

# Liver transplantation in patients with *situs inversus*

Todo S, Hall R, Tzakis A, Starzl TE. Liver transplantation in patients with *situs inversus*.

Clin Transplantation 1990; 4: 5-8.

**Abstract:** Two patients with *situs inversus* and biliary atresia were treated with hepatic transplantation, one with an auxiliary liver and the other with an orthotopic graft which was placed using a piggy-back technique. Both transplants functioned well initially. The auxiliary liver was rejected after 1 ½ months, and the patient died after an attempt at retransplantation many months later. The recipient of the orthotopic liver has perfect liver function 10 months postoperatively.

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**Key words:** liver Tx – *situs inversus* – biliary atresia

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Accepted for publication 9 August 1989

In patients with biliary atresia, other significant anomalies are present in 12 to 27% of cases (1, 2). One such anomaly is *situs inversus*, a condition which may not be rare in the biliary atresia patient population. During the 25 years of our liver transplantation program, more than a dozen patients have been referred to us with both conditions. Almost all of these children were rejected as candidates. However, one such patient was treated with an auxiliary liver transplantation and the other with a modified orthotopic liver transplantation. In both cases, the technical problems were manageable enough to warrant further trials.

## Case Reports

### Case 1

A Caucasian child was born on 27 July 1979 and had laparotomy 2 months later. *Situs inversus* with polysplenia, a bilobed liver, and a preduodenal portal vein were found. Kasai portoenterostomy was performed with externalization of the mid portion of a 45 cm Roux limb of jejunum. There was bile flow and clearing of jaundice, but after taking down the stoma at the age of 8 months, cholangitis developed which persisted despite operative revisions. By 13 April 1983, when auxiliary liver transplantation was performed at the age of 3-2/3 yr, she had developmental retardation (height < 5%), multiple excoriations from pruritis,

splenomegaly, ascites, jaundice (Bilirubin 11.0 mg%) and coagulopathy.

The donor liver, from a 9-month-old infant, had a left hepatic artery coming from the celiac axis and a right hepatic artery from the superior mesenteric artery. A large Carrel patch containing the take-off of both parent vessels was sewed to a short free graft of donor thoracic aorta and this conduit was anastomosed to the terminal recipient aorta after reflecting the colon (Fig. 1).

The graft infrahepatic vena cava was anastomosed end-to-side to the terminal vena cava of the recipient, but because of inadequate length, the recipient terminal vena cava was ligated and divided. This allowed the vena cava to swing over into an appropriate location in the right paravertebral gutter (Fig. 1). The donor suprahepatic vena cava was ligated. Portal inflow was established by anastomosing the donor superior mesenteric vein end-to-side to a large recipient meandering venous collateral (Fig. 1) which previously had been identified by angiography in the right side of the abdomen (Fig. 2). Biliary reconstruction was via cholechojejunostomy to a new 45 cm Roux-limb. Two spleens of medium size in the right upper abdomen were removed. The orientation of the new liver is shown in Fig. 1.

Except for a bout of atelectasis, recovery was uncomplicated and the child was sent home 30 d after transplantation with normal liver function. Eight days later, she was readmitted for cellular rejection. Bilirubin was 2.5 mg%, SGOT 505 IU/l, SGPT 770 IU/l, and alkaline phosphatase

Supported by Research Grants from the Veterans Administration and Project Grant No. DK 29961 from the National Institutes of Health, Bethesda, Maryland.

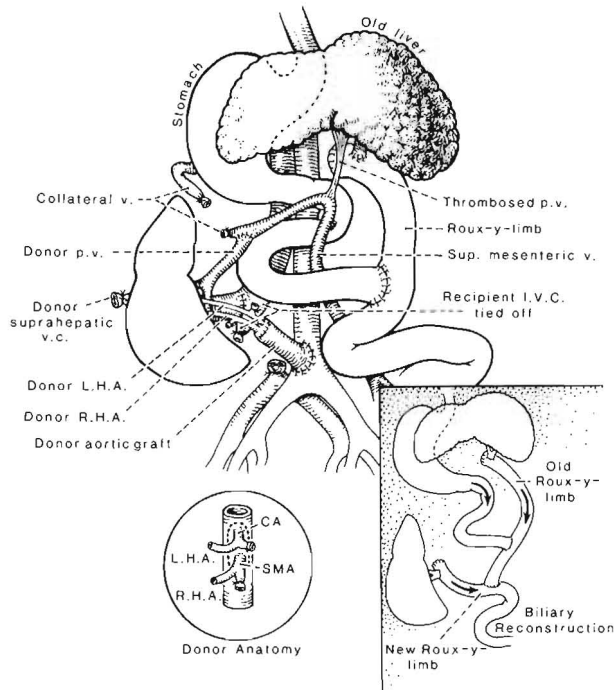


Fig. 1. Technique of auxiliary liver transplantation in Case 1. Note the aortic conduit used to arterialize the graft, and the unusual means of providing a portal venous inflow.

965 IU/l. Angiographic studies proved that the principal hepatic artery from the celiac axis was open although the branch from the superior mesenteric artery was clotted (Fig. 3). However, the distal arterial box in this portion of the right lobe filled from collaterals (Fig. 3). The graft portal vein was perfused from the meandering splanchnic collateral (Fig. 4). The rejection was reversed, but a series of infections including urosepsis, otitis media, wound infection, and pneumocystis pneumonia necessitated a reduction of immunosuppression. Although the infections were controlled, the liver was irreversibly damaged by chronic rejection. Immunosuppression was reduced but not stopped (CyA 75 mg BID, prednisone 5 mg/d) for 6-1/2 months.

Auxiliary graft retransplantation was performed on 5 January 1984. The auxiliary graft was shrunken and fibrotic without recognizable liver tissue grossly or microscopically. The collateral vein used for portal venous inflow was densely adherent to the anterior abdominal wall. The second auxiliary graft was placed in the same location as the first one. It functioned well for 17 d. However, the wound could not be closed. This was accomplished several days later but the patient developed intestinal ischemia, thought to be secondary to compression by the graft, and then hyperkalemic cardiac arrest. The second auxiliary hepatic allograft had been removed following hyperkalemic arrest

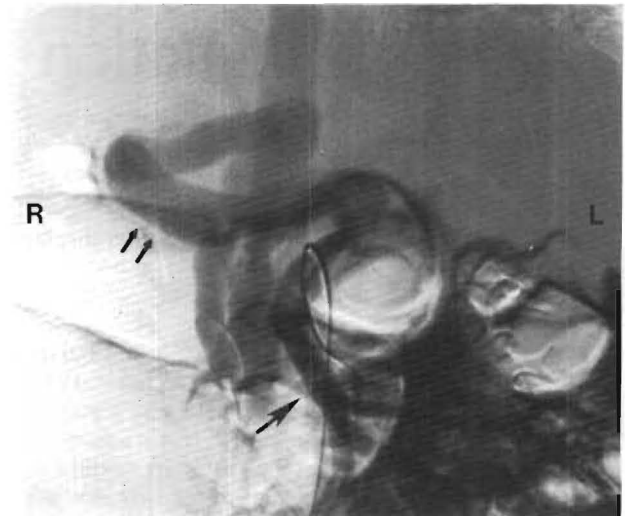


Fig. 2. Venous phase of a superior mesenteric artery angiogram in Patient 1. The superior mesenteric vein (arrow) drains into a large tortuous venous collateral (double arrow) in the right upper abdomen which eventually fills the azygous vein at the level of the right renal vein. The collateral was used to provide portal inflow to the auxiliary hepatic graft (see Fig. 1).

and showed massive infarction with thrombosis in the hepatic artery and portal vein. The subsequent course was complicated by multiple systems deterioration and the child died on 31 January 1984.

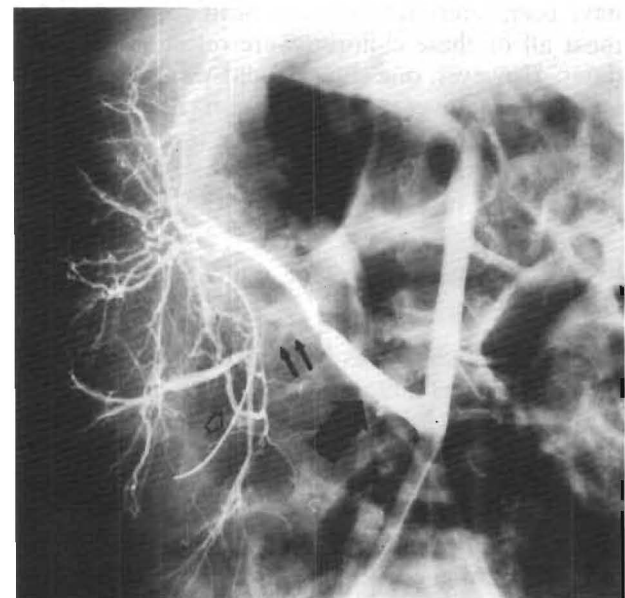


Fig. 3. Postoperative arteriogram via the right iliac artery in Patient 2. The arterial inflow to the auxiliary liver graft is from the recipient infrarenal aorta via an interposition graft of donor thoracic aorta to the donor hepatic artery (large arrow). A curved radio opaque stent catheter in the choledochoenterostomy is seen overlying the auxiliary graft (open arrow). The right hepatic arterial branch and its superior mesenteric artery origin (double arrow) has obstructed but its radicals fill from collaterals.

Autopsy showed a nodular cirrhotic native liver, bacterial (citrobacter, pseudomonas, enterococcus) and fungal peritonitis.

Case 2

A 7-1/2-yr-old female was found at the time of a Kasai procedure at age 2 months to have abdominal *situs inversus* with a midline liver and several small spleens in the right upper quadrant. Palliation from the portoenterostomy was limited and she developed progressive liver disease including jaundice (total bilirubin 11.5 mg%), and clubbing of fingers (PO<sub>2</sub>68) at room air.

Liver transplantation was from a 5-yr-old donor on 24 September 1988, using a piggy-back orthotopic technique (3) which allowed the retrohepatic inferior vena cava to be spared. The original Roux limb for portoenterostomy had been passed behind the stomach to reach to the liver hilum (Fig. 5A). The celiac axis was located at a higher position than normal and its hepatic artery branch ran into the liver hilum directly, passing along the upper border of the pancreas and through the hepatoduodenal ligament (Fig. 5A). The portal vein was post-duodenal. The inferior vena cava was located to the left of the abdominal aorta and ran behind the left-sided pancreas head and duodenum. It then crossed over the aorta and passed through the right diaphragm (Fig. 5B). The right hepatic vein and

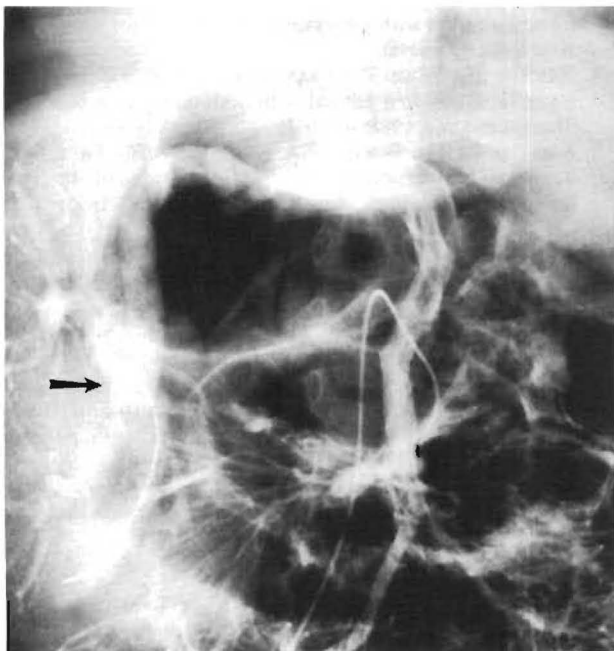


Fig. 4. Venous phase of superior mesenteric artery angiogram showing filling of the auxiliary graft portal system via the portal vein anastomosis (arrow) of the large meandering venous collateral shown in Fig. 2 (see also Fig. 1).

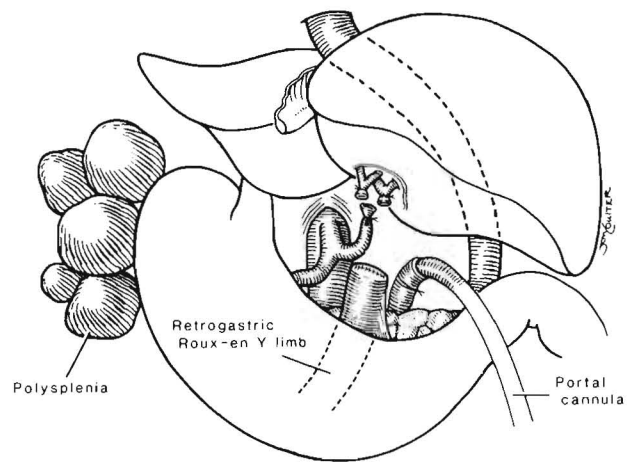


Fig. 5A. Drawing of the anatomical variations of Case 2. Notice veno-venous bypass in place.

left hepatic vein were long enough to make them extrahepatic. In addition, there were more than 10 short hepatic veins draining into the retrohepatic inferior vena cava, which had half of its circumference covered by liver tissue.

A veno-venous bypass was used to decompress the splanchnic venous bed (Fig. 5A). A cloaca was created for the outflow anastomosis (Fig. 5B) at the confluence of the exposed right and left hepatic veins which could be crossclamped without obstructing the inferior vena cava. The liver graft was placed in the midline of the upper abdomen without any effort at rotation. Vascular reconstructions were with an end-to-side anastomosis of the

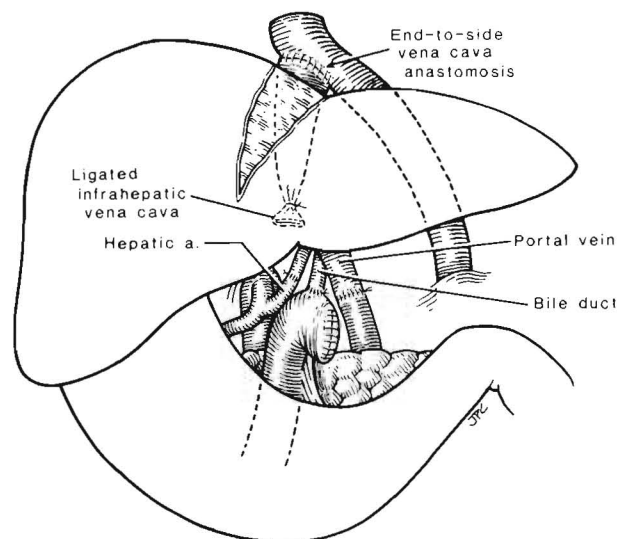


Fig. 5B. Reconstruction after transplant. Note suprahepatic inferior vena cava of the graft was anastomosed to the anterolateral surface of the recipient inferior vena cava. The graft infrahepatic vena cava was ligated.

suprahepatic vena cava and end-to-end anastomoses of the portal vein and the hepatic artery (Fig. 5B). The infrahepatic vena cava was doubly ligated. The previously constructed retrogastric Roux limb of jejunum was used for choledochojejunostomy and this reconstruction was revised 5 d later when the internal stent migrated. The patient was discharged on the 31st postoperative d and is doing well 10 months after transplantation.

### Discussion

To our knowledge, the first patient with *situs inversus* to be treated with liver transplantation was Patient 1 who was given an auxiliary liver. This previously unreported case was mentioned in passing by Raynor et al. (4) who reported the first case of liver replacement under these circumstances.

The auxiliary transplantation was decided upon in Patient 1 because of the extensiveness of the previous operations in the upper abdomen, the loss of small intestine that had occurred during these efforts, and the absence of the portal vein. It was possible to place the liver of a much smaller donor into the right paravertebral gutter, to arterialize the graft from the terminal aorta, and to drain its venous blood into the inferior vena cava. Even more encouraging was the ability to anastomose the graft portal vein to a large splanchnic collateral. Thus, the optimal conditions for auxiliary liver graft revascularization were adhered to as delineated by Marchioro et al. (5) and observed in subsequent efforts at auxiliary transplantation (6–9). Biliary drainage was into a jejunal Roux limb, the construction of which added a further loss to the already shortened intestine.

Rejection of the auxiliary graft after it had functioned for several months was because of undertreatment, particularly with cyclosporine. The patient was cheated thereby of the chance for long-term survival and rehabilitation which has been shown to be feasible with auxiliary liver transplantation (7–9).

The outcome has been better with orthotopic liver transplantation. In the patient of Raynor's group, the liver replacement was carried out in the standard way, including replacement of the retrohepatic inferior vena cava (4). The length of the portal vein and hepatic artery were tailored to allow a crossover of these vessels from the left

sided recipient hilum. The recipient had been followed for 7 months at the time of reporting, and the follow-up is now 2.5 yr (personal communication, form B.W. Shaw Jr, April 1989).

The technical approach to orthotopic transplantation in Patient 2 was different in that the new liver was placed piggyback onto the retained recipient inferior vena cava. This technique which is used by preference in about 1/5 of our orthotopic liver transplantations (3) allows a somewhat greater mobility of the "floating" graft and, with *situs inversus*, this may make adjustments easier for the reconstruction of the hilar vessels. The patient has had complete rehabilitation during the 10 months of postoperative life.

These results demonstrate how quickly advances in transplantation can be applied in a practical way to the treatment of diseases or disease variants which may have seemed in prospect to be beyond help. There would seem to be little justification to consign patients with *situs inversus* to a low priority status merely because of this congenital anomaly.

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