

UNMASKING SINGLE-SYSTEM ECTOPIC URETER IN BOYS: INSIGHTS FROM A DECADE AT KUALA LUMPUR GENERAL HOSPITAL

M Sakhawi M Yusof^{1,2*}, Lynette Loi¹, Poongkodi Nagappan¹

¹Urology Department, Hospital Kuala Lumpur, Jalan Pahang, Wilayah Persekutuan Kuala Lumpur, Malaysia.

²Urology Department, Hospital Selayang, Lebuhraya Selayang-Kepong, Batu Caves, Selangor, Malaysia.

*Corresponding author:

Muhammad Sakhawi Mohd Yusof, Urology Department, Hospital Kuala Lumpur, Jalan Pahang, Wilayah Persekutuan Kuala Lumpur, Malaysia. Email: sakhawiyusof@yahoo.com

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ABSTRACT:

Single-system ectopic ureter is a rare congenital anomaly that primarily affects boys. Despite its low prevalence, it can lead to significant urinary tract complications if left untreated. This proposal aims to investigate the clinical presentation, diagnostic challenges, and optimal management strategies for single-system ectopic ureter among boys. By reviewing existing literature, analysing clinical cases, and exploring advancements in diagnostic imaging and surgical techniques, this research seeks to enhance our understanding of this condition and improve patient outcomes.

Keywords: ureteroscopy; leucocyte esterase; urinary tract infection

INTRODUCTION

Urinary tract with congenital anomalies, such as a single-system ectopic ureter, present considerable diagnostic and therapeutic challenges, particularly in male pediatric patients, where the clinical presentation can be atypical and mimic other lower urinary tract or genital infections[1,2]. These anomalies, collectively known as congenital anomalies of the kidney and urinary tract, are a leading cause of renal morbidity in children and a significant contributor to end-stage kidney disease

in adults. Specifically, maldevelopment of the collecting system leading to urinary tract obstruction is a primary identifiable source of chronic kidney disease in the pediatric population.

Literature Review

Single-system ectopic ureters are rare congenital anomalies, particularly in male patients. Unlike girls, where ectopic ureters often involve duplex systems, boys more commonly present with single-system ectopia, typically draining into

structures such as the vas deferens, posterior urethra, or seminal vesicles. The incidence of ectopic ureters is estimated at approximately 1 in 2,000–4,000 births, but single-system ectopias in males represent a small subset of these cases[3-5]. Clinically, boys with ectopic ureters often present later than girls and may exhibit less obvious symptoms. Continuous urinary incontinence, a hallmark in females, is less frequent in boys due to drainage into internal reproductive structures rather than outside the body. When incontinence is present, it is typically persistent and unresponsive to standard toilet training, prompting further investigation [6].

Diagnostic imaging is crucial for identification. Ultrasonography, voiding cystourethrogram (MCUG), and magnetic resonance urography (MRU) are commonly employed. Ultrasonography may reveal a small, dysplastic kidney or a distended ureter [7]. VCUG is particularly useful for excluding vesicoureteral reflux and to assessing bladder anatomy. MRU provides detailed anatomic information but is not universally available. Nuclear imaging may be used to evaluate renal function, especially since kidneys associated with ectopic ureters are often hypofunctional [8].

Management typically involves surgical correction. Ureteral reimplantation is the treatment of choice when the associated renal unit is salvageable. Nephroureterectomy is considered if the kidney is non-functional. Outcomes following surgery are generally favourable, with significant improvement in continence and preservation of renal function where applicable.

Despite advances in imaging and surgical techniques, delays in diagnosis remain common, emphasizing the need for heightened clinical awareness, particularly in boys with unexplained urinary symptoms [9,10].

METHODOLOGY

This study aimed to investigate the diagnostic challenges, imaging modalities, and management strategies for single-system ectopic ureters in boys,

thereby providing a comprehensive understanding of this rare anomaly and its impact on pediatric urological practice. This retrospective review of medical records from January 2016 to December 2024 was conducted to delineate the subtleties of clinical presentation, the utility of various imaging techniques, and the subsequent surgical and medical interventions, thereby contributing to improved patient outcomes.

RESULT

Case 1

A boy with VACTERL association, tethered cord syndrome, neurogenic bladder, bilateral ectopic ureters, horseshoe kidney, high-grade bilateral vesicoureteric reflux, subcoronal hypospadias, and bifid scrotum was followed since infancy for recurrent epididymitis and scrotal abscesses. Imaging showed bilateral hydronephrosis, a small-capacity, poorly compliant bladder, and progressive renal dysfunction. Despite early surgical recommendations for ureteric reimplantation, parental preference for conservative management and COVID-19 delays postponed intervention. He progressed to stage 4 chronic kidney disease, with urodynamics confirming neurogenic bladder and detrusor overactivity. Cystoscopy revealed a poorly developed posterior urethra, an absent trigone, and bilateral ectopic ureters. Management has remained conservative with clean intermittent catheterization, anticholinergic therapy, and nephrology follow-up, as surgery was deemed unlikely to reverse renal decline.

Case 2

A boy with antenatal urinary tract dilatation was diagnosed postnatally with left grade IV vesicoureteric reflux, later developing bilateral urinary tract dilatation with right-sided obstruction on MAG3 scan. He experienced recurrent febrile urinary tract infections, including ESBL *Pseudomonas aeruginosa*, despite antibiotic prophylaxis. Cystoscopy revealed tight phimosis, poorly developed posterior urethra, underdeveloped trigone, and possible ectopic ureter. Bilateral ureteric reimplantation (Politano-

Leadbetter) was performed in April 2022. Postoperatively, he voided with a good stream and remained infection-free, allowing discontinuation of prophylactic antibiotics six months after surgery.

Case 3

A boy, previously healthy, was diagnosed with type I choledochal cyst and benign multicystic gallbladder, with incidental bilateral acute pyelonephritis. Over the following year, he had recurrent febrile urinary tract infections, including ESBL *Klebsiella urosepsis* complicated by left renal abscess and bilateral renal microabscesses, requiring six weeks of IV meropenem. Imaging demonstrated bilateral renal scarring, a small right kidney, and bilateral single-system ectopic ureters with reflux during voiding. Cystoscopy showed a normal urethra, an underdeveloped bladder trigone, and ectopic ureteric orifices. Since the initiation of low-pressure timed voiding and cephalixin prophylaxis, he remained UTI-free, and antibiotics were discontinued in 2022. With stable renal function and absence of symptoms, ureteric reimplantation was deferred in favor of observation.

Case 4

A term male infant presented on day 8 of life with poor activity, feeding difficulty, and bilateral upper limb hypotonia, found to have acute kidney injury and bilateral urinary tract dilatation (UTD P3) on ultrasound. Persistent candiduria with *Candida albicans* despite IV fluconazole led to suspicion of bilateral renal fungal balls, prompting a switch to IV amphotericin B. MCUG revealed bilateral grade V vesicoureteric reflux with mild bladder trabeculation but no posterior urethral valves. Continuous bladder drainage was discontinued, and the patient was discharged on oral cephalixin and fluconazole. At follow-up, he was clinically well, voiding spontaneously, and feeding adequately; circumcision was discussed but declined, and infectious disease follow-up was arranged for ongoing antimicrobial management.

DISCUSSION

This often includes a multidisciplinary approach involving pediatric urologists, nephrologists, and radiologists to optimize diagnosis and treatment strategies. This integrated approach is crucial for managing the complex presentation of single-system ectopic ureters, which often involves subtle clinical signs, varied anatomical anomalies, and significant renal implications. Early and accurate diagnosis is paramount to prevent irreversible renal damage and improve long-term outcomes [11]. The diagnostic challenges are particularly pronounced in boys due to the less overt symptoms compared to girls, often manifesting as recurrent urinary tract infections, epididymitis, or subtle voiding dysfunction [12].

CONCLUSION

The comprehensive evaluation of single-system ectopic ureter cases in boys offers valuable insights for refining clinical protocols and enhancing early detection, which is crucial for mitigating long-term renal complications. This retrospective case series further emphasizes the need for increased awareness among clinicians regarding the varied presentations of this rare condition and the challenges associated with its definitive diagnosis and subsequent management in a resource-constrained setting. Furthermore, it highlights the necessity of developing more accessible and accurate diagnostic tools to facilitate timely intervention, particularly in regions where advanced imaging may be limited. Future research should focus on developing non-invasive screening methods and refining diagnostic algorithms to improve the identification of this elusive condition in pediatric populations, especially in resource-limited environments.

CONFLICTS OF INTEREST

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