



Large-caliber congenital portocaval shunt and hepatopulmonary syndrome: Limitations of closure and the role of liver transplantation

Emilie Balent Aguirre¹ , José G. Cervantes¹ , Lucía Navarro² , Rosana Pérez Carusi² , Patricia D'Alia³ , Anahí Reartes³ , Carlos Luque⁴ , Miguel Palmeiro⁴ , Fabián Salgueiro⁴ , Fernando R. García Maida⁵ , Marcelo E. Lenz Virreira¹ , Fernando Cairo² , Francisco J. Mattera¹ , Emilio G. Quiñonez¹

ABSTRACT

Congenital portosystemic shunts (CPS) are rare vascular malformations associated with hepatic and extrahepatic complications, including hepatopulmonary syndrome (HPS).

We present a 16-year-old adolescent with no relevant medical history who presented with episodes of abdominal pain, jaundice, and dark urine. Ultrasound revealed a heterogeneous liver with multiple hyperechoic nodules and no visualization of the intrahepatic portal vein. Angiography demonstrated an extrahepatic portocaval shunt at the level of the portal trunk, with no opacification of the intrahepatic portal tree and low portal pressures; liver biopsy revealed regenerative nodular hyperplasia. Subsequently, he developed hypoxemia with orthodeoxygenation and required home oxygen therapy. He was placed on the waiting list and received an orthotopic liver transplant from a deceased donor.

This case highlights the need for timely diagnosis, proper anatomical characterization, and individualized treatment planning in CPS associated with HPS.

Keywords: congenital portosystemic shunt; Abernethy malformation; hepatopulmonary syndrome; liver transplantation.

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¹ Hepatopancreatobiliary Surgery and Liver Transplant Unit, Hospital de Alta Complejidad El Cruce, Florencio Varela, Argentina; ² Hepatology and Liver Transplant Service, Hospital de Alta Complejidad El Cruce, Florencio Varela, Argentina; ³ Gastroenterology and Hepatology Unit, Hospital General de Niños Pedro de Elizalde, Autonomous City of Buenos Aires, Argentina; ⁴ General Surgery Department, Hospital de Niños Ricardo Gutiérrez, Autonomous City of Buenos Aires, Argentina; ⁵ Pathology Department, Hospital de Alta Complejidad El Cruce, Florencio Varela, Argentina.

Correspondence to Emilie Balent Aguirre: emii.balent@hotmail.com

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INTRODUCTION

Congenital portosystemic shunts (CPS) are abnormal connections between the portal system and the systemic circulation that can partially or completely divert splanchnic blood flow. Their prevalence is estimated at 1 in 30 000 to 1 in 50 000 live births.¹ They can be intrahepatic or extrahepatic; the latter are classically referred to as Abernethy malformation. The clinical presentation is variable and includes cholestasis, hyperammonemia, nodular liver lesions, and cardiopulmonary complications, such as hepatopulmonary syndrome.¹

Hepatopulmonary syndrome (HPS) is defined by intrapulmonary vascular dilation with impaired gas exchange in the presence of liver disease or a portosystemic shunt;

According to PaO_2 , it is classified as mild (≥ 80 mmHg), moderate (60-79 mmHg), severe (50-59 mmHg), and very severe (< 50 mmHg).² In severe or irreversible cases, liver transplantation is the curative treatment.²

CLINICAL CASE

A 16-year-old patient with no relevant medical history who, during the two years prior to referral, experienced isolated episodes of abdominal pain, jaundice, and dark urine. Initial laboratory tests revealed elevated alkaline phosphatase (809 U/L) and total hyperbilirubinemia of 3.38 mg/dL, with mildly elevated transaminases (aspartate aminotransferase 34 U/L and alanine aminotransferase 41 U/L). He was evaluated at another institution: eutrophic (weight 51.5 kg; height 163 cm; body mass index [BMI] 19.3 kg/m²; BMI Z-score -0.19 [42nd percentile] and height Z-score -0.81 [21st percentile]), anicteric, and without visceromegaly. Ultrasound revealed a heterogeneous liver with multiple diffuse hyperechoic nodules (largest lesion 40 × 27 mm) and failed to visualize the portal vein.

Angiography/hemodynamics revealed patency of the celiac trunk and its branches, with an enlarged hepatic artery. Indirect portography demonstrated an extrahepatic portocaval shunt at the level of the main portal trunk, without opacification of the portal *carrefour* (Figure 1). Portal and suprahepatic venous pressures were 4 mmHg. A balloon occlusion attempt (30 mmHg) was made but did not result in complete occlusion due to the large diameter of the shunt. A biopsy of the largest nodule was performed and was consistent with regenerative nodular hyperplasia.

Subsequently, a laparotomy was performed at

another institution with the intention of correction (classified as Abernethy type II). The report described a large congenital communication between the portal vein and the inferior vena cava, without a bridge and in apposition, which prevented complete closure. Partial cerclage was performed with two 2-0 silk sutures, and a new liver biopsy was obtained, again consistent with regenerative nodular hyperplasia. This approach was based on an initial interpretation as Abernethy type II, potentially amenable to closure; however, the failure to achieve coaptation with a balloon, the absence of intrahepatic portal opacification, and the mesenteric-splenic confluence with systemic drainage were consistent with type Ib, leading to a change in strategy.

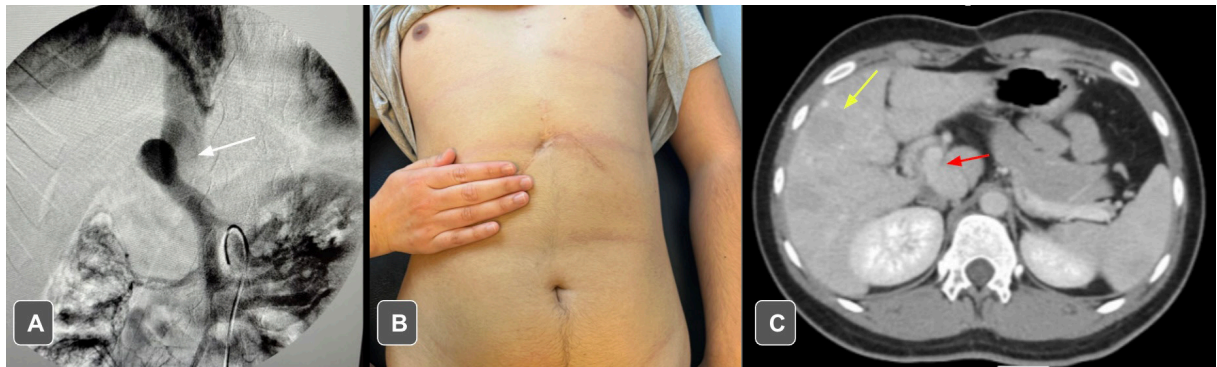
The patient developed progressive hypoxemia. Orthodeoxia was observed (peripheral oxygen saturation of 91% while seated and 89-90% while standing, with tachycardia), so home oxygen therapy was prescribed. Arterial blood gas analysis revealed severe hypoxemia (pH 7.41; PaCO_2 36.3; PaO_2 50 mmHg; SaO_2 85%) with an elevated alveolar-arterial oxygen gradient (A-aO_2 53).

The contrast-enhanced echocardiogram was positive for HPS, and the patient was referred to our center for evaluation by the liver transplant team.

The three-phase CT scan revealed multiple regenerative nodules (the largest in segment VIII, measuring 38 × 36 mm) and portal agenesis, with no cleavage plane between the portal vein and the inferior vena cava at the site of the shunt, and with the superior mesenteric vein and the splenic vein converging into a common portal trunk prior to systemic drainage—findings consistent with Abernethy malformation type Ib (Figure 1).

The FibroScan result was F0. Based on PaO_2 , the clinical picture was consistent with severe HPS. Given the progressive hypoxemia, the unfavorable anatomy for a secure closure, and the absence of a demonstrable portal bed, the case was discussed in a multidisciplinary conference, and it was decided to place the patient on the liver transplant waiting list.

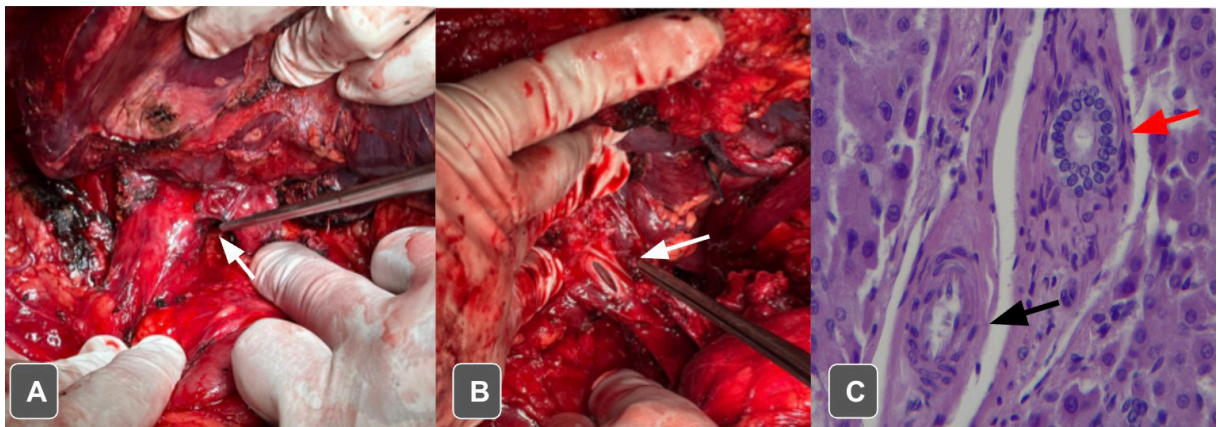
Two months later, he underwent an orthotopic liver transplant with a complete liver graft from a 20-year-old deceased donor. A total hepatectomy was performed with resection of the vena cava (Figure 2) and end-to-end cavo-caval (suprahepatic and infrahepatic), porto-portal, and arterial anastomoses, with end-to-end common bile duct-to-common bile duct

FIGURE 1. Preoperative images

A Indirect portogram: the white arrow points to the portocaval shunt.

B Physical examination on admission, revealing a scar from a previous laparotomy.

C Preoperative computed tomography: the red arrow points to the portocaval shunt, and the yellow arrow points to a hepatic regeneration nodule.

FIGURE 2. Intraoperative images

A Portosystemic shunt indicated by the white arrow.

B Ostium of the portal vein in the inferior vena cava after it has been transected during explantation, indicated by the white arrow.

C Microscopic examination (H&E × 40): portal space with an artery (black arrow) and bile duct (red arrow), without a portal vein.

biliary reconstruction. The cold ischemia time was 263 minutes, and the total surgical time was 270 minutes. He developed reperfusion syndrome with sustained hypotension, which required epinephrine and vasopressor support. Intraoperative Doppler ultrasound confirmed vascular patency and adequate blood flow. Two units of packed red blood cells were transfused.

He was admitted to the intensive care unit on mechanical ventilation and receiving low-dose norepinephrine. Forty-eight hours later, he was extubated; subsequently, he experienced a tonic-clonic seizure accompanied by impaired consciousness, requiring reintubation. The

electroencephalogram showed no epileptiform discharges, and the MRI revealed acute multifocal ischemic lesions and microhemorrhages. Doppler ultrasounds were performed on days 1, 2, and 3 post-transplant (*Table 1*). *Table 2* summarizes selected laboratory parameters. At 21 days, the patient was discharged home with a favorable clinical course on a regimen of methylprednisolone 20 mg/day, tacrolimus 4 mg every 12 hours, and mycophenolate mofetil 1000 mg every 12 hours.

The histopathological examination of the explant revealed regenerative micronodules without fibrous septa, with marked vascular

TABLE 1. Changes in Doppler ultrasound findings on the 1st, 2nd and 3rd days post-transplant

Day after transplantation	Portal vein	Suprahepatic veins	Hepatic artery (RI)
Day 1	Patent, hepatopetal flow; diameter 10 mm; VPMax 68 cm/s	Triphasic flow	Proximal 0.60; hilum 0.45
Day 2	Patent, hepatopetal flow; diameter 10 mm; VPMax 110 cm/s	Triphasic flow	Proximal 0.42; hilum 0.36; left intrahepatic 0.35; right 0.25
Day 3	Patent, hepatopetal flow; 10 mm in diameter; VPMax 88 cm/s	Triphasic flow	Proximal 0.50; hilum 0.51; left intrahepatic 0.36; right 0.43

RI: resistance index; VPMax: maximum portal vein velocity.

TABLE 2. Post-transplant laboratory values

Time	pH	HCO ₃ ⁻ (mmol/L)	Lactate (mmol/L)	AST (U/L)	ALT (U/L)	Total bilirubin (mg/dL)	PT (%)	Factor V (%)
Immediate POP	7.33	22.3	5.7	1,218	918	2.98	21	16
24 h	7.45	25.4	1.8	1,075	1,300	3.21	51	77
72 h	7.41	21.2	0.8	149	484	1.89	77	—*
7 days	7.37	27.1	1.8	167	411	1.02	79	—*

*Not recorded in the registry at that time.

POP: postoperative; HCO₃⁻: bicarbonate; AST: aspartate aminotransferase; ALT: alanine aminotransferase; PT: prothrombin time.

dilation and mild sinusoidal dilation, consistent with regenerative nodular hyperplasia; and abnormal vascular architecture with distortion of portal-central relationships, with stenosis/obliteration of portal veins (*Figure 2*).

Six months after the transplant, the patient is asymptomatic from a respiratory standpoint, with a baseline peripheral oxygen saturation of 95% and a need for nighttime home oxygen therapy; spirometry showed a forced vital capacity of 69% of predicted, and the chest X-ray showed no evidence of pleural or parenchymal involvement. No neurological sequelae.

DISCUSSION

Extrahepatic CPSs are rare and heterogeneous conditions. The anatomical characterization of the shunt and the intrahepatic portal system is essential for guiding treatment and avoiding ineffective interventions.^{1,3,4} The Morgan and Superina classification distinguishes type I (complete shunt)—which is subdivided into Ia (separate drainage of the superior mesenteric and splenic veins) and Ib (confluence into a common trunk)—from type II (partial shunt).⁵ In our patient, the findings were consistent with Abernethy type Ib.

Recent recommendations propose a stepwise approach that uses Doppler ultrasound, CT/

MRI, and targeted hemodynamic evaluation, emphasizing avoidance of unnecessary transplants when closure is feasible.^{4,6,7} The diagnosis of “congenital absence of the portal vein” may be overestimated if venography with transient balloon occlusion is not performed, since occlusion may reveal previously invisible hypoplastic portal branches and allow for closure strategies.⁸ However, when the shunt is large-bore and there is no demonstrable intrahepatic portal bed, closure may cause a sharp increase in portal pressure, with a risk of acute portal hypertension, splenic-mesenteric congestion, intestinal edema, and ischemia; if a portal bed is present, progressive closure is considered based on the hemodynamic response.^{4,6} In our case, balloon occlusion was not achieved due to the large caliber of the shunt, and CT scanning suggested portal agenesis, which limited the feasibility of safe occlusion.

The picture became more complicated with HPS, which can occur in the context of portosystemic shunts even in the absence of cirrhosis.² Although pediatric cases have been reported in which the condition reversed following ligation of the shunt when closure was possible,⁹ in situations where closure is not feasible or the patient develops severe hypoxemia, liver

transplantation is a definitive alternative.^{4,10,11} After liver transplantation, HPS resolves in most cases; in a recent meta-analysis, resolution was observed in approximately 90% of patients at 6 months, with complete resolution within the first year in the patients evaluated.¹²

CPS/Abernethy is also associated with nodular liver lesions—ranging from benign lesions to tumors—linked to the absence of portal flow and a compensatory increase in arterial flow.^{13,14} In our patient, the histological findings of regenerative micronodules, vascular dilation, and portal obliteration reflect this pathophysiology; likewise, a preoperative biopsy showing the absence or marked hypoplasia of portal veins within the portal spaces can serve as a guiding finding in the appropriate context. However, the therapeutic decision should be based primarily on the shunt's anatomy and hemodynamic evaluation.^{4,15}

In summary, the course of action should be determined based on anatomy and hemodynamics: closure if a portal bed is present and there is definitive evidence of occlusion; transplantation in cases of shunts that cannot be occluded, absence of a functional portal bed, or serious complications such as severe or very severe hepatopulmonary syndrome. ■

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