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Complete left sided Duplex system with Upper Moiety Hydroureteronephrosis with Ectopic insertion of upper moiety Ureter Beyond the Sphincter(a rare Phenomenon with unusual presentation): Case Report and Review

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ABSTRACT

Background: Complete duplex systems are rare congenital anomalies of the urinary tract and usually present later in childhood with continuous dribbling or recurrent urinary tract infections (UTIs). Presentation in early infancy is unusual and may be easily overlooked.

Case Presentation: We describe the case of a 3-month-old female infant who presented with high-grade fever, recurrent UTIs, and persistent dribbling of urine despite apparently normal voiding. The mother also observed that the child cried and strained forcefully during micturition, which heightened concern. Imaging revealed a complete duplication of the left renal collecting system with ectopic insertion of the upper-moiety ureter into the urethra, complicated by vesicoureteral reflux (VUR) and moderate hydroureteronephrosis. Laboratory evaluation confirmed sepsis, and preoperative workup revealed deranged coagulation (prolonged PT/INR) secondary to urosepsis which was corrected with culture documented IV antibiotics and coagulopathy got corrected once sepsis part was over. The child underwent modified Politano-Leadbetter ureteric reimplantation after cystoscopy...

Conclusion: Ectopic ureters with insertion beyond the sphincter may present early with continuous dribbling and infections. Early recognition and timely surgical correction are essential to preserve renal function and prevent long-term morbidity.

1. INTRODUCTION

Congenital anomalies of the kidney and urinary tract (CAKUT) account for 20–30% of all prenatally detected anomalies, and among these, ectopic ureters are relatively rare entities [1]. An ectopic ureter refers to an abnormal ureteric opening that drains outside the typical bladder trigone region. The reported incidence in the general population is less than 0.1% [2]. Females are disproportionately affected, with published series showing ratios from roughly two to six females for every male [3]. This sexual dimorphism is explained by the broader anatomical possibilities for ureteral ectopia in females, where insertions may occur into the urethra, vestibule, or vagina, compared to males where the ureter typically inserts into seminal structures [4].

A strong association exists between ectopic ureters and duplex collecting systems, with approximately 80–85% of ectopic ureters arising from a duplicated kidney system [2,5]. In duplex kidneys, According to the classic Weigert–Meyer rule, the upper-pole ureter typically ends ectopically and is vulnerable to obstruction, whereas the lower-pole ureter usually enters the bladder normally but is more susceptible to reflux. [6]. However, as our case demonstrates, VUR can also involve the upper moiety.

The clinical presentation of ectopic ureters varies by age and sex. In female patients, a key clinical clue is persistent leakage of urine between normal voiding episodes, as the abnormal ureteric opening is positioned beyond the urethral sphincter. [7]. Recurrent urinary tract infections (UTIs), abdominal pain, or nonspecific urinary complaints are also common [8]. Presentation in infancy is particularly challenging because symptoms may be non-specific—such as fever, irritability, or

voiding dysfunction—and the diagnosis can easily be missed without high clinical suspicion [9].

Advances in imaging have improved early diagnosis. While ultrasonography can indicate the presence of a duplicated kidney, it generally lacks the precision to show the exact ureteric insertion site. Voiding cystourethrogram (VCUG/MCU) helps detect reflux, while CT or MR urography is often required for detailed anatomic delineation [10,11]. Nuclear renal scans are useful to assess split renal function and guide surgical planning [12].

Surgical management depends on the function of the involved renal moiety. If the upper moiety is poorly functioning, heminephrectomy is commonly performed [13]. In contrast, when function is preserved, reconstructive procedures such as ureteroneocystostomy or ureteroureterostomy are preferred [14,15]. Laparoscopic approaches have increasingly been adopted, with studies reporting shorter hospital stays and less morbidity, though outcomes between open and minimally invasive procedures are comparable [16].

Here we report a rare case of a 3-month-old female presenting with UTI, forceful voiding, and dribbling. Imaging confirmed a complete duplex system with ectopic insertion of the upper-moiety ureter into the vagina and associated VUR. The child was successfully managed with ureteroneocystostomy and DJ stenting, highlighting the importance of timely diagnosis and intervention in such rare infantile presentations

Case Presentation

A 3-month-old female infant was brought to our outpatient department with complaints of persistent urinary dribbling since birth, episodes of forceful micturition, and recurrent fever for 4–5 days prior to presentation. The mother was particularly worried because the child would cry and make a straining, forceful facial expression while passing urine, and despite apparently normal voiding with a good urinary stream, the diaper remained constantly wet in between.

The baby was the second child of non-consanguineous parents. Their first child, born at term, had unfortunately died on the third day of life from non-specified neonatal complications. Both parents were chronic tobacco consumers, though there was no history of consanguinity or known congenital anomalies in the extended family. The present infant was born at term via spontaneous vaginal delivery after an uneventful pregnancy, with a birth weight of 2.6 kg. No antenatal ultrasound had suggested urinary tract anomalies.

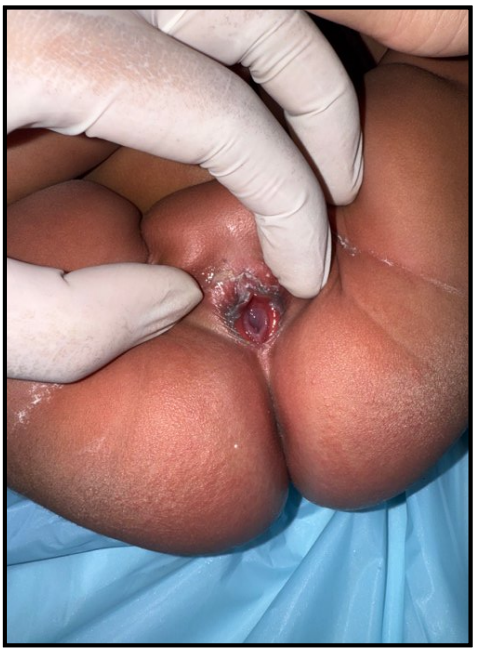


Figure 1: Abdominal Examination **Figure 2**: Perineal Examination

On examination, the infant was febrile (101°F), with stable hemodynamic. Abdominal exam revealed mild fullness in the left flank but no palpable mass. External genitalia were normal, and no abnormal urethral or vaginal openings were visible.

Laboratory investigations demonstrated leucocytosis (WBC count 18,000/µL, neutrophilic predominance) and elevated C-reactive protein. Urine microscopy revealed pyuria. Renal function tests were within normal limits (serum creatinine 0.3 mg/dL, blood urea nitrogen 12 mg/dL).

A significant perioperative concern was a deranged coagulation profile (prolonged prothrombin time and elevated INR). This was corrected by transfusion of fresh frozen plasma (FFP) on two separate occasions, after which coagulation parameters normalized and the patient was cleared for surgery

Imaging studies were undertaken sequentially:

Ultrasound of the abdomen performed outside our institution revealed a left duplex kidney with dilatation of the upper-moiety collecting system and ureter. The lower moiety appeared normal. The right kidney was normal in size and echotexture.

CT urography was done for detailed anatomical assessment. It confirmed complete duplication of the left renal collecting system. The upper-moiety ureter was markedly dilated and tortuous, coursing distally and inserting ectopically in the urethra beyond external sphincter. There was associated moderate hydroureteronephrosis of the upper moiety, while the lower-moiety system drained normally into the bladder. (**Fig. 3)**

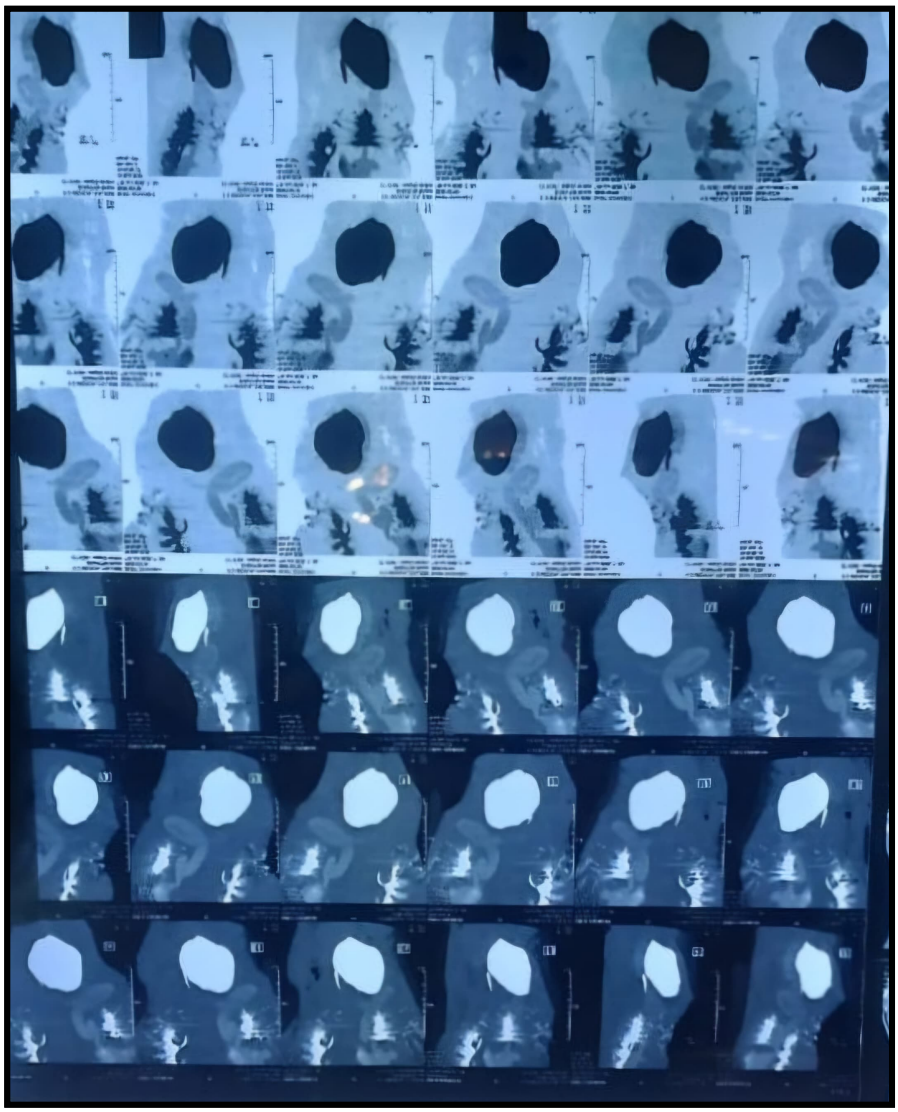


Figure 3: CT Urography

Micturating cystourethrogram (MCU) demonstrated vesicoureteral reflux into the ectopic upper-moiety ureter during micturition, with contrast outlining the tortuous dilated ureter and pelvis. The bladder outline and urethra were otherwise normal. **(Fig-4)**

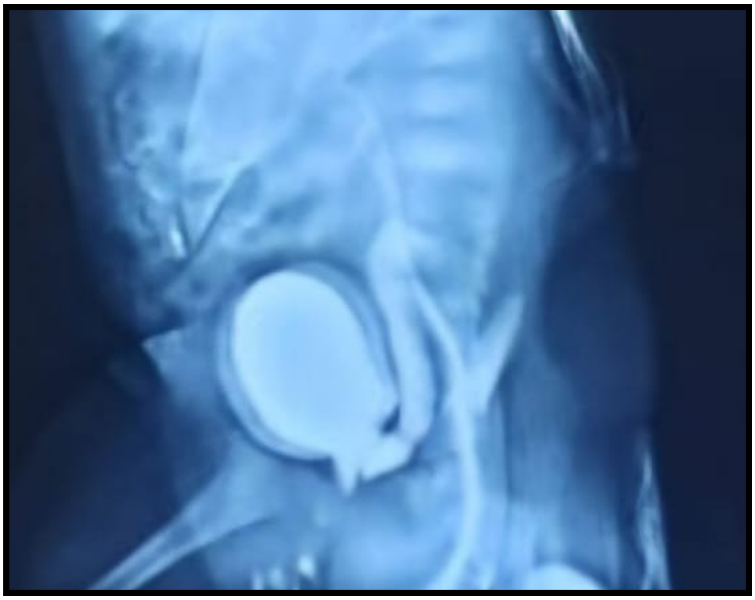


Figure 4: Micturating cystourethrogram (MCU)

DTPA scan was performed under normal room conditions, ensuring adequate hydration and preventing hypothermia. The DTPA revealed a duplex moiety on the left with improper drainage of upper moiety. The uptake was completely normal and there is evidence of delay drainage of upper moiety, However drainage pattern of lower moiety remained normal. Impression: Left complete Duplex system with obstructive pattern of left upper moiety.

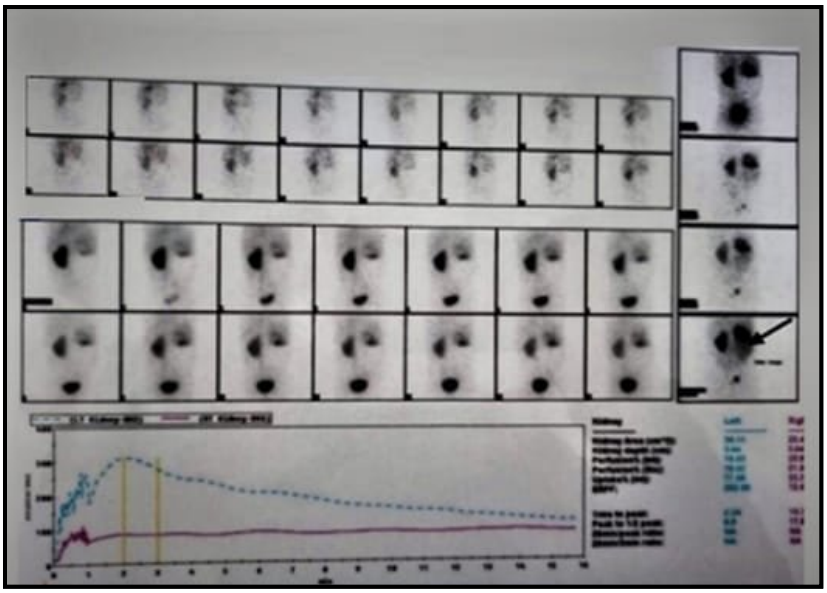


Figure 5: DTPA

Diagnosis: Left duplex kidney with ectopic vaginal insertion of the upper-moiety ureter complicated by vesicoureteral reflux and hydroureteronephrosis**.**

After appropriate counselling of the parents regarding surgical options, the decision was made to proceed with reconstructive surgery, as the upper moiety demonstrated preserved function and the infant was experiencing recurrent infections. The chosen procedure was **Cystoscopy (CPE) followed by Modified Lead-better Politano ureteric implantation.**

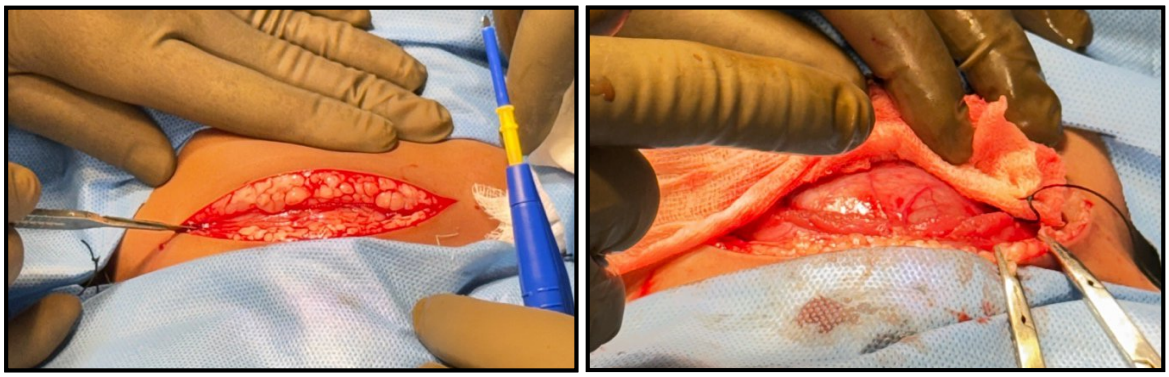
Operative details:

CPE Suggestive of one single opening on perineal examination . Another opening seen in the left lateral surface approximately 1.5 cm away from External urinary meatus.

Following bladder distension with saline, a Pfannenstiel incision was made to access the bladder and ureter. The ectopic ureter was identified and traced distally. Its terminal portion was ligated to prevent residual stump diverticulum formation.

The bladder was opened anteriorly, and a submucosal tunnel was fashioned to create an anti-reflux mechanism. The upper-moiety ureter was reimplanted into this tunnel. A 16 cm, 3.5 Fr double-J stent (both ends open) was placed to ensure patency. The bladder was closed in two layers, a corrugated drain was positioned, and the abdominal wall was closed in layers.

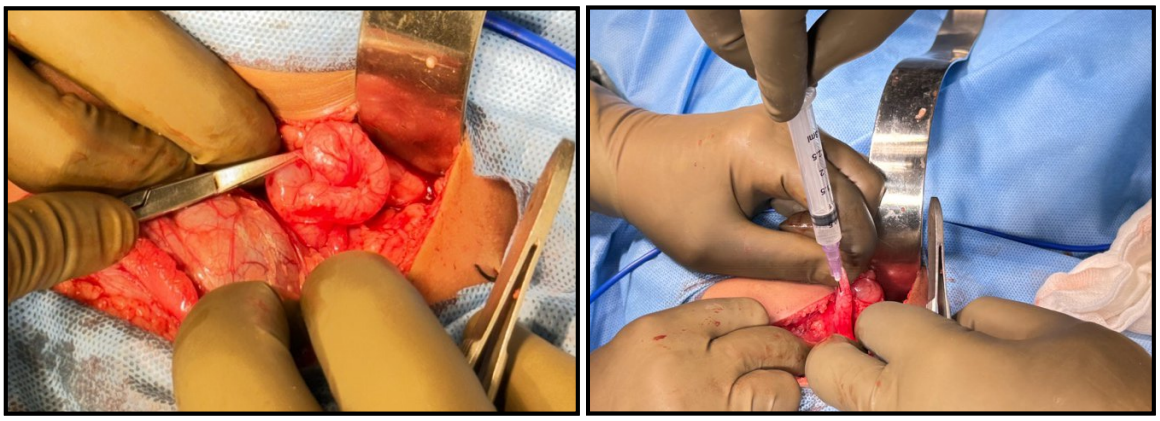
Approach and Bladder Exposure



A **B**

Figure 6: Intraoperative View Showing Surgical Approach(A) and Bladder Exposure(B)

Ureter Identification and Mobilisation



A **B**

Figure 7: Intraoperative View Showing Ureter identification and Mobilisation

Transection of ureter

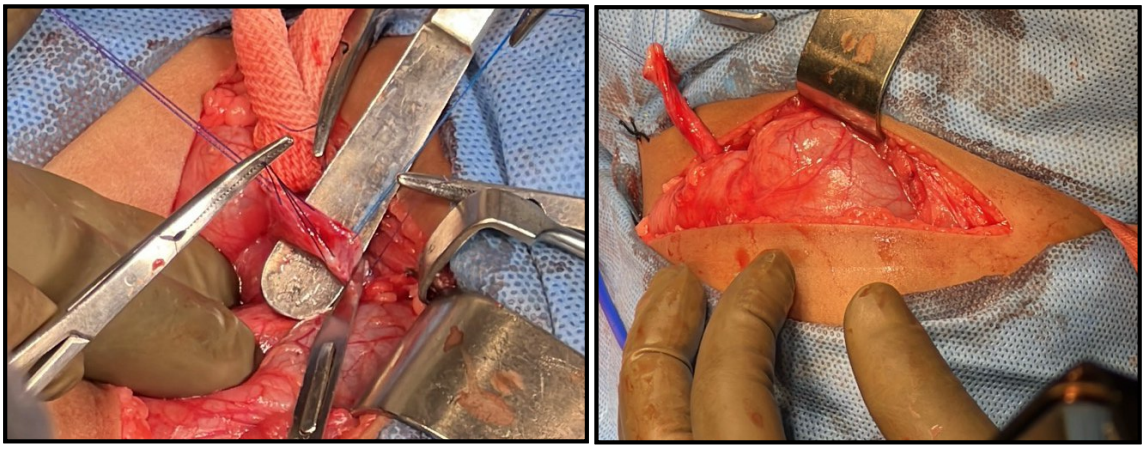


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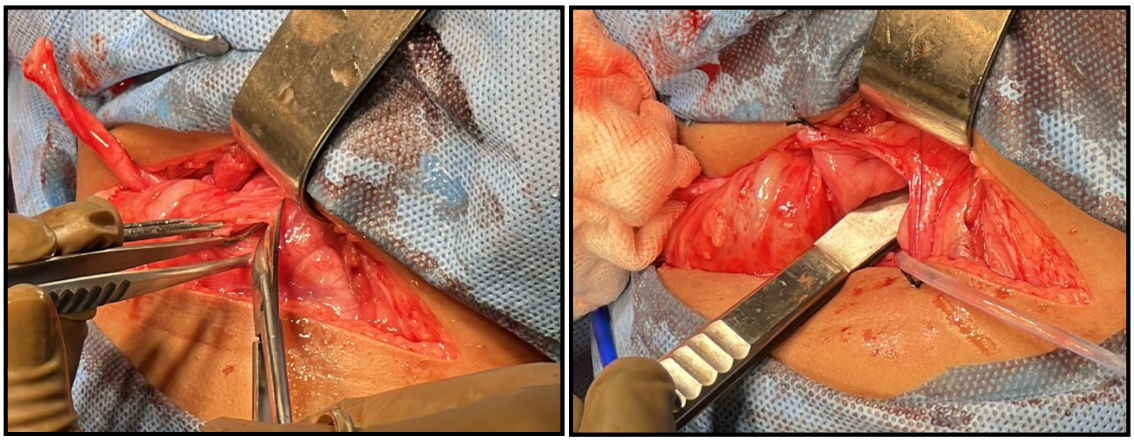
Figure 8: Intraoperative View Showing Transectioning of ureter

Bladder opening



A **B**

Figure 9: Intraoperative View Showing Bladder opening

Submucosal Tunnelling

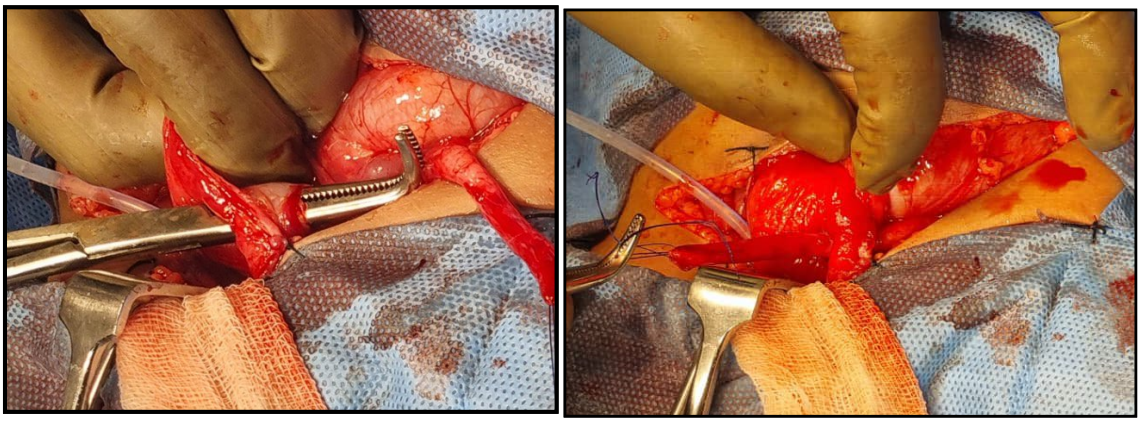


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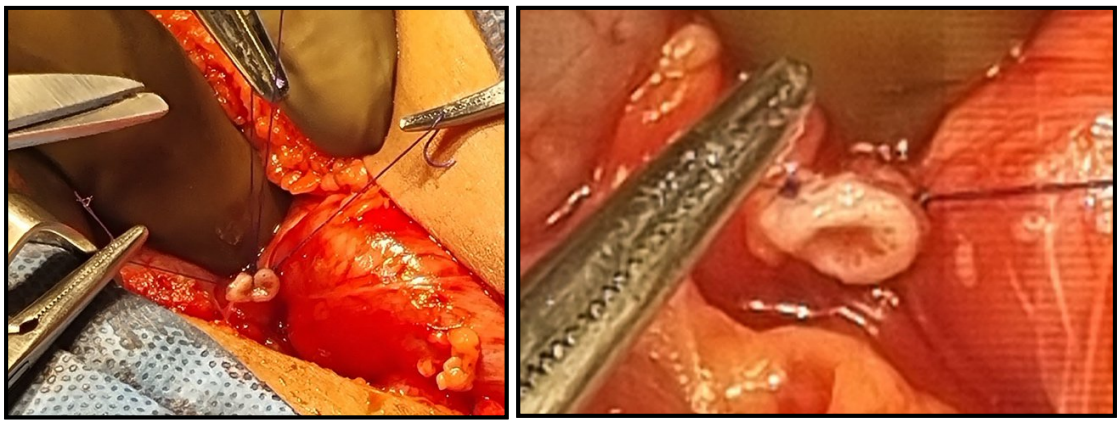
Figure 10: Intraoperative View Showing Submucosal tunneling

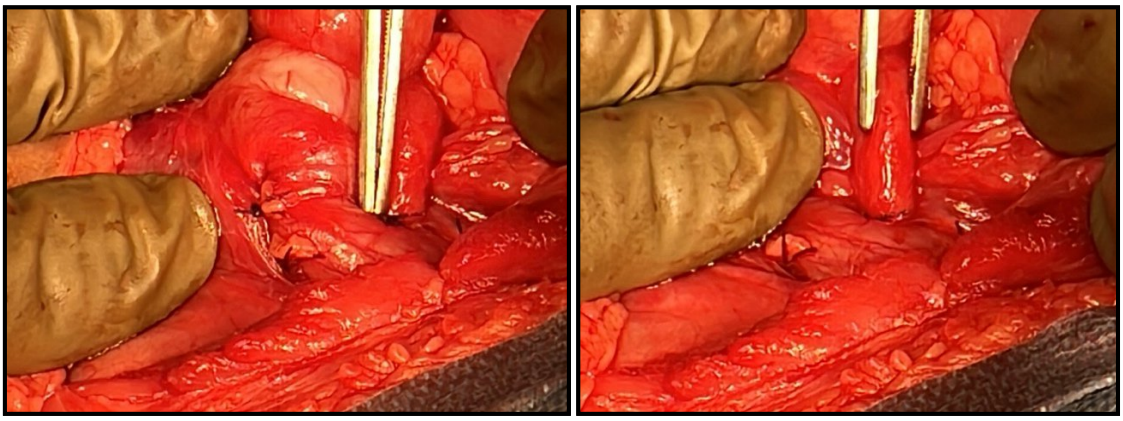
Orificial Fixation



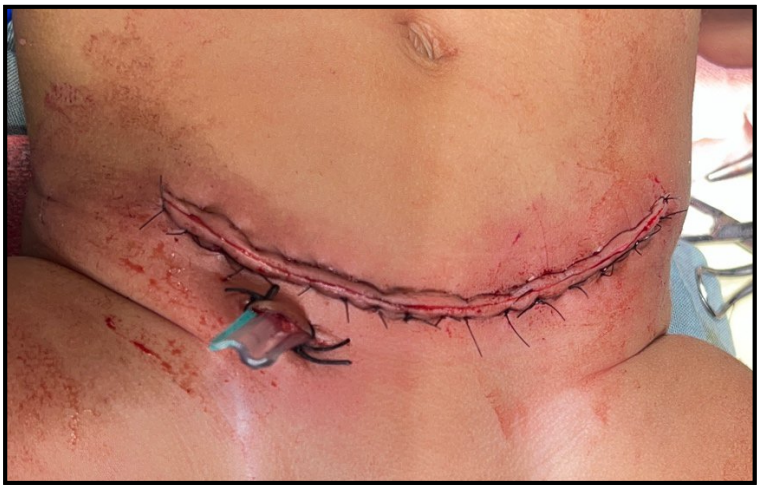
A **B**

Figure 10: Intraoperative View Showing Orificial Fixation

Closure



A B



C

Figure 11: Intraoperative View Showing Bladder closure (A & B) and Abdominal Wall Closure with drain fixation(C)

Postoperative course: The infant was maintained on intravenous antibiotics and analgesics. Postoperatively, the patient developed diuresis, which was managed appropriately with fluid replacement. Oral feeds were resumed within 24 hours. The corrugated drain was removed on postoperative day 3, and the patient was discharged on day 5 with oral prophylactic antibiotics.

2. DISCUSSION

Ectopic ureters present a significant diagnostic and therapeutic challenge in pediatric urology. Beyond their rarity, it is the variability of presentation and deviation from established anatomic rules that complicate management. In our case, the child not only had a complete duplex system with ectopic urethral insertion but also demonstrated vesicoureteral reflux (VUR) into the ectopic upper moiety — a pattern that does not strictly conform to the classical Weigert–Meyer law [6]. Such deviations, though uncommon, have been documented and underscore the importance of individualized assessment [7,8].

Early diagnosis in infants remains difficult, as clinical manifestations often overlap with common pediatric problems such as urinary tract infections, irritability, or failure to thrive. Imaging therefore plays a central role. While ultrasound provides the first clue, its sensitivity is limited in defining ureteral insertion [10]. MCU helps identify reflux but may not delineate ectopic pathways [12]. In our case, CT urography was decisive in confirming ectopic vaginal insertion, consistent with the growing use of cross-sectional imaging for complex anomalies [10,11]. MR urography, though not employed here, has been shown to be especially useful in avoiding radiation exposure in infants while providing superior anatomical detail [11,17]. Nuclear medicine studies such as DMSA or MAG-3 scans are also recommended to objectively assess moiety function and guide surgical choice, particularly when considering heminephrectomy [12,13].

The management of ectopic ureters associated with duplex systems has traditionally revolved around two major approaches: ablative (heminephrectomy) versus reconstructive surgery. Heminephrectomy with ureterectomy is often chosen for poorly functioning moieties [13], but concerns have been raised regarding the risk of damaging the remaining moiety, especially when the vasculature is complex [18]. Conversely, reconstructive procedures, particularly ureteroureterostomy and

ureteroneocystostomy, aim to preserve renal tissue.

Recent comparative studies provide valuable insights. Tao et al. [14] demonstrated that laparoscopic ureteroureterostomy (LUU) offers excellent outcomes in duplicated kidneys, with reduced perioperative morbidity. Bin et al. [15], in a comparative study of 80 pediatric patients, reported shorter operative times and faster recovery with LUU compared to ureteral reimplantation, though both procedures yielded comparable functional outcomes. Importantly, ureteroneocystostomy remains the procedure of choice in cases complicated by reflux, as in our patient, since it directly addresses the anti-reflux mechanism [13,14].

The durability of ureteral reimplantation has also been reaffirmed. Gerwinn et al. [16], in a large follow-up cohort, found that late complications such as recurrent reflux or obstruction were rare, supporting the long-term safety of this technique. Thus, the surgical choice must be individualized, taking into account not only anatomy but also functional status, presence of reflux, and surgeon expertise..

Equally noteworthy are the psychosocial aspects. The parents had previously lost a neonate within three days of birth, leading to heightened anxiety about their surviving child. The mother's observation of straining and crying during micturition was pivotal in prompting evaluation — a reminder of the importance of listening carefully to parental concerns in pediatric practice. Furthermore, both parents were chronic tobacco users, which, though not directly related to the anomaly, raises concerns about household health risks and future well-being. Addressing such issues through counselling is an essential yet often overlooked component of holistic pediatric surgical care.

The strength of this case lies in its early recognition, comprehensive imaging workup, and timely corrective surgery, despite perioperative challenges. The main limitation is the absence of nuclear functional imaging, which would have further quantified upper-moiety function. Nevertheless, the decision to preserve the moiety through reimplantation was justified by clinical and imaging evidence of preserved function.

3. CONCLUSION

Ectopic ureters associated with duplex systems, though rare, may present even in early infancy with subtle yet persistent symptoms such as dribbling and recurrent infections. Our case highlights the importance of maintaining a high index of suspicion, comprehensive imaging, and individualized surgical planning. Successful ureteroneocystostomy with stent placement not only preserved renal function but also provided durable relief of symptoms. Early diagnosis and timely intervention are critical in preventing long-term morbidity, and this case emphasizes that even atypical presentations beyond the sphincter must be carefully evaluated to optimize outcomes

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