

# Prolapsed Ectopic Ureterocele Presenting with Urethral Obstruction and Acute Kidney Injury

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## Abstract

Ureteroceles are cystic dilatations of the terminal ureter which fall within the spectrum of congenital abnormalities of the kidney and urinary tract (CAKUT). They are commonly associated with duplex urinary systems which have a vast clinical presentation and may be accompanied with complications such as urinary tract infections and urinary obstruction. The management of ureteroceles must be individualized and tailored in a logistical model to achieve a favorable outcome. The aim of this report is to highlight this individualized approach as seen in an infant with a prolapsed ectopic ureterocele associated with duplex ureter and presenting with acute kidney injury secondary to urethral obstruction.

**Keywords:** Ectopic ureterocele, Individualized approach, Management criteria.

## Introduction

Ureteroceles are cystic dilatations of the terminal ureter which occur with an incidence of 1 in 4000 live births. The majority of ureteroceles are associated with a complete duplication (duplex) system and are of ectopic location [1]. The clinical spectrum of ureteroceles is vast, from asymptomatic patients to patients presenting with complicated disease due to upper tract deterioration. It is not uncommon for ureteroceles to present with complications as a result of delayed diagnosis which has an impact on the management and outcome of such patients especially in low income countries [2]. The choice of therapy is individualized and depends on the following criteria: the clinical status of the patient, the patient's age; the renal function; the presence of obstruction; the presence of duplex system, the presence of vesicoureteral reflux (VUR) and the location either orthotopic or ectopic ureterocele [3]. The aim of this report is to highlight this individualized approach as seen in an infant with a prolapsed ectopic ureterocele associated with duplex ureter and presenting with acute kidney injury secondary to urethral obstruction.

## Case Presentation

A 5-month old female was referred to Kenyatta National Hospital from a primary care facility with complaints of inability to pass urine and suprapubic fullness. There was a history of occasional difficulty in passing urine and stool prior to this episode. The child was also treated for recurrent urinary tract infection on 4 previous occasions. Notably, the mother reports seeing an intermittent pink swelling on the genitalia on several occasions which retracted spontaneously upon micturition.

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An abdominal ultrasound was done before referral which showed bilateral hydronephrosis with normal morphology of the ureters and bladder. On assessment at our facility, the patient was noted to be sick looking, lethargic and febrile with a temperature of 38.6°C. There was suprapubic fullness and a distinct pink inter-labial cystic mass which completely obliterated the urethral orifice. The vaginal and anal orifices appeared normal externally (Figure 1).

She had a white cell count of  $32.28 \times 10^9/L$  with neutrophilia of 86%, the hemoglobin level was 6.7 g/dL. The urea and creatinine levels were markedly elevated at 41.1 mmol/L and 501.2 mmol/L respectively. The sodium and potassium levels were also deranged at 126.0 mmol/L and 6.45 mmol/L respectively. An urgent abdominopelvic ultrasound showed a 3.2x3.4x3.6 cm cyst within the bladder lumen arising from the left side of the bladder base. The bladder wall was markedly thickened and there was bilateral gross hydro-ureteronephrosis. On suspicion of a ureterocele causing bladder outlet obstruction, we used a urethral catheter to reduce the cystic mass back to the bladder and ballooned the catheter achieving significant drainage of urine. An urgent voiding cysto-urethrogram (VCUG) confirmed the diagnosis of a ureterocele with no vesicoureteral reflux. (Figure 2) Notably, upon withdrawal of the catheter to allow micturition the ureterocele ruptured and drained a significant amount of blood stained urine.

The management plan required a multidisciplinary approach involving the pediatric nephrologist, pediatric intensivist and the pediatric surgeon. Initial management included fluid and electrolyte correction with blood transfusion in addition to prompt administration of antibiotics and antipyretics. In view of the acute kidney injury (AKI), a peritoneal dialysis (PD) catheter was inserted and dialysis was commenced. The urethral catheter was also re-inserted for bladder drainage and monitoring of urine output. After 10 days the child was out of renal failure with normal renal function tests and normal septic screen. A renal nuclear scan was ordered but was not done due to financial constraints. We therefore ordered for a CT scan which revealed bilateral hydro-ureteronephrosis with loss of cortico-medullary tissue especially on the left side but with preserved renal function on both kidneys. The bladder wall was also noted to be thickened. (Figure 3)

After obtaining informed consent, surgery was performed through an open intra-vesical approach which revealed a ruptured left ectopic ureterocele (figure 4). Upon excision of the ureterocele, duplex ureteral orifices were noted. The two ureters on the left shared a common adventitial wall at the hiatal opening (Figure 4). The right ureteral orifice was single and appeared normal in position. We mobilized both ureters on the left as a unit and re-implanted them in a common sheath. The child made full recovery postoperatively with adequate urinary drainage and preserved renal function. She is currently on follow-up at our surgical clinic.

## Discussion

The diagnosis of ureterocele requires a high index of suspicion with investigations tailored in a methodical approach, preferably in specialized centers, before deciding

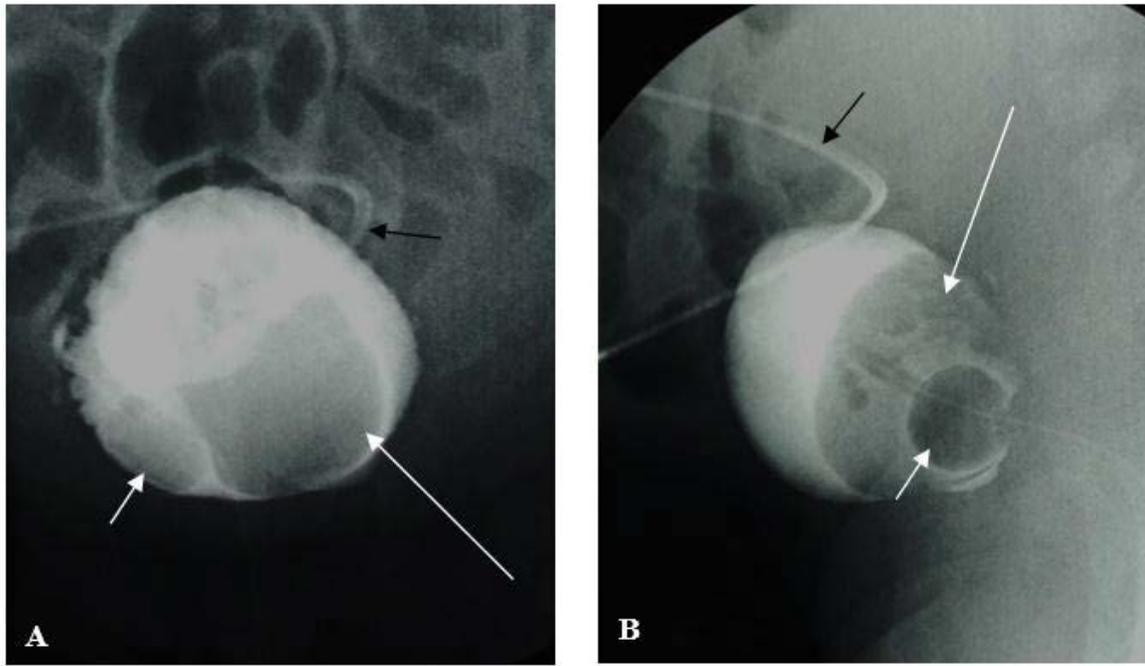
on the optimum management plan [2]. The case reported outlines the challenges faced in a primary care facility with limited diagnostic and specialized services leading to a delay in diagnosis with resultant sequelae of urosepsis and acute kidney injury.

The role of imaging is for definitive diagnosis of the ureterocele; to define the anatomy of the kidney, ureters and bladder and to assess the renal function. The diagnosis is made by ultrasonography which is user dependent and therefore it is possible to have false negatives [4]. The initial ultrasound done on our patient did not detect the ureterocele but detected upper tract involvement which prompted the referral. Further imaging by VCUG, Intravenous urogram (IVU), CT urography and renal nuclear scan are used to determine whether there are associated renal and urologic abnormalities [2]. We were able to perform a VCUG which confirmed the diagnosis and the absence of vesicoureteral reflux. A renal scan is used to evaluate the relative renal function especially in duplex systems which usually have an upper pole moiety with little or no function [3]. Due to financial constraints we could not perform a renal scan and opted for a CT urogram which detected some preserved renal function on the left side and this guided our choice of preserving the kidney in our management plan.

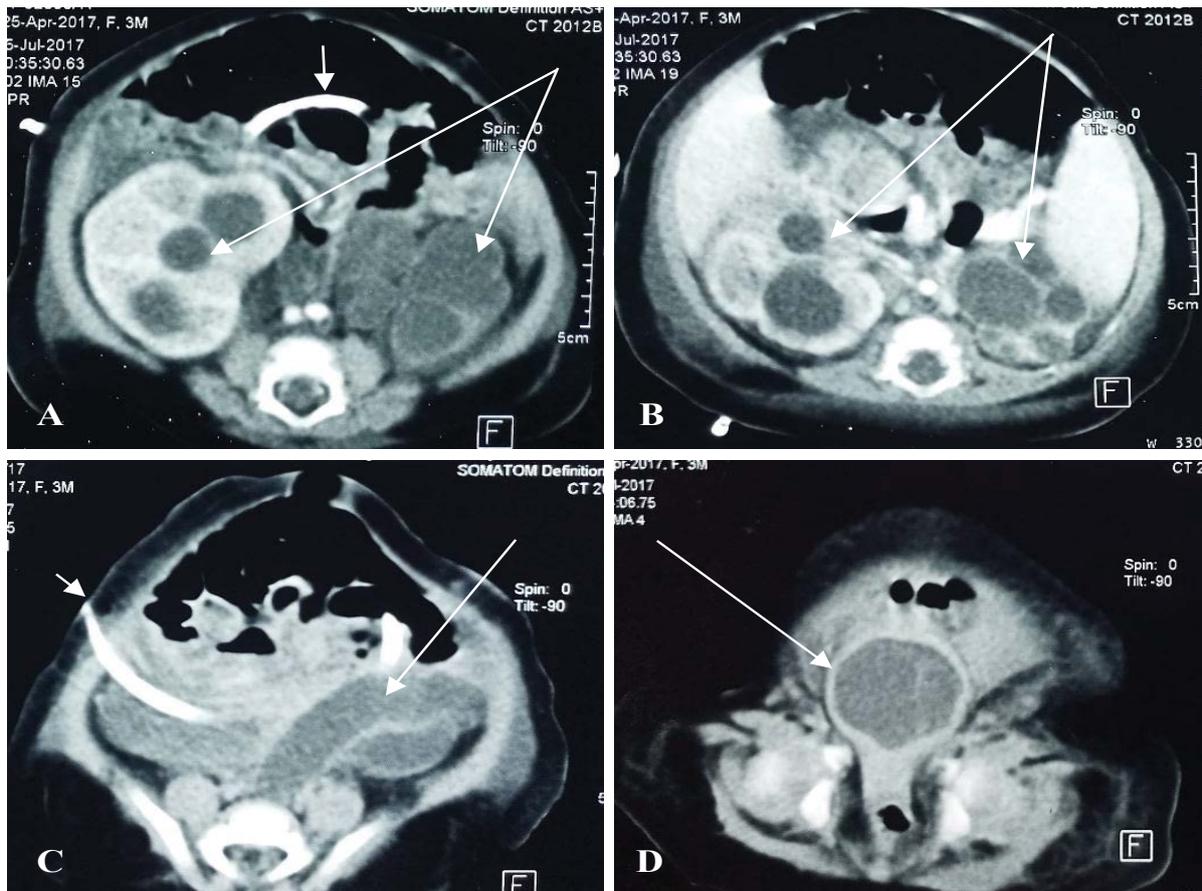
The choice of intervention is dependent on the clinical status. Patients presenting with complete urinary obstruction with or without sepsis require emergent urinary drainage. This is usually achieved via endoscopic incision of the ureterocele [5]. Our case presented a unique scenario as the ureterocele ruptured pre-operatively hence achieving drainage. Additionally, the patient was in AKI and required urgent stabilization with bladder drainage via catheterization and peritoneal dialysis.



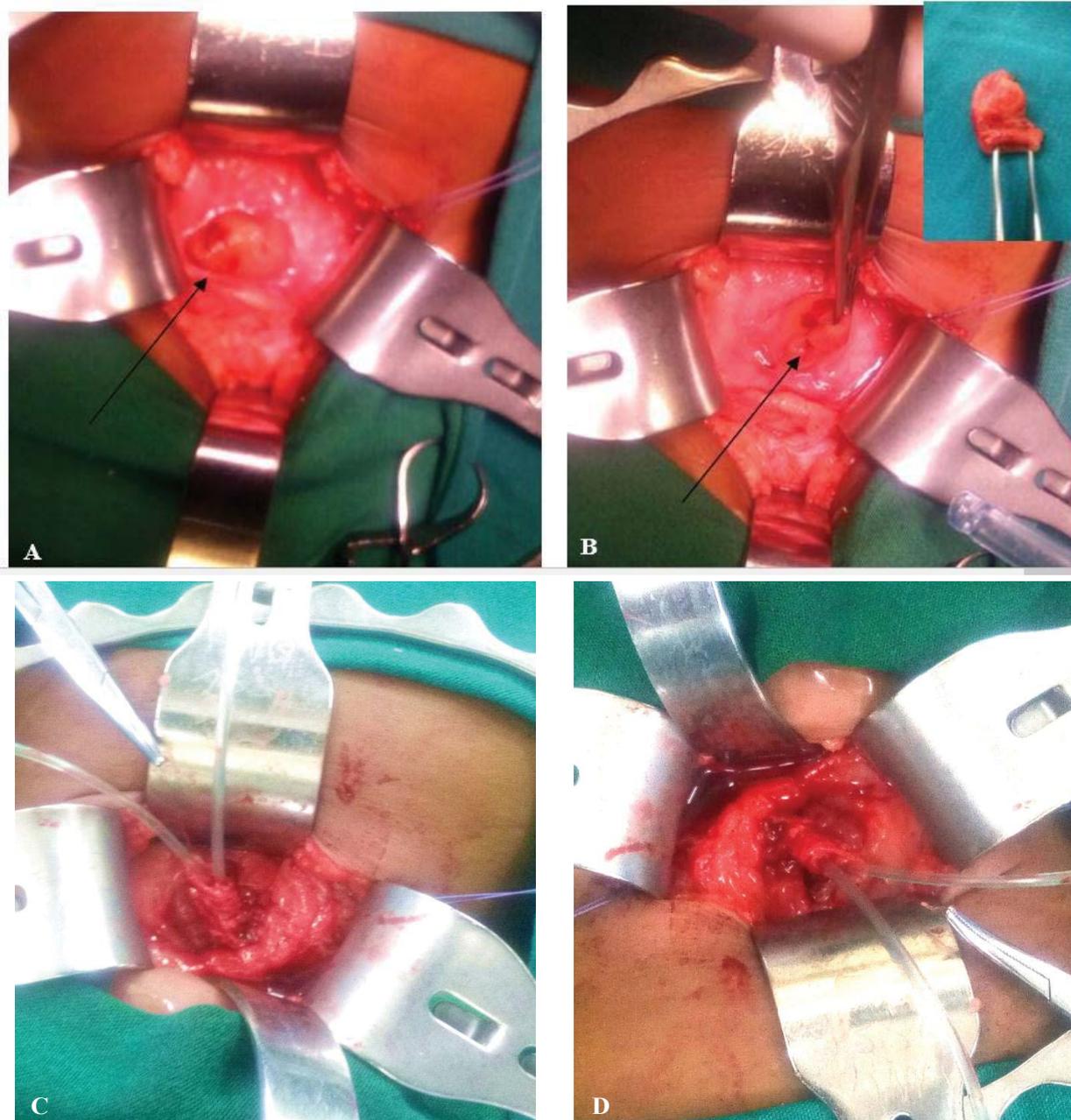
**Figure 1:** Inter-labial cystic mass. Image of infant's perineum showing a distinct inter-labial cystic mass consistent with a prolapsed ureterocele.



**Figure 2:** VCUg showing the ureterocele as a cystic mass within the bladder lumen. A. Antero-posterior view and B. lateral view. Both views showing two fluid filled opacities, the ureterocele (long white arrows) and the urethral catheter balloon (short white arrows). Note the contrast filled bladder with trabeculations. The PD catheter is visualized (short black arrows)



**Figure 3:** CT scan showing bilateral hydro-ureteronephrosis and thickened bladder wall. A and B. Axial images showing bilateral hydronephrosis (long white arrows). The PD catheter can be seen (short white arrow). C. Axial images showing bilateral hydro ureter which is more pronounced on the left side (long white arrow). The PD catheter can be seen (short white arrow). D. Axial scans showing a thickened bladder wall.



**Figure 4:** Intraoperative images showing ureterocele excision and duplex ureters. A. Intraoperative image showing an intra-vesical approach. The ureterocele can be seen upon entry into the bladder (black arrow) B. Image showing the ureterocele just before ureterocectomy (black arrow). The top right image shows the ureterocele after excision. C and D. Images showing duplex ureters intubated with two feeding tubes. Note that the two ureters share a common adventitial wall.

Intravesical (orthotopic) ureteroceles are commonly associated with single ureters and can be managed by endoscopic decompression which is successful in approximately 80 to 90 percent of cases. In contrast, endoscopic treatment is effective in only 25 to 30 percent of ectopic ureteroceles which are commonly associated with duplex systems [6]. Endoscopic decompression is done in emergency cases and facilitates a technically easier subsequent open re-implantation [7]. The spontaneous rupture in our patient negated then need for endoscopic decompression.

Open surgery for ectopic ureteroceles combined with ureteral duplication involves either an upper tract approach,

a bladder-level approach or a combination of both. The upper tract approach consists of partial upper-pole nephroureterectomy, pyelo-ureterostomy, uretero-ureterostomy and upper-pole ureterostomy [8]. In cases of VUR and obstruction with severe hydro-ureteronephrosis, a bladder level approach such as common sheath re-implantation with ureterocectomy or total reconstruction of the upper and lower urinary tracts with ureterocectomy, ureteral re-implantation, and hemi-nephrectomy is employed [9]. The common sheath re-implantation was favorable in our patient due to the common adventitial wall of the duplex ureters which had to be mobilized and re-implanted as a unit to preserve the blood supply. An algorithm on the

management of duplex system ureterocele is well outlined in the EUA guidelines, section on pediatric urology [3].

## Conclusion

In conclusion it is paramount that the management for ureterocele be individualized. A tailored approach of investigations and intervention yields the most favorable outcome. The approach depends upon whether the renal system is duplex or single, the age at presentation, the location of the ureterocele, the function of the involved renal moiety, the presence of reflux and the clinical status of the patient.

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## Patient Consent

Consent for surgery and publishing of this case report was provided in form of written consent, by the patient's mother. However, this report does not contain any personal information that could lead to the identification of the patient.

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## Conflict of Interest

The authors have no financial disclosures and declare that they have no conflict of interest.

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