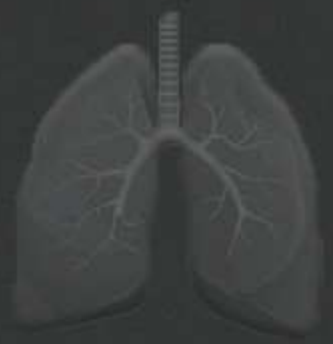


Idiopathic Pulmonary Arterial Hypertension

IPAH

Idiopathic Pulmonary Arterial Hypertension

- Pulmonary Arterial Hypertension (PAH) = Mean Pulmonary Artery Pressure (PAP) > 25 mmHg

- Idiopathic Pulmonary Arterial Hypertension (IPAH) = PAH in the absence of Lung or Left-sided Heart disease

Other Names

- Pre-capillary Pulmonary Hypertension
- Group – I Pulmonary Hypertension (in WHO Classification)
- Primary Pulmonary Hypertension

Idiopathic Pulmonary Arterial Hypertension

- Pulmonary Arterial Hypertension (PAH) = Mean Pulmonary Artery Pressure (PAP) > 25 mmHg
  - Idiopathic Pulmonary Arterial Hypertension (IPAH) = PAH in the absence of Lung or Left-sided Heart disease

Epidemiology

- Rare disorder, with no apparent cause
- Young & Middle-aged women

Prognosis

- Untreated, leads to right heart failure & death

Idiopathic Pulmonary Arterial Hypertension

Presentation

Symptoms

- Progressive dyspnea
- Malaise
- Chest pain
- Exertional syncope

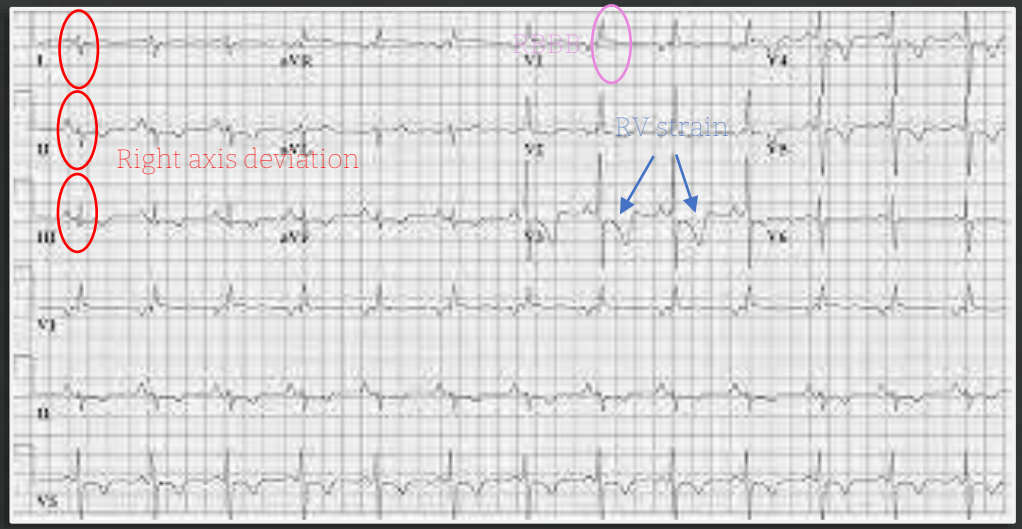
Signs

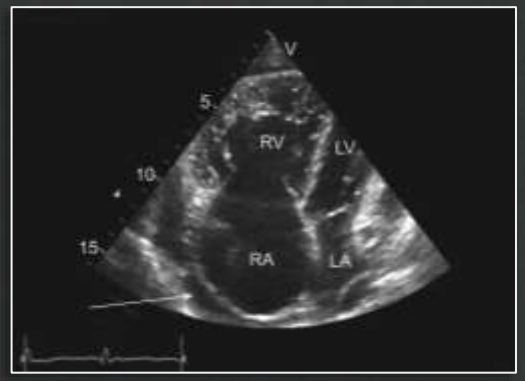
- Tachycardia
- Left parasternal heave (due to Right ventricular lift)
- Loud P2
- Right-sided S3
- Pulmonary &/or Tricuspid regurgitation murmurs
- Right sided heart failure (in advanced disease)
    - Jugular venous distention
    - Peripheral edema
    - Hepatomegaly
    - Ascites

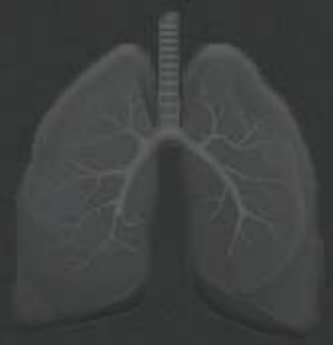
Idiopathic Pulmonary Arterial Hypertension

Investigations

- Electrocardiography (ECG)
  - Right axis deviation
  - Right bundle-branch block
  - Right ventricular hypertrophy or strain
- Echocardiography
  - ↑ right ventricular systolic pressure
  - +/- RV dilation *or* dysfunction
- Right-Heart Catheterization







Idiopathic Pulmonary Arterial Hypertension

Differential Diagnosis

- Pulmonary venous hypertension secondary *Left heart failure due to any reason*
- Sleep apnea & obesity hypoventilation syndrome
- Chronic Thromboembolic disease
- Diffuse Parenchymal lung diseases (DPLD)
- Chronic obstructive pulmonary disease (COPD)
- Sympathomimetic drug-induced pulmonary hypertension
- HIV-associated pulmonary hypertension
- Porto-pulmonary hypertension in Liver cirrhosis with portal hypertension

Idiopathic Pulmonary Arterial Hypertension

Treatment

- *Combinations of agents are generally more beneficial than any agent alone*
- *Clinical benefit versus side effects must be considered*

  - Calcium channel blockers - Response is seen only in patients who respond to vasodilator challenge
  - Endothelin receptor blockers & Phosphodiesterase-5 inhibitors
  - Prostacyclin analogues - in more severe cases
  - Anticoagulation - survival benefit
  - Lung *or* heart-lung transplantation should be considered in appropriate candidates

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