# A Re-transplant case for hepatopulmonary syndrome without liver cirrhosis or portosystemic shunt after living-donor liver transplantation


## Abstract

Hepatopulmonary syndrome (HPS) is a disease of gas exchange caused by intrapulmonary shunting secondary to liver disease–associated intrapulmonary vascular dilation. HPS is characterized by the triad of cirrhosis, chronic liver disease (CLD), or portosystemic shunting (PSS); arterial hypoxemia; and intrapulmonary arteriovenous shunting in the absence of a primary cardiopulmonary anomaly. We encountered a rare case of HPS without liver disease or PSS. The patient was an 8-year-old girl who underwent living donor liver transplantation (LDLT) shortly after developing fulminant hepatitis at 11 months of her age. Eight years after LDLT, hypoxemia and shortness of breath developed. The shunt ratio on $^{99m}$Tc-macroaggregated albumin lung perfusion scintigraphy ($^{99m}$Tc-MAA lung scan) was 32%. She had no cardiopulmonary disease, so we diagnosed her illness as HPS. We did not find cirrhosis, CLD, or PSS as a cause of HPS. We thought the graft was the cause of HPS. Second transplantation was planned. One year after the diagnosis of HPS, the shunt ratio on $^{99m}$Tc-MAA lung scan worsened to 42%, digital clubbing appeared, and hypoxemia was worsening. Thus, we performed second LDLT promptly.

Afterwards, the shunt ratio on $^{99m}$Tc-MAA lung scan normalized (6%) and cyanosis resolved. We determined that the graft was the cause of HPS; the typical causes of HPS were not

clearly revealed in the histological examination of the second liver explant. Acute rejection occurred twice after LDLT, so we speculated that HPS occurred because the graft became stressed over the long-term.