

## CLINICAL IMAGES

# Shortness of breath while sitting up: hepatopulmonary syndrome

Chih-Hsin Lee MD, Shih-Tsung Cheng MD

See related article by Lin and colleagues, page 77

**Competing interests:** None declared.

This article has been peer reviewed.

**Affiliations:** From the Department of Internal Medicine, Buddhist Tzu Chi General Hospital (Lee, Cheng), Taipei, Taiwan; the School of Medicine, Tzu Chi University (Lee), Hualien, Taiwan; and the Graduate Institute of Biomedical Electronics and Bioinformatics, National Taiwan University (Lee), Taipei, Taiwan.

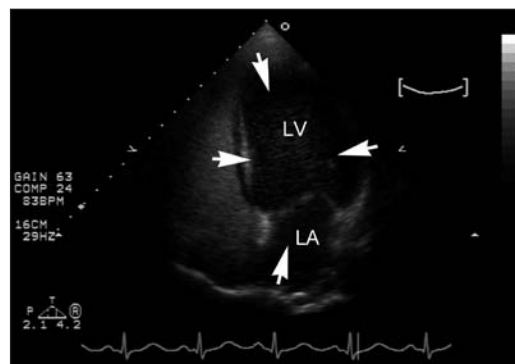
**Correspondence to:** Dr. Chih-Hsin Lee, chlee.tw@gmail.com

CMAJ 2011. DOI:10.1503/cmaj.090576

A 70-year-old man with hepatitis B-associated cirrhosis and hepatocellular carcinoma presented with a two-week history of shortness of breath. He was also experiencing ongoing fatigue, anorexia, edema and exertional dyspnea, which had begun two months earlier after transcatheter arterial chemoembolization. His partial pressure of oxygen was 67 mm Hg, and the alveolar–arterial gradient was elevated at 43 mm Hg when he breathed room air in a supine position. His oxygen saturation decreased from 96% to 91% when he sat upright from a recumbent position. Chest radiography showed increased, diffuse interstitial markings (Appendix 1, at [www.cmaj.ca/cgi/content/full/cmaj.090576/DC1](http://www.cmaj.ca/cgi/content/full/cmaj.090576/DC1)). Contrast echocardiography showed enhancement in the left heart five beats after microbubbles opacified the right heart (Figure 1). Hepatopulmonary syndrome was diagnosed and was managed with oxygen, bed rest, salt restriction and diuretics.

Hepatopulmonary syndrome is characterized by intrapulmonary vascular dilatations and shunting, which lead to arterial deoxygenation in patients with advanced liver disease.<sup>1</sup> The prevalence of the syndrome among such patients ranges from 13% to 47%.<sup>2</sup> Orthodeoxia–platypnea, defined as a decrease in the partial pressure of oxygen of at least 5% or at least 4 mm Hg in an upright position, is highly specific for hepatopulmonary syndrome.<sup>1,3,4</sup> The mortality associated with hepatopulmonary syndrome is high, with a median survival of 10.6 months.<sup>2</sup> The only definitive treatment is liver transplantation, for which a five-year survival rate of 76% has been reported.<sup>4</sup>

A definite diagnosis of hepatopulmonary syndrome requires scintigraphy with technetium-99m-labelled macroaggregated albumin, pulmonary angiography or contrast echocardiography. Contrast echocardiography is more sensitive and less invasive than angiography,<sup>1</sup>



**Figure 1:** Four-chamber view of contrast echocardiography using agitated saline, showing microbubbles opacifying the left heart five beats after they were seen in the right heart. LA = left atrium, LV = left ventricle. See also videos online at [www.cmaj.ca/cgi/content/full/cmaj.090576/DC1](http://www.cmaj.ca/cgi/content/full/cmaj.090576/DC1).

but the latter may be considered when direct arteriovenous communication is strongly suspected. In hepatopulmonary syndrome, the microbubbles during contrast echocardiography pass through the abnormally dilated pulmonary vasculature into the left heart with a delay of three to six heart beats. In an intracardiac shunt, enhancement appears in the left heart within the first three cardiac cycles.<sup>5</sup>

## References

1. Fallon MB, Mulligan DC, Gish RG, et al. Model for end-stage liver disease (MELD) exception for hepatopulmonary syndrome. *Liver Transpl* 2006;12:S105-7.
2. Lange PA, Stoller JK. The hepatopulmonary syndrome. *Ann Intern Med* 1995;122:521-9.
3. Cheng TO. Mechanisms of platypnea-orthodeoxia: What causes water to flow uphill? *Circulation* 2002;105:e47.
4. Rodriguez-Roisin R, Krowka MJ. Hepatopulmonary syndrome — a liver-induced lung vascular disorder. *N Engl J Med* 2008; 358:2378-87.
5. Ho V. Current concepts in the management of hepatopulmonary syndrome. *Vasc Health Risk Manag* 2008;4:1035-41.



See videos online of the contrast echocardiography: [www.cmaj.ca/cgi/content/full/cmaj.090576/DC1](http://www.cmaj.ca/cgi/content/full/cmaj.090576/DC1)