



CASE REPORT

Living-related Donor Nephrectomy in a Patient With Complete Situs Inversus

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In this case, we report the successful transplantation of a kidney from a patient with complete situs inversus (SI) to a recipient with normal lateralization. Radiologic findings as well as intraoperative presentation of the right-sided donor organ and vasculature mirrored that of a normal subject. One-year postoperative follow-up in the recipient was reassuring. As there are few reports of functional or anatomic renal disease in patients with SI, we propose that these patients should remain a viable group for donor nephrectomy, given a normal thorough preoperative work-up.

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1. Introduction

Complete situs inversus (SI) is defined as total transposition of the thoracic and abdominal organs. Complete SI is present in 1/8000 to 1/20,000 persons.^{1,2} In the past, it was thought that SI should be considered an absolute contraindication for organ donation, especially in cases of liver and heart transplantation where lateralization changes operative planning and technique.^{2,3} Although various anatomic anomalies exist in higher percentages in these patients, rarely are the kidneys involved. The most common anomalies include gastrointestinal (>50% of cases), cardiac anomalies such as transposition of the great vessels (40%) and preduodenal portal vein (>40%).³ SI also coexists with Kartagener's syndrome in approximately 10% of cases, which is a deficiency in dynein leading to pulmonary bronchiectasis, chronic sinusitis, and male infertility.^{2,4} There have been few reports of functional or anatomic kidney disease associated with Kartagener's syndrome. In addition, there have been only a handful of documented case reports, suggesting an association between renal cell carcinoma and SI.^{1,4,5,6}

Because of the extreme need for organ donation across the world, it is imperative that we explore patients with alternative anatomic and pathophysiologic processes to determine the feasibility of organ transplantation. For this reason, we would like to document our experience of transplanting a kidney procured from a living-related donor with SI into a family member with normal lateralization.

2. Case report

A 31-year-old female with no significant medical history voluntarily presented for a right-sided, living-related kidney donation. She denied history of renal disease, renal cell carcinoma, rheumatologic disease, polycystic kidney disease, hypertension, diabetes mellitus, coagulopathy, bleeding disorders, or anemia. She had no prior surgical history. Her family history included end-stage renal disease secondary to focal glomerular sclerosis in her mother. Social history was negative for tobacco, drug, or alcohol abuse. Nephrology determined the patient to be eligible for organ donation. Her electrocardiogram showed sinus arrhythmia with a first-degree atrioventricular block.

A preoperative chest X-ray (Figure 1) was performed showing no evidence of active pulmonary disease; however, there was total visceral thoracic and abdominal SI. A spiral computed tomography angiogram of the abdomen with and without iodinated contrast (Figures 2 and 3) was then performed to evaluate anatomy and rule out any disqualifying pathology. The study showed complete SI of the heart and abdominal organs. The great vessels were also reversed in position mirroring normal anatomic distribution.

The patient was taken to the operating room for a right-sided living-related donor mini-open nephrectomy. She was placed in a left-lateral decubitus position. After obtaining control of the renal hilum through dissection and placement of vessel loops, the right renal vein, artery, and ureter were identified. The inferior vena cava and right renal vein were consistent with an anatomic left system as the right gonadal vein and suprarenal veins flowed into the right renal vein rather than directly into the inferior vena cava. The right renal vein was longer than typical as would be expected. Care was taken to ligate and transect the right suprarenal vein, right lumbar L1 vein, and right gonadal veins as they converged into the right

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Figure 1 Preoperative chest X-ray demonstrating situs inversus.

renal vein. The remainder of the procedure was completed without variation from normal lateralization technique.

The recipient at 1-year follow-up had adequate urine output and a baseline creatinine of 1.2 mg/dL.

3. Discussion

To date, there have been few articles discussing kidney donation in patients with complete SI. Polak et al³ described a case in 2006 where bilateral kidneys were procured from a deceased donor and transplanted into two different recipients; however, follow-up was not discussed. Black et al⁷ reported a case of successful hand-assisted

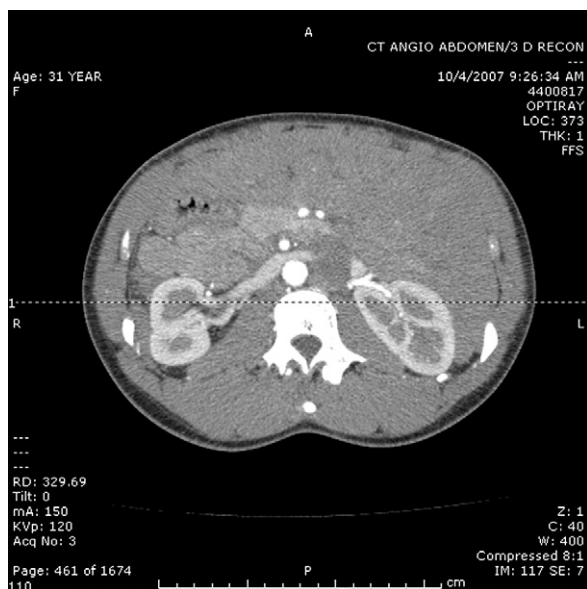


Figure 2 Spiral computed tomography angiogram of abdomen demonstrating long right renal vein.

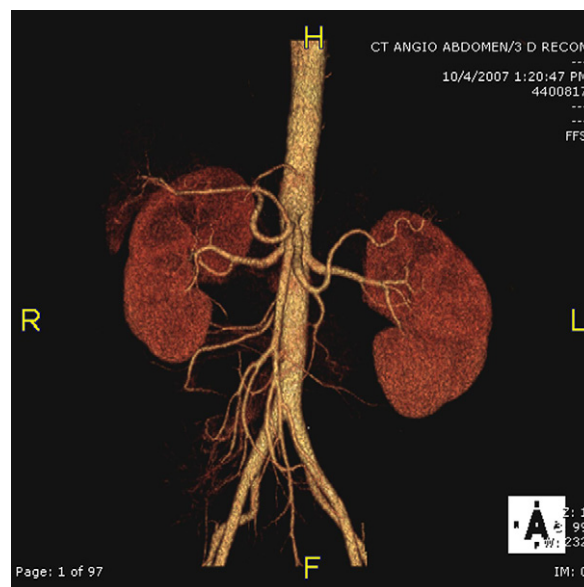


Figure 3 3-D reconstruction of abdominal computed tomography angiogram.

laparoscopic nephrectomy in a live donor with SI. Although it seems intuitive that kidney donation in a patient with SI should be similar to usual technique with the exception of opposite anatomy, transplant surgeons must be aware of the potential anomalies and disease processes that may complicate organ donation. This knowledge will allow for better preoperative preparation, more prudent patient selection, and likely more favorable surgical outcomes. For this reason, complete radiologic evaluation of the intra-abdominal vasculature should be sought before surgery to allow for operative planning and identification of possible anatomic variants that may complicate the procedure.⁸ The limited amount of data published regarding organ transplantation in patients with SI calls for more reporting of such findings so that a consensus can be formed regarding outcomes. This case represents successful kidney procurement in a patient with complete SI and subsequent transplantation with a viable and functioning organ at 1-year follow-up. The great need for organ donation today requires that transplantation centers look for new ways to broaden our selection of patients suitable for organ donation. This must be accomplished without significantly increasing risk to donor or recipient.

In conclusion, successful organ donation from a patient with complete SI transplanted into a patient with normal lateralization is feasible. This diagnosis should not hinder transplantation programs from considering these patients as suitable donors.

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