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Summary

Shortness of breath: Pulmonary vascular complications from liver disease

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Paediatrics | Pancreas | Hepatobiliary

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Hepatopulmonary syndrome and portopulmonary hypertension are distinct pulmonary vascular complications of liver disease requiring active screening in transplant candidates, with liver transplantation curative for HPS but controversial for portopulmonary hypertension.

KEY POINTS

- Hepatopulmonary syndrome resolves in about 95% of cases after liver transplantation, usually within 6 to 12 months, with long-term outcomes equal to patients without HPS, making it an indication for transplant referral with possible extra waiting list points for severe disease.
- The speaker reported that without liver transplant, hepatopulmonary syndrome has a five-year survival of only 23%, and the syndrome doubles mortality risk independent of liver disease severity.
- Hepatopulmonary syndrome diagnosis requires proving vascular dilatations via contrast echocardiography, gas exchange abnormalities with alveolar-arterial oxygen gradient of 15 mmHg or more, and presence of liver disease; arterial blood gas is most sensitive for screening but pulse oximetry misses mild-to-moderate cases.
- The speaker stated there is no current medical treatment for hepatopulmonary syndrome, with small studies including one RCT with sorafenib all failing to show benefit.
- Portopulmonary hypertension occurs in less than 1% of end-stage liver disease patients and has a five-year survival of only 14% if untreated; the speaker recommends vasodilator treatment, with macitentan the only drug tested specifically in an RCT for this condition.
- For portopulmonary hypertension, liver transplant should only be considered when a primary hepatic indication exists, patients are pre-treated with vasodilators to reach safe thresholds, and care is provided at experienced centres, as outcomes are unpredictable with uncontrolled pulmonary pressures.

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