

## Case Report

# Congenital Hepatoportal Arteriovenous Fistula Leading to Segmental Portal Hypertension and Splenic Vein Thrombosis

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### INTRODUCTION

Congenital hepatoportal arteriovenous fistulas are a rare cause of segmental portal hypertension and splenic vein thrombosis (1–4). These may be intrahepatic or extrahepatic in location and present during infancy or early childhood with portal hypertension and gastrointestinal bleeding secondary to varices, ascites, malabsorption, anemia, and congestion of the bowel (5). In childhood, arterioportal fistulas are usually intrahepatic and regarded as congenital. The congenital fistulas are generally due to cavernous hemangioma, hereditary telangiectasia, and Ehlers-Danlos syndrome. The acquired arterioportal fistulas are usually traumatic, iatrogenic (following liver biopsy), or a result of rupture of a hepatic artery aneurysm (6).

### CASE REPORT

A 16-year-old boy presented with a history of abdominal pain and distension for the previous 3 months. There were no complaints of fever, loss of weight, loss of appetite, or gastrointestinal bleeding. There was no history of trauma, liver biopsy, or any other predisposing condition, nor any cutaneous stigmata to suggest Ehlers-Danlos syndrome or hereditary telangiectasia. The hemogram and liver function tests were within normal limits. Ultrasound examination revealed evidence of massive splenomegaly. On color Doppler imaging, there was gross dilation of the left portal vein and the hepatic artery. High-velocity pulsatile flow was encountered in the portal vein, which showed a hepatofugal flow pattern. The splenic vein was visualized only at the splenic hilum. Computed tomography confirmed the aforementioned findings and showed a thrombosed splenic vein with multiple collateral channels at the splenic hilum, fundus of stomach, and around the gastroesophageal junction. The portal vein was dilated with gross ectasia of its left branch (Figs. 1 and 2). The celiac axis and the hepatic

artery were also dilated. These findings were conclusive for portal hypertension as a result of arterioportal fistula, so endoscopy was not performed and the patient was taken directly for angiography. Intraarterial catheter angiography demonstrated the fistulous communication between the right hepatic artery and left portal vein (Figs. 3 and 4). On selective superior mesenteric artery injection in the venous phase, there was opacification of the superior mesenteric vein but the portal vein was not filling up, which was attributed to reversed flow in the portal vein. The mean pressure measurement taken at the time of angiography in the right hepatic artery and left portal vein was 85 mmHg, with no pressure gradient. Because this was thought to be a congenital fistula, the surgical option was considered and the child underwent splenectomy with hepatic artery ligation. He did well postoperatively, and follow-up Doppler examination revealed thrombosis of the hepatic artery with restoration of antegrade flow in the portal vein. The child had no new complaints during 1 year of follow-up.

### DISCUSSION

The earliest description of a hepatoportal arteriovenous fistula was made by Sachs in 1892 in a 60-year-old man who died of bleeding esophageal varices secondary to portal hypertension (7). Congenital intrahepatic hepatoportal arteriovenous fistulas are extremely rare and are generally attributed to hereditary hemorrhagic telangiectasia (ie, Rendu-Osler-Weber syndrome) or Ehlers-Danlos syndrome (8). The acquired fistulas are encountered more frequently than congenital fistulas and have been described after blunt abdominal trauma, penetrating liver injury (including liver biopsy), or in association with hepatocellular carcinoma (5,9).

Patients with hepatoportal arteriovenous fistulas may be asymptomatic or can present with varying clinical manifestations depending on the size, location, and duration of the arterioportal communication. The patients



**FIG. 1.** Axial contrast medium–enhanced computed tomography section through liver shows aneurysmal dilation of the left branch of portal vein (arrow). Note presence of gastric varices as tortuous vascular channels in stomach wall.

with congenital lesions usually present in early childhood with failure to thrive and show features of portal hypertension, hepatosplenomegaly, and malabsorption.

There are various imaging modalities used for the diagnosis of hepatoportal arteriovenous fistula, including Doppler ultrasonography, biphasic computed tomography, magnetic resonance angiography, and catheter angiography. The Doppler examination usually reveals a dilated hepatic artery associated with an arterialized and dilated portal venous system with reversal of flow in the portal vein (10). Dynamic contrast medium–enhanced biphasic computed tomography reveals a dilated hepatic artery and portal vein with early filling of the fistula and opacification of the portal venous system in the arterial phase. Contrast medium–enhanced magnetic resonance angiography



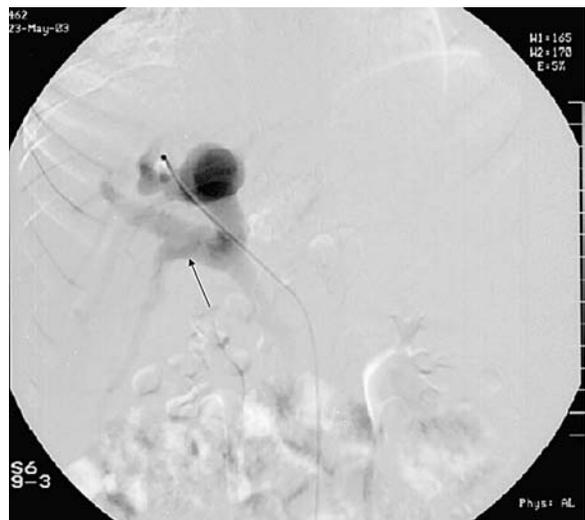
**FIG. 2.** Axial contrast medium–enhanced computed tomography section at the level of coeliac axis shows dilated coeliac axis (white arrow) and hepatic artery (black arrow). Note presence of gross splenomegaly and thrombosed splenic vein (bold white arrow).



**FIG. 3.** Digital subtraction flush abdominal aortogram shows dilated hepatic artery and arterioportal fistula.

offers a new, noninvasive, and nonionizing method of demonstrating hepatoportal arteriovenous fistulas. However, catheter angiography still remains the investigation of choice because it not only helps in confirming the diagnosis but it also offers the option of endovascular treatment. Early visualization of the portal vein after aortic injection or early filling of any part of the portal venous system on selective injection of the hepatic arterial tree is pathognomonic for hepatoportal arteriovenous fistula.

The treatment options available for these rare hepatoportal arteriovenous fistulas are surgery and intraarterial embolization. Although transcatheter coil embolization is considered the treatment of choice for acquired intrahepatic hepatoportal arteriovenous fistulas (11,12), it has



**FIG. 4.** Selective right hepatic arterial angiogram depicts fistula and opacification of portal venous branches (arrow).

shown limited success in the treatment of congenital fistulas as a result of rapid development of other intrahepatic fistulous communications (11,13). Thus, surgical ligation of the hepatic arterial feeding vessels is considered the best treatment for congenital hepatoportal arteriovenous fistulas. In our index case, there was no evidence to suggest an acquired cause of fistula, so the child underwent surgery and was doing well at 1-year follow-up. The present case was unique because there was splenic vein thrombosis associated with hepatic arterioportal fistula. We speculate that splenic vein thrombosis in this patient developed because of gradual enlargement of the fistula, progressive increase in portal venous pressure, and stagnation of flow.

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