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Lower-Pole Ureterocele With Upper-Pole Ureteropelvic Junction Obstruction in a Complete Duplex Collecting System: A Case Report

Asher Wen [1], Bryant Tran [2], Michael Ernst [3]

1. Urology, Stony Brook Medicine, Stony Brook, USA 2. Radiology, Stony Brook Medicine, Stony Brook, USA 3. Pediatric Urology, Stony Brook University Hospital, Stony Brook, USA

Corresponding author: Asher Wen, wenasher1@gmail.com

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Abstract

The duplication of the collecting system is a rare congenital anomaly that is detected prenatally or at birth through ultrasound. We report a six-month-old female patient demonstrating a lower-pole ureterocele with upper-pole ureteropelvic junction obstruction in a complete left duplex collecting system. A voiding cystourethrogram and magnetic resonance urogram showed complete duplication of the collecting system with respective upper and lower moieties of the left kidney. Subsequently, intraoperative cystoscopy and fluoroscopy confirmed the anatomical relation of the ureterocele and its connection to the lower renal moiety. The ureterocele was resected and a pyeloplasty was performed to correct the upper moiety ureteropelvic junction obstruction. Cystoscopy and retrograde pyelography may be necessary to definitively assign duplicated ureters to their corresponding renal moieties when cross-sectional imaging is inconclusive.

Categories: Urology, Pediatrics, Anatomy
Keywords: congenital anomalies of the kidney and urinary tract, duplex collecting system, pediatric urology, ureterocele, ureteropelvic junction obstruction, vesicoureteral reflux, weigert-meyer law

Introduction

Duplication of the collecting system and ureter represents one of the most identified congenital anomalies of the urinary tract [1]. The reported prevalence is approximately 0.7-1% in unselected populations [1,2], though true prevalence is difficult to ascertain because many cases remain asymptomatic and may be discovered incidentally years after.

Duplicated systems occur with similar frequency unilaterally and bilaterally and are approximately twice as common in females as in males [1,2]. While most individuals with a duplicated collecting system remain asymptomatic, clinically significant cases are often associated with vesicoureteral reflux and obstructive pathology. Symptomatic patients may present with recurrent urinary tract infections, hydronephrosis, or renal parenchymal injury [1,3].

Evaluation typically involves a combination of ultrasonography, voiding cystourethrogram (VCUG), functional renal imaging, and cross-sectional imaging to define urological anatomy and drainage patterns. Definitive management is individualized and may include surgical intervention in patients with recurrent infections, impaired drainage, or evidence of progressive renal dysfunction [1,3].

Case Presentation

This is the case of a six-month-old female patient with a previous concern for a congenital renal anomaly detected on prenatal ultrasound, suspected to be a multicystic dysplastic kidney. About a week later, postnatal ultrasound demonstrated a left-sided duplex kidney with severe hydroureteronephrosis (HUN) with a possible ureterocele. A VCUG was subsequently performed and demonstrated a left-sided grade IV vesicoureteral reflux with tortuosity (Figure *1*). Amoxicillin prophylaxis was initiated and continued for three months. Notably, the patient was voiding well according to her parents and had no other medical issues. The patient was monitored closely outpatient for the management of possible symptoms and for continued diagnostic clarification.

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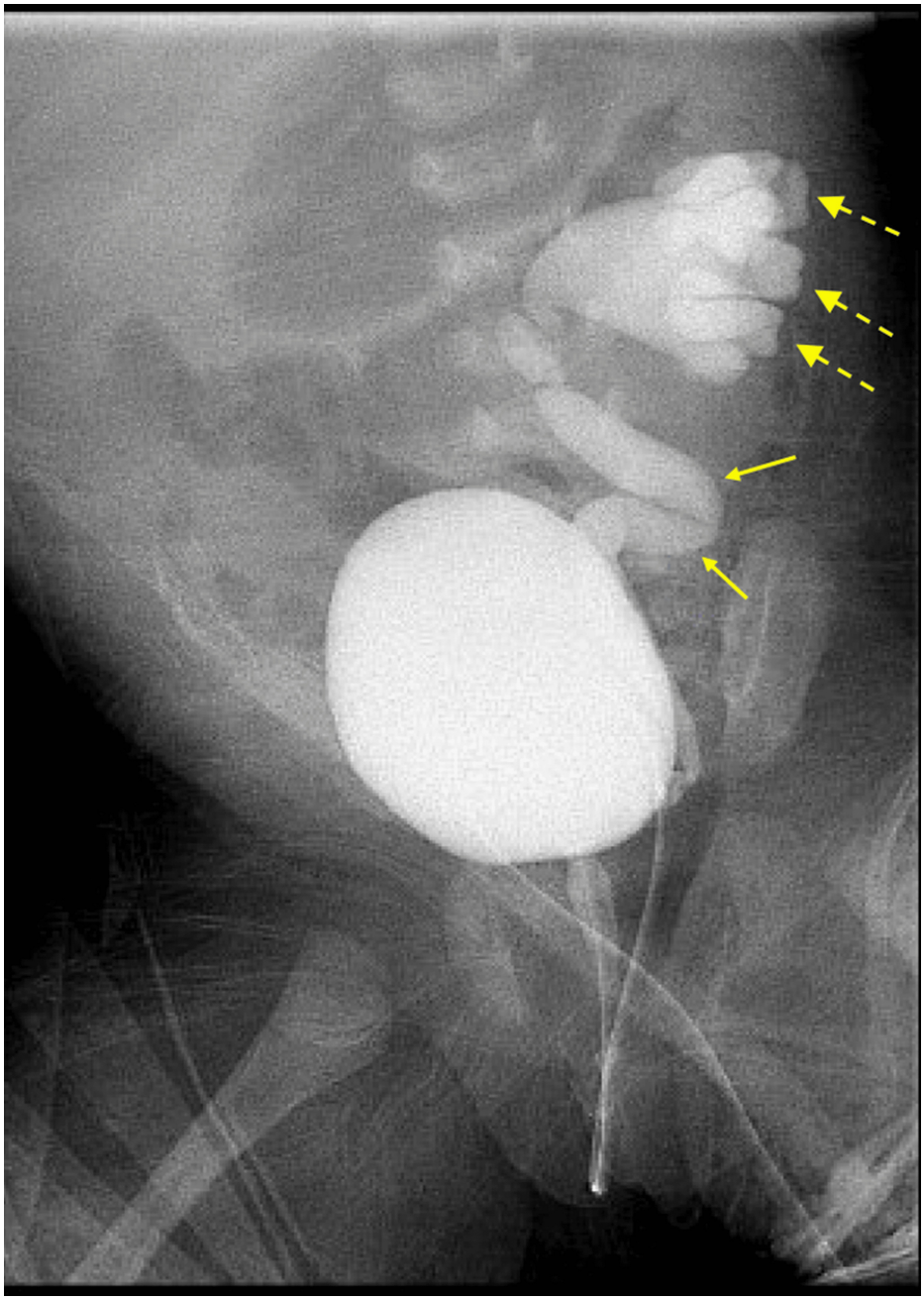


FIGURE 1: Voiding cystogram showing evidence of left grade IV vesicoureteral reflux with tortuosity and moderate dilation of the left ureter (solid arrows) and blunting of the fornices (dashed arrows).

At two months of age, a diuretic renal scintigraphy (Lasix renogram) was obtained to quantitatively assess renal function and urinary drainage. The study demonstrated bilateral hydronephrosis, with high-grade obstruction involving the left kidney and low-grade partial obstruction involving the right kidney. Following Lasix administration, the left duplex kidney demonstrated greater tracer retention and impaired excretion compared with the right kidney. This is reflected by the overall calculated effective renal plasma flow (ERPF) of 145 ml/min. Detailed diuretic renogram findings, including region-of-interest (ROI) curves, are provided in Appendices A-D. In addition, radiology noted a photopenic defect in the upper-pole moiety of the left kidney, raising concern for functional obstruction.

To further delineate the anatomy, a magnetic resonance urogram (MRU) was obtained at four months of age. Imaging showed a duplicated left kidney with suspected ureteropelvic junction obstruction (UPJO) in the upper-pole moiety, which corroborated with previous findings (Figure *2*). The lower-pole moiety ureter was dilated and extended distally to the posterior bladder wall, where a ureterocele-like configuration was identified (Figures *3*-*4*). In comparison, the upper-pole moiety ureter is normal in caliber, but its distal

insertion could not be found. Given the concerns of functional obstruction, the patient was scheduled for robotic-assisted left upper-pole pyeloplasty at six months of age.

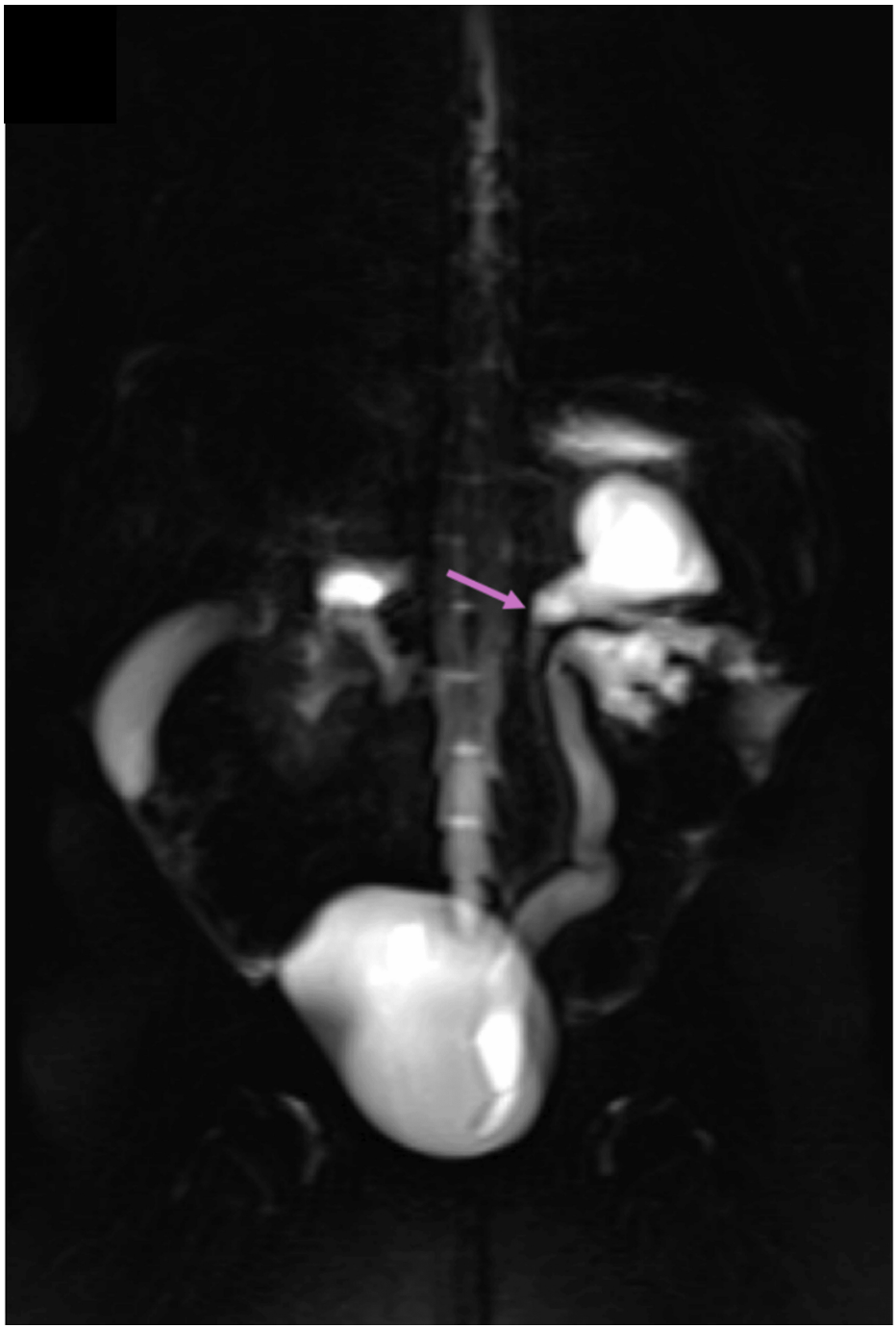


FIGURE 2: T2 MRI coronal image demonstrating duplicated left renal collecting systems with dilation of the upper-pole collecting system and an ureteropelvic junction obstruction in the left upper-pole moiety (pink arrow).

MRI: magnetic resonance imaging



FIGURE 3: T2 MRI sagittal image demonstrating a ureterocele pattern of the lower-pole ureter at its insertion into the bladder base above the neck of the bladder (yellow arrow).

MRI: magnetic resonance imaging

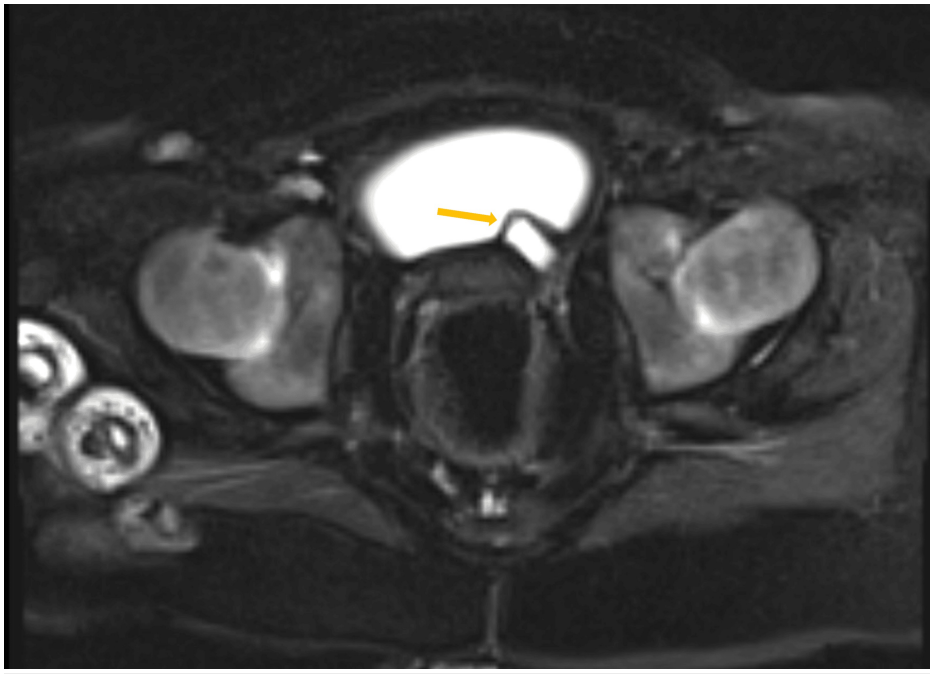


FIGURE 4: T2 MRI axial image demonstrating distal dilation of the lower-pole ureter terminating in an intravesical ureterocele (orange arrow).

MRI: magnetic resonance imaging

Perioperative findings

On cystourethroscopy, a left-sided ureterocele-like structure was identified. The lower-pole ureteral orifice is located within the ureterocele. Contrast injection through this orifice during retrograde pyelography resulted in lower moiety opacification (Figure *5*). Following a transurethral resection of the left ureterocele, a double-J ureteral stent was placed retrograde in the lower-pole ureter. The left upper-pole ureteral orifice was unable to be identified retrograde. UPJO upper moiety pyeloplasty was performed laparoscopically with antegrade left upper-pole moiety stent placement.



FIGURE 5: Intraoperative retrograde pyelogram with contrast dilation of ureterocele demonstrating the opacification of the lower-pole ureter and collecting system.

Postoperative course and follow-up

Postoperative abdominal radiography and renal ultrasonography raised concern for ureteral stent migration, with possible coiling within the upper moiety ureter. Interventional radiology subsequently performed an antegrade nephrostogram and exchanged the ureteral stent for a percutaneous nephroureteral catheter (PCNU). The remainder of the postoperative course was uncomplicated, and the patient was discharged two days after the intervention. After one month, for stent removal, the cystoscopy done for stent removal shows the relevant anatomy with both stents (Figure *6*) and a cystic remnant of a lower moiety ureterocele (Figure *7*) seen.

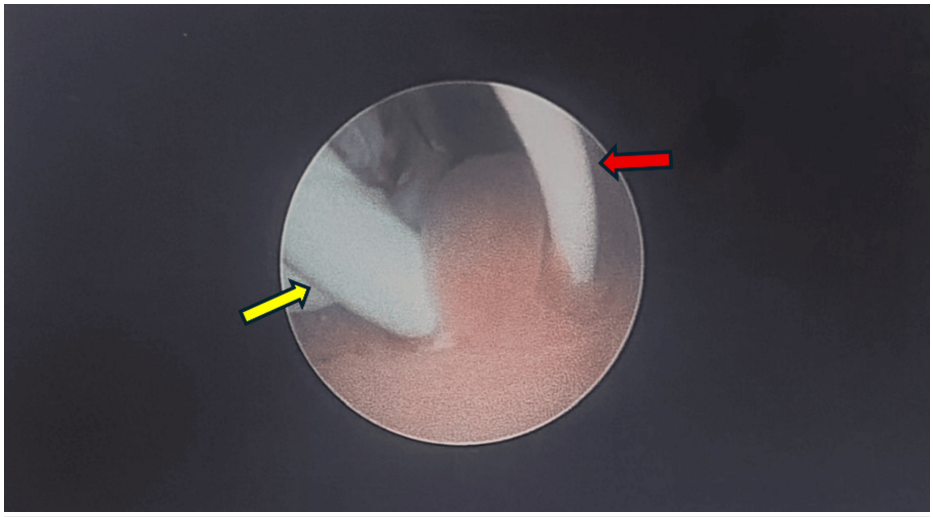


FIGURE 6: One-month postoperative cystoscopy image taken after pyeloplasty and post-transurethral resection of the cystic structure with both stents placed. The leftmost stent (yellow arrow) is placed into the upper moiety, and the rightmost stent (red arrow) is placed into the lower moiety.

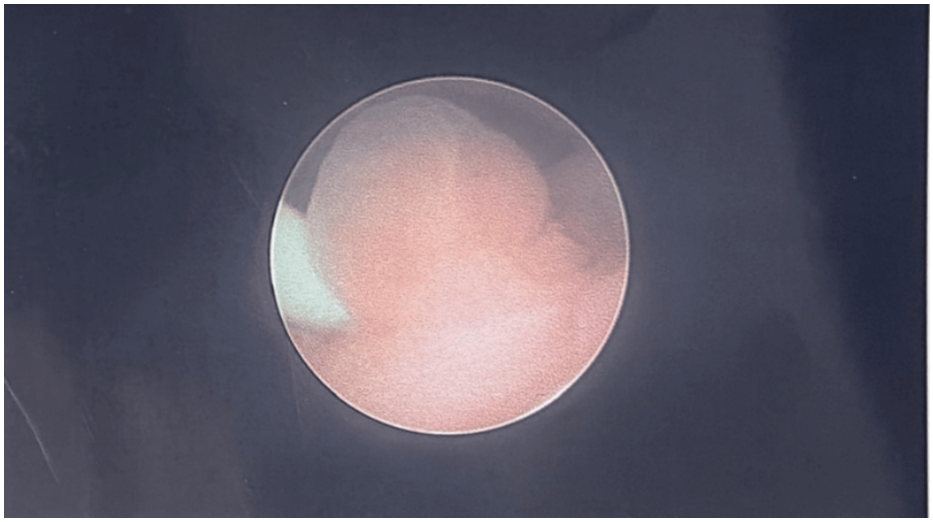


FIGURE 7: One-month postoperative cystoscopy image of a left-sided remnant cystic structure after lower moiety stent removal.

Serial renal ultrasonography and anteroposterior (AP) renal pelvic diameter measurements were used to monitor the severity of hydronephrosis. Preoperatively, the AP diameter measured 1.55 cm in the upper moiety and 0.3 cm in the lower moiety. At one month postoperatively, these measurements were 1.5 cm and 0.4 cm, respectively. By the three-month follow-up, the AP diameter had decreased to 0.6 cm in the upper moiety, while hydronephrosis of the lower moiety had resolved. At the three-month outpatient follow-up, the patient remained clinically well without recurrent urinary tract infections or other reported complications. The PCNU was removed at three months.

Discussion

Duplicated collecting systems result from abnormal ureteric bud development and are commonly explained by the embryologic principles described by the Weigert-Meyer law [4]. In a complete duplex kidney, the upper-pole ureter ectopically inserts inferiorly and medially and is predisposed to obstruction, while the lower-pole ureter inserts more laterally and superiorly and is more commonly associated with vesicoureteral reflux [1].

In the present case, preoperative imaging demonstrated upper-pole UPJO and lower-pole vesicoureteral reflux, along with an additional finding suggestive of a ureterocele. MRU demonstrated a cystic structure at the left vesicoureteral junction with associated distal ureteral dilation, which was radiologically interpreted as arising from the lower-pole ureter. However, the tortuous course of the duplicated ureters made it difficult to trace the structure to its respective renal moiety on preoperative imaging alone. Subsequent intraoperative cystoscopy demonstrated a cystic structure protruding into the bladder lumen, consistent with a ureterocele, with a ureteral orifice identified within the structure. Retrograde pyelography performed through this orifice, prior to ureterocele incision, demonstrated the opacification of the lower renal moiety, which confirmed its communication with the lower-pole collecting system.

Compared to upper moiety ureteroceles, lower moiety ureteroceles have rarely been described in the literature. Arap et al. reported what they identified as the second such case in 1983, followed by Harb et al., who described the third reported case in the English literature in 1999 [5,6]. Harb et al. identified a lower-pole ureterocele associated with grade IV vesicoureteral reflux in a completely duplicated left collecting system. Similar to the present case, retrograde pyelography was used to confirm the association of the ureterocele with the lower-pole moiety. Across the literature identified in our review of duplex collecting systems, the relationship between the ureterocele and the corresponding renal moiety was often not explicitly confirmed by diagnostic evaluation, with ureteroceles instead characterized primarily by their anatomic location [7,8].

Conclusions

This pediatric case describes a lower-pole ureterocele associated with an upper-pole UPJO in a completely duplicated collecting system, in contrast to the more typical association of ureteroceles with the upper-pole moiety. In the setting of a duplex collecting system, the tortuosity and crossing of the ureters may further complicate anatomical assignment. Cystoscopy and retrograde pyelography with renal scintigraphy remain important for definitive anatomical assignment, particularly when imaging findings appear to deviate from established embryologic expectations.

Appendices

Appendix A

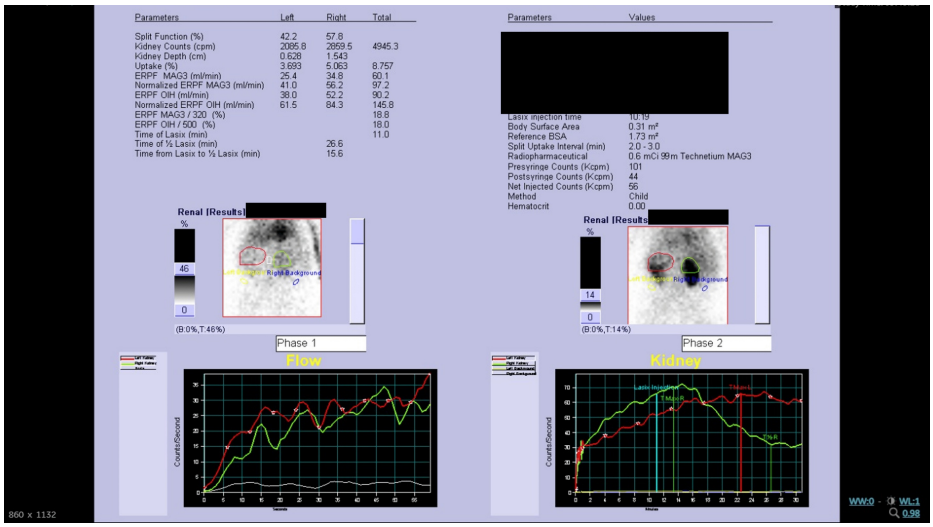


FIGURE 8: Preoperative diuretic renogram and region-of-interest curves at two months.

ERPF: effective renal plasma flow; BSA: body surface area; OIH: orthoiodohippurate; MAG3: mercaptoacetyltriglycine

Appendix B

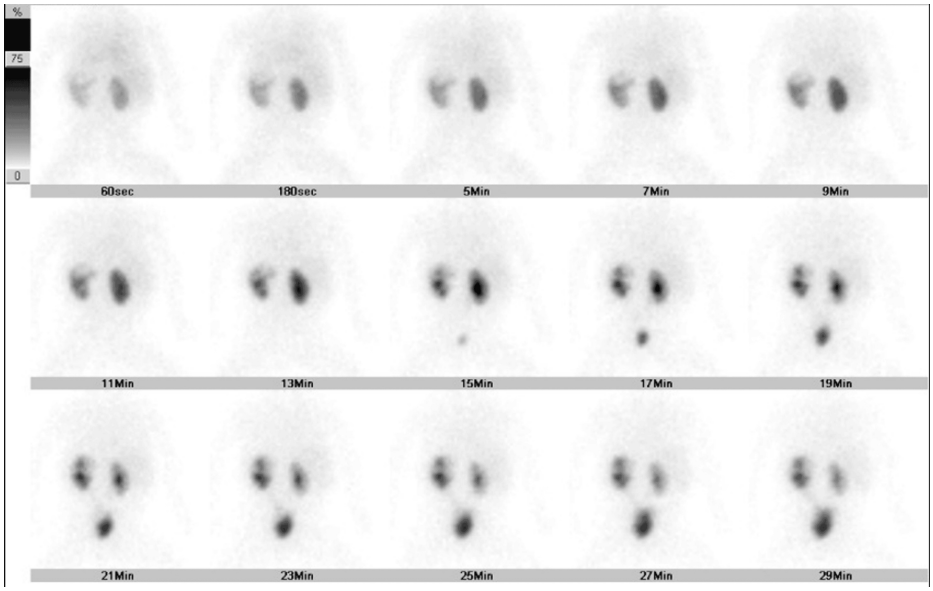


FIGURE 9: Serial renal scintigraphy images from diuretic renogram at two months.

Appendix C

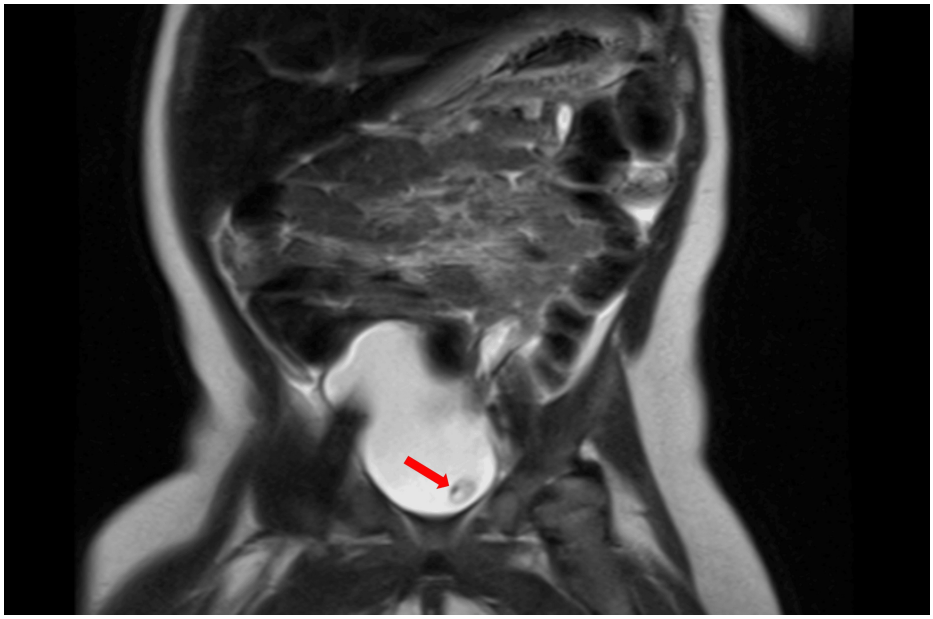


FIGURE 10: Supplemental T2 MRI coronal view demonstrating the relationship of the left-sided cystic structure (red arrow) in relation to the bladder.

MRI: magnetic resonance imaging

Appendix D

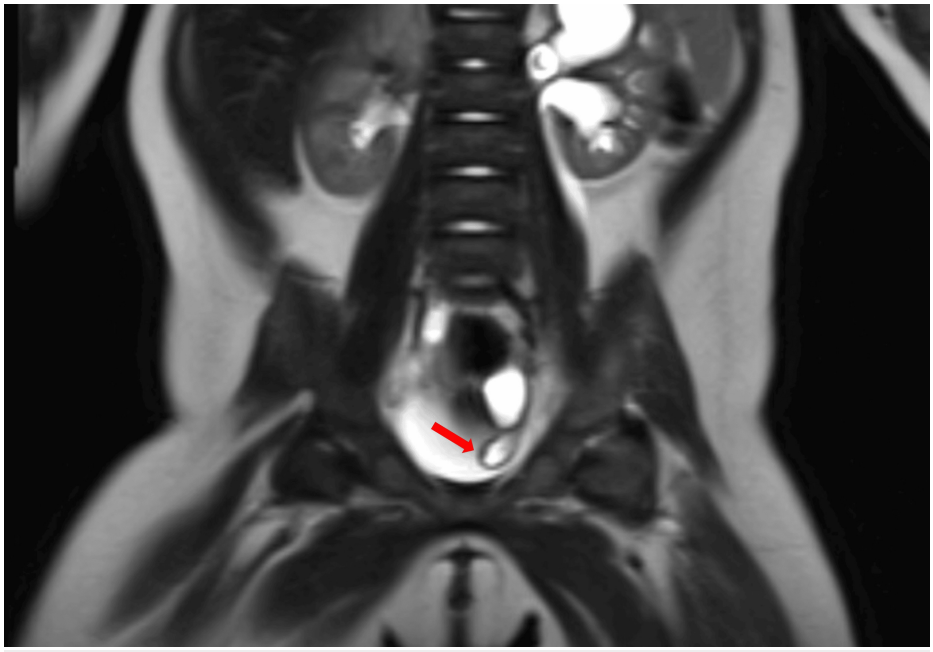


FIGURE 11: Supplemental T2 MRI coronal oblique view demonstrating a left-sided cystic protrusion (red arrow) into the bladder lumen at the inferior portion of the bladder.

MRI: magnetic resonance imaging

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Asher Wen, Michael Ernst

Acquisition, analysis, or interpretation of data: Asher Wen, Bryant Tran

Drafting of the manuscript: Asher Wen

Critical review of the manuscript for important intellectual content: Asher Wen, Michael Ernst, Bryant Tran

Supervision: Michael Ernst

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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