Case Report: First Reported Combined Heart-Liver Transplant in a Patient With a Congenital Solitary Kidney

R.M. Hanna[a,*], M. Kamgar[b], H. Hasnain[c], R. Khorsan[d], A. Nsair[e], F. Kaldas[f], A. Baas[g], S. Bunnapradist[h], and J.M. Wilson[b]

[a]Department of Medicine, Division of Nephrology, UCLA Health – South Bay Nephrology, Rancho Palos Verdes, California, USA; [b]Department of Medicine, Division of Nephrology, UCLA Stone Center Surgical and Consultative Nephrology, Westwood, California, USA; [c]Department of Medicine, Division of Nephrology, UCLA Health-South Bay Nephrology, Torrance, California, USA; [d]Department of Medicine, Division of Nephrology, UCLA-Santa Monica Medical Center, Santa Monica, California, USA; [e]Department of Surgery, Division of Cardiothoracic Surgery, UCLA Ronald Reagan Medical Center, Westwood, California, USA; [f]Department of Surgery-Liver Transplant Surgery, UCLA Ronald Reagan Medical Center, Westwood, California, USA; [g]Department of Medicine-Cardiology, UCLA Ronald Reagan Medical Center, Westwood, California, USA; and [h]Department of Medicine-Transplant Nephrology, UCLA Ronald Reagan Medical Center, Westwood, California, USA

ABSTRACT

We report a case of successful combined heart liver transplant in a patient with a congenital solitary kidney. The patient had normal renal function before combined heart-liver transplantation and developed acute kidney injury requiring slow continuous dialysis and subsequent intermittent dialysis for almost 8 weeks post transplantation. Her renal function recovered and she remains off dialysis now 7 months post transplantation. She only currently has mild chronic renal insufficiency. We believe this is the first reported case of successful heart liver transplant in a patient with a congenital solitary kidney.

TRADITIONALLY there has been concern for increased risks of solitary kidney patients for various renal insults. However, a recently reported large cohort showed the risk for acute kidney injury (AKI), dialysis, or death attributable to contrast-enhanced computed tomography (CT) was similar in patients with a solitary kidney relative to controls with both kidneys [1], although other studies reported nephrectomy as a significant contributor to the development of chronic kidney disease (CKD) in nephrectomized patients with renal cell carcinoma [2]. The majority of effects observed in the patient with a solitary kidney are long-term in nature [3]. The expected pattern of injury is nephron enlargement followed by development of focal segmental glomerulo-sclerosis.

The rate of development of end-stage renal disease (ESRD) and CKD in patients who were cleared and underwent living kidney donation is quite low (90/10,000 patients) and actually lower than the ESRD risk of the general population (326/10,000 patients), yet slightly higher than comparable very healthy age-matched controls (14/10,000 patients) [4]. The risk of developing renal disease in patients with solitary kidneys is very low with selected populations of highly healthy individuals being able to tolerate the stress of a solitary kidney. Therefore, it is likely that other genetic, environmental, or clinical factors influence which subset of patients with a solitary kidney develop chronic renal disease.

The increased complication rate, mortality rate, and resultant CKD after postoperative AKI has been well noted in post–cardiac surgery patients [5]. The renal prognosis of patients who have AKI is worse than those who do not have such an insult. Although prevention is ideal, a more easily attainable goal is identifying who may develop CKD/ESRD postoperatively. The term cardiac surgery–associated AKI (CSA-AKI) is now in use to investigate this subset of kidney injury with its negative short-term and long-term prognostic implications [6,7].

Heart transplantation is associated with a 10% risk of developing ESRD and patients with advanced renal disease prior to heart transplantation are often managed with simultaneous heart-kidney transplantation (HKT) [8–11].

*Address correspondence to Ramy M. Hanna, MD, Clinical Instructor of Nephrology and Internal Medicine, David Geffen School of Medicine, Department of Medicine, Division of Nephrology, UCLA South Bay-Rancho Palos Verdes, Nephrology, 501 Deep Valley Suite #100, Rolling Hills Estates, CA 90274, USA. E-mail: Rhannamd81@yahoo.com

Published by Elsevier Inc.
230 Park Avenue, New York, NY 10169

0041-1345/17
https://doi.org/10.1016/j.transproceed.2017.11.030

HKT for dialysis-dependent pre–cardiac transplantation recipients tends to be less controversial than those with non–dialysis-dependent kidney disease [11]. Liver transplant recipients often have comorbid renal disease at time of transplantation at a rate of about 32% [12]. CKD occurs in about 20%–80% of liver transplant recipients post-transplantation for varied causes [13]. Hence the burden of renal disease in both these populations is high and can be associated with higher costs and worse outcomes including worsening of CKD and progression to ESRD [14]. Patients who have undergone combined heart-liver transplantations (CHLTs) are a small subset of all transplant recipients but studies do show comparable outcomes, despite the increased surgical complexity and acuity of illness present in such patients [15]. In one study between 1987 and 2005, 43 CHLTs were performed for amyloidosis; of these patients, 6 had enough renal disease to warrant a CHLT with a kidney transplantation (triple-organ transplantation). There is a paucity of published data on multi-organ transplantation in patient with a solitary kidney. We present the first case of a congenital solitary kidney patient who underwent a CHLT.

CASE REPORT

The patient was a 39-year-old woman with a history of tricuspid atresia who underwent a Blalock Taussig shunt at the age of 1, which was revised to an extra-cardiac fenestrated Fontan procedure and a MAZE procedure in 2012. She then developed sick sinus syndrome (SSS), and underwent pacemaker placement and later ablation of ectopic loci of electrical activity in the atria in 2014. In 2016 she became acutely ill with a gastrointestinal (GI) duodenal ampullary bleed and had a respiratory arrest with successful resuscitation. She developed ascites and evidence of worsening liver function with frequent paracentesis. A liver biopsy done in Colorado showed advanced fibrosis and cirrhosis due to congestive hepatopathy. Echocardiography confirmed tricuspid atresia and normal size right atrium with a widely patent atrial septal defect. Pulmonary hypertension was noted, and a visual estimate showed normal left ventricular systolic function.

She presented to the University of California Los Angeles for a CHLT evaluation. During work-up she was noted to have a congenital solitary kidney and her serum creatinine level ranged from 0.5-1.1 mg/dL. Her baseline level of proteinuria was only mild with a urine protein to creatinine ratio (UPC) of 0.2 g total protein/g creatinine. Her renal ultrasound revealed a kidney size of 13 cm that was mildly enlarged but structurally normal with no hydronephrosis and with normal cortical thickness and echogenicity. Her renal functions remained stable with no accompanying signs of glomerular disease, allowing her to be scheduled for CHLT without a concomitant renal transplantation.

As her diuresis for volume overload continued, her serum creatinine level increased from a baseline of about 0.5–1.1 to 1–1.5 mg/dL. She was maintained on ionotropic agents and diuretics while awaiting CHLT. She was prepared for transplantation with plasmapheresis due to high sensitization and was induced for transplantation with rabbit anti-thymocyte globulin. She was also given tacrolimus 0.5 mg orally twice a day, intravenous solumedrol and oral steroids tapered down to lower doses, and mycophenolate mofetil at 1 g twice a day. Once anti-thymocyte globulin infusion was finished, the tacrolimus was titrated up to 2 mg orally twice a day to target 8–10 ng/mL with an eventual decrease of target to 5–7 ng/mL 6 months post-transplantation. She was listed as status Ia for a CHLT. Her Model for End Stage Liver Disease (MELD) score on March 21, 2017 was 22.

Postoperatively her serum creatinine level started to slowly increase from 0.9 mg/dL to 1.1–1.62 mg/dL, likely due to hypotension with systolic blood pressure ranging near 80 mm Hg, with mean arterial pressures as low as 52 mm Hg. This necessitated initiation of vasopressors with vasopressin and norepinephrine. There was concern for sepsis due to positive donor blood cultures with lactobacillus rhamnosus, which was treated with intravenous vancomycin.

Urine output decreased from 3.7 L/d to 1.7 L/d despite an attempt for diuresis with a furosemide drip, and the patient remained in positive net fluid balance, which is not optimal for the cardiac graft. Continuous renal replacement therapy (CRRT) with a continuous veno-venous hemodialysis (CVVHD) modality on the NxStage machine was then started on postoperative day 2 to prevent further volume overload and ameliorate any dyselectrolytemias. The patient was extubated 8 days after initial orthotopic heart transplantation (OHT)/orthotopic liver transplantation (OLT), but was still on CRRT and had pressor requirements. Pressors were progressively weaned off and were off by 19 days post–OHT/OLT. The next day (20 days postoperative) the patient received her first session of intermittent hemodialysis and required daily dialysis for some time.

She started having increasing urine output on postoperative day 31 and was challenged with diuretics while maintaining on intermittent dialysis (IHD) when her urine output was not adequate. Her serum creatinine level peaked at 4.85 mg/dL but started to improve on postoperative day 38. Her serum creatinine level continued to decrease thereafter, returning to baseline 53 days post–OHT/OLT. Please see Figure 1 for graphical trend of serum creatinine levels with timing of OHT/OLT, CRRT initiation/discontinuation, IHD initiation/discontinuation, and beginning of diuretic challenge. The patient has now been status post OHT and OLT for nearly 7 months and has a serum creatinine baseline level of 1–1.26 mg/dL without need for hemodialysis. Her creatinine level briefly peaked at 1.36 mg/dL but promptly came down with calcineurin inhibitor (CNI) dose adjustment. Her cardiac and hepatic grafts are working well. She is tolerating CNI therapy without significant renal injury. The target tacrolimus level will be maintained at 5–7 ng/mL for the first year and reduced to around 5 ng/mL thereafter. There has been minimal CKD after her initial AKI observed post-transplantation. Despite the concern of the solitary kidney, this patient was able to recover renal function after a prolonged episode of AKI due to the hemodynamic stresses of a CHLT.

DISCUSSION

The presence of a solitary kidney may only mark a higher risk for certain insults acutely and for development of focal segemental glomerulosclerosis (FSGS) chronically. This case illustrates that a patient with a solitary kidney who has no other markers of kidney disease was able to have an acceptable renal outcome after life-saving OHT and OLT for her systemic disease brought about by congenital tricuspid atresia.

This case report represents, to our knowledge, the first case of a patient who had a dual organ transplantation with