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Abstract The CT findings of a congenital portacaval shunt are presented, and the literature on this unusual anomaly is reviewed.

Keywords Congenital portacaval shunt · CT findings · Literature review

Introduction

Although first described by Abernethy in 1793, congenital vascular shunts between the main portal vein or its main branches and the inferior vena cava are extremely rare [1, 2, 3, 4]. Two types of congenital portacaval shunts have been described. Type I shunts include congenital absence of the portal vein with redirection of portal blood directly into the inferior vena cava. Children with this specific anomaly are almost always girls, who often have multiple concurrent congenital anomalies [4]. Type II anomalies consist of a side-to-side primarily extrahepatic connection between the inferior vena cava and an otherwise normal portal vein. Type II anomalies are seen primarily in male patients and are not usually associated with other anomalies [4]. We report an extremely rare case of a type II congenital portacaval shunt in a 5-year-old girl with multiple congenital anomalies, which was diagnosed on helical CT of the abdomen, and we review the limited literature on this topic.

Case report

A 5-year-old girl with congenital cytomegalovirus was referred to our institution for a CT examination of the abdomen as part of the work-up for elevated liver enzyme levels. The girl was born with an unclear neonatal syndrome, after a full-term gestation which was remarkable for first trimester maternal exposure to cytomegalovirus. Soon after birth, the child was noted to have generalized hypotonia, which spontaneously improved, hypertrophic cardiomyopathy, a persistent ductus arteriosus that resolved over time, and hepatitis. An extensive work-up revealed cytomegalovirus in the urine. Ultrasound and CT examinations performed in the neonatal period demonstrated hepatosplenomegaly, an intrahepatic gallbladder, and abnormal hepatic vascular anatomy which could not be defined with certainty. Although a hepatobiliary nuclear scan showed no evidence of biliary obstruction, no definite gallbladder activity was noted. The child has continued to be icteric since birth, with elevated liver enzyme levels twice that of normal values. A CT scan of the abdomen was obtained at 5 years of age, during the current admission, with oral and IV contrast material. Arterial and portal venous phase 5-mm images of the liver were acquired. These revealed significant dilatation of the subhepatic and intrahepatic inferior vena cava. There was a wide side-to-side communication between the inferior vena cava and the portal vein in the region of the porta hepatis (Fig. 1). A small intrahepatic
collateral vessel also connected the middle hepatic vein and the left portal vein. Additionally, there was malrotation of the intestines associated with inversion of the mesenteric vessels. The presence of an intrahepatic gallbladder was also confirmed.

**Discussion**

A portosystemic venous shunt — i.e., a direct communication between the main portal vein or its proximal branches and the hepatic vein — is a relatively uncommon finding on cross-sectional imaging studies in adults, whereas true congenital shunts directly between the main portal venous system and the inferior vena cava are extremely rare [1, 2, 3, 4]. No known definitive developmental cause has been identified for congenital portacaval shunts [2]. The portal venous system is formed from the vitelline veins, the umbilical veins, and the ductus venosus. Embryologically, the right vitelline vein is instrumental in connecting the extrahepatic portal vein to the intrahepatic portal vein; therefore, any abnormality in development of the aforementioned structures could lead to a congenital portacaval shunt, either intrahepatic or extrahepatic [3, 4]. Persistence of the ductus venosus has been offered as a possible etiology for communication between the left portal vein and the inferior vena cava [5]. Abernethy described the first congenital extrahepatic portacaval shunt in 1793 [4]. On postmortem examination of a 10-month-old female infant, he found absence of the portal vein and complete termination of the portal blood supply into the inferior vena cava. The infant also had multiple congenital anomalies including congenital heart disease and polysplenia [4]. Recently, a classification system for congenital portacaval shunts was proposed by Howard and Davenport, who described two major types [4]. In type I shunts, there is congenital absence of the portal vein with redirection of portal blood directly into the inferior vena cava [4]. Fourteen of the 19 children with congenital portacaval shunts reported in the literature to date have been female, of whom 13 had type I shunts. Of these 13 female patients, 5 had polysplenia, 4 had biliary atresia, and 3 developed liver tumors. Type II shunts, as seen in our patient, consist of a side-to-side primarily extrahepatic connection between the inferior vena cava and an otherwise normal portal vein. Type II anomalies have been described most often in male patients, and are not usually associated with other anomalies [4]. Two of the five boys with type II shunts reported in the literature had a communication between the right portal vein

![Fig. 1a–c](image-url) A 5-year-old girl with a history of congenital cytomegalovirus infection, icterus, and abnormal liver function tests. **a** CT scan during the portal venous phase of IV contrast enhancement shows the inferior vena cava (large arrowhead), the main portal vein (small arrowhead), and a prominent left portal vein (curved arrowhead, a and b). Also note the intrahepatic gallbladder (open arrow, a and b). **b** CT scan at a slightly higher level shows a wide communication between the inferior vena cava (large arrowhead) and the main portal vein (small arrowhead). **c** CT scan just above the level of the portacaval shunt demonstrates a large inferior vena cava (arrowhead)