Pediatric Living Donor Liver Transplantation for Congenital Absence of the Portal Vein With Pulmonary Hypertension: A Case Report

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ABSTRACT

Few reports of liver transplantation exist in patients with congenital absence of the portal vein and pulmonary hypertension. Living donor liver transplantation is usually performed before exacerbation of pulmonary hypertension. A 7-year-old girl (height: 131.5 cm; weight: 27.4 kg) with congenital absence of the portal vein was diagnosed with pulmonary hypertension (mean pulmonary artery pressure 35 mm Hg), and liver transplantation was planned before exacerbation of pulmonary hypertension. We successfully managed her hemodynamic parameters using low-dose dopamine and noradrenaline under monitoring of arterial blood pressure, central venous pressure, cardiac output, and stroke volume variation. Anesthesia was maintained using air-oxygen-sevoflurane and remifentanil 0.1 to 0.6 $\mu g \cdot kg^{-1} \cdot min^{-1}$. It is necessary to understand the potential perioperative complications in such cases and to adopt a multidisciplinary team approach in terms of the timing of transplantation and readiness to deal with exacerbation of pulmonary hypertension.

CONGENITAL absence of the portal vein (CAPV) is a type of portosystemic shunt that can cause hepatic encephalopathy, hepatopulmonary syndrome, and portopulmonary syndrome [1]. CAPV is classified as portal vein deficiency (type 1) and hypoplasia (type 2). Type 1 CAPV requires liver transplantation in cases complicated with pulmonary hypertension or liver failure. Type 2 cases require surgical management or catheter embolization of the shunt.

Only a few reports exist of pediatric liver transplantation in patients with CAPV with pulmonary hypertension [2–7]. We report here the anesthetic management of living donor liver transplantation (LDLT) in a 7-year-old girl with CAPV and pulmonary hypertension.

CASE PRESENTATION

Written informed consent was obtained from the patient and her parents for publication of this case report and accompanying images. A 7-year-old girl (height: 131.5 cm, weight: 27.4 kg) had type 1 CAPV. Although she had no positive family history, a congenital metabolic abnormality test at birth indicated galactosemia, after which an abdominal computed tomography (CT) scan was performed. She had no mental deterioration and had normal exercise tolerance. Our patient was diagnosed with CAPV type 1b, with the splenic vein serving as the portal vein, flowing into the inferior vena cava (IVC) without going through the liver (Fig 1A and B); hence, the patient had no collateral veins and no portal hypertension. Although the patient did not have any clinical symptoms, persistent hyperammonemia was conservatively managed by oral lactulose.

Pulmonary hypertension (mean pulmonary artery pressure of 35 mm Hg) was diagnosed by echocardiography at the age of 7 years, although she did not have any symptoms of dyspnea. Her aspartate aminotransferase and alanine transaminase were only slightly elevated, although portal contrast and abdominal CT scan showed that the native portal vein trunk flowed into infrahepatic IVC as type 1 CAPV. Cardiac catheterization revealed a mean pulmonary artery pressure of 35 mm Hg, pulmonary artery resistance of 600 $dyne \cdot sec \cdot cm^{-5}$, and cardiac index of 2.79 $L \cdot min^{1} \cdot m^{2}$, leading to a diagnosis of mild pulmonary hypertension. Since we anticipated that liver transplantation would be required in the future, our surgical team and pediatric doctors discussed its timing many times.

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Finally, based on several case reports showing that performing transplantation before exacerbation of pulmonary hypertension was the appropriate treatment strategy, we decided to proceed with the liver transplantation.

She underwent LDLT using an extended left liver lobe graft from her mother. In such cases, perioperative issues include the possibility of prolonged drug effects and exacerbation of pulmonary hypertension [4]. Due to the risk of exacerbation of pulmonary hypertension at the time of induction of anesthesia and reperfusion, the required equipment for extracorporeal membrane oxygenation (ECMO) and nitric oxide inhalation was kept available preoperatively, in consultation with the cardiac surgery department. In our preoperative conference (anesthesiologists and surgical teams), we determined that in case of circulatory collapse, the cardiac surgical team would help establish ECMO assistance. Hence, the surgical team preoperatively performed cannulation of the femoral artery and vein in order to facilitate rescue ECMO.

General anesthesia was induced without premedication using air-oxygen-sevoflurane inhalation, with intravenous injection of 50 μg fentanyl and 20 mg rocuronium to facilitate tracheal intubation. After induction, 2 other peripheral venous lines and a triple-lumen central venous line were inserted into the right jugular vein. Anesthesia was maintained using air-oxygen-sevoflurane and remifentanil 0.1 to 0.6 $\mu g \cdot kg^{1} \cdot min^{-1}$. Intraoperatively, we monitored central venous pressure, cardiac output, and stroke volume (Vigileo monitor; FloTrac system; Edwards Lifesciences, Irvine, Calif, United States), in addition to standard monitoring. The estimated blood loss was relatively small during the anhepatic phase, which lasted for 141 minutes. Low-dose dopamine (1.3-4.6 $\mu g \cdot kg^{-1} \cdot min^{-1}$) and noradrenaline (0.01 $\mu g \cdot kg^{-1} \cdot min^{-1}$) were used for maintenance of vital signs before reperfusion. With this management, her hemodynamic parameters could be managed without nitric oxide inhalation and ECMO during and after reperfusion. Our surgical team also prepared for cannulation of the femoral and axial veins, in case veno-venous bypass was needed to prevent circulatory collapse at the time of IVC cross clamping, although the case was successfully managed without using veno-venous bypass. As for the recipient procedure, the native portal vein trunk was originally isolated from hepatoduodenal ligament and flow into infrahepatic IVC. The native hepatectomy was completed with preservation of the inferior vena cava and confluence of portal vein trunk as type 1 CAPV (Fig 2A). In the anhepatic phase, massive hemorrhage from the perihepatic area was almost completely stanched.

Implantation of the left lobe liver graft was started with reconstruction of the hepatic vein. The left and middle hepatic vein of the liver graft was anastomosed to native left and middle hepatic vein by 5-0 prolene running suture as the common channel. Subsequently, the native portal vein trunk was divided from the IVC with the wall of vena cava, which led to a large orifice for anastomosis. The native portal vein was anastomosed to the left portal branch of the liver graft by 6-0 prolene running suture with no size mismatch of the venous orifice. After that, the hepatic artery was reconstructed between the native proper hepatic artery of the recipient and the left hepatic artery of the graft under surgical loupe, followed by duct-to-duct biliary anastomosis (Fig 2B).

Intraoperatively, arterial blood gases were monitored every hour and successfully managed without severe hypoxia, acidosis, or hypercapnia. Body temperature was maintained between 37.2°C and 38.5°C using a forced-air warming system. The estimated total blood loss was 290 mL, and fluids administered included 290 mL of albumin, 520 mL of crystalloids, and 380 mL of colloids. Surgical duration was 7 hours 16 minutes. The patient was transferred to the intensive care unit postoperatively without extubation. Her postoperative course was stable, and she was extubated the following day. She was discharged from the intensive care unit on postoperative day 14 without acute cellular rejection or worsening of pulmonary hypertension. Her average pulmonary artery pressure improved to 26 mm Hg, as seen on thoracic echocardiography. Her portal vein was finally constructed after LDLT after 2 months (Fig 3).

DISCUSSION

Between 1973 and 2019, 84 cases of CAPV were reported, but only 1 of these cases had no clinical symptoms [3]. The reports included cases with airway and tracheal abnormalities due to Goldenhar syndrome, Costello syndrome, and stenosis of the trachea, in addition to congenital heart disease and chromosomal aberrations [4]. Since these conditions are often associated with difficult airways or intubation, thorough preoperative assessment for identification of associated congenital abnormalities is required in patients with CAPV undergoing surgery under general anesthesia. However, our patient did not have any other

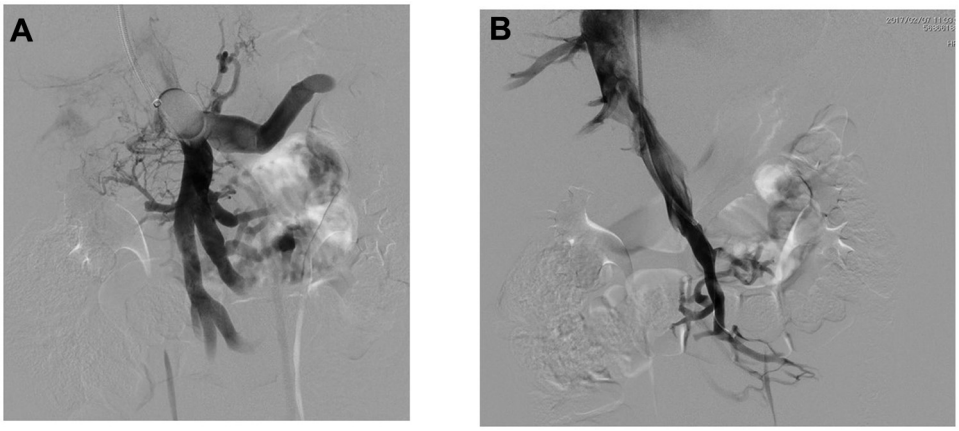

Fig 1. Preoperative venous angiography. **(A)** The intrahepatic portal vein was absent, and the splenic vein served as the portal vein. **(B)** The portal vein flowed into the inferior vena cava without going through the liver.

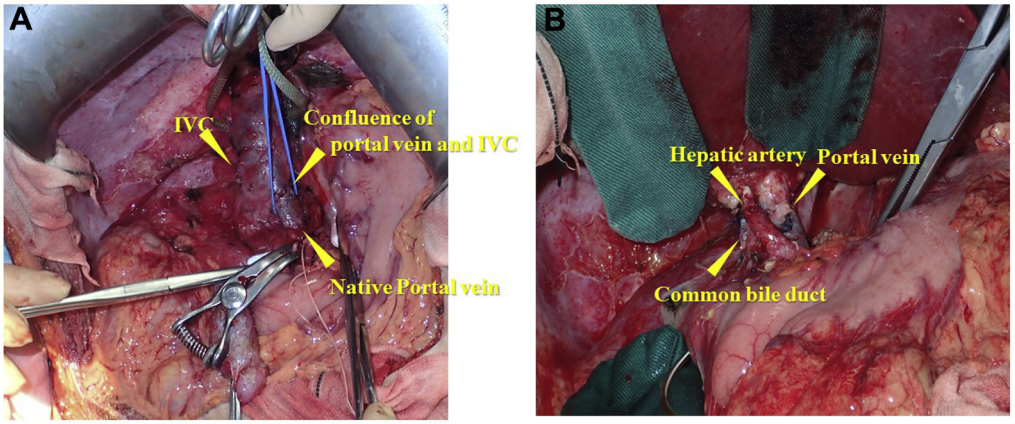

Fig 2. **(A)** After hepatectomy, native portal vein trunk flowing into inferior vena cava (IVC) was completely preserved. Confluence of portal vein and IVC could work as porto-caval shunt during a hepatic phase. **(B)** The final view of the reconstruction of hepatic artery and portal vein.

congenital malformations likely to be associated with a difficult airway and difficult tracheal intubation.

There are 13 previous reports of cases of pediatric liver transplantation for CAPV [5], only 2 of which were performed because of pulmonary hypertension [6,7], as in our case. The reason for liver transplantation in the 13 previous reports was biliary atresia in 4 cases, pulmonary hypertension in 4 cases (2 cases of portopulmonary syndrome and 2 cases similar to ours), portosystemic encephalopathy in 4 cases, and hepatoblastoma in 1 case [5]. In cases with pulmonary hypertension, the timing of liver transplantation is extremely important in terms of reducing mortality. Hence, in previous reports, performance of liver transplantation before exacerbation of pulmonary hypertension was considered appropriate [5–7]. In our case, we believe that performing liver transplantation before exacerbation of pulmonary hypertension was one of the factors that facilitated good perioperative management.

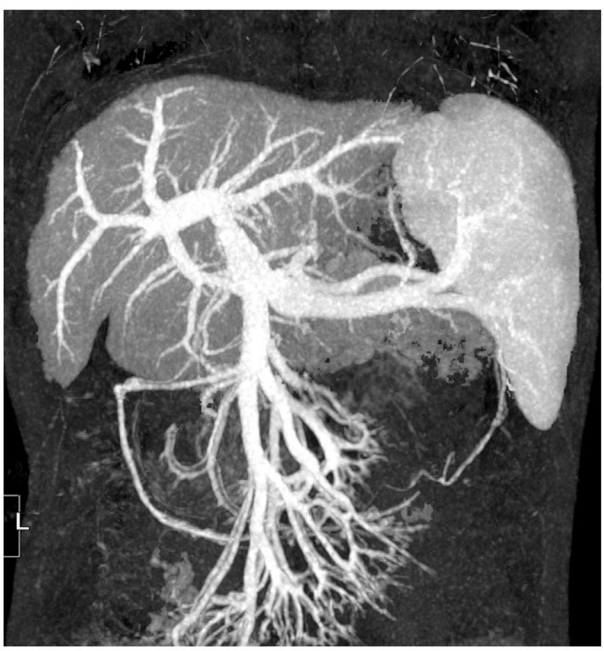

Fig 3. Postoperative coronal contrast computed tomography scan 2 months after living donor liver transplantation. The portal vein was constructed after living donor liver transplantation.

During general anesthesia in children with pulmonary hypertension, various factors can lead to exacerbation of pulmonary hypertension, which requires strict management. Specific exacerbators of pulmonary hypertension include hypoxemia, hypercapnia, acidosis, invasive stress, and hypothermia [8]. It is also necessary to monitor and appropriately maintain fluid balance and acid–base equilibrium. If pulmonary vascular resistance is elevated, it should be lowered by administration of pure oxygen or inhalation of nitric oxide. At the same time, use of catecholamines, such as norepinephrine, vasopressin, dobutamine, and milrinone, should be considered for maintenance of systemic blood flow, including coronary blood flow. In the unlikely event that circulatory dynamics fail, it is also important to consult the surgical and co-medical team to ensure appropriate measures to save the patient's life.

Regarding monitoring of pulmonary hypertension in children, the use of pulmonary artery catheters and transesophageal echocardiography is recommended for cases with severe pulmonary hypertension and low cardiac function [9]. However, in our case, surgery was completed uneventfully without monitoring of pulmonary artery pressure and transesophageal echocardiography, suggesting that the need for invasive monitoring should be tailored for each case in children without severe pulmonary hypertension.

Since our patient was a child with mild pulmonary hypertension, we decided at the outset not to place a pulmonary artery catheter. Also, since upper gastrointestinal

endoscopy was not performed before surgery and esophageal varices could not be ruled out, transesophageal echocardiography was not performed. At our institution, when performing liver transplantation in children, we attempt to keep bleeding to a minimum; hence, we perform transesophageal echocardiography only when exacerbation of pulmonary hypertension during surgery is suspected. In the current case, however, since both surgeons and anesthesiologists performed adequate management and intervention, surgery was accomplished without circulatory collapse. Our experience suggests that the need for invasive monitoring should be tailored for each case and at each facility. Appropriate perioperative management in such cases requires an understanding of the potential complications during the perioperative period and full discussion within the medical team about the timing of transplantation and preparation for exacerbation of pulmonary hypertension.

In conclusion, we successfully managed the case of a 7-year-old girl with CAPV and portopulmonary syndrome. To ensure appropriate perioperative management, it is necessary to understand the potential perioperative complications in such cases and to adopt a multidisciplinary team approach in terms of the timing of transplantation and readiness to deal with exacerbation of pulmonary hypertension.

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