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# Bilateral proximal ureteral and ureterovesical junction obstruction in a child

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ABSTRACT

The most common sites of ureteral obstruction in children are at the level of the ureteropelvic junction (UPJ) and ureterovesical junction (UVJ). Bilateral hydronephrosis or hydroureteronephrosis due to varying degrees of obstruction at the UPJ or UVJ is common in children and typically improves with time. Clinically significant obstruction at both locations in an ipsilateral ureter occurs less commonly and rarely requires both dismembered pyeloplasty and ureteral reimplantation. We believe this case report is the first description of bilateral proximal and distal ureteral obstruction requiring both dismembered pyeloplasty and ureteral reimplantation.

## 1. Introduction

The ureteropelvic junction (UPJ) and ureterovesical junction (UVJ) are the two most common locations in the ureter for obstruction in children. The exact mechanisms involved in this pathologic process have not been elucidated, but the pathophysiology is believed to occur due to failed maturation of fetal ureteric cells at the UPJ and UVJ leading to decreased peristaltic activity at these two sites.[1]

Obstruction typically occurs at only one level in a single ureter. However, obstruction at the UPJ, or less commonly the proximal ureter, and/or UVJ obstruction can present as an ipsilateral or bilateral condition. Bilateral proximal ureteral (PU) and UVJ obstruction are exceedingly rare and to our knowledge have not been reported. We present a case of a child with bilateral UVJ obstruction who developed bilateral PU obstruction following ureteral reimplantation requiring bilateral pyeloplasty.

## 2. Case presentation

A full-term girl was born at 36 weeks gestation. Prenatal sonography demonstrated bilateral mild hydronephrosis without ureteral dilation and an unremarkable bladder with normal amniotic fluid. The child was born with a tracheoesophageal fistula, which was repaired on day of life (DOL) 2. Postnatal renal bladder ultrasound (RBUS) on DOL 6 demonstrated mild hydronephrosis, no ureteral dilation, and a decompressed bladder. The nadir creatinine during the child's initial hospitalization was 0.34 mg/dL. The child was discharged home at 3 weeks of age but lost to follow-up for social reasons.

At 11 months of age, the patient presented to the emergency room with severe dehydration, pyelonephritis, and acute kidney injury marked by an elevated serum creatinine of 2.1 mg/dL. An RBUS revealed severe bilateral hydroureteronephrosis down to the level of the bladder with echogenic debris in both kidneys (Fig. 1). Placement of an indwelling catheter failed to improve the patient's hydroureteronephrosis or creatinine elevation. Bilateral percutaneous nephrostomy tubes were placed which resulted in rapid clinical improvement and return of normal renal function. A subsequent bilateral antegrade nephrostogram demonstrated tortuous, dilated ureters down to the level of the bladder. A mercaptoacetyltriglycine (MAG-3) nuclear renogram with furosemide and clamped nephrostomy tubes demonstrated obstruction at the level of UVJ bilaterally (Fig. 2).

One month later, the patient underwent excision of both aperistaltic segments of ureter with bilateral Politano-Leadbetter ureteral reimplantation without ureteral tailoring. Surgery was uncomplicated with no ureteral stent placed and both nephrostomy tubes were left open to drainage. The early postoperative course was unremarkable. However, the child did not tolerate clamping of the nephrostomy tubes as evidenced by increased irritability, rising serum creatinine, and absence of

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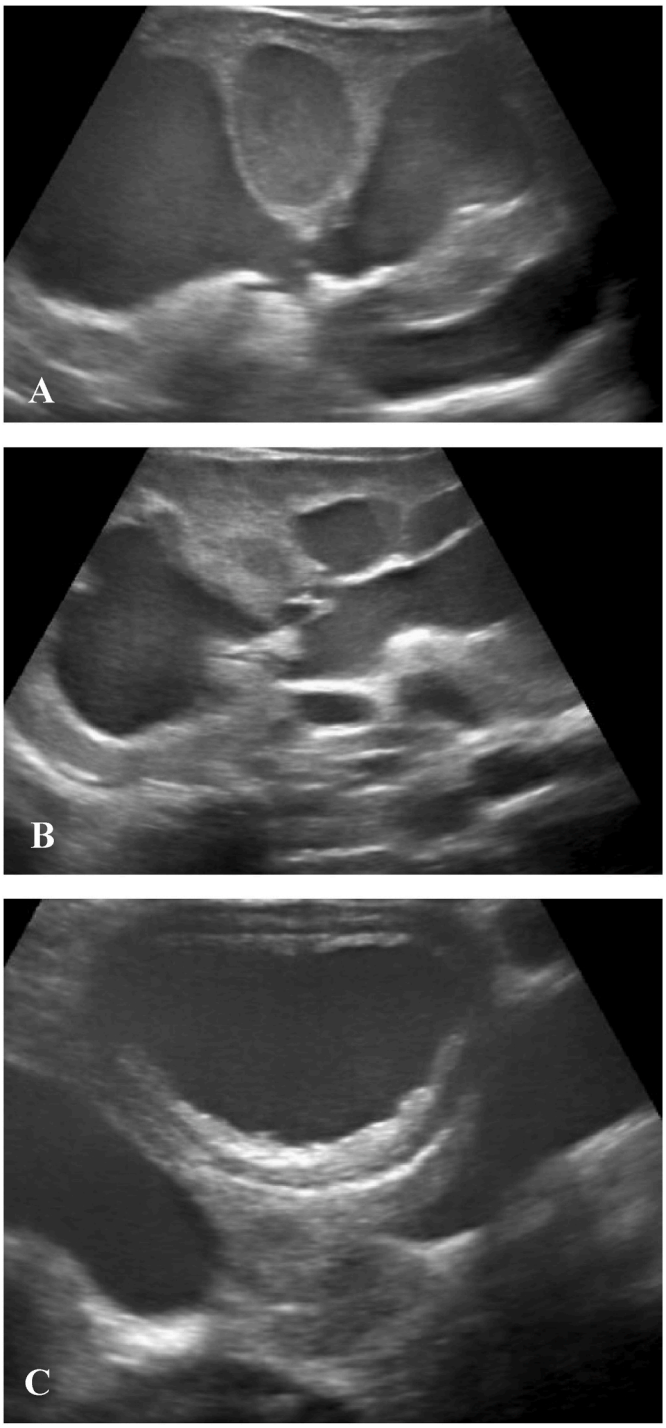


**Fig. 1.** Renal bladder ultrasound demonstrating severe right (A) and left (B) hydroureteronephrosis. Transverse image of bladder (C) showing bilateral distal hydroureters down to the level of the bladder.

urine output from the indwelling bladder catheter. It was presumed the patient had a transient distal ureteral obstruction from edema following the bilateral ureteral reimplantation, which would improve with time.

The child was discharged home with the nephrostomy tubes to gravity drainage and without a Foley catheter. Several capping trials of the nephrostomy tubes at home over a 2-month period were similarly not tolerated. This failure prompted repeat bilateral antegrade nephrostogram demonstrating bilateral PU obstruction (Fig. 3). Again, the child was taken to the operating room and retrograde pyelography demonstrated no evidence of distal ureteral obstruction and confirmed the proximal ureteral issue. Bilateral ureteral stents were placed and the nephrostomy tubes were capped. The patient tolerated this well and had a prompt return of normal voiding.

Three months after ureteral reimplantation surgery, the patient underwent a left dismembered pyeloplasty, followed by a right dismembered pyeloplasty 6 weeks later. A dysplastic, stenotic segment of ureter, distal to the UPJ and was encountered bilaterally. After staged pyeloplasty, the patient has done well as evidenced by normal serum creatinine and resolution of hydronephrosis at 2-year follow-up.

## 3. Discussion

Primary UVJ obstruction presents as a bilateral process in approximately 15% of children.[2] UPJ obstruction has a slightly higher rate of bilaterality with a range of 10–40%.[3] Coexisting ipsilateral UPJ obstruction was reported to occur in 13% of children with unilateral obstructive megaureter.[2] To our knowledge there are no reports of bilateral UPJ/PU and UVJ obstruction requiring pyeloplasty and ureteral reimplantation.

Identifying the location of ureteral obstruction preoperatively is challenging when there is both a proximal and distal impedance to flow. In a scenario with coexisting abnormalities, radiographic evaluation is often incomplete and an accurate diagnosis is made at the time of, or in follow-up after a successful initial surgery.[4] Most commonly in coexisting ipsilateral proximal and distal ureteral obstruction, diagnosis of the UVJ obstruction is unmasked following successful pyeloplasty.[5]

Secondary obstruction at the UPJ has been reported in children with high-grade vesicoureteral reflux (VUR) and a dilated, tortuous ureter. Ureteral reimplantation and correction of VUR may improve the proximal obstruction and obviate the need for pyeloplasty. In contrast, severe tortuosity and dilation of the ureter seen with congenital obstructive megaureter are not thought to be a contributor to proximal ureteral obstruction. Instead, ipsilateral concomitant UPJ/UVJ obstructions are believed to be separate, independent pathologies.[4]

Our patient presented with bilateral UVJ obstruction without a coexisting proximal ureteral issue. Following successful bilateral ureteral reimplantation, the child developed bilateral PU obstruction. This new obstruction occurred within months following surgery and required bilateral pyeloplasty. Our patient did not have concomitant UPJ and UVJ obstruction with a decision to address the UVJ first then the UPJ second as reported as an option.[2]The mechanism that led to the bilateral proximal obstruction has eluded the authors and literature provides no clear insight into a possible etiology. Based on the fairly rapid obstruction following ureteral reimplantation, excision and straightening of the distal ureters may have altered the proximal ureteral course, playing a role in the obstruction.

## 4. Conclusion

Congenital ureteral obstruction is common and can evolve. Typically, obstruction is unilateral and affects either the UPJ or UVJ. Concomitant obstruction at both locations in the ipsilateral ureter can be a diagnostic challenge, and a coexisting second obstructive segment may not be unmasked until after a successful primary repair. Bilateral PU and UVJ requiring bilateral pyeloplasty and ureteral reimplantation can occur in children. Our patient presented with secondary proximal obstruction following successful ureteral reimplantation.

## Consent

Appropriate authorization to use or disclose de-identified health information for publication and educational purposes was obtained.

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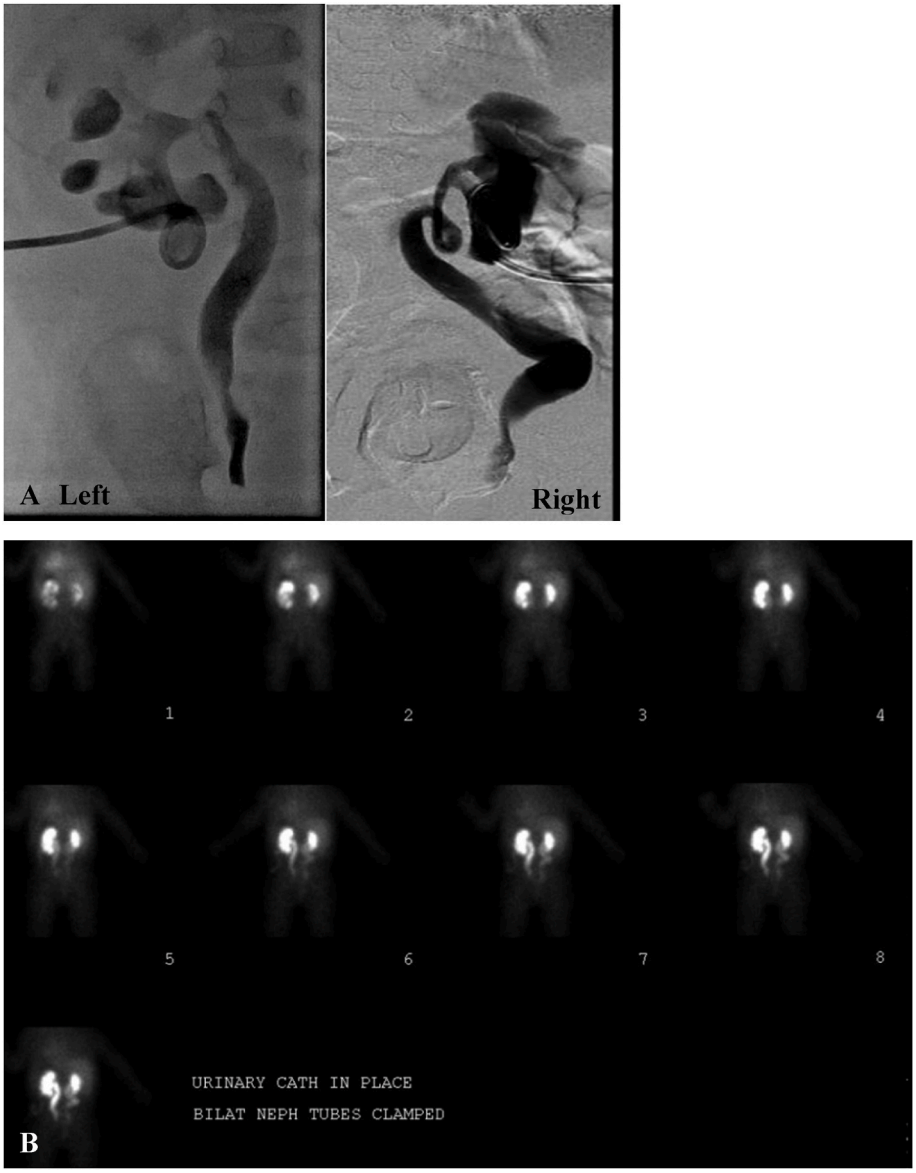


**Fig. 2.** Images from bilateral antegrade nephrostogram (A) and MAG-3 nuclear renogram (B) demonstrating bilateral UVJ obstruction.

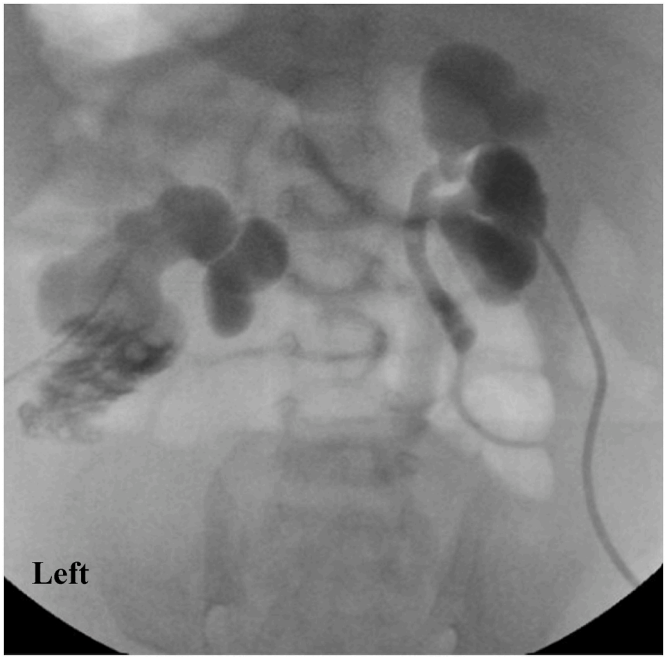


**Fig. 3.** Prone images from bilateral antegrade nephrostogram demonstrating no contrast traversing UPJ/proximal ureter with accompanying severe calyceal dilation.

agencies in the public, commercial, or not-for-profit sectors.

## Author statement

Daniel A. Reich: Conceptualization, Data Curation, Writing - Original draft, Writing - Review & Editing, Christopher E. Bayne: Writing - Original draft, Writing – Review & Editing, Cynthia A. Sharadin: Writing – Original draft, Writing – Review & Editing, Romano T. DeMarco: Conceptualization, Data Curation, Writing – Review & Editing, Visualization.

## Declaration of competing interest

None.

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