

## A Child with Bilateral Hydronephrosis Presenting with Coexistence of Bilateral UPJ and Bilateral UVJ Obstruction: A Case Report

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### Abstract

**Introduction:** Hydronephrosis can be either unilateral or bilateral, often due to ureteropelvic junction (UPJ) or ureterovesical junction (UVJ) obstruction. UPJ and UVJ can coexist in the same ureter, varying in severity, and the presence of both can make appropriate diagnosis more difficult. As far as we know, there are no reports of concurrent bilateral UPJ and bilateral UVJ obstructions.

**Case Presentation:** An 11 month old female with history of congenital hydronephrosis and urethral stenosis presented to the hospital with Klebsiella urosepsis and acute kidney injury. Due to bilateral hydroureteronephrosis, thickened bladder wall, and failed catheterization attempt, a suprapubic catheter was placed to relieve bladder outlet obstruction. However, minimal decompression was observed and the serum creatinine continued to rise to 1.9 mg/dL despite therapy. A MAG-3 scan showed bilateral high-grade UVJ obstruction, for which she underwent bilateral nephrostomy tube placement. With renal decompression, the serum creatinine trended downwards to her new baseline of 0.6 mg/dL. Subsequently, she underwent bilateral ureteral re-implantation at 13 months of age. A repeat antegrade nephrostogram revealed bilateral UPJ obstruction as well.

**Conclusion:** It is important to have a high index of suspicion for coexisting UPJ and UVJ obstruction when evaluating a patient with hydronephrosis. The aim for therapy is to salvage the kidney function. A multi-disciplinary approach with a nephrologist and urologist is also crucial.

**Keywords:** Hydronephrosis, UPJ obstruction, UVJ obstruction, MAG-3 scan, Retrograde pyelogram, CAKUT.

### Introduction

Ureteropelvic junction obstruction (UPJO) is the most common cause of hydronephrosis and affects approximately 1 in 1500 live births [1]. The left side is affected in approximately two-thirds of cases and about one-fourth to one-third of cases of UPJO

are bilateral. Ureterovesical junction obstruction (UVJO) is associated with a primary megaureter and is bilateral in 15-40% of cases [2]. UPJO is more prevalent than UVJO. However, the coexistence of congenital UPJ and UVJ obstruction in the same ureter is a rare occurrence, and has not been extensively reported. Since the obstruction can lead to increase in back-up pressure and progressive decline in renal function, it is important for proper diagnosis and treatment. In this case report, we describe a pediatric patient with congenital bilateral hydroureteronephrosis due to concurrent bilateral UPJ and bilateral UVJ obstruction.

### Case Presentation

An 11 month old Caucasian female with history of congenital hydronephrosis and presumed common urogenital sinus presented to the hospital with Klebsiella urosepsis and acute kidney injury. She was born at 40 wks gestational age to a 24yo gravida 1 para 1 mother. The mother had history of polyhydramnios and substance abuse (methadone and opiates) during pregnancy. Infant was born with esophageal atresia and distal trachea-esophageal fistula (repaired on day of life two), presumed common urogenital sinus, and left sided hydronephrosis with a right sided simple renal cyst. After being discharged from the NICU at one month of age, she was lost to follow-up by the urology service. She presented at 11 months of age to the emergency room with vomiting, diarrhea and persistent fevers for six weeks. Her pediatrician had treated her for viral and bacterial infections with no improvement. Her initial hospital labs showed severe anemia (hemoglobin 4.8 mg/dL, hematocrit 15.6 mg/dL), thrombocytopenia (platelet count of 100,000), elevated CRP 173 mg/L (normal reference range: 0-4.9 mg/L), acute kidney injury (serum creatinine of 1.81 mg/dL, BUN 51 mg/dL), and severe electrolyte abnormalities (potassium 6.1 mmol/L, sodium 126 mmol/L, serum bicarbonate 13 mmol/L). Blood and urine cultures grew Klebsiella pneumoniae. She was treated with intravenous Ceftriaxone for a total of 14 days.

The initial renal ultrasound on first day of admission showed

severe bilateral hydronephrosis with debris in the bilateral collecting systems, compatible with pyonephrosis (see Image 1A). The right kidney measured 7.9 x 4.9 x 3.7 cm. The left kidney measured 9.5 x 5.5 x 3.7 cm. The urinary bladder wall was thickened (see Image 1B). Due to initial concern for bladder outlet obstruction secondary to a presumed urogenital sinus and difficulty passing a urinary catheter, a suprapubic catheter was placed by pediatric urology. However, with a rising serum creatinine of 2.06 mg/dL and concern for persistent hydronephrosis, a Technetium Tc99m mercaptoacetyl-triglycine (MAG3) scan was obtained. The first MAG3 scan showed bilateral obstruction, presumed to be at UVJ, with a differential of 67% on the left and 33% on the right (see Image 1C). Thus, in the presence of a worsening renal function and poor differential, it was decided to proceed with the placement of bilateral percutaneous nephrostomy tubes. Immediately after the placement of the nephrostomy tubes, the serum creatinine decreased to 0.58 mg/dL a day later. Repeat renal ultrasound two days after the surgery showed decrease in the size of the hydronephrosis (right kidney measured 7.4 x 3.7 x 2.9cm and the left kidney measured 7.1 x 4.1 x 2.7 cm) (see Image 1D). Two weeks later, a repeat MAG3 scan confirmed the presence of bilateral UVJ obstruction (see Image 1E). At 13 months of age, she underwent bilateral ureteral re-implantation, while the nephrostomy tubes were still in place at that time. It was observed under anesthesia that she did not have a common urogenital sinus, but a urethral stenosis that required further dilation. A repeat antegrade nephrostogram after ureteral re-implantation interestingly showed concurrent bilateral UPJ obstruction (see Image 1F). The UPJ obstruction has not been corrected yet. Her renal function remains stable with normalization of her electrolyte disturbances.

## Discussion

The coexistence of obstruction at the UPJ and UVJ is rare. Prevalence of having obstruction in the same ureter at both ends varies between 3% to 25% [2]. The degree of severity of obstruction can have intra and inter-individual variation. Many theories exist to explain the development of intrinsic UPJO. Some authors postulate that intrinsic UPJO is thought to be a result of inadequate recanalization in utero at 10-12 weeks of gestation [1] and others associate it with smooth muscle cell apoptosis and defective neural development [3].

To aid in the proper diagnosis of UVJO and/or UPJO, one can utilize one or more of the readily available imaging modalities. An ultrasound can be used to assess pelvicalyceal dilation, renal cortical thinning, ureteric dilation, kidney size, cortical echogenicity, and bladder wall. Diuretic renography (MAG3 scan) is the agent of choice to assess for obstruction. The time required for clearance of 50% of the accumulated radionuclide is a half-life ( $t_{1/2}$ ) less than 10 min, while a  $t_{1/2} > 20$  minutes is suggestive, but not diagnostic, of obstruction [1]. A voiding cystourethrogram (VCUG) helps exclude other causes of upper tract dilation, such as, vesicoureteral reflux (VUR), urethral valves and ureteroceles. The coexistence of VUR and UPJO occurs in 8-14% of cases, and differentiating UVJO from a nonobstructed, nonrefluxing megaureter can be difficult.

Although UPJO is more easily identifiable, it can often obscure a distal narrowing or stricture. Hence, the identification of UVJO is often made post-operatively. McGrath et al. [2] reported the diagnosis of coexisting UPJO and UVJO preoperatively in 3 (21.4%) and postoperatively in 8 (57.1%) of their 14 patients. The correct diagnosis was made preoperatively in 5 of 11 patients (45.4%) by Pesce et al [4], in 5 of 14 patients (35.7%) by Cay et al. [5], and in 10 of 15 patients (66.7%) by Lee et al. [6].

Many cases of UVJO are diagnosed post-operatively due to various reasons. First, the obstruction at one end of the ureter can mask the second obstruction [2]. Second, many of the pediatric cases often resolve spontaneously without need for surgical intervention [7]. Third, the diagnosis may be overlooked. Thus, Cay et al. [5] advises that the presence of a coexisting distal obstruction should always be considered, especially since it can compromise the UPJO repair.

In our patient, the reverse scenario occurred where we identified the UVJO before the UPJO. In the presence of UPJO, where the obstruction is proximal, there is dilation of the renal pelvis with absence of hydronephrosis. Hence, it was logical to assume only UVJO since the initial studies of renal ultrasound showed bilateral hydronephrosis and the MAG3 scan and antegrade nephrostogram showed drainage of contrast in the proximal ureters. Thus, the patient underwent bilateral ureteral re-implantation first. But after the bilateral UVJO repair, the repeat antegrade nephrostogram showed bilateral UPJO as well, which was entirely unexpected. Perhaps the severe UVJO masked the findings of UPJO.

The aim of treatment in UPJO and/or UVJO is to preserve and/or prevent deterioration of the kidney function. Management can be either conservative and/or surgical depending on the severity of the obstruction. A dismembered pyeloplasty approach is the gold-standard treatment with the most favorable surgical outcome for UPJO. The complications of pyeloplasty include urinary tract infections, urinary extravasation and leakage, recurrent UPJO and stricture formation [1]. Ureteroneocystostomy is the more effective treatment for UVJO. However, with coexisting UPJO and UVJO, it can be difficult to decide which surgery is most appropriate first. Simultaneous procedures on the UPJ and UVJ are not generally recommended because of the potential negative effect on ureteral vascularity [6]. A preoperative retrograde pyelogram can help to define the anatomy of the ureter and visualize any other anomalies before the repair is done.

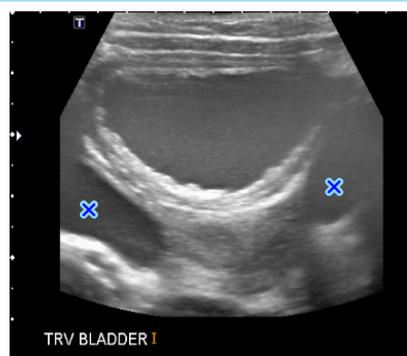
## Conclusions

Hydronephrosis is often due to UPJO and/or UVJO. Rarely, does one have a ureter with both UPJO and UVJO. This is the first reported patient that we know of that has concurrent bilateral UPJO and bilateral UVJO. When both UPJ and UVJ obstruction occur in the same ureter, it can raise dilemma in the diagnosis and management of the patient. However, by having a high index of suspicion for coexisting UPJ and UVJ obstructions, one can properly tailor therapy to salvage the kidney function. A multidisciplinary approach with a nephrologist and urologist remains crucial.

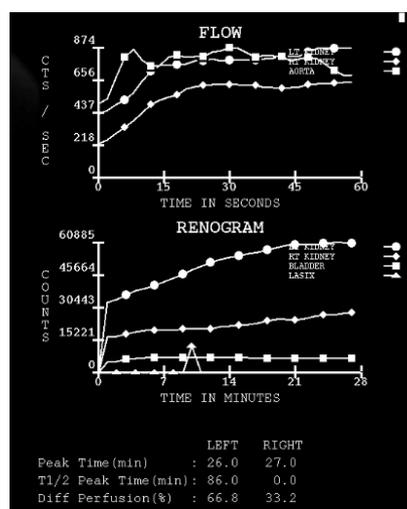
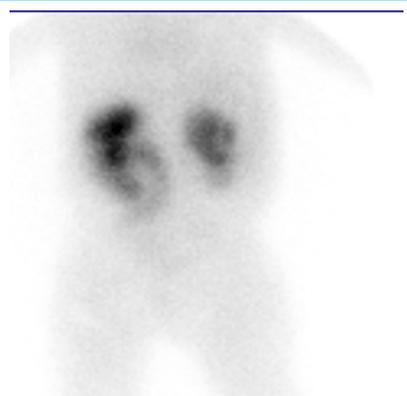
Figure 1: Radiologic Images of our Patient



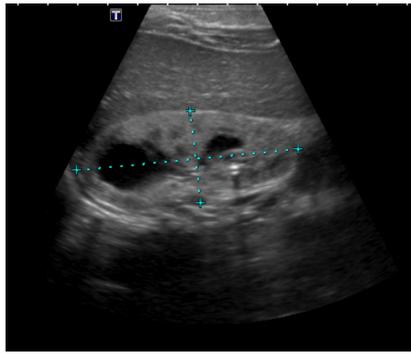
A. Initial renal ultrasound (RUS) with increased echogenicity of the right kidney and pyonephrosis. There is dilation of the proximal ureter (blue x)



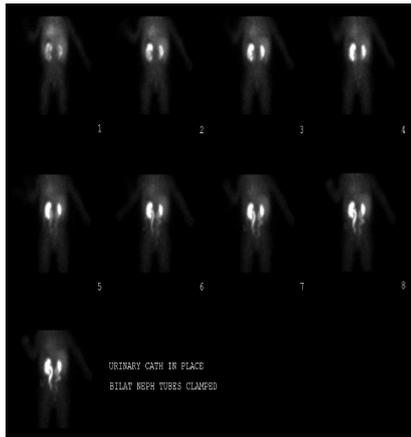
B. Initial bladder ultrasound: Presence of bladder wall thickening with bilateral hydroureters (blue x)



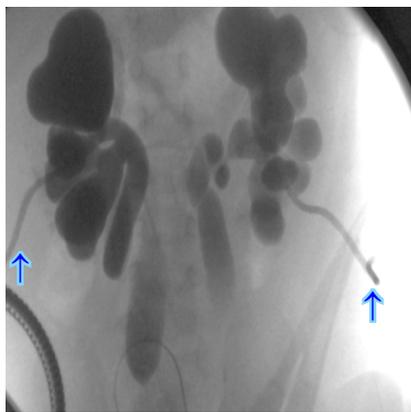
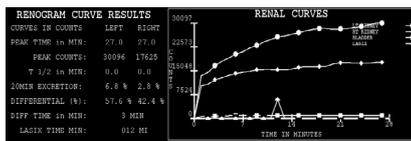
C. Initial Diuretic Renogram (MAG3 scan). Hydronephrotic kidneys, with no significant emptying after Lasix or gravity. Differential function is 67% on the left and 33% on the right.



D. RUS (right kidney) after bilateral nephrostomy tube placement. There is resolution of both hydronephrosis and pyonephrosis.



E. MAG3 scan repeated 2 weeks after nephrostomy tube placement. Image study is consistent with bilateral UVJ obstruction.



F. Anterograde nephrostogram shows bilateral UPJO post-bilateral ureteral re-implantation (clamped nephrostomy tubes are in place, see arrows)

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