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Liver transplantation in patients with situs inversus

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Abstract. Situs inversus has been considered an absolute contraindication to liver transplantation due to technical difficulties. Associated vascular malformation and distorted anatomy may make the procedure even more complicated or impossible. Only three cases of patients with abdominal situs inversus who underwent successful liver transplantation have been reported in the English literature. We describe two additional patients with situs inversus who suffered from biliary atresia and underwent successful liver transplantation. The preoperative evaluation and the operative procedure are presented, and technical difficulties are discussed. Since biliary atresia is associated with polysplenia syndrome, including vascular malformation and visceral malposition, we suggest that each case be extensively evaluated preoperatively to determine the size requirement for the donor liver and the feasibility of reconstruction.

Key words: Liver transplantation, situs inversus – Situs inversus, liver transplantation

Introduction

Situs inversus, a rare congenital anomaly occurring in 0.1%–0.002% of live births [1], is associated with biliary atresia and polysplenia syndrome in 7% of all cases [6]. Situs inversus has been considered a contraindication to liver transplantation due to technical difficulties arising from vascular anomalies. Only three successful liver allografts in patients with abdominal situs inversus have been reported [3, 4, 7].

We report on two additional patients with biliary atresia and situs inversus who underwent successful liver transplantation. The first patient, who had situs inversus totalis and polysplenia syndrome, is the only known successful liver transplant recipient with that constellation of anomalies. The second patient had abdominal situs inversus only.

Case reports

Case 1

A 23-month-old white male with biliary atresia was referred for transplant after multiple episodes of esophageal variceal hemorrhage. At 6 weeks of age he underwent a Kasai portoenterostomy and was found to have intestinal malrotation, with the cecum located in the left upper quadrant, and Ladd's bands. Additional abdominal malformations included situs inversus, polysplenia, and a preduodenal portal vein. On admission, the patient was deeply jaundiced with severe ascites and prominent abdominal wall venous collaterals. Preoperative radiologic evaluation included upper GI series and CT of the abdomen (Figs. 1, 2). The inferior vena cava could not be adequately assessed by ultrasound evaluation; therefore, cardiac catheterization was performed with the catheter entering the suprahepatic caval system at the level of the heart through the femoral and azygos veins. Findings included dextrocardia, which confirmed the diagnosis of situs inver-

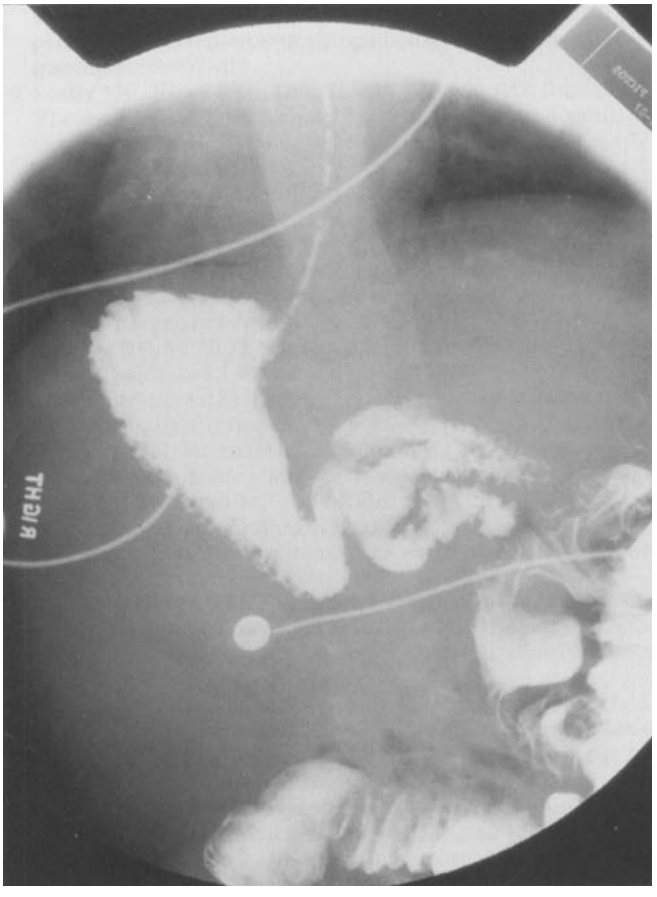

Fig. 1. Right-sided stomach of patient 1 with duodenum sweeping to the left as revealed by upper GI series. The ileocecal valve is located in the left lower quadrant and empties right-to-left into the ascending colon

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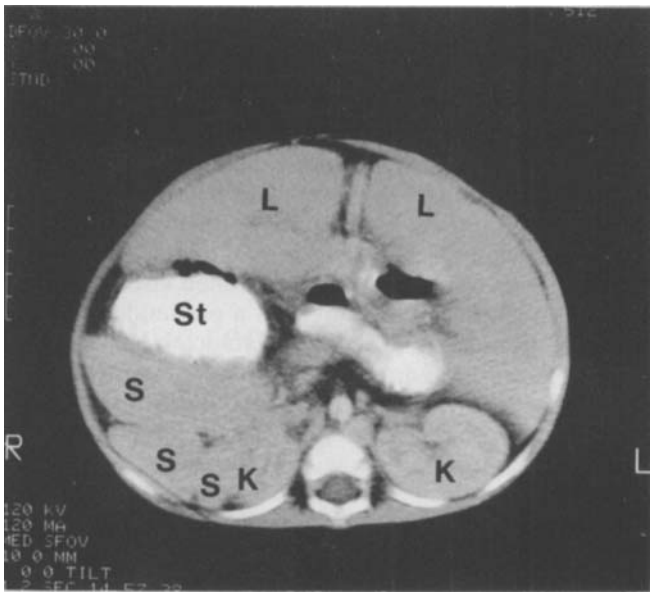

Fig. 2. CT of the abdomen of patient 1 revealing a right-sided polysplenia (*S*) compressing the right kidney. *ST* Stomach; *L* large liver; *K* kidneys

sus totalis, and the absence of the retrohepatic vena cava and its replacement by persistent azygos vein continuation. The hepatic veins drained directly into the functional right atrium (Figs. 3, 4).

On 24 December 1987, the patient underwent liver transplantation. The native liver was in the left upper quadrant extending to the midline and was dissected so that the hepatic veins at the diaphragm were exposed, the portal vein isolated, and the hepatic artery divided. The donor liver was prepared by oversewing the infrahepatic vena cava and anastomosing the donor suprahepatic vena cava to the hepatic venae comitans of the recipient. A standard end-to-end portal vein anastomosis was performed, and recirculation was accomplished via the portal vein. An end-to-end hepatic artery anastomosis was then performed, and a choledochojejunostomy was performed to the pre-existing Roux-en-Y limb of jejunum. Upon completion of the procedure, the right lobe of the liver was in the midline, and the left lateral segment was in the left upper quadrant. Postoperative recovery was uneventful, with discharge on postoperative day 17. At the end of January 1988, a CT of the abdomen showed the liver in the midline as described, but the left lateral segment had expanded to fill the entire left upper quadrant (Fig. 5). Three years and 9 months after liver transplantation, the patient is on minimal immunosuppression and has normal liver function studies.

Case 2

A 7-year-old white female was born with duodenal atresia and jaundice. Shortly after birth, she underwent correction of the duodenal atresia and Kasai portoenterostomy. Concurrent preduodenal portal vein and situs inversus were found. She did well initially but suffered from recurrent episodes of cholangitis and progressive jaundice. Upper gastrointestinal series and CT scan confirmed the diagnosis of abdominal situs inversus. Ultrasound showed a fully preserved inferior vena cava. Magnetic resonance imaging revealed microsplenia, hepatomegaly, and a left inferior vena cava crossing the midline to end at the right atrium. At the time of transplant, the preduodenal portal vein was confirmed and the left-sided liver was excised. Transplantation of the donor liver was accomplished by anastomosing the suprahepatic and infrahepatic vena cavae in the standard fashion, which caused a slight counterclockwise rotation and displacement of the liver to the left. Portal vein and hepatic arterial anastomoses were performed in the standard fashion and recirculation was accomplished without difficulty. The postoperative course was complicated by an ileal perforation and subhepatic abscess requiring surgical drainage. Two and a half years after trans-

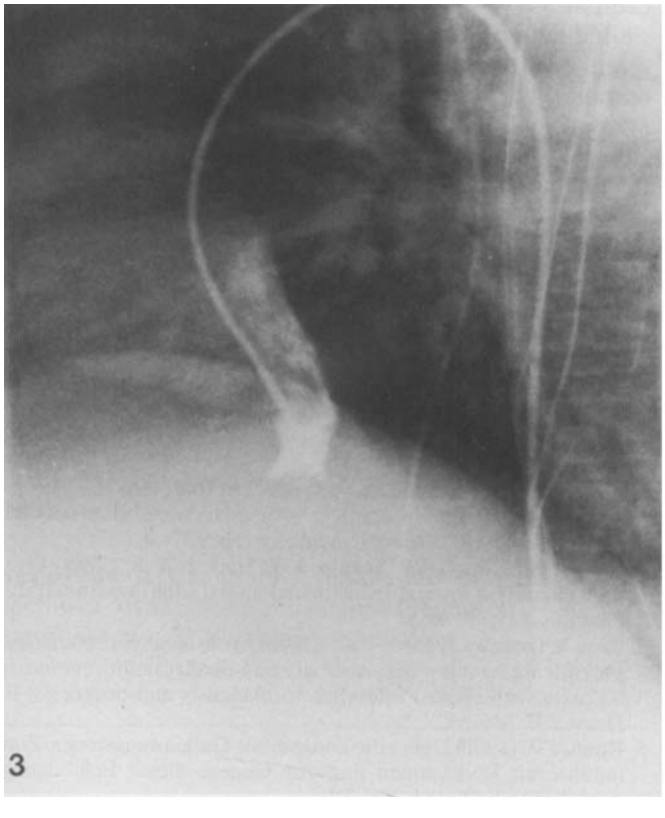

Fig. 3. Femoral catheter of patient 1 rising through the azygos vein entering the suprahepatic caval system at the level of the heart. The infrahepatic cava is absent

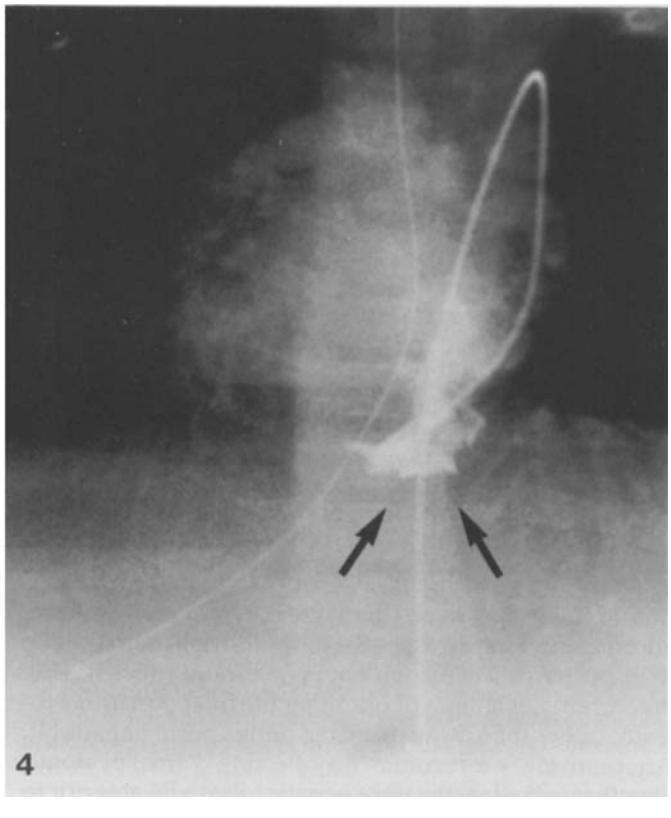

Fig. 4. AP view of Fig. 3 (patient 1) showing dextrocardia and hepatic veins draining into the suprahepatic cava

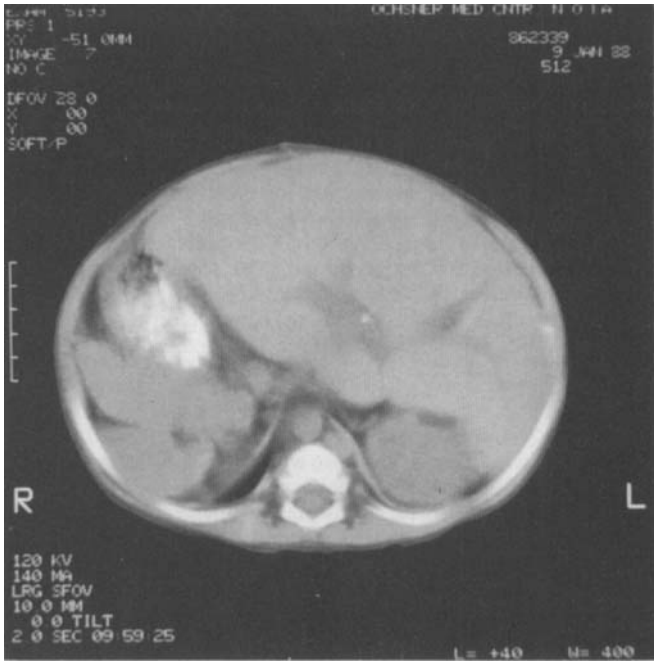

Fig. 5. CT of patient 1, 1 month after liver transplantation: the left lobe of the liver filled the entire left upper quadrant of the abdomen

plantation, liver function studies are normal and the patient is thriving.

Discussion

We report two of the five known cases of situs inversus in which liver transplantation has been successfully attempted. One of our patients had a situs inversus totalis constellation (abdominal situs inversus with dextrocardia). Of the remaining three cases, Raynor et al. reported a case of situs inversus without dextrocardia but with a normally situated suprahepatic vena cava [4], Klien et al. reported a case of abdominal situs inversus with levocardia [3], and Todo et al. described two patients with biliary atresia and situs inversus, only one of whom survived after liver transplantation [6].

Unsuccessful liver transplantation in patients with situs inversus is attributed to (1) the potential difficulties associated with a left-sided suprahepatic vena cava; (2) the difficulty in accommodating a large right lobe in the right upper quadrant, which is already occupied by the stomach and spleen (or multiple spleens); and (3) the potential for kinking of the suprahepatic vena caval anastomosis by the anterior and leftward displacement of the large right hepatic lobe.

Raynor et al. have suggested, but not attempted, rotation of the donor liver 180 degrees on the vertical axis, allowing the anterior aspect of the bulky right hepatic lobe to lie posteriorly in the left upper quadrant, which has the theoretical advantage of orienting the hilar structures toward, rather than away from, the native porta hepatis [4]. Alternatively, we recommend selecting a smaller donor liver that will allow the right hepatic lobe to lie anterior to the vertebral column without excessive anterior displacement of the suprahepatic caval anastomosis or compression of the liver. The left lobe appears to expand to fill the left upper quadrant space formerly occupied by the native liver. Increased experience transplanting smaller livers has eliminated these size and orientation problems.

Furthermore, the partial liver transplant, which has evolved into a clinically useful procedure, alleviates the shortage of donors for those smaller patients and, by using only the left lobe, solves the space problems as well [2].

Patients with situs inversus may exhibit a wide variety of vascular anomalies and malpositioned viscera, such as our first case, in which there was a preduodenal portal vein and an absent infrahepatic vena cava. Unexpected anomalies may require unusual positioning of the liver graft or vascular extension grafting; therefore, it is always important to obtain maximum lengths of suprahepatic and infrahepatic vena cavae with the donor liver and to harvest iliac arterial and venous grafts and possibly carotid and jugular venous grafts. Extension grafting was not required in our first case since adequate venous length could be obtained by dissecting the native hepatic veins. The absence of an infrahepatic vena cava actually simplified the anastomosis by allowing simple ligation of the graft infrahepatic cava. Another note of caution is that the presence of the polysplenia syndrome should alert one to the existence of multiple vascular anomalies and congenital cardiac anomalies. The polysplenia syndrome has been associated with a 50%–60% mortality in the 1st year of life, usually secondary to cardiac complications such as anomalous pulmonary venous return and transposition of the great vessels [5].

In conclusion, situs inversus is not an absolute contraindication to liver transplantation. Situs inversus is, however, an indication that concomitant vascular anomalies should be anticipated. In addition, presence of the polysplenia syndrome further raises the likelihood of intra-abdominal visceral and vascular anomalies and intrathoracic anomalies. When leftward displacement is mandated by situs inversus, using a down-sized donor liver that allows the right lobe to fit in the midline or using only the left lobe of an adult liver contributes to the ease of operation.

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