

Samir Gupta, MD, FRCPC
Canadian Hepatopulmonary Syndrome (HPS) Program
30 Bond Street, Suite 6-044 Toronto, ON M5B 1W8
Appointments/Information: 416-864 6060 x 2252
Fax: (416) 864-5649



HPS
CANADA

Information about Hepatopulmonary Syndrome For Healthcare Providers

Hepatopulmonary syndrome (HPS) is a pulmonary complication of liver disease which affects 5-32% of patients with liver disease (including non-cirrhotic portal hypertension).

It is defined by a triad of features:

- Liver dysfunction or portal hypertension
- Intrapulmonary vascular dilatations (IPVDs) diagnosed on contrast echocardiography
- Abnormal oxygenation (on arterial blood gas: an alveolar-arterial oxygen gradient 15-20 mmHg or $\text{PaO}_2 < 80$ mmHg)

HPS is not limited to patients with severe liver dysfunction; in fact, patients with normal or only mildly deranged synthetic liver function can have severe HPS. While the majority of patients presenting with HPS have known pre-existing liver disease, about 20% initially present with pulmonary complaints.

Chest X-ray and chest CT scan are usually normal in HPS; a lack of radiographic abnormalities does not rule out HPS.

A classic complaint in HPS is platypnea (breathlessness experienced in the sitting/standing position which improves in the supine position). This often corresponds to an objective finding of **orthodeoxia** (a drop of 4 mm Hg or 5% in PaO_2 after moving from the supine to the sitting/standing position).

When to refer for HPS evaluation:

Any patient with liver dysfunction and/or portal hypertension who:

Complains of significant, unexplained dyspnea

Has abnormal saturation on pulse oximetry*

and/or

and/or

Has platypnea/orthodeoxia, clubbing or cyanosis*

Abnormal oxygenation on an arterial blood gas

* Clubbing (positive predictive value of 78%)

* Cyanosis (positive predictive value of 100%)

* In liver disease patients, a saturation of $\leq 97\%$ has a 68% specificity for an alveolar-arterial oxygen gradient > 20 mmHg

The Canadian Hepatopulmonary Syndrome Program is a clinical and research program specializing in the diagnosis and management of patients with HPS.

We are pleased to accept new referrals and/or clinical questions.

For more information about HPS and the Canadian HPS Program, please visit our website hpscure.com