Valvular Lesions of the Heart

In Slide Show mode, to advance slides, press spacebar
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

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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

Stenotic mitral valve

Fusion of chordae

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 $2^\circ$ 
  $\uparrow$ 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 $\rightarrow$ fibrosis
  - Pulmonary hemosiderin deposited in lungs
  - Pulmonary ossification may occur
Effect of Mitral Stenosis on Lungs
Normal chamber pressures

RA  LA

RV  LV

M 1-5  M 5-10

D 1-7
S 17-32

M 15

D 5-12
S 90-140
Effect of Mitral Stenosis
On Lungs

- ↑ pulmonary venous and capillary pressure

<table>
<thead>
<tr>
<th>Condition</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>5-10</td>
</tr>
<tr>
<td>Cephalization</td>
<td>10-15</td>
</tr>
<tr>
<td>Kerley B Lines</td>
<td>15-20</td>
</tr>
<tr>
<td>Pulmonary Interstitial Edema</td>
<td>20-25</td>
</tr>
<tr>
<td>Pulmonary Alveolar Edema</td>
<td>&gt; 25</td>
</tr>
</tbody>
</table>
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
Mitral Stenosis

Convexity from enlarged left atrial appendage
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 left atrial appendage from mitral stenosis

Right atrial enlargement from tricuspid regurgitation

Mitral stenosis/regurgitation with tricuspid regurgitation
<table>
<thead>
<tr>
<th>X-Ray Findings of MS Calcifications</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Calcification of valve--not annulus--seen best on lateral film and at angio</td>
</tr>
<tr>
<td>- Rarely, calcification of left atrial wall 2° fibrosis from long-standing disease</td>
</tr>
<tr>
<td>- Rarely, calcification of pulmonary arteries from PAH</td>
</tr>
</tbody>
</table>
Calcification of mitral valve

Calcification of pulmonary artery

Calcification of left atrial wall
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

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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
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

<table>
<thead>
<tr>
<th>Etiology/Findings</th>
<th>Calcification</th>
<th>Other clues</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital Bicuspid Valve</td>
<td>30’s</td>
<td>Jet effect on aortogram</td>
</tr>
<tr>
<td>Degeneration of Tricuspid Valve</td>
<td>&gt; 65</td>
<td>Coronary artery ca++ Commissures don’t fuse</td>
</tr>
<tr>
<td>Rheumatic dz in Tricuspid Valve</td>
<td>30’s here; teens in 3rd world countries</td>
<td>MS or MR almost always present; commissures fuse</td>
</tr>
</tbody>
</table>
Aortic Stenosis
X-Ray Findings

- Depends on age patient/severity of disease
  - In infants, AS $\rightarrow$ CHF/pulmonary edema
- In adults
  - Normal heart size
    - Until cardiac muscle decompensates
  - Enlarged ascending aorta $\circ ^{2}$ post-stenotic dilatation $\circ ^{2}$ 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
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

- Chest pain
- SOB
- Syncope
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

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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
Pulmonary 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

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Pulmonic Stenosis
Valvular pulmonic stenosis

- Presents in childhood
- Pulmonic click
- Dome-shaped pulmonic valve in systole
- RX: Balloon valvuloplasty
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%)
Concave pulmonary artery segment

Tetralogy of Fallot with subvalvular pulmonic stenosis
Trilogy of Fallot

- Severe pulmonic valvular stenosis
- RV hypertrophy
- ASD with R $\rightarrow$ 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