Summary

Two forms of pulmonary vascular disorders can complicate chronic liver disease: the hepatopulmonary syndrome and portopulmonary hypertension. Both influence survival and candidacy for orthotopic liver transplantation.

The hepatopulmonary syndrome is defined as the triad of liver disease, pulmonary gas exchange abnormalities leading to hypoxaemia, and widespread pulmonary vascular dilatation.

The main pathological feature of hepatopulmonary syndrome is gross dilatation of the pulmonary precapillary and capillary vessels (15 to 100 μm in diameter when the patient is at rest), coupled with an absolute increase in the number of dilated vessels, causing ventilation–perfusion imbalance, increased intra-pulmonary shunt, and diffusion impairment to oxygen.

Dyspnea on exertion, at rest, or both and cyanosis are the most prevalent symptoms in hepatopulmonary syndrome. The diagnosis of hepatopulmonary syndrome is established when the following three points are fulfilled. (1) Chronic liver disease. (2) Arterial hypoxaemia, defined by a reduced partial pressure of arterial oxygen or more accurately by an increased alveolar-arterial difference in the partial pressure of oxygen. (3) Intrapulmonary vascular dilatation, detected either by two dimensional contrast echocardiography or macroaggregated albumin lung perfusion scan.

No clearly effective medical therapy for HPS has been found, and liver transplantation is the only proven therapy for hepatopulmonary syndrome based on the total resolution or significant improvement in gas exchange postoperatively.