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CASE REPORT

Left lobe living donor liver transplantation in an adult patient with situs inversus: technical considerations

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Summary

Situs inversus (SI) is a rare congenital disorder involving a complete mirror image of the thoracic and abdominal organs. Living donor liver transplantation (LDLT) in SI cases poses particular challenges on account of its technical complexity, and only a few cases have been reported. Here, we present an adult with SI who was managed successfully by LDLT using a left lobe graft. Some technical modifications, including triangulated anastomosis of the hepatic vein, were required but no vascular graft was necessary. Graft function and vascular integrity were excellent throughout the postoperative course, although sepsis secondary to wound infection with methicillin-resistant *Staphylococcus aureus* developed. In conclusion, LDLT using a left lobe graft is a feasible procedure for patients with SI, even for adults. Therefore, this condition, while rare, should not be a contraindication for LDLT. Meticulous preoperative simulation and planning of the vascular reconstruction are important steps in LDLT for this rare anomaly.

Introduction

Situs inversus (SI) totalis is a rare congenital anatomic variant involving a complete mirror image of the thoracic and abdominal viscera or the anomaly may be limited to abdominal organs. The incidence of SI is estimated to be <0.005% of the general population [1]. Biliary atresia, the most common indication for liver transplantation, can be associated with SI and up to 28% of SI patients are affected [2,3].

Although SI was once an absolute contraindication for liver transplantation on account of the complexity of the vascular malformations, such as absence of the inferior vena cava (IVC), preduodenal portal vein (PV) and aberrant hepatic artery (HA) anatomy, several successful cases using deceased donor whole liver [4–6] or split and reduced liver grafts [7] have been reported. However, most of these procedures were performed in pediatric patients who are expected to exhibit less graft displacement and hepatic venous torsion on account of their smaller abdominal cavity, even in cases where split and reduced grafts are used. On the contrary, in adults, a large empty space can be formed in the left upper quadrant (LUQ) following recipient hepatectomy, thereby predisposing the patients to lateral displacement with superolateral graft rotation and torsion of the hepatic venous pedicle. Nonetheless, a few successful cases have been reported in adult patients with SI to date.

Living donor liver transplantation (LDLT) is currently a legitimate and established alternative to deceased donor liver transplantation, especially in countries such as Japan and other Asian countries, where deceased donor grafts are hardly available and living donor grafts are the only realistic option. Various technical innovations in LDLT have overcome the associated technical difficulties and helped to create stable results. However, LDLT for patients with SI should pose further technical challenges on account of the limitation of the vascular length as well as the availability of vascular grafts for interposition and the undetermined positioning of partial grafts.

In the present report, we describe a successful case of LDLT using a left lobe graft in an adult with SI and discuss our technical modifications for this particular case. This is the first case report of LDLT using a left lobe graft in an adult with SI.

Case report

A 19-year-old female patient was referred to our outpatient clinic for end-stage liver cirrhosis secondary to hepatitis C infection. In her history, we found that she had undergone closure of a ventricular septum defect at 5 months of age, and SI totalis with dextrocardia was diagnosed. She received a blood transfusion at that time and was followed up by a pediatrician until she reached 18 years of age. She had been well until the summer of 2005, when she developed peripheral edema and pancytopenia. In October 2005, decompensated liver cirrhosis on account of hepatitis C was diagnosed at the hospital and conservative treatment was rendered. However, her liver function continued to deteriorate through 2006, and she was referred to us for the possibility of liver transplantation. The pretransplant work-up, including a three-dimensional abdominal CT scan, confirmed SI totalis and left-sided cirrhotic liver with minimal ascites, and moderate splenomegaly (Fig. 1a). No other anatomical anomalies, such as preduodenal PV, polysplenia, midgut malrotation, anomalous blood supply to the liver or discontinuous suprarenal IVC, were found. Her liver function was Child Class C with a Model for End-Stage Liver Disease (MELD) score of 9. She was accepted for LDLT by our institutional Liver Transplantation Subcommittee in May 2006.

The donor was her 53-year-old father, whose blood type was identical to that of the recipient. The preoperative imaging studies are shown in Fig. 2. The hepatic venous anatomy was normal. The PV anatomy showed trifurcation, in which the right anterior and posterior PVs were independently derived from the main PV trunk (Fig. 2a). The arterial anatomy was normal, and the artery to segment 4 branched from the right HA (Fig. 2b). The estimated graft volume (GV) of the full left lobe measured by CT volumetric analysis was 428 ml, which corresponded to 43.8% of the standard liver volume (SLV) of the recipient. Therefore, the left lobe including the middle hepatic vein (HV) was chosen as a graft according to our selection criteria [8]. The caudate lobe was not procured on account of the anomalous vascular anatomy, where the PV branch to the caudate lobe derived from the main PV trunk (Fig. 2a). A drip-infusion cholangio-CT scan revealed that the posterior right hepatic duct drained into the left hepatic duct. Mild fatty liver was suspected by an abdominal ultrasound but a biopsy was not performed.

The donor procedure was performed using a standard method, which is described elsewhere [9]. The PV was found to be trifurcated as expected, and the left PV was divided distal to the origin of the anterior right portal branch. The caudate lobe and the portal venous branches to the caudate lobe were preserved in the recipient. The actuarial allograft weight was 370 g (GV/SLV, 37.9%), and the graft contained two HAs (left HA and HA to segment 4). The posterior wall of the common orifice of the

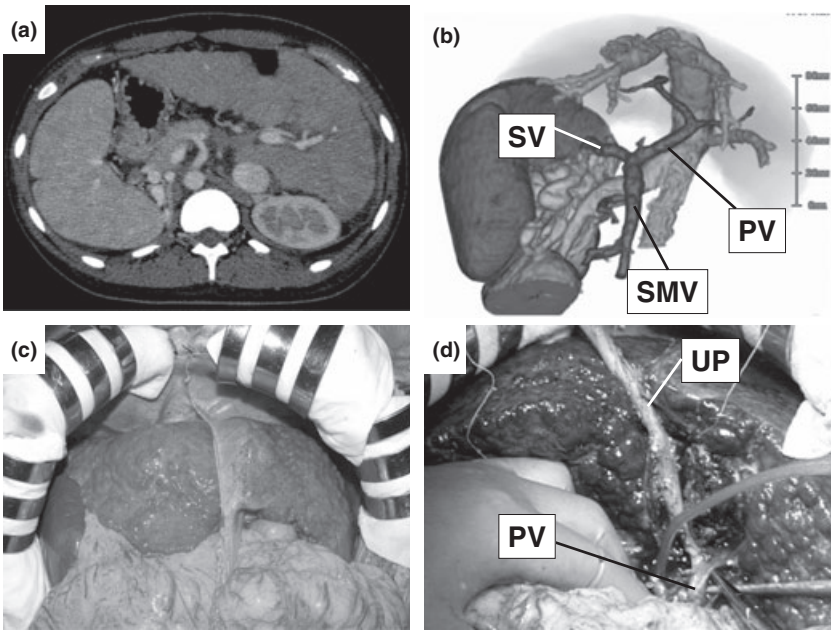

Figure 1 Preoperative imaging and operative findings of the recipient. (a) Abdominal CT scan. (b) Three-dimensional image of the portal vein and hepatic veins. (c) Intra-operative appearance of situs inversus with a cirrhotic liver. (d) The anatomical left portal vein is dissected high up to the umbilical portion to obtain a longer portal vein. SV, splenic vein; SMV, superior mesenteric vein; PV, portal vein; UP, umbilical portion of the portal vein.

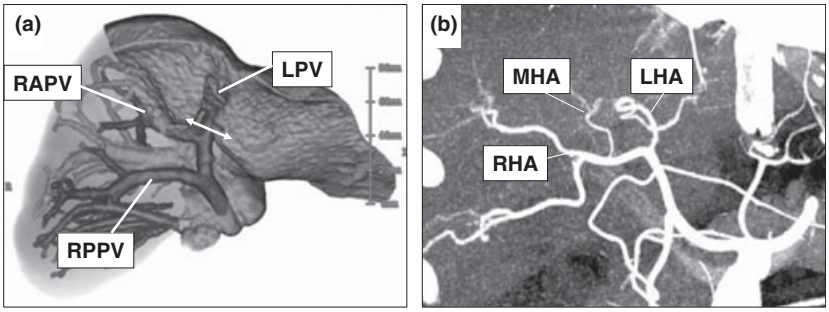

Figure 2 Preoperative three-dimensional CT imaging of the donor vessels. (a) The right anterior and posterior portal veins are independently derived from the main portal vein trunk. The portal vein of the graft is divided at a distal site to the right anterior portal vein takeoff (double arrow). (b) The hepatic artery to segment 4 (middle hepatic artery) is derived from the right hepatic artery. RAPV, right anterior portal vein; RPPV, right posterior portal vein; LPV, left portal vein; RHA, right hepatic artery; MHA, middle hepatic artery; LHA, left hepatic artery.

middle and left HVs was cut longitudinally at the back table in order to make a triangulated and wide orifice ($30 \times 20 \times 20$ mm).

In the recipient, SI was evident on entering the abdominal cavity and showed a markedly cirrhotic left-sided liver with a moderately enlarged spleen in the right upper quadrant (Fig. 1c). In the hepatic hilum, the anatomic left and right HAs were freed as much as possible, ligated with 3–0 silk ties and divided. The common hepatic duct was ligated with 2–0 silk ties and divided. The anatomical right hepatic lobe was mobilized from the left diaphragm and the retroperitoneum. The PV was dissected free beyond the bifurcation. The anatomical left PV was further dissected distally to the round ligament by ligating all the branches to the anatomical segment 2–3 and segment 4 (Fig. 1d) to facilitate later PV anastomosis.

A total hepatectomy was then performed by dividing the anatomical left and right PVs, which were left as long as possible. The common orifice of the anatomical middle and left HVs was further cut longitudinally to the inferior angle of the orifice, creating a wide and triangulated orifice in the recipient in order to make a wide anastomosis and prevent torsion of the anastomotic site. A veno-venous bypass was not used.

The left lobe graft was placed in the midline position and rotated slightly to the left compared to the normal anastomosis in order to align the HV (Fig. 3a). Next, the triangulated HV orifice of the graft was anastomosed to the triangulated anatomical middle/left HV conduit of the recipient in an end-to-side fashion using 5–0 PDS II (Johnson & Johnson Inc., Tokyo, Japan) running sutures. The left PV of the graft, which was shorter than normal, was easily anastomosed to the main PV trunk of the recipient in an end-to-end fashion using 6–0 PDS II running sutures with a growth factor. No tension or torsion was observed at either of the PV and HV anastomotic sites. Immediately after completing the anastomoses, the clamp on the IVC was released, followed by removal of the PV clamp. Reperfusion of the graft was prompt and smooth. The anhepatic, cold and warm ischemic times were 64, 65 and 36 min, respectively. After obtaining hemostasis, arterial reconstructions between the graft middle HA and the recipient anatomical right HA as well as between the graft left HA and the recipient anatomical left HA were

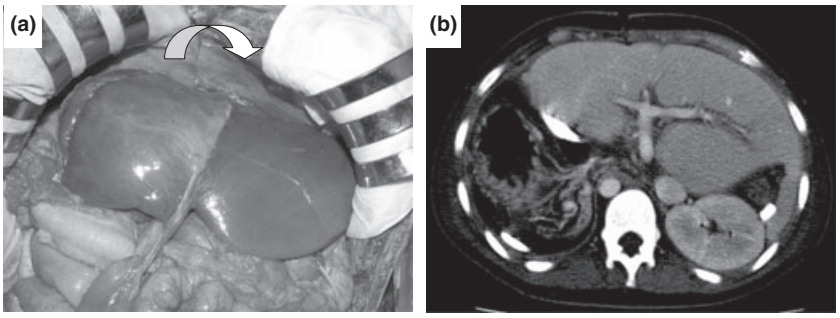

Figure 3 Intra- and postoperative views of the graft. (a) Intra-operative appearance of the graft positioning demonstrating slightly left-sided and clockwise rotation of the graft. (b) An abdominal CT scan at postoperative day 28 reveals a well-accommodated and regenerated graft without any torsion of the anastomosis.

performed under a microscope using interrupted 8–0 Prolene sutures (Johnson & Johnson Inc.). An intra-operative Doppler ultrasound confirmed adequate flow in the PV, HA, and HV. A splenectomy was performed in a standard manner by dividing the surrounding attachment using electrocautery and a LigaSure™ system (Valley Laboratories, Boulder, CO, USA). The splenic pedicle was divided using a stapler (ENDO GIA® Universal; Tyco Healthcare Japan, Tokyo, Japan).

Biliary reconstruction was performed by hepaticojejunostomy over a 2.0-mm tube (RTBD Tube; Sumitomo Bakelite Co. Ltd., Tokyo, Japan) using interrupted 6–0 PDS II sutures. The portal pressure before abdominal closure was 18 mmHg. At the last step, the falciform ligaments were reapproximated to the abdominal wall to prevent torsion of the HV anastomosis and tension on the PV anastomosis.

After implantation of the graft, the right renal vein (RV) was dissected and exposed. A large spleno-renal shunt was found to drain into the right RV, which was carefully dissected, controlled with a tape and then doubly ligated with 2–0 silk ties.

The operative time was 14 h 20 min and the estimated blood loss was 4330 ml for which 14 units of red blood cells, 20 units of fresh frozen plasma and 30 units of platelets were transfused. The immunosuppressive protocol was tacrolimus-based with steroids. A CT scan taken on postoperative day (POD) 28 revealed a well-positioned and regenerated left lobe graft without either kinking or stenosis of the anastomosed vessels (Fig. 3b).

However, the postoperative course of the patient was complicated by the development of sepsis secondary to wound infection with *methicillin-resistant Staphylococcus aureus* at the abdominal wall, for which a re-laparotomy for peritoneal lavage and drainage was necessary on POD 7. The patient subsequently developed wound infection and dehiscence, which resulted in an intestinal fistula within the abdominal wall. The fistula had closed spontaneously by POD 45. The graft function continued to be excellent throughout the postoperative course and the patient was discharged from the hospital on POD 50. Currently, at 1 year after the LDLT, she is enjoying a normal life with normal liver function tests and undetectable hepatitis C virus RNA, as evaluated by polymerase chain reaction, following a combination therapy of weekly PEG-interferon alpha-2b (PEG-Intron®; Schering-Plough K.K., Tokyo, Japan) and daily ribavirin (Rebetol®; Schering-Plough K.K.).

Discussion

The first successful case of liver transplantation in a patient with SI was reported by Raynor *et al.* [9]. Since then, several cases of SI who underwent deceased donor liver transplantation (DDLT) using whole liver [4–6] or reduced-size liver [7] grafts have been reported. Watson *et al.* [10] described seven patients with SI abdominis and one patient with SI totalis who underwent liver transplantation. Seven of these patients had additional abnormalities associated with polysplenia-biliary atresia syndrome. After follow-up periods of 7 months and 5 years, all the patients remained alive. Two of the patients required re-transplantation within the first 3 weeks for primary nonfunction and thrombotic infarction, respectively. Delayed abdominal wall closure was necessary in two patients and all eight patients required a modification of the 'piggy-back' technique of suprahepatic vena cava anastomosis to overcome recipient venous anomalies. Biliary drainage by Roux-en-Y choledochojejunostomy was specifically performed for all of the patients. The authors concluded that, although the procedure is technically challenging, SI is not a contraindication for liver transplantation and the patients should be expected to make a full recovery. Farmer *et al.* [11] summarized their experiences of DDLT in six cases of SI. In their series, the liver graft was placed in a midline position and transplanted. Venous continuity was achieved by donor suprahepatic IVC to the recipient hepatic common orifice and direct end-to-end portal anastomoses. The donor infrahepatic IVC was oversewn. Arterial continuity was restored using either a direct branch-patch anastomosis (3/6) or a supraceliac aortic interposition graft (3/6). The authors concluded that SI in liver recipients requires operative technical modifications, but does not change the outcome.

Living donor liver transplantation has become a standard option for treating end-stage liver disease in pediatric patients as well as in adults. More than 7000 such procedures have been performed worldwide [12]. However, there have only been a few cases of SI reported in the setting of LDLT, especially for adult patients. The potential difficulty associated with performing LDLT for such cases is the vascular reconstruction. Due its nature, a living donor graft has short vessels and the availability of vascular grafts from the donor is very limited, especially for adult patients. Furthermore, the recipient's vessels may be anomalous and the orientation of the hepatic vessels is completely reversed to the opposite side relative to that of the normal counterparts, thereby complicating the procedure. Maggard *et al.* [13] first reported their experience of LDLT using a left lateral segment graft for a pediatric patient with biliary atresia and SI. They claimed that no extraordinary procedures were required for the case and concluded that SI should not be considered as a barrier to the use of split grafts such as LDLT. Furthermore, Matsubara *et al.* [14] described a series of four pediatric patients with SI who underwent LDLT using a left lateral segment

graft ($n = 3$) and a monosegment graft ($n = 1$). Two of these patients developed PV thrombosis after the LDLT, which was probably on account of the recipients' sclerotic and small PVs that required interposition grafts, rather than the technical failure related to the presence of SI. These data suggest that vascular grafts may not be required for vascular anastomoses, even in the setting of LDLT for small children. Nevertheless, graft positioning does not seem to be a major problem for small pediatric patients because the LUQ can be a free space for fitting the left lobe grafts, including a lateral segment graft. Regarding adult cases, we have only found two reports describing right lobe LDLT for patients with SI [15,16]. In one case, a right lobe graft was positioned at the midline directly over the IVC [15]. The right HV of the graft was anastomosed to the recipient HV, which was located slightly more anteriorly than would be expected. The donor right PV was connected to the recipient main PV. Arterial reconstruction was performed between the donor right HA and the recipient right HA. Biliary reconstruction was carried out with a Roux-en-Y hepaticojejunostomy. The second case was LDLT for a patient with SI using the right lobe of a donor with SI (16). This case should not pose any particular anatomical challenges, except for the realization of the complete mirror image of the structures. In the present case, the donor left PV was supposed to be significantly short because the left PV had to be cut distally to the right anterior PV takeoff (Fig. 2a). Therefore, we dissected the recipient anatomical left PV as distally as possible high up to the umbilical portion, thereby preserving a long PV for a secure anastomosis (Fig. 1d). However, the PV anastomosis was easily performed without any tension or difficulty. Therefore, we currently consider that no special techniques are necessary for PV anastomosis for left lobe grafts.

Regarding the anastomosis method for the HV, the optimal procedure for this reconstruction remains controversial. The size of the anastomotic orifice, orientation of the vessels and position of the graft are important determinants for maintaining the patency of the reconstructed HV. We utilized a triangulated suture method that we have previously used for pediatric LDLT. The initial paper by Emond *et al.* [17] reported the efficacy of this technique, especially for small children. We utilized this technique because we believe it to be the most appropriate technique for avoiding torsion and maximally increasing the venous outflow of the graft, thereby preventing outflow blockage of the HV anastomosis on account of rotation of the graft to the left side. Furthermore, we expected positional changes of the graft during the regeneration of the liver parenchyma or the accommodation of the graft in the abdominal cavity, especially in the LUQ. At 1 year after the transplant, a routine Doppler ultrasound and CT scan revealed a patent and wide-open HV anastomosis.

In conclusion, a left lobe LDLT was found to be feasible for an adult patient with SI after some technical modifications. Appropriate preoperative planning is essential and should be individualized, especially in the setting of LDLT. Therefore, technically demanding procedures such as this should only be performed by an experienced team. Further accumulation of cases is warranted for this type of rare anomaly.

Authorship

YS collected the data and wrote the paper. YS, MM, AT and TI were responsible for treatment of the patient. YY, NH, SI, HU, TY and YM contributed reviewing the paper.

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