

Rare Presentation of Pelvi-ureteric Junction Obstruction: A Series of Three Cases

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ABSTRACT

Pelvi-Ureteric Junction Obstruction (PUJO) is one of the most frequent causes of hydronephrosis in both children and adults. It may present with flank pain, vomiting, or progressive renal dysfunction, and if left untreated, may cause irreversible renal damage. Although laparoscopic and robotic techniques are gaining popularity, open pyeloplasty continues to be the gold standard, particularly in anatomically complex cases. A series of three cases of adult PUJO presented here, which were managed with open surgical reconstruction, each with distinct intraoperative findings and outcomes. A 30-year-old male presented with right flank pain and vomiting for two months. Computed Tomography (CT) urography revealed moderate hydronephrosis with a grossly dilated inferomedial calyx, while Diethylenetriaminepenta Acetic Acid (DTPA) scan showed reduced right renal function {24.2%, Glomerular Filtration Rate (GFR) 21.7 mL/min}. Intraoperatively, the dilated lower calyx was compressing a high-insertion ureter. Reduction calyccoplasty with infundibular dilatation and Double J (DJ) stenting was performed. Follow-up DTPA showed a 10% increase in split function and +8 mL/min GFR improvement. A 20-year-old female with dull aching left flank pain for one year had a CT urography suggestive of severe hydronephrosis with cortical thinning. DTPA scan showed left kidney function at 18.3% (GFR 16.1 mL/min). Retrograde pyelography confirmed a grossly dilated pelvis. Anderson-Hynes dismembered pyeloplasty was performed, preserving a crossing systemic vein. Follow-up DTPA demonstrated a 10% rise in split function and +10.3 mL/min GFR gain. A 55-year-old female with intermittent right flank pain and vomiting for one year had Intravenous Pyelography (IVP) and Right Retrograde Pyelogram (RGP) suggestive of PUJO due to Pelvi-Ureteric Junction (PUJ) kink with poor drainage. She underwent standard dismembered pyeloplasty. Postoperative recovery was uneventful, and she remains asymptomatic with preserved renal function. In this case series, all three cases were rare and unique presentations of PUJO.

Keywords: Calyccoplasty, Crossing vessel, Pyeloplasty, Rare cases, Ureteral kink, Ureteropelvic junction obstruction

INTRODUCTION

The PUJO is one of the most common pathologies encountered in urology. PUJO is the most common cause of hydronephrosis in the paediatric age group, but it can also occur frequently in adults [1]. PUJO is mainly of two types: congenital and acquired. Congenital PUJO is the most common, and it can be due to ureteral hypoplasia, high insertion of the ureter, entrapment of the ureter by crossing a vessel, and a malrotated kidney. Acquired causes can be extrinsic, such as retroperitoneal fibrosis, retroperitoneal lymph node, and retroperitoneal mass, and intrinsic, including ureteric stone, ureteric infection, and iatrogenic post-endoscopic procedure [2].

Anderson-Hynes dismembered pyeloplasty is the most commonly performed surgery for the treatment of PUJO using the open method, laparoscopically or endoscopically [3]. This case series presents three cases of unusual presentation of PUJO.

Case 1

A 30-year-old man presented with complaints of right flank pain for two months, dull aching in nature, with intermittent vomiting. Pain was not associated with fever, burning micturition, or haematuria. No other urinary complaints were present, and there were no comorbidities. He had a history of appendectomy five years ago. The patient had no significant family history.

On examination, the patient's vitals were within normal limits and no lumps or masses were felt on palpation. Ultrasonography (USG) of the Kidney, Ureter, and Bladder (KUB) was performed, which suggested a moderately dilated right renal pelvis with abrupt narrowing at the PUJ, suggestive of Right PUJO. For further evaluation, CT urography followed by a Diethylenetriaminepenta Acetic Acid (DTPA) scan was performed to quantify the renal

function. CT urography was suggestive of the right kidney size 98×60×58 mm with moderate hydronephrosis. No hydroureter or calculus was noted; the impression was right PUJO.

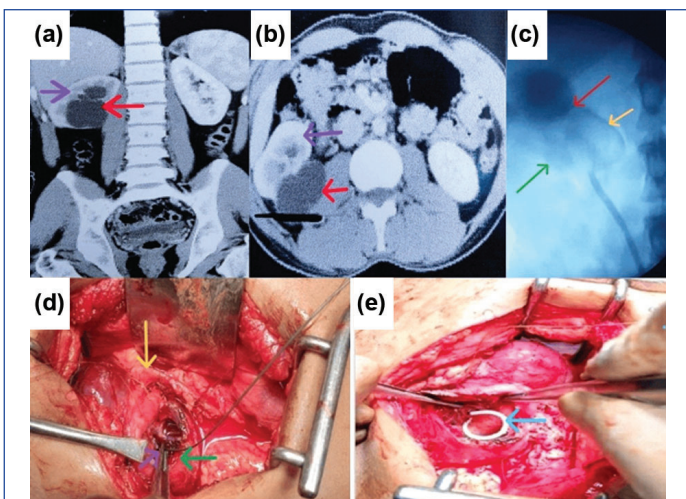
The inferomedial calyx showed disproportionately marked dilatation with paper-thinning of the cortex or calyceal stricture. Coronal CT image depicted a dilated lower inferomedial calyx and the right kidney [Table/Fig-1a]. Axial CT image demonstrated the dilated inferomedial calyx and the right kidney [Table/Fig-1b].

The DTPA scan was suggestive of a normally functioning, non-obstructed left kidney and suboptimally functioning, hydronephrotic, obstructed right kidney. Split function was 75.8% on the left side with a 68.1 mL/min Glomerular Filtration Rate (GFR). On the right side, the split function was 24.2%, and GFR was 21.7 mL/min.

The patient underwent open pyeloplasty. A right Retrograde Pyelogram (RGP) revealed a grossly dilated lower calyx (green arrow) and a high-inserting ureter coursing around the dilated calyx (yellow arrow), as shown in [Table/Fig-1c].

Intraoperatively, the dilated lower calyx compressing the coursing high-inserting ureter was seen. Dilated calyx was opened, infundibular dilatation was done, a 6/26fr Double J (DJ) stent was placed across the infundibulum, and the upper coil was kept in the lower calyx, as shown in [Table/Fig-1d,e]. The redundant calyx was excised, and the remaining part was sutured, amounting to reduction calyccoplasty.

Postoperatively, the patient was doing well and was discharged on postoperative day 5. On follow-up, a DTPA scan was done after three weeks of stent removal, which showed an increase in split function by 10% and GFR by 8 mL/min.



[Table/Fig-1]: a) Coronal CT image showing dilated lower inferomedial calyx (red arrow), right kidney (purple arrow); b) Axial CT images showing dilated inferomedial calyx (red arrow) and right kidney (purple arrow); c) RGP showing dilated lower inferomedial calyx (green arrow), stretched tortuous coursing ureter around the dilated calyx (yellow arrow), Pelvi-ureteric junction (red arrow); d) Opened lower calyx with guide wire in narrow infundibulum after dilatation, opened lower calyx (green arrow), high inserting tortuous ureter coursing around the calyx (yellow arrow), and dilated infundibulum with guidewire inside (purple arrow); e) Stent (sky blue arrow) placed specifically in lower opened calyx.

Case 2

A 20-year-old female presented with a complaint of left flank pain for one year. This was a dull aching pain not associated with nausea, fever, or vomiting. There was no history of haematuria, comorbidity, or surgery.

On examination, the vital signs were within normal limits, and systemic evaluation revealed no abnormalities. CT urography demonstrated a right kidney measuring 8.6×6.4×4.7 cm, normal in position and morphology. The left kidney, measured 13.2×4.9×4.5 cm, appeared bulky and showed severe hydronephrosis with paper-thin parenchyma, which was normal in position. The left ureter showed no significant dilatation or opacification on contrast, suggestive of PUJO. These findings are illustrated in [Table/Fig-2a], which shows an axial CT image with a dilated left renal pelvis and the left kidney and in [Table/Fig-2b], which shows a coronal CT demonstrating the dilated left renal pelvis and the left kidney.

A DTPA scan was performed to assess and quantify the renal function, which was suggestive of an enlarged sub-optimally functioning, hydronephrotic obstructed left kidney, and normal functioning non-obstructed right kidney. The split function of the right kidney was 81.7%, and the GFR was 71.9 mL/min. The split function of the left kidney was 18.3% and 16.1 mL/min GFR. A plan for left open pyeloplasty was made.

Left RGP was performed, which showed a grossly dilated left renal pelvis with ureteric jet, as shown in [Table/Fig-2c]. Intraoperatively aberrant systemic vein was seen crossing the PUJ and causing external mechanical compression, leading to obstruction as depicted in [Table/Fig-2d]. Anderson-Hynes dismembered pyeloplasty was performed, and the crossing systemic vein was spared [Table/Fig-2d-e].

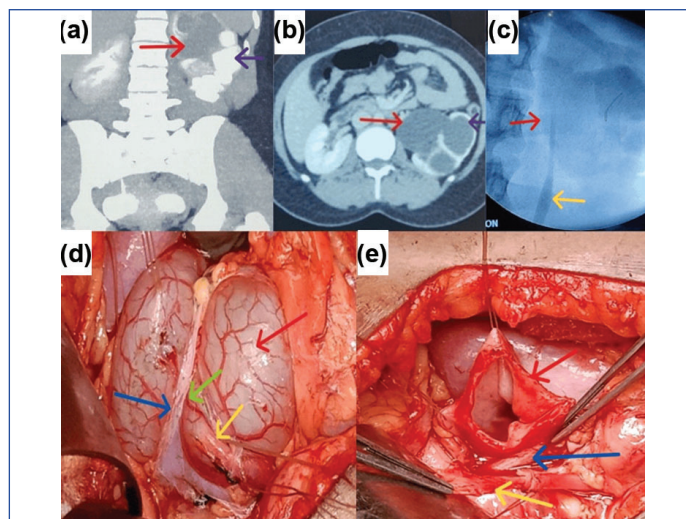
The patient was discharged on postoperative day 4. On follow-up, a DTPA scan was done after six weeks, showing a 10% increase in left-side split function and GFR by 10.3 mL/min.

Case 3

A 55-year-old female presented with complaints of right flank pain on and off over the last year. The pain was occasionally associated with episodes of vomiting and not associated with fever, dysuria, or haematuria.

On examination, the patient was found to be vitally stable and no abnormality was seen on systemic examination.

The USG abdomen pelvis was done, which was suggestive of a right grossly dilated kidney with an abrupt cut-off at the PUJ, suggestive

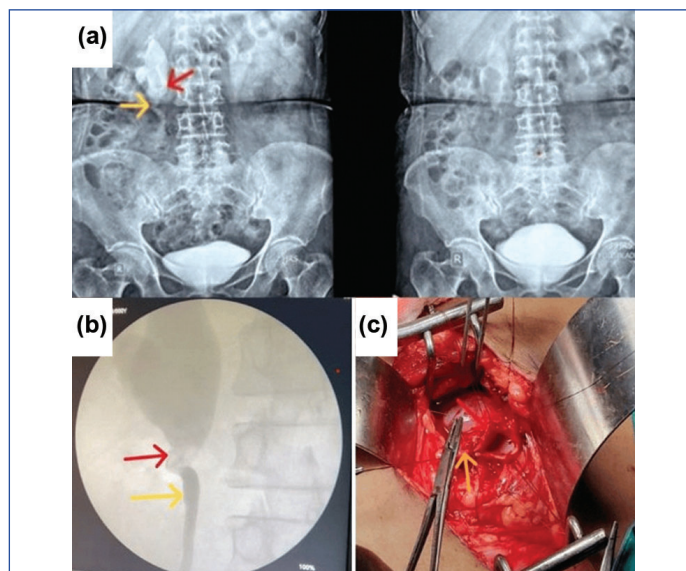


[Table/Fig-2]: a) Coronal CT film showing dilated renal pelvis (red arrow) and left kidney (purple arrow); b) Axial CT film showing dilated renal pelvis (red arrow) and left kidney (purple arrow); c) Retrograde pyelogram of left kidney showing markedly dilated renal pelvis with ureteric jet (red arrow) and ureter (yellow arrow); d) Intraoperative picture showing markedly dilated renal pelvis (red arrow), ureter (yellow arrow), Pelvi-ureteric junction (green arrow) and aberrant crossing systemic vein compressing the pelvi-ureteric junction (blue arrow); e) Anderson-Hynes dismembered pyeloplasty done, dismembered renal pelvis (red arrow), spatulated upper ureter (Yellow arrow) and spared crossing vein (Blue arrow).

of PUJO. Intravenous Pyelography (IVP) suggested a grossly dilated right pelvicalyceal system with a kink in the upper ureter at the pelvi-ureteric junction with minimal excretion of contrast on delayed films, as shown in [Table/Fig-3a].

The patient was scheduled for right open Anderson-Hynes dismembered pyeloplasty. Right RGP was performed, which was suggestive of a kink at the PUJ causing PUJO, as shown in [Table/Fig-3b]. Right dismembered pyeloplasty was done, as shown in [Table/Fig-3c].

The patient was discharged on postoperative day 5, and is currently asymptomatic and on regular follow-up.



[Table/Fig-3]: a) IVP image showing grossly dilated right renal pelvis (red arrow) and Kink at the upper ureter (yellow arrow); b) Retrograde pyelogram showing grossly dilated renal pelvis marked (red arrow) and Kink at the upper ureter near the pelvi-ureteric junction (yellow arrow); c) Dismembered pelvis (red arrow). Spatulated upper ureter being anastomosed to the pelvis (yellow arrow).

DISCUSSION

The PUJO is one of the most common causes of hydronephrosis. It mainly occurs in paediatric patients [4]. The aetiology of PUJO in children has been well studied, but in adults, it is still unclear. In a study carried out by Richstone L et al., it was observed that in 60% of adult cases, the main aetiology was crossing vessels [5]. A PUJO may be caused by a peristaltic or dysplastic segment at the PUJ. In

the first case, the aetiology of PUJO was compression of the PUJ due to a dilated lower calyx secondary to a narrow calyceal infundibulum and high insertion of the ureter in the pelvis. To date, only a few cases of calyceal dilatation and high insertion of the ureter in the pelvis have been reported, leading to PUJO [6]. A similar case report was published by Siddiqui MAK et al., documenting a high insertion of the ureter in a horseshoe kidney causing PUJO, which was managed by open Anderson-Hynes pyeloplasty [7]. But this case was managed uniquely, by infundibular dilatation and reduction calyco-plasty.

In the second case, aetiology of PUJO was an aberrant systemic vein compressing the PUJ. Vessel crossing is considered an etiology of PUJO [8]. These are most commonly caused by aberrant lower polar arteries; abnormal crossing veins are rare [9]. In this case, an aberrant systemic vein was observed compressing the PUJ, resulting in PUJO secondary to external mechanical compression. Dismembered pyeloplasty was performed, and the compressed/aperistaltic segment was excised and sent for Histopathological Examination (HPE). There are only a few reported cases in the literature of systemic veins causing PUJO [10]. A similar case report has been published by Maheshwari PR and Shammam O in 2008 documenting an anomalous ileocolic vein causing obstruction at the PUJ, which was managed by Anderson-Hynes pyeloplasty [11].

In the third case, the etiology was a kink at the PUJ secondary to adhesions around the upper ureter, which is a distinct and rare cause of PUJO. The patient was managed with open Anderson-Hynes dismembered pyeloplasty. In a systematic review of 12 studies comprising 513 patients done by O'Sullivan NJ and Anderson S, 15.3% of cases of PUJO were due to adhesions and kinks at PUJ. All of them were managed by Anderson-Hynes pyeloplasty, either done by open, laparoscopic or robotic approach [6].

In this case series, all three cases were very rare and unique presentations of PUJO. The first case was managed uniquely with infundibular dilatation and reduction calyco-plasty. Two patients were managed with Anderson-Hynes pyeloplasty. This is considered the gold standard for the management of PUJO [12]. Laparoscopic pyeloplasty has emerged as a minimally invasive alternative to open surgery, with reported success rates of 90-95% [13] and advantages such as reduced postoperative pain, shorter hospital stay, and improved cosmesis [14]. Robotic assistance has further improved suturing precision and ergonomics, particularly in paediatric cases. However, laparoscopic management of PUJO becomes technically demanding in anatomically complex situations such as grossly dilated calyces, aberrant systemic crossing veins, or PUJ kinks with adhesions, as seen in our cases. In such settings, open pyeloplasty provides superior exposure, tactile feedback, and easier vascular control, while maintaining a high long-term success rate exceeding 95%. Additionally, the steep learning curve and increased operative time of laparoscopy limit its universal applicability. Thus, while laparoscopy offers clear benefits in routine PUJO, open dismembered pyeloplasty remains the gold standard in rare and challenging anatomical variants.

CONCLUSION(S)

The PUJO itself can present differently in each patient, which necessitates a tailored approach in each case. Open dismembered pyeloplasty remains the treatment of choice and the gold standard in most cases of PUJO. Intraoperative decision-making becomes crucial when altered anatomy or anomalies are encountered, requiring informed decision-making right on the spot. It can be performed laparoscopically or via an open approach. In either approach, the success of the surgery lies in the proper preoperative examination, imaging, meticulous management during surgery, and staying vigilant while dealing with such challenging cases.

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