Pulmonary Vascular Disease
We Have No Relevant Disclosures
References

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• Simonneau, Gérald. “Haemodynamic Definitions and Updated Clinical Classification of Pulmonary Hypertension,” n.d.
Pulmonary Vascular Disease is a Spectrum

Pulmonary Arteries

Right Ventricle
Pulmonary Vascular Disease is a Spectrum

Pulmonary Arteries

Right Ventricle

Pulmonary Veins

Left Ventricle
Pulmonary Vascular Disease is a Spectrum

Parenchymal findings may be clue to microscopic disease!
Pulmonary Arteries

- Dilated
  - Pulmonary Hypertension
  - Aneurysm/Pseudoaneurysm
- Narrowed/Occluded
  - Acquired
  - Congenital
Pulmonary Hypertension

Five Groups*:

1. Pulmonary Arterial Hypertension (PAH)
2. Left heart disease
3. Lung disease/hypoxemia
4. Chronic Thromboembolic Pulmonary Hypertension
5. Unclear/multifactorial

Mean Pulmonary Artery Pressure >20 mmHg

*6th World Symposium on Pulmonary Hypertension 2018
How does the Radiologist help?

Alternative Diagnoses of Patients Referred to PAH Specialty Clinic

- ILD: 5%
- VTE: 5%
- Other: 12%
- Structural Heart Disease: 13%
- OSA: 19%
- LV Dysfunction: 22%
- Obstructive Lung Disease: 24%
- All Alternative Diagnoses: 85%

N= 268

RV Dilation vs Hypertrophy

The RV is very sensitive to changes in pressure.
Group 1 - PAH

- Precapillary disease
- Causes:
  - Idiopathic
  - Heritable
  - Drugs
  - CTD
  - Congenital heart disease (shunt)
  - PAH with overt features of venous/capillaries (PVOD/PCH) involvement
Idiopathic PAH
Idiopathic PAH
PAH from Congenital Heart Disease
Parenchymal Findings in PAH
Parenchymal Findings in PAH

- Mosaic attenuation
- Groundglass centrilobular nodules
- Corkscrew vessels
Parenchymal Findings in PAH

- Mosaic attenuation
- Groundglass centrilobular nodules
- Corkscrew vessels
Atherosclerosis in longstanding PAH

Patent Ductus Arteriosus
PVOD/PCH

PAH + Pulmonary Edema + Normal Left Heart
PVOD/PCH

- Diagnostic dilemma (normal PCWP)
- 5-10% of patients with idiopathic PAH
- Incidence ~2/million per year
- Associations
  - Idiopathic
  - HIV, BMT, CTD, sarcoid, PLCH

16-year-old – PVOD/PCH
19-year-old – PVOD/PCH
Left Heart Failure (not PVOD/PCH)

Dilated Left Ventricle
Group 2 – Left Heart Disease

- LV Systolic Dysfunction
- LV Diastolic Dysfunction
- Valvular Disease
- Congenital heart disease
  - Acquired inflow/outflow tract obstruction
  - Congenital cardiomyopathies
Group 2 – Longstanding Mitral Stenosis
Noncompaction cardiomyopathy

Mean PAP 54 mmHg
Group 3 – Lung Disease and/or Hypoxia

- COPD
- ILD
- Mixed obstructive and restrictive pattern
- Sleep-disordered breathing
- Alveolar hypoventilation
- Chronic exposure to high altitude
- Developmental lung disease
Group 3 – PH in COPD

- Common
- Usually mild but associated with reduced survival
- Lung transplantation is best long-term therapy
Group 3 – Cystic Fibrosis
Combined pulmonary fibrosis and emphysema

- AKA CPFE
- PH more frequent/more severe than IPF or COPD alone
- Can occur with IPF or other fibrosis
Group 4 - CTEPH

- Potentially *curable cause of PH*
- Can occur after only ONE episode of acute PE
  - 4-5% of patients with PE develop CTEPH
  - May have no known episode of acute PE
Acute vs Chronic PE

**Acute**
- Central (unless saddle)
- Convex margins
- Increased caliber if occlusive

**Chronic** (acute PE > 3 months)
- Eccentric
- Concave margins (pouch)
- Decreased caliber if occlusive
- Webs, bands
Worsening Dyspnea 6 months after PE
CTEPH - Thromboendarterectomy
Group 5 – Unclear/Multifactorial

- Hematologic disorders
  - Chronic hemolytic anemia
  - Myeloproliferative disorders
  - Splenectomy
- Systemic disorders
  - Sarcoidosis
  - Pulmonary histiocytosis
  - Lymphangioleiomyomatosis
- Metabolic disorders
- Others
  - Tumoral obstruction
  - Fibrosing mediastinitis
  - Chronic renal failure
  - Segmental PH
Fibrosing mediastinitis

- Confluent soft tissue
- Mass effect on pulmonary vessels
- Enlarged bronchial arteries
Langerhan cell histiocytosis
Locally Dilated PA

- Aneurysm or Pseudoaneurysm
Behçet Disease
Mycotic Pseudoaenurysm
Traumatic PA Pseudoaneurysm
Narrowed/Occluded

- CTEPH
- Vasculitis
- Congenital
CTEPH
Proximal Interruption of the LPA
Proximal Interruption of the RPA
Abnormalities of Vessel Drainage

• Anomalous pulmonary venous connection
• Pulmonary arteriovenous malformation
• Meandering Pulmonary Vein
Scimitar Syndrome
Pulmonary AVM
Pulmonary AVM
Meandering Pulmonary Vein

- Rare vascular anomaly
  - Abnormal course
  - Normal drainage

- Not a shunt → negative bubble study

- Little physiologic significance
Abnormalities of Vessel Lumen

• Filling defect
  – Pulmonary thromboembolism
  – Tumor embolism
  – Pulmonary artery sarcoma
  – Foreign body
Pulmonary Artery Sarcoma
Nonthrombotic Pulmonary Embolism

Any substance in the venous circulation can embolize to the lungs

Cause mechanical obstruction and/or inflammatory cascade

The right ventricle is *very sensitive* to changes in pressure
Macroscopic vs Microscopic

- Bullet
- IVC Filter
- Brachytherapy seed
- Wire
- Methylmethacrylate
- Mercury
- Catheter
- Air
- Excipient
- Amniotic Fluid
- Silicone
- Fat
- Ethiodol
- Chemotherapy

Tumor
- Bacteria/Fungi
Macroscopic – Catheter Fragment
Macroscopic - Methylmethacrylate
Microscopic – Excipient Lung Disease
Microscopic – Excipient Lung Disease

- Tablets crushed and injected intravenously

- Excipient = inert binder of medication

Microscopic – Excipient Lung Disease

- Right heart failure
- Unexplained fever/bacteremia
- Indwelling catheters

20-year-old after a cosmetic procedure
Microscopic – Silicone Embolization
Microscopic – Silicone Embolization
Microscopic – Silicone Embolization

- Chemical pneumonitis
  → vasoconstriction
  → acutely elevated right heart pressures
  → right heart failure
Pulmonary Veins

- Stenosis/Occlusion
Pulmonary Vein Stenosis from Ablation
Referred for “ILD”

Pulmonary vein stenosis from ablation
Small Vessels

• PVOD/PCH
• Vasculitis/Capillaritis
PVOD/PCH

59-year-old with PVOD/PCH overlap on explant
Small Vessels

- Anti-GBM antibody disease (Goodpasture syndrome)
- Primary Vasculitis
  - ANCA-associated
    - Granulomatosis with polyangiitis
    - Microscopic polyangiitis
    - Eosinophilic granulomatosis with polyangiitis (rare)
  - Behçet disease (rare)
  - Henoch-Schönlein purpura (rare)
  - Isolated pauci-immune pulmonary capillaritis
Small Vessels

• Secondary Vasculitis
  – Connective Tissue Disease
    • Systemic lupus erythematosus
    • Rheumatoid arthritis
    • Mixed connective tissue disorder
    • Polymyositis
    • Primary or secondary antiphospholipid antibody syndrome

• Drug-induced hemorrhage
Anti-GBM Antibody Disease
Granulomatosis With Polyangiitis
Granulomatosis With Polyangiitis
Microscopic Polyangiitis
Drug-Induced Hemorrhage
Summary

• Patients with pulmonary vascular disease present with a wide range of imaging abnormalities
• Be familiar with the PH Classification
• Direct findings of large vessel disease usually apparent on CT (or MRI)
• Imaging features of small vessel disease overlap (DAH)