

## Liver Transplantation for the Abernathy Malformation

**TO THE EDITOR:** Previously in the *Journal*, we reported on a 4-year-old boy with the hepatopulmonary syndrome, a condition characterized by intrapulmonary shunting due to dilatation of pulmonary vessels in the setting of liver disease or dysfunction.<sup>1</sup> The Abernathy malformation, a congenital anomaly of the splanchnic vasculature in which portal venous blood is diverted into the inferior vena cava, was diagnosed. This is a follow-up report on the outcome.

In the 2 years after presentation, worsening hypoxemia and exercise intolerance developed, with an oxygen saturation of 70% while the patient was receiving 6 liters of oxygen per minute by nasal cannula; the patient also had progressive weight loss. With the prospect of interrupting the progression of the hepatopulmonary syndrome, a cadaveric liver transplantation was performed. During the procedure, we confirmed the presence of the Abernathy malformation, since the portal vein drained into the inferior vena cava (Fig. 1). We divided the congenital portacaval shunt and repaired the caval defect just before performing portal-vein anastomosis. Pathological evaluation of the explanted liver revealed the

absence of portal veins in the hilum and in the portal tracts, further confirming the Abernathy malformation.

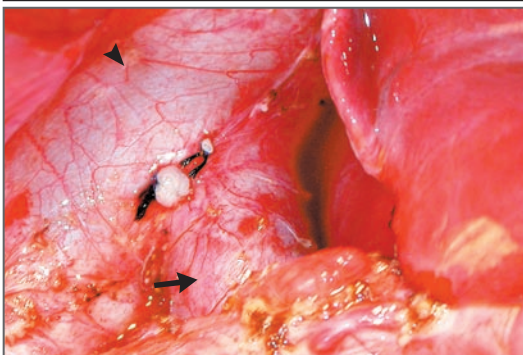
Immediately after the procedure, profound hypoxemia that was only partially responsive to 100% inhaled oxygen developed, implicating postoperative enhancement of pulmonary capillary vasodilatation and intrapulmonary shunting. The patient's condition gradually stabilized after treatment with supplemental oxygen, inhaled nitric oxide, and sildenafil. However, a significant degree of intrapulmonary shunting persisted, and the patient was ultimately discharged while receiving 5 liters of oxygen per minute and sildenafil.

In sharp contrast to the acuity of the patient's condition during the postoperative period, his condition improved dramatically after his discharge from the hospital. Within 4 months after transplantation, the patient's oxygen saturation was 100% while he was breathing ambient air, his exercise tolerance had improved, normal growth had resumed, and the digital clubbing had already begun to resolve. He continues to do extraordinarily well, with no signs or symptoms of pulmonary limitation.

Liver transplantation has reportedly been successful in reversing the hepatopulmonary syndrome due to a variety of liver conditions,<sup>2,3</sup> and we now report on a successful liver transplantation for treatment of the hepatopulmonary syndrome caused by the Abernathy malformation.

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**Figure 1. Intraoperative Photograph of the Abernathy Malformation in a 7-Year-Old Boy.**

During liver transplantation, the portal vein (arrow) in the patient's native liver can be seen draining directly into the infrahepatic vena cava (arrowhead).

1. Case Records of the Massachusetts General Hospital (Case 31-2004). *N Engl J Med* 2004;351:1667-75.
2. Lange PA, Stoller JK. The hepatopulmonary syndrome: effect of liver transplantation. *Clin Chest Med* 1996;17:115-23.
3. Taillé C, Cadranel J, Bellocq A, et al. Liver transplantation for hepatopulmonary syndrome: a ten-year experience in Paris, France. *Transplantation* 2003;75:1482-9.