Miscellaneous Cardiac Diseases

In Slide Show mode, to advance slides, press spacebar or click left mouse button
Sinus of Valsalva Aneurysm
Types

- Congenital
- Inherited
- Acquired
**Sinus of Valsalva Aneurysm**

**Congenital**

- Usual type
- Involves a single cusp
- Most often arise from R coronary sinus
Sinus of Valsalva Aneurysm
Inherited

- Associated with Marfan’s Disease
- All cusps involved
- Produce aortic regurgitation
Sinus of Valsalva Aneurysm
Acquired

- Usually 2° to endocarditis of aortic valve
- Other causes
  - Syphilis
  - Atherosclerosis
  - Dissecting aneurysm
  - Marfan's
## Sinus of Valsalva Aneurysm
### X-ray Findings

- Since aortic root is intracardiac, usual aneurysm is not visible
- Rarely, a large aneurysm of L aortic sinus → bulge L upper heart border in region of LA appendage
## Sinus of Valsalva Aneurysm

### Other X-ray Findings

- Rarely, a large aneurysm of the R aortic sinus → bulge on R heart border
- Usually the aneurysm dilates the aortic ring → AI
Aneurysm of Sinus of Valsalva and Proximal Ascending Aorta = annuloaortic ectasia
Ruptured Sinus of Valsalva Aneurysm
Ruptured Sinus of Valsalva Aneurysm
Congenital vs. Acquired

- Congenital forms (usually R sided) always produce an intracardiac fistula
  - Most congenital aneurysms rupture during third or fourth decade of life
- Acquired forms can produce either intra- or extracardiac fistulae
Ruptured Sinus of Valsalva Aneurysm

**General**

- May rupture → aortic-cardiac fistula
  - L → R shunt usually
- Most ruptures involve R coronary sinus
  - Into R ventricle
- Posterior (non-coronary) aortic sinus ruptures occasionally
  - Into R atrium
Ruptured Sinus of Valsalva Aneurysm

General

- Rupture of aneurysms of L sinus are very rare
  - May rupture into the pericardial space
Ruptured Sinus of Valsalva Aneurysm
Clinical

- Symptoms due to sudden onset of massive aortic regurg or L → R shunt
  - Acute onset of SOB
  - Chest pain
  - Acute onset of murmur
  - CHF
  - Death
Ruptured Sinus of Valsalva Aneurysm

X-ray Findings

- Acute CHF
- Followed by L → R shunt
- With rupture of L sinus, LA may suddenly enlarge
Two cases of rupture of right coronary sinus into RV
Congenital Defect in the Pericardium
Congenital Pericardial Defect
Embryogenesis

- Premature atrophy of left duct of Cuvier (cardinal vein)
- Failure of nourishment of left pleuro-pericardial membrane → failure of pericardium to develop
## Congenital Pericardial Defect

### General

- Male:female ratio of 3:1
- May be detected at any age
  - Most common in low 20’s
<table>
<thead>
<tr>
<th>Location</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Foraminal defect on left side</td>
<td>35%</td>
</tr>
<tr>
<td>Complete absence of left side gives levoposition of heart</td>
<td>35%</td>
</tr>
<tr>
<td>Diaphragmatic surface</td>
<td>17%</td>
</tr>
<tr>
<td>Total bilateral absence</td>
<td>9%</td>
</tr>
<tr>
<td>Right sided</td>
<td>4%</td>
</tr>
</tbody>
</table>
Congenital Pericardial Defect
Associations

- Bronchogenic cysts
- VSD, PDA, mitral stenosis
- Diaphragmatic hernia
- Sequestration
Congenital Pericardial Defect
Clinical

- Mostly asymptomatic
- May have:
  - Tachycardia
  - Palpitations
  - Right bundle block
  - Positional discomfort lying on left side
  - Chest pain
Congenital Pericardial Defect
X-ray Findings

- Focal bulge in area of main pulmonary artery
- Sharply marginated
- Lung may interpose between heart-left hemidiaphragm
- Increased distance between sternum and heart 2° absence of sternopericardial ligament
Congenital Pericardial Defect
X-ray Findings-Continued

- Levoposition of heart
- Pneumopericardium following pneumothorax
Congenital Defect in the Pericardium
Congenital Pericardial Defect

Treatment

- Since herniation and strangulation of left atrial appendage or herniation of LA/LV may occur
- Foraminal defect requires surgery
Cardiac Malpositions

Heterotaxy

Syndromes
Trilobed and Bilobed Lungs

- Trilobed lung
- Bilobed lung
- Liver
- Spleen
## Naming Rules

- Since anatomic side (i.e. “left” or “right”) in complex lesions is frequently reversed or indeterminate

- Naming conventions for
  - Atria
  - AV valves
  - Ventricles
  - Ventricular outflow tracts
<table>
<thead>
<tr>
<th>The Rules</th>
</tr>
</thead>
<tbody>
<tr>
<td>How the atria are named</td>
</tr>
</tbody>
</table>

- **Anatomic right atrium is on side of trilobed lung and liver**
  - Shape of atrial appendage - broad and pyramidal
  - Same side as IVC
- **Anatomic left atrium is on side of bilobed lung and spleen**
  - Shape of atrial appendage - thin c narrow neck
  - Same side as aortic arch
The Rules
How the ventricles are named

- Anatomic right ventricle is trabeculated ventricle
  - Coarse in both systole and diastole
  - Has tricuspid AV valve
- Anatomic left ventricle is smooth-walled ventricle
  - In diastole; fine trabeculations in systole
  - Has bicuspid AV valve
Anatomic Ventricles

Trabeculated ventricle-Anatomic Right

Smooth ventricle-Anatomic Left
<table>
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</thead>
<tbody>
<tr>
<td>Mitral and tricuspid valves</td>
</tr>
</tbody>
</table>

- **Tricuspid valve** belongs to anatomic right ventricle
  - Not right atrium

- **Mitral valve** belongs to anatomic left ventricle
  - Not left atrium
**AV Connections**

**Concordance**

- Ventricles are concordant to the atria
  - When R atrium connects to R ventricle
  - L atrium connects to L ventricle
- Ventricles are discordant to the atria
  - When R atrium connects to L ventricle
  - When L atrium connects to R ventricle
- With atrial isomerism, AV connections are ambiguous
## The Rules

### Aortic and pulmonic valves

- Pulmonic valve is part of pulmonary artery
  - Not anatomic right ventricle
- Aortic valve is part of aorta
  - Not anatomic left ventricle
- Pulmonic infundibulum is part of anatomic right ventricle
Anatomic R atrium is on side of trilobed lungsame side as IVC

Anatomic L atrium is on side of bilobed lung—same side as Ao arch

Tricuspid valve belongs to anatomic RV

Mitral valve belongs to anatomic LV

Pulmonic infundibulum belongs to anatomic RV

Aortic valve belongs to aorta

Pulmonic valve belongs to pulmonary artery

Anatomic R ventricle is trabeculated

Anatomic L ventricle is smooth
Situs
Definitions

- Describes position of asymmetric organs in body
  - Lungs
  - Liver
  - Spleen
  - Stomach
**Situs Solitus**

- Normal anatomic relationships
  - **Right side**
    - Trilobed lung
    - Eparterial bronchus
    - Anatomic right atrium
    - Liver
  - **Left side**
    - Bilobed lung
    - Hyparterial bronchus
    - Anatomic left atrium
    - Spleen
Situs Solitus

- Trilobed Lung
- Venous Atrium
- Liver

- Bilobed Lung
- Arterial Atrium
- Stomach
- Spleen
### Situs Solitus

**Hyparterial/Eparterial Bronchi**

<table>
<thead>
<tr>
<th>Eparterial bronchus-</th>
<th>Hyparterial bronchus-</th>
</tr>
</thead>
<tbody>
<tr>
<td>First branch of Right mainstem bronchus is above pulmonary artery</td>
<td>First branch of Left mainstem bronchus is below pulmonary artery</td>
</tr>
</tbody>
</table>
Situs Solitus
0.6 - 0.8% CHD
## Situs Inversus

- **Reversed anatomic relationships**
  - **Right side**
    - Bilobed lung
    - Hyparterial bronchus
    - Anatomic left atrium
    - Spleen
  - **Left side**
    - Trilobed lung
    - Eparterial bronchus
    - Anatomic right atrium
    - Liver
Situs Inversus

- Situs Inversus
  - Bilobed Lung
  - Trilobed Lung
  - Arterial Atrium
  - Venous Atrium
  - Stomach
  - Liver
  - Spleen
Situs Ambiguous

- Lungs and abdomen are symmetric so right and left sides can’t be defined
  - Isomerism—both atria have the same features
    - Either right or left
  - Two kinds of situs ambiguous
    - Bilateral right-sidedness
    - Bilateral left-sidedness
Situs Ambiguous Heterotaxy Syndrome

- **Bilateral right-sidedness**
  - Since, spleen is usually on left side
  - No spleen
    - Asplenia syndrome

- **Bilateral left-sidedness**
  - Since, spleen is usually on left side
  - Many spleens
    - Polysplenia syndrome
Situs Ambiguous
Cardiac Positions

- Position of cardiac apex
  - Levocardia on the left
  - Dextrocardia on the right
  - Mesoccardia in the midline

- Cardiac malposition
  - Anything other than situs solitus with levocardia
Cardiac Malpositions
Types

- Situs solitus with dextrocardia
- Situs inversus with levocardia
- Situs inversus with dextrocardia
Situs Solitus with Dextrocardia

Test Alert

95% chance of CHD of which 80% corrected transposition
If cyanotic with ↑ flow, then tricuspid atresia.
If cyanotic with ↓ flow, then corrected transposition.
If asplenia, then 100% have common ventricle.
Interrupted IVC common.
Situs Solitus with Dextrocardia
Coarctation of Aorta
Situs Inversus with Levocardia

Rare, but 100% CHD
If asplenia, 100% have common ventricle
Interruption of IVC common
Situs Inversus with Levocardia
Situs Inversus with Levocardia Transposition
Situs Inversus with Dextrocardia

3-5% CHD
Most common is Corrected Transposition
Kartgener’s
Situs Inversus with Dextrocardia

Trilobed lung
Situs Solitus
Malposition of the Stomach

R/O asplenia
Most have CHD (L → R shunt)
Most with polysplenia have azygous continuation of IVC
Situs Solitus with Malposition of the Stomach
Situs Inversus
Malposition of the Stomach

Situs Inversus with Malposition of the Stomach
95% CHD of which 80% are Corrected Transposition
Situs Ambiguous
Heterotaxy Syndrome

- Bilateral right-sidedness
  - Since, liver is usually on right side
  - No spleen
    - Asplenia syndrome

- Bilateral left-sidedness
  - Since, spleen is usually on left side
  - Multiple spleens
    - Polysplenia syndrome
Asplenia
Bilateral Right-sidedness

- Male
- Cyanotic
- High risk of infection
- Severe cardiac abnormalities
  - Transposition
  - TAPVR
## Polysplenia

### Bilateral left-sidedness

- Female
- Abnormalities are more benign
  - Azygous continuation of IVC
  - Bilateral superior vena cava
  - PAPVR
  - ASD
Situs Ambiguous-polysplenia
Situs Ambiguous-polysplenia
Approach to Cardiac Malpositions

- Which side is heart on
- Which side is trilobed lung on
- Which side is arch on
- Which side is stomach bubble on
- Check for asplenia
  - Midline liver
  - Minor fissures in both lungs
Another Approach

**CARDIAC MALPOSITIONS**

- **Situs solitus**
  - Aortic arch and LA are always on the same side except with isolated R arch 0.6-0.8% CHD.

- **Situs inversus with dextrocardia**
  - 3-5% CHD
  - Most common is corrected transposition (Kartagener)

- **Situs inversus with levocardia**
  - Rare, but 95-100% CHD; no specific prevalence. If asplenia, 100% have common ventricle, interruption of IVC common.

- **Situs solitus with malposition of the stomach**
  - R/L asplenia
  - Most have CHD (L->R shunt)
  - Most with polysplenia and azygos continuation of IVC

- **Situs inversus with malposition of the stomach**
  - 95% CHD of which 80% are corrected transposition. If cymosis with ductus, then atroventricular. If cymosis and IVC, then corrected transposition. If asplenia, 100% have common ventricle, interrupted IVC common.
Situs Inversus with Levocardia

Rare, but 100% CHD
If asplenia, 100% have common ventricle
Interruption of IVC common
Situs ambiguous
Asplenia with dextrocardia
Complex CHD
Nine Lesions Which Produce 75% of All Severe Congenital Heart Lesions In the Neonate

- **Decreased flow**
  1. Tetralogy of Fallot
  2. Tricuspid Atresia
  3. Severe Pulmonic Stenosis
  4. Ebstein’s

- **Increased Flow**
  5. Transposition
  6. VSD
Nine Lesions Which Produce 75% of All Severe Congenital Heart Lesions In the Neonate

- Pulmonary venous hypertension
  - 7. Hypoplastic left heart
  - 8. Coarctation of the aorta
  - 9. TAPVR with infradiaphragmatic obstruction

- What’s left
  - Left-to-right shunts
    - ASD
    - PDA
  - Truncus arteriosus
The End