Pulmonary Vascular Disease

any condition that affects the blood vessels along the route between the heart and lungs.

Blood travels from the heart, to the lungs, and back to the heart. Venous blood poor with Oxygen returns from the body's tissues through the veins back to the right side of the heart. The right heart pumps oxygen-poor blood through the pulmonary arteries into the lungs. This blood becomes filled with oxygen in a process of Gas Exchange.

The oxygen-rich blood returns from the lungs back to the left side of the heart. The left heart pumps the oxygen-rich blood into the body through the aorta and many other arteries.

Any part of the heart-lung blood circuit can become damaged or blocked, leading to pulmonary vascular disease.

Causes of Pulmonary Vascular Disease

1. Pulmonary Arterial Hypertension: Increased blood pressure in the pulmonary arteries (carrying blood away from the heart to the lungs). In which the pulmonary mean arterial pressure is greater than 25mmHg at rest, or greater than 30mmHg during exercise

The symptoms of pulmonary hypertension include the following:

* [Shortness of breath](https://en.wikipedia.org/wiki/Shortness_of_breath)
* [Fatigue](https://en.wikipedia.org/wiki/Fatigue_(medical))
* [Chest pain](https://en.wikipedia.org/wiki/Chest_pain)
* [Palpitations](https://en.wikipedia.org/wiki/Palpitations) ([heartbeat](https://en.wikipedia.org/wiki/Pulse) rate increased)
* Right-sided abdominal pain
* Poor [appetite](https://en.wikipedia.org/wiki/Appetite)
* [Lightheadedness](https://en.wikipedia.org/wiki/Lightheadedness)
* [Fainting](https://en.wikipedia.org/wiki/Syncope_(medicine))
* [Swelling](https://en.wikipedia.org/wiki/Swelling_(medical)) (legs/ankles)
* [Cyanosis](https://en.wikipedia.org/wiki/Cyanosis)

Causes

**WHO Group I** – Pulmonary arterial hypertension (PAH)

* [Idiopathic](https://en.wikipedia.org/wiki/Idiopathic)
* [Heritable](https://en.wikipedia.org/wiki/Heritable_disease) ([BMPR2](https://en.wikipedia.org/wiki/BMPR2), [ALK1](https://en.wikipedia.org/wiki/ALK1), [SMAD9](https://en.wikipedia.org/wiki/SMAD9), [caveolin 1](https://en.wikipedia.org/wiki/Caveolin_1" \o "Caveolin 1), [KCNK3](https://en.wikipedia.org/wiki/KCNK3) mutations)
* Drug- and toxin-induced (e.g., [methamphetamine](https://en.wikipedia.org/wiki/Methamphetamine), [amphetamine](https://en.wikipedia.org/wiki/Amphetamine), or [cocaine](https://en.wikipedia.org/wiki/Cocaine) use )
* Associated conditions: [Connective tissue disease](https://en.wikipedia.org/wiki/Connective_tissue_disease), [HIV infection](https://en.wikipedia.org/wiki/HIV_infection),  [Congenital heart diseases](https://en.wikipedia.org/wiki/Congenital_heart_disease), [Schistosomiasis](https://en.wikipedia.org/wiki/Schistosomiasis" \o "Schistosomiasis).

**WHO Group II** – Pulmonary hypertension secondary to [left heart](https://en.wikipedia.org/wiki/Left_heart) disease

* Left ventricular [systolic and diastolic dysfunction](https://en.wikipedia.org/wiki/Systolic_dysfunction)
* [Valvular heart disease](https://en.wikipedia.org/wiki/Valvular_heart_disease)

**WHO Group III** – Pulmonary hypertension due to [lung disease](https://en.wikipedia.org/wiki/Lung_disease), chronic [hypoxia](https://en.wikipedia.org/wiki/Hypoxia_(medical))

* [Chronic obstructive pulmonary disease (COPD)](https://en.wikipedia.org/wiki/Chronic_obstructive_pulmonary_disease)
* [Interstitial lung disease](https://en.wikipedia.org/wiki/Interstitial_lung_disease)
* [Sleep-disordered breathing](https://en.wikipedia.org/wiki/Sleep-disordered_breathing)

**WHO Group IV** – chronic pulmonary arterial obstruction

* Chronic [thromboembolic](https://en.wikipedia.org/wiki/Thromboembolism) pulmonary hypertension

**WHO Group V** – Pulmonary hypertension with unclear or [multifactorial](https://en.wikipedia.org/wiki/Multifactorial_inheritance) mechanisms

* [Hematologic diseases](https://en.wikipedia.org/wiki/Hematologic_disease): chronic [hemolytic anemia](https://en.wikipedia.org/wiki/Hemolytic_anemia) (including [sickle cell disease](https://en.wikipedia.org/wiki/Sickle_cell_disease))

Pathogenisis

The pathogenesis of pulmonary arterial hypertension (WHO Group I) involves the narrowing of blood vessels connected to the lung and within the lungs. This makes it harder for the heart to pump blood through the lungs,. Over time, the affected blood vessels become stiffer and thicker, in a process known as **fibrosis**. these changes result in an increased workload for the right side of the heart making it to enlarge and lead to failure.

Diagnosis

Clinical examination, sign of heart failure, elevated Jugular Venous Pressure

ECG, P- pulmonal and tachycardia

CXR finding of lung disease like COPD, increase cardiac size

Echo study revealing Dilated Right Side and pulmonary hypertension

CT scan finding of lung fibrosis , or pulmonary embolism.

Treatment is anticoagulant, venodilaters like endothelin receptor antagonist eg: **bosentan** , Phosphodiesterase type 5 inhibitors like **sildenafil**, diuretics, oxygen.

1. **Pulmonary Venous Hypertension**: Increased blood pressure in the pulmonary veins (carrying blood from the lungs, to the heart). Pulmonary venous hypertension is most often caused by congestive heart failure. A damaged mitral valve in the heart (mitral stenosis or mitral regurgitation) may contribute to pulmonary venous hypertension.

Diagnosis by clinical and physical examination, ECG and Echo study

Treatmant is treating the causes like valve replacement and treating the fluid overload with diuretics

**Pulmonary Embolism**:

closure of pulmonary artery with a foreign substances from another part of the venous system, most common is blood clots, Rarely, the embolism can be a large bubble of air, or ball of fat, or vegetation .

Symptoms include dyspnea (shortness of breath), tachypnea (rapid breathing), chest pain of a "pleuritic" nature (worsened by breathing), cough and hemoptysis (coughing up blood). About 15% of all cases of sudden death are attributable to PE.

As smaller pulmonary emboli tend to lodge in more peripheral areas without collateral circulation, they are more likely to cause lung infarction and small effusions (both of which are painful), but not hypoxia,. Larger PEs, which tend to lodge centrally, typically cause dyspnea, hypoxia, dyspnea, hemodynamic instability such as tachycardia low blood pressure, and fainting,

Causes

About 90% of emboli are from proximal leg deep vein thrombosis (DVTs) or pelvic vein thrombosis

Risk factors includes

**First**- surgical complication as in orthopedic surgery at or below the hip without prophylaxis. This is due to immobility during or after the surgery, as well as venous damage during the surgery.

**Second**- Pancreatic and colon cancer patients .This is due to the release of procoagulants.

**Third**- Pregnant individuals Due to "hypercoagulable state" in which increased expression of factors VII, VIII, X, Von Willebrand, and fibrinogen.

Diagnosis

Clinical, ECG sinus tacchycardia, CXR, Echo study. CT pulmonary Angiography

Treatment

* + - 1. Ant coagulant with heparin
      2. Thrombolytics like actylase