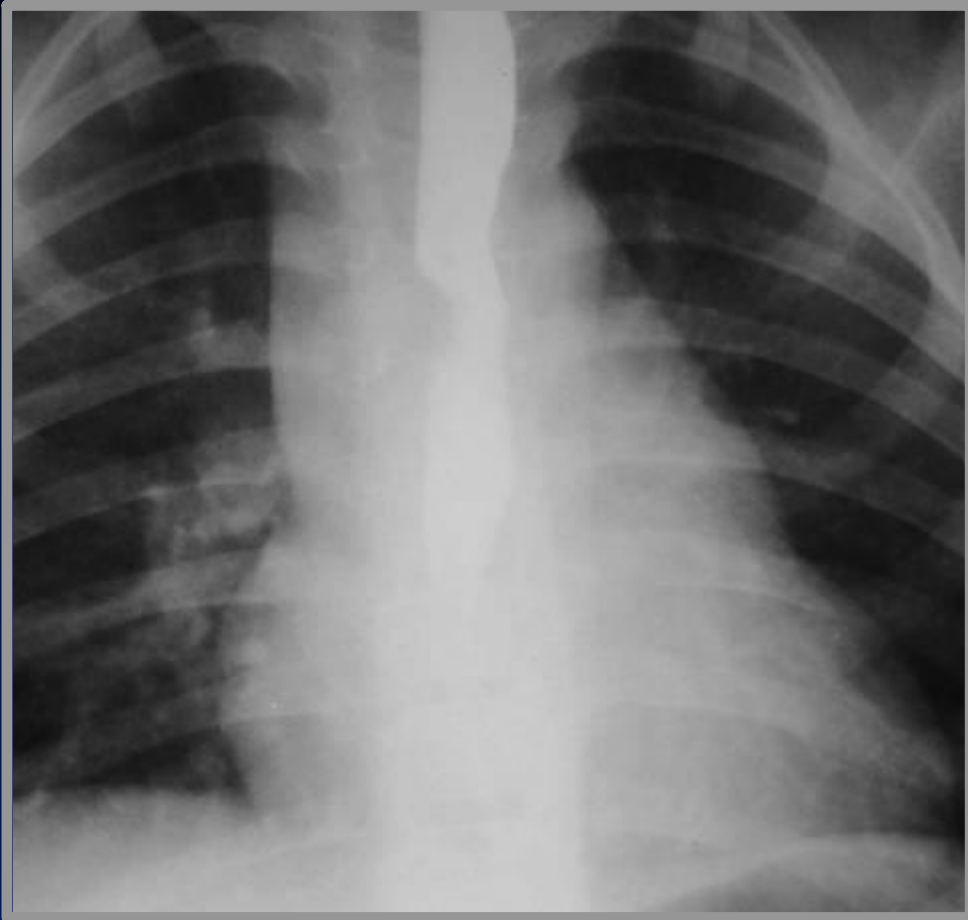
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Pulmonary Sling

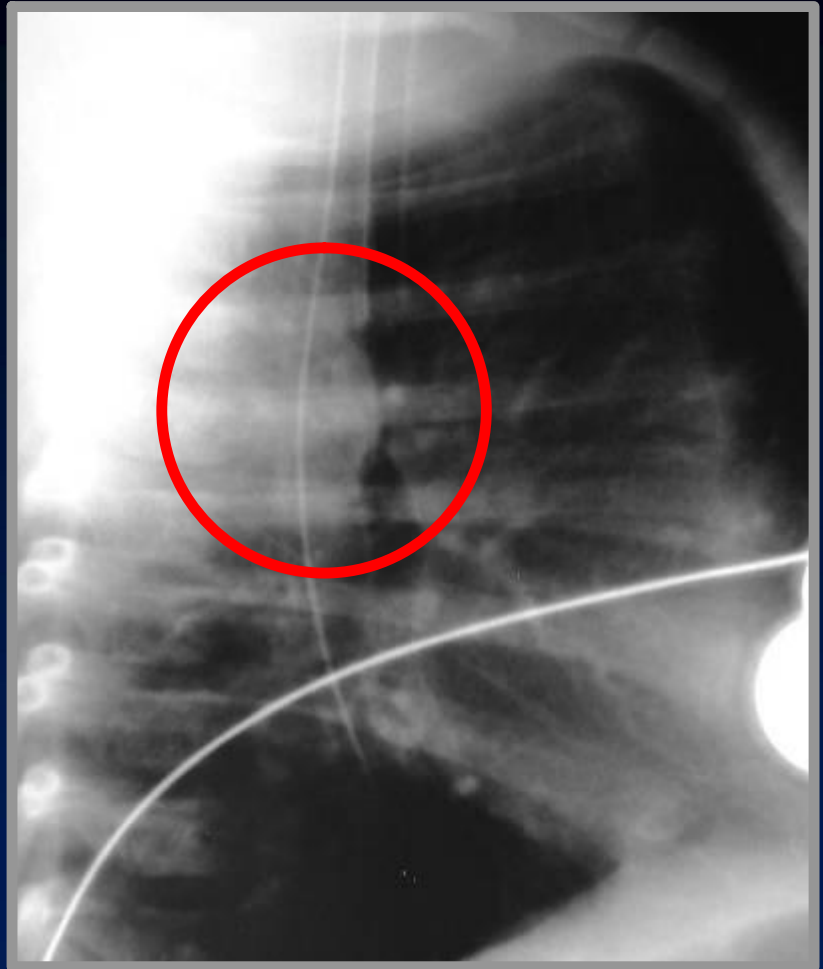
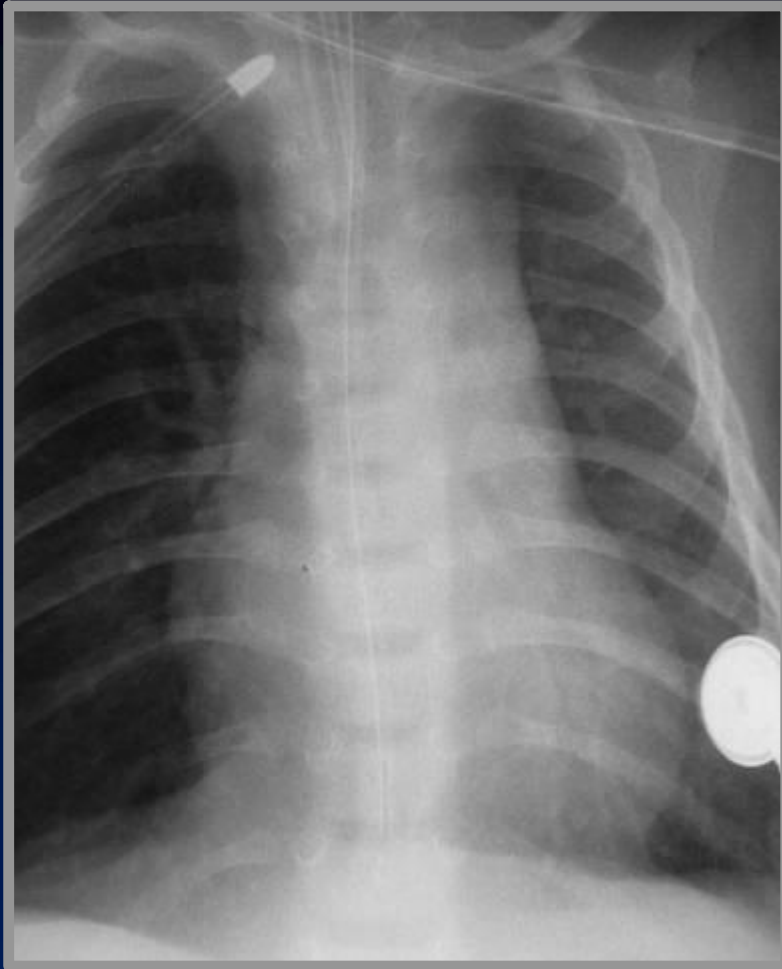
Pulmonary Sling

DDX

- **Only vascular malformation to pass between esophagus and trachea**
- **Bronchial cyst may produce same finding on esophagus/trachea**



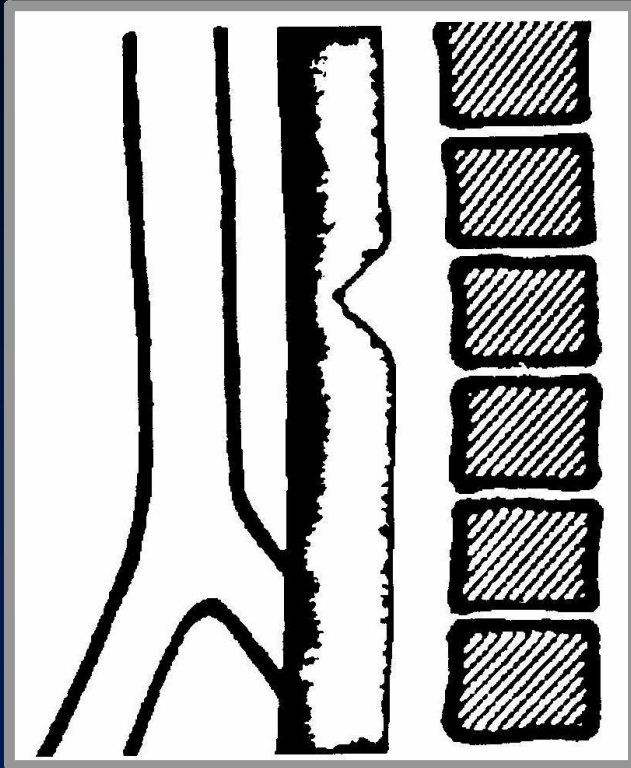
Pulmonary Sling



Pulmonary Sling

Tracheal Impressions

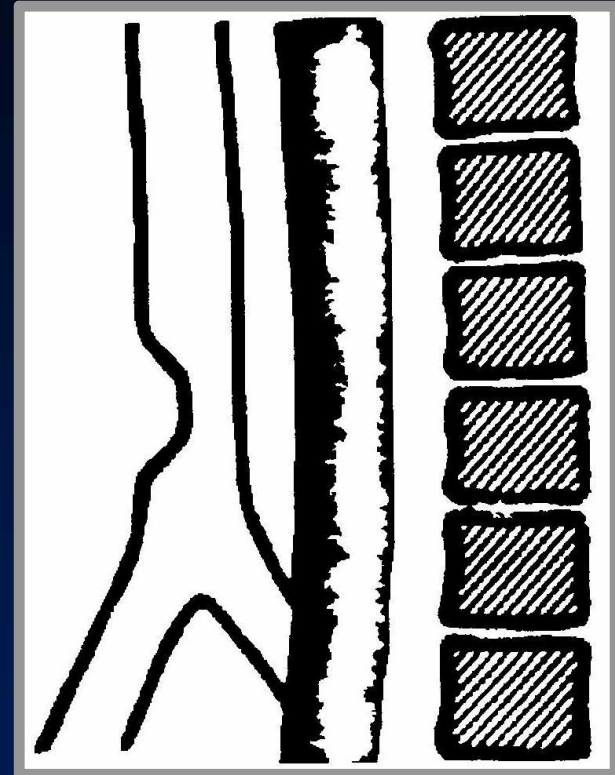
Posterior Esophagus



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Left Ao Arch with Aberrant R SCA
Right Ao Arch with Aberrant L SCA

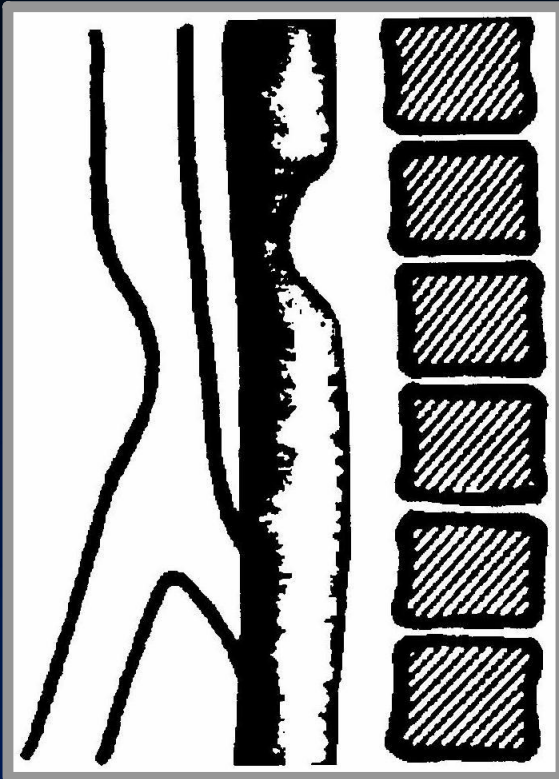
Anterior Trachea



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Isolated anomalies
BCA arising too distal
CCA arising too proximal
CCA and BCA arising together

Anterior trachea and Posterior Esophagus

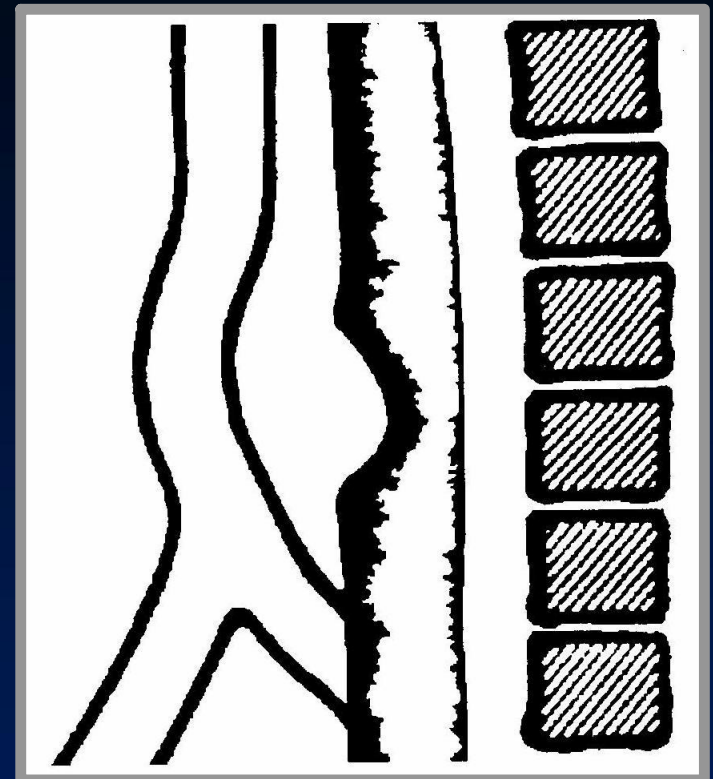


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Double Aortic Arch

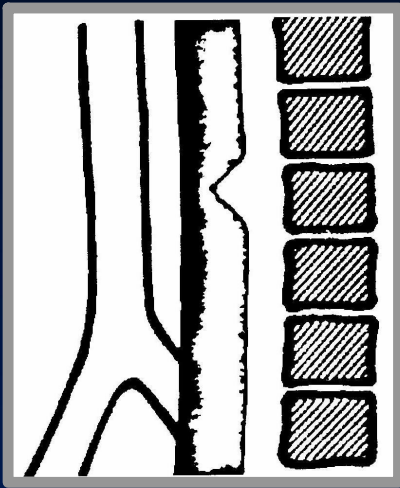
R Ao Arch with Aberrant LSCA + L ductus
L Ao Arch with Aberrant RSCA + R ductus

Posterior trachea and Anterior Esophagus

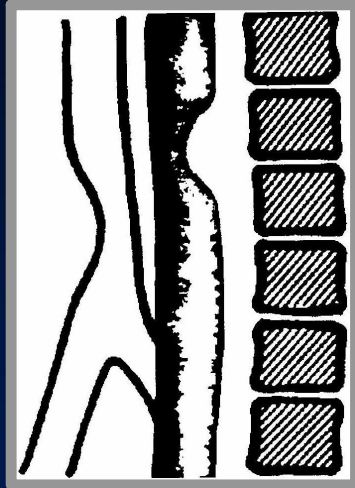


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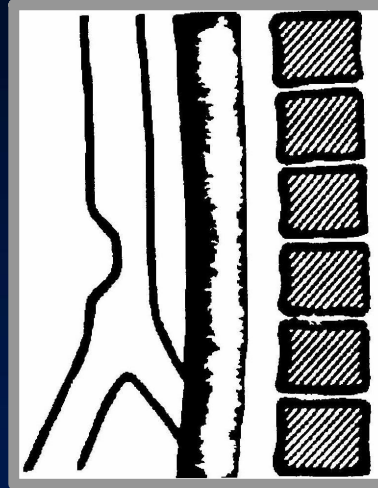
Pulmonary Sling



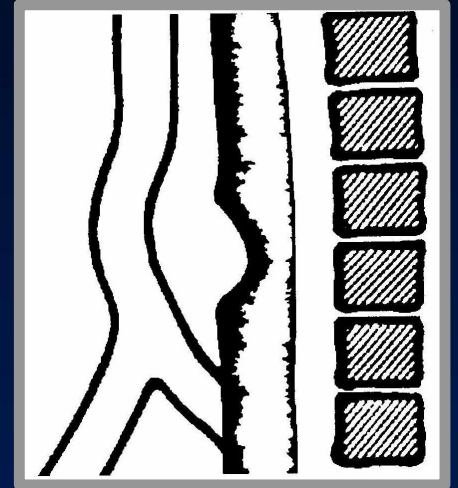
Aberrant SCA



Double Ao Arch



Isolated Anomalies



Pulmonary Sling

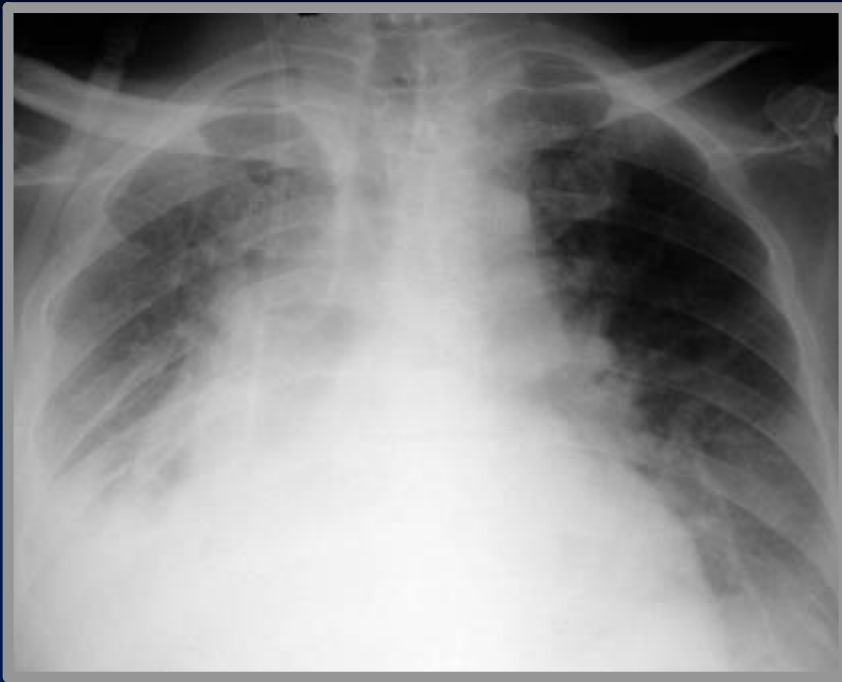
© Dahmert Lippincott Williams & Wilkins

Venous Anomalies

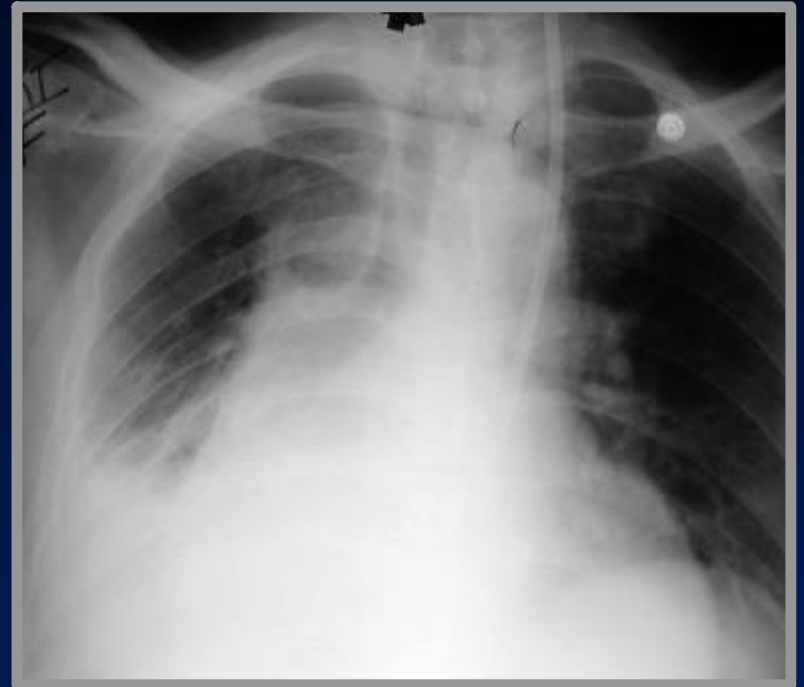
Persistent Left SVC

Persistent Left SVC

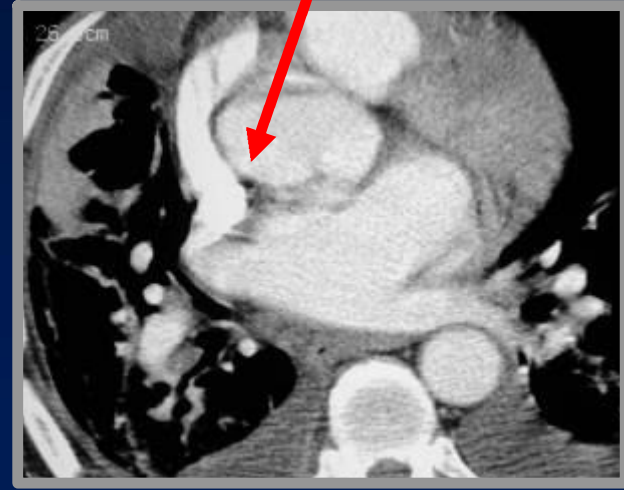
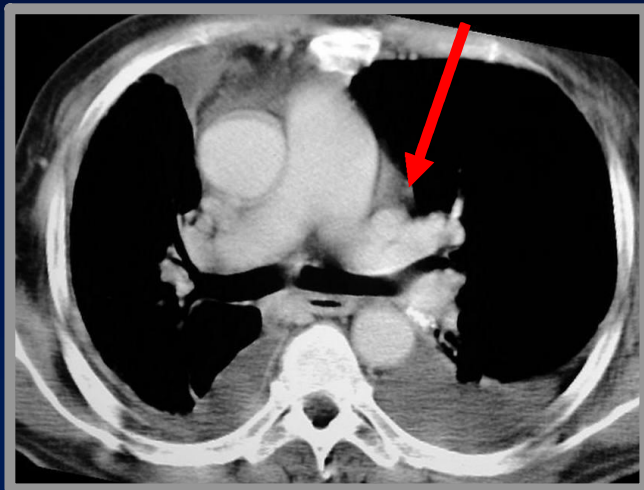
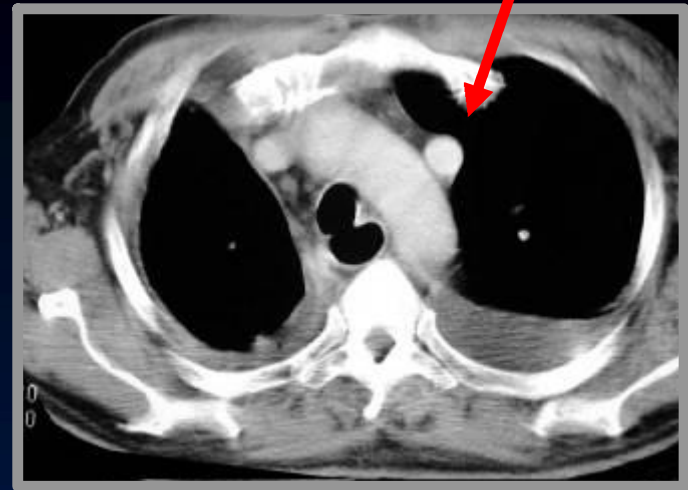
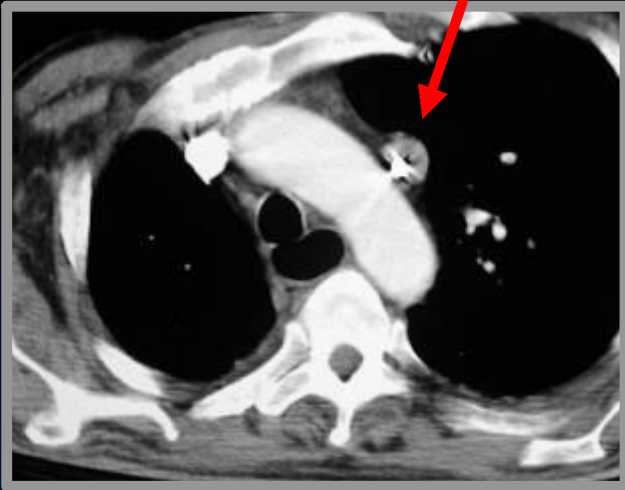
- Occurs in less than 0.5% of people
 - Failure of regression of L common and Ant. Cardinal veins
- Drains left jugular and left subclavian v
- Most patients also have right-sided SVC
- Drains into dilated coronary sinus → R atrium



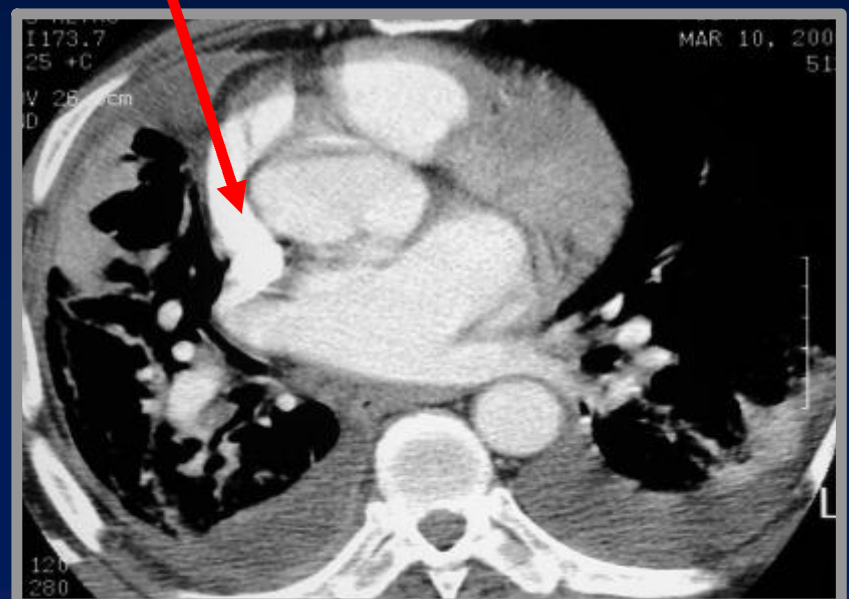
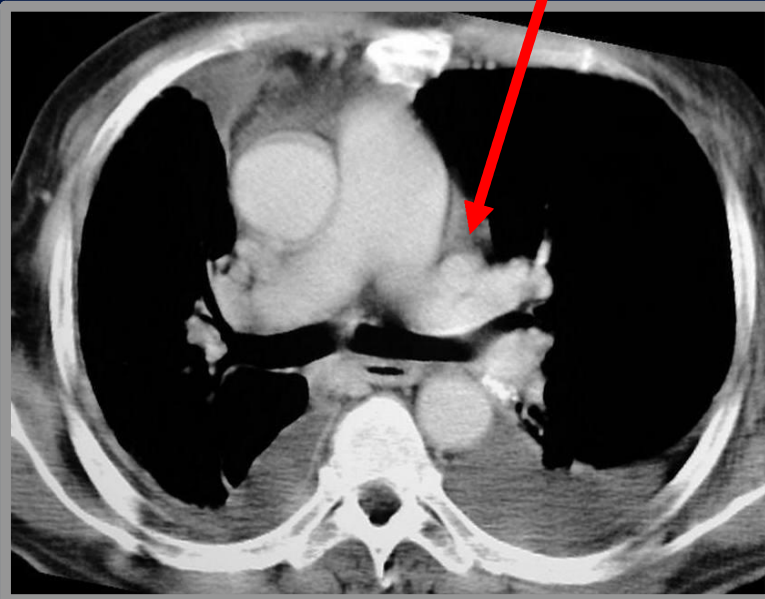
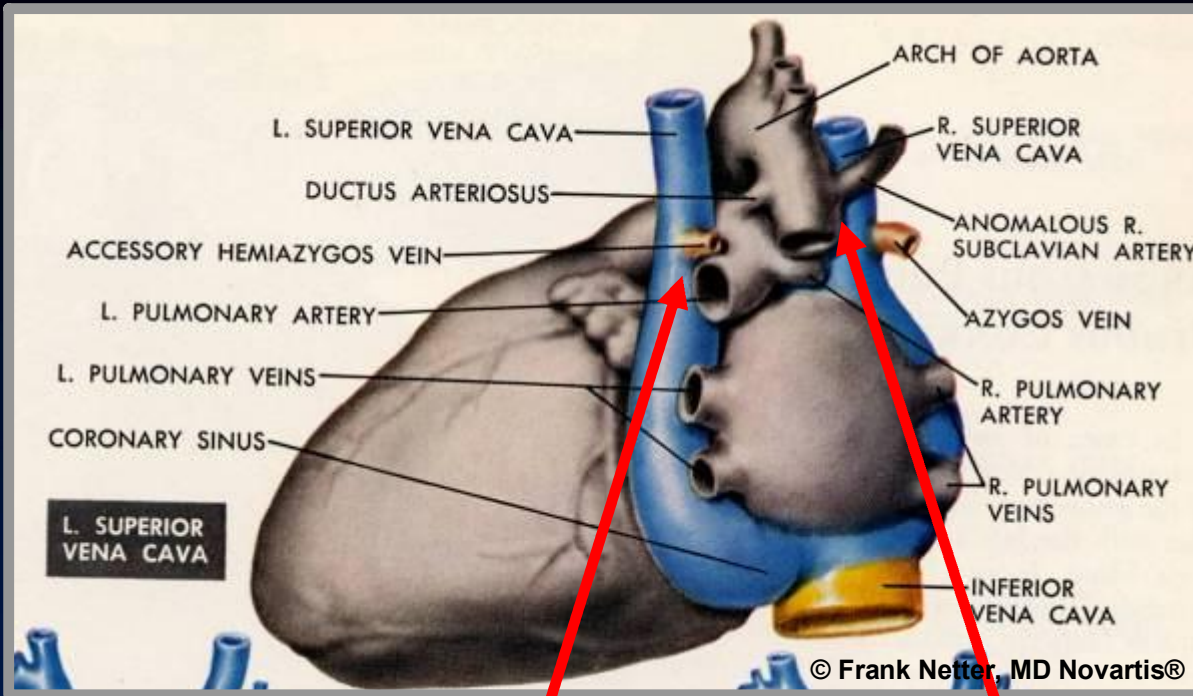
Post-op day 3



Post-op day 6



Persistent Left SVC



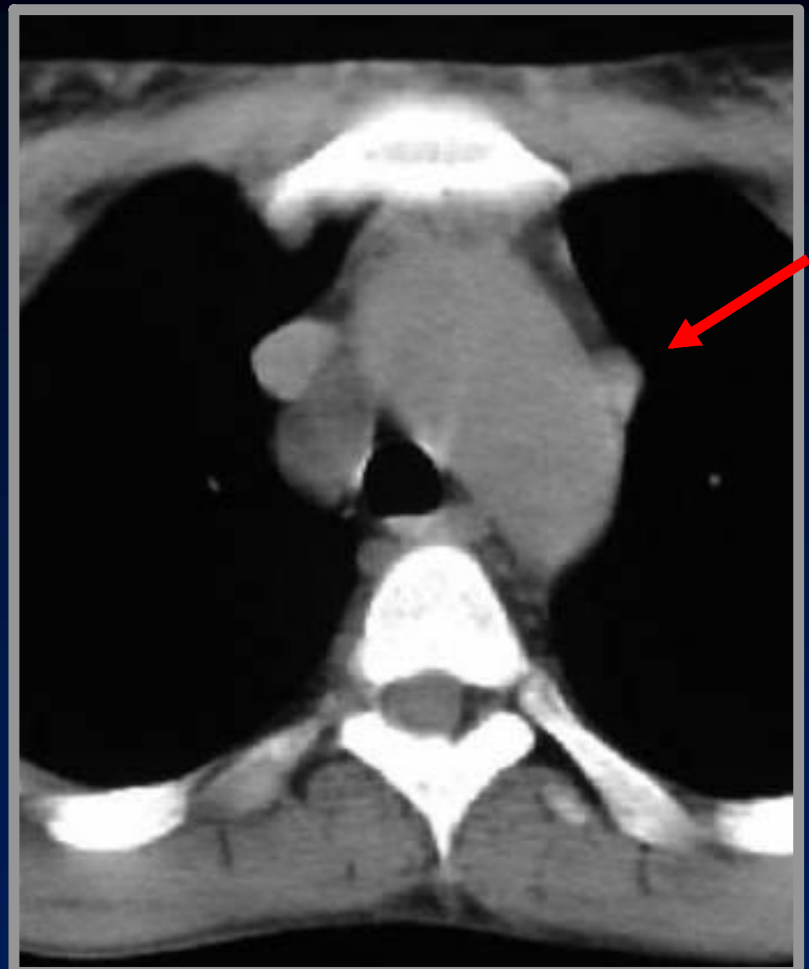
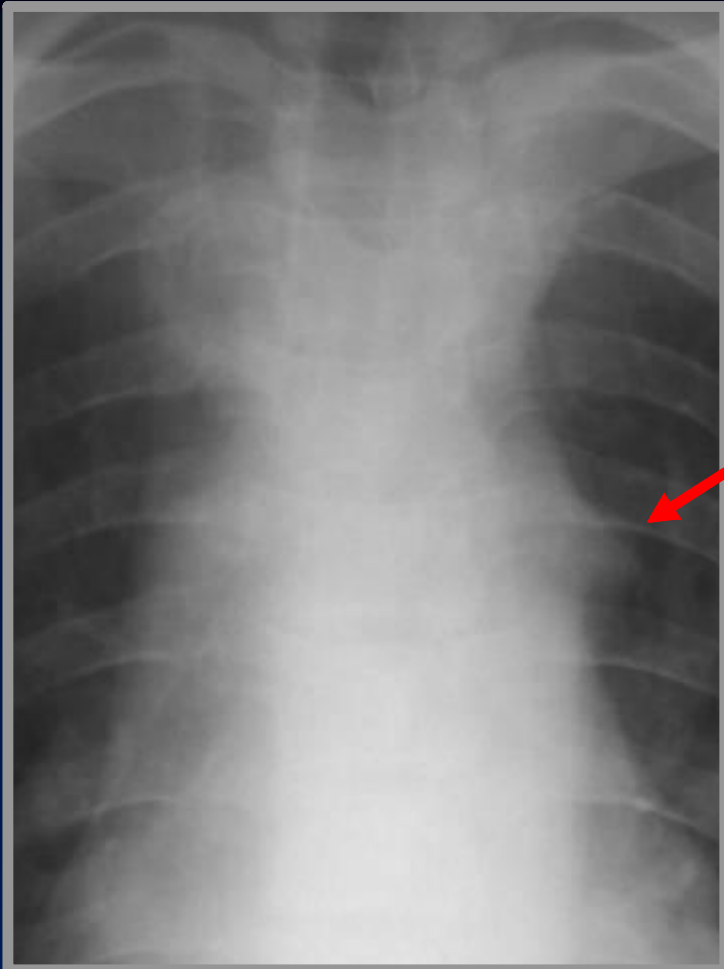
Diseases of the Not-So-Great Vessels

**Left Superior
Intercostal Vein
Aortic Nipple**

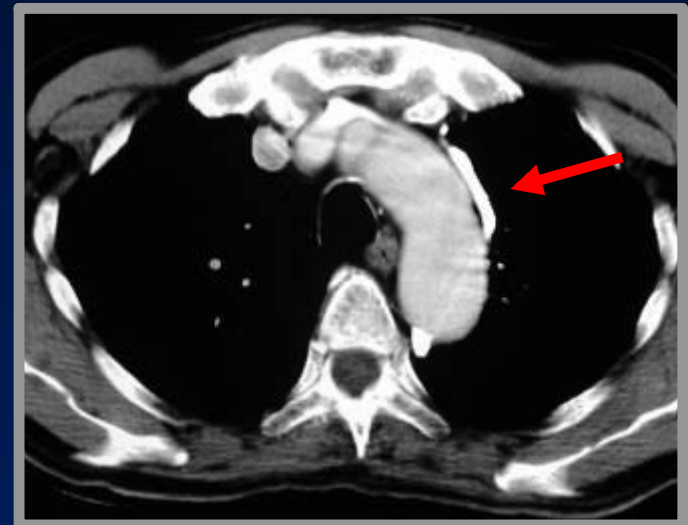
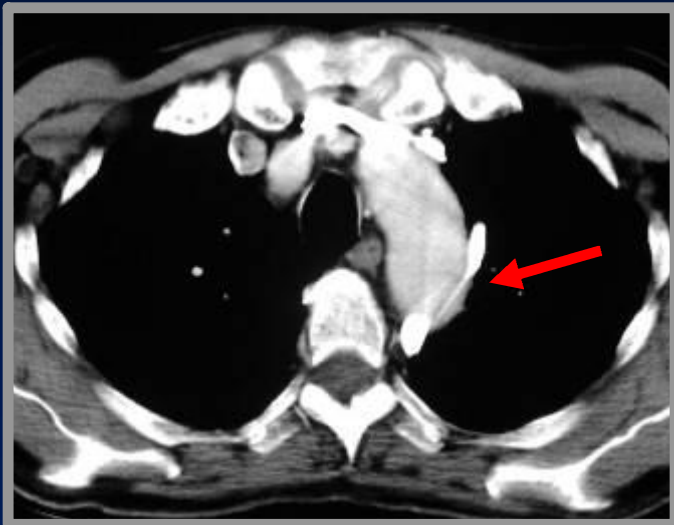
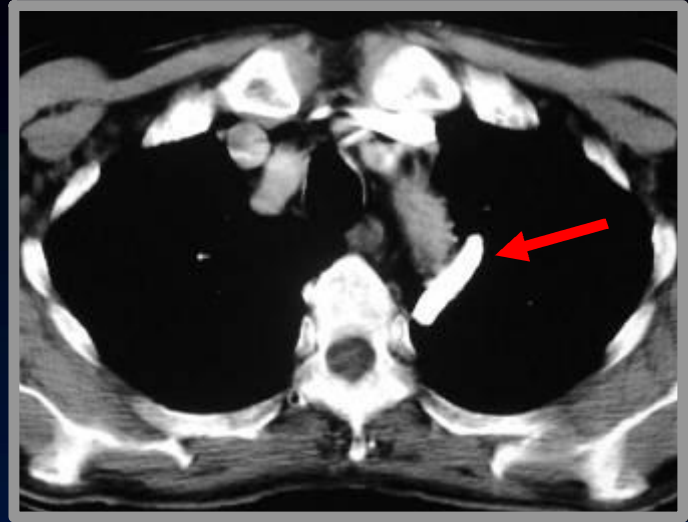
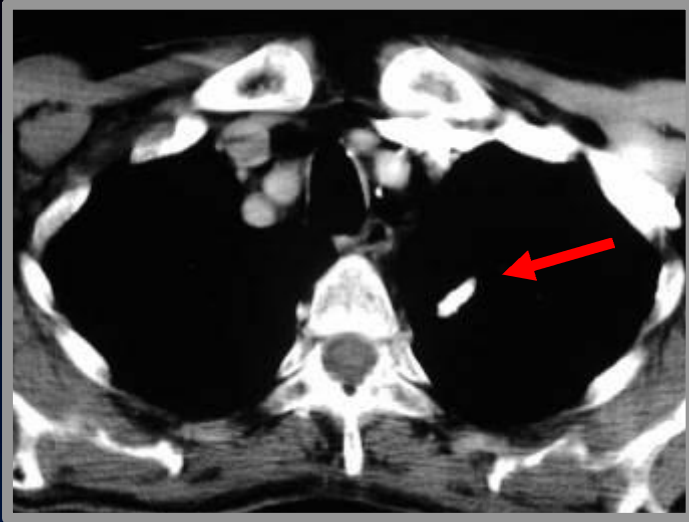
Left Superior Intercostal Vein

The Aortic Nipple

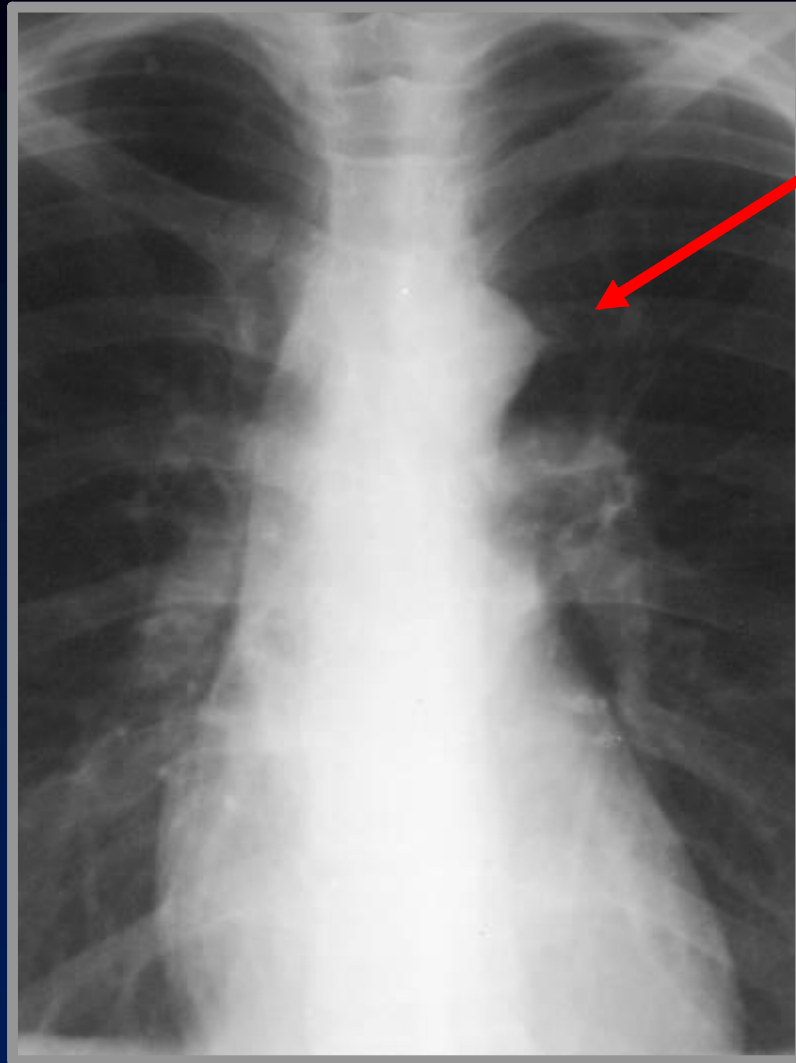
- **Visible in 5% of people**
- **Should not be mistaken for mass**



**Aortic Nipple-Left Superior
Intercostal Vein**



Aortic Nipple-Left Superior Intercostal Vein



**Aortic Nipple-Left Superior
Intercostal Vein**