

Case Report

Pulsatile Hepatofugal Flow in the Portal Vein: Hallmark of a Congenital Hepatoportal Arteriovenous Fistula

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Congenital fistulae between the hepatic artery and portal vein, also known as hepatoportal arteriovenous fistula (HPAVF), are extremely rare and are usually caused by vascular malformations (1–5). The key diagnostic finding for this entity is a pulsatile (inverted) flow in the portal vein (1,3,4). We present a case with diffuse and multiple HPAVF complicated by a necrotizing enterocolitis.

CASE REPORT

After an uneventful pregnancy and perinatal period, the second child of healthy nonconsanguineous parents was admitted at the age of 6 weeks because of failure to thrive. On physical examination, he had massive ascites and splenomegaly. He had mild normochrome anemia (hemoglobin, 5.4 mmol/L; normal, >5.9 mmol/L); blood smear and platelet count were normal, as were liver function tests. Screening for metabolic disorders was negative. The serum/ascites ratio was 0.16 both for total protein and lactate dehydrogenase, classifying the ascites as a transudate. Broad coagulation studies failed to demonstrate any signs of thrombophilia.

During diagnostic workup, the boy developed clinical and radiological signs of necrotizing enterocolitis with positive blood cultures for *Escherichia coli*.

Only then a marked thickening of his bowel walls with intramural air and intrasplenic air inclusions was noted on ultrasound. Duplex Doppler investigation showed pulsatile hepatofugal Doppler signals in the portal vein and its major branches (Fig. 1). On magnetic resonance velocity phase map, simultaneous peak flow in opposite

directions was demonstrated in arterial and portal vessels (Fig. 2). Digital subtraction angiography confirmed the diagnosis of multiple HPAVFs, by simultaneous opacification of the vascular territory of the hepatic arteries and portal veins (Fig. 3).

A short time after this diagnosis, our patient successfully underwent orthotopic liver transplantation. Pathological findings were thickening of the portal vein intima compatible with an arteriovenous fistula. In addition, a small bowel segment with covered perforations and ulcerations was consistent with necrotizing enterocolitis.

DISCUSSION

Hepatoportal arteriovenous fistulae are mostly observed in older patients as a complication of liver trauma, hepatic malignancy, liver biopsy or liver cirrhosis. Congenital fistulae between the hepatic arterial and portal venous vasculature are extremely rare and represent vascular malformations. Hepatoportal arteriovenous fistulae have also been reported in Rendu-Osler-Weber disease (1,2,4).

The high systemic blood pressure causes arteriportal shunting, leading to severe portal hypertension. Vascular resistance is too high in the hepatic sinusoids to allow sufficient drainage toward the hepatic veins, and the compliance of the extrahepatic portal system is higher than the intrahepatic portion. This leads to the characteristic pulsatile hepatofugal flow pattern in the portal veins, which can easily be demonstrated, on duplex Doppler studies, as in the case presented herein. The differential diagnosis of this Doppler finding only includes advanced liver cirrhosis in which the strong arterial pulsations in the hepatic artery can induce a pulsatile hepatofugal flow pattern in the adjacent portal vein (2,4,6).

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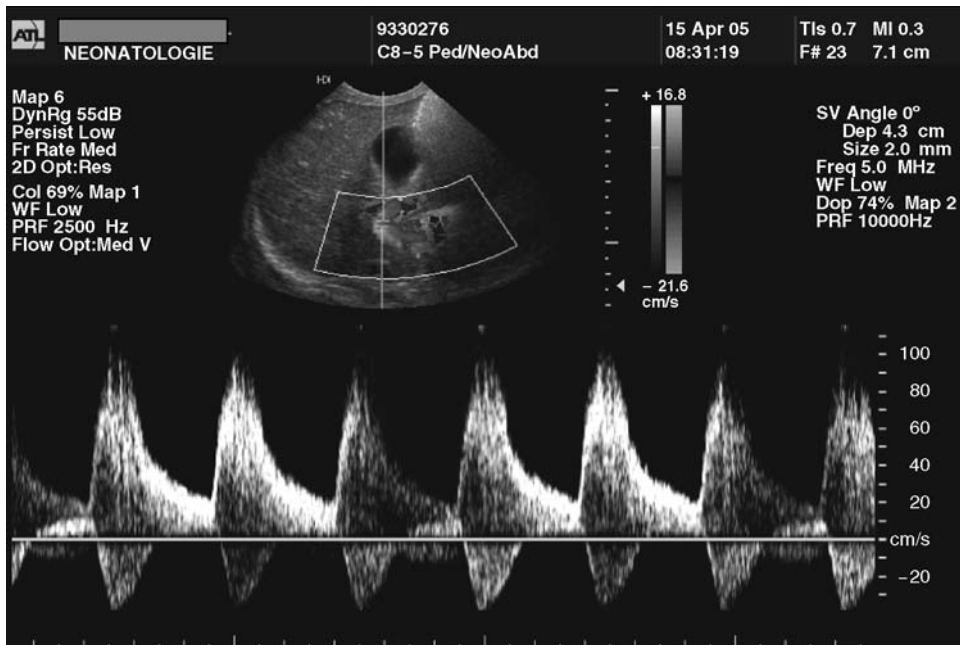


FIG. 1. Duplex-Doppler sonography showing pulsatile flow in the portal vein.

The severe portal hypertension from HPAVF causes splenomegaly and ascites (transudate) and severely impairs venous return from the small intestine. The

latter manifests clinically as edema of the bowel wall and potentially leads to necrotizing enterocolitis, as in our patient. Unlike necrotizing enterocolitis in

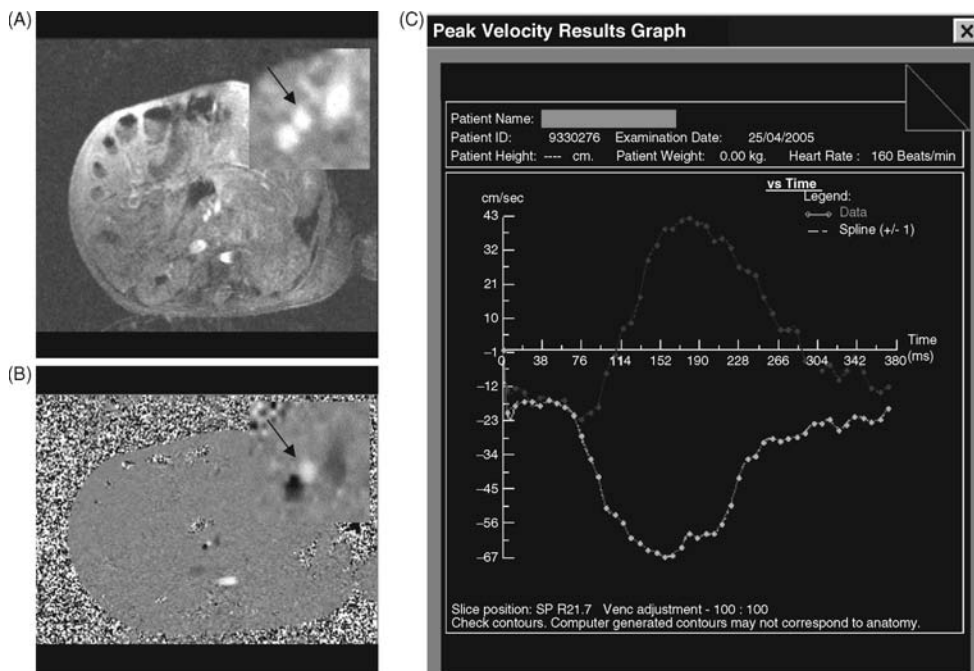


FIG. 2. A, Magnitude reconstruction of echo-planar data acquired with 1 excitation. The selected region includes both hepatic arteries and the portal vein (arrow). B and C, Velocity phase map showing simultaneous peak flow in opposite directions of portal and arterial vessels.

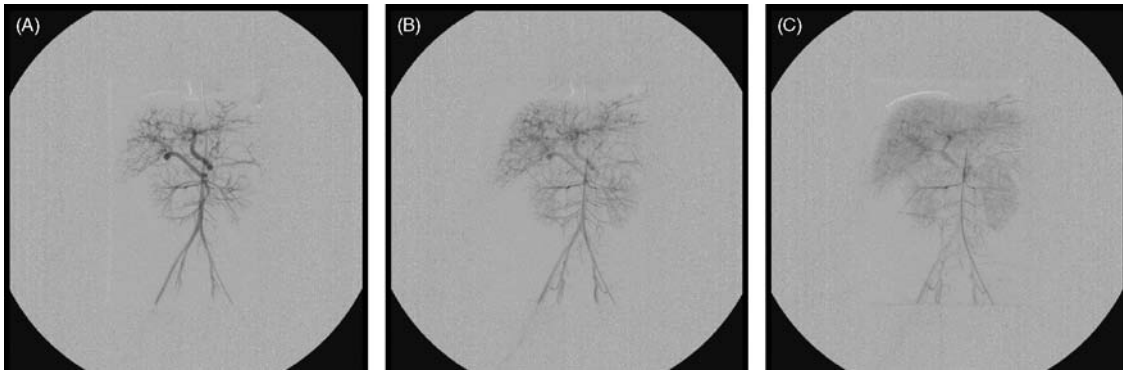


FIG. 3. A to C, Digital subtraction angiogram showing fast portal centrifugal filling in arterial phase with diffuse and multiple arterioportal shunting. Note aberrant hepatic artery from superior mesenteric artery.

premature babies, this form of enterocolitis is not expected to improve unless the underlying cause, that is, the arterioportal shunt, is relieved (2).

Angiogram shows the site and size of the fistula. Simple HPAVF can effectively be treated by percutaneous transcatheter embolization even in young children (2). However, in the present case, multiple and diffuse arterioportal shunting made endovascular embolization impossible (4). Therefore, our patient was handed in for liver transplantation.

In conclusion, demonstration of pulsatile hepatofugal flow in the portal vein without signs of parenchymal liver disease is highly suggestive of an arterioportal fistula.

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