PULMONARY HYPERTENSION AND COR PULMONALE

Definition
- Normal pulmonary artery pressure = 25/10 with mean 15mmHg.
- PAH: at sea level systolic pulmonary arterial pressure >30mmHg OR, Mean pulmonary artery pressure > 25mmHg at rest or > 30mmHg during exercise.

Severity of Pulmonary Hypertension
Degree of disease severity:
- Mild (MPAP =25– 40 mmHg)
- Moderate (MPAP= 41– 55 mmHg)
- Severe (MPAP >55 mmHg)

Mechanisms and types
Pulmonary blood pressure = pulmonary flow × pulmonary vascular resistance.
So, pulmonary hypertension occurs when there is increase in flow and/or resistance.

Increased pulmonary blood flow as in (↑Preload) | Increased pulmonary vascular resistance (↑ Preload)
--- | ---
- Left to right shunt **added flow**: hyperdynamic pulmonary hypertension.
- Left sided heart failure **stagnant flow**: passive pulmonary hypertension.

Wall
- Contraction :-Hypoxemia/ Drugs: vasoconstrictive pulmonary hypertension.
- Thickening:- Pulmonary fibrosis: obliterative pulmonary hypertension.

Lumen
- Intravascular thrombosis: obstructive pulmonary hypertension.

N.B: Pulmonary hypertension could be encountered without evident causes, so termed idiopathic pulmonary hypertension OR primary pulmonary hypertension (PPH).

Causes of pulmonary hypertension
A) Primary pulmonary hypertension
- Arteriolar: Dietary and familial primary pulmonary hypertension.
- Venous: Pulmonary veno-occlusive disease.
- Capillary: Pulmonary capillary hemangiomatosis.

B) Secondary pulmonary hypertension
- COPD
- Parechymal diseases
- IPF - collagen diseases
- Pulmonary embolism - Bilharziases.
- Kyphoscoliosis - obesity hypoventilation syndrome.
- VSD - ASD - PDA.
- Mitral & aortic valve diseases.
- Left ventricular failure.
- Cardiomyopathy.
- Left atrial myxoma.

Clinical manifestations
<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Signs</th>
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<tbody>
<tr>
<td>1. Symptoms due to underlying pulmonary or cardiac disease</td>
<td>No signs</td>
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<tr>
<td>2. Dyspnea on exertion (insidious or gradual onset usually unexplained, so; Dx is delayed)</td>
<td>Congested Neck veins: Elevated JVP</td>
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<td>3. Angina- like chest pain</td>
<td>Cyanosis</td>
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<tr>
<td>4. Syncope</td>
<td>Precordial examination</td>
</tr>
<tr>
<td>5. Peripheral edema or ascitis</td>
<td>Signs of pulmonary artery dilatation.</td>
</tr>
<tr>
<td>6. Hoarseness of voice (Ortner syndrome)</td>
<td>Signs of right ventricular hypertrophy</td>
</tr>
<tr>
<td>7. Hemoptysis (rare)</td>
<td>Auscultation:</td>
</tr>
</tbody>
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Stigmata of causes of secondary PAH: scleroderma, cirrhosis, HIV, OSA/OHS

Investigations
Aiming to document the presence of pulmonary hypertension and disclose its etiology:
1. Electrocardiogram.
2. Chest x-ray, and CT- chest.
3. Echocardiography.
4. Cardiac catheterization.
5. Lung biopsy.

Treatment
1. Pulmonary vasodilators
   - Calcium channel blockers
   - Prostacyclin
   - Bosentan
   - Oxygen therapy (most effective pulmonary vasodilator in cases of hypoxia)
   - Inhaled nitric oxide

2. Diuretics to decrease preload.
3. Anticoagulant (specially for primary pulmonary hypertension).
4. Phlebotomy when the hematocrits above 55 to 60%.
5. Lung heart transplantation.
**COR PULMONALE**

**Definition**
- Cor pulmonale is right ventricular enlargement with or without failure resulting from pulmonary diseases after exclusion of left sided heart failure and congenital heart diseases.

**Causes of Cor Pulmonale**
- The same causes of pulmonary hypertension
- After exclusion of cardiac causes

**Pathogenesis of cor pulmonale**
1. The normal right ventricle can sustain its output in the face of abrupt increments in mean pulmonary arterial pressure of up to 50mmHg.
2. The right ventricle can cope with much higher after loads if they are applied gradually so that the right ventricle can undergo hypertrophy.
3. In time, if the pressure load continues, the right ventricle will fail.

**Types of Cor Pulmonale (according to onset)**

<table>
<thead>
<tr>
<th></th>
<th>Acute</th>
<th>Subacute</th>
<th>Chronic</th>
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</thead>
<tbody>
<tr>
<td><strong>Acute</strong></td>
<td></td>
<td></td>
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<tr>
<td>• Acute massive pulmonary embolism.</td>
<td></td>
<td>• Lymphangitis carcinomatosis</td>
<td>• COPD</td>
</tr>
<tr>
<td>• Tension pneumothorax.</td>
<td></td>
<td>• Recurrent minor pulmonary embolism.</td>
<td>• IPF</td>
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<tr>
<td>• Acute massive collapse.</td>
<td></td>
<td></td>
<td>• kyphoscoliosis</td>
</tr>
<tr>
<td><strong>Subacute</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Chronic</strong></td>
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</table>

**Clinical picture of cor pulmonale**
- Anorexia and discomfort in the right upper quadrant of the abdomen due to hepatic engorgement.
- Tricuspid insufficiency
- Pansystolic murmur.
- A prominent v wave appears in the jugular pulse.
- The liver often also shows expansive pulsations that are synchronous with the heart beat.
- Pedal edema.
- Arrhythmias.

**Diagnosis of cor pulmonale**
It requires the following:
- Diagnosis of pulmonary hypertension by the characteristic signs, symptoms and investigations.
- Exclusion of left sided heart failure and congenital heart diseases.
- Diagnosis of the cause of cor pulmonale.

**Treatment**
1. Pulmonary vasodilators as calcium channel blockers and prostacyclin, Inhaled nitric oxide.
2. Diuretics.
3. Oxygen therapy.
4. Anticoagulants

**Differentiation between Bilharzial and hypoxic cor pulmonale**

<table>
<thead>
<tr>
<th></th>
<th>Bilharzial cor pulmonale</th>
<th>Hypoxic cor pulmonale</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Etiology</strong></td>
<td>2ry to end arteritis oblitrans due to bilharzial granuloma</td>
<td>2ry to hypoxic vasoconstriction of pulmonary vessels.</td>
</tr>
<tr>
<td><strong>Symptoms</strong></td>
<td>low cardiac out put</td>
<td>hypoxia and or hypercapnia</td>
</tr>
<tr>
<td><strong>Complexion</strong></td>
<td>Pale</td>
<td>Cyanotic</td>
</tr>
<tr>
<td><strong>Pulse</strong></td>
<td>Thready</td>
<td>Pounding</td>
</tr>
<tr>
<td><strong>Hand</strong></td>
<td>Cold</td>
<td>Warm</td>
</tr>
<tr>
<td><strong>Spleen</strong></td>
<td>Enlarged</td>
<td>Normal or enlarged</td>
</tr>
<tr>
<td><strong>Cardiac auscultation</strong></td>
<td>Manifestations of pulmonary hypertension</td>
<td>Manifestations of pulmonary hypertension masked by emphysema</td>
</tr>
<tr>
<td><strong>Chest shape</strong></td>
<td>Normal</td>
<td>Barrel shaped</td>
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</tbody>
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