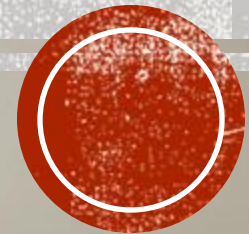


PULMONARY ARTERIAL HYPERTENSION



By Shahaana Shirin

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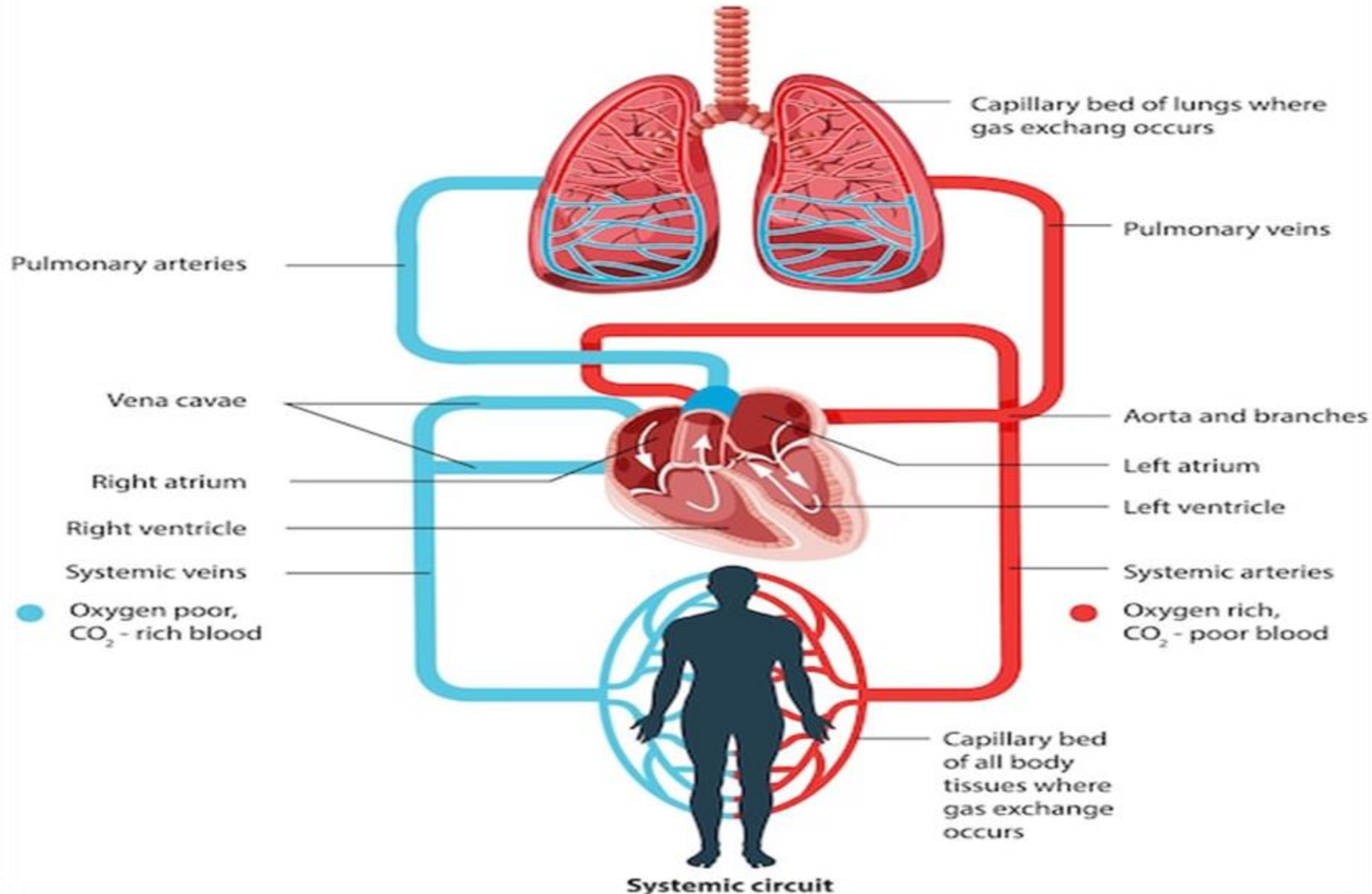
21st batch

PULMONARY CIRCULATION

- The pulmonary circulation is the vascular system that conducts blood from the right to left side of the heart through the lungs. Pulmonary arteries are very thin walled and distensible.
- Pulmonary vascular resistance (PVR) is a measure of the impedance to flow in the pulmonary vasculature
- PVR Depends on pulmonary artery pressure ,left atrial pressure and the cardiac output



PULMONARY CIRCUIT



DEFINITION

- Pulmonary hypertension (PH) is defined as an increase in blood pressure in pulmonary circulation (either in the arteries, or both in arteries and veins).
- Normal pressure as measured at right heart catheterization is 14-18 mm Hg at rest and 20-25 mm Hg on exercise. Hemo-dynamically it is defined as an increase in mean pulmonary arterial pressure to 25 mm Hg at rest.
- Definition may be refined by giving consideration of the pulmonary wedge pressure; the cardiac output and the transpulmonary pressure gradient (mean PAP - mean PWP).



ETIOLOGY

TABLE 3.39: Causes of secondary pulmonary HTN

Causes of pulmonary HTN	Mechanism
<i>Disorders of ventilation</i> • Obstructive sleep apnea • Morbid obesity (Pickwickian syndrome) • Cerebrovascular disease	They produce hypoxia and pulmonary vasoconstriction leading to pulmonary HTN
<i>Cardiac disorders</i> • Mitral valve disease (stenosis and regurgitation) • Left ventricular failure • Left atrial myxoma • Congenital heart disease with Eisenmenger syndrome	They cause pulmonary HTN either by increased left atrial pressure (mitral valve disease, LVF, myxoma) or by increased blood flow through pulmonary circulation (Eisenmenger syndrome)



TABLE 3.39: Causes of secondary pulmonary HTN (contd.)

Causes of pulmonary HTN	Mechanism
<i>Pulmonary vascular disorders</i> • Acute and chronic pulmonary thromboembolism • Multiple pulmonary artery stenoses • Pulmonary veno-occlusive disease • Parasitic infection, e.g. schistosomiasis	They cause increased resistance to blood flow and hence hypertension
<i>Diseases of the lung and parenchyma</i> • COPD (most common cause) • Interstitial lung diseases • Other chronic lung disorders (bronchiectasis, bilateral fibrocavity)	They decrease the surface area of pulmonary vasculature and hypoxia leading to pulmonary HTN
<i>Musculoskeletal and neurological disorders</i> • Kyphoscoliosis • Poliomyelitis • Myasthenia gravis	They cause chronic hypoventilation leading to hypoxia, pulmonary vasoconstriction and pulmonary HTN
<i>Miscellaneous</i> • Appetite-suppressant drugs, e.g. dexfenfluramine • Type 1 glycogen storage diseases • Lipid storage diseases, e.g. Gaucher's disease • Connective tissue diseases, e.g. SLE, scleroderma, sarcoidosis • Cirrhosis of liver • Sickle cell disease • HIV infection	They cause increased resistance to pulmonary blood flow and also endothelial damage leading to pulmonary HTN



PATHOPHYSIOLOGY

- Pulmonary hypertension leads to right ventricular hypertrophy and right heart failure . Right ventricular failure leads to peripheral edema . Right ventricular dilatation leads tricuspid annular dilatation leading to functional TR.



Table 7.97: WHO classifications of pulmonary hypertension-2008 (Dana point).

- **Pulmonary arterial hypertension**
 - Idiopathic pulmonary arterial hypertension (IPAH)
 - Familial: BMPR2, ALK 1, unknown
 - Associated with PAH
 - Connective tissue disease: Scleroderma, SLE, rheumatoid arthritis
 - Congenital heart disease
 - Portal hypertension (5–7% of patients)
 - HIV (0.5% of patients)
 - Drugs/toxins: aminorex, dexfenfluramine, or fenfluramine-containing products, cocaine, methamphetamine
 - Others: Hereditary hemorrhagic telangiectasia, hemoglobinopathies, myeloproliferative disorders, splenectomy
 - Associated with venous/capillary involvement
 - Pulmonary veno-occlusive disease (evidence of pulmonary vascular congestion)
 - Pulmonary capillary hemangiomatosis
 - Persistent PH of newborn
- **Pulmonary hypertension owing to left heart disease:** Systolic dysfunction, diastolic dysfunction and valvular disease.
- **PH secondary to chronic hypoxemia:** Chronic obstructive lung disease, interstitial lung disease, sleep disordered breathing, alveolar hypoventilation disorders, chronic exposure to high altitude, developmental abnormalities
- **Chronic thromboembolic pulmonary hypertension (CTEPH):** Thromboembolic obstruction of proximal or distal pulmonary arteries
- **Miscellaneous (usually extrinsic compression of pulmonary arteries):** Sarcoidosis, histiocytosis X, lymphangiomyomatosis, compression of pulmonary vessels (adenopathy, tumor, fibrosing mediastinitis, thyroid disorders, glycogen storage disease, Gaucher's disease)

CLASSIFICATION



SYMPTOMS

Cyanosis,
peripheral
edema

Raised JVP

Left
parasternal
heave

Large pulsatile
liver

Dyspnea

Syncope

Chest pain

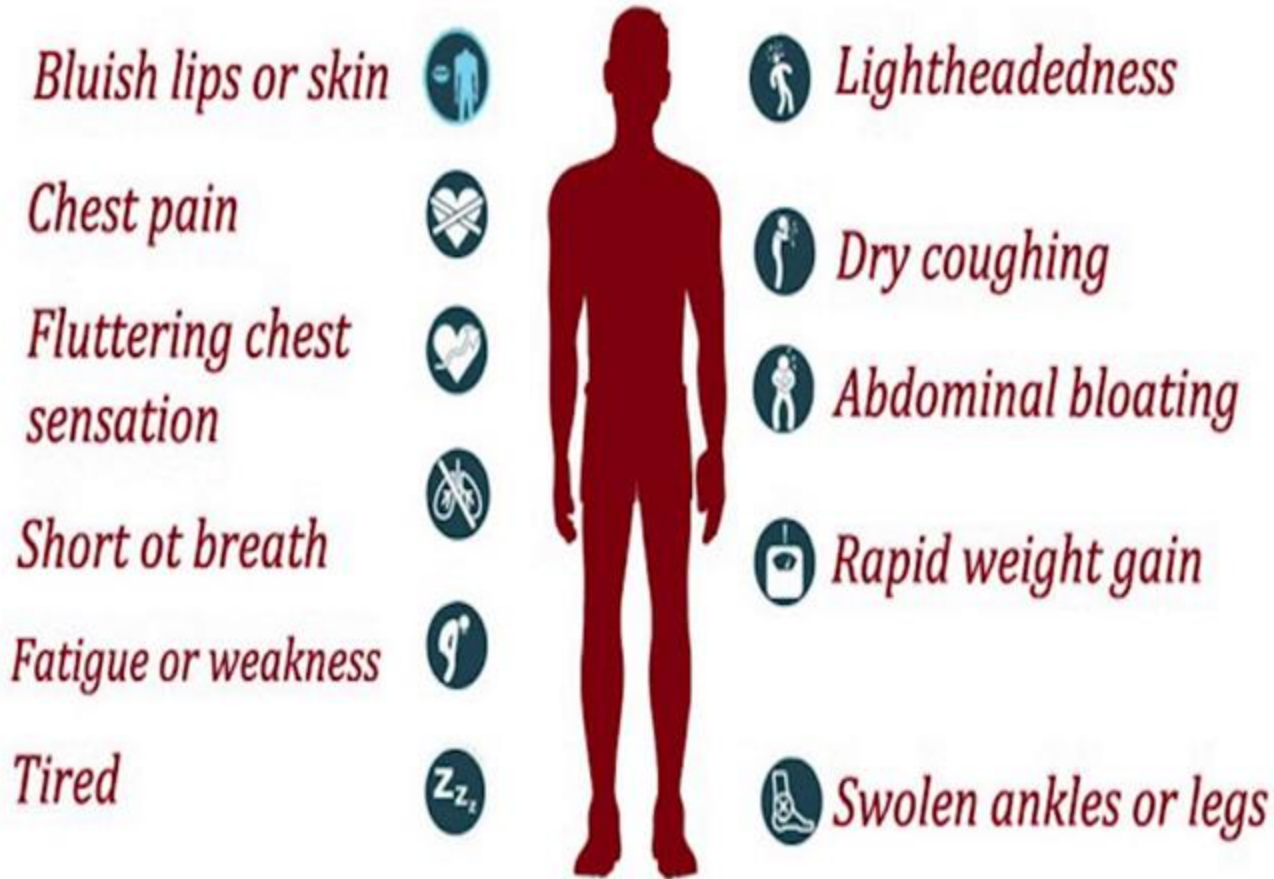
Palpitations

edema

Hoarseness
of voice



Pulmonary Hypertension Symptoms



SIGNS

- JVP raised with a prominent 'a' wave.
- A right ventricular (parasternal) heave is present, and a loud P2 is heard.
- Other findings include a right ventricular fourth heart sound, and an early diastolic murmur due to pulmonary regurgitation (Graham Steell murmur).
- This murmur is heard over the second and third left intercostal spaces, close to the sternum.

Tricuspid regurgitation may develop and indicates right ventricular failure. If TR develops, there is a pansystolic murmur and a large jugular 'v' wave.

Features of right ventricular failure such as ascites, peripheral edema and hepatomegaly may be present.



AUSCULTATORY SIGNS

- Pulmonary ejection sound (ES)
- Abnormal second heart sound (S_2): Loud P_2 , narrow splitting
- Right atrial fourth heart sound (S_4)
- Right ventricular third heart sound (S_3)
- Pulmonary ejection systolic murmur (ESM)
- Pulmonary early diastolic murmur (EDM): Graham Steel murmur
- Tricuspid pansystolic murmur (PSM)



INVESTIGATIONS

- Chest X-ray

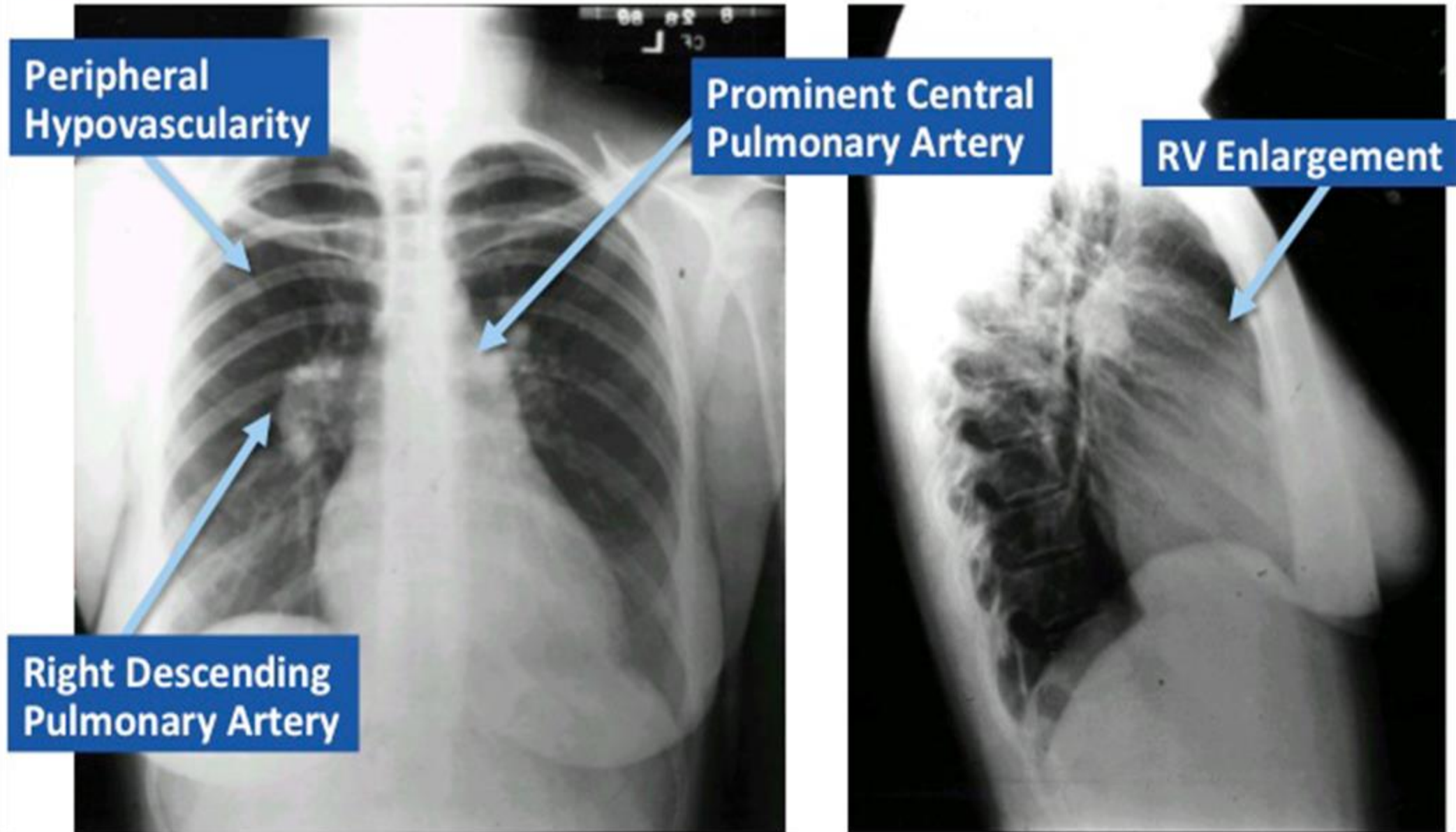
1. - Enlargement of the pulmonary artery and its main branches
2. - Peripheral "pruning" of vascular shadows
3. - Enlarged right atrium, and right ventricle

- Pulmonary ejection sound (es)

1. Abnormal second heart sound (S₂): narrow splitting
2. Right atrial fourth heart sound (S₄)
3. Right ventricular third heart sound (S₃)
4. Pulmonary ejection systolic murmur (ESM)
5. Pulmonary early diastolic murmur (EDM)



Figure 1. Characteristic X-Ray of a Patient with PAH



- Steel murmur

1. Tricuspid pansystolic murmur (PSM)
2. - Findings of underlying lung or cardiac pathology
3. Electrocardiogram: Shows right axis deviation, R/S wave ratio greater than one in lead VI, right bundle branch block (incomplete or complete) and increased P wave amplitude in lead II (due to right atrial enlargement).
4. Echocardiography: Most useful for detecting pulmonary hypertension and excluding cardiac disease. It shows tricuspid regurgitation, right atrial and ventricular hypertrophy, flattening of interventricular septum, small LV dimension, dilated pulmonary artery and secondary causes (if any).
5. Transthoracic echocardiography
6. Right heart catheterization: Mean PAP pressure at rest >25 mm Hg, with exercise >30 mm Hg, wedge pressure < 15 mm Hg.



- Other tests
- - Complete blood counts, prothrombin time, partial thromboplastin time and liver function tests.
- - Autoantibodies if autoimmune disease is suspected
- .- HIV-ELISA - Arterial blood gas
- - Sleep studies if sleep apnea suspected.
- - Pulmonary function tests
- - High-resolution computed tomography of chest.
- .- Helical CT
- - Ventilation-perfusion scanning to differentiate chronic thromboembolism from primary pulmonary hypertension.



TREATMENT

- Early recognition and treatment of the underlying cause.
- Medical: Diuretics mainly to treat edema from right heart failure, digoxin (for atrial tachyarrhythmias), oxygen (keep oxygen saturation $\geq 90\%$) and anticoagulants (IPAH) .
- PAH specific therapy- Calcium channel blockers: Indicated in patients who respond to vasodilators during catheterization.
 - Oral endothelin receptor antagonists (ERAs). Liver function be monitored monthly throughout the duration of use
- Phosphodiesterase type 5 inhibitors (PDE 5-1): For example, sildenafil, tadalafil,. Pneumococcal and influenza vaccination.
- Surgical therapy- Atrial septostomy (the creation of a right-to-left shunt).- Lung transplantation- Pulmonary thromboendarterectomy for chronic proximal thromboembolic pulmonary hypertension.



THANK YOU!

