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Evolution of congenital malformations of the umbilical-portal-hepatic venous system

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Abernethy malformation

Abstract

Objective: The objective of this study is to describe the evolution of 8 cases of congenital malformations of the umbilical-portal-hepatic venous system diagnosed before the first month of life.

Materials and methods: All cases of congenital malformation of the portal and hepatic venous system diagnosed prenatally or during the first month of life in our institution were systematically reviewed since November 2000. Clinical features, imaging, and anatomical findings were reviewed, focusing primarily on clinical and radiologic evolution.

Results: Eight cases of congenital malformation of the umbilical-portal-hepatic venous system were studied. Fifty percent of these malformations were diagnosed prenatally. We report 4 portosystemic shunts. Three involuted spontaneously, and the fourth one required surgical treatment. We report a variation of the usual anatomy of portal and hepatic veins that remained asymptomatic, an aneurysmal dilatation of a vitelline vein causing portal vein thrombosis that needed prompt surgical treatment with good result, a complex portal and hepatic venous malformation treated operatively, and a persistent right umbilical vein that remained asymptomatic.

Conclusion: Prenatal diagnosis of malformations of the umbilical-portal-hepatic venous network is uncommon. Little is known about the postnatal prognosis. Clinical, biologic, and radiologic follow-up by ultrasonography is essential to distinguish pathologic situations from normal anatomical variants.

Prenatal diagnosis of a congenital malformation of the umbilical-portal-hepatic venous system by ultrasonography is uncommon. The hepatic venous system initiates its formation by the fifth gestational week. The terminal part of the inferior vena cava (IVC), hepatic veins, and the portal veins is derived from the umbilical and vitelline veins. The portal venous system originates from a selective involution of the anastomotic network formed by umbilical and vitelline veins around the duodenum [1]. Malformations and anomalies during the development of this venous network are rare but highly varied. Efforts have been made during the recent decades to describe and classify these malformations, but little is known about their postnatal outcome. Multidisciplinary management (gynecology, neonatology, and

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pediatric surgery) is needed for the prenatal and postnatal management of these malformations. Because they may be asymptomatic, such malformations can be diagnosed in childhood or even in adulthood. When the diagnosis is made prenatally, careful counseling is essential because evolution of the umbilical-portal-hepatic venous system is yet unpredictable. We report 8 cases of malformations of the umbilical-portal-hepatic venous system, focusing primarily on clinical and radiologic evolution.

1. Materials and methods

All cases of congenital malformation of the portal and hepatic venous system followed in our institution since November 2000 were systematically reviewed.

Radiology reports that included one or more of the following terms were selected: Abernethy malformation; congenital portosystemic shunt, intrahepatic shunt; extrahepatic shunt; absent portal vein; and venous malformation.

Inclusion criteria were diagnosis of a congenital malformation of the portal, hepatic, or umbilical veins and age younger than 1 month. Instances of iatrogenic malformations, portal cavernomas, and persistent ductus venosus were excluded.

For each case, the gestational age at diagnosis, biometric data, hemodynamic data, associated anomalies, clinical features, imaging, and anatomical findings were reviewed, focusing primarily on clinical and radiologic evolution.

2. Results

Between 2000 and 2011, 4 fetuses (antenatal diagnosis between 22 and 24 weeks of gestation) and 4 newborns (1 day, 1 day, 7 days, and 1 month old) were diagnosed with a malformation of the umbilical-portal and hepatic venous system. Fifty percent of these malformations were diagnosed prenatally by ultrasonography.

There were 4 portosystemic shunts. Cases 1 and 2 (Fig. 1) were intrahepatic portosystemic shunts; case 3 and 4 (Fig. 2) were extrahepatic portosystemic shunts.

Case 5 was a malformation or the portal and hepatic venous network, associated with a complex angioma in the right hepatic lobe, diagnosed at 1 month.

Case 6 was a newborn with an aneurysmal dilatation of a vitelline vein causing portal vein thrombosis.

Case 7 was a persistent right umbilical vein diagnosed at 7 days of age.

Case 8 was a variation of the usual anatomy of portal and hepatic veins, first considered as a portosystemic shunt (Fig. 3).

All 8 cases of congenital malformations of the umbilical-portal-hepatic venous system were studied. The results are summarized in Table 1.

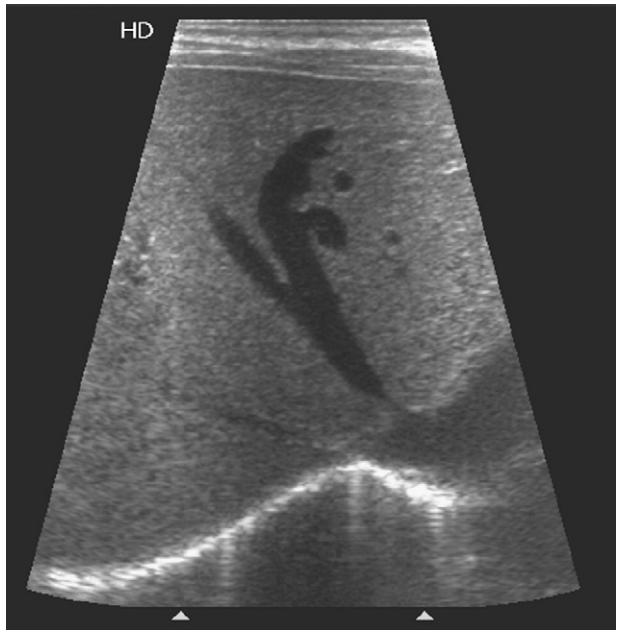

Fig. 1 Ultrasound axial scan in patient 2 at the level of the hepatic veins showing the end of the fistula arising from the left portal vein.

3. Discussion

Malformations of the umbilical-portal-hepatic venous system are rare and highly varied. We report 8 cases of such malformations with different clinical histories and outcomes. When the malformation is diagnosed prenatally and in the absence of severe associated malformation,

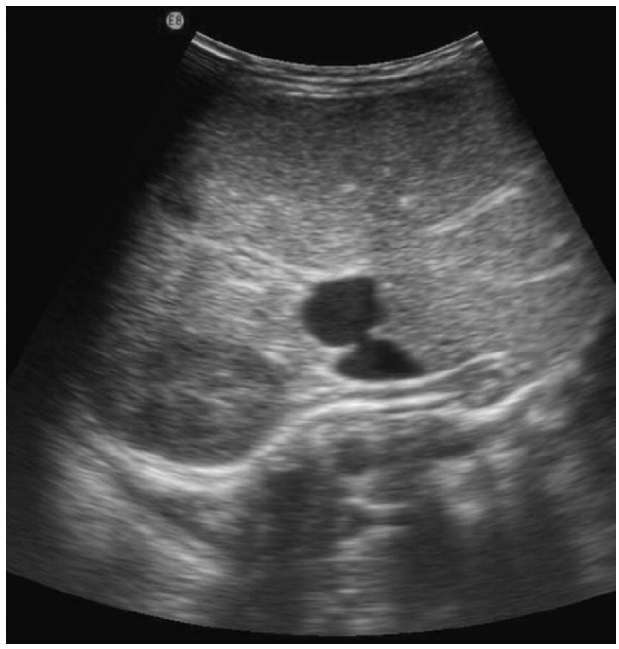

Fig. 2 Ultrasound axial scan of patient 4 at the level of the right kidney upper pole, showing the direct communication between the IVC and the dilated main portal vein.