

Congenital Hepatoportal Arteriovenous Fistula associated with Intrahepatic Portal Vein Aneurysm and Portohepatic Venous Shunt: Case Report ¹

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A congenital hepatoportal arteriovenous fistula associated with an intrahepatic portal vein aneurysm is a rare type of arteriovenous malformation. Only 14 pediatric cases have been reported to the best of the authors' knowledge. An intrahepatic shunt between the portal and systemic veins is also relatively rare. We report a case of a congenital hepatoportal arteriovenous fistula associated with an intrahepatic portal vein aneurysm and a portohepatic venous shunt in a neonate who presented with tachypnea and melena.

Index words : Congenital hepatoportal arteriovenous fistula

Portal vein, aneurysm

Portohepatic venous shunt

Ultrasound (US)

Angiography

An intrahepatic arteriovenous fistula is usually acquired as a result of trauma, including a liver biopsy, or in association with hepatic tumors and ruptured aneurysms of the hepatic artery (1). A congenital hepatoportal arteriovenous fistula is extremely rare and there are very few reports. The common clinical manifestations are the features of portal hypertension such as bowel congestion, gastrointestinal bleeding, malabsorption, ascites and splenomegaly (2). An intrahepatic portovenous shunt is also rare (3). Duplex Doppler sonography is the modality of choice for both diagnosing and evaluating abnormal vascular communications along with their hemodynamic consequences in the liver, which can be confirmed by angiography (4). We describe a case of a male neonate who was diagnosed by

duplex Doppler sonography with a congenital hepatoportal arteriovenous fistula and portohepatic venous shunt associated with an intrahepatic portal vein aneurysm, which was confirmed by angiography.

Case Report

A 3740-g male neonate that was born after 41 weeks' gestation via a normal spontaneous vaginal delivery was referred to this hospital for an assessment of his tachypnea. A chest radiogram at birth showed a mild cardiomegaly, and the echocardiography revealed grade III tricuspid regurgitation and a mild right-side heart enlargement. At one week of age, the baby exhibited melena, which required transfusions. There was no history of jaundice and the examination showed no hepatosplenomegaly and cyanosis. The liver function tests were normal, but the hemoglobin level was 6.4 g/dl. Gray-scale ultrasonography of the abdomen demonstrated a 1.5 cm in diameter saccular dilatation of the right portal vein (Fig. 1A), and the color Doppler exami-

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nation showed mixed colors and turbulent flow (Fig. 1B). The blood flow in the portal vein was hepatopetal but the duplex Doppler examination revealed an arterialized pattern (Fig. 1C). There were no features of portal hypertension including hepatosplenomegaly, venous

collaterals and ascites. A diagnosis of an intrahepatic portal vein aneurysm and arteriportal shunt between the portal vein aneurysm and the hepatic artery was established. Digital subtraction abdominal aortography confirmed an intrahepatic arteriportal fistula with a

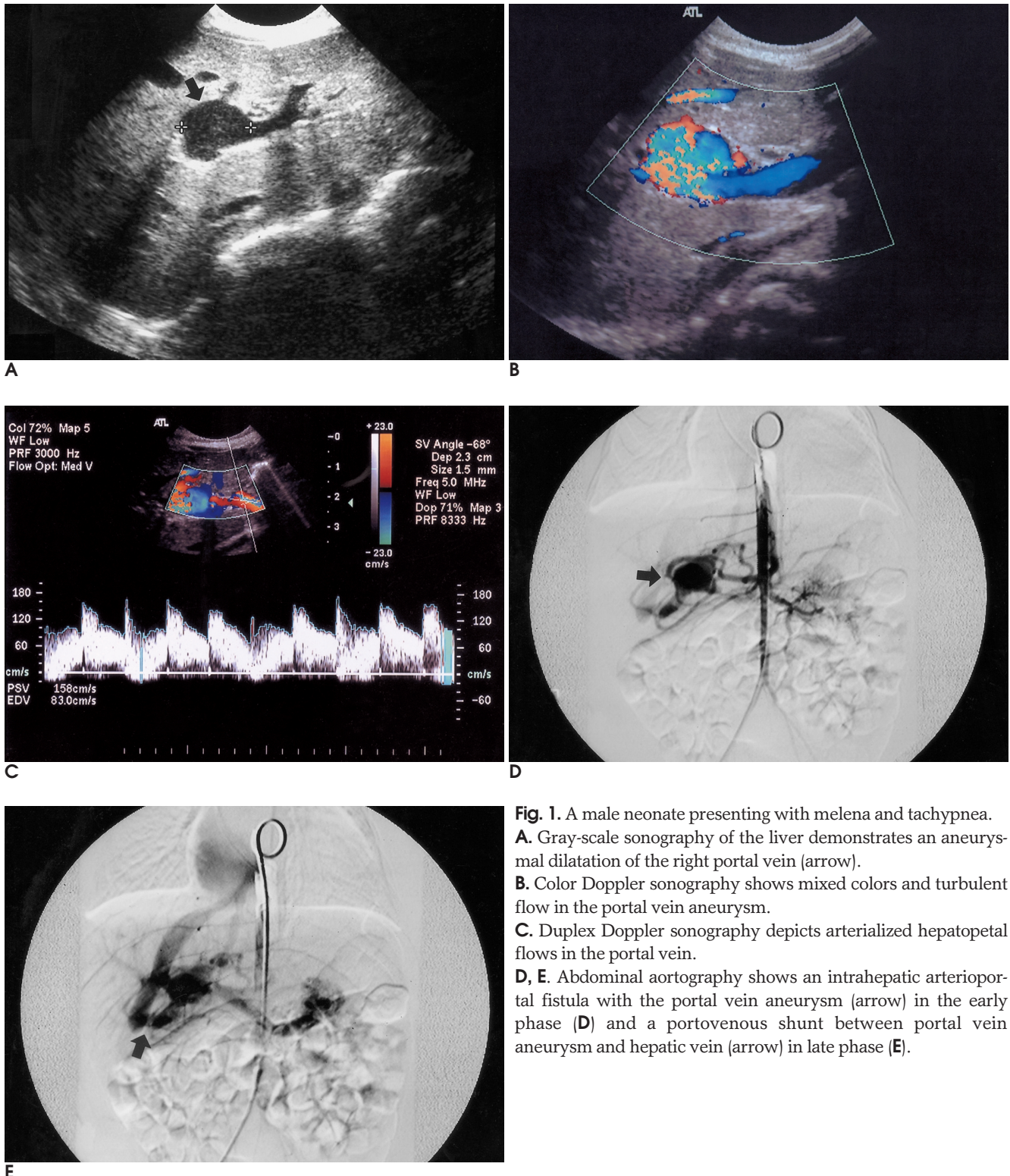


Fig. 1. A male neonate presenting with melena and tachypnea. **A.** Gray-scale sonography of the liver demonstrates an aneurysmal dilatation of the right portal vein (arrow). **B.** Color Doppler sonography shows mixed colors and turbulent flow in the portal vein aneurysm. **C.** Duplex Doppler sonography depicts arterIALIZED hepatopetal flows in the portal vein. **D, E.** Abdominal aortography shows an intrahepatic arteriportal fistula with the portal vein aneurysm (arrow) in the early phase (**D**) and a portovenous shunt between portal vein aneurysm and hepatic vein (arrow) in late phase (**E**).

portal vein aneurysm (Fig. 1D). A portovenous shunt between the portal vein aneurysm and the hepatic vein was also shown on late phase of angiography (Fig. 1E). An attempt was made to embolize the feeder right hepatic artery using microcoils but the hepatic angiogram immediately after embolization showed additional feeding vessels communicating with the portal vein aneurysm.

Discussion

A hepatoportal arteriovenous fistula may be congenital or acquired. The acquired type is more common with congenital lesions being extremely rare. A congenital hepatoportal fistula is a very rare cause of infantile portal hypertension (2). Most of pediatric patients with congenital arterioportal fistula show symptoms such as watery diarrhea, melena and features of portal hypertension, including esophageal varices, splenomegaly and ascites (1). Poor circulation and necrotic changes in the small bowel mucosa are believed to be due to a steal phenomenon from the superior mesenteric artery to the arterioportal venous fistula (5). Our patient exhibited melena, but did not show the signs of portal hypertension. None of the previously reported patients had heart failure, which might be due to the flow restrictions as a result of the hepatic sinusoids interposed between the lesion and the right heart. Our case exhibited mild tachypnea and mild cardiomegaly but did not show the signs of heart failure.

A portal vein aneurysm is also rare and the etiology of this lesion is believed to be congenital or secondary to portal hypertension, or it is associated with abnormal weakness of the vein wall (6). In our case, the portal vein aneurysm was demonstrated by gray-scale sonography, and duplex Doppler sonography identified the arteriovenous fistula to be an etiologic factor. Therefore, the portal vein aneurysm was considered to be secondary to the hemodynamic changes in the portal venous system by the hepatoportal arteriovenous fistula.

Duplex Doppler sonography is a rapid and noninvasive initial diagnostic imaging modality (7). It may demonstrate a saccular dilatation of the portal vein and show high flow velocities with turbulence, an arterial feeder from an enlarged hepatic artery branch with a high flow velocity, and pulsatile reversed hepatofugal flow in the portal vein. Gray-scale sonography demonstrates the features of portal hypertension, which include splenomegaly, venous collaterals and ascites (4).

Hepatic arteriography confirms the diagnosis, and demonstrates the vascular anatomy as well as the precise characteristics and location of the fistula (2).

Various therapeutic approaches for an arterioportal fistula have been reported. Many believe that direct surgical closure is the most suitable technique for treating extrahepatic fistulas, and embolization is a safe and effective method for treating intrahepatic fistulas (4). In our case, we failed to embolize the feeding hepatic artery on account of new additional feeding vessels communicating with the portal vein aneurysm. In our opinion, the sac of the portal vein aneurysm may be embolized using the Guglielmi detachable coils (GDCs) in the same manner used for the cerebral aneurysm embolization.

Pediatric portovenous fistulas are always associated with many other malformations (8). Park et al. (9) arbitrarily categorized portohepatic venous shunts into four different morphologic types. The first and most common type is a single large tube with a constant diameter, which connects the right portal vein to the inferior vena cava. Most patients with this type of intrahepatic portosystemic shunt show clinical evidence of liver cirrhosis and portal hypertension. The second type is a localized peripheral shunt in the branches of the portal and hepatic veins in a single hepatic segment. The third type is the aneurysmal type, where the peripheral portal and hepatic veins are connected through the aneurysm. The fourth type has diffuse multiple communications between the peripheral portal veins and the hepatic veins in both lobes. Our case was the third type because the portal aneurysm preceded the venous shunt and ruptured in the hepatic vein to make the communication, and a hepatoportal arteriovenous fistula caused the portal aneurysm.

In summary, even though this is a rare anomaly in infants, a congenital hepatoportal arteriovenous fistula may be included in a differential diagnosis when an infant presents with the features of portal hypertension. Therefore, in order to evaluate any intrahepatic abnormal vascular communications, it may be necessary to perform a careful sonographic examination, including both gray-scale and duplex Doppler sonography.

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