58M with chronic liver disease, SOB, hypoxia

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Hepatopulmonary Syndrome (pulmonary-to-systemic shunting)
Tc-99m MAA, 4.2 mCi, IV

Normal lung perfusion

Uptake of radionuclide in the kidneys, brain, or both suggests shunting through the lung caused by an intrapulmonary shunt.
Using total lung radioactivity, correct for percentage of systemic circulation to the brain (20%)
~ 5% pulmonary-to-systemic shunting.
Hepatopulmonary Syndrome

Diagnosis facilitated by $^{99m}$Tc-MAA lung scan lung perfusion and brain uptake quantification:

- particles are efficiently trapped by the pulmonary capillaries such that normally only about 3% of the activity is seen outside the lungs
- Brain or kidney uptake indicates arteriovenous shunt (R-L shunt)
- R-L shunts develop in the lungs secondary to portal hypertension
  * Probably due to increased levels of circulating vasodilators, likely nitric oxide
  * Dilated precapillary and capillary vessels; pleural/pulmonary arteriovenous shunts, portopulmonary anastomoses
- Percentage R-L shunt is expressed as the fraction of perfusion reaching the body outside the lungs to the total body perfusion including the lungs
  * Percentage brain uptake > 6% is considered abnormal and suggestive of presence of an anatomic shunt in the lungs
Hepatopulmonary Syndrome

- Hepatopulmonary syndrome (HPS) triad:
  1. hepatic dysfunction (cirrhosis)
  2. hypoxemia (alveolar-arterial $O_2$ gradient of $>$15 mmHg; $>$20 mmHg in $>$64 years old patients)
  3. peripheral pulmonary arterial dilatation (due to right to left micro-shunts)

- Presentation: progressive dyspnea, cyanosis, clubbing in a patient with established cirrhosis

- Rx:
  - Mild to moderate: Observation $\pm$ oxygen
  - Severe HPS: Oxygen, liver transplantation
References


3. Statdx