Valvular Lesions of the Heart

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Mitral Stenosis

Left Atrial Outflow Obstruction

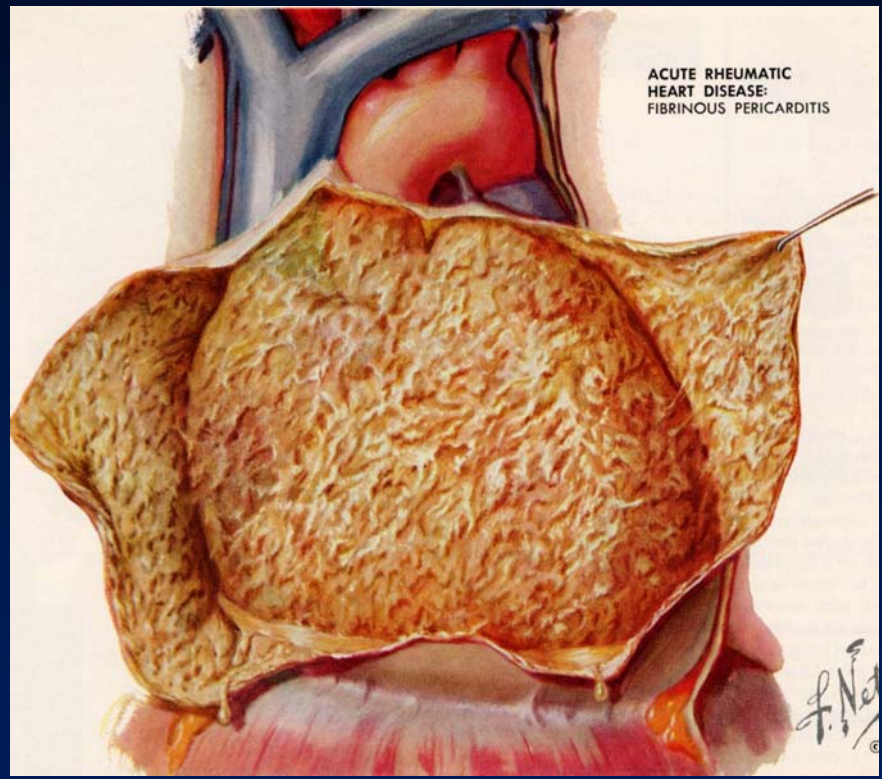
Mitral Stenosis
Rheumatic Valvular Heart Disease

- Rheumatic heart disease causes mitral stenosis in 99.8% of cases

Acute Rheumatic Valvulitis

Pathophysiology

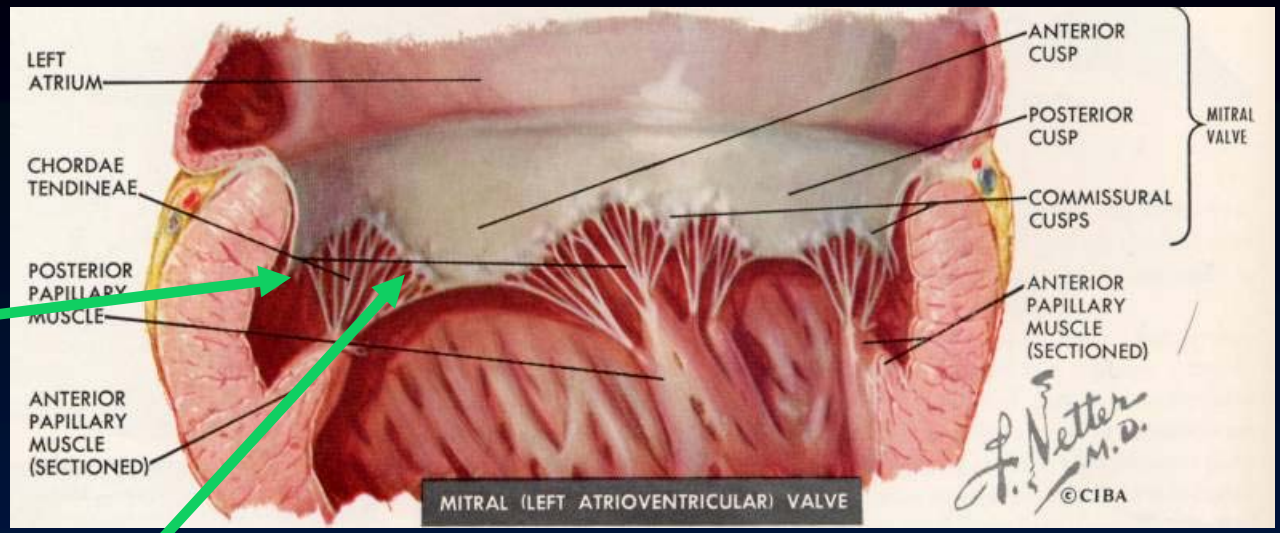
Multiple episodes of Acute Rheumatic Fever (ARF) first → pancarditis



Acute Rheumatic Valvulitis
Pathophysiology

- Acute phase subsides
  - Fibrosis alters leaflet and cusp structure
  - Results in leaflet or cuspal thickening along valvular margins of closure
- Valves affected
  - Most often mitral valve alone
  - Then most often mitral and aortic together
  - Lastly aortic alone

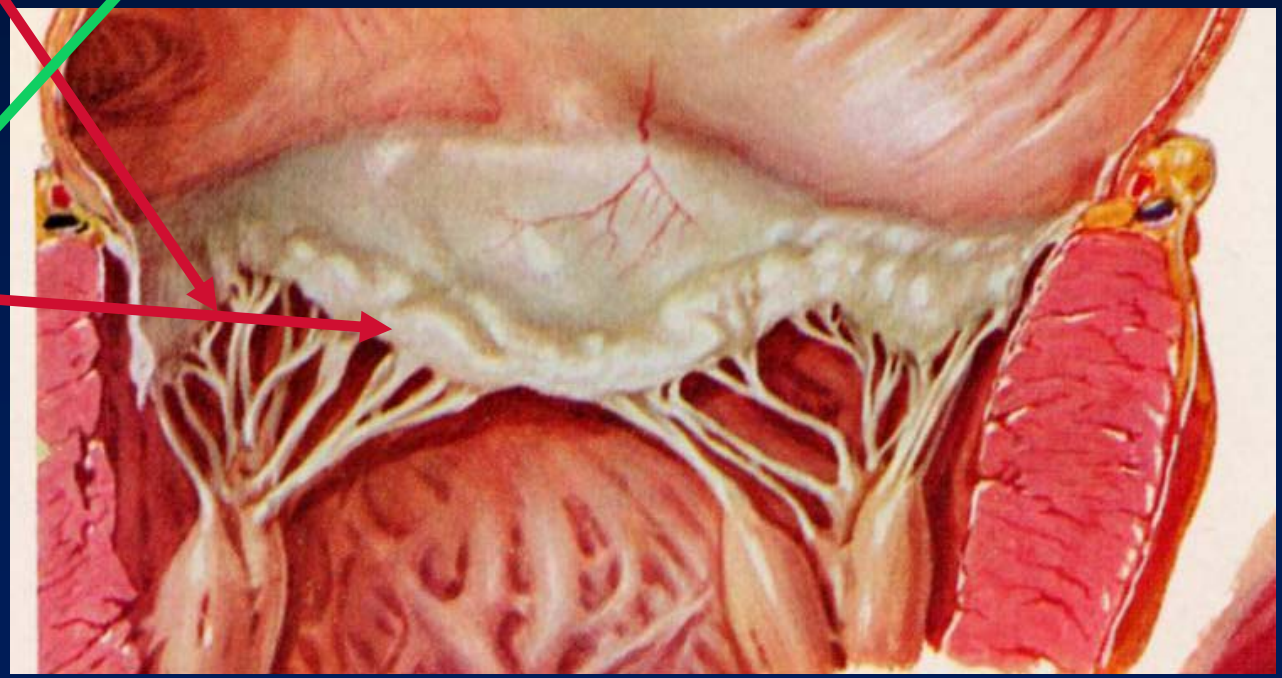
Normal mitral valve



Fusion of chordae

Stenotic mitral valve

Thickening of cusps



Chronic Mitral Stenosis
Pathophysiology

- Mitral orifice becomes smaller ➜
  - Two circulatory changes
    - To maintain LV filling across narrowed valve, left atrial pressure ↑
    - Blood flow across mitral valve is ↓ which ➜ to ↓ cardiac output

Effects of Mitral Stenosis

- On heart
- On lungs
- On right ventricle

Effect of Mitral Stenosis
On Heart

- **Left atrium** hypertrophies and dilates 2° ↑ pressure
  - Atrial fibrillation and mural thrombosis follow
- **Left ventricle** is "protected" by stenotic mitral valve
  - LV usually normal in size and contour

Effect of Mitral Stenosis
On Heart

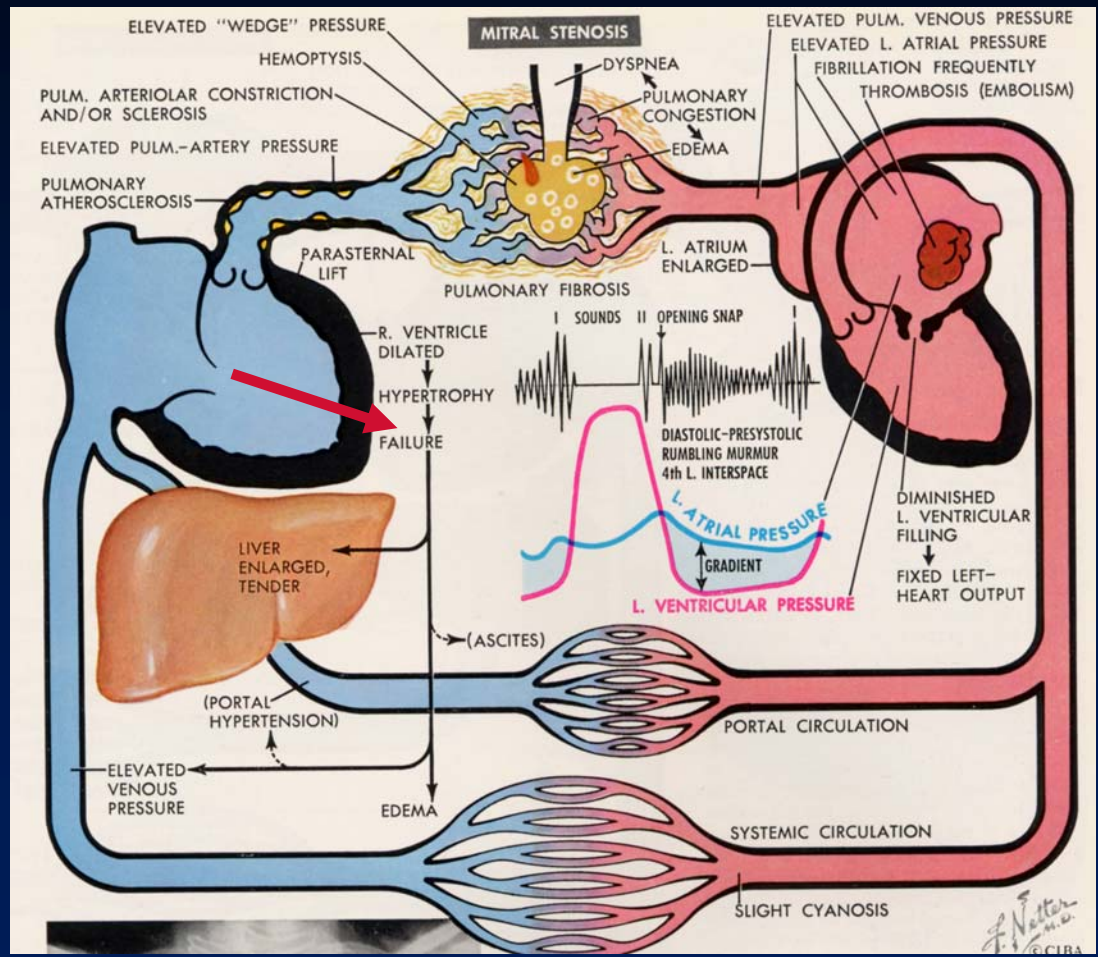
- Pulmonary arterial pressure ↑
  - Intimal and medial hypertrophy of pulmonary arteries ➜ ↑ pulmonary vascular resistance
- Right ventricle dilates from pressure overload
  - Main pulmonary artery dilates ➜ pulmonary valve regurgitation

Effect of Mitral Stenosis
On Heart

- Tricuspid regurgitation develops
  - 2° dilated RV
- Right atrium dilates 2° volume overload
  - Right heart failure

Time course of MS in adult

- Mitral stenosis occurs
- Left atrial pressure ↑
- Left atrium enlarges
- Cephalization
- PIE
- PAH develops
- PVR increases
- RV enlarges
- Pulmonic regurg develops
- Tricuspid annulus dilates
- Tricuspid insufficiency
- RV failure



Effect of Mitral Stenosis
On Lungs

- Pulmonary arterial hypertension develops
  - First passively
- Then 2° muscular hypertrophy and hyperplasia → increased pulmonary vascular resistance

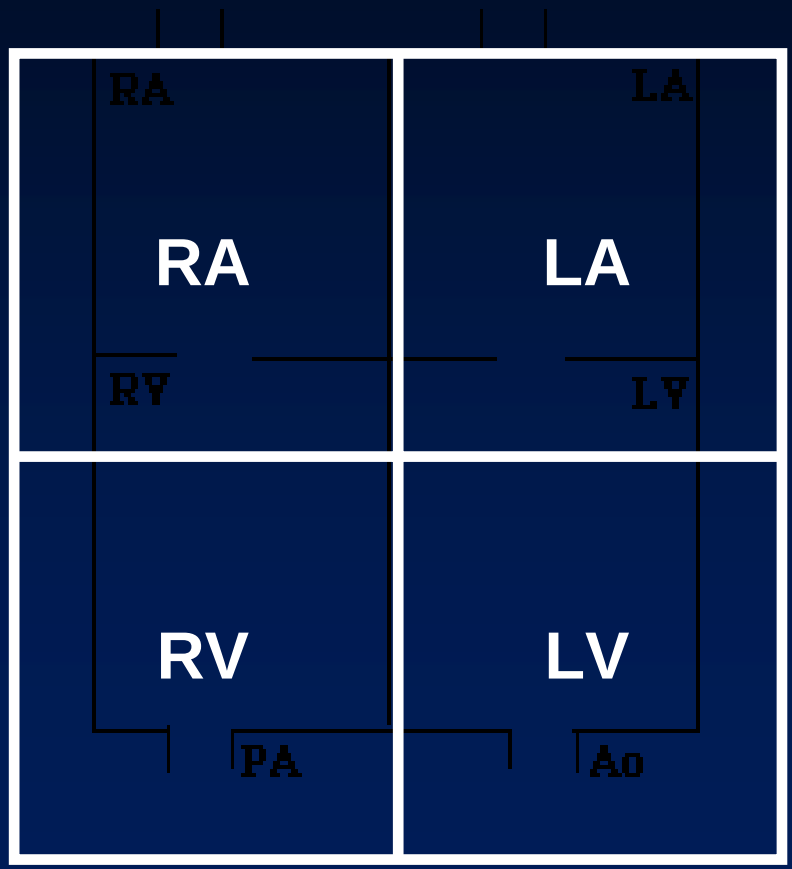
Effect of Mitral Stenosis
On Lungs

- Chronic edema of alveolar walls → fibrosis
  - Pulmonary hemosiderin deposited in lungs
  - Pulmonary ossification may occur

Effect of Mitral Stenosis on Lungs
Normal chamber pressures

M 1-5

D 1-7
S 17-32
M 15



M 5-10

D 5-12
S 90-140

Effect of Mitral Stenosis
On Lungs

- ↑ pulmonary venous and capillary pressure

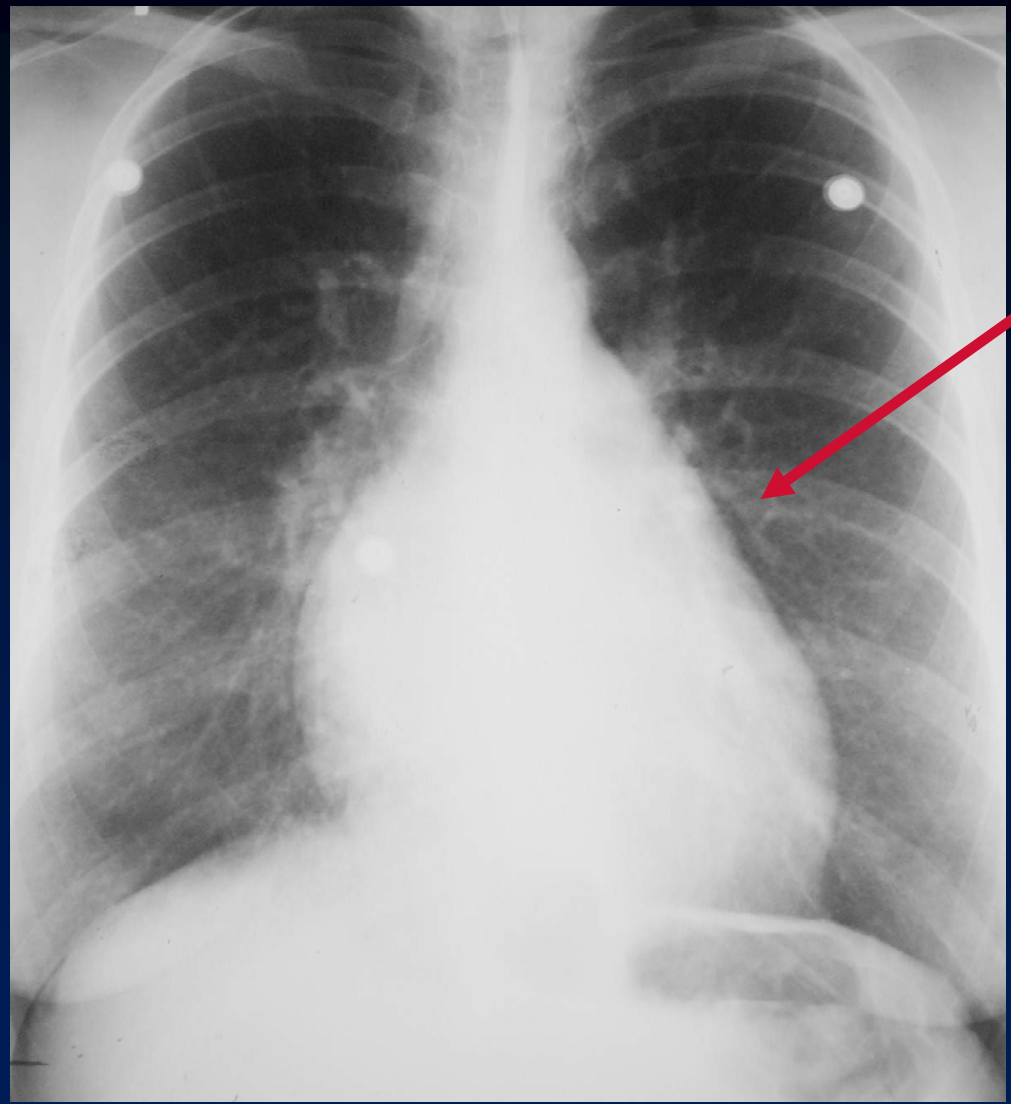
Normal	5-10 mm Hg
Cephalization	10-15 mm
Kerley B Lines	15-20
Pulmonary Interstitial Edema	20-25
Pulmonary Alveolar Edema	> 25

Effect of Mitral Stenosis
On Right Ventricle

- RV hypertrophies in response to increased afterload
- Eventually RV fails and dilates
  - Causes dilation of tricuspid annulus ➜ tricuspid regurgitation

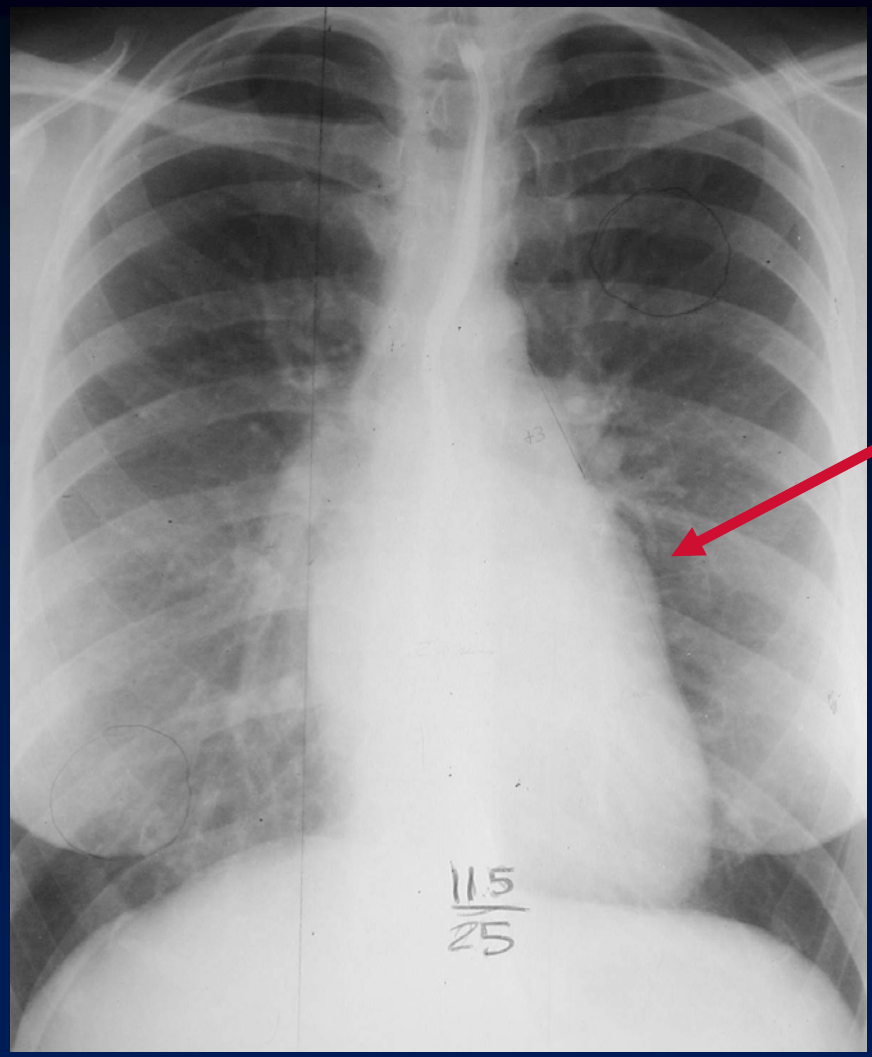
X-Ray Findings of MS
Cardiac Findings

- Usually normal or slightly enlarged heart
  - Enlarged atria do not produce cardiac enlargement; only enlarged ventricles
- Straightening of left heart border
- Or, convexity along left heart border 2° to enlarged atrial appendage
  - Only in rheumatic heart disease



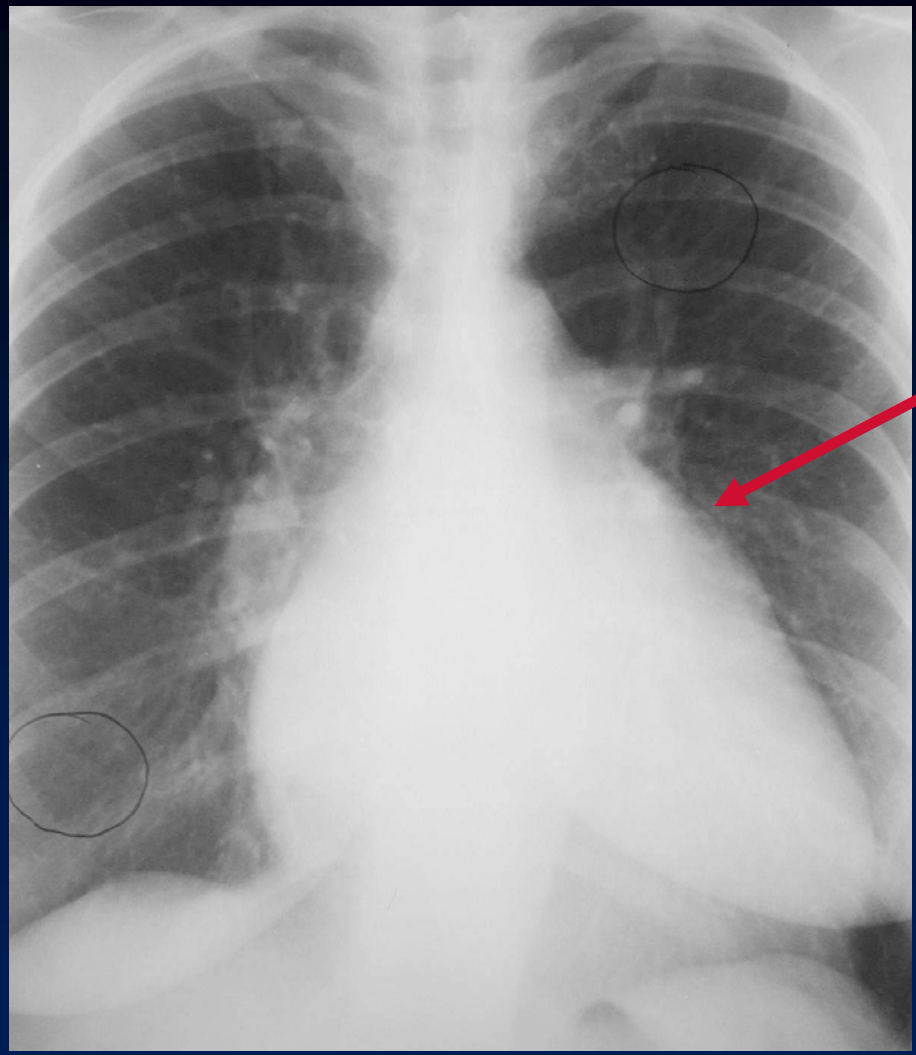
“Straightening” of left heart border

Mitral Stenosis



Convexity from enlarged left atrial appendage

Mitral Stenosis

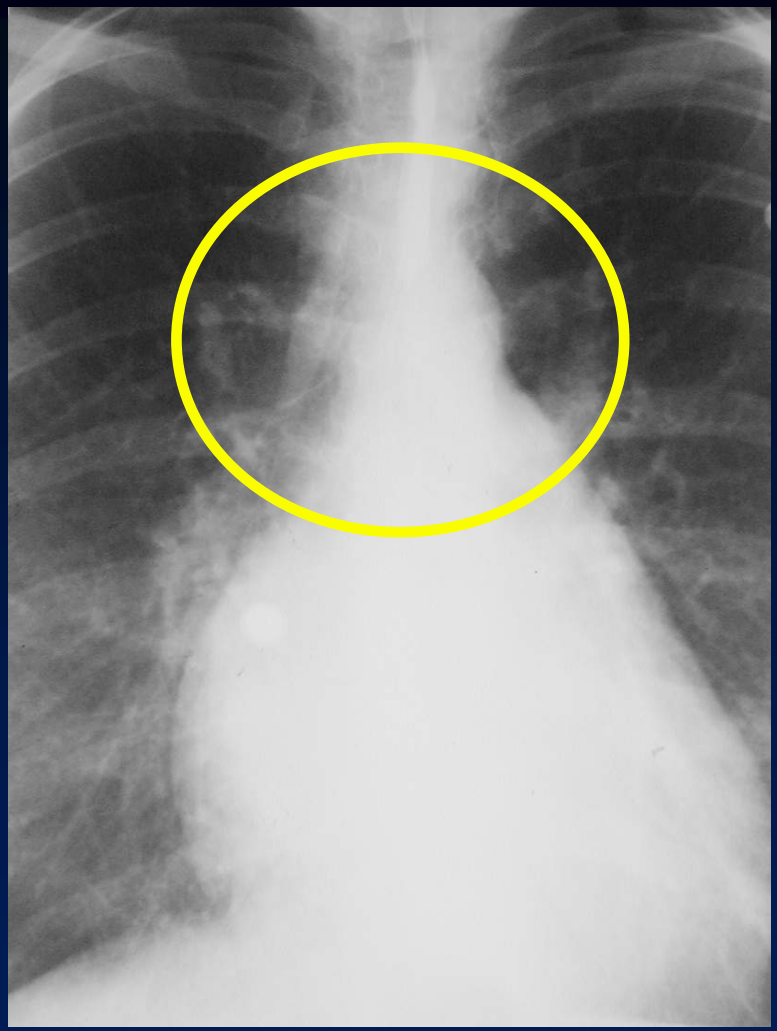


Convexity from enlarged left atrial appendage

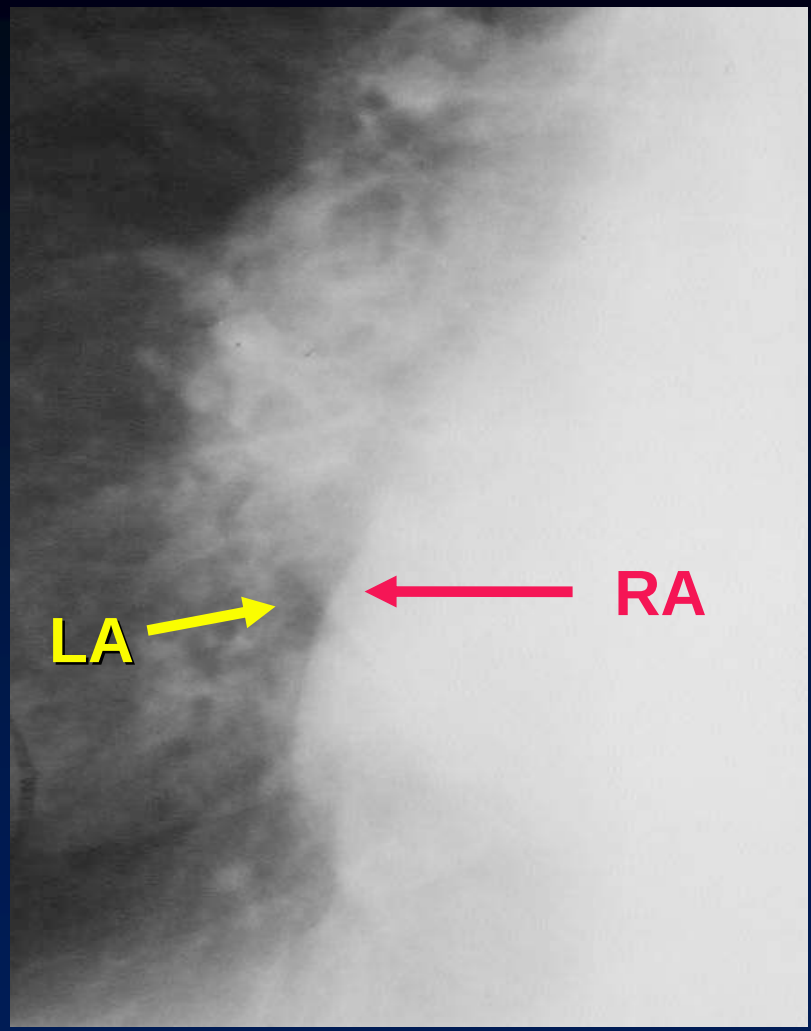
Mitral Stenosis

X-Ray Findings of MS
Cardiac Findings

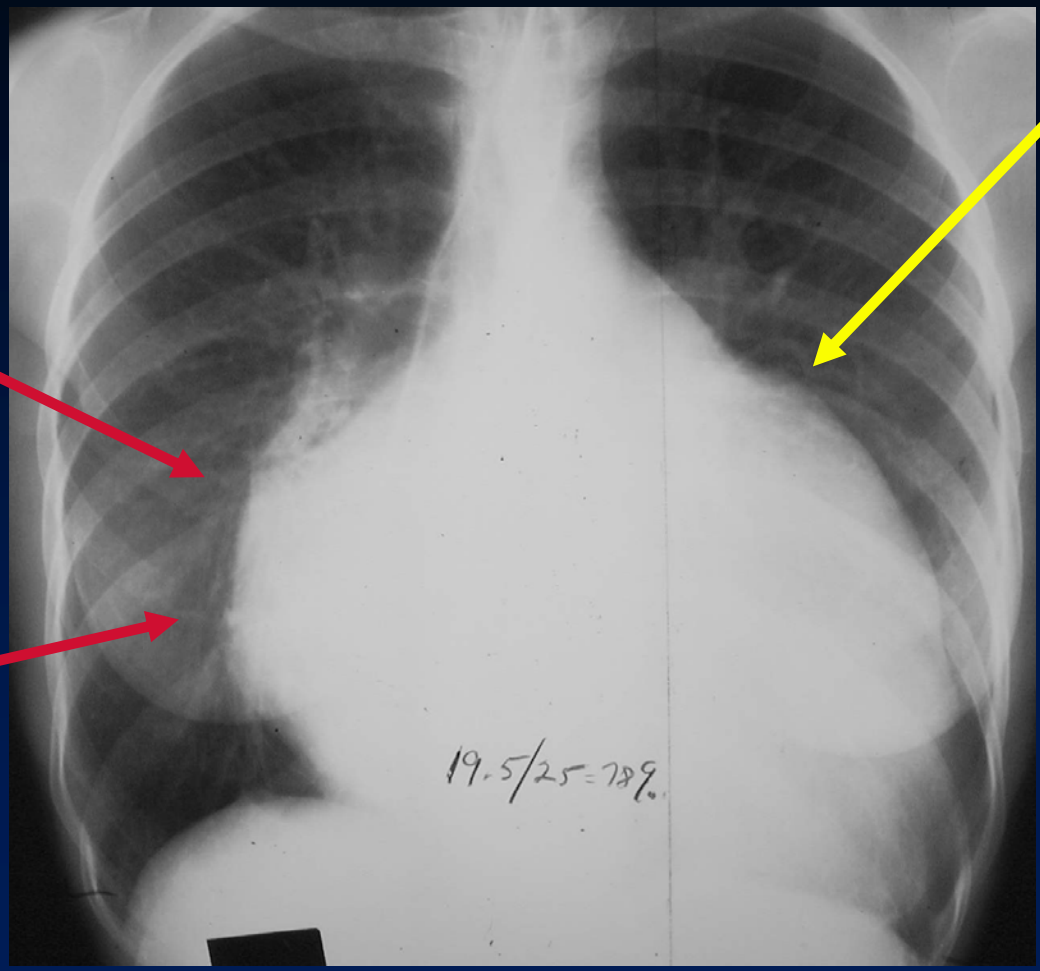
- Small aortic knob from decreased cardiac output
- Double density of left atrial enlargement
- Rarely, right atrial enlargement from tricuspid insufficiency



Small aorta from ↓ cardiac output



“Double density”



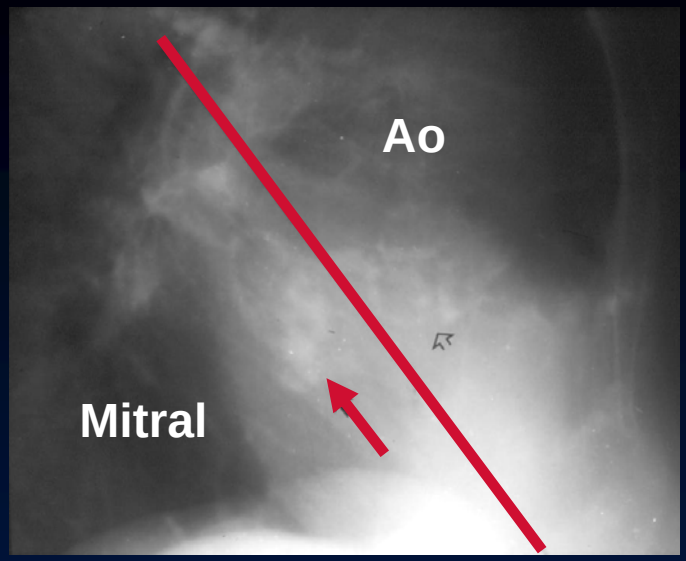
Enlarged L atrial appendage from mitral stenosis

Right atrial enlargement from tricuspid regurgitation

Mitral stenosis/regurgitation with tricuspid regurgitation

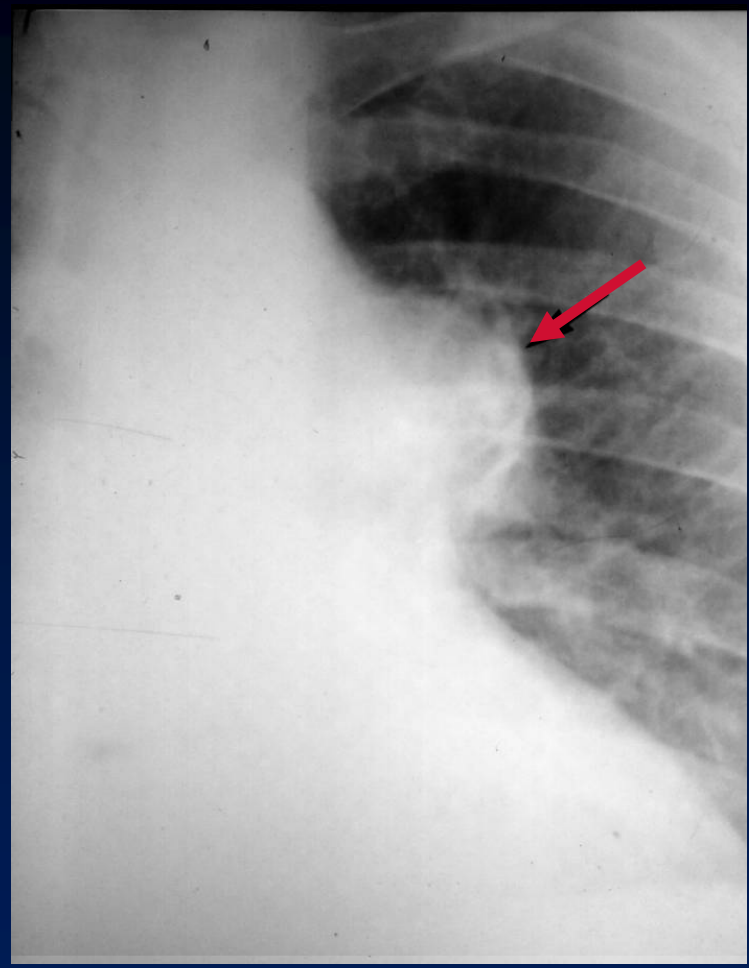
X-Ray Findings of MS
Calcifications

- Calcification of valve--not annulus--seen best on lateral film and at angio
- Rarely, calcification of left atrial wall 2° fibrosis from long-standing disease
- Rarely, calcification of pulmonary arteries from PAH



Calcification of mitral valve

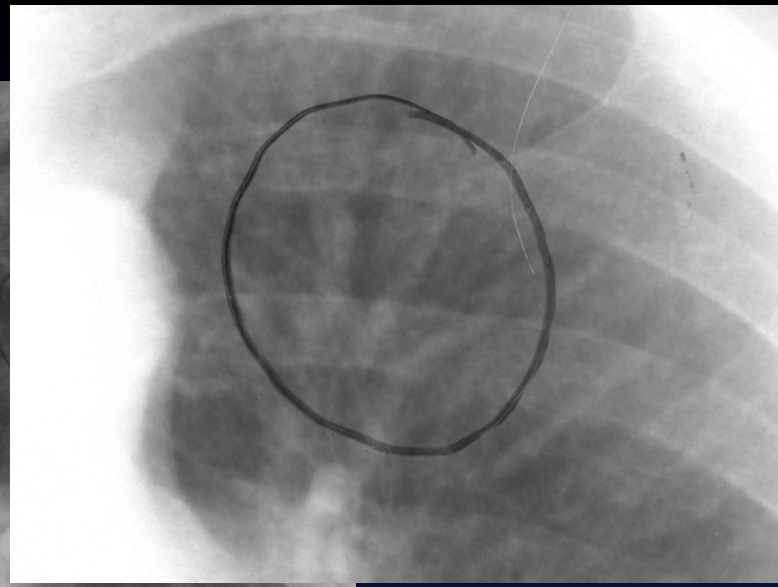
Calcification of left atrial wall

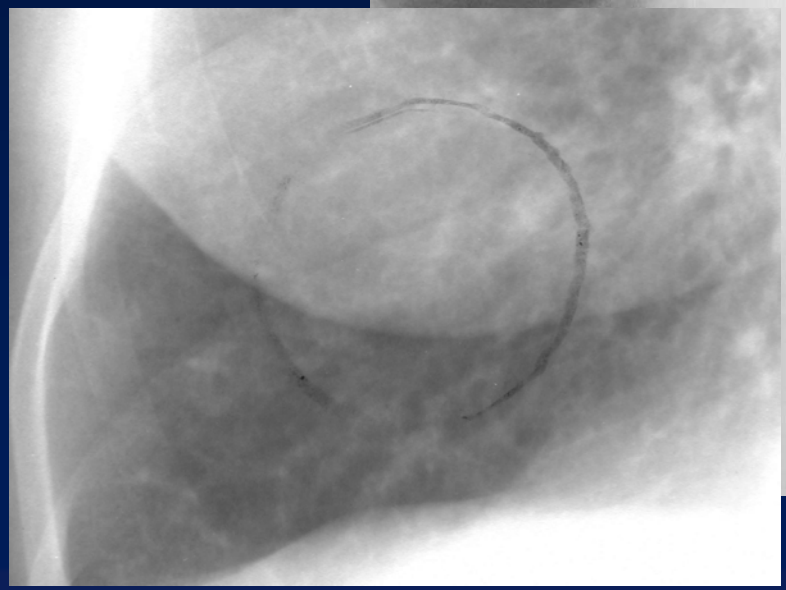


Calcification of pulmonary artery

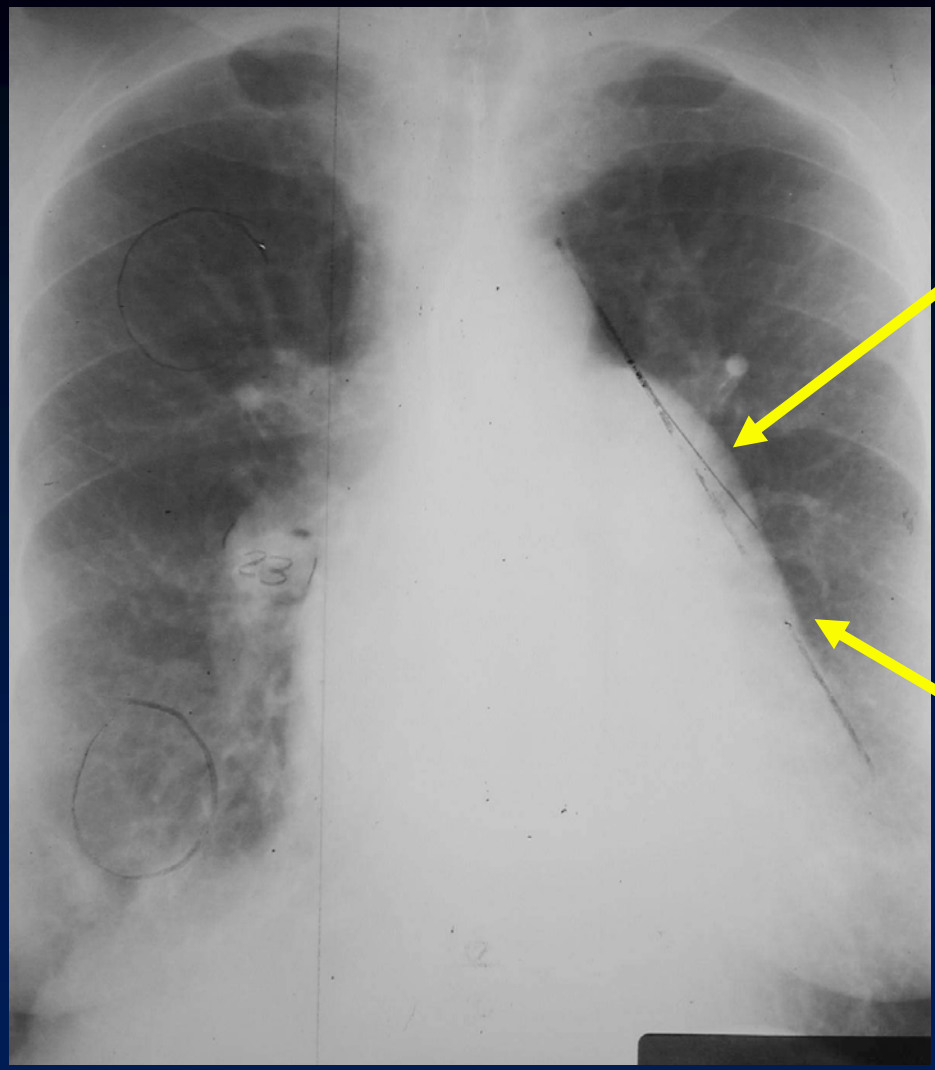
X-Ray Findings of MS
Pulmonary Findings

- Cephalization
- Elevation of left mainstem bronchus (especially if 90° to trachea)
- Enlargement of main pulmonary artery 2° pulmonary arterial hypertension
  - Severe, chronic disease
- Multiple small hemorrhages in lung
  - Pulmonary hemosiderosis





Upper lobe vessels equal to or larger than size of lower lobe vessels = Cephalization



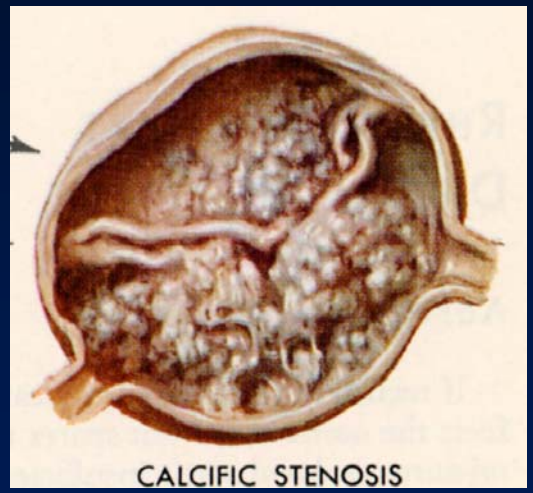
Enlarged MPA segment from severe pulmonary arterial hypertension

Straightening of left heart border from ↑ LA

Mitral Stenosis with severe PAH

Mitral Valve Calcification

- Presence indicates MS
- Calcium usually deposited in clumps on valve leaflets
- Heavier calcific deposits in men than women



Mitral Annulus Calcification

- Calcification of mitral annulus does not signify presence of mitral valve disease
  - Occurs in older women
  - Usually asymptomatic
  - Rarely → Mitral Stenosis

Mitral Stenosis
Other Causes

- MS 2° rheumatic disease 99.8% of cases
  - Congenital mitral stenosis
  - Infective endocarditis
  - Carcinoid syndrome
  - Fabray's Disease
  - Hurler's syndrome
  - Whipple's Disease
  - Left atrial myxoma

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

LA Myxoma

- Most common form of primary cardiac tumor
- 86% of myxomas found in left atrium
- 90% of myxomas are solitary
- Usually occur around fossa ovalis

MS and MR

- Rheumatic mitral stenosis occurs with varying degrees of mitral regurgitation
- When MS is severe, MR is relatively unimportant

Mitral Regurgitation

Mitral Regurgitation
Causes

- Thickening of valve leaflets 2° rheumatic disease
- Rupture of the chordae
  - Posterior leaflet more often-Trauma, Marfan's
- Papillary muscle rupture or dysfunction
  - Acute myocardial infarction
- LV enlargement → dilatation of mitral annulus
  - Any cause of LV enlargement
- LV aneurysm → valvular dysfunction
  - Acute myocardial infarction

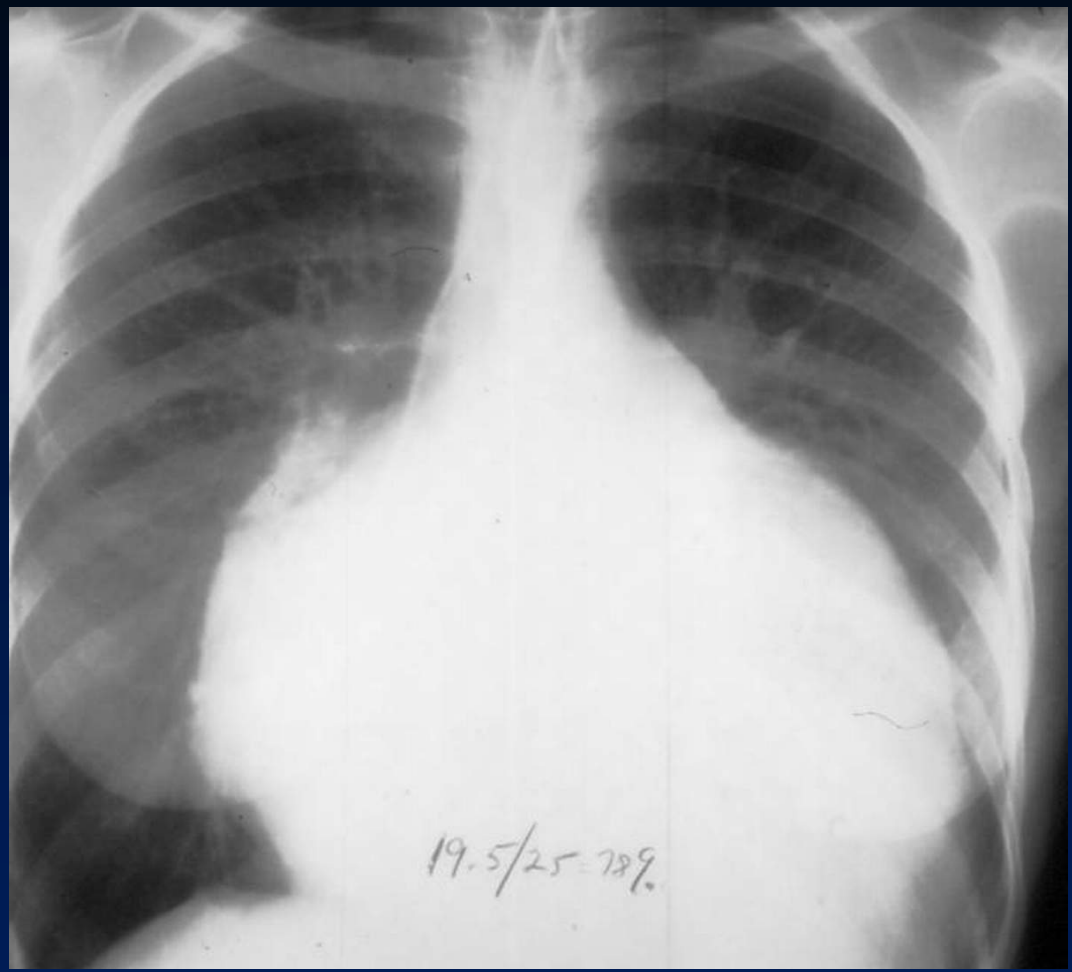
Mitral Regurgitation
General

- The acute lesion of rheumatic fever is mitral regurgitation, not stenosis
- The largest left atria *ever* are produced by mitral regurgitation, not mitral stenosis

Mitral Regurgitation
X-ray Findings

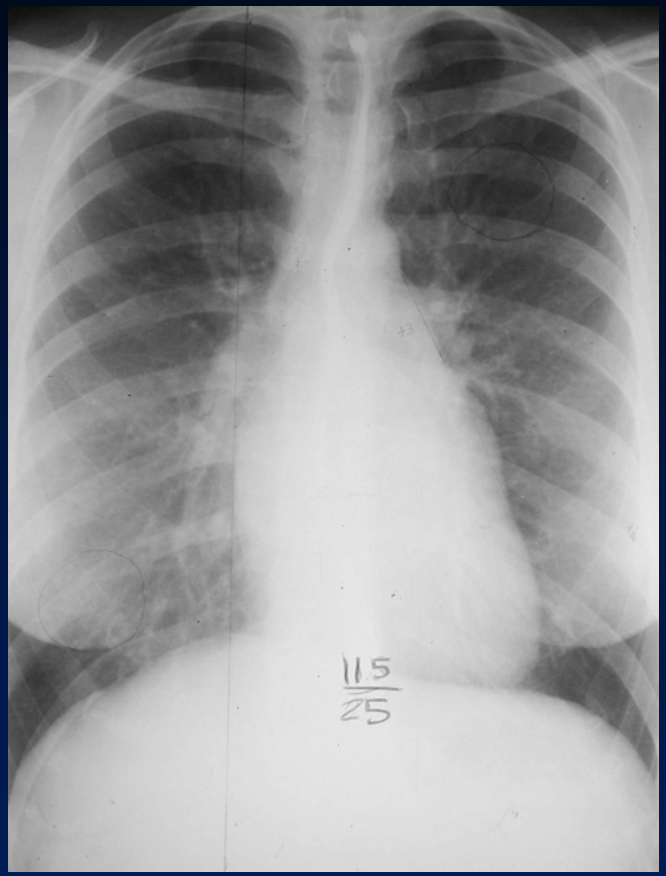
- In acute MR
  - Pulmonary edema
  - Heart is not enlarged
- In chronic MR
  - LA and LV are markedly enlarged
    - Volume overload
  - Pulmonary vasculature is usually normal
    - LA volume but not pressure is elevated

Mitral regurgitation



Mitral regurgitation

Difference in heart size – MS and MR



Mitral Stenosis

Mitral Regurgitation

Aortic Stenosis

Aortic Stenosis
Frequency of Causes

- Most often as result of degeneration of bicuspid aortic valve
- Less commonly, 2° to degeneration of tricuspid aortic valve in person > 65
- Even less commonly, 2° rheumatic heart disease in tricuspid valve

Aortic Stenosis
Locations

- Supravalvular
- Valvular
- Subvalvular

Valvular Aortic Stenosis Congenital

Congenital Valvular Aortic Stenosis
General

- Bicuspid aortic valve is the most common congenital cardiac anomaly
  - 0.5 –2%
- Usually not stenotic during infancy
- More prone to fibrosis and calcification than normal valve

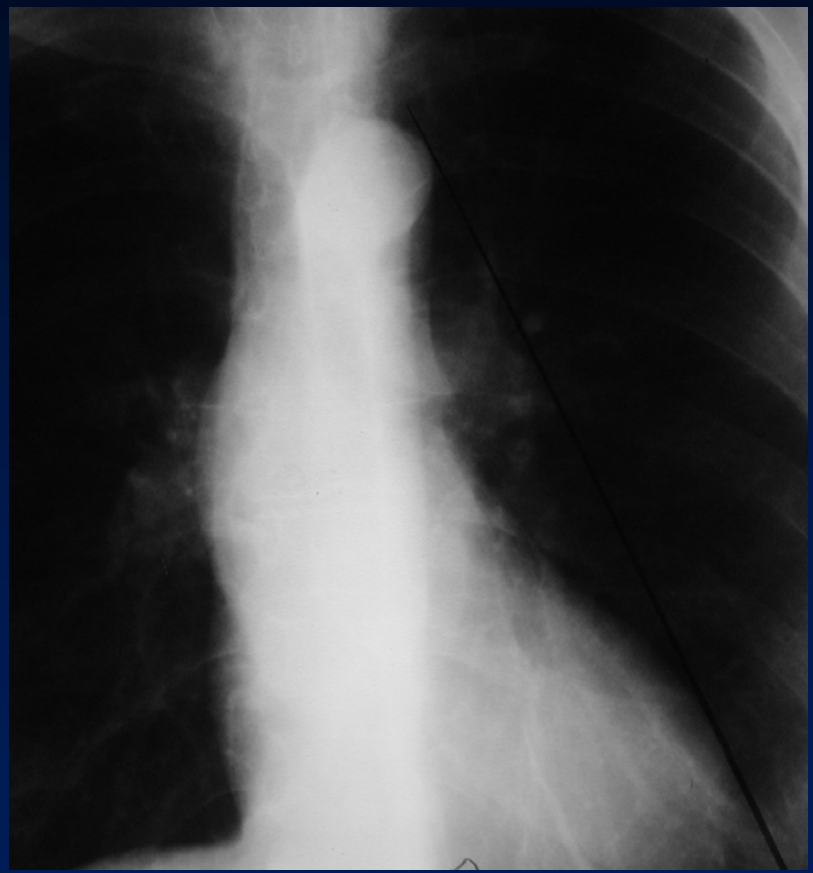
Congenital Valvular Aortic Stenosis
Associations

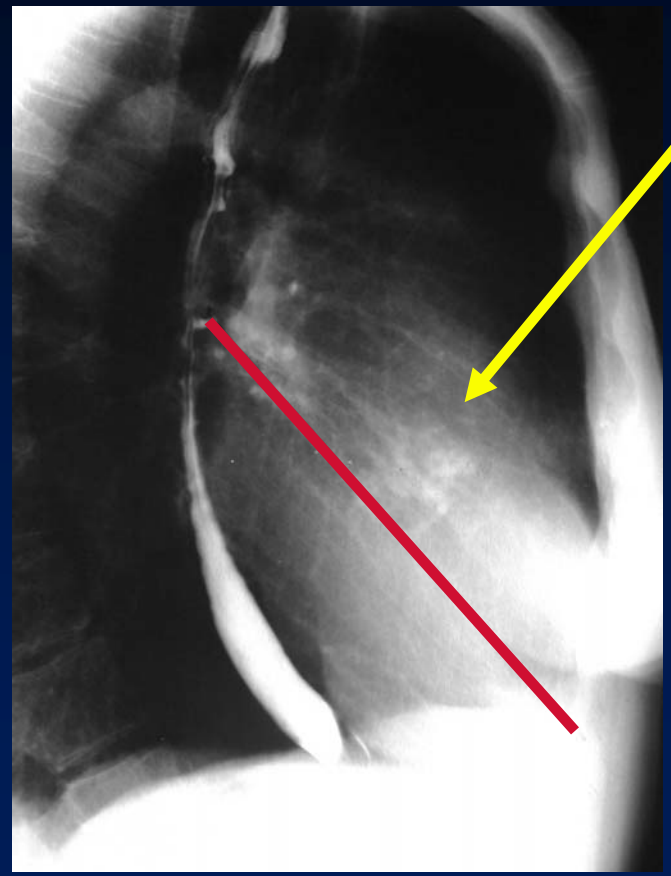
- Many malformations of aorta and/or LV are associated with bicuspid valve
  - 50% with coarctation of aorta
  - Hypoplastic left heart syndrome
  - Interruption of aortic arch

Congenital Valvular Aortic Stenosis
Calcification

- Bicuspid valves are most apt to calcify
- Calcification begins earlier (4th decade) than in degenerated tricuspid Ao valve (>65)
  - Early calcification can also occur with Rheumatic heart dz

Calcification of Aortic Valve



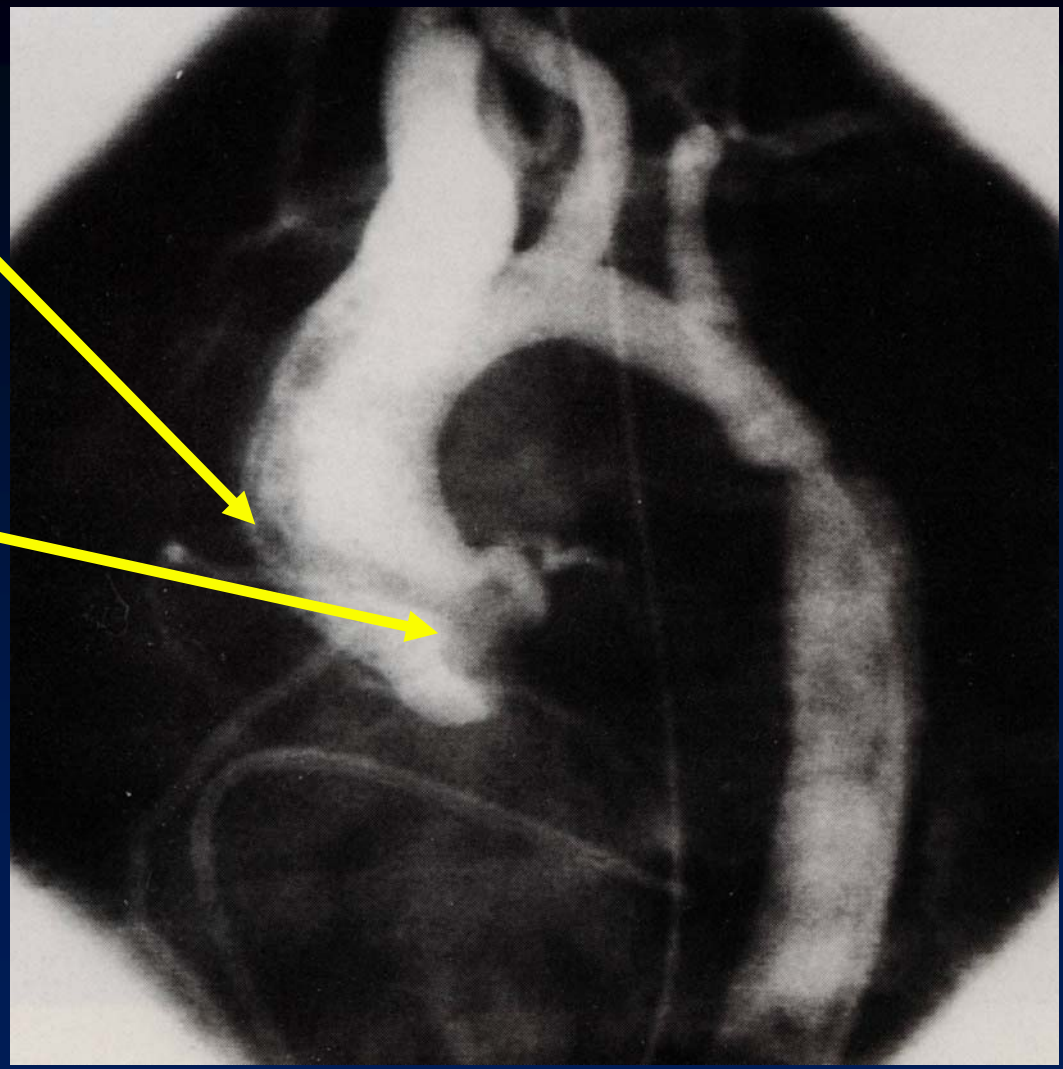


Congenital Valvular Aortic Stenosis
Angiographic findings

- A non-calcified, bicuspid valve reveals thickening and doming of valve leaflets in systole
- A jet of non-opacified blood is visible through stenotic bicuspid valve
  - Does not occur with acquired AS

Unopacified jet stream through a bicuspid aortic valve

Leaflets are “domed” on systole



Acquired aortic stenosis would not demonstrate this jet stream because severe deformity of valve ➜ turbulent flow

Congenital Valvular Aortic Stenosis
Angiographic findings

- Congenitally bicuspid valves usually have 2 aortic sinuses
  - 3 sinuses in acquired AS
- In rheumatic disease, aortic valve commissures usually fuse
  - Don’t fuse in degenerated tricuspid valve

Valvular Aortic Stenosis Acquired

Acquired Valvular Aortic Stenosis
Causes

- Fusion, thickening or calcification of a tricuspid valve
  - Degenerative process
  - Rheumatic heart disease

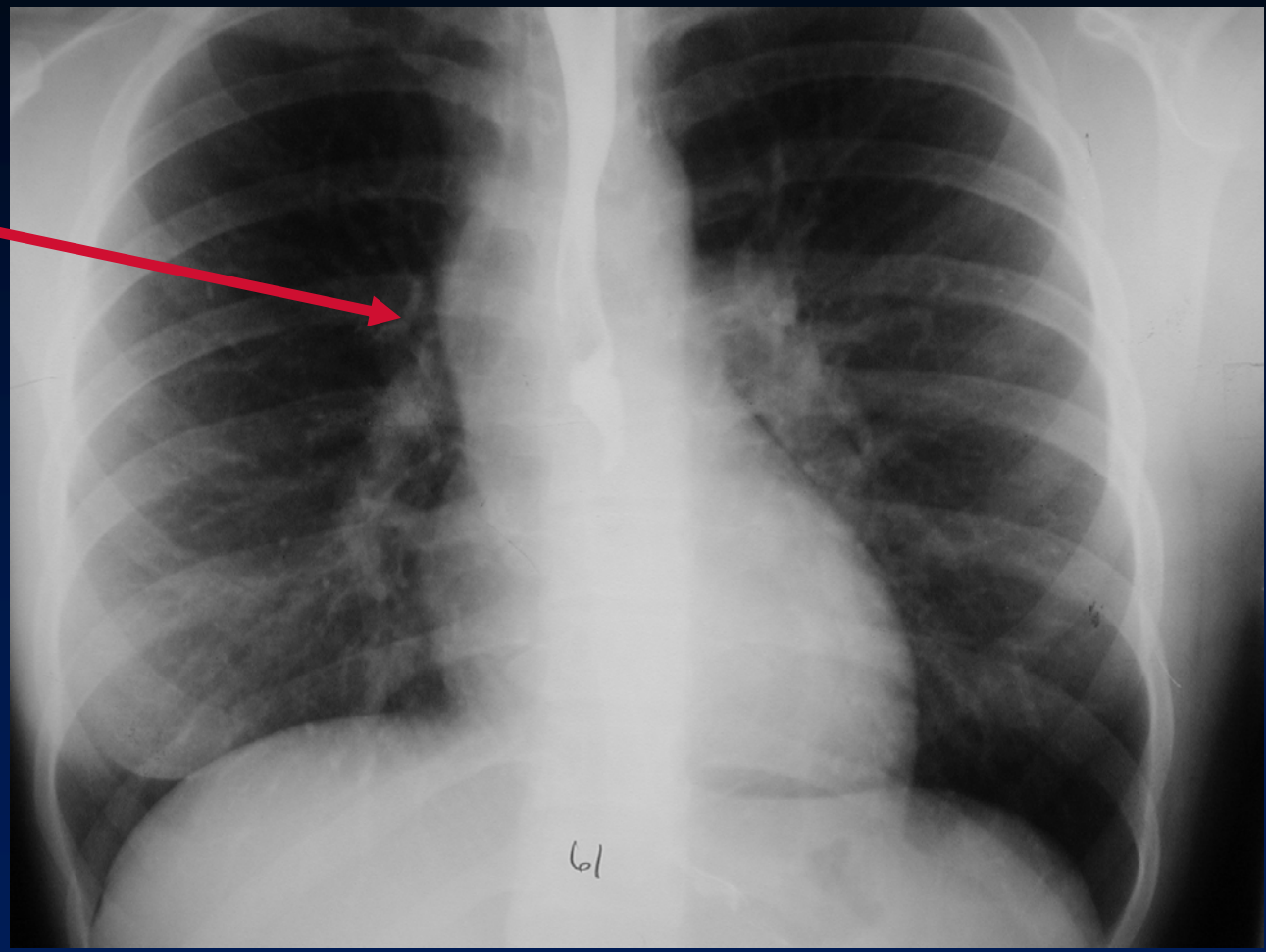
Valvular Aortic Stenosis
Differentiating Features

Etiology/Findings	Calcification	Other clues
Congenital Bicuspid Valve	30's	Jet effect on aortogram
Degeneration of Tricuspid Valve	> 65	Coronary artery ca++ Commissures don't fuse
Rheumatic dz in Tricuspid Valve	30's here; teens in 3rd world countries	MS or MR almost always present; commissures fuse

Aortic Stenosis
X-Ray Findings

- Depends on age patient/severity of disease
  - In infants, AS ➜CHF/pulmonary edema
- In adults
  - Normal heart size
    - Until cardiac muscle decompensates
  - Enlarged ascending aorta 2° post-stenotic dilatation 2° turbulent flow
  - Normal pulmonary vasculature

Prominence of ascending aorta from post-stenotic dilatation

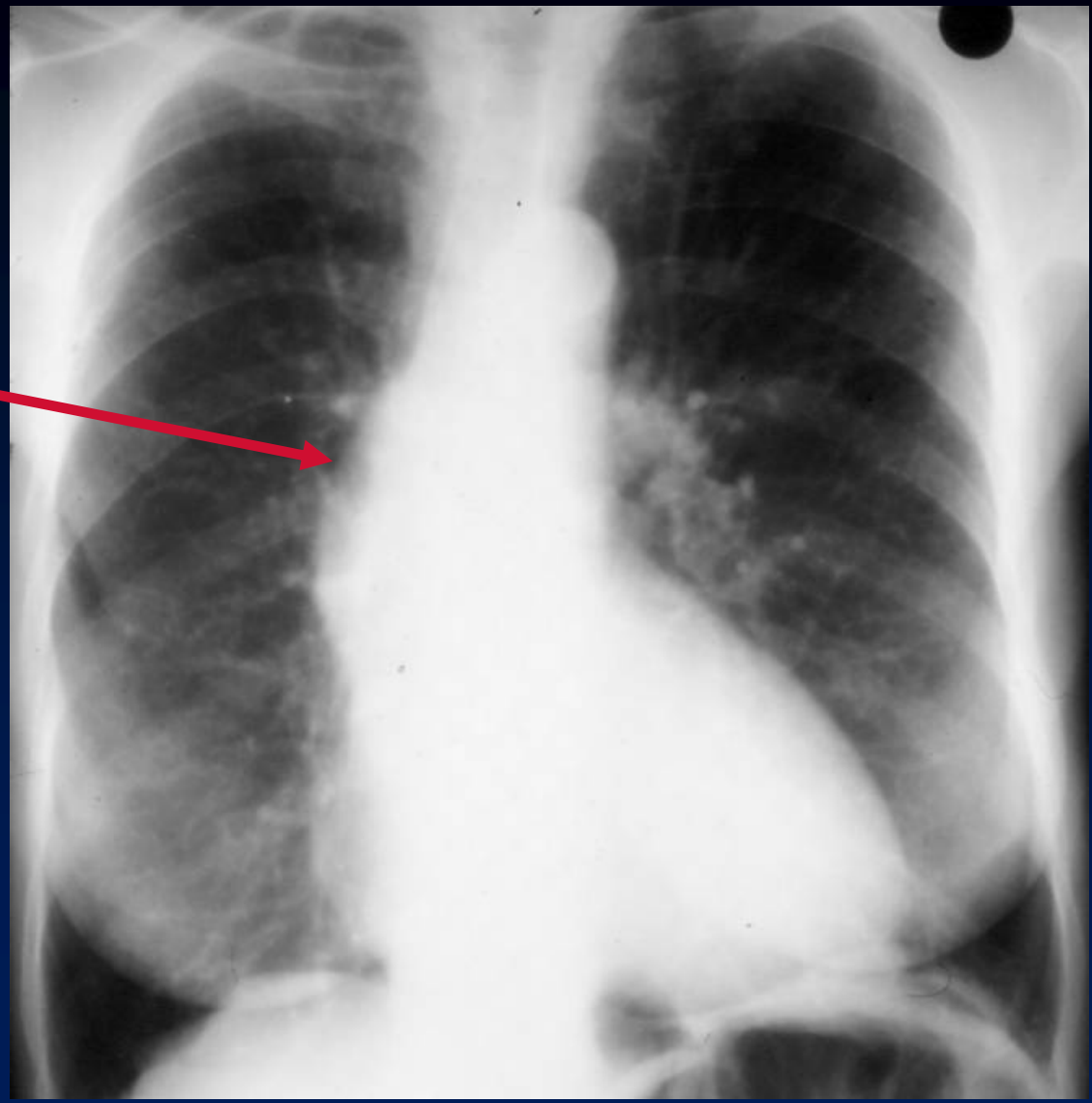


Aortic stenosis

Post-stenotic Dilatation of Aorta

- From turbulent flow just distal to any hemodynamically significant arterial stenosis
  - Jet effect also plays role
- Occurs mostly with valvular aortic stenosis
  - May occur at any age

Prominence of ascending aorta from post-stenotic dilatation



Aortic stenosis

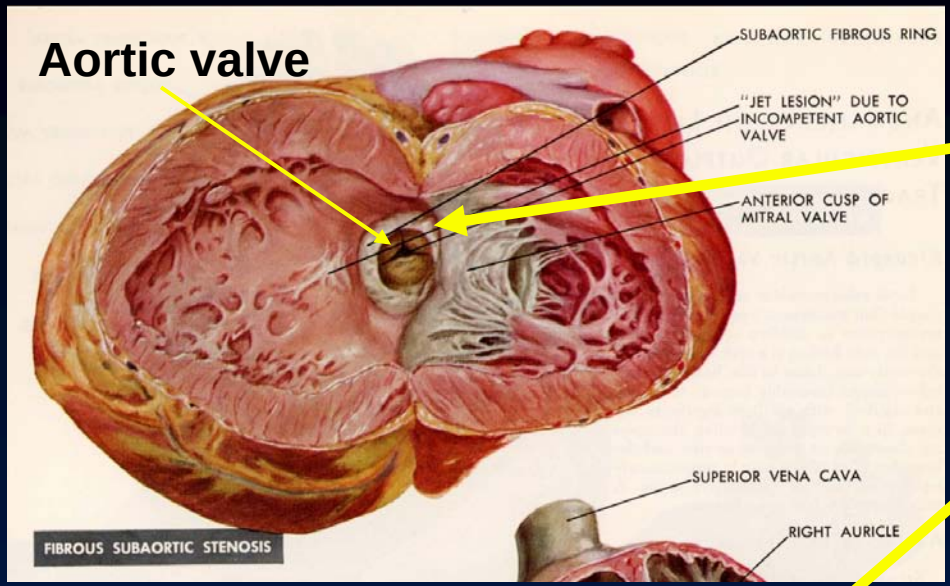
Aortic Stenosis
Calcification of Valve

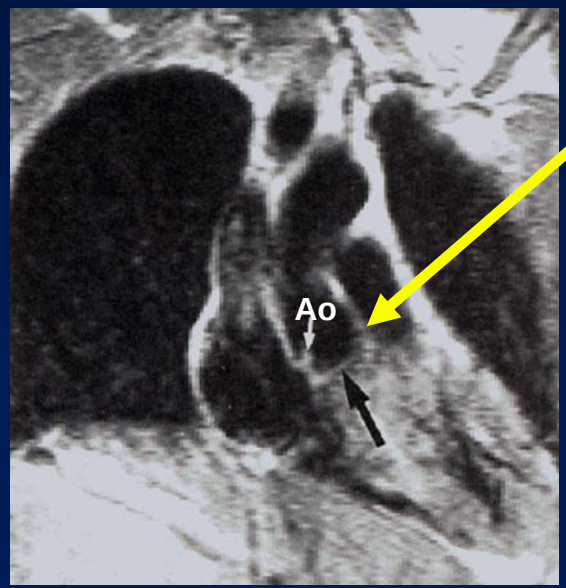
- In females, usually indicates hemodynamically significant AS
- Calcification of valve usually indicates gradient across valve of > 50mm Hg

Subvalvular Aortic Stenosis

Subvalvular Aortic Stenosis
Subaortic Stenosis

- Associated with
  - Subaortic fibrous membrane
  - Hypoplastic left heart syndrome
  - Idiopathic Hypertrophic Subaortic Stenosis





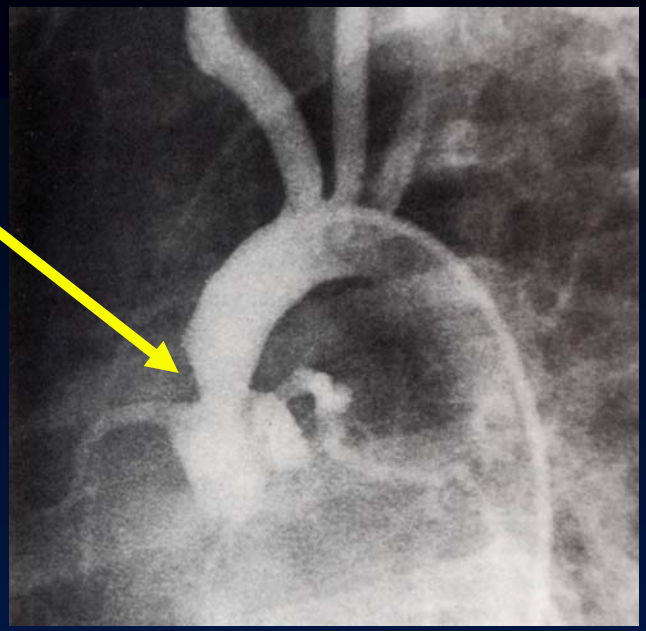
Subaortic Fibrous Membrane

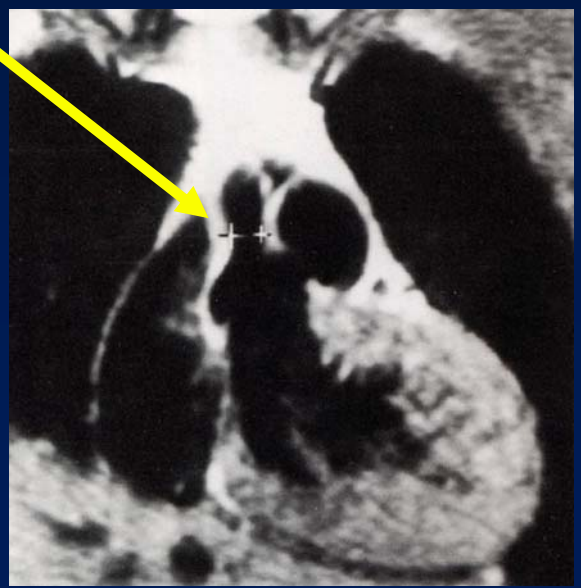
- About 15% of patients with congenital obstruction to LVOF
- Membrane just below aortic valve
- May attach to anterior leaflet of mitral valve
  - Mitral regurg
  - Aortic regurg

Supravalvular Aortic Stenosis

Supravalvular Aortic Stenosis
General

- Uncommon
- Types
  - Hourglass
  - Membrane
  - Hypoplasia of entire ascending aorta
- Associated lesions in 2/3
  - William’s syndrome

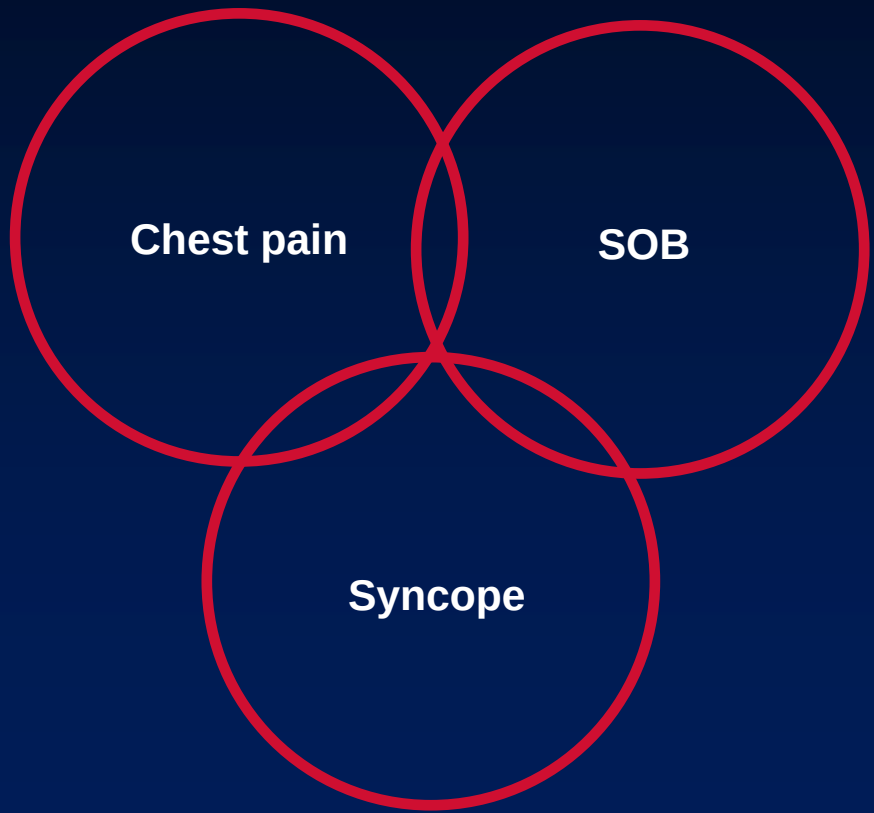




Supravalvular Aortic Stenosis

- William's syndrome
  - Supravalvular aortic stenosis
    - Hypercalcemia
    - Elfin facies
    - Pulmonary stenoses
    - Hypoplasia of aorta
    - Stenoses in
      - Renals, celiac, SMA

Aortic Stenosis
Clinical Triad



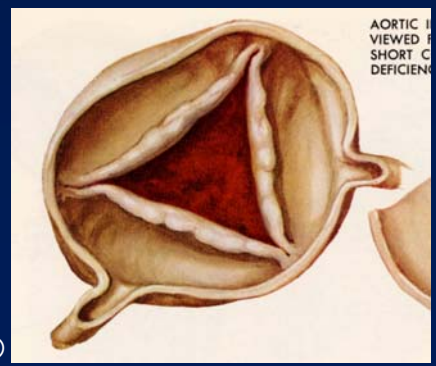
Aortic Regurgitation
(Aortic Insufficiency)

Aortic Regurgitation
Causes

- Rheumatic heart disease
- Marfan’s
- Luetic aortitis
- Ehlers-Danlos syndrome
- Endocarditis
- Aortic dissection

Aortic Regurgitation
Rheumatic Heart Disease

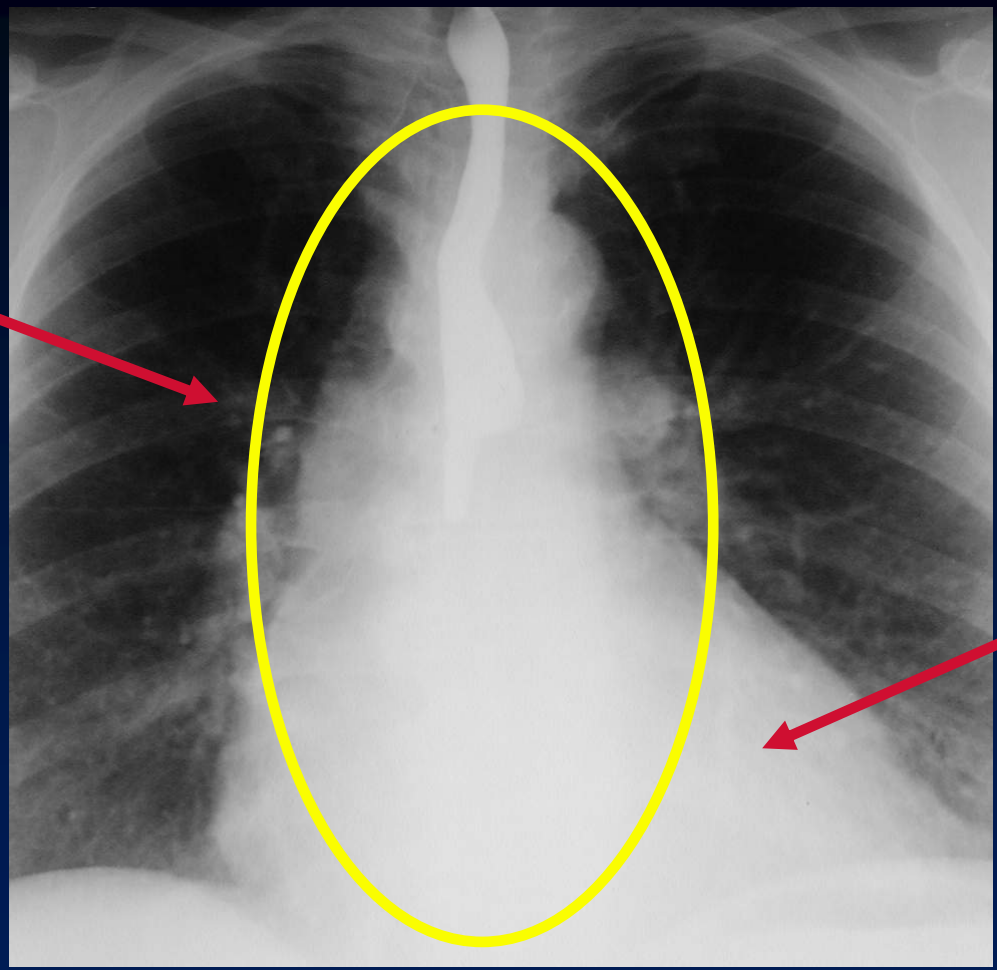
- Thickened cusps
- May have commissural fusion
  - In degenerative Ao regurg, no commissural fusion
- Regurgitant jet is usually central
  - In degenerative, usually not discrete jet



Aortic Regurgitation
Imaging Findings

- X-ray hallmarks are
  - Left ventricular enlargement
  - Enlargement of entire aorta
- Cine MRI (gradient refocused MRI)
  - “White blood” technique
  - Signal loss coming from Ao valve into LV during diastole
- Color Doppler is also diagnostic

Enlargement of entire aorta



Enlarged left ventricle

Aortic Regurgitation

Pulmonic Stenosis

Pulmonic Stenosis
General

- Without VSD = 8% of all CHD
- Mostly asymptomatic
- When symptomatic
  - Cyanosis and heart failure
  - Cor pulmonale
- Loud systolic ejection murmur

Pulmonic Stenosis
Types

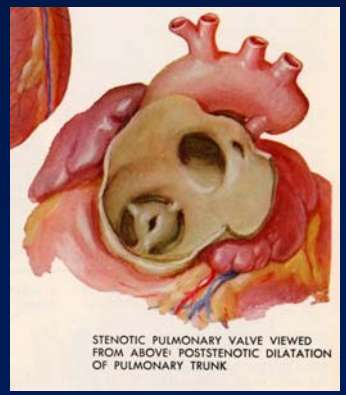
- Subvalvular
- Valvular
- Supravalvular

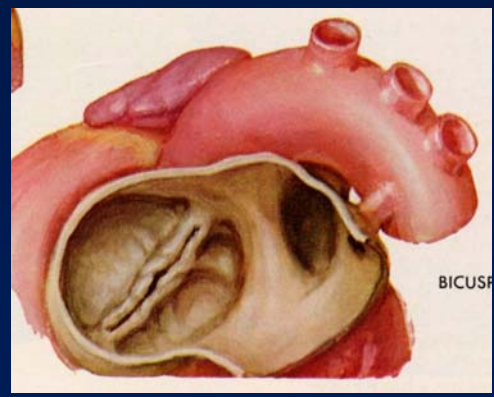
Pulmonic Stenosis
Valvular Pulmonic Stenosis

- Classic pulmonic stenosis (95%)
  - Congenital in origin
  - Associated with metastatic carcinoid syndrome
    - Tricuspid valve dz as well
  - Associated with Noonan Syndrome
    - ASD
    - Hypertrophic cardiomyopathy

Pulmonic Stenosis
Valvular Pulmonic Stenosis

- Morphology of abnormal valve
  - Membrane with central opening, or
  - Fusion of pulmonary cusps





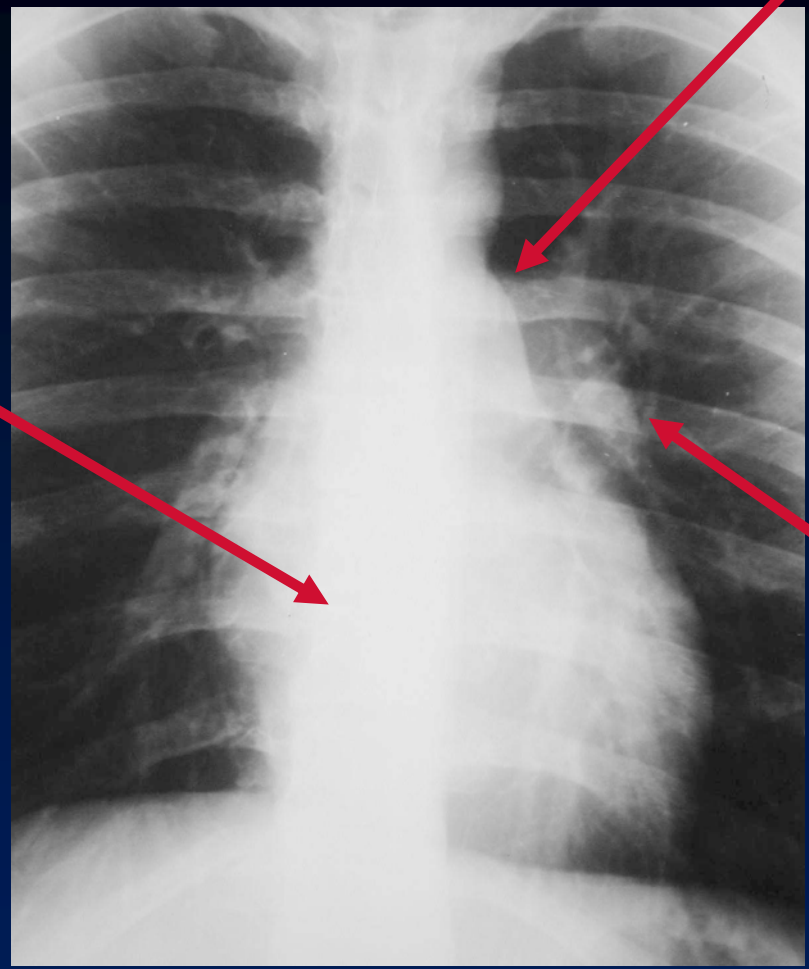
Pulmonic Stenosis
Valvular pulmonic stenosis

- Presents in childhood
- Pulmonic click
- Dome-shaped pulmonic valve in systole
- RX: Balloon valvulo-plasty

Pulmonic Stenosis
X-ray Findings

- Enlarged main pulmonary artery
- Enlarged left pulmonary artery (jet effect)
- Normal to decreased peripheral pulmonary vasculature
- Rare calcification of pulmonary valve in older adults

Prominent main pulmonary artery segment



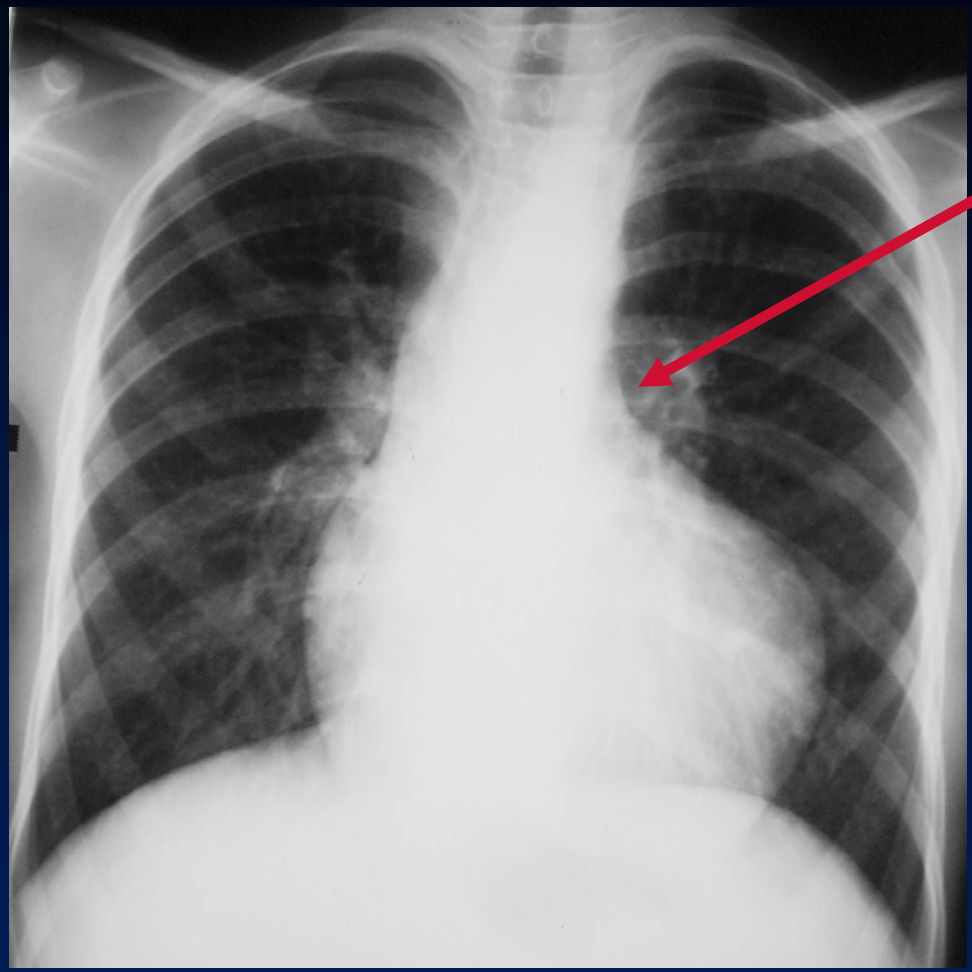
Normal-sized heart

Enlargement of left pulmonary artery

Pulmonic Stenosis

Pulmonic Stenosis
Subvalvular pulmonic stenosis

- **Infundibular pulmonic stenosis**
  - **Typically in Tetralogy of Fallot**
    - **50% of pts with TOF also have bicuspid pulmonic valves**
    - **50% of patients with TOF also have valvular pulmonic stenosis**
- **Subinfundibular pulmonic stenosis**
  - **Associated with VSD (85%)**



Tetralogy of Fallot with subvalvular pulmonic stenosis

Trilogy of Fallot

- Severe pulmonic valvular stenosis
- RV hypertrophy
- ASD with R ➜ L shunt

Supravalvular Pulmonic Stenosis
General

- May be either tubular hypoplasia or localized with poststenotic dilatation

Supravalvular Pulmonic Stenosis
Associated CV abnormalities

- Valvular pulmonary stenosis
- Supravalvular aortic stenosis
- VSD, PDA
- Systemic arterial stenoses

Supravalvular Pulmonic Stenosis
Associated Syndromes

- Williams Syndrome
  - Pulmonic Stenosis
  - Supravalvular AS
  - Peculiar facies
- Post-rubella syndrome
- Carcinoid syndrome with liver mets
- Ehlers-Danlos syndrome

The End