Hepatopulmonary syndrome (HPS) is characterized by the presence of liver dysfunction, intrapulmonary vascular dilatation (IPVD), and gas exchange abnormalities, varying from increased alveolar-arterial oxygen gradient to severe hypoxia not explained by underlying cardiopulmonary disease.

- This syndrome usually occurs with cirrhosis but also has been described with noncirrhotic portal hypertension.
- The clinical manifestations are nonspecific and include dyspnea, platypnea, orthodeoxia, clubbing, cyanosis, and spider nevi.

Almost half of liver transplantation candidates have gas exchange abnormalities.

- Hypoxemia secondary to HPS is present in 13% to 15% of patients with end-stage liver disease.
- An imbalance in the expression of pulmonary vasodilating and vasoconstricting factors has been implicated in the pathogenesis of this phenomenon.
- Nitric oxide is thought to be responsible for the vasodilatation and the blunted hypoxic pulmonary vasoconstriction seen in HPS.

- Over 80% of patients have resolution or marked improvement, although time to resolution of HPS is quite variable and may take more than a year. The degree of preoperative hypoxia is not predictive of reversibility.
- Liver transplantation remains the only currently available treatment for HPS and should be considered in patients with HPS having PaO2 less than 60 mm Hg.