

Abernethy malformation type I (congenital absence of the portal vein) in a patient with chronic kidney disease

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A 31-year-old male patient with chronic membranoproliferative glomerulonephritis (confirmed by biopsy) treated with immunosuppression (steroids, cyclosporine, mycophenolate mofetil) was admitted to the hospital due to nephrotic syndrome with chronic kidney disease stage 3 and coma (rigid pupils with bilateral Babinski sign). Blood tests revealed elevated concentrations of ammonia (288 $\mu\text{mol/l}$), serum creatinine (258 $\mu\text{mol/l}$), urea (31.2 mmol/l), serum albumin (27 g/l), alanine aminotransferase (37 U/l), aspartate aminotransferase (30 U/l), bilirubin (24 $\mu\text{mol/l}$), γ -glutamyltranspeptidase (138 U/l), cholinesterase (3678 U/l), ceruloplasmin (0.26 g/l), and alkaline phosphatase (205 U/l). Serological markers for hepatitis A, B, C, G viruses,

as well as immunological markers were negative. On the basis of electroencephalography and head computed tomography (CT), the patient was diagnosed with nonconvulsive epilepsy (treated with valproic acid). An abdominal CT scan revealed polysplenia (FIGURE 1A) and the absence of the portal vein (FIGURE 1B). It also showed that the splenic and intestinal venous blood flowed via the splenic and mesenteric veins to the inferior vena cava and to the left renal vein without passing through the liver (FIGURE 1C). On portovenography with the occlusion test (FIGURE 1D), the liver was not perfused with portal blood and the superior mesenteric vein and splenic vein did not join to form confluence (abernethy malformation type IB).

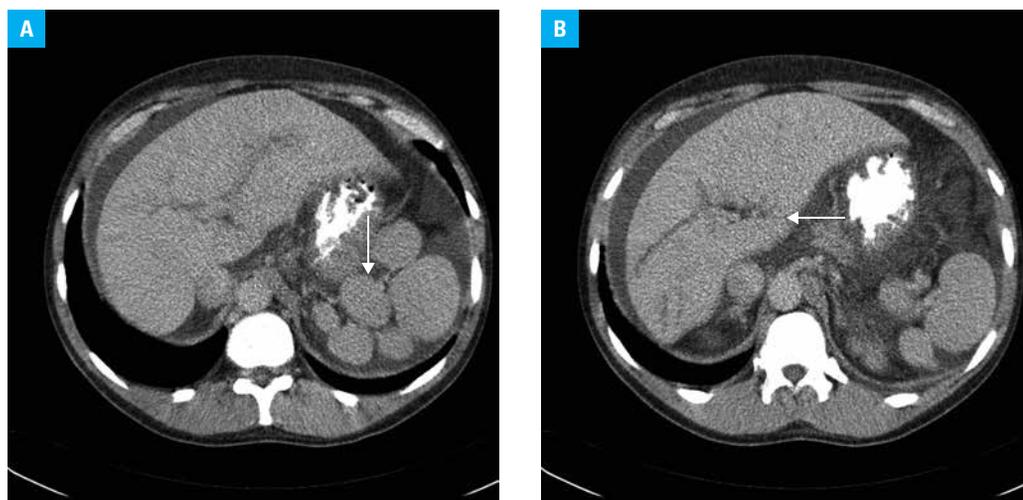


FIGURE 1 A – computed tomography scan: multiple spleens (white arrow); B – computed tomography scan: absence of the portal vein (white arrow); C – computed tomography scan: splenic vein (white arrow) and inferior mesenteric vein (short black arrow) connect with the left renal vein (long blue arrow) to form a common confluence (short blue arrow) entering the inferior vena cava (long black arrow); D – image showing no contrast enhancement of the portal vein

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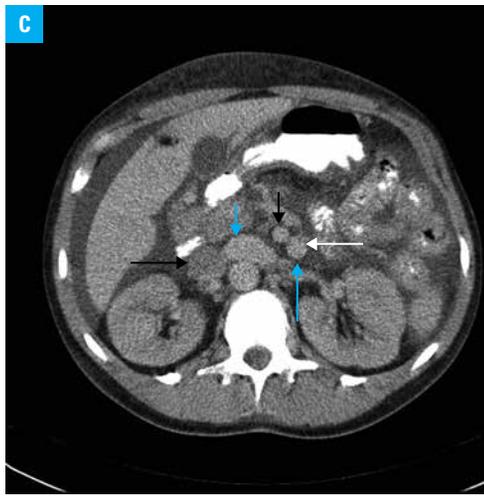
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The congenital absence of the portal vein is a very rare portal venous blood system malformation that has been reported in 101 patients since 1793 (first described by John Abernethy). In this malformation, splanchnic venous blood partially or completely bypasses the liver and drains directly into the inferior vena cava or to the left renal vein as a result of the congenital absence of the portal vein or the hypoplastic portal vein.¹

The CAPV is frequently associated with other congenital cardiac, gastrointestinal, genitourinary, or skeletal defects. The lack of portal flow can result in the development of nodular regenerative hyperplasia or hepatocellular adenoma, rarely hepatocellular carcinoma or hepatoblastoma.² Portosystemic encephalopathy is rare in patients with congenital absence of the portal vein, and it is more common in older patients because of increased brain sensitivity to ammonia or disorders of homeostatic control mechanisms that develop with aging. A serious although rare complication may be hepatopulmonary syndrome due to humoral factors (in normal conditions metabolized by the portal passage through the liver) shunted into the systemic and pulmonary venous circulation.^{3,4}

Six months later, the patient was started on hemodialysis due to exacerbation of chronic kidney disease (serum creatinine, 720 $\mu\text{mol/l}$; urea, 33 mmol/l). A dialysis regimen with 4 sessions per week was introduced along with protein restriction (0.6 g/kg/d), with administration of intravenous aminoacids and albumin supplementation twice a month. The patient's clinical condition significantly improved. The recurrence of portosystemic encephalopathy was not observed and ammonia levels decreased to a range of 60 to 90 $\mu\text{mol/l}$. The patient was placed on the waiting list for kidney transplantation, which will be the best treatment option.⁵

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