PORTOPULMONARY HYPERTENSION


Take home points:
1. Don’t confuse portopulmonary hypertension (the development of pulmonary hypertension in a cirrhotic patient with portal hypertension) with hepatopulmonary syndrome (shunting and V/Q mismatch due to arteriovenous malformations in the lung).
2. The most common symptom of portopulmonary hypertension are dyspnea on exertion.
3. Previously thought to be a contraindication to liver transplantation, now patients with portopulmonary hypertension can be safely transplanted if they are pre-treated with IV prostacyclin (Flolan®) and can achieve mean pulmonary artery pressures (PAP) < 40 mmHg.

Definition: look for “pre-capillary” pulmonary hypertension in the setting of pre-existing portal hypertension
- Mean pulmonary artery pressure > 25 mmHg at rest
- Pulmonary vascular resistance > 120 dyne s cm⁻⁵
- Pulmonary capillary wedge pressure (PCWP) < 15 mmHg
- Portal hypertension (portal pressure > 10 mmHg)

Clinical features:
- Portopulmonary hypertension occurs in 2-5% of all cirrhotics
- No gender predilection, mean age of presentation 5th decade of life (as compared to primary pulmonary hypertension which affects females > males and presents in the 4th decade of life).
- Symptoms: dyspnea on exertion (most common), syncope, chest pain, fatigue, hemoptysis, orthopnea
- Poor prognosis (if left untreated): mean survival 15 months; median survival 6 months.

Differential diagnosis:
- Hepatopulmonary syndrome (causing hypoxemia)
- Hepatic hydrothorax (causing hypoxemia, dyspnea)
- Other causes of liver disease + secondary pulmonary hypertension: anti-phospholipid syndrome, mixed connective tissue disease, schistosomiasis, sarcoidosis, systemic lupus erythematosus (SLE), microangiopathic hemolytic anemia (MAHA), HIV, toxins/drugs

Portopulmonary hypertension (PPHTN) vs. hepatopulmonary syndrome (HPS):

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<tr>
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<th>PPHTN</th>
<th>HPS</th>
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<tbody>
<tr>
<td>Pathophysiology</td>
<td>Similar to primary pulmonary hypertension with intense vasoconstriction of pulmonary capillaries as well as remodeled, thickened pulmonary vasculature</td>
<td>Arteriovenous (AV) shunting in the lung, predominantly at the bases. “Spider angiomata” in the lung.</td>
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<td>Symptoms</td>
<td>Dyspnea on exertion, syncope, orthopnea</td>
<td>Platypnea</td>
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<td>Physical findings</td>
<td>Loud P₂, RV heave, TR murmur, hypoxemia worsens with exercise (normal O₂ saturation at rest is common)</td>
<td>Hypoxemia, orthodeoxia</td>
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<td>Echocardiogram</td>
<td>Elevated pulmonary artery systolic pressure</td>
<td>Positive bubble study</td>
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<td>Treatment</td>
<td>Same as primary pulmonary hypertension; if mean PAP &lt; 40 mmHg, can safely undergo liver transplantation.</td>
<td>Liver transplantation</td>
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Note on treatment of PPHTN: patients with pulmonary hypertension are routinely anticoagulated because of the risk of in-situ pulmonary thrombi due to sluggish flow through the pulmonary circulation. However, patients with portal hypertension are at risk for variceal bleeding. Therefore, the risks and benefits of anticoagulation should be addressed in each patient individually; if anticoagulation is started, keep the INR on the low range of therapeutic.