# THE TRIPLEX ECTOPIC KIDNEY ASSOCIATED WITH BICORNUATE UTERUS

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### ABSTRACT

We present a rare case of young unmarried women who presented with complaints of left lumbar pain and burning urination for one month. The IVP and CTU demonstrated right ectopic triplex kidney and left duplex kidney. The left kidney shows staghorn calculus in lower moiety causing moderate hydronephrosis. It is associated with bicornuate uterus with single cervix and vagina confirmed on MRI Pelvis. To the best of author's knowledge, the triplex ectopic kidney has not been described earlier in the literature.

Keywords: Triplex kidney; ureteral triplication; IVP; CTU; bicornuate uterus;

# Introduction \_

Ureteral triplication is a rare congenital anomaly which was first described by Wrany in 1870.1 A review of the literature showed that there have been only about 100 reported cases of this condition. Diagnostic tools include Ultrasonography, Intravenous Pyelography, CT Urography, Retrograde Pyelography, MRU and Cystoscopy. This case report discusses a patient with ectopically located type 3 triplex kidney with concurrent left renal duplication and bicornuate uterus.

#### Case Presentation \_\_\_\_

24 yr old female was referred to Civil hospital Karachi, with left lumbar pain and burning urination for one month, not associated with fever. Her rest of history and general physical examination were normal except for mild pallor. Her routine hematological and other biochemical investigations were within normal limits. She was referred to Radiology department for X-Ray IVP which showed staghorn calculus in left renal area

in scout film. Further series revealed left sided duplex pelvi-calyceal system with staghorn calculus in lower moiety causing moderate hydronephrosis of upper and lower moiety. Right kidney is ectopic, located in the pelvis, appears distorted with a short ureter draining normally into urinary bladder (Fig. 1). Renal ultrasound confirmed empty right renal fossa occupied by bowel loops and a dysmorphic kidney in the right iliac fossa. There was an incidental finding of two

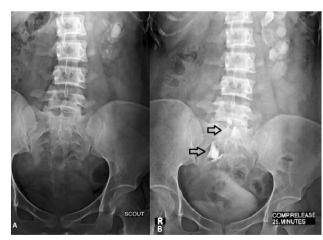


Figure 1: Scout film of X-Ray IVP (A) showing left renal calculi and right distorted kidney (arrows in B)

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uterine horns which was further evaluated by MRI which revealed bicornuate unicollis uterus with two separate functional uterine horns, single cervix and vagina (Fig. 2).

Subsequently, Triple-Phasic CT Urography with IV contrast administration (rate 3 ml/sec) was performed on 16 slice Toshiba Aquilion scanner slice thickness





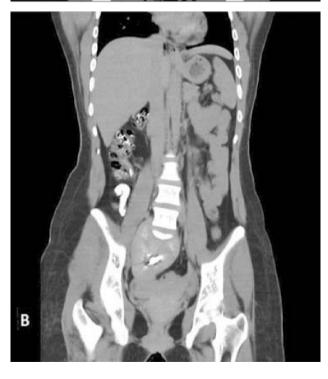
**Figure 2:** MRI Pelvis axial T2W image **(A)** and coronal STIR image **(B)** showing bicornuate uterus and dysmorphic pelvic kidney.

was 3 mm. The Axial, Multiplanar Reconstruction (MPR) and 3-Dimensional (3D) images were acquired at 1 mm reconstructed slice thickness in the excretory phase. There was prompt excretion of contrast through three pelvi-calicyceal systems uniting into single short

ureter, which shows orthotopic insertion on right side of urinary bladder (Fig. 3). There was duplex Left kidney with staghorn calculus and incomplete duplication of the ureter uniting at the level of L2 transverse process. (Fig. 4).

Our patient was referred to the urology department for nephrolithotripsy with double J stent placement for the passage of residual stones.





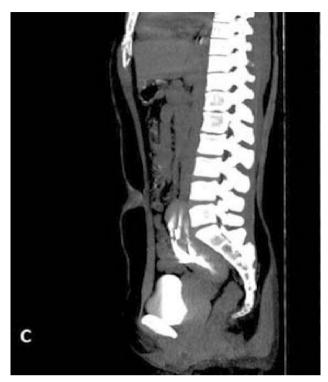


Figure 3: CTU excretory phase images showing triplex ectopic kidney.

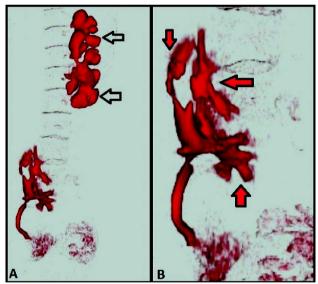


Figure 4: CTU MPR images showing left duplex (arrows in A) and right triplex (arrows in B) collecting systems.

#### Discussion \_\_\_

Ureteral duplication has an incidence of 0.8%. The triplex system is much rarer. It has previously been reported to be associated with other urological anomalies.

Ureteral buds develop as a diverticulum from the distal part of the mesonephric duct in the fourth and fifth weeks of embryological life. The distal end grows dorsally and then cranially, makes contact with the metanephros and differentiates into the renal pelvis and major and minor calyces.<sup>3</sup>

In triplication of ureter, the three ureteric buds could arise independently from the mesonephric duct or from early fission of one or more ureteric buds to join the metanephros.

The landmark article by Smith<sup>4</sup> in 1946 proposed the following classification system (Fig. 5).

- 1. Type I: Completely separate ureters where all three ureters open into the bladder or one opening may be ectopic.
- 2. Type II: Double ureters with one bifid.
- 3. Type III: Trifid ureters with one ureteric orifice.
- 4. Type IV: Inverted Y formation of one ureter with a separate ureter.

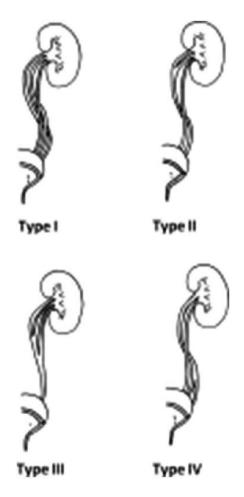


Figure 5: Smith classification of the ureteric triplex.

Clinical signs of ureteral triplication are non-specific, and duplication and triplication are often incidental findings on investigation. They consist of abdominal or back pain, hematuria, fever and urine storage and voiding symptoms.<sup>5</sup>

Triplication of ureter is reported to be associated with an increased incidence of congenital anomalies as well as a predisposition to infection and calculus formation. Haluk Soylemez at al<sup>6</sup> reported a case with vesico-ureteral reflux in a contralateral duplex system. Mills et al<sup>7</sup> reported a rather unique case of ureter triplication with a blind ending bifid ureter. Ander H et al<sup>8</sup> in 1997 reported a case of type 1 variant of ureteral triplication associated with vesicoureteral reflux into lower and mid pole ureters in a solitary kidney.

CT urography is the corner stone for the diagnosis of complex renal anomalies since both the renal parenchyma and urothelium can be evaluated with a single comprehensive examination. Ali SN et al<sup>9</sup> in 2014 presented a case of 10 year old male who presented with a history of recurrent urinary tract infections and right loin pain. CT Urogram was performed which showed a triplicate right ureter with gross hydroureter of the upper moiety. There was a duplex system present on the left side - comparable to our case. The patient underwent right upper pole hemi-nephroureterectomy.

# Conclusion \_

In this article, we present type 3 triplex kidney, according to Smith classification. To the best of author's knowledge, the triplex ectopic kidney with bi-cornuate uterus has not been described earlier in the literature. This case report signifies the role of multi-modality imaging techniques in reaching diagnosis of extremely rare renal anomaly.

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