Liver Transplantation Technique from A Cadaveric Donor with Situs Inversus Totalis. Review and a New Case

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1. Abstract
Because of the shortage of organs available for transplantation, bodies that may be valid, such as those coming from donors with situs inversus totalis, should not be ruled out. This is a rare anatomical abnormality that results in dextrocardia, mirror imaging of normal abdominal organs, and other congenital abnormalities. These organs are technical challenges for surgeons during liver transplantation because of problems associated with the vascular anatomy and graft positioning. We herein describe the technique used in a case and review the limited number of published reports.

2. Introduction
Donors with *situs inversus totalis* (SIT) are considered to be a technical challenge in Liver Transplantation (LT). This is a rare congenital anomaly with a prevalence rate of 1:4,000 to 1:20,000 births and an incidence rate of 0.01% worldwide. All thoracoabdominal and retroperitoneal organs are symmetrically positioned on the midline, a condition often termed mirror-image reversal [1]. Although SIT in recipients was historically considered to be an absolute contraindication to LT, several surgical solutions have been proposed and successfully performed, and SIT is no longer considered to be a contraindication in such cases [2, 3].

However, because of the infrequent occurrence and complexity of this clinical situation, there is little experience with the use of cadaveric donors with SIT. Some transplant groups still rule out these grafts despite the low number of donors and the high number of recipients on waiting lists [4].

A review of published articles revealed eight cases of deceased donors with SIT whose grafts were successfully implanted and in which different technical solutions were proposed [5-12].

The aim of this article is to review all the current experience and analyze it against a new case.

3. Case Description and Methods
3.1. Donor Information and Surgical Technique
The deceased donor was a 41-year-old female (blood type A; weight, 65 kg; height, 165 cm) with a history of dyslipidemia and subclinical hypothyroidism. The etiology of death was cerebral hemorrhage secondary to rupture of an aneurysm of the middle cerebral artery. In the initial assessment of different organs to be transplanted, a chest X-ray revealed dextrocardia and an abdominal ultrasound confirmed SIT. Due to technical difficulties, the cardiac procurement team discarded the heart and procured only the valves.

SIT was observed, with the Inferior Vena Cava (IVC) and liver on the left side and the aorta, spleen, stomach, and heart on the right side (Figure 1a). A left hepatic artery from the left gastric artery was also observed. The abdominal organs were flushed with Celsior solution and procured with the Starlz standard procurement technique. The kidneys and lungs were also procured and successfully implanted.
3.2. Back Table Surgery

Standard back table surgery was performed. The surrounding fatty tissue and diaphragm muscle attached to the liver surface were removed, and the phrenic veins were ligated. The vascular structures were dissected as far as the hilum and prepared for anastomosis with the recipient structures. The left hepatic artery stemming from the left gastric artery was preserved. Cholecystectomy was performed and the bile duct was flushed with 150 mL of Celsior solution. The liver was preserved at 4°C until implantation (Figure 1b, c).

3.3. Recipient Information and Surgical Technique

The recipient was a 52-year-old male with hepatitis C virus cirrhosis (Child-Pugh score, C10; Model for End-stage Liver Disease score, 22). He had severe portal hypertension, splenomegaly (15 cm), and ascites.

A right subcostal incision was created and extended to the left. Approximately 9.5 L of ascites were suctioned. The hepatectomy was performed following the standard procedure with preservation of the IVC (piggy-back technique) and no setbacks. The right, middle, and left hepatic veins were joined to create a common ostium in the hepatic veins (Figure 1d).

A double running suture with 3/0 polypropylene was placed in the IVC stump, which was kept open to wash out the liver just before reperfusion.
The right liver lobe was positioned on the left side of the recipient and the left liver lobe was placed into the recipient liver fossa. Portal vein anastomosis was performed with a 5/0 polypropylene running suture in the posterior face and an interrupted suture pattern in the anterior face. Before reperfusion, a wash-out with recipient blood was performed through the infrahepatic IVC stump. The stump was closed and the clamps were removed from the hepatic and portal veins.

The cold and warm ischemia times were 300 and 55 minutes, respectively. The duration of time until artery anastomosis was 45 minutes. The artery anastomosis was performed between a patch created with the right and left hepatic arteries of the recipient and the common hepatic artery of the donor (running suture of 7/0 polypropylene). No problems associated with the length of the vessels were observed.

Biliary reconstruction was performed by end-to-end anastomosis between the recipient common hepatic duct (5 mm) and the donor common bile duct with a running suture of 6/0 monofilament polyglyconate in the posterior face and an interrupted suture pattern in the anterior face. A rubber T tube was then placed (Figure 2b).

The procedure lasted 270 minutes. (Figure 2c) illustrates the final position of the transplanted graft. The patient had excellent postoperative graft function. The T tube was removed 2 months after transplantation. Six months later, a CT scan showed no vascular problems (Figure 2d).

**Figure 2a:** Vascular anastomosis of the hepatic veins

**Figure 2b:** T-tube cholangiography

**Figure 2c:** Implanted liver

**Figure 2d:** Computed tomography scan at postoperative month 6

4. Discussion

SIT is characterized by dextrocardia and mirror-image distribution of the abdominal organs. It may coexist with other malformations such as an underdeveloped portal vein and IVC, polysplenia syndrome, and congenital biliary atresia [1]. Because the incidence of this syndrome is very low, there is little experience in the use of these organs. Thus, these types of donors are often ruled out as effective donors [4].
We found only eight reported cases of LT from deceased donors with SIT [5-12] (Table 1). There is no consensus on the strategy for implantation of these organs into the recipient, and different modifications of transplantation techniques to overcome the problems associated with the vascular anatomy and graft positioning have been described.

**Table 1: Features of published cases involving deceased donors with situs inversus totalis**

<table>
<thead>
<tr>
<th>Author</th>
<th>Technique (hepatectomy and implant)</th>
<th>Venous outflow</th>
<th>Biliary reconstruction</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asfar et al. (1995)</td>
<td>• Piggy-back hepatectomy • Heterotopic 90° rotation</td>
<td>End-to-side IVC infrahepatic</td>
<td>Hepatico-jejunostomy</td>
<td>Dead</td>
</tr>
<tr>
<td></td>
<td></td>
<td>donor-IVC recipient</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Herrera et al. (1996)</td>
<td>• Piggy-back hepatectomy • Orthotopic</td>
<td>IVC donor-right hepatic</td>
<td>Choledocho-choledochostomy</td>
<td>Alive (2 years)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>vein recipient</td>
<td>(with T tube)</td>
<td></td>
</tr>
<tr>
<td>Braun (1998)</td>
<td>• Piggy-back hepatectomy • Orthotopic</td>
<td>End-to-side IVC</td>
<td>Choledocho-choledochostomy</td>
<td>Alive (17 months)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>donor-IVC recipient</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pomposelli et al. (2007)</td>
<td>• Piggy-back hepatectomy • Heterotopic 180° rotation along axis</td>
<td>IVC donor-right middle</td>
<td>Hepatico-jejunostomy</td>
<td>Not available</td>
</tr>
<tr>
<td></td>
<td></td>
<td>hepatic veins recipient</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dou (2010)</td>
<td>• Classical • hepatectomy • 15° clockwise rotation</td>
<td>End-to-end IVC donor-IVC recipient</td>
<td>Choledocho-choledochostomy</td>
<td>Alive (10 months)</td>
</tr>
<tr>
<td>Sun et al. (2013)</td>
<td>• Piggy-back hepatectomy • Orthotropic (minimum rotation)</td>
<td>IVC donor-left middle hepatic</td>
<td>Choledocho-choledochostomy</td>
<td>Alive (36 months)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>veins recipient</td>
<td></td>
<td></td>
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<tr>
<td>Manzia et al (2014)</td>
<td>• Piggy-back hepatectomy • Orthotopic</td>
<td>Side-to-side IVC donor-IVC recipient</td>
<td>Choledocho-choledochostomy</td>
<td>Alive (6 months)</td>
</tr>
<tr>
<td>Lópe-Mendújar (2014)</td>
<td>• Piggy-back hepatectomy • Orthotopic</td>
<td>End-to-end IVC donor-IVC recipient</td>
<td>Choledocho-choledochostomy</td>
<td>Alive (8 years)</td>
</tr>
<tr>
<td>Reimondez et al. (2019)</td>
<td>• Piggy-back hepatectomy • Orthotopic</td>
<td>End-to-end IVC donor-IVC</td>
<td>Hepatico-jejunostomy</td>
<td>Not available</td>
</tr>
<tr>
<td></td>
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<td>IVC</td>
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</table>

We have herein added a new case and provided a step-by-step description of a successful liver implantation technique that could be extrapolated to other cases. Since the first liver transplant, the details of many surgical techniques have been improved and adopted to achieve a better outcome. The piggy-back technique is currently the first choice for hepatectomy and is feasible in most cases. In the piggy-back technique, in contrast to the conventional technique, the native IVC remains in situ during hepatectomy. Several studies, including a randomized trial comparing the results of both techniques, have shown a shorter operation time, warm ischemia time, and anhepatic phase as well as reduced blood loss and procedure costs in association with the piggy-back technique [13]. For this reason our standard technique is piggy-back hepatectomy and that is why we also did it in this case with donor SIT as in other similar cases described in the literature [5-8, 10-12] but not always [9].

The “Achilles heel” of the piggy-back technique is venous outflow reconstruction to avoid caval stenosis or thrombosis, major graft congestion, acute or chronic Budd-Chiari syndrome and torsion or stenosis of the anastomosis, with a reported frequency of 2.5-4.6% [14]. There is no clear evidence of the advantages of one technique over another. In the prospective study by Hesse et al. they concluded that in terms of outflow complications, the classical piggy-back technique is better than side-to-side anastomosis. In our group, the usual outflow reconstruction technique is the classical piggy-back between the end of the suprahepatic IVC of the graft and the common orifice created by the junction of the three hepatic veins and it was the technique of choice even with the graft from the donor SIT.

The most important handicap when a liver graft coming from a donor with SIT is to fit it to the hepatic fossa in the recipient. Some authors prefer to rotate the graft to better adapt it to the cavity; however, this requires a number of changes to the classical piggy-back technique. Asfar et al. [5] described 90° counterclockwise on its coronal plane rotation of the liver, which means that the right lobe is placed in the right hepatic fossa and the left lobe goes into the paracolic gutter. Using this technique, the suprahepatic IVC is ligated and an end-to-side anastomosis is performed between the infrahepatic IVC and the recipient vena cava.

Pomposelli et al. [8] reported a liver graft rotated backward 180° along the IVC axis, which allowed for a large portion of the left side of the liver to be located in the abdominal cavity. Furthermore, the suprahepatic vena cava of the graft was directly anastomosed to the junction of the middle and right hepatic veins; no cavoplasty or re-
medial procedure was required.

Herrera et al. [6] and Sun et al. [10] also used the piggy-back technique and implanted an entire liver graft without rotation. Herrera et al. [6] performed an anastomosis of the suprahepatic IVC from the donor to the right hepatic vein of the recipient, and Sun et al. [10] involved the junction of the left and middle hepatic veins by broadening the ostium to the IVC of the recipient. Braun [7] performed an end-to-side anastomosis between the donor suprahepatic IVC and recipient vena cava.

In our case, we chose the standard piggy-back technique with anastomosis to the join of the three hepatic veins and implanted the entire liver graft without rotation. The exact position of the graft was determined by perfect alignment between the IVC of the liver graft and the ostium of the three hepatic veins of the recipient; we usually perform a patch with the three veins with good outflow results, as reported by other groups [14]. A mandatory premise is to have enough space so to avoid any problems derived from a lack of space, we recommend allocating these grafts to recipients with ascites and greater abdominal perimeters, as we did in the present case. In our opinion these grafts should be considered as extraordinary and be allocated in a more flexible way to recipients with the above mentioned features. Arterial and portal reconstruction was not a problem in any of the published cases regardless of whether the graft was implanted with rotation. Our recommendation is to perform end-to-end anastomosis as usual, without the use of vascular grafts, assuring that the anastomosis is long enough in both the donor and the recipient.

Various techniques have been described for biliary reconstruction. Choledochocholedochostomy is generally preferred because it creates physiological bile flow to the duodenum by preserving the function of the sphincter of Oddi, it allows endoscopic diagnostics and interventions, and it avoids intestinal contamination of the bile ducts. Since hepaticojejunostomy is mandatory when the graft is rotated [5, 7], to avoid it in this case with the liver implanted without rotation, we used a T tube based on a previous demonstration of the benefits of its use [16].

To prevent a kinking that could compromise the outflow, we consider it very important to fix the graft to the diaphragm and falciform ligament and we also filled the liver fossa of the recipient with his omentum, and the right colon and implanted liver were attached to the diaphragm through the falciform ligament.

In conclusion, although the scarce successful experience, probably the only published, the use of grafts from SIT donors should be considered in appropriate and selected recipients.

References