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Case Report

# Bilateral hydroureteronephrosis: A neonatal presentation of bladder duplication ☆,☆☆

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ABSTRACT

In a male neonate with bilateral hydroureteronephrosis, the most common surgical diagnosis is posterior urethral valves. This case report describes a male infant with the same presentation, but caused by a very uncommon congenital anomaly.

The summation of different imaging modalities allowed a multidisciplinary team of colleagues to define the anatomy: bilateral duplex kidneys draining into separate urinary bladders. Only one of the bladders had an outlet, hence the obstructive uropathy to the right kidney led to total loss of function. The distended tortuous ureters produced a mass effect at presentation. This case acts as a reminder that complex congenital anomalies can mimic the presentation of more common conditions, and that they often require input from various specialists to diagnose the condition and guide its management.



## Introduction

Posterior urethral valves (PUV) are a serious cause of bladder outlet obstruction in male neonates and require urgent intervention. This is known to affect 1 in 4000-6000 male infants and is usually suspected by antenatal scans showing echogenic kidneys, with hydronephrosis and a small thickened bladder [1]. Postnatal ultrasound would confirm these findings, and in some patients there is associated urinary ascites.

Duplications of the urinary tract are well-recognized and occur in 1-3% of people. The most common variant is a duplex kidney which can range from a duplex collecting system connected to a single ureter (partial duplex), to duplex systems extending all the way into the bladder [2,3].

Bladder duplication is much less frequently reported, only really featuring in the literature within case reports [4,5]. It was

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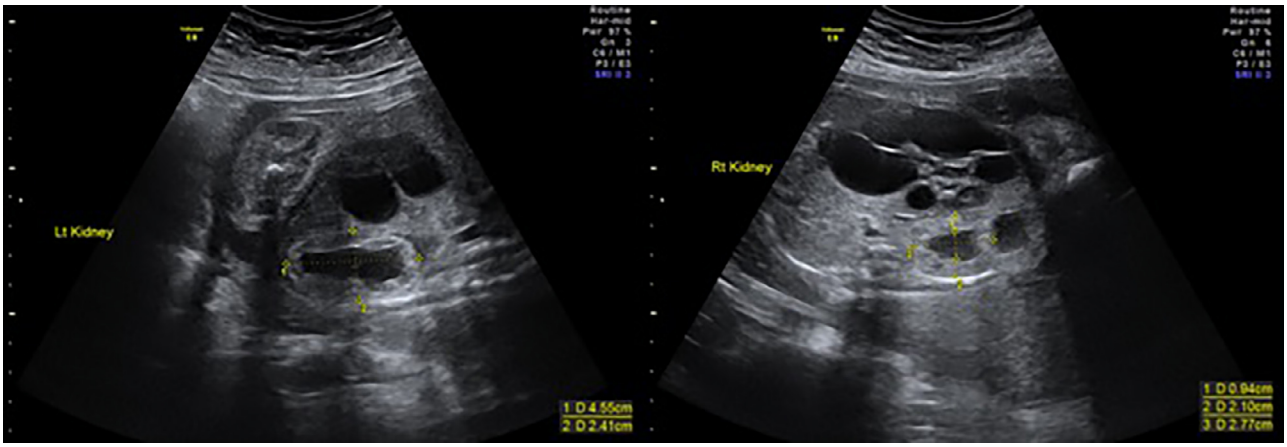


**Fig. 1 – (a) Antenatal ultrasound at 28 weeks gestation showing left hydronephrosis. (b) Antenatal ultrasound at 28 weeks gestation showing right hydronephrosis.**

first reported in 1961 by Abrahamson who described the variants and proposed a classification system [6]. From the sparse literature on this rare condition, it is seen more commonly in males than females at a ratio of 2.5:1 [7]. Bladder duplication can be identified on antenatal ultrasound scans, but the degree of detail that can be gathered at this point is variable [8]. The orientation of the duplication can be in the coronal plane, with one bladder in front of the other. It can also be duplicated in the sagittal plane, and in this variant the ipsilateral kidney drains into each of the bladders, which then drain via separate urethrae [9].

Bladder duplication is most commonly seen as a part of a wider lower uro-digestive tract anomaly [10]. In males this can include phallus and urethral duplication. In females this can include vaginal and uterine duplication. In both sexes it can include anorectal duplications. As it can be related to more clinically obvious anomalies, bladder duplication is most commonly discovered on screening imaging done in the neonatal period.

Scholtmeijer and Molenaar reported a series of duplication cases, the third of their cases demonstrated loss of function in the duplicated system that was obstructed [11].

To our knowledge, this report is the first to describe bilateral duplicated upper tracts draining into separated noncommunicating bladders, with only one side of the body having anatomical drainage through a urethra.

## Case report

A 36-week gestation male neonate was transferred to our institution with an antenatal sonographic diagnosis of bilateral hydroureteronephrosis (Fig. 1). His abdomen was markedly distended and he was unable to tolerate enteral feeds. His external genitalia had a normal appearance.

The 2 main unifying differential diagnoses for this presentation were posterior urethral valves with urinary ascites, and an intestinal duplication cyst causing a fluid filled mass and obstructing urinary drainage. The antenatal ultrasound findings could also have been consistent with bilateral high grade vesico-ureteric reflux, or bilateral vesico-ureteric junction obstruction with duplex kidneys.

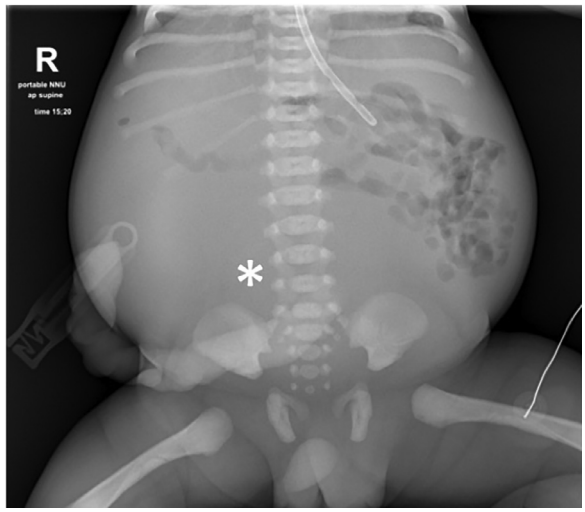


**Fig. 2 – Plain abdominal radiograph on day 0 of life demonstrating a right-sided abdominal mass displacing the bowel to the patients left (asterix at the center of the opacity).**

A plain abdominal radiograph showed a bowel gas pattern suggesting a right sided mass (Fig. 2). In view of the antenatal diagnosis of bilateral hydronephrosis, a urinary catheter was inserted and urinary tract ultrasound was performed. This demonstrated bilateral duplex kidneys (Fig. 3a), bilateral hydroureteronephrosis and a cystic structure posterior to the orthotopic catheterized bladder (Fig. 3b+c).

To better delineate the anatomy an MRI scan was performed. This showed that the mass effect came from distended tortuous right sided ureters, appearing to enter a separate cystic structure posterior to the orthotopic urinary bladder (Fig. 4). The differential of an enteric duplication cyst was therefore dismissed.

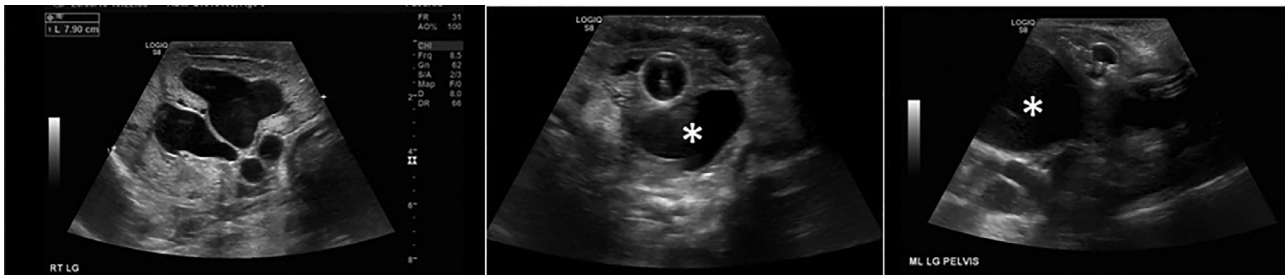


**Fig. 3 – (a) Longitudinal ultrasound image showing the duplex right kidney, and suggesting a loss of cortico-medullary differentiation. (b) Axial ultrasound image showing a section of the cystic mass (asterix) posterior to the catheterized orthotopic bladder. (c) Sagittal ultrasound image showing the ectopic bladder (asterix) not decompressing despite urethral catheterization, which is visualized superiorly.**

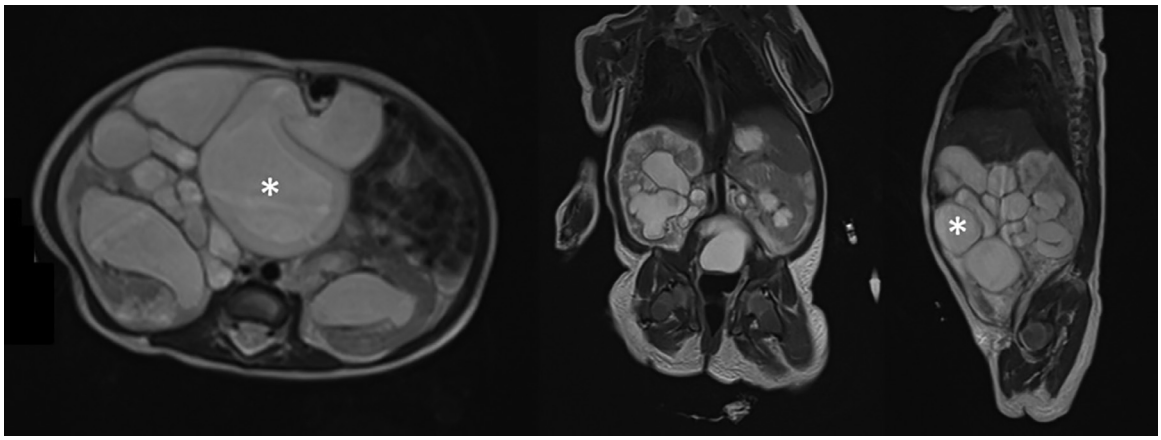

**Fig. 4 – Axial, coronal, and sagittal sections of a T2-weighted MRI showing the duplex right kidney and the mass effect of the dilated tortuous right sided obstructed ureters (asterix on axial and sagittal images).**

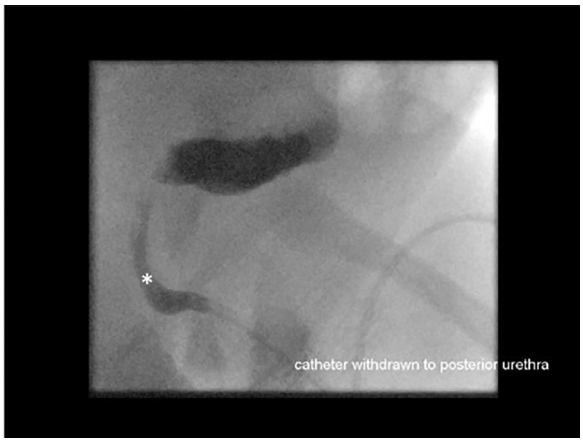


**Fig. 5 – Lateral view of the micturating cystourethrogram, showing contrast in the orthotopic bladder, and the normal caliber urethra (asterix).**

A micturating cystourethrogram (MCUG) showed a urethra of normal caliber, which excluded PUV's (Fig. 5) and it also did not show any flow into the posterior cystic structure. There was reflux of contrast into the left ureter. His right kidney and ureter remained distended despite drainage of the orthotopic bladder. He had a cystoscopy to assess the right ureteric orifice and potentially to improve the drainage from the right kidney, but the ureteric orifice could not be identified within the orthotopic bladder. We concluded that the posterior cystic structure was a second bladder into which the right ureters were draining. No connection could be seen between the orthotopic and the duplicated bladders. A vesicostomy was therefore formed from the ectopic bladder to facilitate decompression of the right sided system.

Ultrasound surveillance showed that the right sided duplex kidney remained hydronephrotic, therefore nephrostomies were inserted to drain the two moieties and to allow antegrade assessment. Nephrostograms (Fig. 6) showed that each moiety of the right sided duplex system drained into the ectopic bladder only, with no contrast passing into the orthotopic bladder. A retrograde contrast study completed urethrally showed contrast passing into the orthotopic bladder and refluxing into the left ureter, this time showing no contrast passing into the ectopic bladder. The lack of connection was confirmed with instillation of methylene blue via the urethral catheter into the orthotopic bladder, and no dye was seen to come out through the vesicostomy.

The patient was discharged with the vesicostomy draining the right sided system, and passing urine urethrally from his

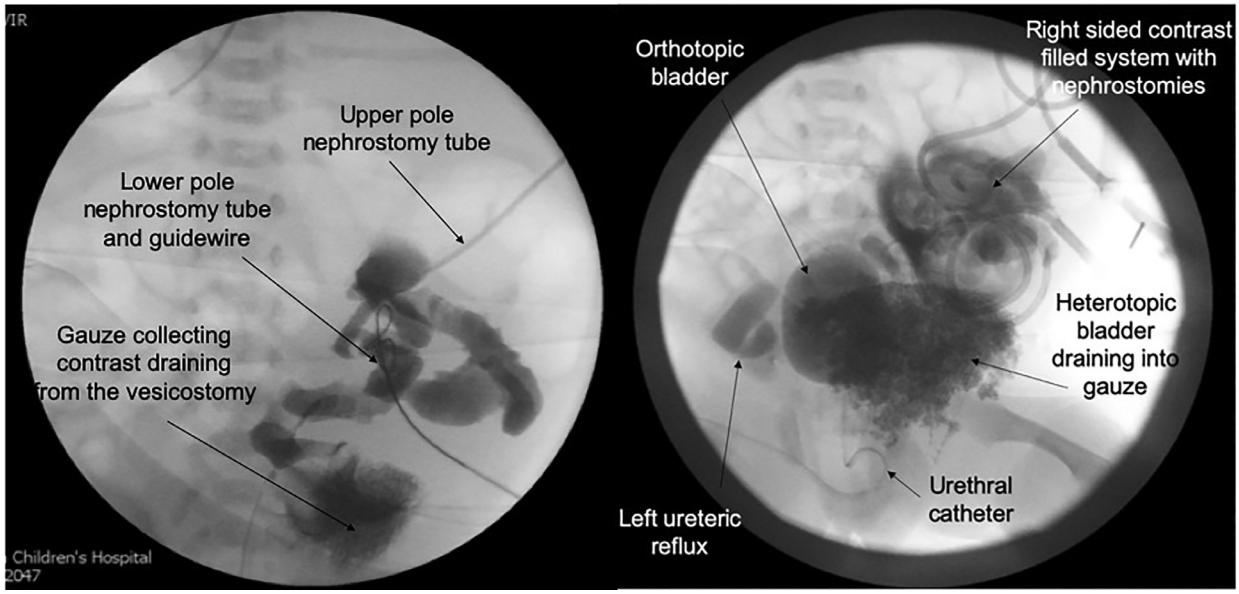


**Fig. 6 – (a) Right kidney antegrade nephrostogram (prone): guidewires in each moiety, with contrast draining into the ectopic bladder and then draining out via vesicostomy. (b) Retrograde cystogram (prone): right upper and lower pole nephrostomies in situ, retrograde cystogram through the urethral catheter, showing reflux into the left ureter.**

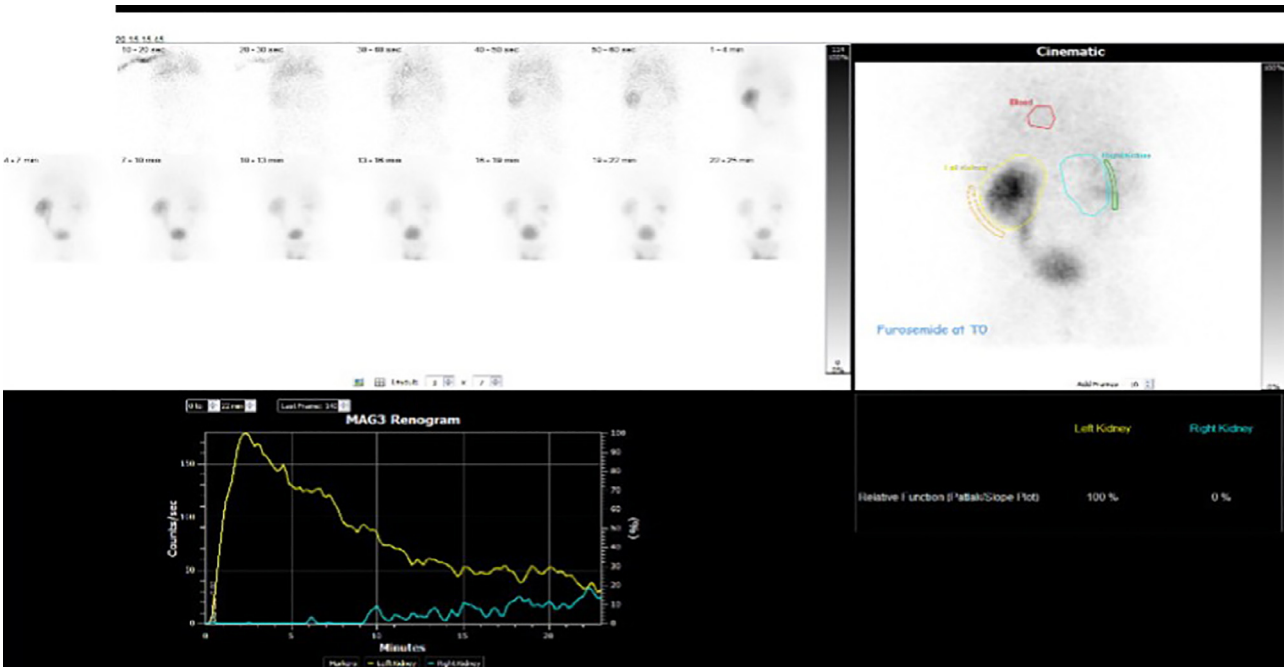


**Fig. 7 – MAG3 renogram with left/right differential function, showing no functional activity in the right kidney (0%), and adequate drainage of the left kidney on the drainage curve.**

functional orthotopic left sided system. At 6 months of age a MAG3 (mercaptoacetyltriglycine) renogram confirmed that the right kidney had no function (Fig. 7) likely due to antenatal right sided obstructive uropathy.

The key surgical intervention performed for the patient was formation of a vesicostomy which allowed decompression of the right sided system. As there was no function in the right kidney, he was planned for a laparoscopic right nephroureteroectomy. In addition, he will also have the ectopic bladder excised via a transperitoneal laparoscopic route.

## Discussion

Bladder duplication is a rare congenital anomaly that can be diagnosed antenatally, symptomatically, or incidentally on

postnatal imaging. It can be seen as a part of wider uro-genito-digestive duplications, but can also be limited to an anomaly of the abdominal urinary tract.

This case demonstrates that complex congenital anomalies require multiple imaging techniques to be able to adequately delineate the structure and function of the urological system. Investigations were able to reveal bilateral duplex kidneys draining into separate bladders. The nonfunctioning right kidney was draining into a blind-ending, ectopic bladder and we plan to remove this complete system in due course. It was only through the close working between radiologists, interventional radiologists, and urologists, that we were able to gather the information we required to inform the management of this infant.

When investigating infants with urinary tract duplications, clinical examination can reveal duplications of the external genitalia such as penile duplication. In those patients without genital duplication, such as in this case report, imaging is essential to define the extent of the duplication.

Ultrasound is useful for antenatal imaging and may demonstrate hydronephrosis, or a pelvic cystic structure. Postnatal ultrasound is readily available, can be repeated over time, and gives anatomical information about the kidneys, ureters, and bladders. This would be our suggested first-line imaging modality. MCUG is a useful test to exclude other diagnoses such as vesico-ureteric reflux and posterior urethral valves. It also would demonstrate most connections between bladders if present. If the anatomy remains unclear, MRI provides detail of the anatomy of the urinary tract by using either T2 weighted, or intravenous contrast sequences as a magnetic resonance urogram to improve clarity. In infants this will require general anesthesia to ensure the child doesn't move, thereby improving the quality of the images. Antegrade and retrograde pyelograms can be performed, to demonstrate the anatomy and drainage of the urinary tract. Facilitating this by the placement of nephrostomies in neonates and in duplex systems however requires a skilled interventional radiologist.

Although not performed in this case, intravenous urography (IVU) can be considered as a dynamic imaging modality which would outline the anatomical relationship of functioning elements in the urinary tract. Finally, nuclear imaging with MAG3 renograms can be used to determine function and drainage from the kidneys. This provides essential information that will guide further treatment.

Treatment for bladder duplication depends on the extent of the anomaly as they can be a part of a complex urinary/genital/digestive duplication. We would therefore advise discussion in a multidisciplinary team before any surgical management is undertaken. After functional imaging is completed, if the duplicated bladder subtends a nonfunctioning kidney, then a cystectomy and nephro-ureterectomy would be the advised management. If the bladder is connected to a functioning kidney, then initial drainage needs to be established, followed by later ureteric reimplantation into the orthotopic bladder and excision of the duplicated bladder.

## Learning points

- Urinary tract duplications are seen often in a tertiary pediatric urology service, but bladder duplications remain a rare but recognized variant.
- Posterior urethral valves remain the predominant cause of bilateral hydronephrosis in a male neonate that would need early intervention, and should therefore be investigated with a MCUG.
- Investigating rare congenital urinary tract anomalies is reliant on a close working relationship between pediatric diagnostic radiologists, pediatric interventional radiologists, and pediatric urologists.

## Informed consent

Written informed consent was obtained from the patient(s) for publication of this case report, including accompanying images.

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