

Research Article

Bilateral Congenital Mid-Ureteral Valves in a Child Managed with Robotic Reconstruction

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Abstract

Background & presentation: Congenital ureteral valves are rare intrinsic causes of ureteral obstruction. Bilateral involvement is exceedingly uncommon. A 7-year-old boy presented with a two-year history of intermittent left flank pain and six months of recurrent right scrotal pain and swelling. CT urography demonstrated bilateral hydroureteronephrosis with multiple areas of ureteral narrowing. Diuretic renography showed preserved differential renal function; voiding cystourethrography excluded vesicoureteral reflux.

Keywords: Congenital ureteral valve; Bilateral ureteral valve; Pediatric hydroureteronephrosis; Robotic surgery; Boari flap; Ureteral obstruction.

Findings & outcome

Bilateral retrograde ureterography identified obstructive lesions in the lower mid-ureters bilaterally, consistent with congenital ureteral valves. The patient underwent robotic-assisted reconstruction: left side required Heineke–Mikulicz ureteroplasty followed by excision and ureteroneocystostomy; right side underwent excision with robotic Boari flap ureteral reimplantation. Histopathology confirmed a Type II congenital ureteral valve. Postoperative follow-up demonstrated complete resolution of hydronephrosis bilaterally.

First reported case of robotic Boari flap ureteral reimplantation for congenital ureteral valve disease.

Congenital ureteral valves are among the rarest intrinsic causes of ureteral obstruction in children. Bilateral involvement is exceedingly uncommon, with fewer than seventy cases of ureteral valve disease reported in the literature and only isolated instances of bilateral disease. We present a 7-year-old boy with bilateral congenital mid-ureteral valves successfully managed with robotic-assisted reconstruction, including — to our knowledge — the first reported robotic Boari flap ureteral reimplantation for this condition.

Introduction

Congenital ureteral valves represent an uncommon developmental anomaly causing intrinsic ureteral obstruction and progressive upper urinary tract dilatation. Since the initial description in the nineteenth century, fewer than seventy cases have been reported in the literature. Bilateral ureteral valves are exceptionally rare, with only isolated cases documented worldwide. The rarity of bilateral disease, combined with the nonspecific nature of its clinical and radiological presentation, creates a substantial diagnostic challenge for the treating clinician.

The condition frequently mimics other causes of pediatric hydroureteronephrosis, including congenital ureteral strictures, primary obstructive megaureter, ureteropelvic junction obstruction, and vesicoureteral reflux. As a result, definitive preoperative diagnosis is infrequently achieved, and the majority of cases are confirmed intraoperatively and subsequently validated by histopathological examination. This diagnostic ambiguity has important implications for surgical planning, particularly when bilateral disease complicates an already uncommon clinical entity.

We report a rare case of bilateral congenital mid-ureteral valves in a child successfully managed using robotic-assisted reconstruction. The case highlights the indispensable role of retrograde ureterography

in achieving accurate preoperative diagnosis and demonstrates, for the first time in the literature, the technical feasibility and clinical efficacy of robotic Boari flap ureteral reimplantation in the context of congenital ureteral valve disease. This report contributes meaningfully to the limited body of literature on this subject and offers a model for the management of complex bilateral ureteral anomalies in the pediatric population.

• <70 Cases reported

Total congenital ureteral valve cases in world literature since 19th century

• 3% Bilateral Involvement

Proportion of ureteral valve cases with bilateral disease

• 60% Left-Sided Predominance

Left lateralization in reported ureteral valve cases

• 1st Robotic Boari Flap

First reported robotic Boari flap for congenital ureteral valve disease

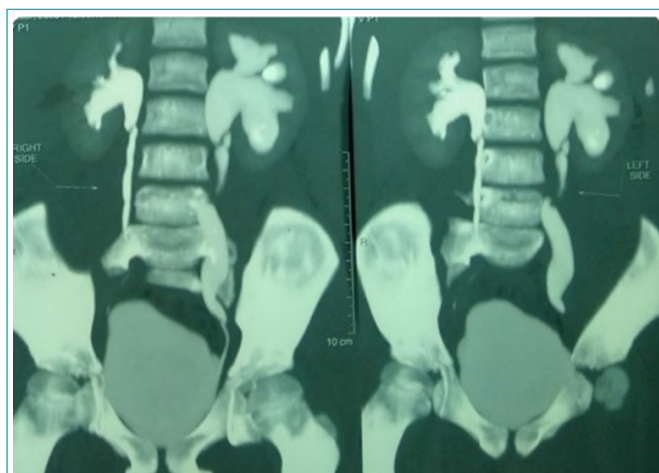


Figure 1: CT urogram (Coronal reconstruction). Reconstructed coronal CT urogram images demonstrating right mild hydronephrosis and left moderate hydronephrosis. The left mid-ureter lower portion shows a distinct narrowing with proximal ureteral dilatation, tortuosity, and a normal-caliber distal segment — a pattern highly suggestive of an intrinsic obstructive lesion.

Case presentation: Clinical history & initial workup

A 7-year-old boy from Bangladesh presented with intermittent left flank pain for two years and recurrent episodes of right testicular pain and swelling for six months. He had previously been treated empirically for epididymo-orchitis and was noted to have left-sided hydronephrosis on ultrasonography at an outside institution; however, the underlying etiology remained unidentified despite prior evaluation. No prior surgical history was documented.

On presentation to our institution, contrast-enhanced computed tomography urography demonstrated moderate left hydronephrosis and mild right hydronephrosis. The left ureter exhibited multiple areas of circumferential and eccentric narrowing involving the upper, mid, and distal ureter, raising clinical suspicion for congenital ureteral webs or valves. The right ureter demonstrated mild dilatation without a clearly defined obstructive lesion on cross-sectional imaging alone. These findings underscored the limitations of CT urography in characterizing the precise morphology of intrinsic ureteral obstruction.

Diuretic DTPA renography revealed split renal function of 39% (left) and 61% (right). Voiding cystourethrography excluded vesicoureteral reflux as a contributing etiology.

Diagnostic imaging: Renography & retrograde ureterography

Diuretic DTPA renography was performed to assess differential renal function and evaluate for obstructive drainage patterns. The study revealed preserved but asymmetric renal function, with the left kidney contributing 39% and the right kidney 61% of total split renal function. This finding confirmed functionally significant obstruction on the left side while indicating adequate preservation of overall renal reserve. Voiding cystourethrography did not demonstrate vesicoureteral reflux in either ureter, effectively excluding a reflux-related etiology.

Given the inconclusive findings on cross-sectional and nuclear medicine imaging, the decision was made to proceed with cystoscopy and bilateral retrograde ureterography. This modality provided critical information regarding the number, location, length, and morphological characteristics of the obstructive lesions — information that CT and MR urography had failed to adequately delineate. Retrograde studies revealed bilateral lower mid-ureteral obstructive lesions with characteristic proximal ureteral dilatation. The left ureter additionally demonstrated significant tortuosity involving the proximal and mid-ureteral segments.

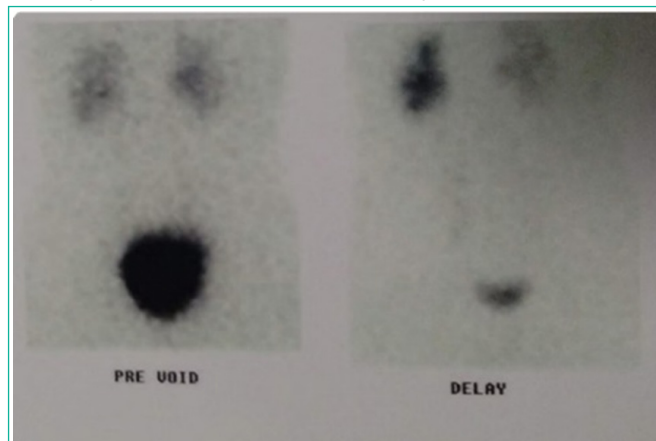


Figure 2: Diuretic DTPA renogram. Pre-void and delay images demonstrating left moderate hydronephrosis with mildly impaired function (39%) and mild right hydronephrosis with preserved function (61%).



Figure 3: Left retrograde ureterogram. Left retrograde ureterogram demonstrating a narrow distal ureter, an obvious constriction at the lower portion of the mid-ureter, and proximal ureteral dilatation with tortuosity -consistent with a congenital ureteral valve at this level.

Retrograde ureterography: Right side & surgical planning

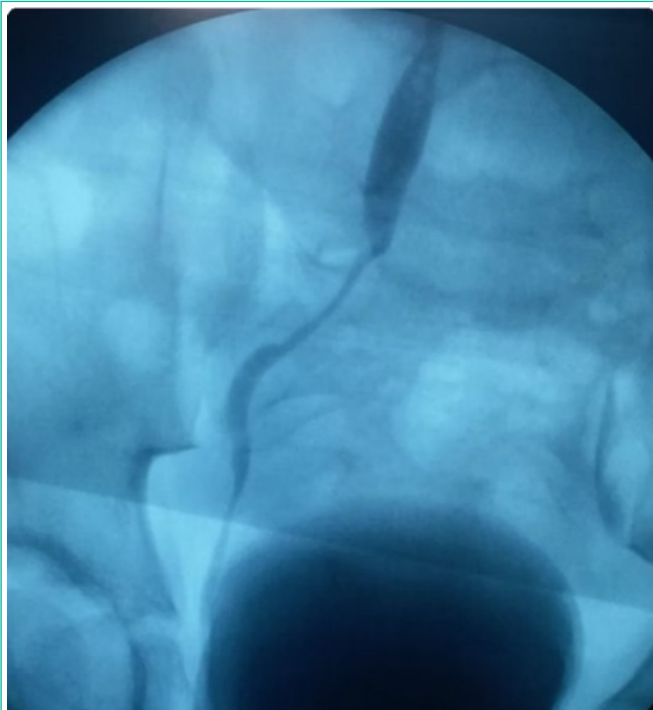


Figure 4: Right retrograde ureterogram. Right retrograde ureterogram showing a narrow distal ureter with an obvious constriction at the lower mid-ureteral level and proximal ureteral dilatation, confirming bilateral obstructive disease and directing the surgical approach for the right side.

The combination of CT urography, diuretic renography, voiding cystourethrography, and bilateral retrograde ureterography established a definitive preoperative diagnosis of bilateral congenital mid-ureteral valves with left proximal ureteral tortuosity. Notably, the initial provisional diagnosis based on cross-sectional imaging alone was left multiple ureteral valves; retrograde ureterography fundamentally revised this to bilateral lower mid-ureteral valves, directly influencing operative planning. Based on the comprehensive preoperative workup, robotic-assisted surgical reconstruction was planned. The surgical strategy was individualized to each side, accounting for the differing degree of ureteral involvement, tortuosity, and proximity to the bladder. The left side presented the greater reconstructive complexity owing to proximal tortuosity in addition to the distal obstructive lesion. Retrograde ureterography changed the preoperative diagnosis and altered the surgical plan — underscoring its role as an indispensable adjunct in the evaluation of pediatric hydronephrosis of unclear etiology.

1. Left proximal narrowing

Heineke-Mikulicz ureteroplasty performed to address proximal ureteral narrowing identified on retrograde ureterography and intraoperative inspection.

2. Left distal stenosis

Following ureterolysis, the lower mid-ureter remained severely stenotic and unsuitable for in-situ preservation. Segmental excision was performed.

Robotic surgical reconstruction: Operative technique



Figure 5: Intraoperative: Left lower mid-ureteral narrowing. Intraoperative robotic view demonstrating the focal stenosis at the left lower mid-ureteral segment prior to reconstruction. The degree of narrowing and surrounding fibrotic tissue precluded in-situ repair, necessitating segmental excision.

3. Left reconstruction

Ureteroneocystostomy completed over a double-J ureteral stent, re-establishing unobstructed urinary drainage from the left renal unit to the bladder.

4. Right reconstruction

Right stenotic segment excised; robotic Boari flap ureteral reimplantation performed over a double-J stent 4 the first reported use of this technique for ureteral valve disease. Gross examination of the excised specimen demonstrated a distinct intraluminal valve-like structure with an eccentric opening. The histopathological findings are consistent with the Rabinowitz classification of Type II ureteral valves and satisfy the diagnostic criteria as modified by Gosalbez et al., who consider the presence of smooth muscle throughout the entire fold unnecessary for definitive diagnosis. Double-J ureteral stents were removed three weeks postoperatively. At one-month follow-up, the patient was asymptomatic and ultrasonography demonstrated significant bilateral resolution of hydronephrosis.

Robotic boari flap: Intraoperative images & histopathology



Figure 6: Boari flap creation (Right Side). Intraoperative robotic view demonstrating the creation of a Boari flap from the bladder wall on the right side. The flap provides adequate length to bridge the ureteral defect following excision of the stenotic segment, enabling tension-free anastomosis with the proximal ureter.

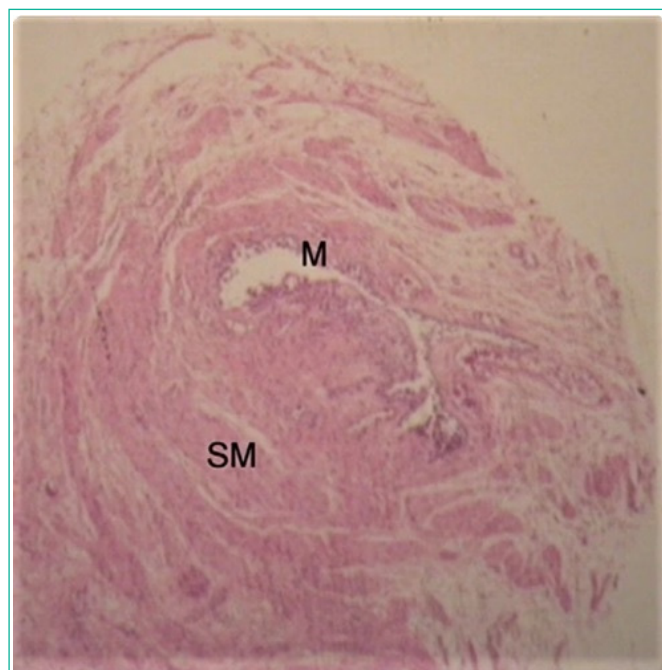


Figure 7: Histopathological examination (H&E, ×100). Hematoxylin and eosin stain at 100× magnification of the excised ureteral specimen. The section demonstrates urothelial mucosa (M) overlying fibrous stroma with smooth muscle (SM) hypertrophy concentrated at the base of the intraluminal fold — fulfilling the modified Wall and Wachter criteria for a Type II congenital ureteral valve, in which smooth muscle is present at the base of the valve leaflet only, rather than throughout the fold.

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Discussion: Pathology, Diagnosis & Surgical Management

Congenital ureteral valves are defined by the Wall and Wachter criteria (1952): (1) histological demonstration of transverse ureteral mucosal folds containing smooth muscle fiber bundles, (2) evidence of proximal obstructive uropathy with a normal distal ureter, and (3) exclusion of alternative mechanical or functional causes. This was subsequently modified by Gosalbez et al., who considered the presence of smooth muscle throughout the entire fold to be unnecessary, requiring only smooth muscle at the base. Rabinowitz classified the entity into Type I (smooth muscle within the leaflet) and Type II (smooth muscle at the base only); the latter was confirmed in the present case.

The embryological basis of ureteral valve formation remains incompletely understood. Proposed mechanisms include incomplete recanalization of the ureteric lumen during fetal development, localized developmental arrest or ischemia caused by extrinsic vascular compression, intrauterine ureteritis, and persistence of

physiological fetal ureteral folds beyond the expected period of regression. Approximately 40% of reported cases have associated genitourinary anomalies, including complete or incomplete renal duplication, vesicoureteral reflux, horseshoe kidney, renal agenesis, and ureteropelvic junction obstruction. Side localization shows a left predominance (60%), with right-sided disease in 37% and bilateral involvement in approximately 3%. By anatomical location, 43% of valves are in the upper ureter, 32% in the lower ureter, 16% in the mid-ureter, 7% are multiple, and 2% occur at the renal pelvis.

Surgical options

- **Heineke-mikulicz ureteroplasty**

For focal, short-segment narrowing amenable to longitudinal incision and transverse closure without resection.

- **Boari flap reimplantation**

For cases requiring bridging of a long ureteral defect after excision; bladder flap provides tension-free reconstruction.

- **Segmental excision & ureteroneocystostomy**

When the stenotic segment is unsuitable for in-situ repair and adequate ureteral length permits direct reimplantation.

- **Nephroureterectomy**

Reserved for poorly or non-functioning renal units where reconstruction is neither feasible nor indicated.

- **Emerging endoscopic options**

Endoscopic incision and endoscopic laser ablation have been described in carefully selected patients with favorable anatomy and preserved renal function. These techniques represent an evolving alternative to open or robotic reconstruction but require further validation in the pediatric population.

Why robotic surgery?

The enhanced three-dimensional visualization, articulated wristed instrumentation, tremor filtration, and superior intracorporeal suturing precision offered by the robotic platform make it ideally suited for complex pediatric ureteral reconstruction. These attributes are especially advantageous when bilateral disease demands multi-quadrant dissection and individualized reconstructive strategies performed in a single operative setting.

Conclusion & Learning points

This case represents a rare and instructive example of bilateral congenital mid-ureteral valves in a pediatric patient successfully managed with robotic-assisted reconstruction. The diagnostic pathway illustrates the limitations of cross-sectional imaging in characterizing intrinsic ureteral obstructive lesions and affirms the clinical value of retrograde ureterography as an indispensable adjunct in selected children with unexplained hydronephrosis. Histopathological examination confirmed a Type II congenital ureteral valve and remains the gold standard for definitive diagnosis. The bilateral robotic reconstruction — including the first reported robotic Boari flap ureteral reimplantation for this condition — demonstrates the feasibility, safety, and efficacy of minimally invasive approaches for complex ureteral valve disease. This report expands the limited global literature on bilateral congenital ureteral valves and provides a reproducible framework for diagnosis and management.

- **Rare but important**

Congenital ureteral valves — especially bilateral — must remain in the differential diagnosis of unexplained pediatric hydroureteronephrosis.

- **Retrograde ureterography**

Retrograde studies significantly improve preoperative diagnostic accuracy when CT and MR urography are inconclusive.

- **Histopathology is gold standard**

Histopathological confirmation with identification of smooth muscle at the valve base remains essential for definitive diagnosis.

- **Robotic reconstruction**

Robotic surgery offers superior visualization and precision for complex bilateral ureteral reconstruction in the pediatric setting.

- **First robotic boari flap**

This case represents the first reported robotic Boari flap ureteral reimplantation for congenital ureteral valve disease worldwide.

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