Living Donor Liver Transplantation Using a Liver Graft With Congenital Intrahepatic Portosystemic Shunt

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Background. Despite of recent development of imaging modalities, congenital intrahepatic portosystemic shunt (IPSS) is rarely diagnosed. Therefore, living donor liver transplantation using a liver graft with IPSS has not been previously published. Materials and Methods. We report a 28-year-old male patient with end-stage liver disease secondary to Wilson disease. His 26-year-old brother was a potential living donor, who had an IPSS of 25 mm in diameter at segment 6 as shown by computed tomography. Liver function tests were normal, and blood ammonia concentration was in the upper limit of normal. Results. Living donor liver transplantation was uneventfully performed. After surgery, a recipient liver function tests showed a quick recovery, and serum ammonia levels were consistently normal. Although thrombosis inside the IPSS was confirmed by computed tomography on postoperative day 21, this thrombosis disappeared at 3 months posttransplant with anticoagulants. Currently (12 months posttransplant), the patient has fully recovered, and the IPSS is still the same size. Conclusions. Based on our experience, liver allografts with IPSS can be accepted as potential liver allografts.

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Case Description

A 28-year-old man with end-stage liver disease secondary to Wilson disease was referred for LDLT. The patient’s features of decompensation included marked ascites and hepatic hydrothorax, which frequently required thoracentesis. The patient’s model for end-stage liver disease score was 18. His LDLT donor candidate was his 26-year-old, ABO blood type identical brother. Volumetry of the whole liver and graft was analyzed by three phases of a 1-mm slice, dynamic computed tomography (CT) scan and intervention planning software by MeVis Distant Services (Bremen, Germany). The estimated right lobe graft weight and graft-to-recipient body weight ratio was 713 g and 1.06%, respectively. The projected graft-to-recipient body weight of left lobe graft was 0.75. Because the estimated volume of his left lobe was inadequate, transplant using a right lobe graft was considered. An abdominal ultrasound (Figure 1A) and dynamic CT scan (Figure 1B) showed IPSS of 25 mm in diameter located at segment 6. Three-dimensional CT images clearly showed the communication between P6 and V6 (Figure 1C). Blood test results, including liver function tests were normal and blood ammonia concentration was upper limit of normal range. The cardiopulmonary assessment including echocardiographic evaluation showed no abnormalities, such as pulmonary hypertension. Pulse-dye densitometry (DDG-2001; Nihon Kohden, Tokyo, Japan) was used to measure blood indocyanine green (ICG) concentrations for the evaluation of the shunt ratio. The basic principle has been described in detail elsewhere.1 The elimination rate constant (ICG-K) was automatically calculated based on blood ICG concentrations. The calculated ICG-K value was 0.16, which
was within a normal range, suggesting that the shunt flow through the IPSS should be low enough to not induce hyperammonia leading to hepatic encephalopathy. Although the use of a liver graft with IPSS increased the risk for this recipient, we selected the right-lobe graft to decrease donor risk because IPSS in the remnant liver could cause hepatic encephalopathy, and large IPSS can even cause liver fibrosis in the long term. The remnant liver volume was estimated to be 42%. We decided that using this graft with an IPSS was the best option for this recipient who had no other potential living donors and a very low likelihood of receiving a brain-death donor organ in Japan.

Procurement of the right liver graft was performed using standard techniques without any complications related to IPSS. The actual graft weight and graft-to-body weight ratio were 720 g and 1.17%, respectively. After total hepatectomy, the hepatic vein and portal vein (PV) were reconstructed in our standard fashion. After reperfusion, a discolored lesion appeared on the liver surface at the site of IPSS (Figure 2A). Hepatic artery and biliary reconstruction were performed in our standard fashion. The PV pressure (PVP) was measured via the small jejunal vein branch with an 18-G central venous catheter during transplantation. PVP at the time of reperfusion of the liver graft was 25 mm Hg. However, after the completion of hepatic arterial reconstruction, PVP was 20 mm Hg. Because the recipient had splenic arterial aneurism, splenectomy was carried out during transplantation. After splenectomy, PVP decreased to 14 mm Hg, which is considered an acceptable portal pressure during an LDLT. The cold, warm ischemic, and total operative times were 36, 39, and 685 minutes, respectively. Blood loss was 9185 mL. Intraoperative portal venography performed at the end of transplantation demonstrated the expected aneurysmal communication between right posterior PV and right hepatic vein (Figure 2B). Color Doppler Sonogram of liver graft performed immediately after operation showed no increase in the size of the IPSS despite the elevation of portal pressure that occurred during the LDLT (Figure 3A). For posttransplant management, a standard immunosuppression protocol was applied. The immediate posttransplant course was uneventful. Serum aspartate aminotransferase, alanine aminotransferase, and international normalized ratio levels quickly returned to normal. Serum bilirubin and ammonia levels returned to normal on postoperative days 1 and 6, respectively. No asterixis or hepatic encephalopathy was present postoperatively. The patient was started on intravenous heparin for splenic vein thrombosis, which was shown by CT scheduled for follow-up of IPSS on postoperative day 7. Thrombosis was also found inside the IPSS with a CT scan on postoperative day 21 (Figure 3B). As expected with the systemic anticoagulation used to treat the splenic vein thrombosis, the thrombosis in the IPSS disappeared by 3 months after transplant. He was discharged home on postoperative day 38. Currently (12 months posttransplant), the patient has fully recovered with no episodes of hepatic encephalopathy, and the size of IPSS is unchanged (Figure 3C). His liver function test and blood ammonia concentration have been consistently normal.

**DISCUSSION**

Optimal use of liver grafts with extended criteria is a necessity in the era of organ shortages for transplantation. To the best of our knowledge, this is the first report of LDLT using...
a liver graft with a large-sized IPSS. In our case, LDLT was successfully performed, and adverse events related to IPSS in the graft never occurred.

An IPSS can develop in patients with cirrhosis, after trauma or rupture of a PV aneurysm, or can be congenital. Although the prevalence of congenital IPSS is unknown, it is increasingly recognized using ultrasound, dynamic CT scans, or magnetic resonance imaging as an incidental finding in asymptomatic patients.9

The clinical importance of IPSS is controversial. Whether patients exhibit symptoms of hepatic encephalopathy depends on the shunt ratio and the sensitivity of the brain to ammonia, as well as liver function. Cerebral tolerance and/or liver function for hepatotoxic substances gradually decrease with age and increase the patient’s risk for hepatic encephalopathy, particularly after middle age.10-13 Other pathophysiological changes that may have clinical relevance relate to the degree of shunting. A large fistula can theoretically cause adverse systemic hemodynamic effects or even cardiac failure with development of a hyperdynamic systemic circulation. The potential of longstanding diversion of portal flow away from the sinusoids to adversely affect the liver (eg, development of fibrosis) has been suggested.14 The small size of most intrahepatic portal venous shunts means that they are unlikely to have a significant clinical effect.

In the current case, the recipient developed considerable portal hypertension when transplanted. Little is known regarding whether IPSS in the graft is affected by portal hypertension and/or hepatic hemodynamic changes during liver transplantation and in the early phase after transplantation.15 Therefore, we were concerned that the IPSS might grow in size and its shunt ratio might increase after being

![Image of liver graft and intrahepatic portal venous shunt](image1)

**FIGURE 2.** Intraoperative findings of liver graft after reperfusion. A, Discolored lesion was shown on the surface of liver at the site of intrahepatic portal venous shunt. B, (arrow) Intraoperative portal venography performed at the end of surgery showing RPPV feeding intrahepatic portal venous shunt (arrowheads) that flows into RHV was shown. RPPV, right posterior portal vein.

![Image of posttransplant images](image2)

**FIGURE 3.** Posttransplant images in the recipient (A) Color Doppler Sonogram of liver graft in the recipient immediately after the transplantation showed communicating color signals entering the lesion from PV and draining into RH-V was shown. The size of intrahepatic portal venous shunt (arrowheads) remained same. B, An abdominal contrast-enhanced CT scan of recipient at postoperative day 21. Thrombosis inside the intrahepatic portal venous shunt was demonstrated. C, An abdominal Contrast-enhanced CT scan of recipient at 12 months after LDLT. The intrahepatic portal venous shunt (arrow) remained the same in size. PV, portal vein.
transplanted to the recipient. Coiling for IPSS might be an option for treatment when a patient develops any complications related to IPSS. Fortunately, there was no change in size of the IPSS during transplantation and posttransplant in our case, and the patient experienced no episodes related to IPSS, including hepatic encephalopathy.

This case demonstrates the fine balance between the risk of portal hyperperfusion and portal hypoperfusion, both of which can have disastrous consequences after LDLT. Splenectomy decreases portal flow, and therefore decreased the risk of both portal hyper perfusion of the graft and the risk of worsening shunt through the IPSS. At the same time, the decreased portal flow carried by the splenectomy likely contributed to the SV and IPSS thrombosis. It is well established that splenectomy carries a significant risk of PV thrombosis due to the reduction in portal flow, so it should not be performed in every LDLT recipient. The splenic artery aneurysm was the indication for splenectomy in this patient. If splenectomy had not been performed, this patient’s outcome may have been different, as there would have been much greater flow through the IPSS. Another form of graft inflow modification might have been required.

From the long-term perspective, we expect that any possibility of an increase in size or untoward effect of IPSS would diminish as portal hypertension resolves. However, the patient should be carefully monitored because little is known about the long-term outcome of IPSS in the transplanted liver. Treatment, including dietary control, embolization, or surgical removal of the shunt, should be considered if this patient develops symptoms related to the IPSS.16,17

Based on our experience, liver graft with IPSS from brain-death donor can be usable unless it is too large. When the donor surgeon identified an IPSS during the procurement, Color Doppler ultrasound is appropriate to make differential diagnosis from any tumors. It is unknown whether IPSS in the remnant liver could cause untoward effects on the donor, but theoretically, it would increase the risk of hepatic encephalopathy. Thus, we would select the right liver graft to avoid an increasing concern about the safety of the donor, even if estimated volume of left liver was large enough as graft.

In our case, we could successfully select right lobe graft with sufficient graft volume. However, in a certain donor-recipient combinations, there is a possibility such as a small-for-size (SFS) left lobe graft with IPSS. If so, we suggest either option: (a) use SFS left lobe graft with IPSS, although there is a risk of SFS syndrome in recipient or (b) find another donor. Most importantly, we need to limit the risk we expose the donor to.

In conclusion, a liver with IPSS can be considered as a potential liver graft based on our experience. However, careful graft selection is important because large IPSS may affect a recipient’s postoperative course. Even though this recipient has an IPSS that is stable in size, normal liver function and no hepatic encephalopathy, long-term follow-up of any graft containing an IPSS is probably warranted.

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