Neonatal Cholestasis as Initial Presentation of Portosystemic Shunt: A Case Report

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Congenital intrahepatic portosystemic shunts are rare in children. Portosystemic venous malformations are characterized by extreme clinical variability. We report a full-term 33-day-old male infant presenting with neonatal jaundice. On physical examination, he had generalized icterus and the liver was palpable 3.5 cm below the right costal margin. He had no other symptoms. Laboratory tests showed AST 632 U/L, ALT 198 U/L, total bilirubin 12.1 mg/dL, conjugated bilirubin 10.2 mg/dL, alkaline phosphatase 753 U/L, GGT 47 U/L and glucose 67 U/L. Colour Doppler ultrasonography showed the left portal vein was more dilated than the right portal branch and communication with dilated left hepatic vein. There was no evidence of portal hypertension, heart failure, hepatopulmonary syndrome and encephalopathy during his hospital stay, so he was discharged from the pediatric department and his parents advised to attend monthly follow-up. Congenital portosystemic shunts are rarely observed in the childhood period.[1] Depending on anatomic characteristics they may be intrahepatic or extrahepatic.[2] Intrahepatic portosystemic shunts (PSS) are observed between the portal vein and hepatic vein or vena cava inferior.[3,4] Small shunts may close themselves before the age of 2 years.[5] With the increase in use of imaging methods, diagnosing PSS has become easier, with an increase in the number of cases reported.[6] Neonatal cholestatis is a frequent complication of PSS.[1] We present a case presenting with neonatal cholestasis diagnosed with congenital intrahepatic PSS. (J Clin Exp Hepatol 2016;6:331–334)

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CASE

A 33-day old male applied to the pediatric gastroenterology clinic due to complaint of jaundice. It was learned that jaundice began at 4 days old, with occasional white (acholic) stools. The case was born at term by cesarean section weighing 3500 g and no previous known disease history was found for him or his family. Physical examination found weight 4600 g (50–75 percentile), length 53 cm (25–50 percentile), skin and sclera icterus, the liver was palpable 3.5 cm below the right costal margin, and spleen could not be palpated. Neuromotor development was normal. Laboratory studies showed a serum aspartate transaminase 632 U/L, alanine transaminase 198 U/L, total bilirubin 12.1 mg/dL, conjugated bilirubin 10.2 mg/dL, alkaline phosphatase 753 U/L, LDH: 515 U/L, glucose 67 mg mg/dL, PT: 14.2 s, INR: 1.26, stool steatocrit +3, TSH: 3.8 uU/mL and FT4: 1.3 ng/ml. Stool samples had acholic appearance. Considering metabolic disease, there was no hypoglycemia or septic appearance. Eye examination found no cataracts. Examination for infectious etiologies was negative (toxoplasma, cytomegalovirus, Epstein–Barr virus, hepatitis A, B, C). Alpha 1-antitrypsin (PiMM) and cystic fibrosis mutations were not detected. Eye examination was normal in terms of metabolic diseases. Abdominal sonography showed hepatomegaly (80 mm) with normal contours and echo pattern of the liver. The gall bladder and intrahepatic biliary system were normal. The main and right portal veins were normal on Doppler ultrasound examination (DUSI). The left portal vein was more dilated than the right portal branch. It was linked to the dilated left hepatic vein through a 0.8 mm aberrant venous connection (Figure 1). The left portal vein showed a continuous waveform spectrum on DUSI, whereas the left hepatic vein showed a mild

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Abbreviations: AST: aspartate transaminase; ALT: alanine transaminase; DSUG: Doppler ultrasound; GGT: gamma glutamyl transferase; LDH: lactate dehydrogenase; PSS: portosystemic shunts
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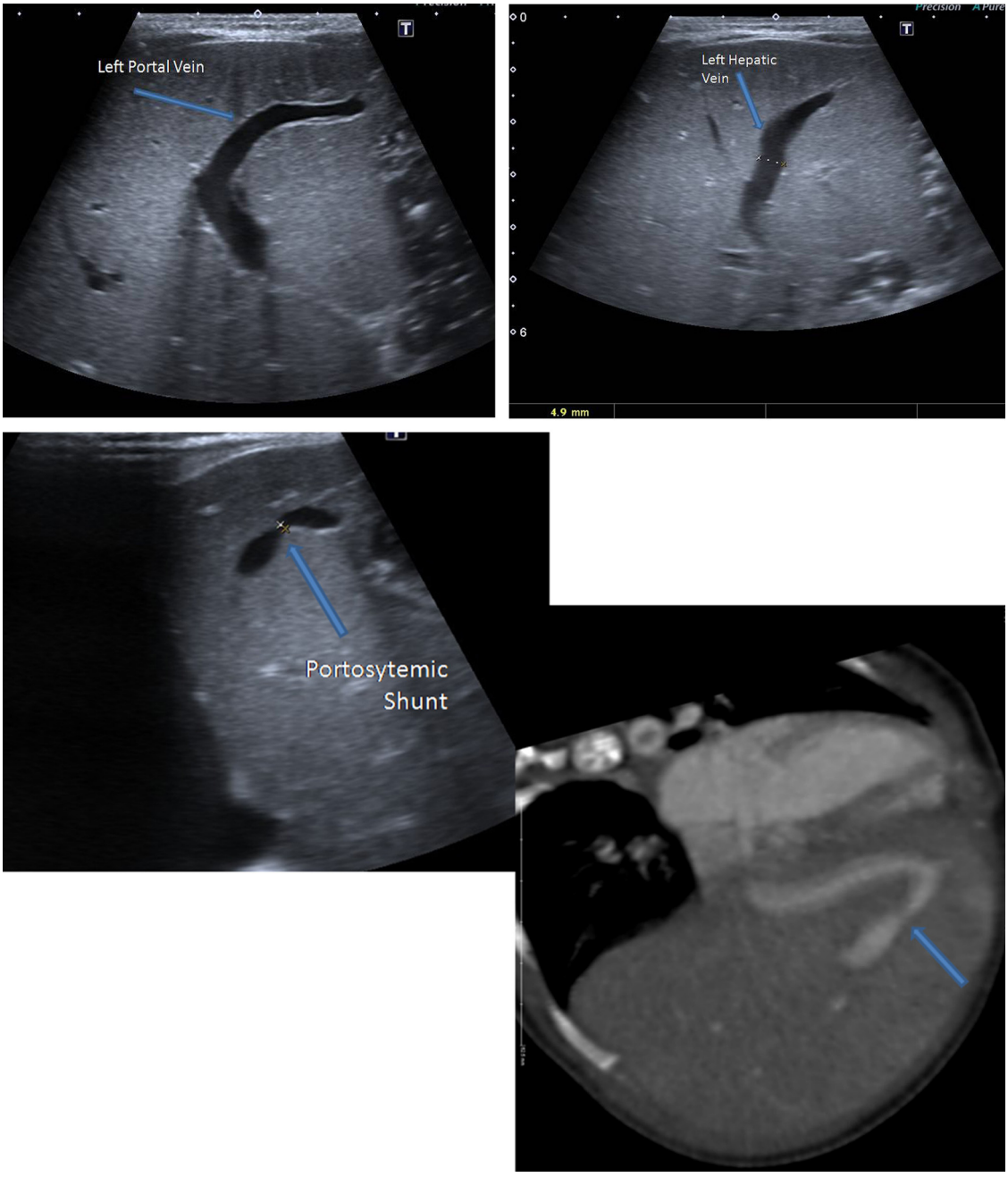

Figure 1 Ultrasonography and reformatted contrasted CT imaging show connection between dilated vena porta and left hepatic vein (white arrow).

increased turbulent flow. Other hepatic veins had normal phasic flow pattern and diameter. As cardiac pathologies may occur in shunt patients, echocardiography was performed for screening purposes. It was interpreted as normal. Echocardiographic evaluation was normal. Neurological examination found no evidence of encephalopathy. (Evaluating encephalopathy in this patient was not as easy as with older children. However the patient was active, mobile, with lively appearance, had no difficulty feeding, and had normal sleep habits for his age, so encephalopathy was not primarily considered.) Due to cholestasis, the patient was begun on ursodeoxycholic acid treatment. During hospital stay, no complications such as portal hypertension, heart failure and hepatopulmonary syndrome developed. The family was informed of complications that might develop. The patient was discharged after an 8-day hospital stay, with good activity, nutrition and weight gain and called for regular follow-up. The patient was monitored for 5 months. On follow-up the patient's hemogram, biochemistry and coagulation values

