Journal of
Medical Case Reports

CASE REPORT

Open Access

Liver and pancreas transplantation in adult donor and recipients with situs inversus totalis: a case series and review of the literature

Alireza Shamsaeefar[1], Fatemeh Masjedi[2], Jamshid Roozbeh[2], Sahar Sohrabi Nazari[1], Edalat Zarei[1], Mehran Jafari[1], Sara Farifteh[1], Mohammad Alikhani[1], Mohammad Eslamian[1], Maryam Mardani[1], Reyhaneh Naseri[3] and Hamed Nikoupour[1*]

Abstract

Background Situs inversus totalis is a rare congenital anomaly characterized by a mirror-image orientation of abdominal, and in some cases, thoracic organs. Here, we report our situs inversus totalis transplantation experience and further review liver transplantations in adult recipients and donors with situs inversus totalis.

Case presentation We describe three cases with situs inversus totalis. The first case was liver transplantation in a recipient (a 61-year-old Iranian man) with situs inversus totalis, the second was a liver transplantation from a donor (a 52-year-old Iranian woman) with situs inversus totalis, and finally, for the first time, a simultaneous pancreas and kidney transplantation in a recipient (a 26-year-old Iranian man) with situs inversus totalis. In patient one, hepatectomy could be performed according to the standard method and on the basis of preoperative studies. Hepatic vein and arterial anastomosis were performed as in every other patient without situs inversus totalis. To prevent biliary complications, a Roux-en-Y hepaticojejunostomy was performed. In patient two, implantation time, suprahepatic vein, portal vein, arterial, and biliary reconstruction could be done as in any other case without situs inversus totalis. Plication of the right-sided diaphragm and fixation of the falciform ligament was done for our patient. In patient three, systemic drainage was preferred to portal flow for establishing the outflow drainage of the pancreas compared with otherwise normal patients.

Conclusion Although situs inversus totalis is a rare condition, our reported techniques are suitable, considering advantages such as easier accessibility, more acceptable placement of the implanted organs regarding vascular variations, and the appropriate location of the allograft in the proximity of other organs.

Keywords Situs inversus totalis, Liver, Pancreas, Transplantation, Vascular anastomosis, Case report

Introduction

Orthotopic liver transplantation (LTx) has become the standard and final treatment option for end-stage liver diseases. Because of the shortage of organs and the increasing number of patients who require transplantation, maximum effort is made to procure and transplant organs appropriately. Therefore, facing anatomical diversity and anomalies in the donor and recipient is inevitable [1, 2]. One of these anomalies is situs

*Correspondence:
Hamed Nikoupour
nikoupour@gmail.com
[1] Shiraz Organ Transplant Center, Abu-Ali-Sina Hospital, Shiraz University of Medical Sciences, Shiraz, Iran
[2] Shiraz Nephro-Urology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran
[3] Thoracic and Vascular Surgery Research Center, Shiraz University of Medical Sciences, Shiraz, Iran

inversus totalis (SIT). It is a rare congenital abnormality characterized by a mirror-image orientation of the abdominal, and in some cases, thoracic organs relative to the midline. The incidence of SIT is about 5–25 in every 100,000 births [3]. Implanting a mirror-image organ may pose a technical challenge to a transplant surgeon because of the reversed position and orientation of the graft and vascular system. Some of the potential risks include vascular torsion and hepatic venous outflow kinking, inferior vena cava (IVC) compression, need for additional vascular reconstruction, and pressure effect on the stomach due to the placement of the greater right lobe in the recipient's hepatic fossa [4].

In addition, SIT is often associated with other anomalies such as biliary atresia (BA), polysplenia syndrome (interrupted IVC with azygous drainage, confluent hepatic veins to the right atrium, preduodenal portal vein, hypoplastic or atretic portal vein, and aberrant hepatic arterial anatomy), congenital heart disease, Kartagener's syndrome, and midgut malrotation that would eventually make the transplantation more complex [3, 5, 6].

The most documented association of SIT is with BA, which is a cause of end-stage liver disease among pediatrics. Approximately 28% of children with SIT have BA [7]. Accordingly, most cases of transplantation in SIT are reported in this group.

Although most people with SIT have a normal life without complications, this condition could be problematic when they require surgery, especially LTx [8]. Due to these difficulties, SIT was previously considered a contraindication for LTx. Advances in surgical technique and experience have overcome these limitations [9, 10].

However, experiences with SIT are still limited, and different techniques have been used according to each individual's circumstances. Here, we first describe a case of LTx in a SIT recipient, then a successful LTx from a donor with SIT, and finally, for the first time, we report on a pancreas transplantation in a SIT recipient.

Case presentation

We collected medical records of patients diagnosed with SIT who underwent transplant surgery from January 2018 to September 2023, affiliated with Shiraz Organ Transplant Center, Abu-Ali-Sina Hospital, Shiraz University of Medical Sciences. The Shiraz University of Medical Sciences Ethics Committee approved this study (IR.SUMS.REC.1402.570). Three written informed consents were obtained from patients for the publication of these case reports and any accompanying images.

Case 1

A 61-year-old Iranian man with a known case of alcoholic liver cirrhosis with complete situs inversus was referred to our center with severe ascites and grade II encephalopathy. He had stopped drinking alcohol a year before. During the previous 6 months, he suffered from severe ascites and required multiple peritoneal paracentesis, resulting in three emergency department admissions. Because of the increase in creatinine level (up to 2 mg/dL) and an existing hepatorenal syndrome, the patient was on the waiting list for LTx. Computed tomography showed that the arterial blood supply to the liver originated from the celiac axis with no evidence of the replaced hepatic artery. He had no associated vascular and visceral anomalies (Fig. 1). In other words, there were no signs of intestinal malrotation or intermittent IVC. The model for end-stage liver disease (MELD) score was 24 at admission, and his height and body weight were 178 cm and 95 kg, respectively.

In April 2022, he had LTx and received an ABO-compatible whole liver from a 75 kg, 44-year-old donor whose graft weighed 1.4 kg. Dissection of the hepatic hilum was performed without unexpected difficulty, but hepatectomy was done with standard technique (resection of recipient's IVC with liver). The liver was directly placed into the recipient's left upper quadrant without rotating adjustment; the right hepatic lobe was positioned on the recipient's left side, and the left liver lobe was placed in the recipient's previous liver fossa. The hepatic superior and inferior vena cava were directly anastomosed to the recipient's IVC with 3–0 vascular sutures. Portal veins were anastomosed

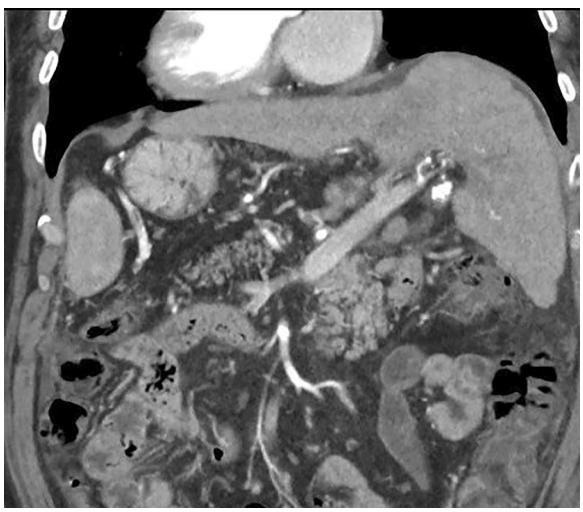

Fig. 1 Pre-transplant contrast-enhanced abdominal computed tomography findings showing a complete situs inversus with a mirror-image positioning of abdominal organs

end-to-end with 6–0 polypropylene sutures. The cold ischemia time was 4 hours.

Arterial reconstruction was done using an end-to-end technique with a 7–0 continuous suture. Biliary reconstruction was performed by duct-to-duct anastomosis between the recipient's common bile duct and the donor's common hepatic duct with polydioxanone sutures (PDS); however, seemingly, there was some tension on the anastomosis site. Finally, to prevent the rotation of the transplanted liver, diaphragm plication and fixation of the falciform ligament to the diaphragm were carried out. The total operation time was 6.5 hours, and the patient was extubated 12 hours after the operation; no veno-venous bypass was used during the anhepatic phase. On postoperative day 12, the patient's serum bilirubin levels increased to 5 mg/dL, and biliary fluid passed through the incision site, so exploration was done for the patient. Exploration showed that the anterior side of the previous common bile duct anastomosis was disrupted. Accordingly, we took the last biliary anastomosis and reconstituted it by a Roux-en-Y hepaticojejunostomy. The patient was discharged from the hospital on day 25 post-transplantation.

Considering the anatomical differences between the recipient with situs inversus and the recipient with normal anatomy, the transplantation technique seems modified to prevent complications following LTx. As shown in Fig. 2A, the direction of the recipient's bile duct and portal vein had an acute angle to the donor's liver. Due to this anatomical difference, performing Roux-en y hepaticojejunostomy would potentially result in fewer complications than duct-to-duct anastomosis. Furthermore, for portal vein anastomosis, the length of the portal vein should be as short as possible to prevent kinking and future portal vein thrombosis. Given that the left lobe of the donor's liver is located in the space that was previously the location of the right lobe of the recipient's liver, this mismatch can cause a possible rotation of the transplanted liver. To prevent this complication, ipsilateral diaphragm plication and fixation of the falciform ligament to the diaphragm were carried out (Fig. 2B).

Case 2

A 52-year-old Iranian woman was admitted due to intracerebral brain hemorrhage (ICH) and was diagnosed with brain death 2 days after admission. The patient was prepared for organ donation. Dextrocardia was discovered in chest radiography, and SIT was confirmed after abdominal ultrasonography. During the procurement procedure, we found the liver, pancreas, spleen, and stomach on the opposite side; the aorta was on the right side, and IVC was on the left side and anterior to the spine (Fig. 3A). There was no problem during the procedure; however, the operation time was longer than usual due to meticulous dissection. During bench surgery, the common bile duct was on the left side of the hilum, and the proper hepatic artery was on the right side. Still, no anatomical variation was seen in the hepatic artery, and portal vein was in the posterior position. Arterial structures were preserved with an aorta patch, and IVC was excised to the longest length possible. The donor's liver weighed 1.2 kg.

The recipient was a 48-year-old man with a history of decompensated cirrhosis due to non-alcoholic steatohepatitis (NASH) and was a candidate for LTx. Hepatectomy was performed with the piggyback method, and the implantation was done with the liver placement in the middle part of the upper abdomen. The right lobe of the liver graft was placed toward the recipient's left side. Outflow was established by anastomosis of the donor's suprahepatic vena cava to the recipient's common funnel of hepatic veins. The distal end of the donor cava was ligated. The portal vein was anastomosed using the end-to-end technique. The liver perfusion was established without any problems. Arterial reconstruction was done using the end-to-end technique by anastomosis of the

Fig. 2 Completion of the suprahepatic, portal, and arterial reconstructions in the recipient with situs inversus totalis following orthotopic liver transplantation (patient 1) (**A**) and schematic orientation of the liver transplant inside the recipient's body with situs inversus totalis (**B**)

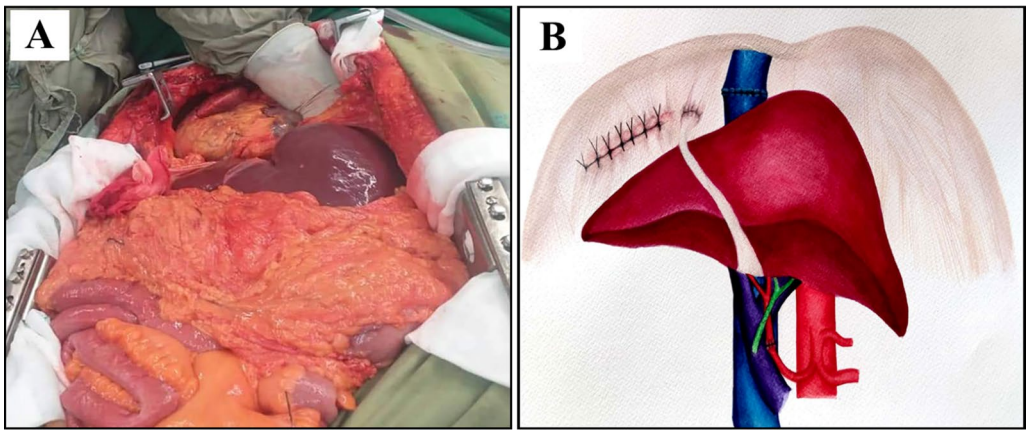

Fig. 3 Liver anatomy in deceased donor with situs inversus totalis (patient 2) (**A**) and schematic orientation of the liver transplant inside the recipient's body with normal anatomy (**B**)

donor's hepatic artery to the recipient's common hepatic artery. Finally, duct-to-duct anastomosis was done without tension. To prevent the rotation of the liver and to eliminate the empty space on the right side, we plicated the right diaphragm and the falciform ligament (Fig. 3B). The patient was discharged without any complications.

Case 3

A 26-year-old Iranian man with diabetic nephropathy was referred to our clinic for pancreas transplantation. He was on continuous insulin therapy without proper glycemic control from 10 years ago. He had also lost his vision due to diabetic retinopathy. During previous imaging studies, SIT was identified, and associated vascular anomalies were documented.

The donor was a 36-year-old man who developed brain death due to subdural hematoma following a traumatic brain injury. Grafts were in good condition with normal function.

During bench surgery, we made a combined patch of the celiac trunk and superior mesenteric artery (SMA) from the donor's aorta to avoid separate anastomosis or using a Y-graft. The donor's portal vein behind the pancreas was also prepared for venous outflow.

Laparotomy was done through a midline incision. The aorta and left-sided IVC were localized. Considering the SIT, the surgeon was on the right side of the patient. The prepared portal vein was anastomosed to the IVC as an outflow pathway, and the Carel patch was anastomosed to the recipient's common iliac artery. The donor's duodenum was anastomosed to the nearest loop of the jejunum from the recipient (Fig. 4).

No unexpected event occurred following declamping, and normal blood sugar levels were achieved quickly. The kidney was transplanted on the right side to achieve better access to the iliac vessels. The hospital course was uneventful except for an episode of rejection diagnosed with elevated creatinine levels. The patient received one course of methylprednisolone and recovered completely.

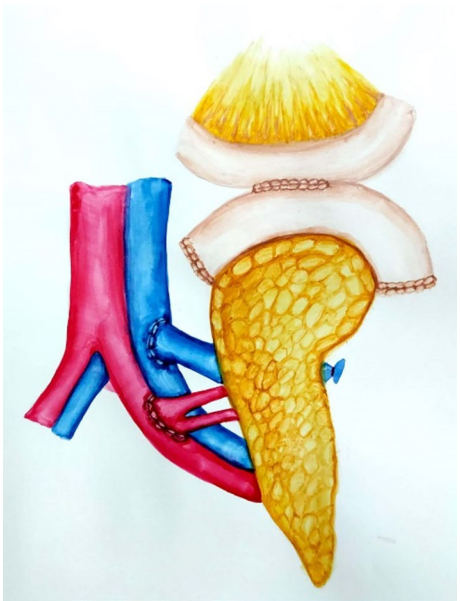

Fig. 4 Pancreas anatomy following transplantation in recipient with situs inversus totalis (patient 3)

Discussion

In cases of SIT, organ transplantation may be complicated. Both donors and recipients can have SIT, although matched donor-recipient combinations are unlikely to be found due to their rarity. However, there are several reports on children and adults with SIT undergoing LTx. Literature on LTx in SIT is presented in Table 1.

Table 1 Published studies on liver transplantation in adults (donors, recipients) with situs inversus

Author	Year	Age (years)	Sex	Diagnosis	Implant technique	Anastomosis			Complications	Outcome
						Biliary	Arterial	Vena cava		
Recipients with situs inversus										
Barone *et al.* [1]	1992	17	F	CHF	Midline shift position	Duct to duct	HA end to end	Piggyback	None	Alive, 6 months
Klintmalm *et al.* [15]	1993	45	F	ETOH	90° clockwise rotation	Duct to duct	HA end to end	End to side	None	Alive, 18 months
Watson *et al.* [10]	1995	35	n/a	CC	Orthotopic	Duct to duct	Celiac patch-hepatic artery	Piggyback	Biliary leak	Alive, 7 months
Pereira-Lima *et al.* [24]	2000	18	F	SBC	Midline shift position	RY HJ	HA end to end	Piggyback	None	Alive, unknown
Heimbach *et al.* [25]	2005	62	M	PSC	Midline shift position, omentum, and hepatic flexure to hepatic fossa	RY HJ	HA end to end	End to end	Biliary stricture	Alive, 4 months
Wente *et al.* [26]	2006	48	M	ETOH	Orthotopic	Duct to duct	HA to GDA	Piggyback	None	Alive, 24 months
Tucker *et al.* [13]	2006	41	M	ETOH	Midline shift position	Duct to duct	HA end to end	Piggyback	None	Alive, 17 months
Hoyos *et al.* [14]	2006	41	M	CC	90° clockwise rotation	Duct to duct	HA end to end	End to side	None	Alive, 21 months
Tang *et al.* [27]	2008	45	M	SBC	45° clockwise rotation	Duct to duct	Celiac patch-hepatic bifurcation	End to side	None	Alive, 40 months
Soejima *et al.* [5]	2008	19	F	HCV CLD	Midline shift position, slight clockwise rotation, falciform ligament fixation	RY HJ	HA end to end	HV to HV	Wound infection-fistula	Alive, 12 months
Rayhill *et al.* [21]	2009	53	F	PBC	180° rotation around the vertical axis (flip, facing backward, reversed)	Duct to duct	HA end to end	Reversed cavoplasty	None	Alive, 36 months
Kim *et al.* [8]	2010	54	F	HBV CLD	Orthotopic	Duct to duct	HA end to end	HV to HV	None	Alive, 8 months
Kamei *et al.* [19]	2014	60	M	HBV CLD	180° rotation around the vertical axis (flip, facing backward, reversed), diaphragm plication	RY HJ	HA end to end	HV to HV	None	Alive, 11 months
Fernandez Soria *et al.* [28]	2015	34	M	ESLD secondary to BA	Orthotopic	RY HJ	HA end to end	HV to RA	None	Alive, unknown

Table 1 (continued)

Author	Year	Age (years)	Sex	Diagnosis	Implant technique	Anastomosis			Complications	Outcome
						Biliary	Arterial	Vena cava		
Yankol *et al.* [29]	2015	18	F	AIH	Orthotopic, slight clockwise rotation, triangulated suture technique of hepatic vein anastomosis	Duct to duct	HA end to end	HV to HV	None	Alive, 20 months
Sankarankutty *et al.* [12]	2015	44	M	CC	90° clockwise rotation	RY HJ	HA end to end	End to side	None	Alive, 36 months
Tabrizian *et al.* [6]	2016	23	F	ESLD secondary to BA	Midline shift position	RY HJ	HA end to end	End to side	None	Alive, 15 months
Kauffmann *et al.* [30]	2017	59	M	HCV CLD	Heterotopic, 90° counter-clockwise rotation	RY HJ	Celiac patch-hepatic artery	Piggyback	None	Alive, 36 months
Kristek *et al.* [2]	2018	58	F	HCC	90° clockwise rotation	Duct to duct	HA end to end	End-to-side	None	Alive, 27 months
King *et al.* [31]	2019	72	F	ESLD secondary to NASH	Orthotopic	Duct to duct	HA end to end	Piggyback	None	Alive, unknown
Na *et al.* [32]	2020	42	M	HBV CLD	Orthotopic	Duct to duct	HA end to end	Piggyback	None	Alive, 30 months
Donors with situs inversus										
Asfar *et al.* [23]	1995	63	M	ARLD	Heterotopic, 90° counter-clockwise rotation	RY HJ	HA end to side	Piggyback	ARDS, biliary leak, sepsis	Dead
Herrera *et al.* [33]	1996	40	M	Cirrhosis	Orthotopic	Duct to duct	HA end to side	Piggyback	None	Alive, 30 months
Braun *et al.* [9]	1998	56	F	ARLD	Orthotopic	Duct to duct	HA end to side	Piggyback	None	Alive, 17 months
Pomposelli *et al.* [20]	2007	41	M	HCV CLD and HCC	Heterotopic, 180° rotation around the vertical axis	RY HJ	Celiac patch-hepatic artery	Piggyback	None	Alive, unknown
Chun *et al.* [18]	2009	53	M	ARLD	Heterotopic, 180° rotation around the vertical axis	Duct to duct	HA end to end	HV to HV	None	Alive, 12 months
Dou *et al.* [34]	2010	50	M	End-stage Budd–Chiari syndrome	15° clockwise rotation	Duct to duct	HA end to end	End to end	None	Alive, 10 months
Sun *et al.* [11]	2013	58	M	HBV CLD	Orthotopic (minimum rotation)	Duct to duct	HA end to end	Piggyback	None	Alive, 36 months

Table 1 (continued)

Author	Year	Age (years)	Sex	Diagnosis	Implant technique	Anastomosis			Complications	Outcome
						Biliary	Arterial	Vena cava		
Moon *et al.* [35]	2013	50	M	ARLD	150° counter-clockwise rotation along the axis of the IVC groove and additionally rotated upward about 80–90° along the coronal axis	Duct to duct	HA end to end	Reversed HV	None	Alive, 34 months
Manzia *et al.* [16]	2015	57	M	HCC	Orthotopic	Duct to duct	Celiac patch-hepatic artery	Piggyback	None	Alive, 8 months
Selvakumar *et al.* [36]	2016	44	M	HCV CLD	180° counter-clockwise rotation	Duct to duct + RY HJ	HA end to end	RHV to IVC	None	Alive, 58 months
	2016	25	M	HBV CLD	180° counter-clockwise rotation	Duct to duct	HA end to end	RHV to IVC	None	Alive, 48 months
	2016	46	M	CC	180° counter-clockwise rotation	Duct to duct	HA end to end	RHV to IVC	None	Alive, 6 months
Reimondez *et al.* [37]	2019	69	M	ESLD	Orthotopic	RY HJ	HA end to end	Piggyback	None	Alive, unknown
Lopez-Andujar *et al.* [38]	2021	52	M	HCV CLD	Orthotopic	Duct to duct	HA end to end	Piggyback	None	Alive, unknown
Both recipient and donor with situs inversus										
Sugawara *et al.* [39]	2001	2 years, 11 months	M	Hepatic failure secondary to BA	Orthotopic	RY HJ	HA end to end	End to end	None	Alive, 20 months
Yu *et al.* [3]	2014	53	M	HCC secondary to HBV	SI graft, reTx, ABOi, piggyback End to side	Duct to duct	HA end to end	Piggyback	None	Alive, 11 months

F female, *M* male, *HA* hepatic vein, *HV* hepatic vein, *n/a* not available, *mo* month(s), *reTx* retransplantations, *ABOi* ABO incompatible, *RY HJ* Roux en Y hepaticojejunostomy, *CHF* congenital hepatic fibrosis, *EOTH* alcoholic cirrhosis, *CC* cryptogenic cirrhosis, *SBC* secondary biliary cirrhosis, *PSC* primary sclerosing cholangitis, *HCV CLD* hepatitis C virus chronic liver disease, *PBC* primary biliary cirrhosis, *HBV CLD* hepatitis B virus chronic liver disease, *ESLD* end-stage of liver disease, *BA* biliary atresia, *AIH* autoimmune hepatitis, *HCC* hepatocellular carcinoma, *NASH* non-alcoholic steatohepatitis, *ARLD* alcohol-related liver disease

Despite SIT being a rare developmental defect during embryogenesis, it is not considered a contraindication for organ transplantation, whether as a donor or recipient. Further, we should keep in mind that the risk of surgical complications may increase because of anatomical variation in the positioning of the vasculature.

The liver is an asymmetric organ that is always affected by SIT-related anatomic abnormalities. As a result, donation from a donor with SIT and transplantation for a recipient with SIT are complicated surgical procedures. Because of the technical challenges associated with the unusual vascular architecture and concerns related to proper graft location, SIT was formerly considered a contraindication for LTx [11]. The two main problems for orthotopic LTx in SIT seem to be (i) having to place the graft's greater right lobe over the right-sided stomach and (ii) the presence of a large empty space in the left upper quadrant, which predisposes the graft to lateral displacement and kinking of the hepatic veins [12].

Even though several successful strategies have been suggested, there has yet to be agreement on a standard technique. These techniques include (i) plication of the left diaphragm combined with graft fixation with a percutaneously inserted gastric balloon of the Sengstaken–Blakemore tube [13]; (ii) transplantation of a portion of the liver from a living donor (that is, reduced size) [8]; (iii) different degrees (15–90°) of lateral rotation of the graft (clockwise in SIT recipients and counter-clockwise in SIT donors) using standard piggyback, end-to-side, or side-to-side cavo-caval anastomosis [2, 8, 14–17]; and (iv) 180° flip of the graft (retroversus or backward facing) [18–21] (see Table 1). Regarding living donations, segmental LTx allows the transplant surgeon greater flexibility in determining the best choice. New technologies, such as three-dimensional (3D) printing, can aid surgery planning if there is enough time for preoperative planning [22].

In our case (patient 1), performing Roux-en-Y hepaticojejunostomy potentially has fewer complications compared with duct-to-duct anastomosis. In addition, for portal vein anastomosis, the length of the portal vein should be as short as possible to prevent kinking and future portal vein thrombosis. To prevent the rotation of the transplanted liver, ipsilateral diaphragm plication and fixation of the falciform ligament to the diaphragm were used.

Moreover, there were few reports on using a liver from a donor with SIT. The most challenging part of these cases is the venous outflow reconstruction. In 1995, Asfar and Ozcay [23] reported transplantation with a liver from a SIT donor. They rotated the liver 90° counter-clockwise and performed an end-to-side anastomosis between the donor's intrahepatic vena cava and the recipient's IVC.

In other reports, authors mentioned different methods, such as backward rotation of the liver 180° along the axis of the IVC or T-shape anastomosis between the left and middle hepatic veins and IVC. In our case (patient 2), we performed the piggyback technique for hepatectomy, IVC was preserved, and the liver was implanted with its original orientation. Anastomosis between the donor's suprahepatic vena cava and the recipient's common funnel of hepatic veins was performed, and there was no need for significant rotation of the liver. Another issue that should be considered is the empty space on the right side of the upper abdomen. In our case, we plicated the diaphragm, but in some case reports, the omentum has been used. Fixation of the falciform is also helpful. A smaller graft might be more logical for preventing compression on the IVC or stomach.

To the authors' knowledge, no previous reports existed for pancreas transplantation in SIT. For this, a systemic approach must be selected due to technical difficulties that may be encountered using conventional portal-portal anastomosis. In SIT cases, the aorta is localized far from the portal system, so using iliac vessels and the superior mesenteric vein (SMV) leads to inappropriate angulation and puts the graft at risk of vascular compromise. In addition, in the method used for this case (patient 3), the graft is positioned perpendicularly, taking a better position in the abdomen with reversely placed organs. Our center's policy is to place the left kidney in the left retroperitoneum for simultaneous kidney and pancreas transplantations. In this case, we used the left kidney on the right side as it provided us with better exposure and vascular anatomy for implantation. Although we encountered complexities, our technique presented a suitable option for this anomaly due to advantages such as easier accessibility, better placement of the implanted organs regarding vascular variations, and the appropriate location of the allograft in the proximity of other organs.

Conclusion

On the basis of previous literature and our experience, we advocate applying specific techniques, which are as follows: our recommendations in organ recipients with SIT: (1) ergonomy; the transplant surgeon should be on the left side of the patient during the operation; hepatectomy can be done as a standard or piggyback method; however, we recommend using the standard technique if the patient has a good cardiopulmonary reserve that can tolerate total clamping of the IVC; (2) implantation; hepatic vein and arterial anastomosis can be performed as with normal patients, the portal vein should be shortened as much as possible, and to prevent biliary complications, we suggest Roux-en-Y

hepaticojejunostomy and organ rotation are not mandatory in all cases; and (3) plication of the diaphragm and fixation of the liver are recommended in the recipient.

Our recommendations in deceased donors with SIT anatomy are as follows: (1) hepatectomy can be carried out using a standard or piggyback method based on preoperation studies; (2) suprahepatic vein, portal vein, and arterial and biliary reconstruction can be done similarly to normal patients, and implantation time can be similar; whether a liver rotation is performed depends on the surgeon's preference; and (3) plication of the right-sided diaphragm and fixation of the falciform ligament are advised in all patients.

Our recommendations in simultaneous pancreas and kidney transplantation with SIT anomaly are as follows: (1) standing on the left side of the patient results in more comfortable access during operation; (2) for establishing outflow drainage, systemic drainage is preferred to portal drainage; (3) for anastomosis of arterial graft, if the procurement team of the liver and pancreas is the same, the patch of the celiac trunk and SMA is to be given to the pancreas, and this patch is directly anastomosed to the aorta; and (4) kidney is placed in the right side in the retroperitoneal space, and anastomosis can be done as usual.

Finally, more acceptable outcomes can be achieved with meticulous perioperative planning, a complete anatomic understanding of the liver and pancreas in the donor and recipient, and the use of novel procedures.

Abbreviations

BA	Biliary atresia
IVC	Inferior vena cava
LTx	Liver transplantation
MELD	Model for end-stage liver disease
NASH	Non-alcoholic steatohepatitis
SIT	Situs inversus totalis
SMV	Superior mesenteric vein
SPK	Simultaneous pancreas kidney transplantation

Acknowledgements
The authors wish to thank Ms. Sheryl Thomas-Nikpoor, Language Editor at Springer Publications, for her invaluable comments in editing this manuscript.

Author contributions
FM, SSN, EZ, MJ, SF, ME, and MM wrote the main manuscript text, and MA prepared Figs. 2, 3, 4. All authors reviewed the manuscript. AS, JR, and HN contributed to the conception and design of the study. FM and RN contributed to the interpretation of data. All authors contributed to the article and approved the submitted version.

Funding
The study was funded by the Vice Chancellor of Research Affairs, Shiraz University of Medical Sciences (ID: 29887), Shiraz, Iran.

Availability of data and materials
The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
All procedures performed in studies involving human participants were in accordance with the ethical standards of the local Medical Ethics Committee of Shiraz University of Medical Sciences (IR.SUMS.REC.1402.570) and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Consent for publication
Written informed consents were obtained from the patients for the publication of these case reports and any accompanying images. A copy of the written consents is available for review by the Editor-in-Chief of this journal.

Competing interests
All the authors declare no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Received: 26 December 2023 Accepted: 12 July 2024
Published online: 29 August 2024

References

1. Barone GW, Henry ML, Elkhammas EA, Tesi RJ, Ferguson RM. Orthotopic liver transplantation with abdominal situs inversus and dextrocardia. Am Surg. 1992;58(10):651–3.
2. Kristek J, Kocík M, Chlupac J, Froněk J. Orthotopic liver transplantation with reduced and rotated graft in adult situs inversus recipient: a case report and a review of reported cases. Rozhl Chir. 2018;97(12):568–75.
3. Yu S, Guo H, Zhang W, Yu J, Yan S, Wu J, *et al*. Orthotopic liver transplantation in situs inversus adult from an ABO-incompatible donor with situs inversus. BMC Gastroenterol. 2014;14:46.
4. Raynor SC, Wood RP, Spanta AD, Shaw BW Jr. Liver transplantation in a patient with abdominal situs inversus. Transplantation. 1988;45(3):661–3.
5. Soejima Y, Meguro M, Taketomi A, Ikegami T, Yamashita Y, Harada N, *et al*. Left lobe living donor liver transplantation in an adult patient with situs inversus: technical considerations. Transpl Int. 2008;21(4):384–9.
6. Tabrizian P, Joseph TT, Radkani P, Cohen E, Facciuto M. Liver transplantation in an adult recipient with situs inversus totalis: case report and review of the literature. Transpl Proc. 2016;48(9):3163–6.
7. Millis JM, Brems JJ, Hiatt JR, Klein AS, Ashizawa T, Ramming KP, *et al*. Orthotopic liver transplantation for biliary atresia: evolution of management. Arch Surg. 1988;123(10):1237–9.
8. Kim BW, Bae BK, Xu W, Wang HJ, Kim MW. Living donor liver transplantation for an adult patient with situs inversus totalis. World J Gastroenterol. 2010;16(18):2311–3.
9. Braun F, Rodeck B, Lorf T, Canelo R, Wietzke P, Hartmann H, *et al*. Situs inversus of donor or recipient in liver transplantation. Transpl Int. 1998;11(3):212–5.
10. Watson CJ, Rasmussen A, Jamieson NV, Friend PJ, Johnston PS, Barnes ND, *et al*. Liver transplantation in patients with situs inversus. Br J Surg. 1995;82(2):242–5.
11. Sun XY, Qin K, Dong JH, Li HB, Lan LG, Huang Y, *et al*. Liver transplantation using a graft from a donor with situs inversus totalis: a case report and review of the literature. Case Rep Transpl. 2013;2013: 532865.
12. Sankarankutty AK, Cagnolati D, Kemp R, Souza FF, Teixeira AC, Mente ED, *et al*. How to do liver transplantation in situs inversus totalis: a simple technique. ANZ J Surg. 2015;85(3):187–8.
13. Tucker O, Prachalias A, Kane P, Rela M. Graft positioning at liver transplantation in situs inversus. Liver Transpl. 2006;12(11):1720–2.
14. Hoyos S, Guzmán C, Correa G, Restrepo JC, Franco H, Cárdenas A. Orthotopic liver transplantation in an adult with situs inversus: an easy way to fit the liver. Ann Hepatol. 2006;5(1):53–5.
15. Klintmalm GB, Bell MS, Husberg BS, Holman MJ, Goldstein RM, Ramsay MA, *et al*. Liver transplant in complete situs inversus: a case report. Surgery. 1993;114(1):102–6.

16. Manzia TM, Tariciotti L, Angelico R, Toti L, Anselmo A, Tisone G. Side-to-side caval anastomosis in a patient receiving a liver graft from a marginal donor with situs inversus totalis. Int J Surg Case Rep. 2015;6:222–5.
17. Kawamoto S, Strong RW, Lynch SV, Ong TH, Pillay SP, Yamanaka J, *et al*. Liver transplantation in the presence of situs inversus totalis: application of reduced-size graft. Liver Transpl Surg. 1995;1(1):23–5.
18. Chun JM, Jung GO, Choi GS, Park JB, Kwon CH, Kim SJ, *et al*. Living donor liver transplantation using a graft from a donor with situs inversus totalis. Liver Transpl. 2009;15(6):666–9.
19. Kamei H, Onishi Y, Ogawa K, Uemoto S, Ogura Y. Living donor liver transplantation using a right liver graft with additional vein reconstructions for patient with situs inversus. Am J Transpl. 2014;14(6):1453–8.
20. Pomposelli J, DaCosta M, McPartland K, Jenkins R. Retroversus implantation of a liver graft: a novel approach to the deceased donor with situs inversus totalis. Am J Transpl. 2007;7(7):1869–71.
21. Rayhill S, Scott D, Orloff S, Horn JL, Schwartz J, Zaman A, *et al*. Orthotopic, but reversed implantation of the liver allograft in situs inversus totalis—A simple new approach to a difficult problem. Am J Transpl. 2009;9(7):1602–6.
22. Ishii T, Fukumitsu K, Ogawa E, Okamoto T, Uemoto S. Living donor liver transplantation in situs inversus totalis with a patient-specific three-dimensional printed liver model. Pediatr Transpl. 2020;24(3): e13675.
23. Asfar S, Ozcay N, Grant D, Wall W. Transplantation of the liver from a donor with complete situs inversus and dextrocardia. Transplantation. 1995;59(3):442–3.
24. Pereira-Lima L, Waechter FL, Sampaio JA, Pinto RD, Álvares-da-Silva MR, de Magalhães FSCF. Liver transplantation in situs inversus. HPB. 2000;2(2):91–3.
25. Heimbach JK, Menon KV, Ishitani MB, Nyberg SL, Jankowski CJ, Lindor KD, *et al*. Living donor liver transplantation using a right lobe graft in an adult with situs inversus. Liver Transpl. 2005;11(1):111–3.
26. Wente MN, Thorn M, Radeleff B, Dei-Anane G, Mehrabi A, Sauer P, *et al*. A routine liver transplantation in a patient with situs inversus: a case report and an overview of the literature. Clin Transpl. 2006;20(2):151–5.
27. Tang DN, Wei JM, Liu YN, Qiao JC, Zhu MW, He XW. Liver transplantation in an adult patient with situs inversus: a case report and overview of the literature. Transpl Proc. 2008;40(5):1792–5.
28. Soria NF, Novoa MG, Polo JR, Sellés CF, de LosÁngelesVázquez M, Milagros MM, *et al*. Orthotopic liver transplantation in an adult with biliary atresia, situs inversus, and inferior cava vein absence: a case report. Transpl Proc. 2015;47:2407.
29. Yankol Y, Mecit N, Kanmaz T, Acarlı K, Kalayoğlu M. Living donor liver transplantation in an adult patient with situs inversus totalis. Ulus Cerrahi Derg. 2015;31(4):232–4.
30. Kauffmann R, Karp SJ, Wright JK, Geevarghese SK. Retroversus implantation of a situs solitus deceased donor liver into a situs inversus totalis recipient. Am Surg. 2017;83(4):e120–2.
31. King DC, Neill JC, Anderson CD, Earl TM, Koller FL. Combined liver and kidney transplantation in a patient with situs inversus. Am Surg. 2019;85(7):e328–9.
32. Na BG, Hwang S, Ahn CS, Moon DB, Song GW, Jung DH, *et al*. Deceased donor liver transplantation in an adult recipient with situs inversus totalis: a case report of 10-year clinical sequences following primary and repeat transplantation. Ann Hepatobil Pancreat Surg. 2020;24(3):319–25.
33. Herrera LA, Castillo J, Martino E, Rabanal JM, Fleitas MG. Orthotopic liver transplantation from a donor with abdominal situs inversus. Transplantation. 1996;62(1):133–5.
34. Dou J, Yang T, Cao JL, Gao QJ, Su YL, Zhu ZJ. Classical orthotopic liver transplantation from a donor with situs inversus totalis using slight rotation of the liver graft. Chin Med J (Engl). 2010;123(10):1353–5.
35. Moon DB, Lee SG, Ha TY. Novel technique of implantation for reversed modified right lobe graft from a donor with situs inversus totalis: new challenge in adult living donor liver transplantation. Transpl Int. 2013;26(10):e88-90.
36. Selvakumar N, Goyal N, Nayeem M, Vohra S, Gupta S. Does situs inversus totalis preclude liver donation in living donor liver transplantation? A series of 3 cases from single institution. Int J Surg Case Rep. 2016;21:23–5.
37. Reimondez S, Alcaraz Á, Giordano Segade E, Agüero Ramaccioni N, Casarez Díaz G, Maraschio MA. Liver transplant from a donor with situs inversus totalis using orthotopic and retroversus technique: a case report. Transpl Proc. 2019;51(2):585–8.
38. Lopez-Andujar R, Andrea B, Javier MH, Cristina B, David C, Vicente I, *et al*. Liver transplantation technique from a ca-daveric donor with situs inversus totalis review and a new case. Japanese J Gstro Hepato. 2021;6(21):1–5.
39. Sugawara Y, Makuuchi M, Takayama T, Yoshino H, Mizuta K, Kawarasaki H. Liver transplantation from situs inversus to situs inversus. Liver Transpl. 2001;7(9):829–30.

Publisher's Note