Brief Reports

Exploiting Phenotypic Plasticity for the Treatment of Hepatopulmonary Shunting in Abernethy Malformation

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An 11-year-old boy presented with exercise intolerance due to chronic hypoxemia. Work-up revealed a diagnosis of hepatopulmonary syndrome (HPS) secondary to a congenital extrahepatic portal-venous shunt (Abernethy malformation). Plasticity in the developing liver was exploited as a strategy for the treatment of HPS. With use of a staged endovascular approach, the portosystemic vascular circuitry was modified in a manner that facilitated progressive growth and development of the severely hypoplastic and underdeveloped intrahepatic portal venous system. After completion of the final procedure, the patient's intrahepatic portal veins were normal in appearance; 2 months later, signs and symptoms of HPS completely resolved. The patient remains free of HPS stigmata after 2 years.

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Abbreviations: HPS = hepatopulmonary syndrome, TIPS = transjugular intrahepatic portosystemic shunt

THE Abernethy malformation is a rare congenital condition characterized by abnormal development of the vitelline venous system that manifests as a spectrum of congenital extrahepatic portosystemic shunts (1–3). Two variants have been described on the basis of the nature of the portosystemic shunt and the developmental status of the intrahepatic portal venous system (4). In type I, the extrahepatic portal vein is completely diverted to the systemic circulation with complete absence of the intrahepatic portal venous system. In type II, which is less common, there is an abnormal communication between the extrahepatic portal venous system and the systemic circulation in the presence of an intact intrahepatic portal venous system.

Hepatopulmonary syndrome (HPS) is a hypoxemic condition resulting from diffuse pulmonary arterial dilatation in the setting of liver disease that occurs in up to 30% of patients with chronic liver disease (5). A common feature in HPS, similar to the Abernethy malformation, is diversion of portal blood from hepatic first-pass metabolism, usually due to underlying liver disease (6,7). Given their similarities, it is not completely surprising that the Abernethy malformation is also a rare but recognized cause of HPS (8–10). Accordingly, liver transplantation currently serves as definitive therapy for both conditions (7–16).

Herein, we describe a case of complete reversal of HPS in a boy with an apparent type II Abernethy malformation by exploiting the concept of plasticity through use of a staged endovascular approach. Phenotypic plasticity is a structure or system's ability to remodel or alter its phenotype in response to environmental change (17,18). Specifically, we exploit portal venous plasticity in the pediatric liver by staged modulation of portosystemic venous hemodynamic forces as a means to induce structural development of the severely hypoplastic portal venous system. In this case, plasticity was exploited for therapeutic reversal of HPS.

CASE REPORT

Under existing institutional guidelines, institutional review board approval is not required for case reports.

An 11-year-old boy presented with a 9-year history of shortness of breath and fatigue with minimal exertion. He demonstrated resting oxygen saturations of 85% on room air and postexercise saturations of 77%. He was a 5th grade student keeping up with peers academically, although he was described as disruptive. He was an only child, the product of a full-term uncomplicated pregnancy. There is no family history of congenital liver or heart disease or vascular anomalies. His chest radiograph was normal. At age 10 years, transthoracic contrast medium–enhanced echocardiography revealed a positive test but no

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structural anomaly. This was confirmed with cardiac catheterization, which showed no evidence of congenital cardiac anomalies; however, pulmonary angiography was suggestive of diffuse pulmonary arteriovenous malformations and helped confirm pulmonary vein oxygen saturations of 75%–80%. Physical examination revealed a noncyanotic, normal-appearing 140-cm-tall boy weighing 39.5 kg (50th percentile for weight and 25th percentile for height). His examination was notable for clubbing of the digits. Liver span and position was normal. Laboratory values are shown in the **Table**. Initial high-resolution axial computed tomography (CT) of his chest helped confirm the presence of diffuse pulmonary congestion but was otherwise unremarkable (**Fig 1a**).

Summary of Laboratory Values

Variable	Measurement
Total bilirubin (mg/dL)	2.1
Aspartate aminotransferase (IU/L)	43
Alanine aminotransferase (IU/L)	39
Alkaline phosphatase (IU/L)	152
Total protein (g/dL)	6.5
Albumin (g/dL)	3.4
Hemoglobin (g/dL)	14.7
Hematocrit (%)	44.3
Ammonia (μmol/L)	52

With the cardiac catheterization showing no cardiac congenital anatomic anomaly yet a positive pulmonary shunt and the lung CT scan revealing no pulmonary arteriovenous malformations but suggesting increased pulmonary vascularity, possible subdiaphragmatic congenital abnormalities needed to be ruled out—necessitating contrast-enhanced abdominal CT. This revealed a large 14-mm-diameter vascular communication between the extrahepatic portal circulation and the inferior vena cava (**Fig 1b**). The shunt had the appearance of an end-to-side portocaval fistula without evidence for an intrahepatic portal venous system. Multiple nonspecific low-attenuation defects were scattered throughout the liver, and these were suggestive of mild underlying chronic liver disease with nodular regenerative hyperplasia. The hepatic venous system was unremarkable; however, the proper hepatic artery was enlarged, measuring 5.5 mm in diameter. There was no secondary evidence of portal hypertension such as varices, splenomegaly, or splenorenal shunts. Given these findings and, in particular, given the absence of an identifiable intrahepatic portal venous system at cross-sectional imaging, an initial diagnosis was made of HPS secondary to type I Abernethy malformation.

Selective catheter angiography was subsequently performed, confirming the presence of the portacaval fistula. However, upon direct catheter interrogation of the portal vein, an irregular minute intrahepatic portal venous network with submillimeter portal veins was recognized (**Fig 1c**). Hemodynamic assessment demonstrated systemic venous pressures measuring 4 mm Hg at the level of the superior vena cava; pressure measurements obtained via the shunt did not demonstrate a gradient, with an equilibrated portal pressure of 4 mm Hg as well. However, at temporary balloon occlusion of the shunt the portal pressure increased to 26 mm Hg, yielding a portosystemic gradient of 22 mm Hg. Nearly stagnant mesenteric venous flow was also noted at portography at the time of balloon occlusion.

A percutaneous liver biopsy was performed to assess liver histology. The major feature was absence of portal veins in most of the visualized portal triads (**Fig 1d, 1e**). Portal veins were present in only four of the 13 portal triads. Despite this finding, given the visualization of an extremely hypoplastic portal venous network at previous catheter angiography, a new diagnosis of HPS secondary to an Abernethy type II malformation was made.

Treatment approaches were discussed among specialists in interventional radiology, pediatric hepatology, pulmonology, and transplant surgery. Isolated surgical ligation of the fistula was not considered possible given the marked and sustained elevation in portal pressures observed during the balloon occlusion test and the near-stagnant flow in the mesenteric vein, reflecting the disproportionate size of the shunt relative to the miniscule intrahepatic portal veins present. Liver transplantation was also an option and is generally accepted as the definitive therapy for treating HPS in patients with cirrhosis and Abernethy type 1 malformations.

An additional option included endovascular closure of the Abernethy malformation, although, to our knowledge, there has been no previously published experience with this approach. The endovascular strategy we devised was based on the hypothesis that his HPS was a deleterious emergent property that ultimately arose from the lack of hepatic first-pass metabolism of mesenteric blood through a dormant but otherwise functionally intact liver due to the portacaval shunt. Accordingly, our primary treatment objective was to facilitate normal hepatic processing of mesenteric blood to simulate a healthy liver by redirecting mesenteric flow through the liver. However, this could only be achieved by encouraging profound development of his severely underdeveloped and abnormal intrahepatic portal venous system while simultaneously avoiding fulminant prehepatic edema and/or thrombosis of the mesenteric circulation. Accordingly, we hypothesized that adaptive development of the intrahepatic portal venous system by regulated increments in intrahepatic portal flow and pressure might result in resolution of HPS (**Fig 2 a–c**).

Endovascular therapy was chosen because it was the least-invasive option, and transplantation could be undertaken if catheter-based intervention failed. The repair was performed in three stages.

STAGE 1

Portal and systemic venous pressure measurements at this time both measured 11 mm Hg. To mitigate potential portal venous hypertension and mesenteric stasis upon endovascular occlusion of the congenital shunt, a secondary portal venous outflow tract was established by creating a transjugular intrahepatic portosystemic shunt (TIPS) from the right hepatic vein directly to the portal end of the portacaval fistula. A 10 mm × 8-cm–covered stent dilated to 8 mm was used to establish the TIPS (Via-

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