HEPATOPULMONARY SYNDROME


Take home points:
1. Don’t confuse hepatopulmonary syndrome (shunting and V/Q mismatch due to arteriovenous malformations in the lung) with portopulmonary hypertension (the development of pulmonary hypertension in a cirrhotic patient with portal hypertension).
2. Look for platypnea and orthodeoxia; however, patients may be asymptomatic with only hypoxemia as a clue.
3. Treatment = lung transplantation which cures hepatopulmonary syndrome in 80% of patients. However, resolution can take up to 14 months.

Definition: look for the triad of liver disease, increased A-a gradient, and evidence of intrapulmonary vascular dilations.
- Note that the liver dysfunction need not be severe.
- Intrinsic cardiopulmonary disease must be excluded.
- Pleural effusions and airflow obstruction can co-exist with the hepatopulmonary syndrome.

Clinical features:
- 8% of cirrhotics manifest true clinical features of the hepatopulmonary syndrome (PaO2 < 60 mmHg).
- The patient may be asymptomatic (only 18% have dyspnea). Look for hypoxemia which can lead to clubbing and cyanosis.
- Platypnea: the opposite of orthopnea. Dyspnea improves when lying flat.
- Orthodeoxia: hypoxemia worsens upon sitting up and improves when lying flat.
- Platypnea and orthodeoxia occur because the pulmonary AVMs occur predominantly in the bases of the lung. Therefore, when sitting up or standing, blood pools at the bases of the lung with resultant increased AV shunting.

Pathophysiology:
- Hypoxemia due to intrapulmonary vascular abnormalities akin to spider angiomata found on skin of cirrhotics. This leads to AV shunting and V/Q mismatch.
- Note: unlike intracardiac shunting, 100% oxygen will (at least partially) correct the hypoxemia because shunting is not the only factor leading to low oxygen saturation. Dilated vasculature also leads to hypoxemia because of diffusion impairment. Increasing the oxygen level can overcome this problem.

Hepatopulmonary syndrome (HPS) vs. portopulmonary hypertension (PPHTN):

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<tr>
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<th>HPS</th>
<th>PPHTN</th>
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<tr>
<td><strong>Pathophysiology</strong></td>
<td>Arteriovenous (AV) shunting in the lung, predominantly at the bases. “Spider angiomata” in the lung.</td>
<td>Similar to primary pulmonary hypertension with intense vasoconstriction of pulmonary capillaries as well as remodeled, thickened pulmonary vasculature</td>
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<td><strong>Symptoms</strong></td>
<td>Platypnea</td>
<td>Dyspnea on exertion, syncope, orthopnea</td>
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<td><strong>Physical findings</strong></td>
<td>Hypoxemia, orthodeoxia</td>
<td>Loud P2, RV heave, TR murmur, hypoxemia worsens with exercise (normal O2 saturation at rest is common)</td>
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<td><strong>Echocardiogram</strong></td>
<td>Positive bubble study</td>
<td>Elevated pulmonary artery systolic pressure</td>
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<td><strong>Treatment</strong></td>
<td>Liver transplantation</td>
<td>Same as primary pulmonary hypertension; if mean PAP &lt; 40 mmHg, can safely undergo liver transplantation.</td>
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