# Rare cause of neonatal pulmonary hypertension: Congenital intrahepatic portosystemic shunt through an aneurysm

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## ABSTRACT

**Congenital portosystemic shunt is a rare congenital anomaly with abnormal communication between portal venous and systemic venous systems. It can be intrahepatic or extrahepatic. Typically, the intrahepatic shunts are managed conservatively as many of them close spontaneously. We present and discuss clinical, radiological findings of an intrahepatic shunt showing the early occurrence of pulmonary arterial hypertension in the neonatal period which required therapeutic intervention.**

**Keywords:** Aneurysm, congenital intrahepatic portosystemic shunt, neonate, pulmonary hypertension

## INTRODUCTION

Congenital intrahepatic portosystemic shunt is a rare anomaly. In humans, the incidence of the portosystemic shunt is estimated at 1/30,000 births. The intrahepatic portosystemic shunt in association with an aneurysm is not reported in neonates as most case reports mention such shunt in the adult age group.[1,2] Furthermore, the early occurrence of pulmonary hypertension in neonatal age is very unusual.

## CASE REPORT

A late-preterm male newborn (35 weeks) delivered by cesarean section was referred to our center for further management as the baby developed respiratory distress. The initial chest radiograph showed cardiomegaly with bilateral diffuse lung opacities. In view of increasing oxygen requirement, the baby was intubated and the surfactant was administered. Echocardiography on day 2 of the life showed dilated right and left ventricles with moderate tricuspid regurgitation and severe pulmonary arterial hypertension (PAH) (mean pulmonary artery pressure ~58 mm Hg) for which baby was started on intravenous sildenafil infusion. The baby maintained oxygen saturation; however, there was persistent tachypnea. Initial liver function tests on day 2 of life showed elevated bilirubin levels with normal liver enzymes. Bilirubin levels showed progressively decreasing values with time. Blood ammonia levels measured on day 8 of life were elevated (~163.6 μmol/lit, Laboratory reference value was 1–35 μmol/lit).

Ultrasound of the abdomen on day 2 of life showed a Park *et al.* type 3[3] congenital intrahepatic portosystemic shunt through an aneurysm with associated mild ascites and diffuse subcutaneous edema [Figures 1-5]. Initially, the shunt was managed conservatively using intravenous sildenafil and optimizing ventilator settings based on serial radiographs. However, tachypnea and severe PAH failed to respond to conservative management. Echocardiography done on day 7 of life showed persistent severe PAH (mean pulmonary artery pressure ~58 mm Hg). Persistent severe PAH

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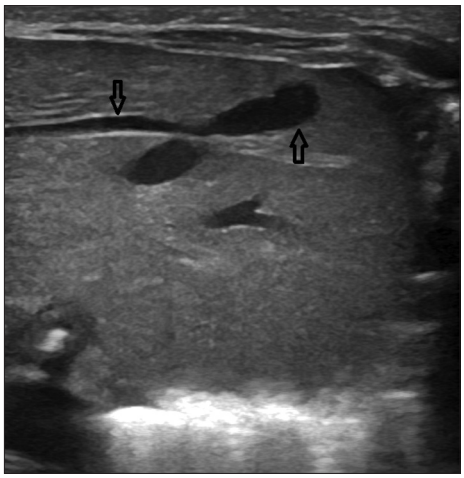


**Figure 1: Grayscale ultrasound image of the left lobe of the liver showing the left branch of the portal vein (down arrow) communicating with anechoic aneurysm (up arrow)**

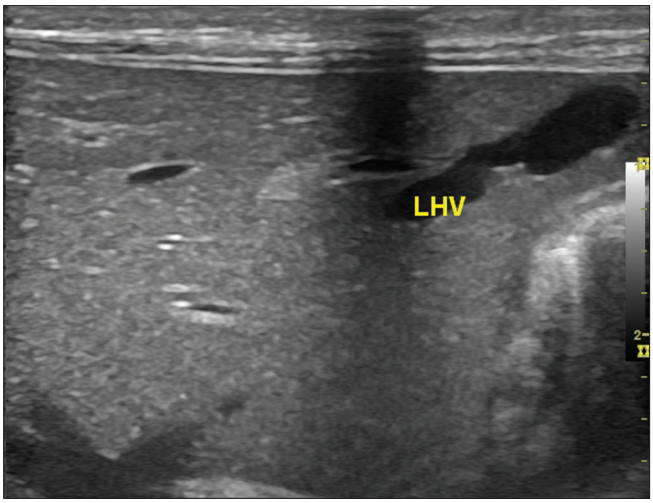



**Figure 2: Grayscale ultrasound image of the left lobe of the liver showing left hepatic vein communicating with an aneurysmal shunt**

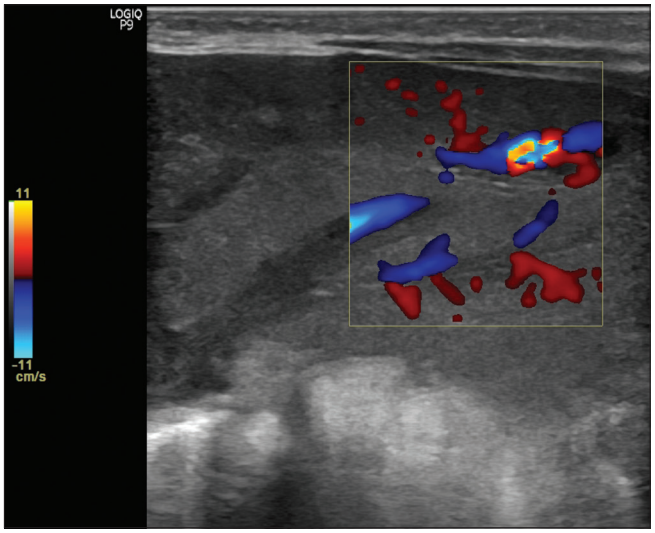



**Figure 3: Color Doppler image showing flow with color aliasing in the shunt**

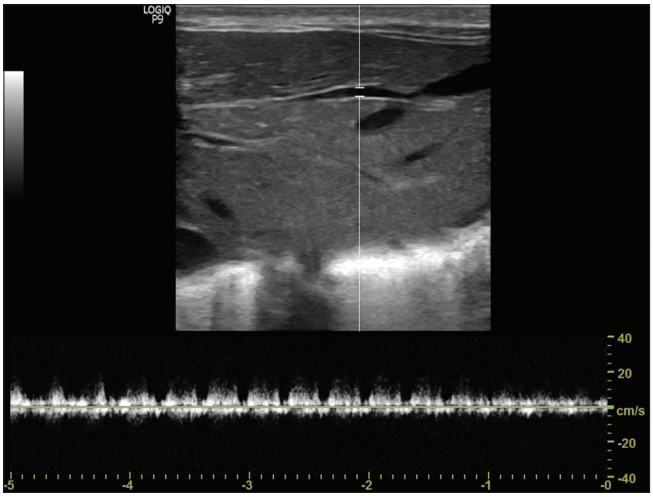



**Figure 4: Spectral Doppler image showing pulsatile flow in the left branch of the portal vein, probably due to transmitted pulsations from the left hepatic vein through the shunt**

was attributed to intrahepatic shunt and percutaneous shunt closure was planned. Contrast-enhanced computed tomography of the abdomen was acquired before shunt closure which confirmed a tubular vascular channel connecting the left branch of the portal vein and left hepatic vein through an aneurysm [Figure 6].

Percutaneous shunt closure was performed on day 14 of the life. In the catheterization laboratory, after securing the right internal jugular vein access, left hepatic vein injection reconfirmed the diagnosis of type 3 intrahepatic portosystemic shunt connected through aneurysm in accordance with the sonogram and CT scan [Figure 7]. Assessment of pulmonary hemodynamics revealed pulmonary hypertension before the shunt closure. Aneurysmal sac measured ~6 mm × 6 mm and shunt measured ~4 mm in width. The shunt was successfully closed by deploying V-Trak microvention coils (Terumo Corporation, Japan) into the aneurysmal sac. Closure of the shunt was confirmed in postembolisation hepatic venous contrast run. The child showed clinical improvement after the procedure and was discharged subsequently in a stable condition. Liver function tests including blood ammonia levels measured after the procedure showed normal values. Ultrasound of the abdomen showed no residual flow in the portosystemic shunt. During the 6 months postprocedure follow-up period, the child remained asymptomatic with normal liver function and complete resolution of pulmonary hypertension.

## DISCUSSION

During prenatal life, congenital intrahepatic portosystemic shunt on ultrasound can cause fetal growth restriction in

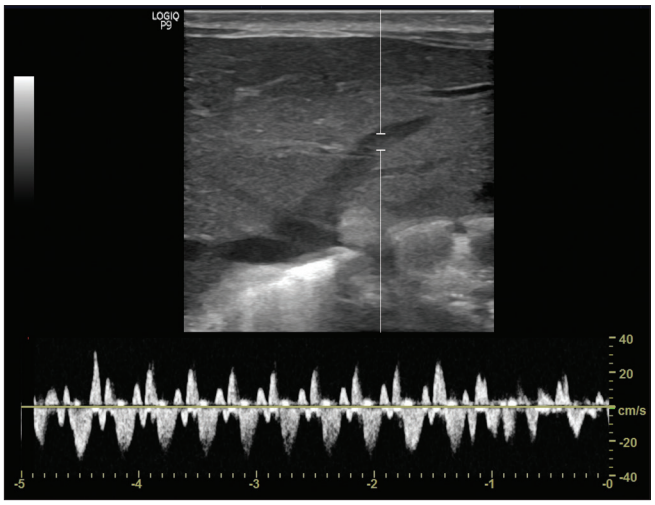
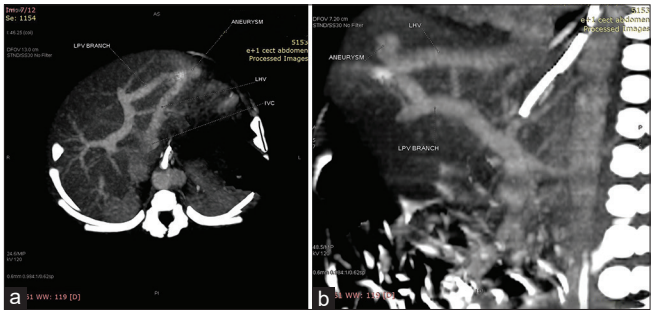



**Figure 5: Tetraphasic spectral waveform in the left hepatic vein**




**Figures 6: (a and b) Axial and sagittal contrast-enhanced computed tomography of the abdomen with maximum intensity projection images of the liver showing the left branch of the portal vein communicating with left hepatic vein through an aneurysm. IVC: Inferior vena cava**

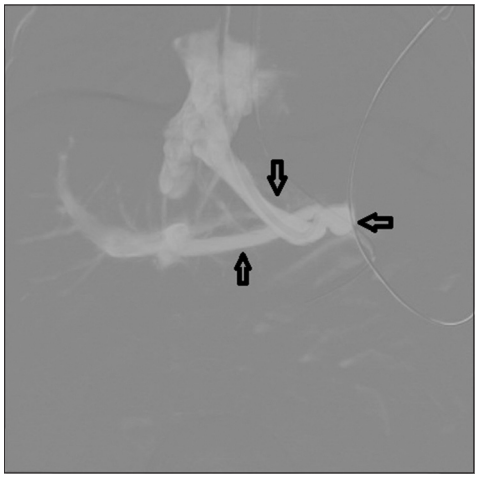


**Figure 7: Digital subtraction angiography image with catheter passing through inferior vena cava and left hepatic vein. The left branch of the portal vein (up arrow) showing aneurysmal communication (left arrow) with the left hepatic vein (down arrow)**

the absence of other identifiable maternal or fetal causes. In the neonatal period, abnormal galactosemia tests or cholestasis are common presenting signs. However, few of the cases are also detected incidentally. Beyond the neonatal age group, patients usually present from complications of the shunt.[4]

Various complications such as hepatic encephalopathy, neonatal cholestasis, liver tumors, PAH, and hepatopulmonary syndrome have been described in the literature. Bernard *et al.* reported the incidence of pulmonary hypertension in 30 out of 180 children at ages ranging from the neonatal period to 15 years (mean: 5 years 4 months) with all anatomic types of the shunt.[5] Congenital portosystemic shunt commonly coexists with congenital malformations, such as cardiovascular anomalies, polysplenia, annular pancreas, and biliary atresia. Some small intrahepatic portosystemic shunts located between the portal branches and hepatic veins disappear spontaneously by the age of 1–2 years.[4,6]

Park *et al.*[3] categorized intrahepatic portosystemic shunts arbitrarily into four different morphologic types. The first and most common type is a single large tube of a constant diameter that connects the inferior vena cava to the right portal vein. The second type includes single or multiple communications between peripheral branches of hepatic and portal veins in one hepatic segment. The third type is the rarest, with aneurysmal communication between the peripheral portal vein and hepatic vein. The fourth type has a peripheral portal and hepatic veins showing multiple communications diffusely involving both lobes.

Chevallier *et al.*[7] classified intrahepatic portosystemic venous shunts into four categories based on their clinical and anatomic features: Type I includes paraumbilical veins, such as those encountered in portal hypertension; Type II involves a connection between a portal branch and hepatic vein in adjacent liver segments; Type III comprises portal and hepatic vein connections between nonadjacent liver segments, and Type IV includes any connection between the right portal branch and inferior vena cava. Findings in our case were consistent with the Type III shunt per Park *et al.*[3] and Type II shunt per Chevallier *et al.*[7]

Grayscale sonography demonstrated a tubular anechoic channel connecting the portal vein branch with the hepatic vein. Color Doppler sonography demonstrated the presence of blood flow within the shunt including the direction of flow. De Gaetano *et al.*[8] have reported low velocity, bidirectional, or helical flow in the aneurysm on Doppler sonography. Multidetector computed tomography with contrast or magnetic resonance imaging of the abdomen with contrast can be used for delineation of shunt anatomy before intervention, excluding associated anomalies, especially if the ultrasound evaluation is suboptimal.

The vascular anomaly may regress spontaneously during infancy.[9] The definitive treatment for

portohepatic shunts can be considered in patients beyond the 1st year of life or those presenting with life-threatening complications related to the shunt. Most authors also agree that patients with symptomatic congenital portosystemic shunt should be treated.[10]

As there are no well-established guidelines, treatment protocols may vary between various centers.

Endovascular intervention is minimally invasive and usually the preferred treatment modality for shunt closure. Prompt reduction in the symptoms with correction of biochemical abnormalities can be noted following endovascular closure of shunt. Various endovascular embolization materials such as coils, balloons, and vascular plugs can be used depending on the shunt.[11] Surgery is preferred in cases with large shunts with a risk of inadvertent migration of embolic agents during embolization or after failed embolization with shunt recurrence or persistence.[11] Surgical options include ligation of the portal vein, resection, or even lobectomy. Treatment must be tailored according to the clinical presentation, type, and hemodynamics of the shunt, and availability of the treatment modalities.

## CONCLUSION

The portosystemic shunt can present with varied clinical manifestations. It is crucial to suspect it in case of unexplained pulmonary hypertension, as in most of the cases, it is treatable either by surgical or catheter interventions.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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