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Case Report

Prenatal Diagnosis of Intrahepatic Portosystemic Shunt

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Introduction

Extrahepatic or intrahepatic congenital portosystemic shunt (Abernerthy malformation) is a very rare vascular anomaly (1:30,000 births). It is characterized by the passage of splanchnic venous blood into the systemic circulation, which gives rise to serious complications such as encephalopathy due to hyperammonemia, which can produce anything from coma episodes to severe intellectual retardation if it is persistent over time; development of liver tumors with a risk of malignant degeneration; hepato-pulmonary syndrome; development of pulmonary hypertension etc. [1,2].

Keywords: Neonatology; extrahepatic or intrahepatic congenital portosystemic shunt

Case Report

We present a male neonate, son of a 41-year-old primigravida mother. Prenatal ultrasound from week 26 revealed a hepatic vascular anomaly, including an anomalous ductus with the presence of an aberrant suprahepatic vein that empties into the right atrium and a single umbilical artery (Figure 1a). GH-array and amniocentesis were performed without alterations and delivery was induced by RIC type I at 37 weeks. From the first hours of life, the newborn presents hypoglycemia and jaundice, with phototherapy range at 17 hours of life. In analytical control highlights indirect bilirubin 14.75mg/dl, direct 2.14mg/dl and normal ammonia. The urine is coluric without the presence of urobilinogen or red blood cells. Neurological examination is normal with adequate tone, primitive reflexes present and good suction-swallowing coordination, normal electroencephalogram, and brain monitoring. Maternal serologies for hepatotropic viruses (Erythrovirus, CMV, Parvovirus, HSV and EBV) and toxic negative. Alpha 1 antitrypsin was normal. Normal metabolic endocrine screening. An abdominal and cardiac ultrasound was performed at 48 hours of life (Figure 1b) where the persistence of the intrahepatic shunt between the portal and right suprahepatic veins is visualized, with a slight decrease in flow to the right hepatic lobe. Patent ductus venosus and patent foramen ovule. In controls after 14 days of life, a decrease in intra-shunt flow caliber was observed with complete disappearance of the Shunt at one and a half months of life (Figure 1c).

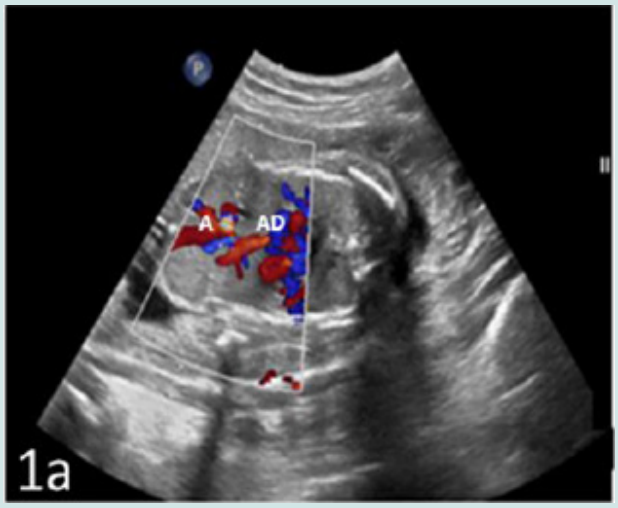

Figure 1a: Prenatal ultrasound (CGE 28+1): presence of hepatic vascular malformation (A) with abnormal drainage to the right atrium (RA).

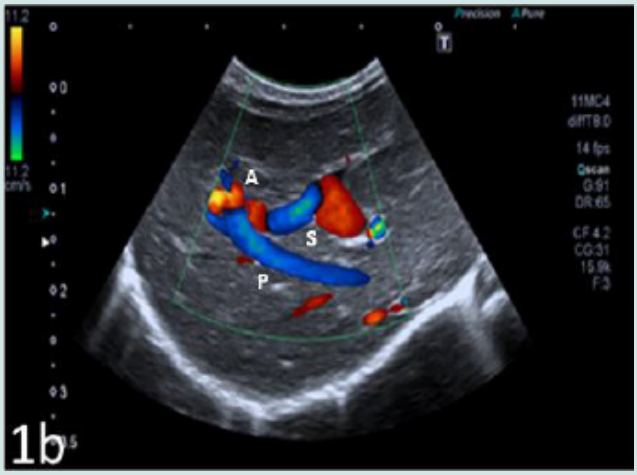

Figure 1b: Postnatal ultrasound (14 days): intrahepatic shunt between the portal vein (P) and the right suprahepatic vein (S), with a slight decrease in flow to the right hepatic lobe.

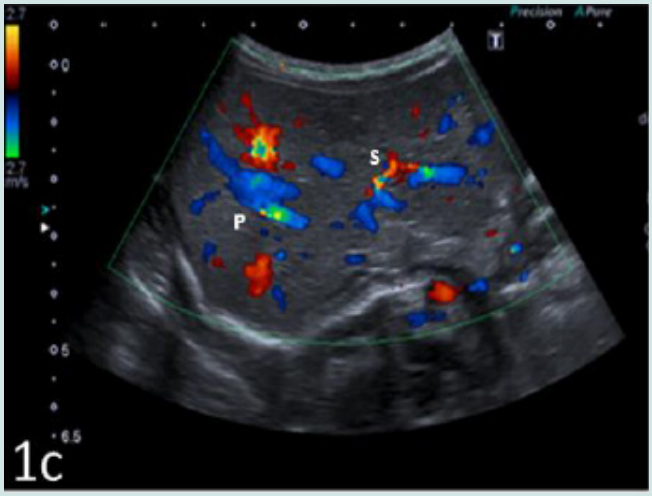

Figure 1c: Postnatal ultrasound (45 days): disappearance of the shunt between the right portal vein and the right suprahepatic vein.

Conclusions

Prenatal detection is very important in the diagnosis of congenital vascular anomalies. In our patient, intrauterine growth retardation [3] associated with the presence of aberrant portal circulation, anomalous ductus venosus, patent foramen ovule and single umbilical artery; together with the existing metabolic alterations (hypoglycemia due to hyperinsulinism), the presence of choluria and cholestasis with jaundice, makes us think of the presence of SPSC [4]. Some small caliber intrahepatic shunts can close spontaneously during the first two years of life, although this usually does not occur as early as in our patient [5]. Several authors suggest the need for early treatment, before the presence of complications by closing the shunt by interventional radiology or open surgery, if it cannot be carried out. Before closing the shunt, the tolerance of the intestine to clamping should be checked to avoid acute, fatal portal hypertension. Shunt occlusion allows the development of intrahepatic portal circulation even in patients with severe hypoplasia or portal agenesis, being able to avoid and/or reverse the complications derived from it.

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