Alterations in Pulmonary Vascularity
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Objectives
1. Identify normal vascular anatomy
2. Detect alterations in pulmonary vascularity on radiographs
3. Describe physiologic basis for alteration of pulmonary vascularity
4. Create differential diagnosis based on abnormal pattern of vascularity

Financial Disclosures
None

Pulmonary Vascularity
Increased
- Over circulation
- Pulmonary venous hypertension

Decreased
- Emphysema
- Ebstein anomaly
- Westermark sign
- Swyer-James-McLeod

Normal Pulmonary Vascularity

Pulmonary Vascularity
Congenital vascular
- Scimitar syndrome
- Pulmonary AVM

Main pulmonary arteries
- Pulmonary arterial HTN
- Pulmonic stenosis
Increased Vascularity
- Overcirculation
  - Left-right shunt
  - Admixture lesions
  - High-flow states
  - Thyrotoxicosis
  - Anemia
  - Pregnancy
  - Peripheral AV fistula

Overcirculation Radiograph
- Enlarged pulmonary arteries
  - RLPA upper limit = 17 mm
- Enlarged pulmonary veins
- Shunt must be at least 40%

Pulmonary Venous Hypertension
- Most common vascular abnormality
- Causes
  - Obstruction/stenosis proximal to mitral valve
  - Mitral valve disease
  - Left ventricular failure or elevated pressure
    - Ischemia
    - Aortic stenosis

Pulmonary Venous Hypertension
- Elevated pressure
  - Pulmonary veins
  - Capillary bed
  - Pulmonary arteries

Pulmonary Venous Hypertension
- Physiologic & anatomic changes
  - Redistribution of blood to upper lung
  - Interstitial edema
  - Alveolar edema

Pulmonary Venous Hypertension
- Chest radiograph
  - Equalization of upper and lower lobe vessels
  - Prominence of the hilum
  - Loss of normal hilar angle
### Pulmonary Vascularity

- **Increased**
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  - Pulmonary venous hypertension

- **Decreased**
  - Emphysema
  - Ebstein anomaly
  - Westermark sign
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### Emphysema

- **Centrilobular** (proximal acinar)
- Panacinar
- Paraseptal
- Cicatricial

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### Emphysema

- **Centrilobular**
- Panacinar
- Paraseptal
- Cicatricial

- Vessels present but narrowed
- Normal axial pathway – fewer side branches
- Absent vascularity

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### Emphysema

- **Secondary alteration of the vasculature**
- **Hyperinflation of affected lung**

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### Emphysema

- **Radiographic Findings**
  - Hyperinflation
  - Vascular alterations
    - Diminished vascularity
    - Vessel distortion
    - Increased branching angles
    - Loss of normal sinuosity
Emphysema

- Centrilobular
  - Upper lung predominance

- Panacinar
  - Lower lung predominance


Localization of Emphysema

- Ventilation of upright lung base
  - Ventilation greater
  - Alveoli are smaller
    - Less negative pleura pressure due to weight of lung
    - Greater change in volume of basal alveoli


Localization of Emphysema

- Perfusion of upright lung base
  - Perfusion greater
  - Affect of gravity
  - Low pressure pulmonary arterial system

- Perfusion of upright lung apex
  - Pulmonary arterial pressure just sufficient to supply blood to apex
  - Ventilation:perfusion ratio is greater at apex


Centrilobular Emphysema

- Associated with smoking
- Affects areas where ventilation:perfusion ration is highest
- Emphysema greatest in upper lung


Panacinar Emphysema

- Associated with alpha-1-antitrypsin deficiency
- Follow distribution of inhaled agent
  - Follows isolated ventilation gradient
- Leukocytes with protease follow blood flow
- Emphysema greatest in lung bases


Ebstein Anomaly

- Congenital malformation tricuspid valve
- Downward displacement
  - Posterior and septal valve leaflets
- Small right ventricle
- Tricuspid regurgitation
- Right atrial enlargement
- Decreased pulmonary vascularity

**Westermark Sign**
- Localized oligemia distal to thrombus
- Occlusion of lobar or semental pulmonary artery
- Widespread small vessel occlusion

**Westermark Sign**

**Swyer–James Syndrome**
- Symptoms
  - None
  - Dyspnea on exertion
  - Hx of childhood respiratory infection
    - Viral
- Histopathology
  - Constrictive bronchiolitis
  - Chronic bronchiolitis
  - Bronchiectasis

**Swyer–James Syndrome**
- Chest Radiograph at full inspiration
  - Volume normal or slightly decreased
  - Increased lucency of affected lung
    - Due to decreased perfusion
  - Hilum may be small in size
- Expiratory Radiograph
  - Air trapping
- Main DDx
  - Central obstructing bronchial lesion

**Bronchial Obstruction**
- Decreased ventilation to affected lung
- Limited gas exchange
- Vascular constriction due to hypoxia
Pulmonary Vascularity

Congenital vascular
• Scimitar syndrome
• Pulmonary AVM

Main pulmonary arteries
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• Pulmonic stenosis

Hypogenetic Lung (Scimitar) Syndrome
• Hypoplastic right lung
• Hypoplasia right pulmonary artery
• Systemic arterial supply to right lung
• Pulmonary venous drainage to IVC
  • Suprahepatic IVC
  • Hepatic veins
  • Azygos vein
  • Coronary sinus
  • Right atrium

Pulmonary Venolobar Syndrome
Hypogenetic Lung Syndrome
• Partial anomalous pulmonary venous return
  • Right lung drains to IVC
  • Hypoplastic/aplastic pulmonary artery
  • Hypoplastic/aplastic bronchi

Scimitar Vein
• PAPVR right lung
• Drainage inferiorly
  • IVC
  • Coronary sinus
  • Azygos vein
  • Portal vein
  • Hepatic vein

Pulmonary Arteriovenous Malformation
• Congenital
  • Idiopathic
  • Hereditary hemorrhagic telangiectasia

• Acquired
  • Hepatopulmonary syndrome
  • Late post-surgical complication of congenital cyanotic heart disease
  • Mitral stenosis
  • Trauma

Congenital Pulmonary Arteriovenous Malformation
• Abnormal connection between pulmonary artery & vein
• Right-to-left shunt
  • Hypoxemia not corrected with 100% O2
Pulmonary Arteriovenous Malformation

- Persistent fetal anastomotic capillaries
- Size of malformation increases over time
- 35–65% associated with Hereditary Hemorrhagic Telangiectasia
  - aka Osler-Weber-Rendu syndrome

Pulmonary Arteriovenous Malformation

- 2/3 lower lobes
- 35% multiple lesions
- 10–30% bilateral lesions

Pulmonary Arteriovenous Malformation

- Simple – 80%
  - Single feeding artery
  - Single draining vein
- Complex – 20%
  - ≥ 2 feeding arteries
  - ≥ 2 draining veins

Pulmonary Arteriovenous Malformation Symptoms

- Epistaxis
  - Most common symptom of patients with Hereditary Hemorrhagic Telangiectasias
- Dyspnea
- Hemoptysis

CNS Symptoms

- Brain abscess
- Embolic stroke
- Transient ischemic attacks
- Hemorrhagic brain AVM

Less common symptoms

- Chest pain
- Cough
- Dizziness
- Syncope
- Polycythemia
- CNS symptoms
Physical Exam

- Mucocutaneous telangiectasias
  - Hereditary hemorrhagic telangiectasia
- Bruit over AVM

Pulmonary AVM Imaging

- Chest radiograph
  - Well-defined nodule
    - Round
    - Oval
    - Lobulated
    - Serpentine
  - Feeding artery & draining vein

Pulmonary AVM Imaging

- CT
  - Nodular density
    - Feeding artery
    - Enlarged draining vein
  - Feeding vessel sign
    - Mets
    - Septic emboli

Pulmonary AVM Imaging

- CT
  - Dynamic contrast bolus
    - Rapid enhancement
    - Rapid washout
  - Detects small lesions
  - Allows for treatment planning

Pulmonary AVM Treatment

- Selective embolization if AVM ≥ 3 mm
- Follow-up CT or MRA
  - Ensure thrombosis
- Periodic screening
  - Patients with hereditary hemorrhagic telangiectasia

Pulmonary AVM

- Don’t mistake adjacent vessel for AVM
  - Look for enlarged draining vein
  - Slab MIP images
- AVMs grow over time
  - HHT patients need periodic screening
- If patient has HHT
  - Screen family members for disease

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**Pulmonary Arterial HTN**
- Enlarged central pulmonary arteries
- Rapid tapering
- Peripheral decreased vascularity

**Pulmonic Stenosis**
- Dilated pulmonary trunk
- Dilated left pulmonary artery
- Otherwise normal vascularity

**Pulmonic Stenosis**
- Congenital
- Cone shaped valve
- Post–stenotic jet causes dilatation of
  - Main pulmonary artery
  - Left pulmonary artery
  - Right pulmonary artery is spared