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2025-01
Elsevier
https://hdl.handle.net/10539/43843
Mohammed Salah E. Khalifa Salem, Abdul Alherek, Daniel DaSilva, Jacob Varghese, Alain Mwamba Mukendi, Unprecedented Association: Bilateral UPJ obstruction with grade 3 hydronephrosis caused by Type 2 circumcaval right ureter and left lower pole crossing vessels, Urology Case Reports, Volume 59, 2025, 102958, ISSN 2214-4420, https://doi.org/10.1016/j.eucr.2025.102958.

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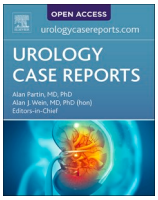
Urology Case Reports 59 (2025) 102958



Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr



Functional Urology



Unprecedented Association: Bilateral UPJ obstruction with grade 3 hydronephrosis caused by Type 2 circumcaval right ureter and left lower pole crossing vessels

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ARTICLE INFO

Keywords:
Circumcaval ureter
Hydronephrosis
Vascular anomalies
UPJ obstruction
Crossing vessels

ABSTRACT

Bilateral ureteropelvic junction obstruction resulting from distinct vascular anomalies on each side, with a pre-ureteric vena cava on the right and crossing vessels on the left, has not been previously documented in the literature. Even more intriguing is the association between a grade 3 hydronephrosis and a type 2 circumcaval ureter. This unprecedented report discusses this association and its management.

1. Introduction

Ureteropelvic junction (UPJ) obstruction is a common urological disorder, occurring in 1 in 1000–1500 live births, although its incidence in adults is not well-defined. Patients can present at any age, typically with flank pain, though it may also be detected incidentally.[1,2]

UPJ obstruction encompasses various obstructive processes, classified as intrinsic or extrinsic. Intrinsic causes include stenotic or aperistaltic segments within the ureter, while extrinsic causes, such as an accessory lower-pole renal artery, compress the UPJ externally. The role of crossing vessels in causing obstruction remains controversial.[1]

Bilateral ureteropelvic junction obstruction is a rare and complex condition often associated with significant morbidity.[2]

A retrocaval ureter is a rare congenital condition where the ureter is abnormally positioned posterior to the inferior vena cava (IVC). This anomaly arises from the development of the inferior vena cava rather than the urinary system.[3,4] Accessory renal arteries, representing persistent mesonephric arteries, are common and arise from the aorta. Numerous cases in the literature have reported that aberrant or accessory vessels often enter the lower pole of the kidney, positioning themselves in a way that obstructs urine outflow at the UPJ.[5]

Vascular abnormalities of different origin and nature as concomitant causes of UPJ obstruction have not been previously described. This case report presents a unique instance of bilateral UPJ obstruction caused by two distinct vascular anomalies: a type 2 pre-ureteric vena cava with grade 3 hydronephrosis on the right and crossing vessels on the left.

2. Case presentation

A 21-year-old female with a history of retroviral disease, treated since birth and virally suppressed, also has a history of congenital renal conditions or anomalies of which she was unable to recall details due to her young age at the time of diagnosis and was subsequently lost to follow-up.

Presented in 2022 to the Urology department with bilateral flank pain. Laboratory tests indicated mild renal dysfunction with urea levels at 7.2 and creatinine at 107. A renal ultrasound revealed bilateral hydronephrosis assessed as moderate on the right and mild to moderate on the left. A CT Urogram performed demonstrated a right retrocaval ureter classified as a Type 2 with proximal hydronephroureterosis (Fig. 1a–f), accessory left renal artery and vein supplying and draining the lower pole, respectively, with these vessels crossing the proximal left ureter, likely causing obstruction of the left kidney. Additionally, a retro aortic left renal vein was noted.

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https://doi.org/10.1016/j.eucr.2025.102958
Received 27 December 2024; Received in revised form 15 January 2025; Accepted 20 January 2025
Available online 22 January 2025
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Subsequently, a cystoscopy and bilateral retrograde pyelogram were performed, revealing bilateral proximal ureteric narrowing with moderate hydronephrosis. Bilateral JJ stents were placed. 6 months later, renal function improved and hydronephrosis resolved therefore stents were removed under local anaesthesia. A MAG-3 renogram conducted a year later, due to a long waiting list and limited imaging modalities and facilities, showed that both kidneys had suboptimal function, with slow drainage on the left and minimal drainage on the right. The differential function was nearly equal between the kidneys, with the left at 50.2 % and the right at 49.8 %.

In early 2024, due to increasing symptoms and worsening renal function, another retrograde pyelogram and bilateral JJ stent insertion were performed, with plans for an explorative laparotomy and bilateral pyeloplasty. Six months later, an explorative laparotomy and bilateral pyeloplasty were carried out through a midline transperitoneal abdominal incision. On the right side, dissection of the ascending colon was performed. The inferior vena cava (IVC) and the ureter were identified (Fig. 2). The ureter was dissected and cut at the UPJ before being placed anterior to the IVC, followed by a simple pyeloplasty (Heineke), which was performed with a double-J (DJ) stent in place (Fig. 3). On the left side, the descending colon was dissected, and the ureter was identified. The crossing vessels were then identified (Fig. 4), and the stenotic segment was resected. A reanastomosis was performed with a DJ stent using the Anderson-Hynes technique. The patient had an uneventful hospital stay and was discharged home with a transurethral catheter, with both drains removed. Her urine output, renal function and septic markers were normal at discharge and remained within normal ranges on subsequent follow-up, with no symptoms reported.

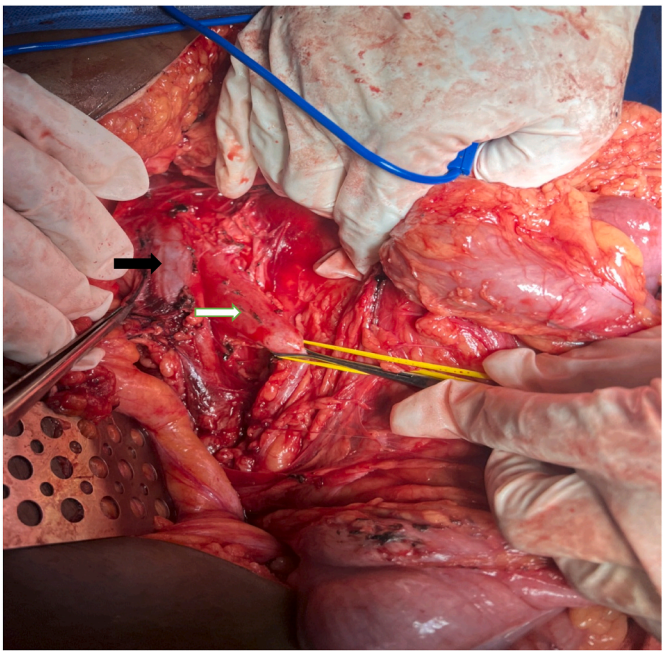


Fig. 2. Intraoperative Images Before Pyeloplasty. Illustrating the right proximal ureter (white arrow) emerging from behind the inferior vena cava (Black arrow), characteristic of a type 2 retrocaval ureter..

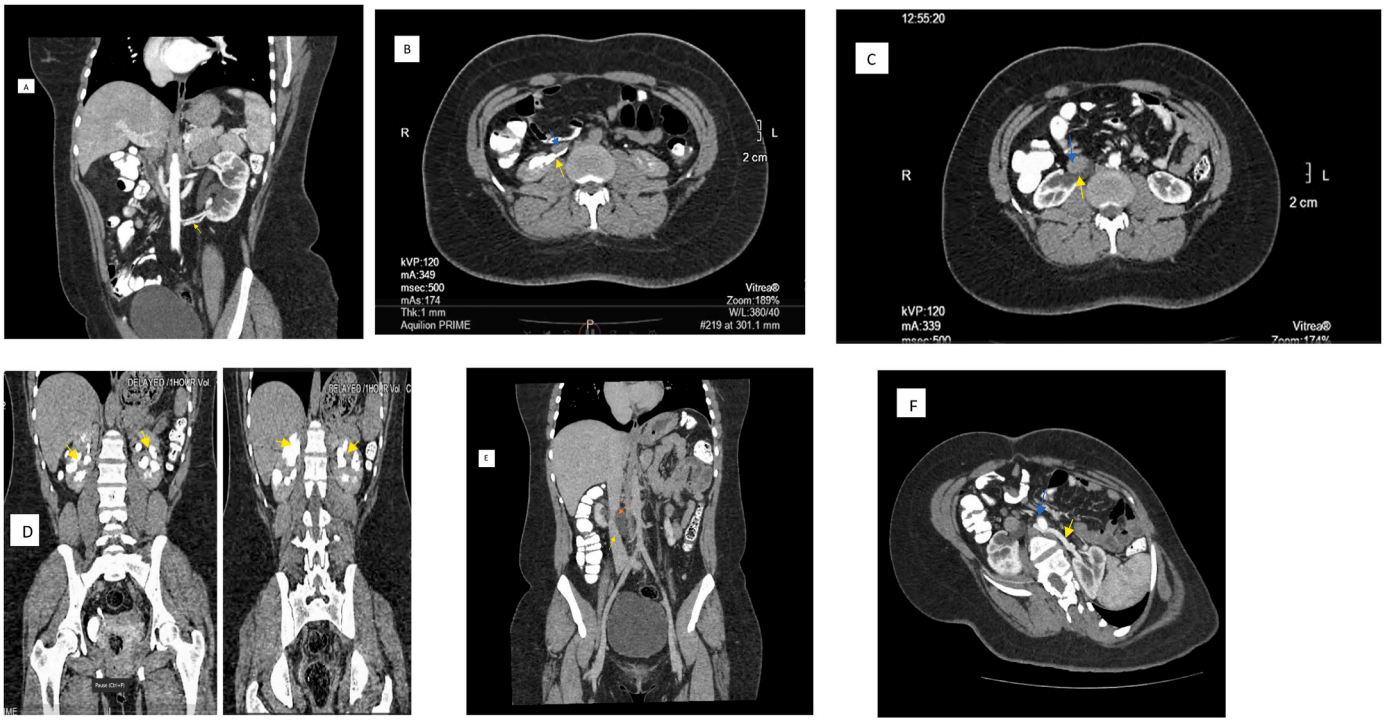


Fig. 1. a-f: Preoperative computed tomography images. Fig. 1a: Coronal view - arterial phase of CT abdomen with intravenous and oral contrast showing left lower pole accessory renal vessels (yellow arrow) crossing the left ureter anteriorly. Fig. 1b: Axial view - delayed phase of CT abdomen with oral and intravenous contrast showing right renal pelvis (yellow arrow) coursing behind the IVC (blue arrow) consistent with type 2 retrocaval ureter. Fig. 1c: Axial view – arterial phase of CT abdomen with oral and intravenous contrast showing right renal pelvis (yellow arrow) coursing behind the IVC (blue arrow) consistent with type 2 retrocaval ureter. Fig. 1d: Coronal view – delayed phase of CT abdomen with oral and intravenous contrast showing bilateral hydronephrosis Grade 3 (yellow arrows). Fig. 1e: Coronal view – venous phase of CT abdomen with oral and intravenous contrast, showing the IVC (yellow arrow) anterior to the renal pelvis (orange arrow). Fig. 1f: Axial view – arterial phase of CT abdomen with oral and intravenous contrast illustrating left retroaortic renal vein (yellow arrow) coursing behind the aorta (blue arrow)..

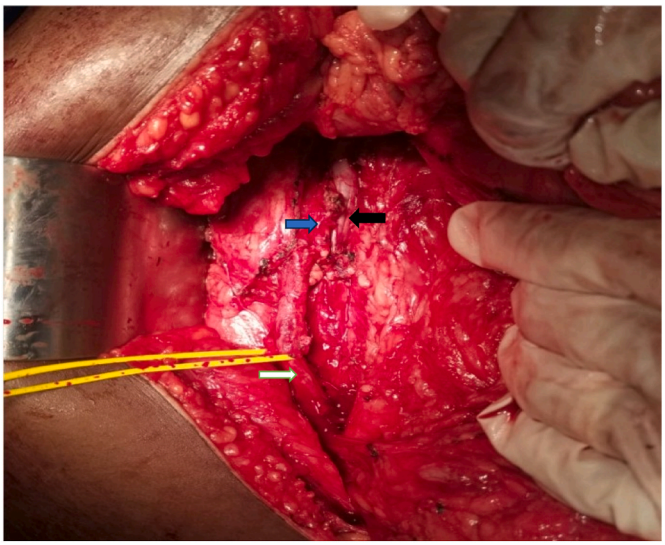


Fig. 3. Intraoperative Images after transection and transposition of right ureter and Pyeloplasty. Depicting the right ureter (white arrow) anterior to the inferior vena cava (Black arrow) and the site of pyeloplasty (blue arrow)..

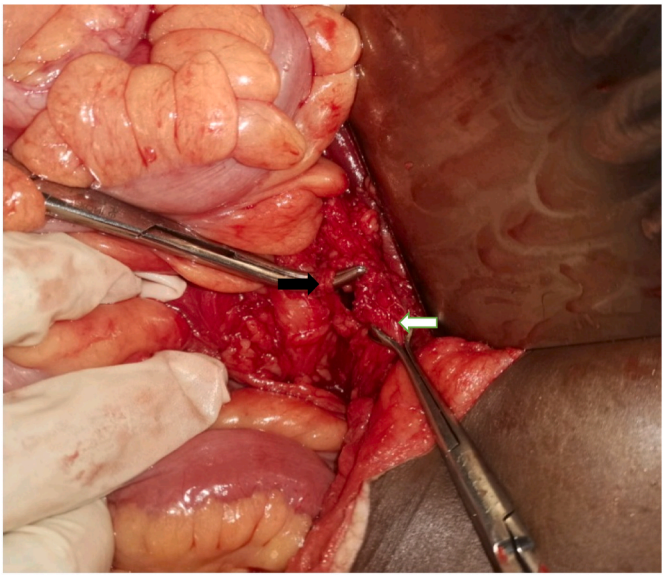


Fig. 4. Intraoperative Images Before Left Pyeloplasty. Depicting the left ureter (white arrow) and a crossing aberrant vessel (black arrow)..

3. Discussion

UPJ obstruction is a common issue in children and young adults, but it can occur at any age and is often associated with complications due to late presentation. These complications include pyelonephritis, hydronephrosis, stone formation and varying degrees of renal function impairment. Bilateral UPJ obstruction is rare and carries a high morbidity rate.[1,2] Our patient initially presented in childhood with a congenital urological condition of the kidney and was lost to follow-up, re-presenting in young adulthood with loin pain, hydronephrosis, and mild renal impairment. The re-presentation in young adulthood led to the revelation of significant anatomical abnormalities, including a right retrocaval ureter and an obstructed left kidney due to accessory vessels. These findings are rare and pose unique challenges in both diagnosis and management.

A retrocaval ureter, also known as circumcave or postcave ureter, is a rare developmental anomaly of the IVC where it is abnormally positioned anterior to the ureter. The IVC is formed from three bilateral venous systems: the posterior cardinal veins, the subcardinal veins, and the supracardinal veins. The persistence of the posterior cardinal vein leads to the formation of a retrocaval ureter.[3,4] While some cases are discovered incidentally, most retrocaval ureters present with symptoms related to upper urinary tract obstruction and its complications. In 20 % of cases, upper urinary tract infections reveal the condition, particularly in children. In adults, right flank pain is a more common presenting symptom.[3,4] This anatomical variant primarily affects the right ureter. However, the left ureter can also be involved if there are co-existing anomalies like situs inversus or a duplicated IVC.[3]

There are two main types of retrocaval ureter, with a reported prevalence of approximately 1.3 in 1000 or 1 in 800, and a higher occurrence in males. **Type 1 (low loop)** is more common, occurring in 94 % of cases, and results in a dilated proximal ureter that spirals medially and upwards, creating an S-shaped or 'fish-hook' appearance. A change in the ureter's caliber is typically observed at the L3/L4 level. Symptoms usually do not appear until the 3rd or 4th decade of life. If left untreated, the obstruction can lead to complications such as pyelonephritis, stone formation, and hematuria. **Type 2 (high loop)** involves a near-horizontal course of the proximal ureter at the level of the renal pelvis, intersecting the IVC and forming a 'sickle-shaped curve.' This type is usually non-obstructive.[3,4] In our exceptional case, the patient had a type 2 postcave ureter with unusually high-grade hydronephrosis on the right, a rare cause of UPJ obstruction. Additionally, crossing accessory vessels on the left also caused UPJ obstruction. Accessory crossing vessels, typically an accessory lower-pole renal artery, are the most common extrinsic cause of UPJ obstruction, leading to external compression of the UPJ. While crossing vessels are frequently found in the unobstructed population, there is ongoing debate about how often they are the true cause of obstruction in patients with UPJ obstruction.[1,5] Our case is unique as it combines vascular anomalies of different origins and natures, namely a retrocaval ureter, accessory crossing vessels, and a retroaortic left renal vein. The retroaortic left renal vein is formed due to the persistence of the dorsal limb, rather than the ventral limb, in the circum-aortic collar of veins.[5] The use of advanced imaging techniques, such as CT Urogram and Mag-3 renogram, was crucial in delineating the anatomical and functional status of the kidneys.

The treatment of a retrocaval ureter in the setting of obstruction primarily depends on the degree of obstruction. Pyeloplasty remains the standard of care for UPJ obstruction, with success rates of approximately 90 %, and can be performed as an open, laparoscopic, or robot-assisted procedure. Endoscopic alternatives, such as balloon dilatation and endopyelotomy, offer less invasive options for select patients, though their success rates are lower than those of pyeloplasty. In cases where a retrocaval ureter is discovered incidentally without significant obstruction, therapeutic abstention and radioclinical follow-up are justified. Nephrectomy is necessary in rare cases where the kidney is severely damaged. For symptomatic cases, conservative surgery is preferred. Type 1 involves an intrinsic anomaly in the retrocaval segment of the ureter, while Type 2 involves extrinsic compression of a normal ureter in its retrocaval portion. The typical approach for Type 1 is resection-anastomosis of the retrocaval segment, and for Type 2, it is ureteral uncrossing with direct plasty and end-to-end uretero-ureteral anastomosis.[1–4] The management strategy in our case initially involved JJ stenting and ultimately, bilateral pyeloplasty at the same sitting through a midline incision. The decision to perform a dismembered pyeloplasty (Anderson-Hynes) after ureteral uncrossing on the left, and a simple pyeloplasty (Heineke) after repositioning the ureter anterior to the IVC on the right to prevent future compression at the PUJ, was based on the specific anatomical challenges presented by each kidney. The successful outcome, with normal renal function and septic markers post-surgery, highlights the effectiveness of tailored surgical interventions in managing complex urological conditions.

4. Conclusion

This case report highlights the complexity of managing bilateral ureteropelvic obstruction due to distinct vascular anomalies. The patient's journey from childhood diagnosis to adult re-presentation emphasizes the importance of continuous follow-up in congenital urological conditions. The combination of a type 2 circumcaval ureter on the right and high-grade hydronephrosis, along with crossing vessels on the left, presented unique diagnostic and therapeutic challenges. The successful management of this case highlights the effectiveness of a tailored, multidisciplinary approach. The patient's positive outcome, with normal renal function and no symptoms on follow-up, underscores the potential for successful treatment in complex cases. This report contributes to the limited literature on rare conditions and emphasizes the need for ongoing research and collaboration in urology to improve patient outcomes.

CRediT authorship contribution statement

Mohammed Salah E. Khalifa Salem: Writing – original draft, Conceptualization. **Abdul Alherek:** Writing – original draft. **Daniel DaSilva:** Writing – original draft. **Jacob Varghese:** Writing – original draft. **Alain Mwamba Mukendi:** Writing – review & editing, Writing – original draft, Resources, Conceptualization.

Patient consent

Consent to publish the case report was obtained from the patient.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Funding

No funding or grant support

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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