



RETROCAVAL URETER IN AN ELEVEN-YEAR-OLD MALE CHILD: A RAREST CASE REPORT

Nizi Alexander¹ | Rahil S K² | Sam Jeeva Kumar* | Prof (Dr.) Shaiju S Dharan⁴ |

1. Pharm D Intern, Ezhuthachan College of Pharmaceutical Sciences, Marayamuttom, Neyyattinkara, Thiruvananthapuram.
2. Pharm D Intern, Ezhuthachan College of Pharmaceutical Sciences, Marayamuttom, Neyyattinkara, Thiruvananthapuram.
3. Associate Professor, Department of Pharmacy Practice, Ezhuthachan College of Pharmaceutical Sciences.
4. Principal/HOD- Pharmacy Practice, Ezhuthachan College of Pharmaceutical Sciences, Marayamuttom, Neyyattinkara, Thiruvananthapuram.

ABSTRACT

Retrocaval ureter is a rare congenital abnormality in which the ureter abnormally courses posterior to the inferior vena cava (IVC), causing obstruction of the upper urinary tract. While it typically presents in the third or fourth decade of life, pediatric cases are extremely rare. In here we report a rare case of an 11-year-old male who presented with intermittent right-sided abdominal pain, vomiting, and one episode of loose stool. Ultrasound revealed right-sided hydronephrosis, and further imaging with CT urogram and intravenous pyelography (IVP) showed mild to moderate hydroureteronephrosis with a characteristic fishhook deformity of the mid-ureter, indicative of Type I retrocaval ureter. Surgical correction was performed via retroperitoneal exploration and anterior transposition of the ureter with uretero-ureterostomy. The postoperative course was uneventful, with improved urinary drainage and complete symptom resolution. This case underscores the importance of considering retrocaval ureter in the differential diagnosis of unexplained right hydronephrosis in children. Early diagnosis through imaging and prompt surgical intervention are essential for optimal outcomes.

KEY WORDS: Retrocaval ureter, Pediatric urology, Hydronephrosis, Fishhook deformity, Ureteral obstruction, Type I retrocaval ureter.

INTRODUCTION:

Retrocaval ureter, also referred to as circumcaval ureter, is a rare congenital malformation in which the ureter passes posterior to the inferior vena cava (IVC), leading to compression, obstruction, and ultimately causing hydronephrosis. Its congenital in origin, but often presents later in life due to the progressive nature of ureteral obstruction. The anomaly results from an abnormal development of the IVC during embryogenesis. Normally, the infrarenal segment of the IVC is formed by the dorsal supracardinal vein, which allows the ureter to descend anterior to it. In retrocaval ureter, however, the ventral subcardinal vein persists abnormally and becomes the IVC. This altered course traps the ureter behind the vein, predisposing it to compression and chronic obstruction^[1]. Its incidence is very low – on the order of 0.06–0.17% of the population (roughly 1 in 1,000)^[3]. Classically, the ureter is symmetrically dilated proximal to the IVC level, producing an “Sshaped” or inverted-J (“fishhook”) deformity on intravenous urography (IVU). Clinically, retrocaval ureter predominates in males (~3:1) and almost always involves the right ureter^[7]. It usually presents in adulthood (third–fourth decades) with right flank pain, urinary tract infection or hematuria, and is very unusual in children^[8].

Retrocaval ureters are classified by Bateson and Atkinson into two radiographic types,

Type I (low-loop): The dilated renal pelvis and upper ureter descend nearly horizontally before sharply angling upward behind the IVC. On IVU this produces the classic “fishhook” or inverted J deformity, often with marked hydronephrosis. This is the common form.

Type II (high-loop): The proximal ureter descends more vertically and passes behind the IVC at a higher level, forming a smooth “sickle”-shaped curve with generally less obstruction^[9]. Awareness of this anomaly is important in any patient (even a child) with unexplained right hydronephrosis, Since timely surgical correction can relieve obstruction and preserve renal function^[10].

CLINICAL PRESENTATION:

An 11-year-old boy presented with a 1-month history of intermittent, crampy right lower abdominal pain associated with nausea and vomiting; he also had one episode of loose stool. There was no fever, dysuria or visible hematuria. Physical examination was unremarkable. Laboratory tests (complete blood count, serum creatinine, urinalysis) were normal.



Fig 1: Abrupt narrowing of the right mid ureter with fish hook deformity and medialization of the distal ureter.

Our patient underwent right retroperitoneal exploration. Intraoperatively, the upper right ureter was found coursing posterior to the IVC with a constrictive loop. The redundant retrocaval segment was excised, and an end-to-end uretero-ureterostomy was performed, repositioning the ureter anterior to the IVC. A double-J ureteral stent was placed. The postoperative course was uneventful. The stent was removed after 4 weeks. Follow-up imaging (ultrasound/IVU) at 6 months showed resolution of hydronephrosis and normal urinary drainage; the patient's pain and symptoms had completely resolved.

DISCUSSION:

Retrocaval ureter is an uncommon cause of hydronephrosis, particularly in children. Most reported cases occur in young adults, so our 11-year-old patient represents a rare pediatric presentation. The clinical features – right-sided abdominal/flank pain with varying degrees of hydronephrosis – were typical. Notably, there was no hematuria or infection, similar to other pediatric series where flank pain is often the sole symptom^[7]. The male sex and right-sided location also fit the usual pattern (males are affected about three times more often, and right-side cases predominate). In our case, no other congenital anomalies were found; this concurs with series such as Sun et al., where none of the eight pediatric cases had associated malformations^[12]. (In general, about 20% of retrocaval ureter cases have concomitant cardiovascular or genitourinary anomalies^[13], so careful screening is recommended.

The imaging in this case was quintessential. Ultrasonography and CT raised suspicion by showing right hydronephrosis. The IVP then provided the pathognomonic appearance: a dilated proximal ureter that descends then turns medially behind the IVC, creating an “S” or inverted-J curve^[6]. This “fishhook” deformity on IVP has been described in classic reports and is virtually diagnostic. Figure 1 illustrates this finding. Spiral CT scan is often cited as a modern diagnostic choice, since it can outline both the ureter and IVC without invasive injection. Magnetic resonance urography is another non-radiative option to confirm the anatomy. In our patient the IVP alone sufficed to make the diagnosis, consistent with many reports that still rely on urography and retrograde pyelography.

Our case was Type I retrocaval ureter (low-loop, fishhook type). Interestingly, Sun et al. found all eight of their pediatric patients to have Type II (high-loop) morphology ^[2], suggesting variability among series. In contrast, Acharya et al. and Bhutani et al. have also reported Type I cases in children. Thus our finding underscores that both types can occur in childhood ^[8].

Treatment of symptomatic retrocaval ureter is surgical, aimed at relieving obstruction. The standard correction is excision of the retrocaval segment with ureteroureterostomy (or pyeloureterostomy) after transposing the ureter anterior to the IVC ^[7]. In our patient, a retroperitoneal open repair was performed with excellent results. Minimally invasive (laparoscopic or robotic) approaches have been increasingly reported as safe and effective, even in children ^[9], but open repair remains an option. In series, postoperative outcomes are uniformly good: renal drainage normalizes and symptoms resolve. We observed complete pain relief and radiographic resolution of hydronephrosis, as expected ^[10].

In summary, this case illustrates the key points of retrocaval ureter: a rare congenital IVC anomaly that causes right-sided hydronephrosis in the young, diagnosed by the classic “fishhook” ureter on imaging. Comparison with the literature shows our case is consistent with reported pediatric cases (right male, flank pain) but highlights a Type I variant. Clinicians should include retrocaval ureter in the differential for unexplained right hydronephrosis, even in children. Prompt surgical repair is important to prevent renal damage and generally leads to excellent outcomes ^[9].

CONCLUSION:

Retrocaval ureter is a rare but important cause of obstructive hydronephrosis. In pediatric patients presenting with recurrent flank pain and hydronephrosis, careful imaging can reveal the characteristic ureteral course behind the IVC. Our case of an 11-year-old boy reinforces that early recognition and surgical transposition of the ureter result in symptom relief and preservation of renal function. Awareness of this anomaly and correlation with IVU/CT findings are essential, as emphasized by previous reports ^[2,8]. This case adds to the small number of pediatric series and underscores that retrocaval ureter – though congenital – may present in childhood and should be treated surgically once identified.

REFERENCES:

1. Campbell-Walsh Urology, 12th Edition; Ashcraft's Pediatric Surgery, 6th Edition.
2. Sun JS, Zhang G, Lin T. Retrocaval ureter in children: a report of eight cases. *West Indian Med J.* 2016;64(4).
3. Kajal P, Rattan K, Sangwan V, Bhutani N. Retrocaval ureter presenting at 6 years of age in a girl child – an extreme rarity. *Asian J Urol.* 2016;3(2).
4. Acharya SK, Jindal B, Yadav DK, Singhal S, Bagga D. Retrocaval ureter: a rare cause of hydronephrosis in children. *J Pediatr Surg.* 2009;44(5).
5. Atawurah H, Maison POM, Owusu-Ansah M, Asante-Asamani A. Retrocaval ureter: report of two cases. *Case Rep Urol.* 2019;2019:2815748.

6. Athanasopoulos A, Papachristou I, Pneumatikos I, et al. Retrocaval ureter and associated abnormalities. *Int Urol Nephrol*. 2002;34(1).
7. Bateson EM, Atkinson D. Circumcaval ureter: a new classification. **Clin Radiol**. 1969;20(2):173–7.
8. Campbell MF, Wein AJ, Kavoussi LR, Partin AW, Peters CA. **Campbell-Walsh Urology**. 12th ed. Philadelphia: Elsevier; 2021.
9. Smith DR, Tanagho EA, McAninch JW. **Smith’s General Urology**. 18th ed. New York: McGraw-Hill; 2013. Chapter 11, Congenital anomalies of the ureter.
10. Wein AJ, Kavoussi LR, Partin AW, Peters CA, editors. **Pediatric Urology**. 2nd ed. Philadelphia: Elsevier; 2016. Chapter on ureteral anomalies; p. 164–72.
11. Holcomb GW, Murphy JP, Ostlie DJ. **Ashcraft’s Pediatric Surgery**. 6th ed. Philadelphia: Elsevier; 2014. Chapter 72, Congenital anomalies of the ureter; p. 935–40.
12. Raj GV, Tracey J, Webster GD. Retrocaval ureter: clinical features and management. **Urology**. 2004;63(2).
13. Singh S, Misra D, Jaiswal S, Sankhwar SN. Laparoscopic management of retrocaval ureter in children: a case report and review of literature. **J Indian Assoc Pediatr Surg**. 2010;15(3).
14. Karan SC, Goel A, Singh V, Bera MK. Retrocaval ureter: our experience and a review of the literature. **Urol Ann**. 2016;8(4).

