



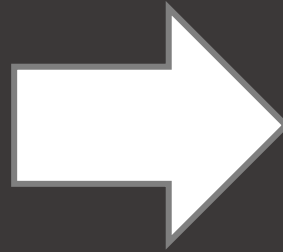
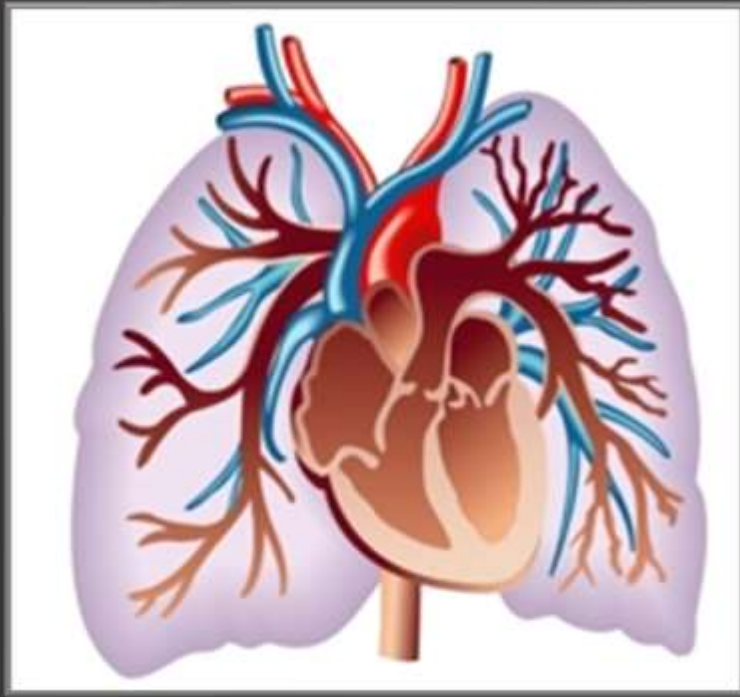
Pulmonary Hypertension

Classification, Investigations & Treatment

Pulmonary Hypertension (PH) =

Mean Pulmonary artery pressure

- At rest > 25 mmHg
- During exercise > 30 mmHg



CLINICAL FEATURES

Symptoms

- Dyspnea
- Chest pain
- Fatigue
- Palpitations
- Syncope

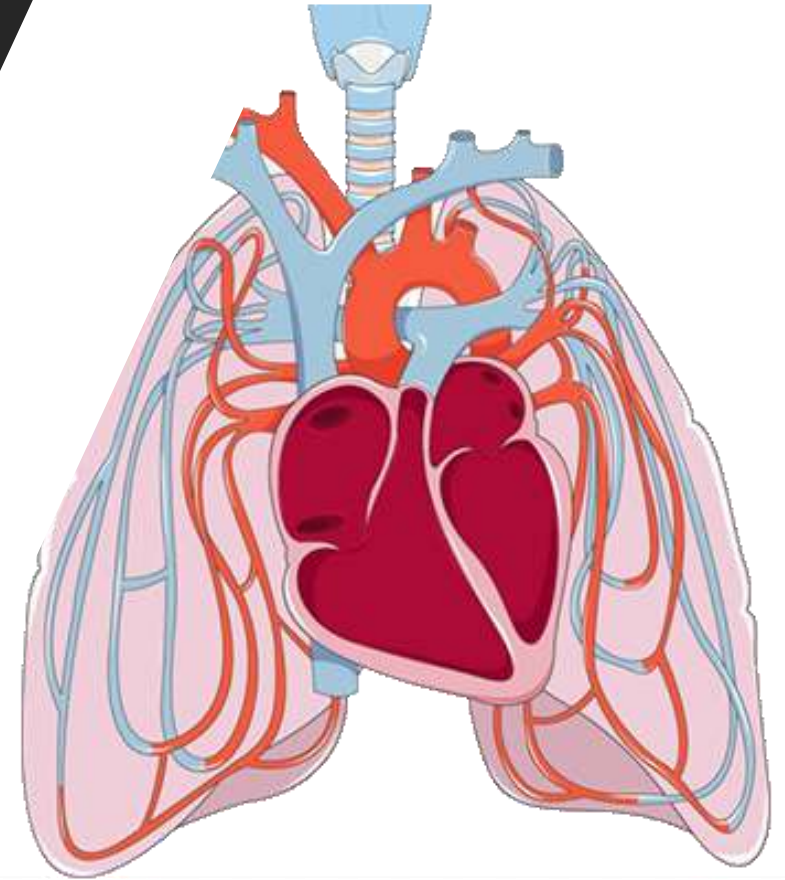
Signs

- Raised JVP
- Left parasternal heave
- Loud P2
- S3
- Signs of Right heart Failure
 - Oedema
 - TR
- Signs of interstitial lung disease or cardiac, liver or connective tissue disease

CLASSIFICATION

5 Groups

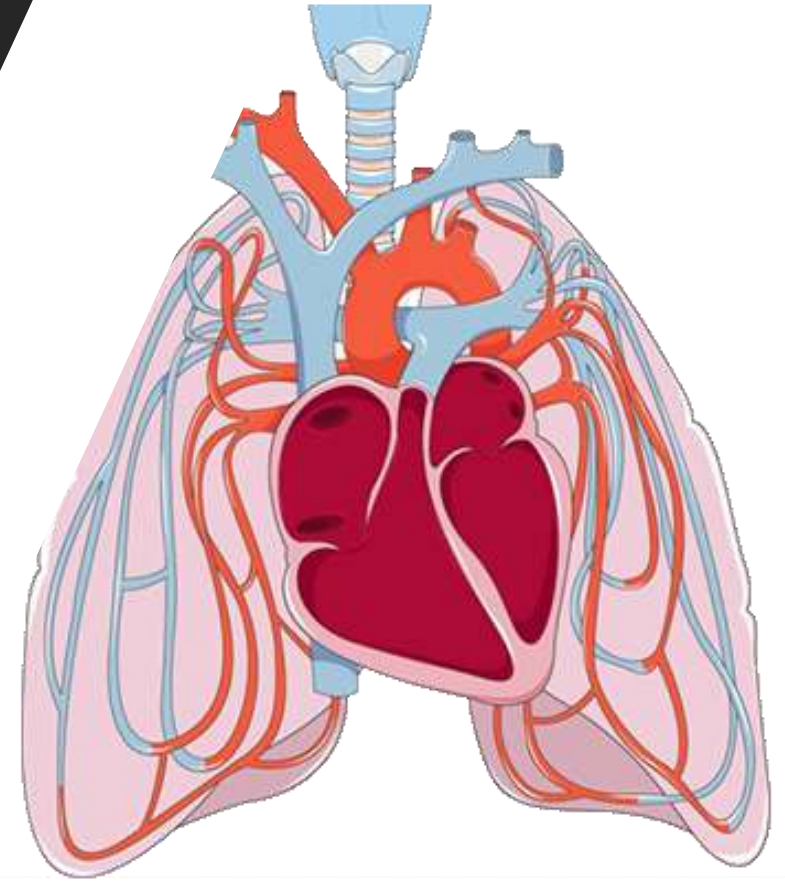
1. Pulmonary Arterial Hypertension (PAH)
2. PH due to Left heart disease
3. PH due to Lung diseases or Hypoxemia
4. PH due to Chronic venous thromboembolism
5. Others



Group 1

1. Pulmonary Arterial Hypertension (PAH)

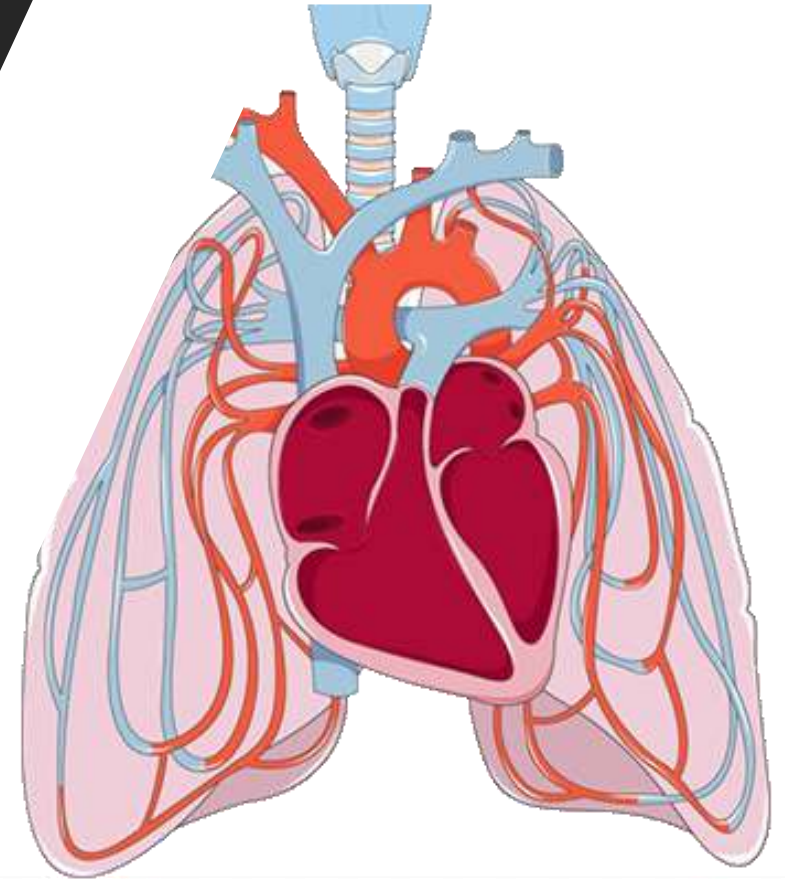
- a) Primary Pulmonary Hypertension (PPH)
- b) Secondary Pulmonary Hypertension
 - Connective Tissue Disease
 - congenital systemic to pulmonary shunts
 - Portal hypertension
 - HIV infection
 - Exposure to various drugs or toxins
 - Persistent pulmonary hypertension of newborn



Group 2

2. PH due to Left heart disease - Pulmonary Venous Hypertension

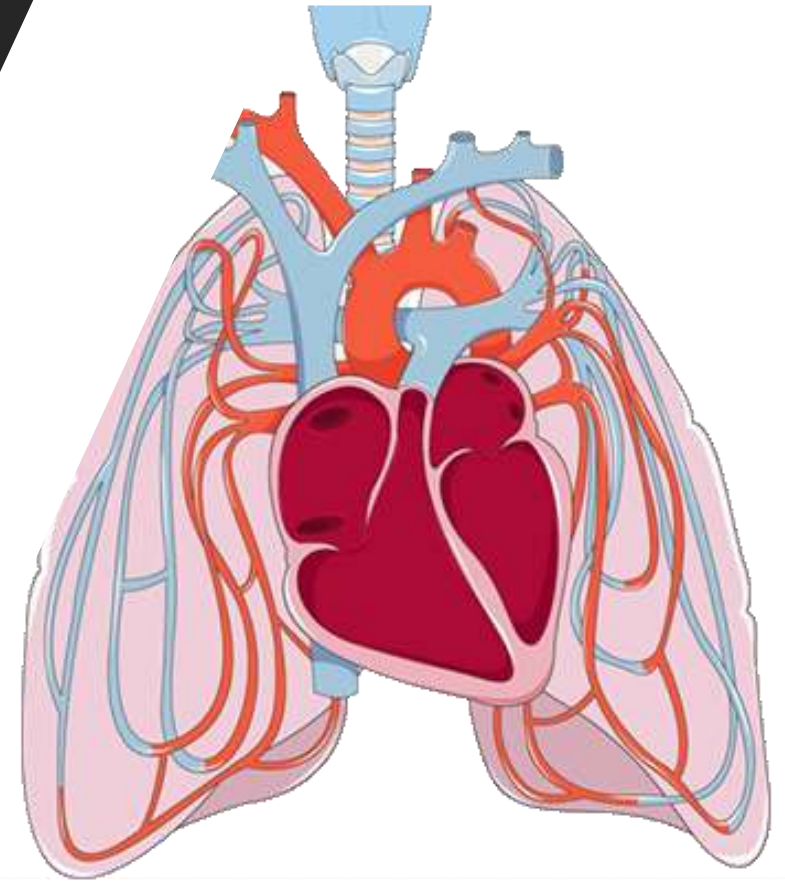
- Left-sided atrial or ventricular heart disease
- Left-sided valvular heart disease
- Pulmonary veno-occlusive disease
- Pulmonary capillary haemangiomatosis



3. Pulmonary hypertension associated with disorders of the respiratory system and/or hypoxaemia

- Chronic obstructive pulmonary disease
- Diffuse parenchymal lung disease
- Sleep-disordered breathing
- Alveolar hypoventilation disorders
- Chronic exposure to high altitude
- Neonatal lung disease
- Alveolar capillary dysplasia
- Severe kyphoscoliosis

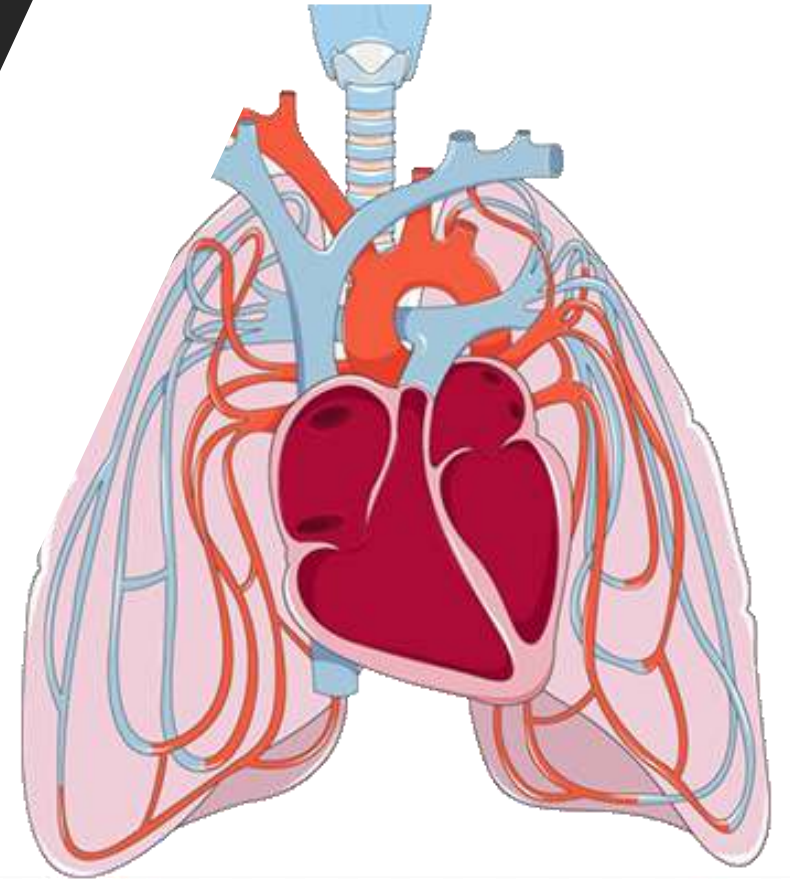
Group 3



Group 4

4. Pulmonary hypertension caused by chronic thromboembolic disease

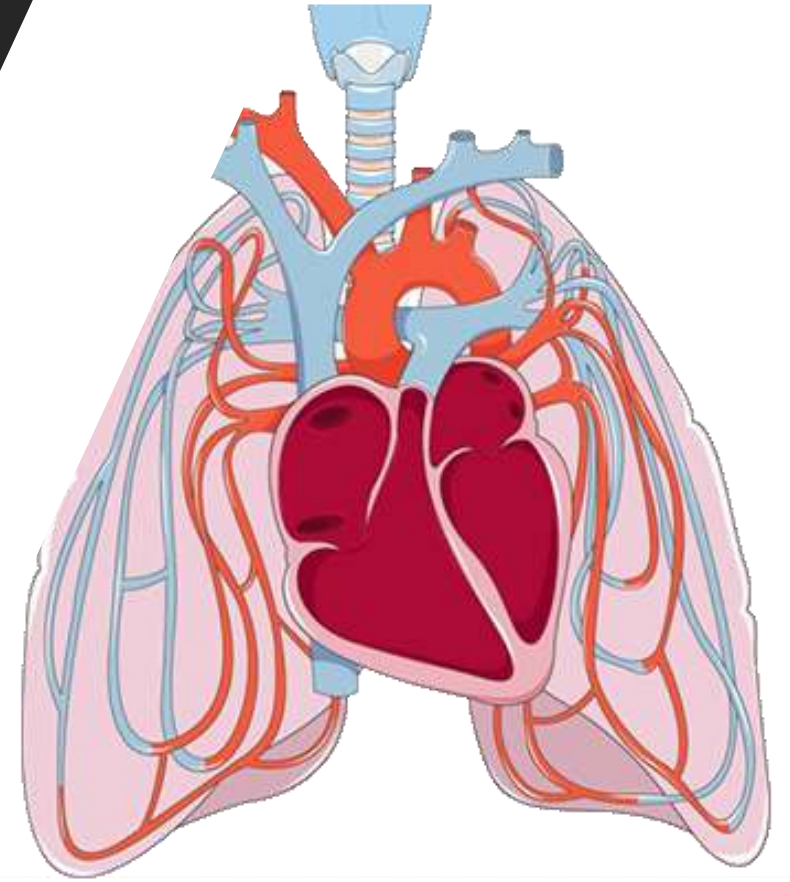
- Thromboembolic obstruction of the proximal pulmonary arteries
- In situ thrombosis
- Sickle-cell disease



Group 5

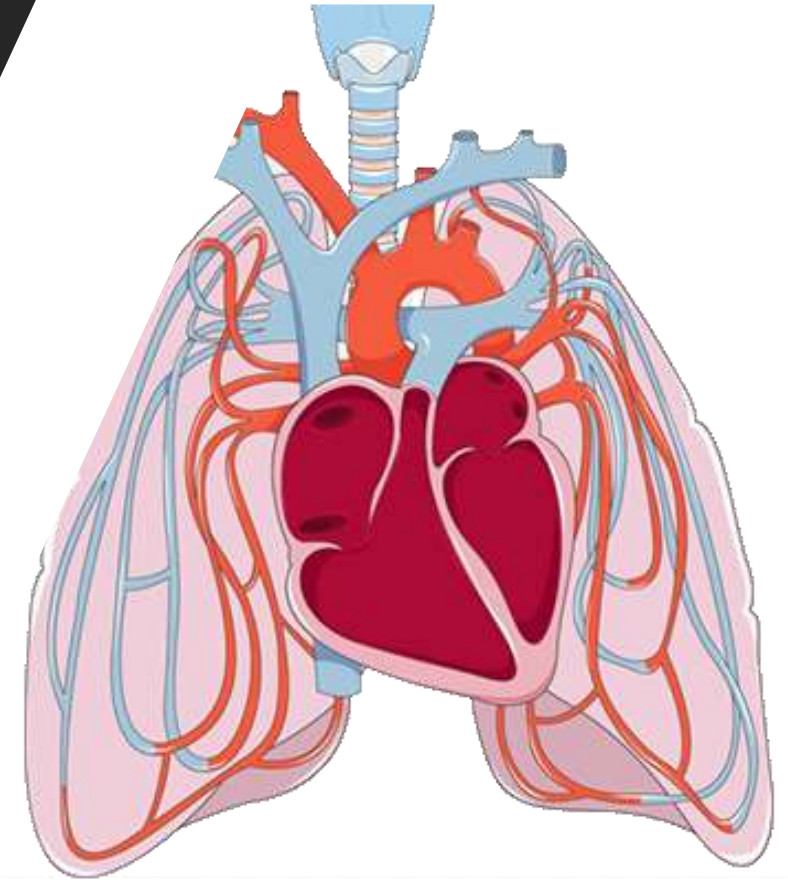
5. Miscellaneous

- Systemic disorders – Sarcoidosis, Lymphangiomyomatosis
- Blood disorders – Chronic hemolytic anemia, myeloproliferative disorders
- Extrinsic compression of central pulmonary veins



5 Groups

1. **Pulmonary Arterial Hypertension (PAH)**
 - Primary Pulmonary Hypertension (PPH)
 - Secondary Pulmonary Hypertension
2. **PH due to Left heart disease** - Pulmonary Venous Hypertension
3. **PH due to Lung diseases** or Hypoxemia
4. **PH due to Chronic venous thromboembolism**
5. **Others –**
Sarcoidosis, Myeloproliferative disorders, chronic hemolytic anemia



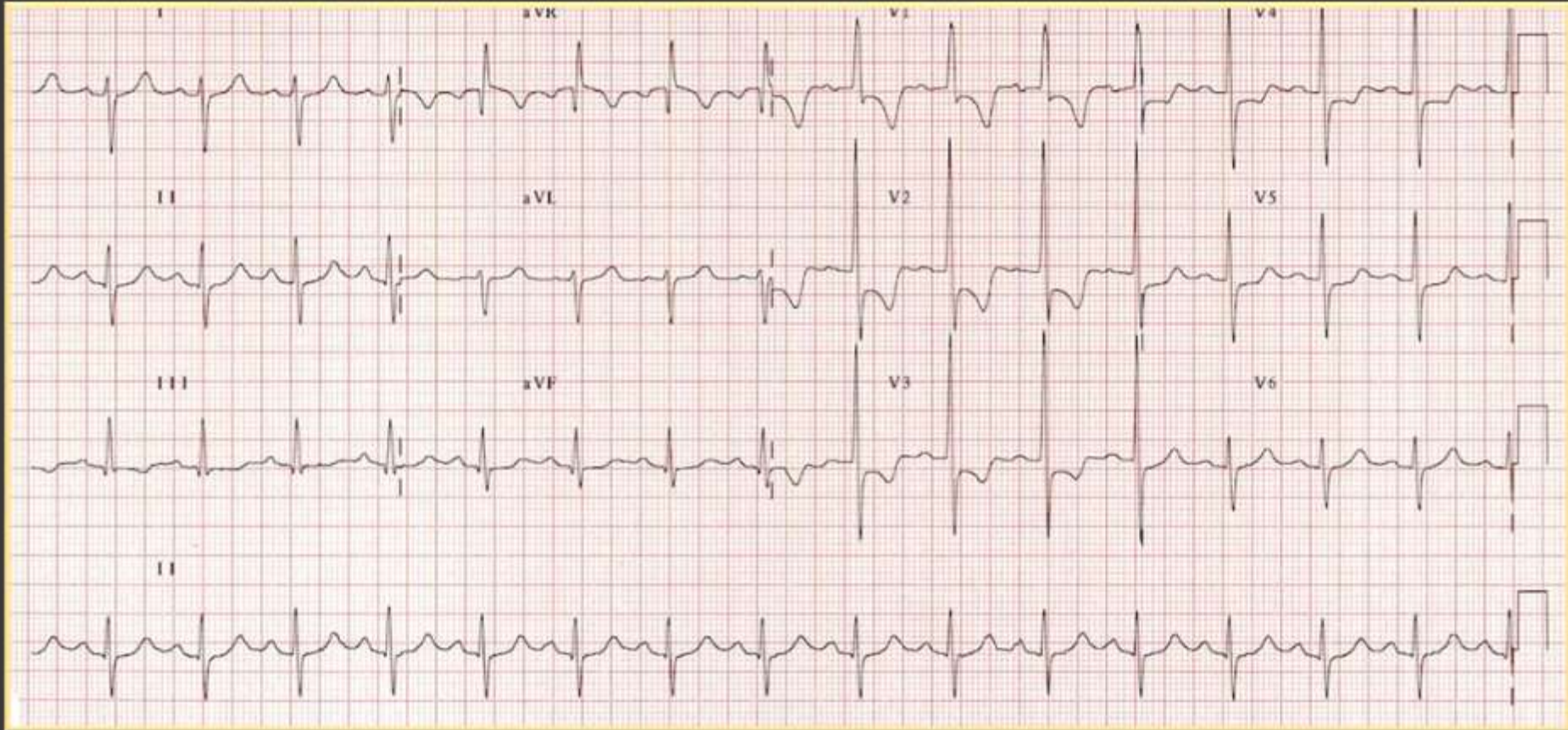
Pathology

- **Increased vasoconstrictors** – Thromboxane, Endothelin I
- **Decrease vasodilators** prostacyclin, NO
- **Vascular wall remodeling** – endothelial and smooth muscle proliferation, chronic inflammation
- Injured endothelium leading to **thrombosis in situ**

INVESTIGATIONS

ECG

- Right ventricular hypertrophy
- Right ventricular strain pattern



Chest X-ray

Enlarged pulmonary arteries,
peripheral pruning and right
ventricle enlargement



Echocardiography

Estimated PASP = $4 \times (\text{velocity of tricuspid regurgitant blood})^2 + \text{CVP}$

Right Heart Cathetrization

- Confirmation of diagnosis
- Pulmonary capillary wedge pressure (PCWP) = Left atrial pressure (< 18 mmHg)
- Vasodilator response

TREATMENT

General Steps

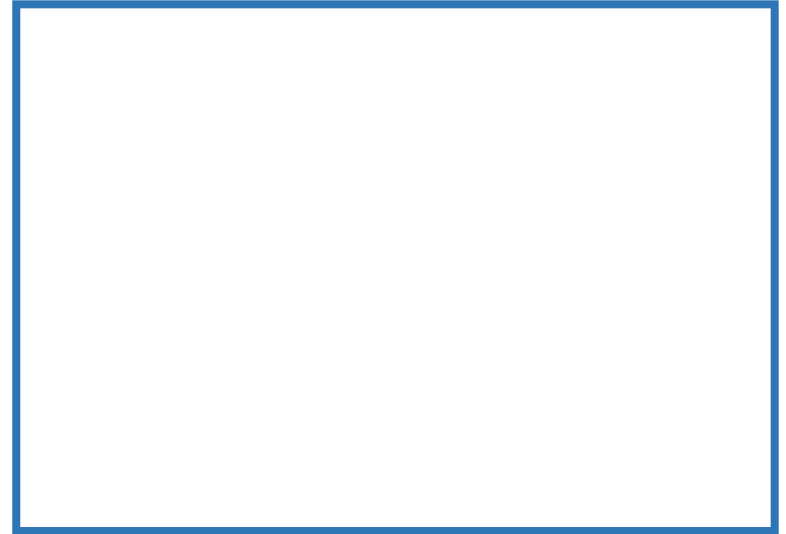
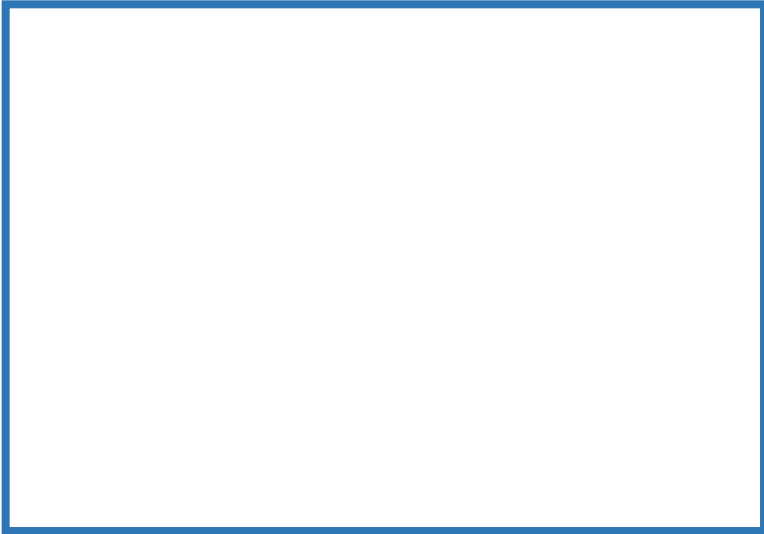
- Encourage to remain active
- **Diuretics** – Right heart failure
- **Oxygen** – maintain $\text{paO}_2 > 60$ mmHg
- **Anticoagulation**
- **Digoxin** – Atrial Tachyarrhythmias
- Pneumococcal and Influenza **vaccination**

Specific

- **High-dose calcium channel blockers** may be appropriate in those with an acute vasodilator response
- **Prostacyclin** infusions of epoprostenol or treprostinil or nebulised iloprost
- **Endothelin Receptor Antagonist** –Bosentan, Ambrisentan or Macitentan
- **Phosphodiesterase –V inhibitors** – Sildenafil, Tadalafil
- **Guanylate cyclase stimulator** - Riociguat

Selected cases

- Double-lung transplantation
- Pulmonary thrombo-endarterectomy
- Atrial septostomy



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