

## Case report

# Lower moiety pelvic-ureteric junction obstruction (PUJO) of the duplex kidney presenting with pyonephrosis in adults

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**Abstract.** The most common congenital abnormality of the urinary tract is a duplex kidney. Pelvic-ureteric junction obstruction (PUJO) is a rare association that can affect the lower moiety of incomplete duplex kidneys. We report two adult cases of PUJO of the lower moiety in a duplex kidney that both presented with pyonephrosis. This late presentation of lower moiety PUJO with pyonephrosis has not been described previously. We describe the imaging appearances of this rare association and highlight this important diagnostic consideration in lower moiety hydronephrosis of the adult patient.

Duplex kidney is the most common congenital abnormality of the urinary tract with a reported urographic incidence of around 2% [1]. It is an important condition to recognise in childhood because of its frequent association with other urinary tract anomalies such as an ectopic ureteric orifice, vesicoureteric reflux (VUR) and uretero-coele [2]. These associations predispose the urinary tract either to recurrent urinary tract infection and its complications, or to obstruction, both of which may cause long-term renal damage.

Pelvic-ureteric junction obstruction (PUJO) is a rare association and is known to affect the lower moiety of incomplete duplex kidneys [3, 4]. This may also occur in complete duplex kidney (where there are totally separate ureters), and can be overlooked when there is coexisting VUR in children [5]. It is important to recognise this rare association since early surgery may be required to preserve renal function and provide symptomatic relief.

We report two cases of PUJO of the lower moiety in a duplex kidney, both presenting with pyonephrosis in adult life. Such late presentation of lower moiety PUJO with pyonephrosis has not been described previously.

### Case 1

A 17-year-old female presented with a 1 day history of left loin pain with no previous history of urinary tract infections. The patient had left loin tenderness and was afebrile, but urine dipstick revealed a trace of leucocytes and moderate red blood cells (3+), and there was a raised white blood cell (WBC) count of  $24 \times 10^3 \text{ mm}^{-3}$ . Serum creatinine was within normal limits ( $50\text{--}100 \mu\text{mol l}^{-1}$ ).

Acute intravenous urography (IVU) showed a left duplex kidney with a hydronephrotic lower moiety, and a normal right kidney (Figure 1). Ultrasound confirmed the IVU findings (Figure 2). Because of the combination of raised WBC count and an obstructed urinary system, percutaneous nephrostomy of the left lower moiety was

performed under ultrasound guidance. Cloudy urine was drained, microscopy of which revealed WBCs ( $>30$  per high power field). There was no significant growth on



**Figure 1.** Acute intravenous urography shows a left duplex kidney with a hydronephrotic lower moiety.



**Figure 2.** Ultrasound shows a left duplex kidney with marked hydronephrosis of the lower moiety.

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subsequent culture, but she had already been started on oral ciprofloxacin 500 mg twice daily.

A subsequent nephrostogram revealed a markedly dilated pelvicalyceal system and renal pelvis with a narrow PUJ and slender ureter. The appearance was of a PUJO of the lower moiety of the left duplex kidney (Figure 3). A micturating cystogram confirmed the absence of VUR, and a MAG-3 renogram indicated minimal renal function in the left lower moiety (Figure 4a, b).

The patient opted for a left lower moiety heminephrectomy, and pathological examination of the resected specimen showed chronic pyelonephritis secondary to PUJO.

## Case 2

A 48-year-old female patient presented with a 5 week history of recurrent left loin pain associated with increased urinary frequency and nausea. She had no previous history of urinary tract infections. There was left loin tenderness but she was afebrile. Urine dipstick revealed a trace of red cells and leucocytes. There was borderline raised white



**Figure 3.** Antegrade study reveals a dilated pelvicalyceal system and renal pelvis with a narrow pelvic-ureteric junction and slender ureter. The appearance is of a pelvic-ureteric junction obstruction of the lower moiety left duplex kidney.

blood cell count of  $11 \times 10^3 \text{ mm}^{-3}$ , and serum creatinine was within normal ( $50\text{--}100 \mu\text{mol l}^{-1}$ ).

An acute IVU revealed a duplex left kidney with a hydronephrotic lower moiety (Figure 5). Ultrasound examination showed that the hydronephrotic lower moiety contained internal echoes (Figure 6), which in combination with the raised WBC count, led to a high index of suspicion for pyonephrosis. An ultrasound guided nephrostomy was performed and cloudy urine was drained. A subsequent nephrostogram indicated the presence of a hydronephrotic lower moiety with PUJO (Figure 7).

The midstream specimen of urine (MSU) sample collected 2 days prior to nephrostomy insertion subsequently grew coliforms, whereas the urine sample taken via the nephrostomy contained abundant white cells ( $>30$  per high power field) but culture was negative. However, the patient had been commenced on oral ciprofloxacin 500 mg twice-daily 2 days prior to the nephrostomy insertion and was also given a bolus dose of intravenous gentamicin as a prophylactic antibiotic immediately before the procedure.

Percutaneous antegrade ureteric stenting was subsequently performed with a view to renography prior to the consideration of surgery.

## Discussion

Duplex kidney is a relatively common normal variant that may remain concealed in the absence of associated complications. However, it frequently occurs in association with other urinary tract anomalies such as ureterocele causing upper moiety obstruction, or a defective vesicoureteric junction valve mechanism causing VUR to the lower moiety [2]. Both situations may present on imaging with a hydronephrotic moiety [4].

PUJO of the lower moiety in the duplex kidney is the most unusual of the associated anomalies [5] with a 2% reported incidence of PUJO in children with duplex kidneys [6]. Late presentation, in adult life, of lower moiety PUJO with pyonephrosis has not been described previously.

Pyonephrosis is a urological emergency since infection in an obstructed urinary tract will lead to rapid irreversible renal damage, and patients are at risk of systemic infection. It is important to recognise this condition in patients who may present with relatively innocuous symptoms such as loin pain, an abdominal mass or urinary tract infection, as urgent percutaneous nephrostomy for relief of obstruction and drainage of infected urine is indicated.

The diagnosis of duplex kidney is usually made by intravenous urography, complemented by delayed films. In difficult cases ultrasound, micturating cystogram, retrograde and antegrade pyelography may be helpful [7]. Presentation of a duplex kidney with lower moiety PUJO in adult life is frequently associated with renal damage and function of the affected moiety can be assessed by a static or dynamic radioisotope renogram [8, 9].

Interestingly, PUJO and ipsilateral VUR may coexist in the children with duplex kidneys. The ureter draining the PUJO moiety is usually dilated in the presence of VUR [5]. It has been postulated that the PUJO is secondary to the VUR which has led to kinking and inflammation around the PUJ of the lower moiety ureter [10].

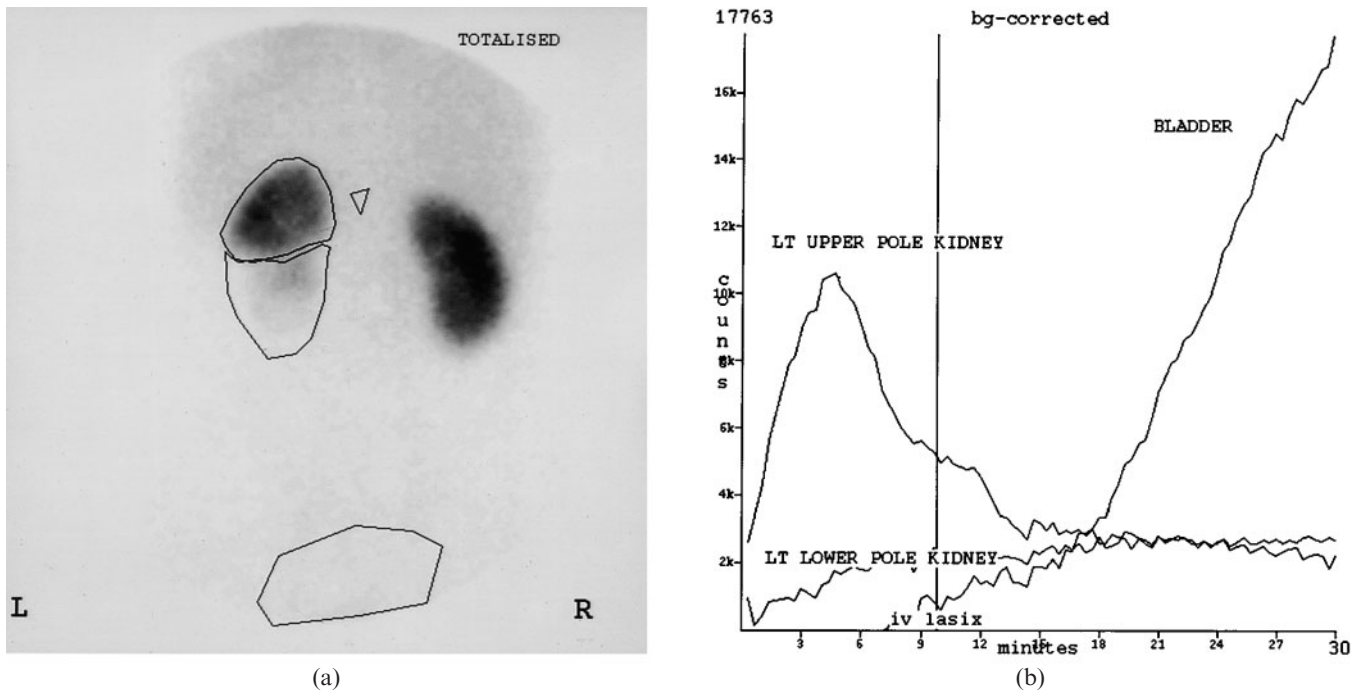


Figure 4. (a, b) Dynamic MAG-3 renogram indicates minimal renal function in the left lower moiety.



Figure 5. Acute intravenous urography shows a left duplex kidney with hydronephrotic lower moiety.

Surgical management of the lower moiety PUJO is tailored accordingly to the particular anatomical configuration and renal function. Treatment options include



Figure 6. Ultrasound examination shows a hydronephrotic lower moiety containing internal echoes (immediately prior to ultrasound guided nephrostomy insertion).

pyeloplasty, pyeloureterostomy, endopyelotomy or heminephrectomy. Ureteric stenting is a temporary procedure to relieve obstruction and symptoms. The patient in our first case was treated via heminephrectomy in view of minimal function of the lower moiety, and the patient in the second case had a ureteric stent inserted pending further investigation.

### Conclusion

PUJO is an unusual cause of hydronephrosis in the lower moiety of a duplex kidney, and it rarely presents in



**Figure 7.** Antegrade study shows the presence of a hydronephrotic lower moiety with pelvic-ureteric junction obstruction.

adult life. These two cases illustrate the imaging appearances and also highlight that it is an important albeit rare

diagnostic consideration in an adult with lower moiety hydronephrosis presenting as pyonephrosis.

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