### **Case report**

# Orthotopic liver transplantation with reduced and rotated graft in adult situs inversus recipient: a case report and a review of reported cases

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#### Summary

Introduction: Situs inversus is a rare congenital anomaly characterized by a mirror-image orientation of abdominal and mostly also thoracic organs. Liver transplantation in these patients is a demanding procedure due to the difficulties pertaining to positioning of the graft and the presence of frequently associated vascular abnormalities. Several reports have been published regarding successful liver transplantation in adult *situs inversus* recipients with different proposed positions of the graft. Relevant experience remains limited.

*Case report:* In this paper we present a case of successful transplantation of a reduced-size cadaverous left hemi-liver graft to an adult *situs inversus* recipient in a 90-degree clockwise rotation. A complex arterial reconstruction was established. A review of published liver transplantations in adult *situs inversus* recipients along with the techniques employed is provided.

*Results:* No vascular or spatial problems were encountered using this technique. The graft function is perfect at 27 months from the transplant procedure. The first liver transplantation with a reduced-size left hemi-liver graft from a *situs solitus* cadaveric donor to the *situs inversus* adult recipient is presented.

*Conclusion:* The devised method of 90-degree clockwise rotation provides perfect spatial adjustment. Relatively smaller grafts are to be preferred as they allow maximum flexibility. Vascular conduits should be readily available.

Key words: situs inversus - liver transplant - reduced graft - position

#### Souhrn

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### Ortotopická transplantace jater redukovaným a rotovaným štěpem dospělému příjemci se situs inversus: kazuistika a přehled publikovaných případů J. Křístek, M. Kočík, J. Chlupáč, J. Froněk

Úvod: Situs inversus je vzácná vrozená anomálie charakterizovaná zrcadlovým umístěním orgánů v dutině břišní a ve většině případů i orgánů hrudníku. Transplantace jater u těchto pacientů je velmi náročná z důvodu časté přítomnosti přidružených cévních abnormalit. Dosud bylo publikováno jen několik příspěvků týkajících se úspěšné transplantace jater dospělému příjemci se situs inversus. Tyto kazuistiky navrhovaly různé polohy štěpu. Zkušenost s danou problematikou je chudá.

Kazuistika: V tomto článku prezentujeme kazuistické sdělení o úspěšné transplantaci redukovaného kadaverozního štěpu levé poloviny jater dospělému příjemci rotovaného o 90° ve směru hodinových ručiček. Bylo nutné provést komplexní rekonstrukci arteriálního zásobení. Kazuistika je doplněna přehledem dosud publikovaných případů transplantace jater dospělému příjemci spolu s použitými technikami.

Výsledky: Použití této techniky nebylo spojeno s žádnými cévními ani prostorovými komplikacemi. 27 měsíců po transplantaci má štěp výbornou funkci.

Závěr: Je prezentována první transplantace levou polovinou redukovaného kadaverozního štěpu jater od dárce se situs solitus příjemci se situs inversus. Použitá metoda 90° rotace štěpu ve směru hodinových ručiček poskytuje výborné prostorové uspořádání. Preferovány jsou relativně menší štěpy, protože poskytují více možností napolohování štěpu. Cévní štěpy by měly být vždy k dispozici. *Klíčová slova:* situs inversus – transplantace jater – redukovaný štěp – poloha štěpu

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### **INTRODUCTION**

The orthotopic liver transplantation (LT) has become a standardized and refined treatment option for various types of liver diseases, both acute and chronic. As an increasing number of patients require and undergo transplantation, more anatomical anomalies of the liver and adjacent vasculature are encountered [1]. These abnormalities may potentially pose technical difficulties and endanger the outcome of both the graft and the patient. One such anomaly is *situs viscerum inversus totalis* (SI). This rare congenital condition represents the mirror-image orientation of both visceral and vascular abdominal and thoracic structures relative to the midline [2]. The anatomically correct position is called *situs viscerum solitus* (SS) [3]. *Situs inversus* presents a major technical challenge to a transplant surgeon for several reasons. Firstly, with the liver being a chiral organ and the donor liver graft most likely coming from a normally arranged donor, there arises a spatial problem with placing the organ in the anatomically non-complementary site of *situs inversus* recipient's hepatic fossa. Such a setting constitutes a risk of hepatic venous outflow kinking and consequential thrombosis [4]. Secondly, *situs inversus* is often related with other, more complex anomalies which may make the situation even more problematic [5], e.g. biliary atresia and other features characteristic for a complex of anomalies called the

polysplenia syndrome (interrupted inferior vena cava with azygous drainage, confluent hepatic veins to the right atrium, preduodenal portal vein, hypoplastic or atretic portal vein, aberrant hepatic arterial anatomy, etc. [2,5,6]. In view of the fact that biliary atresia is the most frequent indication for LT in children and its high co-incidence with SI [7,8], one can easily understand why most of LTs in SI recipients have been performed in children [5,8,9,10]. LT procedures in adult situs inversus recipients are extremely rare [8,9].

Although abdominal *situs inversus* was once considered a contraindication to LT on account of the complexity of the potentially present vascular malformations [2,6,11], refinements of techniques and greater experience with LT have led to reconsideration of this paradigm. Although successful LTs in patients with SI have been described, and it is not deemed a contraindication any more [7,12], the experience with this procedure in SI recipients remains scarce and the related literature is limited. In this paper, we present a successful case of reduced liver graft transplantation to a recipient with *situs viscerum inversus totalis* and a review of the literature concerning the topic.

### **CASE REPORT**

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A 58-year-old Caucasian woman with hepatitis C (HCV) cirrhosis (Child-Turcotte-Pugh A) and situs viscerum inversus totalis presented with a solitary liver tumor consistent with the diagnosis of hepatocellular carcinoma (HCC). The tumor was located in liver segment VII and had a diameter of 25 mm. She had a known history of unsuccessful hepatitis C (HCV) treatment with PEG-interferon alpha-2B and ribavirin; the antiviral therapy was discontinued due to a side effect (depression). Following the diagnosis of HCC she underwent another course of antiviral therapy with sofosbuvir+simeprevir regimen. No signs of generalized malignancy disease or any medical contraindications to LT were revealed. Due to the presence of portal hypertension and liver cirrhosis of a very high degree (45 kPa on shear wave elastography), liver resection was contraindicated. With sustained virologic response at 12 weeks, the patient fulfilled the Milan criteria and was listed for liver transplantation.

In preoperative CT angiography of the recipient, the presence of complete thoracic and abdominal visceral as well as vascular mirror-image orientation was confirmed. The arterial blood supply to the liver originated typically from the celiac axis with no evidence of accessory or replaced hepatic artery. Besides the complete *situs viscerum inversus* including a left-sided *vena cava inferior* and *dextrocardia* (Fig. 1), she had no associated vascular and visceral anomalies. In other words, there were no signs of preduodenal portal vein, intestinal malrotation or intermittent inferior vena cava (IVC). One accessory spleen of 10 mm in diameter was revealed.

At the time of transplantation, her Body Mass Index (BMI) was 20.6 kg/m<sup>2</sup> (height, 162 cm; weight, 54 kg) and her Model for End-Stage Liver Disease score to-

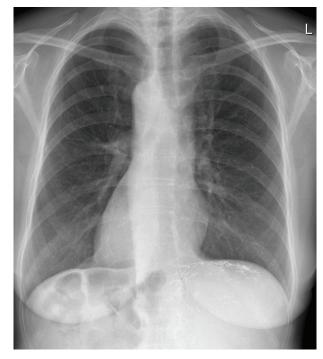


Fig. 1: Postoperative chest X-ray showing dextrocardia (thus complete situs inversus)

Note the hemoclips at the resection plane of the liver under the left portion of the diaphragm.

taled 10. After 6 days of being on the waiting list, an acceptable ABO compatible (A RhD+) 38-year-old male cadaveric SS donor was accepted with BMI of 23 kg/m<sup>2</sup> (height, 178 cm; weight, 73 kg). Due to the donor-recipient body size discrepancy (donor-to-recipient weight ratio 1.35:1) and the presence of SS graft for SI recipient and thus impending complications concerning positioning of the graft, the donor liver was reduced to anatomic left hemi-liver graft (segments II+III+IV). Graft-to-recipient weight ratio was 1% (reduced graft weight 540 g, volume 607 mL). The total donor liver volume before reduction was 1777 mL. In preoperative planning, there were anomalies of hepatic arterial blood supply revealed in cross-sectional contrast imaging studies of the donor. A replaced thick-caliber right hepatic artery (HA) arising from the superior mesenteric artery (SMA), a thin proper hepatic artery supplying segment IV, and a thick-caliber replaced left hepatic artery arising from the left gastric artery were found. The right HA supplied the biliary duct. There were no anomalies regarding hepatic veins and the portal vein (PV). The procurement was done at our institution. The liver was reduced using the *in situ* technique (Fig. 2). The preservation solution used was HTK Solution.

The transplantation itself was begun with a left-sided subcostal incision. Abdominal exploration confirmed the preoperative anatomical findings. The liver was cirrhotic (Fig. 3); no ascites was present. The hepatic hilum structures were carefully dissected. The HA and bile duct were ligated and divided. The PV was temporarily preserved. The liver was dissected free from its attachments. The PV and hepatic veins were divided; the recipient's IVC was preserved and the hepatectomy

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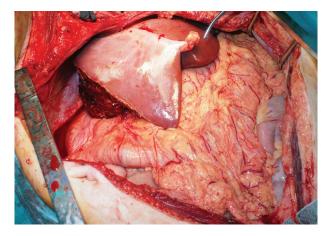


Fig. 2: In situ-reduced left hemi-liver graft during the procurement period

completed (Fig. 4). The graft was tentatively placed in the hepatic fossa rotated 90-degree clockwise. I.e., the resection plane was located under the diaphragm, the left lateral segment pointed to the left hypogastrium, and the hepatic hilum was directed medially. The alignment of the hila in this orientation was perfect (Fig. 5). The cavocavostomy was accomplished using modified piggyback end-to-side technique. The suprahepatic end of the graft IVC was oversewn. The infrahepatic IVC constituted the outflow of the graft, and was sutured to the recipient's left-sided IVC end-to-side. The portal veins were easily approximated without tension and anastomosis was carried out in a standard end-to-end fashion. Reperfusion was homogenous. No venovenous bypass was used during the anhepatic phase. The IVC was side-occluded by Satinsky clamp. Major hemodynamic instability with ventricular tachycardia followed and was present for 10 min. During the back-table period the arterial graft reconstruction was carried out. The celiac axis with Carrel patch had been anastomosed to the SMA, thus all three hepatic arteries of the graft were about to be supplied by the SMA. Due to the fact that there was a significant discrepancy of diameter between the recipient HA and the donor SMA (approximately 1:3), we were forced to construct subdiaphragmatically the aortohepaticostomy, i.e. in fact aortomesentericostomy (Fig. 6). Having cross-clamped the aorta and performed the requisite endarterectomy, the construction of the anastomosis was started with a running Prolene 5-0 suture. However, having finished one side of the anastomosis, we found out that the stitch cut through the aorta. Having started over, we performed the anastomosis with a 4-0 Prolene suture. We observed no perianastomotic bleeding following the aortic declamping. The anastomosis was secured using BioGlue®. The biliary anastomosis was constructed without any problems in the end-to-end fashion with a stent. The abdomen was packed with pads and closed. Neither venovenous bypass nor any adjunctive fixation technique of the graft was used in course of the procedure. The transplantation took 3 hours and 37 minutes. The planned second look operation was done on postoperative day 2. It revealed a source of

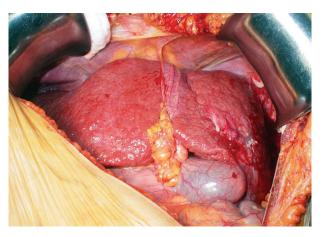


Fig. 3: The situs inversus cirrhotic liver of the recipient before explantation

bleeding at a resection plane and a biliary leak from the biliary anastomosis, both of which were readily taken care of. In further course, there were no significant disturbances to the patient's recovery apart from a temporary renal function impairment that necessitated transitory volume and diuretic therapy; no dialysis was necessary. The patient's liver function recovered quickly and did not show any signs suggestive of the small-for-size syndrome. The patient was discharged on postoperative day 12 with maintenance immunosuppressive regimen comprising of Tacrolimus, Mycophenolate Mofetil (MMF) and Prednisone. No induction immunosuppressive therapy was used.

The explanted liver histology confirmed moderately differentiated (grade 2) hepatocellular carcinoma (pT-2N0MX) with the proportions of 25 x 20 x 20 mm, with microvascular invasion, and with no apparent spread-

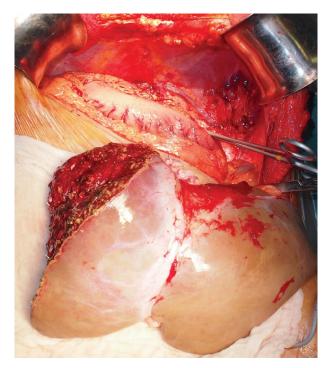


Fig. 4: Obvious spatial discrepancy between the reduced situs solitus graft and the situs inversus hepatic fossa during the anhepatic phase

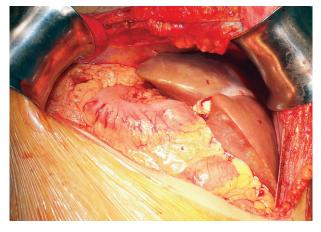


Fig. 5: Probatory placement of the rotated reduced graft in the hepatic fossa

The resection plane is located under the diaphragm; the left lateral segment points to the left hypogastrium. The picture was taken before reperfusion of the graft.

ing through the tumor capsule. No signs of metastatic involvement of lymph nodes were present.

At 5 months follow-up pancytopenia was revealed. Myelotoxic medication including MMF was discontinued and a few courses of granulocyte-colony stimulating factor were applied. The liver enzymes were transitorily elevated. Despite thorough investigation, including a liver biopsy, the causal factor of the liver enzymes elevation was not identified. Control CT an-



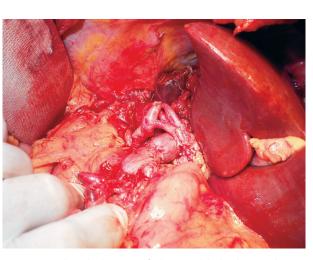


Fig. 6: A detailed view of the established portal vein anastomosis and aortohepatic bypass

The picture was taken after reperfusion of the graft. The biliary duct anastomosis is not clearly visible in this image.

giography confirmed perfectly strong arterial blood supply to the graft. Shortly, the liver tests normalized. In further course, chronic kidney disease grade III (CKD-EPI 34 mL.min-1) developed, probably secondary to calcineurin inhibitor renal toxicity.

At the time of publication, i.e., 27 months after the LT, the patient is in an excellent condition with normal liver function tests, negative HCV viremia and no signs of HCC recurrence (Tab. 1).

Variable	Value of variable				
Diagnosis	HCC, HCV cirrhosis				
MELD score	10 pts				
CTP score	A (5 pts)				
SWE of the liver	F4 (45.3 kPa)				
Blood group, the donor's/the recipient's	A RhD+/ A RhD+				
Cold ischemia time	3 h, 33 min				
Manipulation time (anhepatic phase)	0 h, 34 min				
Surgery length (backtable+explantation+anhepatic+implantation)	3 h, 37 min				
Recipient body characteristics	Weight 54 kg (119 lb), height 162 cm (5 ft 4 in)				
Donor body characteristics	Weight 73 kg (161 lb), height 178 cm (5 ft 10 in)				
Whole donor liver volume according to CT volumometry	1777 mL				
Donor hemi-liver (segments II+III+IV) volume	607 mL				
Donor hemi-liver (segments II+III+IV) weight	540 g				
Graft-to-body weight ratio	0.01 (1%)				
CMV status (recipient/donor)	lgG+/lgG+				
EBV status (recipient/donor)	lgG+/unknown				
Number of PRBC given (in course of the transplant procedure)	2				
Intraoperative blood loss	Approx. 1200 mL				
Volume of autotransfusion	600 mL				
Vasopressoric (Norepinephrine) support	Up to 0.7 mcg/kg/min				
LOS at the hospital	12 days				
LOS at the ICU	6 days				
Length of orotracheal intubation	3 days				
Induction immunosuppression	None				
Maintenance immunosuppression	Tacrolimus-Mycophenolate Mofetil-Prednisone				

Notes: CM – cytomegalovirus; CT – computed tomography; CTP – Child-Turcotte-Pugh; EBV – Epstein-Barr virus; h – hour(s); HCC – hepatocellular carcinoma; HCV – hepatitis C virus; ICU – Intensive Care Unit; LOS – length of stay; mcg, microgram(s); MELD – Model for End-Stage Liver Disease; min – minute(s); PRBC – packed red blood cells; pts – points; SWE – shear wave elastography.

### DISCUSSION

According to a review of Farmer and Busuttil [9], there have been overall 90 transplantations in recipients with SI and/or polysplenia syndrome described in English written literature until 2015. Most of the recipients were children with biliary atresia and associated SI. In a mere 12 of these 90 cases the recipients were adults with situs inversus. In other words, LT procedures in adult SI recipients are extremely rare [8,9]. Since the Farmer's and Busuttil's publication, there have been 5 more case reports of LT in SI adult recipients published [2,6,8,13,14]. In 5 out of these 17 cases, the adult SI recipients received a reduced-size graft. However, all of these grafts came from living-related donors (Tab. 2). To the best of our knowledge, this is the first case that describes a liver transplantation with a reduced-size graft from a cadaveric SS donor to an adult SI recipient.

In the presented case, the patient suffered from HCC in a cirrhotic liver. Due to the presence of portal hypertension and a very high degree of liver cirrhosis, liver resection was contraindicated. The patient met the Milan criteria for LT. The patient was on the list for not more than a few days when a potentially suitable donor was identified. However, there was a size discrepancy present between the donor and the recipient. In regard to the recipient's relatively small body proportions (weight 54 kg, height 162 cm) and her diagnosis of HCC, it could be tricky to wait for an optimal whole graft not requiring a reduction. Of course, another possibility may have been to treat the patient with locoregional therapy while she was waiting for the adequate full-size graft. However, we preferred to accept this relatively large graft for this recipient and to reduce it to a left hemi-liver graft.

Since patients with SI make up a heterogeneous group with respect to often accompanying abdominal and/or cardiopulmonary malformations, there is no standard surgical technique of LT in this setting. For this reason, one of the most important technical aspects of LT in patients with SI remains resourcefulness of the surgeon [5]. The performing surgeon must be ready to apply any of a multitude of possible reconstructive techniques during the procedure. All in all, smaller grafts permit flexibility and donor-to-recipient weight ratio of <1 has been described as optimal for whole-organ donors. Also segmental grafts (either a deceased-donor, or living-related) are frequently used and allow easier placement in the abdominal cavity [5]. Thus, careful donor selection including precise size matching is of paramount importance [5]. According to the literature, pediatric patients are expected to exhibit less graft displacement and hepatic venous torsion on account of their relatively smaller abdominal cavity, even in cases where split and reduced grafts are used [5]. On the contrary, in adults following a hepatectomy, a large empty space can be formed in the left upper quadrant which predisposes the graft to lateral displacement with supero-lateral rotation and torsion of the hepatic venous pedicle [5,15]. A particularly crucial point to be noted

is that vascular conduits should always be available [6], which is a significant advantage of cadaverous grafts and at the same time, a considerable limitation to living donor grafts. In our case, although we had to construct an arterial bypass there was no need of using vascular conduits. Due to the discrepancy between the caliber of the recipient's and graft's HA (the risk of thrombosis) we had to establish an aortohepaticostomy. A perfect patency of the anastomosis without signs of stenosis was confirmed with CT angiography 5 months later. There are several other aspects worth mentioning regarding the preoperative planning. Due to the high incidence of associated cardiopulmonary abnormalities, evaluation of the functional reserve should be considered. An extensive preoperative recipient anatomy work-up including contrast-enhanced cross-sectional imaging is a matter of course, as well as the previously mentioned meticulous size-matching [16]. The hemodynamic instability in course of the anhepatic phase we observed in this case may have been preventable by establishment of a temporary venovenous bypass.

The abovementioned will probably not provoke any discussion, but it is the placement and orientation of the SS liver graft in an SI recipient which is a matter of debate. To date, several possible techniques have been described in the literature. By and large, the location of the recipient's suprahepatic IVC or hepatic veins (left- or right-sided in relation to the vertebral column) will most likely determine the position of the graft. In other words, positioning of the graft will most probably differ in the recipients with dextrocardia from those with levocardia. Most of the patients with situs inversus have dextrocardia and left-sided IVC (situs inversus totalis), which means – in the case of using the midline position ("orthotopic") method – that the right lobe of the SS whole graft will occupy the midline, where the space is scarce due to the presence of the vertebral column and the stomach. Surprisingly and somewhat paradoxically, in most of the cases reported, the graft was placed in this way. The problem with this position is that the anatomical right lobe overlies the recipient's stomach and could possibly cause IVC compression and, most importantly, rotation of the graft with aforementioned consequences. The second most common technique reported used a segmental allograft, either from a living related donor or a cadaveric one. Other alternatives have been infrequent. Klintmalm et al. ingeniously invented a technique of placing the graft rotated 90-degrees clockwise around its anteroposterior axis [17]. Using this method, the larger right lobe fits better the hepatic fossa; the left lateral segment points to the left lower quadrant and the hepatic hilum is in favorable syntopy with the structures of the hepatoduodenal ligament. A modified hepatic outflow needs to be set up between the graft's infrahepatic and recipient's IVC in the end-toside fashion. In contrast with the midline position, using the Klintmalm's 90-degree rotation method, the risk of the laterodisplacement of the graft and consequent kinking of the venous outflow is minimized.

#### Tab. 2: All published experience with liver transplant in situs inversus adult recipients

Image: Probability of the second se	rab. 2. All published experience with liver transplant in situs inversus adult recipients											
2 Wente et al. 20 2006 48 M orthotopic ETOH CAD SS, whole none   3 Tucker et al. 15 2006 41 M midline shift position, Sengstaken-Blakemore tube support, Giaphragm plication, faic-form ligament fixation ETOH CAD SS, whole none   4 Barone et al. 1 1992 17 F midline shift position CHF CAD SS, whole none   5 Kilintmalm et al. 17 1993 45 F 90° clockwise rotation ETOH CAD SS, whole none   6 Farmer et al. 9 N/A 37.5 M N/A HCV unk unk none   7 Rayhill et al. 10 2005 53 F 90° clockwise rotation CCC CAD SS, whole none   9 Hoyos et al. 21 2006 41 M 90° clockwise rotation CCC CAD SS, whole none   10 Tang et al. 12 2008 54 F orthotopic HBV </th <th></th> <th>Team</th> <th>Reference number</th> <th>Year</th> <th>Age</th> <th>Sex</th> <th>Technique, adjunct maneuvers</th> <th>Diagnosis</th> <th>Donor type</th> <th>Graft type</th> <th>Complica- tions</th> <th>Outcome</th>		Team	Reference number	Year	Age	Sex	Technique, adjunct maneuvers	Diagnosis	Donor type	Graft type	Complica- tions	Outcome
3 Tucker et al. 15 2006 41 M midline shift position, sengstaken-Blakemore tube support, diaphragm plication, falci-form ligament fixation ETOH CAD SS, whole none   4 Barone et al. 1 1992 17 F midline shift position CHF CAD SS, whole none   5 Klintmalm et al. 17 1993 45 F 90° clockwise rotation ETOH CAD SS, whole none   6 Farmer et al. 9 N/A 37.5 M N/A HCV unk. unk. none   7 Rayhill et al. 10 2009 53 F 180° rotation around vertical axit (flip, facing backwards, reversed) PEC CAD SS, whole none   9 Hoyos et al. 21 2006 41 M 90° clockwise rotation CSC CAD SS, whole none   9 Hoyos et al. 21 2006 41 M 90° clockwise rotation CCC CAD SS, whole none   10 Tang et al. 12 2006 54 <th>1</th> <th>Watson et al.</th> <td>19</td> <td>1995</td> <td>35</td> <td>unk.</td> <td>orthotopic</td> <td>CC</td> <td>CAD</td> <td>SS, whole</td> <td></td> <td>A, 7 mo</td>	1	Watson et al.	19	1995	35	unk.	orthotopic	CC	CAD	SS, whole		A, 7 mo
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5 Klintmalm et al. 17 1993 45 F 90° clockwise rotation ETOH CAD SS, whole none   6 Farmer et al. 9 N/A 37.5 M N/A HCV unk. unk. unk. none   7 Rayhill et al. 10 2009 53 F 180° rotation around vertical axis (flip, facing backwards, reversed) PBC CAD SS, whole none   8 Heimbach et al. 7 2005 62 M midline shift position, oronentum and hepatic flexure to hepatic fossa PSC LRD SS, whole none   9 Hoyos et al. 21 2006 41 M 90° clockwise rotation CCC CAD SS, whole none   10 Tang et al. 12 2008 45 M 45° clockwise rotation SBC CAD SS, whole none   11 Soejima et al. 5 2008 19 F midline shift position, faciform ligament fixtula HCV LRD SS, RPS HVOO   13 Kim et al. 2 2016 <th< th=""><th>3</th><th>Tucker et al.</th><th>15</th><th>2006</th><th>41</th><th>Μ</th><th>Sengstaken-Blakemore tube support, dia- phragm plication, falci-</th><th>ETOH</th><th>CAD</th><th>SS, whole</th><th>none</th><th>A, 17 mo</th></th<>	3	Tucker et al.	15	2006	41	Μ	Sengstaken-Blakemore tube support, dia- phragm plication, falci-	ETOH	CAD	SS, whole	none	A, 17 mo
6Farmer et al.9N/A37.5MN/AHCVunk.unk.unk.none7Rayhill et al.10200953F180° rotation around vertical axis (flip, facing backwards, reversed)PBCCADSS, wholenone8Heimbach et al.7200562Mmidline shift position, omentum and hepatic flexure to hepatic fossaPSCLRDSS, Rbiliary strx9Hoyos et al.21200641M90° clockwise rotationCCCCADSS, wholenone10Tang et al.21200845M45° clockwise rotationSBCCADSS, LLOBEwound infection, intestinal fistulation11Soejima et al.5200819Fmidline shift position, slight clockwise rotationHCVLRDSS, LLOBEwound infection, intestinal fistulation12Kim et al.4201054ForthotopicHBVLRDSS, RPSHVOO13Kamei et al.13201460M180° rotation around vertical axis (flip, facing backwards, reversed), claphragm plicationrecurrent cholis post LT, primarilyGADSS, wholenone14Yu et al.2201453MSI graft, reTx, ABOi, reversed), claphragm plicationBAAGADSS, wholenone15Tabrizian et al.6201534MorthotopicBAGADSD, wholenone16Soria et al. <th>4</th> <th>Barone et al.</th> <td>1</td> <td>1992</td> <td>17</td> <td>F</td> <td>midline shift position</td> <td>CHF</td> <td>CAD</td> <td>SS, whole</td> <td>none</td> <td>A, 6 mo</td>	4	Barone et al.	1	1992	17	F	midline shift position	CHF	CAD	SS, whole	none	A, 6 mo
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Image: Section of the secting of the secting of the secting of th	6	Farmer et al.	9	N/A	37.5	М	N/A	HCV	unk.	unk.	none	A, 50 mo
Image: Section of the section of th	7	Rayhill et al.	10	2009	53	F	vertical axis (flip, facing	PBC	CAD	SS, whole	none	A, 36 mo
10Tang et al.12200845M45° clockwise rotationSBCCADSS, wholenone11Soejima et al.5200819Fmidline shift position, slight clockwise rotationHCVLRDSS, L LOBE (SI-HII)wound infection, intestinal fistula12Kim et al.4201054ForthotopicHBVLRDSS, RPSHVOO13Kamei et al.13201460M180° rotation around vertical axis (flip, facing bckwards, reversed), diaphragm plicationHBVLRDSS, R RDS, RPNone14Yu et al.2201453MSI graft, reTx, ABDi, piggyback E-Srecurrent robiggyback E-SCAD (DBD)SI, wholeNone15Tabrizian et al.6201523Fmidline shift positionBA RDCAD (DBD)SS, wholenone16Soria et al.14201534MorthotopicBA RDCAD (DBD)SS, wholenone15Tabrizian et al.6201523Fmidline shift positionBA RDCAD (DBD)SS, wholeabdominal compart- ment sy, pulmonary herroring herroringSS, wholesoftinia abbcos singitis primarily HCC in HBVSS, wholeabdominal compart- ment sy, pulmonary herroring herroring herroring abbcos singitisSS, wholesoftinia abbcos singitis pulmo abbcos singitis14Soria et al.1	8		7	2005	62	Μ	omentum and hepatic flexure to hepatic fossa	PSC	LRD	HEMI-LIV- ER (SV-VIII)	biliary strx.	A, 6 mo
11Soejima et al.5200819Fmidline shift position, slight clockwise rota- tion, falciform ligament fixationHCVLRDSS, L LOBE (SII+III)wound infection, infection, 	9	Hoyos et al.	21		41	М	90° clockwise rotation	CC	CAD		none	A, 21 mo
Image: Series of the series	10	Tang et al.	12	2008	45	М	45° clockwise rotation	SBC	CAD	SS, whole	none	A, 24 mo
13Kamei et al.13201460M180° rotation around vertical axis (flip, facing backwards, reversed), diaphragm plicationHBVLRDSS, R HEMI-LIV- R (SV-VIII)none14Yu et al.2201453MSI graft, reTx, ABOi, piggyback E-Srecurrent chol- angitis post LT, primarily HCC in HBVCAD (DBD)SI, wholenone15Tabrizian et al.6201523Fmidline shift positionBACAD (DBD)SS, wholenone16Soria et al.14201534MorthotopicBACAD (DBD)SS, whole abdominal compart- ment sy, pulmonary hemorrhage intra-abdom inal abscess strokeSS, whole abdominal compart- ment sy, pulmonarySS, whole abdominal compart- ment sy, pulmonary	11	Soejima et al.	5	2008	19	F	slight clockwise rota- tion, falciform ligament	HCV	LRD		infection, intestinal	A, 12 mo
Image: series of the series	12	Kim et al.	4	2010	54	F	orthotopic	HBV	LRD	SS, RPS	HVOO	A, 8 mo
Image: Serie and Series	13	Kamei et al.	13	2014	60	М	vertical axis (flip, facing backwards, reversed),	HBV	LRD	HEMI-LIV-	none	A, 50 mo
Image: Soria et al. 14 2015 34 M orthotopic BA CAD (DBD) SS, whole (DBD) abdominal compart- ment sy, pulmonary hemorrhage intra-abdom inal abscess stroke	14	Yu et al.	2	2014	53	Μ		chol- angitis post LT, primarily HCC in		SI, whole	none	A, 11 mo
(DBD) compart- ment sy, pulmonary hemorrhage intra-abdom inal abscess stroke	15	Tabrizian et al.	б	2015	23	F	midline shift position	BA		SS, whole	none	A, 20 mo
17 Vankol et al. 8 2015 18 E orthotopic slight AIH LRD SS LLORE none	16	Soria et al.	14	2015	34	Μ	orthotopic	BA		SS, whole	ment sy, pulmonary hemorrhage, intra-abdom- inal abscess,	A, unk.
Notes: A – alive; ABQi – ABQ incompatible; AIH – autoimmune hepatitis; BA – biliary atresia; CAD – cadaveric donor; CC – cryptogenic c	17	Yankol et al.	8			F	triangulated suture technique of hepatic vein anastomosis	AIH	LRD		none	A, 20 mo

Notes: A – alive; ABOi – ABO incompatible; AlH – autoimmune hepatitis; BA – biliary atresia; CAD – cadaveric donor; CC – cryptogenic cirrhosis; CHF – congenital hepatic fibrosis; DBD – donor after brain death; E-S – end-to-side; ETOH – ethylism; F – female; HA – hepatic artery; HBV – hepatitis B virus; HCC – hepatocellular carcinoma; HV – hepatic vein; HVOO – hepatic venous outflow obstruction; LLOBE – left lobe; LRD – living related donor; LT – liver transplant; M – male; mo – month(s); n – normal; N/A – not available; PBC – primary biliary cirrhosis; PV – portal vein; R – right; reTx – retransplantation; RHV – right hepatic vein; R LOBE – right lobe; RPS – right posterior sector; SBC – secondary biliary cirrhosis; SI – situs inversus; SS – situs solitus; SII+III – liver segments II and III; SV-VIII – liver segments V-VIII; sy – syndrome; unk. – unknown ۲

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Also the recipient's stomach is not compromised by the donor right lobe. In our case, we made use of this method and it proved very helpful. We did not face any problems concerning this position. The hepatic hila were perfectly aligned and cavocavostomy was also constructed conveniently. Another technique described by Todo et al. mentions an auxiliary LT in a patient with complex vascular anomalous anatomy [18]. A fascinating solution to this problem was devised by Rayhill et al. placing the graft orthotopically, but reversely, i.e. 180-degree rotated around the axis of the IVC (facing backwards) [10]. The hepatic hilum faced anteriorly, well aligned with the recipient SI hepatoduodenal structures. Hepatic outflow was established using reversed cavaplasty (end-to-side anastomosis between suprahepatic IVCs).

The potentially most threatening peril of the midline position in SI LT recipients is the lateral displacement or a rotation of the graft and torsion of the venous pedicle with consequent thrombosis and impending failure of the graft. According to the literature, in an attempt to solve this anatomically unfavorable condition of empty hepatic fossa, several maneuvers have been proposed and used: 1) Plication of the diaphragm [15]; 2) Temporary placement of the inflated Sengstaken-Blakemore tube in the hepatic fossa [15]; 3) Fixation of the falciform ligament to the diaphragm [15]; 4) Filling the fossa up with omentum [7,15]; and 5) Mobilizing the hepatic flexure and positioning it in the relatively empty hepatic fossa [7,15]. We feel obliged to mention that we have no experience with using any of these methods apart from the method number 3.

Postoperative management of these patients does not differ significantly from that required for other patients undergoing LT. However, medical teams must be aware of the anatomical variety present. This is of importance in postoperative radiographic studies and, first of all, in invasive procedures such as liver biopsy.

### CONCLUSION

In this paper, we present the first liver transplantation with reduced-size left hemi-liver graft from a situs solitus cadaveric donor to the situs inversus adult recipient. The graft was placed orthotopically 90-degree clockwise rotated and a complex reconstruction of the arterial blood supply was performed. There were no difficulties regarding positioning of the graft, nor were any vascular abnormalities in the postoperative course encountered. Generally speaking, the experience with this type of operation is rare. In our opinion, smaller grafts are to be preferred as they allow maximum flexibility. Vascular conduits should be readily available. The situs inversus is not a contraindication to liver transplantation. However, thorough preoperative planning is a matter of course because recipients with this anomaly are often afflicted with other cardiac and non-cardiac malformations.

List of abbreviations

- BMI body mass index
- CT computed tomography
- HA hepatic artery
- HCC hepatocellular carcinoma
- HCV hepatitis C virus
- IVC inferior vena cava
- LT liver transplantation
- MMF mycophenolate mofetil
- PV portal vein
- SI situs viscerum inversus
- SMA superior mesenteric artery
- SS situs viscerum solitus

#### Conflict of interests

The authors declare that they have not conflict of interest in connection with the emergence of and that the article was not published in any other journal.

#### REFERENCES

- Barone GW, Henry ML, Elkhammas EA, et al. Orthotopic liver transplantation with abdominal situs inversus and dextrocardia. Am Surg. 1992;58:651–3.
- Yu S, Guo H, Zhang W, et al. Orthotopic liver transplantation in situs inversus adult from an ABO-incompatible donor with situs inversus. BMC Gastroenterol 2014;14:46.
- Tawfik AM, Batouty NM, Zaky MM, et al. Polysplenia syndrome: a review of the relationship with viscero-atrial situs and the spectrum of extra-cardiac anomalies. Surg Radiol Anat 2013;35:647–53.
- Kim BW, Bae BK, Xu W, et al. Living donor liver transplantation for an adult patient with situs inversus totalis. World J Gastroenterol 2010;16:2311–3.
- Soejima Y, Meguro M, Taketomi A, et al. Left lobe living donor liver transplantation in an adult patient with situs inversus: technical considerations. Transpl Int 2008;21:384–9.
- 6. Tabrizian P, Joseph TT, Radkani P, et al.

Liver transplantation in an adult recipient with situs inversus totalis: Case report and review of the literature. Transplant Proc 2016;48:3163–6.

- Heimbach JK, Menon KV, Ishitani MB, et al. Living donor liver transplantation using a right lobe graft in an adult with situs inversus. Liver Transpl 2005;11:111–3.
- Yankol Y, Mecit N, Kanmaz T, et al. Living donor liver transplantation in an adult patient with situs inversus totalis. Ulus Cerrahi Derg. 2015;31:232–4.
- Farmer DG, Busuttil RW. Transplantation: situs inversus and polysplenia syndrome. In: Busuttil RW, Klintmalm GB. Transplantation of the liver. Third edition. ed. Philadelphia, PA: Elsevier/ Saunders 2015.
- Rayhill SC, Scott D, Orloff S, et al. Orthotopic, but reversed implantation of the liver allograft in situs inversus totalis-a simple new approach to a difficult problem. Am J Transplant

2009;9:1602-6.

- 11. Lilly JR, Starzl TE. Liver transplantation in children with biliary atresia and vascular anomalies. J Pediatr Surg 1974;9:707–14.
- Tang DN, Wei JM, Liu YN, et al. Liver transplantation in an adult patient with situs inversus: a case report and overview of the literature. Transplant Proc 2008;40:1792–5.
- Kamei H, Onishi Y, Ogawa K, et al. Living donor liver transplantation using a right liver graft with additional vein reconstructions for patient with situs inversus. Am J Transplant 2014;14:1453–8.
- 14. Fernandez Soria N, Garcia Novoa MA, Rivas Polo JI, et al. Orthotopic liver transplantation in an adult with biliary atresia, situs inversus, and inferior cava vein absence: A case report. Transplant Proc 2015;47:2407–9.
- 15. Tucker O, Prachalias A, Kane P, et al. Graft positioning at liver transplantation in situs inversus. Liver Transpl

2006;12:1720-2.

- Farmer DG, Shaked A, Olthoff KM, et al. Evaluation, operative management, and outcome after liver transplantation in children with biliary atresia and situs inversus. Ann Surg 1995;222:47–50.
- Klintmalm GB, Bell MS, Husberg BS, et al. Liver transplant in complete situs inversus: a case report. Surgery 1993;114:102–6.
- Todo S, Hall R, Tzakis A, Starzl TE. Liver transplantation in patients with situs inversus. Clin Transplant 1990;4:5–8.
- Watson CJ, Rasmussen A, Jamieson NV, et al. Liver transplantation in patients with situs inversus. Br J Surg 1995;82:242–5.
- Wente MN, Thorn M, Radeleff B, et al. A routine liver transplantation in a patient with situs inversus: a case report

and an overview of the literature. Clin Transplant 2006;20:151–5.

- 21. Hoyos S, Guzman C, Correa G, et al. Orthotopic liver transplantation in an adult with situs inversus: an easy way to fit the liver. Ann Hepatol 2006;5:53–5.
- 22. Raynor SC, Wood RP, Spanta AD, et al. Liver transplantation in a patient with abdominal situs inversus. Transplantation 1988;45:661–3.

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### Nekrolog

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### Vzpomínka na primáře Vladimíra Černého

Dne 25. 7. 2018 po dlouhé nemoci zemřel ve věku nedožitých 83 let MUDr. Vladimír Černý. Odešla tak významná osobnost plzeňské a západočeské chirurgie, nesmírně obětavý lékař, jemný a precizní chirurg, který ovlivnil několik generací svých žáků.

MUDr. Černý se narodil v r. 1935 v Topolčanech na Slovensku. Absolvoval Gymnázium Jaroslava Vrchlického v Klatovech a rozhodl se pro stadium medicíny. V roce 1954 nastoupil na Lékařskou fakultu Univerzity Karlovy v Plzni. Po jejím absolvování začal pracovat na chirurgickém oddělení sušické nemocnice, kde se pod vedením primáře J. Vovsa aktivně podílel na zavádění progresivních metod v anestezii, chirurgii i pooperační péči. Po smrti primáře převzal s úspěchem vedení oddělení.

Po invazi v r. 1968 odešel z politických důvodů ze sušického primariátu do Plzně, kde se záhy začal specializovat na dětskou chirurgii. V r. 1978 z ní složil atestaci a v r. 1979 se stal primářem oddělení dětské chirurgie. Politická situace v období normalizace mu však nedovolila plně rozvinout jeho vědecké ambice, zabránila vycestovat za zkušenostmi do zahraničí a nedostalo se mu ani uznání za jeho úspěšné pedagogické působení. Až listopadové události 1989 umožnily primáři Černému plně rozvinout jeho občanskou angažovanost. Aktivně a svědomitě působil v České lékařské komoře. Začátkem devadesátých let výrazně pomáhal při konsolidaci personální situace na plzeňské klinice.

V roce 1993 přijal místo primáře chirurgie Mulačovy nemocnice Plzeň, kde se věnoval zavádění a rozvoji moderních operačních technik, především laparoskopických. Svoje působení v Mulačově nemocnici ukončil až v roce 2009, tedy ve svých 74 letech.

V posledním roce života poznal zdravotnictví z druhé strany. Svoji neuroonkologickou diagnózu nesl velmi statečně.

Vladimír Černý byl člověk širokého rozhledu a řady zájmů. Miloval literaturu, hudbu, především jazz a swing, a uměl překvapit, když vzal do ruky tenor saxofon, na který v mládí hrával. Byl nejen výjimečný lékař a chirurg, ale i přítel a učitel, vzácný člověk s velkým srdcem, kterého lze bez rozpaků označit za "umělce života".

Čest jeho památce

MUDr. Jaroslav Špatenka, CSc. prof. MUDr. Jiří Valenta, DrSc. MUDr. Vladislav Frühauf

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