

## Case Report

# Iatrogenic Megaureter of Ipsilateral Native De-Functionalized Ureter Following Kidney Transplantation for Posterior Urethral Valves

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## Abstract

We present a novel case in which a PUV patient who had undergone a living-related renal transplant developed recurrent episodes of intermittent frank pyuria due to his native ureter. It ultimately developed into an iatrogenic megaureter impinging on the transplant structures. For such an immuno compromised patient, this unanticipated outcome leads to complications with clinical sequelae that may have jeopardized his renal graft.

**Keywords:** Megaureter; Pediatric; Iatrogenic megaureter; Posterior urethral valves; Renal transplant; Vesicoureteral reflux

## Introduction

Posterior Urethral Valves (PUV) leads to End-Stage Renal Disease (ESRD) in 15 to 20 percent of patients, and in severe cases, this requires renal replacement therapy in infancy [1]. These patients may undergo native nephrectomy with sparing of their ureter, as the native ureter has demonstrated usefulness for ureterocystoplasty, urinary catheterizable channels, and ureteral substitution for complications associated with the transplant ureter [2,3]. Alternatively, the native ureter can be clipped without the need for formal nephrectomy [4,5].

## Case Presentation

OB presented to our nephrology service at 5 weeks of age with known chronic kidney disease secondary to obstructive uropathy and a functioning vesicostomy. Surgery was performed at 2 weeks of age due to lower urinary tract obstruction and a rising creatinine. He had confirmed PUV and bilateral grade 5 vesicoureteral reflux on voiding cystourethrogram. At two months of age, he underwent a G-tube and peritoneal dialysis catheter placement, after which he was subsequently admitted for peritonitis and several times for hyperkalemia. He underwent left nephrectomy, creation of a catheterizable channel using the left ureter in addition to valve ablation and vesicostomy closure at six months at an outside institution. Recurrent episodes of febrile urinary tract infections secondary to *Pseudomonas* prompted

surgery. The patient did well with peritoneal dialysis but developed a vesicocolonic fistula requiring surgical intervention at eight months and institution of hemodialysis until reaching a target weight of 10 kg for renal replacement therapy. At eleven months, the patient received a living donor right renal transplant with extravesical reimplantation of the transplant ureter on the posterior bladder without an advancing stitch in addition to native ureter-sparing right nephrectomy. The surgeon left the right ureter in situ for future reconstruction, if it became necessary. Following this operation, the patient did well on anti-rejection medications and had normal blood pressure recordings on Amlodipine. Renal Bladder Ultrasound (RBUS) one month post-transplant showed increased caliber of the tortuous right native ureter. The subsequent RBUS four months later confirmed no hydronephrosis and a fluid-filled serpentine structure with maximum caliber of 3.3 cm in the right lower abdominal quadrant (Figure 1). Two months later, repeat study showed mildly dilated distal right ureter and definitive absence of previous serpentine structure. The patient's mother reported that two days prior to this RBUS study, her son's urine had become cloudy for the first time since transplantation although there was an unremarkable urinalysis at that time. A post-transplant VCUG study displayed grade III VUR into the transplanted ureter but no reflux into the right native ureter (Figure 2). Nine months post-transplantation, OB had complaints of pain with nighttime catheterization, right lower abdominal mass effect, and an ultrasound confirmed a dilated tortuous right native ureter with maximum caliber of 3 cm. Anticholinergic therapy and bladder catheterizing twice per day was recommended.

The patient's surgeon relocated from the area with the patient transferred to another outside institution. He underwent video urodynamic evaluation one year post-transplant demonstrating no evidence of reflux in the transplanted kidney. He was maintained on oxybutynin and twice-daily catheterizations along with gentamicin irrigation of his bladder daily. Fourteen (14) months post-transplant, referral of OB to the Children's Hospital of the King's Daughters Urology Division occurred. His anticholinergic therapy was gradually weaned, since his prior urodynamic study showed a compliant

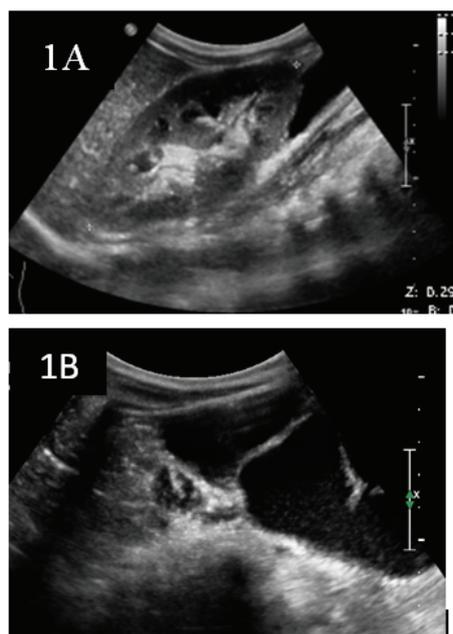
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**Figure 1:** 1A: Right transplant kidney without hydronephrosis after transplantation; 1B: Serpentine structure consistent with dilated native ureter.



**Figure 2:** VCUG shows grade 3 vesicoureteral reflux into the transplant ureter at bladder dome.

bladder but incomplete bladder emptying. He developed 5 days of cloudy urine, with a RBUS confirming a markedly dilated tortuous right native ureter with copious accumulation of debris throughout the ureter (Figure 3). The urine culture was positive for enterococcus despite intravesical gentamicin irrigation. A MRU urography further revealed the abnormal anatomy. There was dilation of proximal to mid transplant ureter without filling defects and distal transplant ureter compressed with concern of impingement by a tortuous massively dilated native right ureter measuring up to 4 cm (Figure 4).

In consultation with the nephrology service, it was recommended that OB undergo a right ureterectomy due to concerns of recurrent infections. In the interim, OB was spontaneously voiding, with the catheterized volumes oftentimes being negligible. On the day of surgery, an intraoperative cystogram demonstrated a faint wisp of reflux in the transplant ureter at a volume of 100 ml (patient's wt=14 kg, predicted bladder capacity of 100 ml). The ureterectomy

was performed *via* a Pfannenstiel incision, which provided excellent exposure to the distal ureter, which was ligated and over sewn at the uretero-vesical junction extravasically. The tortuous dilated ureter, more so distally than proximally, was removed without compromising the transplanted ureter.

Gross pathology of the native ureter and microscopic findings confirmed luminal variations with compensatory hypertrophy and inflammation (Figure 5).

OB returned to the urology clinic 6 weeks following surgery. His RBUS showed grade 2 hydronephrosis of the transplant kidney with no ureteral dilation. He continued with the twice-daily catheterization without anticholinergic treatment. There has not been large volumes with catheterization nor episodes of cloudy urine or pyuria.

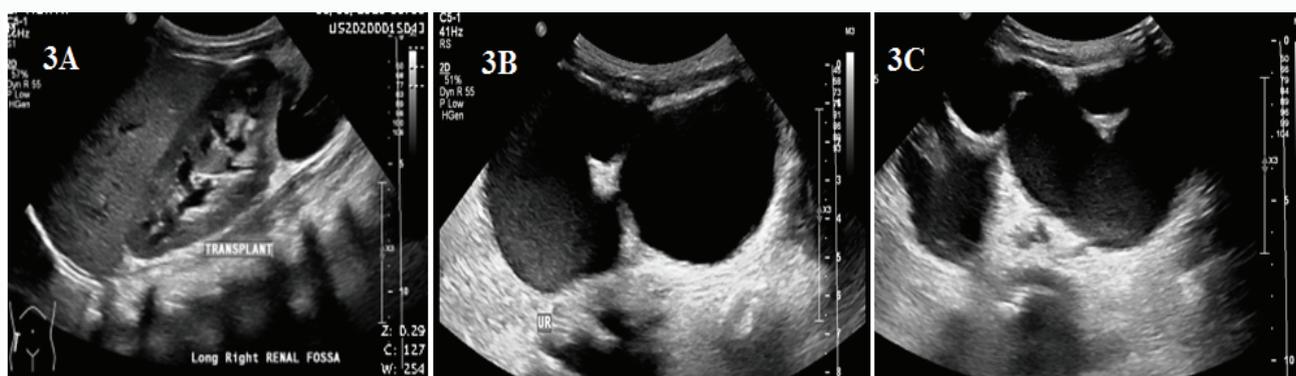
## Discussion

Historically, ureteral stump syndrome (clinically seen as febrile urinary tract infections, lower quadrant pain, and hematuria) occurs post-nephrectomy. Empyema of the ureteral stump after nephrectomy is a rare (0.8% to 1%), but potentially devastating complication [6]. In our patient, a dilated tortuous ureter with empyema of the entire stump led to recurrent frank pyuria and compromise to drainage of the transplant ureter. These rare cases have been reported in adults, for whom nephrectomy was performed for non-malignancy related conditions [7]. The majority of patients with remnant native ureter do not have any issues. Surgeons often leave the native ureter to minimize the surgical dissection and maintain an extra-peritoneal approach with one incision.

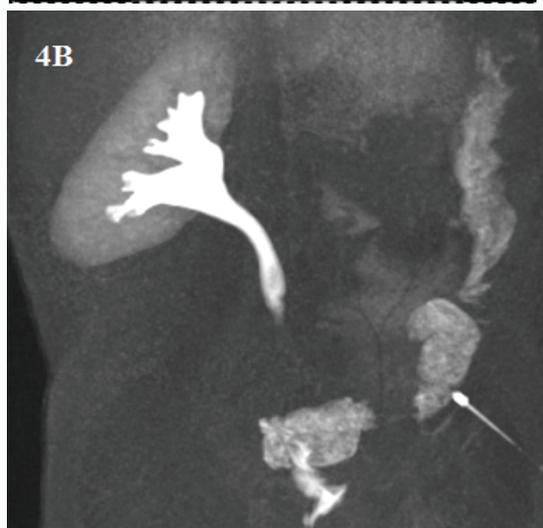
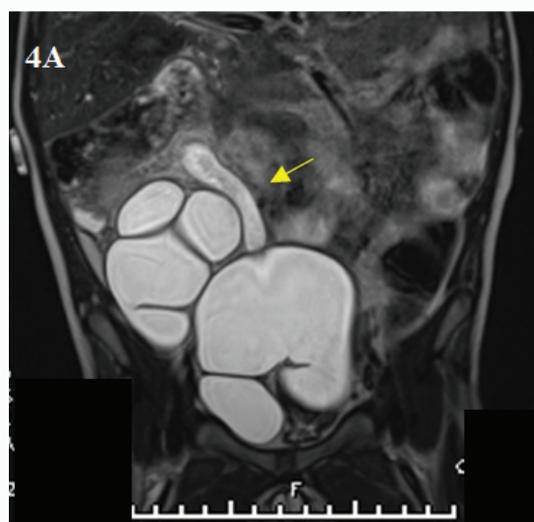
A consideration for leaving a native ureter in patients undergoing nephrectomy is for potential future reconstructive procedures. This is beneficial for pediatric patients who may require bladder augmentation, urinary diversion, or catheterizable channels. This was the rationale for our patient. However, the ureteral stump may serve as a nidus for infection, as was the case in our index patient [8]. The remnant ureter is still peristaltic and may lead to creation of a loculated type of bladder diverticulum.

Following renal transplantation, patients are at risk for a variety of ureteral complications including vesicoureteral reflux. This is especially true in the pediatric population, and in fact, the patient in our case demonstrated VUR into his transplant ureter but not the native ureter. Transplant patients with VUR into the transplanted kidney can be successfully managed with bladder training and temporary antibiotic prophylaxis so long as they are not developing recurrent febrile Urinary Tract Infections (UTI) [9]. Those with recurrent febrile UTI typically require surgical intervention, either re-implantation or the use of Deflux [10,11].

The native ureter is typically de-functionalized and not a source of pathology. From extensive review of the literature, there is one case report of a megaureter requiring intervention in an adult following childhood transplant as well as a case of urothelial carcinoma developing in a de-functionalized native ureter [12]. To the best of our knowledge, we report the first pediatric case of iatrogenic induced megaureter of the native ureter that served as both a nidus of recurrent pyuria with clinical symptomatic UTI and structural impediment to the functional transplant kidney/ureter. It remains to be seen if more complications of the native ureter will arise with the contemporary practice of ureteral ligation of the native ureter without nephrectomy. To date, this has not been the case with this surgical practice.



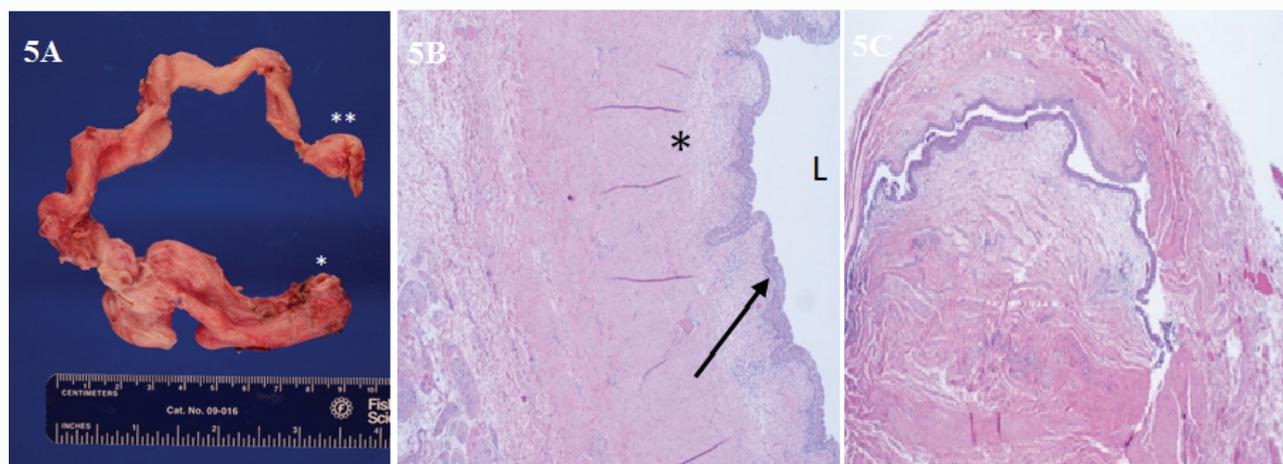
**Figure 3:** 3A: Transplant kidney with mild pelviectasis; 3B: Ureteral dilation of 3.1 cm distally adjacent to bladder with debris; 3C: Native ureter with dilation of 1.3 cm proximally and dependent debris within acquired mega ureter.



**Figure 4:** 4A: MRI urogram showing severely dilation of the native ureter with dilation of the transplant ureter (arrow); 4B: MRI nephrogram with abrupt termination of spontaneous drainage of the transplant ureter, due to marked dilated native ureter.

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**Figure 5:** 5A: Native ureter: 21 cm markedly coiled with adhesions around the coils, stretched length of 23 cm. The decompressed specimen had a proximal (\*\*) ureter of 0.5 cm and the distal (\*) end measured 1.3 cm with other portions varying up to 2 cm diameter; 5B: Microscopic cross-section with dilated ureter showing lumen (L) and muscle (\*) and inflammation at the luminal edge (arrow); 5C: Microscopic cross-section with absence of lumen secondary to severe hypertrophy.