

# Congenital extrahepatic portosystemic venous shunt: imaging features

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## ABSTRACT

**Congenital extrahepatic portosystemic venous shunt (CEPS) is a rare anomaly. It causes metabolic derangements and is often associated with liver tumours and other anomalies. Imaging plays an important role in the diagnosis of CEPS. However, it may be misleading in determining the type of shunt. We present a six-year-old girl with CEPS to illustrate the importance of histology in determining the presence of portal veins in the portal triad, which were too small to be seen on imaging.**

**Keywords: computed tomography, congenital extrahepatic portosystemic venous shunt, hepatic shunts, liver anomalies, portosystemic shunts, ultrasonography**

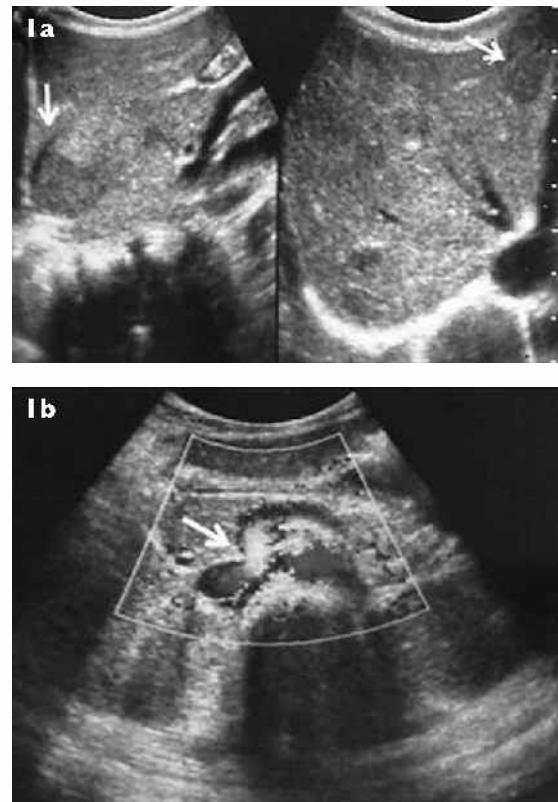
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## INTRODUCTION

Congenital extrahepatic portosystemic venous shunt (CEPS) is an extremely rare anomaly of the splanchnic venous system. Depending on the presence or absence of intrahepatic portal venous supply, CEPS has been divided in two types.<sup>(1)</sup> Recognition of the type of shunt has vital clinical implications, as the treatment options for these two types of shunts are entirely different. We present a case of CEPS in which a liver biopsy helped to determine the presence of portal venous radicals. These were too small to be visualised on imaging.

## CASE REPORT

A six-year-old girl presented to our hospital with anasarca, poor growth and loss of appetite for the last two years. Her liver enzymes were mildly elevated (aspartate aminotransferase 70 IU, alanine aminotransferase 50 IU), bilirubin level was normal and serum proteins were diminished (4.8 g/dL). The clinical diagnosis of chronic liver disease was made. On colour Doppler abdominal ultrasonography (US), the main portal vein was seen to drain entirely into the inferior vena cava. In addition, a few small well-defined



**Fig. 1** (a) US images show multiple well-defined hypoechoic nodules in the liver (arrows). (b) Colour Doppler US image shows the fistulous communication between the portal vein and the inferior vena cava (arrow).

hypoechoic lesions were seen in the liver parenchyma (Fig.1). Computed tomography (CT) (Fig. 2) and magnetic resonance (MR) imaging (Fig. 3) confirmed the presence of a large fistulous communication between the portal vein (just beyond its formation) and the inferior vena cava. Beyond the shunt, the main portal vein and its intrahepatic branches were not visualised on any of the imaging modalities; this is suggestive of a complete end-to-side extrahepatic portocaval shunt. The spleen was normal in size and there were no ascites.

There were no associated anomalies and no neurological signs of encephalopathy, although serum ammonia level was elevated at 68 mg/dL (normal range < 40 mg/dL). Technitium-99m-labelled red blood

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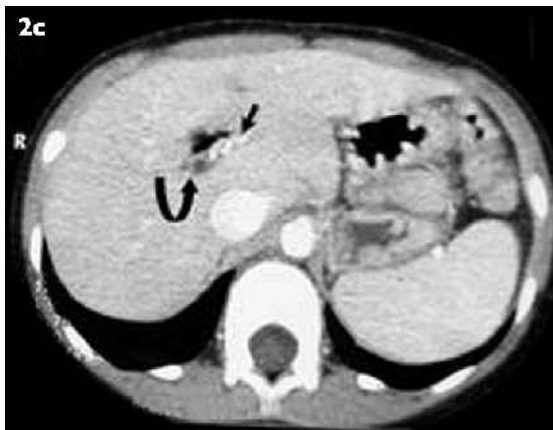
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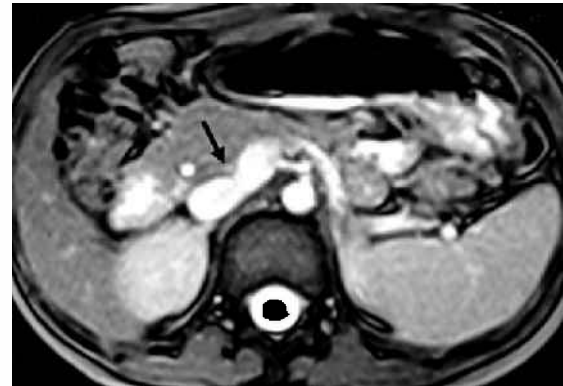
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**Fig. 2** Contiguous contrast-enhanced axial CT images taken from caudal to cranial (a–c) show a complete end-to-side portocaval fistula (a, b: arrow). At the level of the hepatic hilum (c), only the hepatic artery (short arrow) and the bile duct (curved arrow) are seen while the portal vein and its branches are not visualised.

cell scintigraphy (Fig. 4) showed markedly-reduced hepatic perfusion in initial dynamic images. Static images after 15, 30, 45 minutes also showed decreased tracer uptake. Based on the imaging findings, a diagnosis of complete end-to-side portocaval fistula was made. However, on liver biopsy, the portal venules were seen



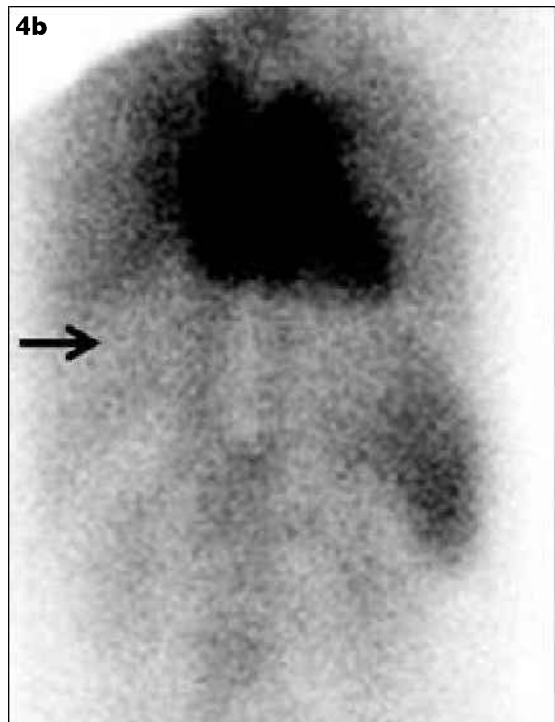
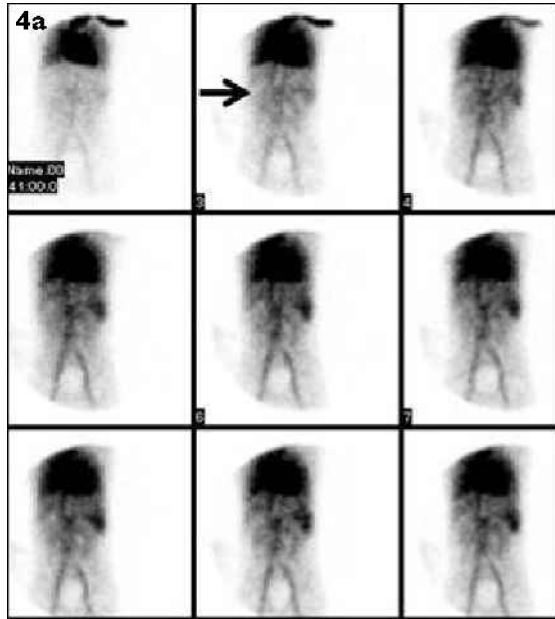
**Fig. 3** Axial balanced-steady-state gradient-echo MR image shows fistulous communication (arrow) between the portal vein and the inferior vena cava.

in the portal tracts (Fig. 5), in addition to arterioles and bile ducts. The final diagnosis of type II CEPS was made. The child was managed conservatively and is currently on follow-up.

#### DISCUSSION

Since the first description of CEPS in 1793 by Abernethy,<sup>(2)</sup> there have been only about 61 cases reported in the English literature.<sup>(3)</sup> These shunts allow the portomesenteric venous blood to drain directly into the systemic circulation, most notably into the inferior vena cava.<sup>(3)</sup> The portal venous system develops from the vitelline veins in a complex process which involves selective involution of these venous loops. Excessive involution can result in the absence of the portal vein. Inferior vena cava develops close to the portal vein from the subcardinal vein as a result. The vitelline and subcardinal veins have anastomotic channels between them in the early stages of embryonic development, which permits the formation of portocaval shunts in rare cases.<sup>(4,5)</sup>

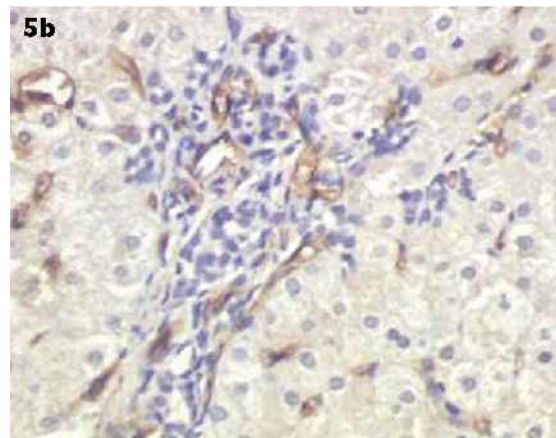
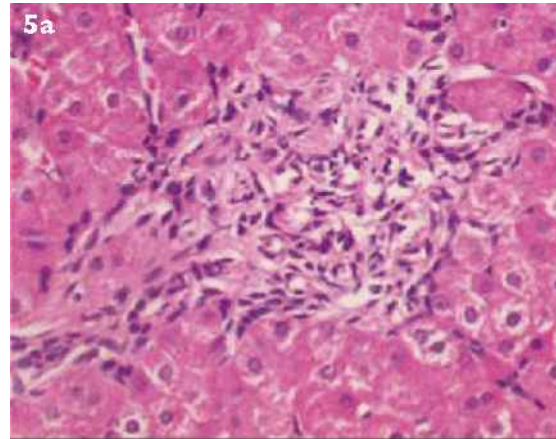
Morgan and Superina classified CEPS into two types.<sup>(1)</sup> Type I CEPS is also called congenital absence of the portal vein, and is an end-to-side shunt between the portal vein and the systemic circulation, such that all the splanchnic venous return enters the systemic circulation and the liver is not perfused with portal venous blood at all. Type II CEPS is a partial side-to-side shunt between the portal and systemic vein. In this type, only a fraction of the splanchnic venous return bypasses the liver parenchyma. The clinical manifestations of CEPS are diverse, and are the results of metabolic derangements and various associated anomalies. In CEPS, serum levels of ammonia, galactose and other toxic metabolites are elevated due to systemic diversion of the portal venous blood. Newborn screening for high galactose levels is a



**Fig. 4** Tc-99m RBC blood pool scan shows decreased perfusion of the liver (arrows) in the (a) initial dynamic, and (b) static images.

useful indicator for the presence of CEPS.<sup>(3)</sup> Metabolic derangements can result in hepatic encephalopathy, which is more often seen in older patients, possibly because the ageing brain is more susceptible to these harmful metabolic products. Patients with congenital absence of the portal vein typically do not have features of portal hypertension.<sup>(3)</sup>

Nodular liver lesions are very commonly seen in cases with CEPS. Most of these are benign



**Fig. 5** (a) Photomicrograph of liver biopsy shows one of the portal tracts with prominent portal venous radicals (Haematoxylin and eosin,  $\times 200$ ). (b) Immunohistochemical stain for endothelium (marker for Factor VIII) highlights the portal venous radicals ( $\times 200$ ).

lesions (such as focal nodular hyperplasia, nodular regenerative hyperplasia and hepatocellular adenoma).<sup>(6)</sup> Regenerative liver lesions have been attributed to hepatic ischaemia and a compensatory increase in the hepatic arterial flow.<sup>(7)</sup> A few small hypoechoic liver nodules were also seen in our case (Fig. 1). Since the lesions were multiple, appeared benign on imaging, and because regenerative nodules are well described in CEPS, we refrained from obtaining tissue diagnosis from these nodules. Cardiac anomalies are particularly common in CEPS, and are seen in nearly a third of the reported cases. The other known associations of CEPS include biliary atresia, skeletal and renal tract anomalies.

The clinical profile and management of the two types of CEPS are different. Patients with type I shunts have a striking female predominance, and are typically young at the time of presentation, as they often have associated anomalies. Shunt ligation is not feasible in a type I shunt since it represents the only exit channel for portomesenteric venous return. Liver transplantation,

though technically challenging, is however possible, and represents the only definitive treatment in these cases. Type II shunt does not present female predominance. Since the incidence of associated anomalies is much less in this group, the patients typically present late in middle age, with symptoms of hepatic encephalopathy. Patients with type II shunts can be treated using ligation or coil embolisation of the shunt vessel.<sup>(3)</sup>

A type I shunt is characterised by the absence of portal venules within the portal triads upon liver biopsy. It is notable that biopsy confirmation of the absence of intrahepatic portal venules has been made in only eight out of the 39 reported cases of type I shunts.<sup>(3)</sup> In our case, as the intrahepatic portal venous branches were not visualised on imaging, this was suggestive of type I CEPS. However, portal venous channels were evident on liver biopsy, indicating type II CEPS. Only two such cases have been reported in the literature where intrahepatic portal venous radicals were seen only on biopsy.<sup>(3,8)</sup> Shunt ligation was not performed in either of these cases. The vast majority of CEPS cases reported in the literature do not have biopsy confirmation of the type of shunt. Since, in the presence of a large shunt, the distal portal venous tree gets attenuated, it may be missed on imaging. It is therefore reasonable to infer that some type II shunts may have been misdiagnosed as type I shunts, if they were based on imaging findings alone.

Ikeda et al reported the results of shunt ligation in four patients with type II shunts who had attenuated portal venous radicals. These were visualised on preoperative MR imaging, US and angiography.<sup>(9)</sup> Even the apparently-attenuated portal system was capable of accommodating the portal venous inflow following shunt closure without development of portal

hypertension. They concluded that poor development of the portal venous tree does not preclude curative shunt ligation. Whether the very small portal venous radicals which are detected only on liver biopsy, as in our case, will also be able to tolerate ligation of the shunt is as yet unknown.

In conclusion, although CEPS is a rare anomaly, it must be recognised early to prevent the consequences of metabolic derangements by applying the appropriate surgical treatments. Imaging plays a vital role in the diagnosis of CEPS. The limitation of imaging in determining the type of shunt in some cases should be recognised, and a histopathological confirmation of the type of shunt may be crucial in deciding the treatment course. In many cases, the detection of tiny intrahepatic portal venous radicals may be beyond the resolution limits of the available imaging methods.

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