Emergency living donor liver transplantation under extracorporeal membrane oxygenation in an infant with biliary atresia-polysplenia syndrome

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Received October 2, 2021
Revised October 5, 2021
Accepted October 7, 2021
Published online October 19, 2021

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Biliary atresia-polysplenia syndrome (BAPS) is diagnosed in a small number of patients with biliary atresia (BA). We present a case of emergency living donor liver transplantation (LT) successfully performed in an infant with BAPS undergoing extracorporeal membrane oxygenation. The recipient was a 10-month-old boy who did not undergo Kasai portoenterostomy due to rapid progression of liver cirrhosis. Co-existing malformations included heterotopic inferior vena cava without hepatic communication, direct hepatic vein drainage into the right atrium, polysplenia, intestinal malrotation, truncated pancreas, and preduodenal portal vein and annular pancreas. Patient condition deteriorated rapidly after pulmonary hemorrhage, and thus emergency living donor LT was performed after starting veno-venous extracorporeal membrane oxygenation (ECMO) with a pediatric end-stage liver disease score of 32. A left lateral section graft obtained from his father showed a graft-to-recipient weight ratio of 3.2%. The recipient surgery was performed according to standard procedures of pediatric LT. The graft hepatic vein was directly anastomosed with the suprahepatic confluence of the recipient hepatic veins. An external iliac vein homograft was interposed for portal vein reconstruction. Multiple portal collateral veins were ligated and intraoperative portography was performed to secure portal vein inflow. The patient was weaned off ECMO and ventilator were weaned off on 17 days and 65 days respectively after transplantation. The patient stayed at the intensive care unit for 3 months before and after transplantation. Our pediatric patient with BAPS manifested various anatomical malformations. Successful LT requires comprehensive preoperative and intraoperative assessment of these anomalies, adoption of customized reconstruction techniques of LT, and careful posttransplant monitoring.

Keywords: Prediportal portal vein; Inferior vena cava; Intestinal malrotation; Vascular complication; Pediatric transplantation

INTRODUCTION

Biliary atresia (BA) is a rare disease, but it is the most common indication for liver transplantation (LT) in children. A small number of patients with BA are diagnosed with biliary atresia-polysplenia syndrome (BAPS) [1-6]. BAPS symptoms include BA and other malformations such as polysplenia, preduodenal portal vein, situs inversus, duodenal or small bowel atresia, midgut malrotation, cardiovascular anomalies, interrupted or absent infrahepatic vena cava with azygous or hemiazygous continuation, and atypical hepatic arterial supply. BAPS is regarded as a high-risk factor for LT because of technical difficulties associated with anomaly in LT operation and frequent posttransplant vascular complications [4-8]. Extracorporeal membrane oxygenation (ECMO) is a form of life support used for patients with life-threatening heart and/or lung problems [9-11]. We present a case of emergency living donor liver transplantation (LDLT) using a left lateral section graft successfully performed in a 10-month-old patient with BAPS under ECMO.

CASE PRESENTATION

The recipient was a 10-month-old boy who did not undergo Kasai portoenterostomy due to rapid progression of liver cirrhosis (Fig. 1). A review of pretransplant computed tomography (CT) findings showed overt liver cirrhosis and concurrent abnormalities including heterotopic inferior vena cava (IVC) without communication with the liver, direct hepatic vein drainage into the right atrium, polysplenia, intestinal malrotation, truncated pancreas, preduodenal portal vein, and annular pancreas (Fig. 2). These findings suggested a diagnosis of BAPS. The patient's general condition and liver function deteriorated progressively. Pulmonary hemorrhage occurred suddenly during the planning for LDLT. Patient condition worsened rapidly and led to acute respiratory distress syndrome, warranting veno-venous ECMO for oxygenation support and emergency LDLT was performed (Fig. 3). At the time of LDLT operation, the patient's height and body weight were 64 cm and 10 kg with a Pediatric End-stage Liver Disease score of 32 (total bilirubin 25.3 mg/dL, albumin 2.9 g/dL, prothrombin international normalized ratio 1.96, and growth failure).

The donor was 35-year-old father of the patient. The left

Fig. 1. Pretransplant computed tomography (CT) scans. (A) CT taken at 3 months of age reveals overt liver cirrhosis. (B) CT taken at 6 months of age shows progression of liver cirrhosis. (C, D) CT taken at 10 months of age indicates marked progression of liver cirrhosis with formation of ascites.

Fig. 2. Images of vascular reconstruction of the retransplant computed tomography taken 3 days before liver transplantation. (A) Two anomalous superior mesenteric veins (arrows) are joined to make a single portal vein. (B) Umbilical collateral veins (arrow) are visibly drained at the upper abdominal wall. (C) Anomalous course of the hepatic artery (arrow) is visible. (D) The heterotopic inferior vena cava (arrow) has no hepatic vein branch from the liver.

Fig. 3. Peritransplant simple x-ray images. (A) Pulmonary hemorrhage occurred before liver transplantation, interfering pulmonary oxygenation. (B) Veno-venous extracorporeal membrane oxygenation (arrow) was initiated through the jugular vein catheters before transplantation and continued until recovery of pulmonary function.

lateral section graft weighed 320 g, which is equivalent to a graft-to-recipient weight ratio of 3.2%.

During recipient hepatectomy, we identified agenesis of

the retrohepatic IVC and the native IVC was buried deep inside and close to the vertebral bodies. There was a strange diversion of superior mesenteric veins, which joined as a preduodenal portal vein (Fig. 4). Polysplenia and intestinal malrotation were also identified. A large umbilical hernia was present with massive ascites. The concurrent malformations in this patient are summarized in Table 1 [1-5].

The umbilical collateral veins drained along the upper abdominal wall into the chest and the lower abdominal wall into the pelvis. The umbilical collateral veins draining toward the pelvis were transected and those serving the chest were preserved during liver dissection. The liver was severely cirrhotic and stony hard. A rudimentary gallbladder was identified. The hepatic hilum was markedly adherent and therefore very meticulous dissection was performed to prevent unnecessary excessive bleeding. The gallbladder was removed, but the common bile duct was absent. The main portal vein was isolated without dissection of the preduodenal portion. The hepatic artery was also meticulously dissected because of its anomalous supply from the lesser curvature of the stomach. Because the retrohepatic IVC was absent, the suprahepatic hepatic vein stump was

Fig. 4. Intraoperative photographs showing the combined vascular anomalies. (A) The liver was markedly cirrhotic and shows large umbilical collateral veins (arrow). (B) The portal vein runs along the ventral surface of the duodenum, forming the preduodenal portal vein (arrow). (C) The isolated portal vein (blue loop) and hepatic artery (yellow loop) are visible after completion of hilar dissection. No common bile duct was identified. (D) The retrohepatic inferior vena cava was absent (arrow) and the hepatic vein stump is directly drained into the right atrium.

Table 1. Comparison of the anatomical malformations described in the literature and in the present case [1-5]

Malformations	Frequency in literature	Case findings
Splenic malformation	100%	Polysplenia
Situs inversus	37%	Absent
Portal vein anomaly	40%	Predudodenal portal vein
Intestinal malrotation	60%	Present
Agenesis of the IVC	70%	Heterotopic IVC
Cardiac anomaly	45%	Present
Pancreatic anomaly	11%	Present

IVC, inferior vena cava.

directly isolated (Fig. 4). Intraoperative ultrasonography revealed that the transverse diameter of the suprahepatic hepatic vein stump was approximately 15 mm.

A left lateral section graft was harvested after completion of recipient liver dissection. The graft hepatic vein resembled figure 8, and thus the 15 mm and 5 mm-sized hepatic vein openings were unified with 5-0 polydioxanone sutures, making a single 20 mm-sized single orifice (Fig. 5).

At the start of recipient hepatectomy, the umbilical collateral veins draining toward the chest were transected. The portal vein and hepatic artery were transected. After the

lower portion of the right atrium was clamped with a large vascular clamp, the hepatic vein stump was transected at the level of hepatic parenchyma. The septum at the hepatic vein stump was cut to make a single orifice measuring 15 mm in width (Fig. 5). The recipient portal vein was hypoplastic, thus an iliac vein homograft was anastomosed as an interposition conduit (Fig. 6).

Graft implantation was initiated with size-matched anastomosis to the recipient hepatic vein stumps (Fig. 5). The preduodenal portal vein was rather redundant, thus the majority of the interposed iliac vein homograft was removed to prevent excessive redundancy during portal vein reconstruction (Fig. 6). The portal vein blood flow was rather lower than expected, thus the retrosplenic collateral veins were completely ligated after portal reperfusion (Fig. 7). To secure portal blood flow, intraoperative portography was performed via a small branch of the superior mesenteric vein, in which no residual collateral veins with good portal flow through the two diverted superior mesenteric vein branches were not identified (Fig. 7). The graft hepatic artery was reconstructed under surgical microscopy. Finally, Roux-en-Y hepaticojejunostomy was performed for biliary reconstruction. The umbilical hernia was directly repaired, followed by appendectomy.

The explant liver showed BA-associated liver cirrhosis

Fig. 5. Intraoperative photographs showing reconstruction of the graft hepatic vein. (A) The suprahepatic hepatic vein stump was exposed after deep clamping of the right atrium. (B, C) The septum at the hepatic vein stump was cut to make a single orifice (arrow). (D) The graft hepatic vein stump was unified (arrow) to match with the recipient hepatic vein stump. (E, F) Graft hepatic vein was reconstructed through a size-matched anastomosis.

with marked bile ductular proliferation and inflammatory obliteration of the perihilar bile ducts (Fig. 8).

Posttransplant recovery of the patient was markedly delayed. The patient was weaned of ECMO at 17 days post-transplantation. Ventilator support continued due to failure of early ventilator weaning, thus tracheostomy was performed. The patient was weaned of ventilator at 65 days post-transplantation. The early follow-up CT scan showed no vascular complications (Fig. 9). The patient recovered slowly from LT operation. The patient stayed at the intensive care unit for 82 days after transplantation. The patient is currently staying at the general ward with normal liver function and improved general condition at 4 months after LT.

DISCUSSION

BA is an obstructive neonatal cholangiopathy caused by inflammation resulting in progressive fibrosis and obliteration of the intrahepatic and extrahepatic bile ducts [12]. BA is the most common cause of neonatal cholestasis and biliary obstruction and occurs in 1 in 15,000 to 20,000 live births. A majority of infant patients with BA undergo Kasai portoenterostomy, but the present patient was not indicated because liver cirrhosis was already developed early at 2 months of age. Thus, the present patient was planned for LT at the age of 1 year.

Associated vascular and visceral malformations have

Fig. 6. Intraoperative photographs showing reconstruction of the graft portal vein. (A) The interposed iliac vein conduit (arrow) connected to the preduodenal portal vein appears to be excessively redundant. (B) Portal vein anastomosis was performed after removal of the majority of the interposed iliac vein homograft. (C, D) A small segment of interposed iliac vein homograft (arrow) was left to enlarge the diameter of the native portal vein.

Fig. 7. Findings of intraoperative portography. (A) Multiple retrosplenic collateral veins were ligated immediately after portal reperfusion. (B) Two anomalous superior mesenteric veins (arrows) are joined to make a single portal vein. No anastomotic stenosis of the portal vein is detected. (C, D) Multiple collateral veins are visible around spleen, but all of them were interrupted by multiple sutures.

been reported in 10% to 20% of patients with BA [7,13]. BA syndrome with splenic malformation has been identified as a subset with distinct etiology. In BAPS, BA coexists with extrahepatic malformations of which the most common is polysplenia, which may be associated with thoraco-abdominal heterotaxy, such as intestinal malrotation and vascular disorders [2].

Fig. 8. Gross photograph of the explant liver.

Early trials of LT in children with BAPS were disappointing because of the high rates of vascular complication attributed to patients' abnormalities [3,4]. The main cause of mortality and morbidity is related to technical challenges involving such cases [5-8]. By contrast, recent studies reported that the long-term outcomes of LT in children with BAPS may be as good as in the usual non-syndromic BA [6,7].

Absence of the retrohepatic IVC at the time of LT operation does not pose significant problems with hepatic outflow reconstruction regardless of the graft types because the confluence of the recipient hepatic veins used for graft outflow vein anastomosis is effective for outflow reconstruction, as shown in our previous case [6]. In contrast, the present patient had a unique heterotopic IVC, which was not connected with the liver. The hepatic vein stump was rather hypoplastic, thus we paid special attention to prevent graft outflow hepatic vein stenosis.

The preduodenal portal vein is a common component of BAPS. For this reason, a high incidence of portal vein thrombosis is reported in direct anastomosis with the preduodenal portal vein, which might be associated with portal vein hypoplasia in BA and abnormal anatomical position of the recipient portal vein in PS [7]. This risk of portal

Fig. 9. Posttransplant computed tomography scan taken 21 days after transplantation. (A, B) Graft hepatic vein outflow (arrows) was patent. (C, D) The preduodenal portal vein and graft portal vein (arrows) were reconstructed without anastomotic stenosis.

vein thrombosis or insufficiency in the present patient was managed via a vein homograft interposition to enlarge the portal vein diameter, which effectively offset the anatomical malposition of the reconstructed portal vein [6,14].

Hepatic artery anomaly is also reported to be common in BAPS. In the present patient, the course of the hepatic artery was anomalous, entailing very meticulous dissection to obtain a sufficient length suitable for arterial reconstruction.

In the present patient, sudden occurrence of pulmonary hemorrhage led to acute respiratory distress syndrome, warranting veno-venous ECMO to enhance oxygenation before LDLT [9,10]. ECMO was continued for 17 days until recovery of oxygenation through ventilator support. The patient was weaned off ventilator at 65 days after transplantation. The present patient was the second case of pediatric LDLT using intraoperative ECMO in our institutional experience with pediatric LT [11].

In conclusion, our pediatric patient with BAPS manifested various anatomical malformations. Successful LT requires comprehensive preoperative and intraoperative assessment of these anomalies, adoption of customized reconstruction techniques of LT, and careful posttransplant monitoring.

FUNDING

There was no funding related to this study.

CONFLICT OF INTEREST

All authors have no conflicts of interest to declare.

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Conceptualization: SH, JMN. Data curation: JMN, SHO, KMK. Methodology: JMN, GCP, HK. Visualization: SH. Writing - original draft: JMN, SH. Writing - review & editing: All.

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