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Congenital absence of the portal vein in a boy

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Abstract Congenital absence of the portal vein (CAPV) is a malformation that is generally thought to be limited to females. We encountered an 11-year-old boy with this malformation. In 17 previously reported cases of CAPV, 2 were male. Three male patients, including our case, were Abernethy type Ib malformation. They had no associated liver tumour and two had no additional congenital abnormality. Conversely, 13 of the 15 female patients had congenital abnormalities and 6 had liver mass lesions.

Introduction

Congenital absence of the portal vein (CAPV) is a rare congenital abnormality that is generally considered limited to females [1, 2]. We report an 11-year-old boy with this malformation.

Case report

An 11-year-old boy was admitted to our hospital because of an abnormal shadow found incidentally on US at a local hospital. Height and weight were within normal limits. He had no cardiac or skeletal abnormality. The patient had been followed up for 4 years without evidence of encephalopathy. Laboratory data were notable for an ammonia level of 82 μg/dl (normal < 50), total bilirubin 2.3 mg/dl (normal 0.3–1.3), alkaline phosphatase 783 IU (normal 135–310), creatine phosphokinase 193 mU/ml (normal 10–100); other liver chemistry was normal. Indocyanine green excretion test showed moderate impairment (15-min retention was 26.0 %).

No portal vein was identifiable on US. MRI demonstrated that the splenic vein and superior mesenteric vein joined before emptying into the suprarenal inferior vena cava (IVC; Figs. 1, 2). Angiography confirmed this and showed no portal drainage to the liver (Fig. 3).

Discussion

Howard and Davenport defined the Abernethy malformation as congenital diversion of portal blood away from the liver by either end-to-side or side-to-side shunt [1]. This was based on the first report of the portosystemic anomaly by Abernethy in 1793 [3]. Morgan and Superina [2] proposed the following classification of portosystemic anomalies: type I – liver not perfused with portal blood because of a complete shunt (e. g. congenital absence of the portal vein); type II – liver perfused with portal blood in the presence of a partial shunt (e. g. porto-hepatic venous anastomoses). Howard and Davenport [1] and Morgan and Superina [2] reported that all patients with CAPV were female. Our case was a boy and of the 17 previously reported patients with CAPV, only 2 were male [4, 5]. The patients are summarised in Table 1 [6–17].

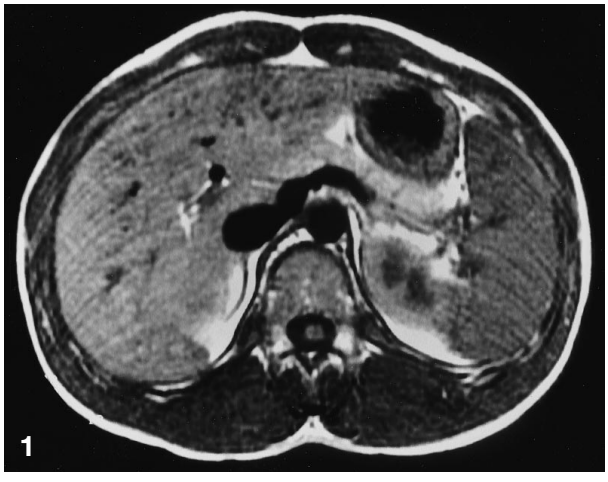
Table 1 Congenital absence of the portal vein, Abernethy malformation type I (*PDA* patent ductus arteriosus, *VSD* ventricular septal defect, *ASD* artrial septal defect, *CA* cardiac anomalies, *BA* biliary atresia, *LD* liver dysfunction, *HCC* hepatocellular carcinoma, *FNH* focal nodular hyperplasia)

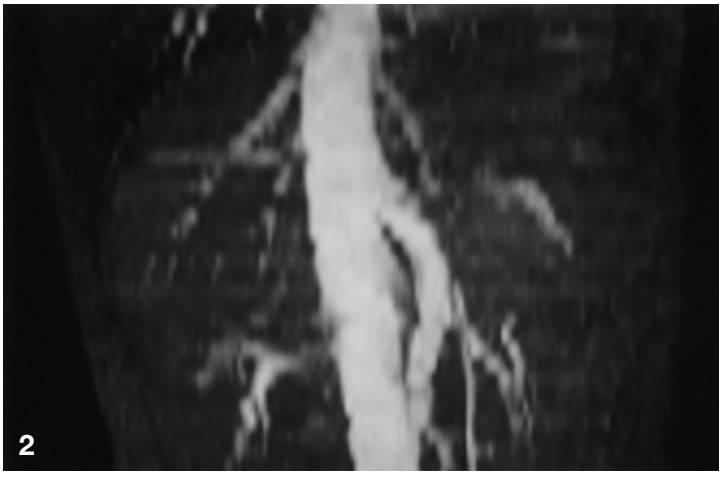
Study	Age/sex	Subtype and drainage site	Other anomalies	Liver diseases
Abernethy	10/F	Ib, Suprahepatic IVC	Dextrocardia, polysplenia	
Kienan	13/F	–, IVC		
Hellweg	0/F	Ib, Suprahepatic IVC	PDA, skin hemangioma	
Olling	50/F	Ib, Suprarenal IVC		LD
Kalifa	4/M	Ib, IVC	Goldenhar syndrome	
Marois	4/F	Ia, Lt. Renal vein	VSD	LD, hepatoblastoma
Morse, Barton	8/F	Ia, Suprarenal IVC	ASD, Goldenhar syndrome	LD, hepatoblastoma
Joyce	10/F	Ib, Suprarenal IVC	Coarctation of aorta	HCC
Okuda	17/F	Ib, Lt. Hepatic vein	Mental retardation	FNH
Nakasaki	14/F	Ia, Lt. Renal vein	ASD	LD, liver adenoma
Woodle	10/F	Ia, Lt. IVC	VSD, BA, polysplenia	LD
Matsuoka	22/F	Ib, Suprarenal IVC	Hemivertebra	LD, FNH
Morgan	0/F	Ib, IVC	CA, BA, polysplenia	LD
	1/F	Ib, Retrohepatic vein	CA, BA, polysplenia	LD
Komatsu	30/M	Ib, Iliac vein		LD
Laverdiere	0/F	–, Lt. renal vein	ASD, PDA	LD
Howard	9/F	–, Azygous vein	Situs inversus, polysplenia	LD
Present Study	11/M	Ib, Suprarenal IVC		LD

CAPV (type I Abernethy malformation) is subclassified into types Ia and Ib [2] based on the anatomy of the portal vein. When the superior mesenteric vein (SMV) and splenic vein (SV) join before emptying into the systemic vein, that vein is called the portal vein. In four cases, the SMV and SV did not join, and thus there was no anatomical portal vein (type Ia). These patients were all female. The SMV of this type drained into the IVC or left renal vein. In 11 cases, the SMV and SV joined before draining into the systemic vein (type Ib). All males had this type malformation. In 7 of 11 cases, the SMV drained into the suprarenal or suprahepatic IVC. Other draining veins were the left hepatic vein, a retrohepatic vessel and the internal iliac vein. The key to understanding this anomaly is the presence of the subcardinohepatic anastomosis. It connects the vitelline vein (which develops into the portal system) and the right subcardinal vein (which develops into the renal

Fig. 1 Axial MRI shows the splenic vein joining the superior mesenteric vein before emptying into the inferior vena cava

Fig. 2 MR angiogram demonstrates the superior mesenteric vein draining to the IVC





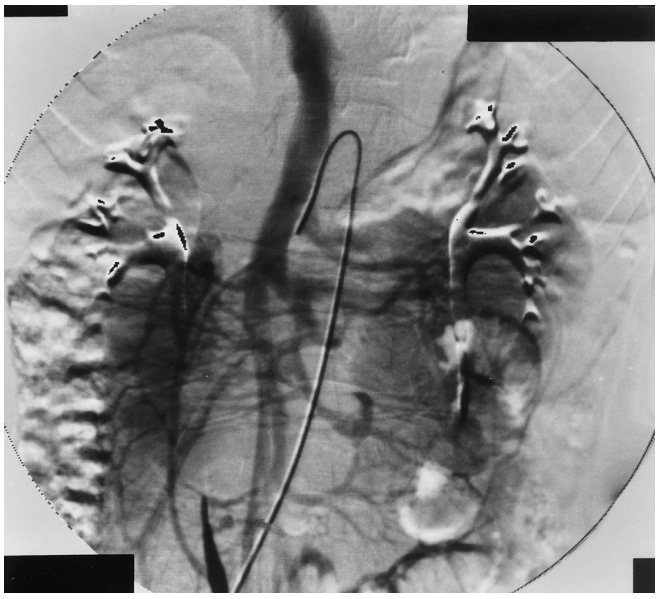


Fig. 3 Venous phase of superior mesenteric arteriography reveals no portal flow into the liver

segment of the IVC) and forms the hepatic segment of the IVC. It may account for the high incidence of draining points at the suprarenal IVC.

Additional anomalies are common in CAPV. Thirteen of the 15 female cases had congenital anomalies, the most frequent of which was a congenital cardiac anomaly. Two patients had Goldenhar syndrome. All but one male patient were free of any other associated anomaly.

In a review of the modern literature, which included our case, all but one had liver dysfunction or a mass. There were two cases of hepatoblastoma, two cases of focal nodular hyperplasia, one hepatocellular carcinoma and one adenoma, all in females. No males had a liver mass.

In conclusion, CAPV may present in the male and have a variable anatomical appearance, but is not associated with a liver mass.

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