



## Case Report

# Liver Transplantation Experience in Two Children Diagnosed with Abernethy Type 1B Congenital Extrahepatic Portosystemic Shunt

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### Abstract

Congenital extrahepatic portosystemic shunt (CEPS), also known as Abernethy malformation, is a rare vascular anomaly characterized by diversion of portal venous blood away from the liver into the systemic circulation. Type 1 malformations are defined by the complete absence of intrahepatic portal venous flow and require liver transplantation as definitive treatment. We report two pediatric patients diagnosed with Abernethy type 1B malformation. The first patient presented with progressive cholestasis, growth retardation, and impaired liver synthetic function. Due to clinical deterioration and absence of intrahepatic portal flow, living donor liver transplantation was performed with portal vein reconstruction using an interposition graft. The second patient presented with hyperammonemia and neurocognitive impairment. Imaging confirmed type 1B CEPS, along with a focal hepatic lesion consistent with a benign regenerative nodule. The patient underwent successful living donor liver transplantation with standard portal reconstruction. Both patients had uneventful postoperative courses. During follow-up (18 and 24 months), liver function normalized and growth parameters improved, with no vascular complications observed. Liver transplantation remains the only definitive treatment for Abernethy type 1B malformation. Early diagnosis and meticulous surgical planning, particularly regarding portal inflow reconstruction, are essential for optimal outcomes.

**Keywords:** Abernethy malformation, liver transplantation, pediatric, portosystemic shunt

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Congenital extrahepatic portosystemic shunts (CEPS), first described by Abernethy, are rare vascular malformations in which portal venous blood bypasses the liver and drains directly into the systemic circulation.<sup>[1]</sup> These anomalies may result in significant metabolic disturbances and multisystem involvement in pediatric patients.<sup>[2,3]</sup> CEPS are classified into two main types based on the presence or absence of intrahepatic portal venous flow.<sup>[1,4]</sup> Altered hepatic perfusion may lead to a variety of radiological findings and the development of hepatic nodules.<sup>[5]</sup> In type 1 malformations, there is a complete absence of intrahepatic portal venous flow, and liver transplantation is considered the only definitive treatment.<sup>[6]</sup> In contrast, type 2 malformations involve partial shunting with preserved portal perfusion and may be managed with alternative approaches depending on anatomical and clinical features.<sup>[7]</sup> Here, we present two pediatric cases of Abernethy type 1B malformation successfully treated with living donor liver transplantation.

## Case Reports

### Case 1

A pediatric patient was referred with persistent jaundice, growth retardation, and biochemical evidence of impaired liver function. Laboratory evaluation revealed elevated bilirubin levels, abnormal transaminases, and decreased albumin levels, indicating impaired hepatic synthetic function. Imaging studies, including Doppler ultrasonography and contrast-enhanced computed tomography, demonstrated absence of intrahepatic portal venous branches and a portosystemic shunt consistent with Abernethy type 1B malformation.

Given progressive cholestasis, impaired liver function, and failure to thrive, liver transplantation was indicated. A living donor liver transplantation was performed.

**Surgical Technique:** Portal vein reconstruction was achieved using an interposition venous graft between the superior mesenteric vein–splenic vein confluence and the donor portal vein, ensuring adequate portal inflow.

The postoperative course was uneventful. Liver function normalized within weeks, and significant catch-up growth was observed during follow-up.

### Case 2

The second patient presented with hyperammonemia and neurocognitive symptoms. Laboratory tests revealed elevated serum ammonia levels with mildly abnormal liver enzymes.

Radiological evaluation confirmed Abernethy type 1B malformation with absence of intrahepatic portal venous flow.

A focal hepatic lesion detected on imaging was characterized as a benign regenerative nodule, likely secondary to altered hepatic perfusion.

Due to persistent metabolic abnormalities and risk of complications, living donor liver transplantation was performed with standard portal vein reconstruction.

The postoperative course was uneventful. Serum ammonia levels normalized, and neurocognitive symptoms improved. Follow-up imaging showed no recurrence of hepatic lesions.

## Discussion

Abernethy malformation is a rare but clinically significant vascular anomaly of the portal venous system.<sup>[1,4]</sup> Its anatomical characteristics and embryological basis have been further detailed in anatomical studies.<sup>[8]</sup>

Patients may present with a wide spectrum of clinical manifestations, including cholestasis, hepatic dysfunction, hepatopulmonary syndrome, and neurocognitive impairment due to hyperammonemia.<sup>[2,3]</sup> In addition, altered hepatic perfusion may predispose patients to the development of benign regenerative nodules or malignant hepatic tumors.<sup>[5]</sup>

The classification of CEPS is essential for determining management strategies. In type 1 malformations, the absence of intrahepatic portal venous flow precludes shunt closure, making liver transplantation the only definitive treatment option.<sup>[6]</sup> Additional anatomical classifications have been proposed to guide surgical planning and optimize portal inflow reconstruction.<sup>[11]</sup> Liver transplantation restores hepatic function, corrects metabolic abnormalities, and reduces the risk of malignant transformation.<sup>[12,13,14]</sup> A critical technical aspect of transplantation in these patients is the establishment of adequate portal inflow, which may require advanced surgical techniques.<sup>[11]</sup> Various surgical approaches, including the use of interposition grafts, have been described to achieve optimal portal vein reconstruction.<sup>[12,13]</sup> Our cases support previous reports demonstrating excellent outcomes following liver transplantation in pediatric patients with type 1B CEPS.<sup>[15]</sup>

## Conclusion

Abernethy type 1B malformation is a rare condition that requires early diagnosis and timely intervention. Liver transplantation remains the only definitive treatment. Careful preoperative evaluation and appropriate surgical techniques, particularly for portal inflow reconstruction, are essential to achieve successful outcomes.

## Disclosures

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