Specificities of transplantation of kidneys procured from donors with situs inversus totalis – A case report and review of the literature


*Clinic of Nephrology, †Clinic of Anaesthesia and Intensive Care, ‡Clinic of Urology, †Clinic of Vascular and Endovascular Surgery, Military Medical Academy, Belgrade, Serbia; †Faculty of Medicine of the Military Medical Academy, University of Defence, Belgrade, Serbia

Abstract

Introduction. Situs inversus totalis (SIT) represents a total vertical transposition of the thoracic and abdominal organs which are arranged in a mirror image reversal of the normal positioning. We presented a successful pre-dialysis kidney transplantation from a living sibling donor with SIT and the longest donor follow-up period, along with analysis of the reviewed literature. Case report. The pair for pre-dialysis kidney transplantation included a 68-year-old mother and 34-year-old daughter at low immunological risk. Comorbidities evidenced in kidney donors with previously diagnosed SIT, included moderate arterial hypertension and borderline blood glucose level. Explantation of the left donor kidney and its placement into the right iliac fossa of the recipient were performed in the course of the surgical procedure. A month after nephrectomy, second degree renal failure was noticed in the donor. A 20-month follow-up of the donor’s kidney and graft in the recipient proved that their functions were excellent.

Conclusion. In donors with previously diagnosed SIT the multidisciplinary approach, preoperative evaluation of the patient and detection of possible vascular anomalies are required to provide maximum safety for the donor.

Key words: situs inversus; kidney transplantation; tissue donors.

Correspondence to: Milica Petrović, Clinic of Nephrology, Military Medical Academy, Crnotravska 17, 11 000 Belgrade, Serbia.
Phon.: +381 65 2511 964, E-mail: drmilicapetrovic@gmail.com
in reverse position. When all organs change their positions, the connections and communication between them remain undisturbed, and thus SIT patients are mostly asymptomatic and have normal lifespan 1–7.

SIT was an absolute contraindication for organ donation, particularly for donation of the liver and heart, having in mind associated anomalies of blood vessels appearing in more than 40% of cases 8. Our paper presented a SIT patient as a living kidney donor as well as our previous experiences with kidney transplantations involving donors with SIT.

Case report

Pre-transplantation evaluation of the kidney donor – the recipient’s 68-year-old mother, was commenced after initial examination performed in transplantation outpatient unit, revealing no immunological complications (the same blood group A Rh D, 5/9 HLA match and negative crossover match according to complement-dependent cytotoxicity (CDC). As for the comorbidities, the donor had arterial hypertension corrected by angiotensin-converting enzyme inhibitor (ACE) and borderline blood glucose levels. Physical examination performed on admission evidenced body mass index (BMI) 29.1 kg/m², with normal findings according to organ systems. The obtained results of laboratory tests were within reference values, kidney function was normal as well as virological tests, including hepatitis and HIV markers (Table 1).

The diagnosis of SIT was confirmed by heart and lung radiography (Figure 1), electrocardiography (Figure 2) and echocardiography. Abdominal ultrasonography (US) revealed the spleen below the right costal arch and the liver below the left costal arch, as well as the kidneys of normal size and echogenicity, without hydronephrosis and calculosis. Renal and pelvic multislice computed tomography (MSCT) angiography evidenced two arteries of the right kidney with ostial stenosis of the lower pole artery. The left kidney was vascularized by a single artery. One renal vein was present on each side. Pelvic blood vessels were free of any anomalies (Figure 3). Dynamic scintigraphy of the kidneys with separate creatinine clearance evi-

### Table 1

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Before nephrectomy</th>
<th>One month after nephrectomy</th>
<th>One year after nephrectomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum creatinine [μmol/L]</td>
<td>62</td>
<td>82</td>
<td>103</td>
</tr>
<tr>
<td>Serum urea [mmol/L]</td>
<td>5.9</td>
<td>8.9</td>
<td></td>
</tr>
<tr>
<td>GFR MDRD [mL/min/1.73 m²]</td>
<td>85.5</td>
<td>64.2</td>
<td>62.3</td>
</tr>
<tr>
<td>24-hours proteinuria [g]</td>
<td>0.032</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Microalbuminuria [mg/mL]</td>
<td>10</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

GFR MDRD – glomerular filtration rate according to modification of diet in renal disease

Fig. 1 – Chest radiography: dextrocardia.

Fig. 2 – Electrocardiogram-dextrogram.

Fig. 3 – Donor’s multislice computed tomography with angiography.
The kidney recipient was a 34-year-old female patient with preterminal renal failure resulting from chronic glomerulonephritis [serum creatinine 508 μmol/L, urea 32 mmol/L, glomerular filtration rate (GFR) according to modification of diet in renal disease (GFR MDRD) 12.6 mL/min/L·73 m²].

On the pre-transplantation meeting it was decided to explant the left kidney of the donor and implant it in the right iliac fossa of the recipient. Standard explantation of the left kidney was performed and it was perfused with 1000 mL of Euro-Collins solution. Warm ischemia lasted for 2 minutes, while cold ischemia and rewarmed time lasted 20 minutes. The kidney was positioned in the recipient’s right iliac fossa using the standard surgical procedure: end-to-end anastomosis of the renal and hypogastric arteries, end-to-side anastomosis of the renal vein and external iliac vein and ureterocystoneostomy performed using the method of two parallel incisions along with “JJ” probe placement.

Postoperatively, the donor was in good general condition, with daily diuresis of 2,000 mL, normotensive with regular antihypertensive therapy, without complaints. Laboratory tests performed in early postoperative period on day 0 revealed the serum creatinine value of 58 μmol/L, being 118 μmol/L on the postoperative day 14 – on discharge. A month after the nephrectomy, second degree renal failure was noticed (Table 1). Abdominal ultrasound examination revealed normal solitary kidney and other organ findings. Regular nephrologic follow-up examinations did not evidence deterioration of the renal function within 1-year period after donor nephrectomy (Table 1). Follow-up US examination performed after one year revealed the following: the remaining right kidney was sized 11.3 × 4.8 cm, with 1.7 cm thick parenchyma, normal echogenicity, without stasis or calculosis.

Postoperative recipient's diuresis was 16,500 mL on the day of transplantation to be gradually decreased and maintained at 4,600 mL at the average, over the 2-week postoperative period. In the 14-day posttransplantation period, during hospitalization, serum creatinine values ranged between 102 μmol/L and maximal 176 μmol/L, without any signs of acute transplant rejection. Acute exacerbation of the chronic renal failure with transient increase in the serum creatinine coincided with an episode of urinary infection caused by Klebsiella species that was managed by susceptibility test-based antibiotic therapy, i.e. carbapenem in a total dose of 15 grams.

Quadruple immunosuppressive therapy was applied for prevention of rejection. Induction was performed by intraoperative administration of antithymocyte globulin (ATG) in the dose of 8 mg/kg, along with the triple therapy according to immunosuppression protocol: glucocorticoids, tacrolimus and mycophenolate mofetil. Ultrasound examination of the transplanted kidney (graft) performed on discharge, on the postoperative day 14, revealed normal findings (Figure 4). The kidney was sized 11.6 × 4.4 cm, with homogenous 1.7 cm thick parenchyma, without hydronephrosis or calculus. Vasculature was well-defined up to the smallest branches on the periphery, with the resistive index (RI) in the interlobar and iliac arteries of 0.66.

Laboratory tests, indicative of renal function parameters performed by the end of the first year confirmed the optimal graft function (serum creatinine 114 μmol/L, GFR MDRD 61.29 mL/min/1.73 m², Biuret 0.115 g/24h/ proteinuria). Ultrasound examination of the transplanted kidney/graft evidenced identical parameters to those obtained upon the previous examination with somewhat less pronounced arterial circulation which was normally tracked up to the level of Malpighian pyramids, and more difficultly peripherally, with refractive index at the level of interlobar arteries of 0.72, considered to be normal finding for transplanted kidney (Figure 5). Stasis was not evidenced in the excretory system.

**Discussion**

SIT was one of the absolute contraindications for organ donation till 1988. SIT is a rare, congenital anomaly that may be associated with vascular and visceral abnormalities and intestinal malformations. It is also associated with other anomalies such as polysplenia, Ivemark-Kartagener syndrome and biliary atresia. Due to a possible onset of complications within the associated anomalies, a particular attention was also paid to it in other surgical interventions, particularly in abdominal surgery. However, the attitude has been changed over the last several years. Few case reports on patients with situs inversus who had undergone cholecystectomy, distal gastrectomy...
due to the gastric carcinoma using the laparoscopic method, gastrectomy with cholecystectomy, liver resection, splenectomy and lung tumor have been published so far\textsuperscript{17-24}.

In transplantation medicine, SIT represents a potential risk for possible complications, particularly in case of liver and heart transplantsations. In 1988 Raynor et al.\textsuperscript{25} described liver transplantation in a patient with \textit{situs inversus}. Thereafter, other successful liver transplantations were described as well\textsuperscript{25,26}. In 1990 Doty et al.\textsuperscript{27} described cardiac transplantation in a patient with SIT, while Rabago et al.\textsuperscript{28} described in 1996 heart-lung transplantation in a patient with SIT.

A review of the reference literature indicated that the total of 6 transplantations were performed with the kidney procured from donors with SIT\textsuperscript{8,29-31} (Table 2). The first case of kidney transplantation from a donor with SIT was published in 2003\textsuperscript{29}. Four transplantations were performed with kidneys obtained from living donors with SIT while two were performed with organs procured from cadaveric donors with SIT (Table 2). Polak et al.\textsuperscript{8} in 2006 reported a case of successful kidney transplantation from the cadaveric donor with SIT.

In all cases successful kidney transplantation from donors with SIT was described owing to good preoperative evaluation aimed at timely detection of blood vessel anomalies present in these donors\textsuperscript{8}. Surgical approach may include open nephrectomy, hand-assisted laparoscopic donor nephrectomy and laparoscopic nephrectomy\textsuperscript{29,32}. Most of the authors failed to indicate the duration of the donor and recipient follow-up period, except for two authors who reported follow-up periods of 6 and 12 months, respectively\textsuperscript{31,32}. It may be observed that greater experience of surgeons leads to higher number of laparoscopic donor nephrectomies\textsuperscript{29,31,32}. We reported a case of successful predialysis kidney transplantation from a donor with SIT without major associated blood vessel anomalies or associated syndromes appearing in 20–25\% of SIT cases\textsuperscript{8,33}. Second degree renal failure was evidenced in our donor, which is not unusual after nephrectomy, but was not previously reported\textsuperscript{34}. At the time of writing this report, renal function was satisfactory in both donor and recipient for 20 months.

A large disproportion between the available organs for transplantation and long waiting lists oblige us to increase the number of transplantations by careful selection and evaluation of patients previously considered unsuitable candidates for organ donation. Since donor’s safety is the utmost priority, based on the review of the reported cases, it is clear that each individual case necessitates multidisciplinary approach to patients with SIT. Careful preoperative evaluation which includes the methods of visualization of organs and their vascularization may reveal possible abnormalities relevant for the surgeon\textsuperscript{30}. Since these cases are rare, recommendations are necessary for the transplantation experts to do appropriate preoperative evaluation and postoperative follow-up of donors with SIT.

### Table 2

<table>
<thead>
<tr>
<th>Authors</th>
<th>Number of kidneys procured</th>
<th>Outcome</th>
<th>Characteristics</th>
<th>Follow-up period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polak WG et al.\textsuperscript{8}</td>
<td>2</td>
<td>Successful</td>
<td>Cadaveric donor - 2 recipients</td>
<td>Unknown</td>
</tr>
<tr>
<td>Black PC et al.\textsuperscript{29}</td>
<td>1</td>
<td>Successful</td>
<td>Living donor</td>
<td>Unknown</td>
</tr>
<tr>
<td>Hoffmann D et al.\textsuperscript{30}</td>
<td>1</td>
<td>Successful</td>
<td>Hand assisted laparoscopic</td>
<td>Unknown</td>
</tr>
<tr>
<td>Berber I et al.\textsuperscript{31}</td>
<td>1</td>
<td>Successful</td>
<td>Right donor nephrectomy</td>
<td>1 year</td>
</tr>
<tr>
<td>van Dellen et al.\textsuperscript{32}</td>
<td>1</td>
<td>Successful</td>
<td>Open nephrectomy</td>
<td>6 months</td>
</tr>
</tbody>
</table>

IVC – inferior vena cava

Conclusion

This case report on a donor with \textit{situs inversus} and a successful kidney transplantation indicates that it is not an absolute contraindication for organ donation any more. This rare case also confirms the necessity of the multidisciplinary approach and team work (nephrologist, vascular surgeon, urologist, radiologist and anesthesiologist) in order to achieve satisfying results, and maximum safety for the donor.

Acknowledgments

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REFERENCES


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