Challenges in the Management of Bilateral Single-system Ectopic Ureters in Male Infants

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OBJECTIVE To describe our experience of managing bilateral single-system ectopic ureters in boys.

METHODS We discuss difficulties in diagnosis, importance of appropriate preoperative imaging, and the individualized surgical management for this rare congenital malformation.

RESULTS The first patient aged 3 months presented with urosepsis—subsequent imaging demonstrated bilateral single-system ectopic ureters, which were reimplanted at age 7 months. The second case was an antenatal diagnosis of bilateral hydronephrosis. Postnatal imaging identified bilateral ectopic single-system ureters. A suprapubic catheter was inserted to ensure optimal bladder drainage. He underwent a Cohen reimplantation at age 7 months because of recurrent urinary tract infections. The third patient was born with a duodenal atresia (repaired in the neonatal period) and a complex anorectal malformation, including a pouch colon with a colourethral fistula, a dilated, ectopic left ureter, and a hypoplastic urethra. He was initially managed with a colostomy and suprapubic catheter. At age 14 months, he underwent a left nephrouretectomy for a nonfunctioning kidney and posterior sagittal anorectoplasty. The right ureter was noted to be ectopic at this time and was reimplanted.

CONCLUSION Our series highlights the many challenges for diagnosis and management in boys with this condition because of the diverse presentation and need for appropriate preoperative imaging. Treatment is by bilateral ureteric reimplantation at an appropriate age. UROLOGY 83: 1373–1377, 2014. © 2014 Elsevier Inc.

An ectopic ureter draining a single-system kidney is an uncommon congenital malformation. The bilateral form is exceptionally rare, with a lower incidence in male patients.[1-3] The ureteric orifices may open intravesically into the bladder neck or extravesically into the urethra.[4] Vesicoureteric reflux (VUR) or obstruction may be associated with the anomalous location and structure of the orifices, and these children are prone to urinary tract infections (UTIs).[2,4] Unlike female patients, male patients with this condition do not generally exhibit urinary incontinence[5]; however, voiding dysfunction may occur because of abnormalities of the bladder trigone.[1,4] The mainstay of surgical management is bilateral ureteric reimplantation,[4] although the specific nature and timing of imaging and treatment should be tailored to the individual patient in every case.

The authors report their experience of managing 3 male infants with bilateral single-system ectopic ureters.

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PATIENTS AND METHODS

Case 1

A term male infant with no previous medical history presented to the emergency department at 3 months of age with a 1-week history of vomiting and lethargy. He was severely dehydrated and unresponsive and required a 20 mL/kg intraosseous fluid bolus. Initial investigations showed leukocytosis of 39×10^9/L with a neutrophilia of 19.7×10^9/L. His biochemical picture confirmed severe dehydration with urea 19.7 mg/dL, sodium 111 mmol/L, and creatinine 0.8 mg/dL. He had a marked metabolic acidosis (pH 7.04, bicarbonate 10 mEq/L) and an elevated lactate of 5.4 mmol/L.

Ultrasound scan (US) of his abdomen and renal tract raised the possibility of bilateral hydronephrosis, and a pediatric urology opinion was sought. A subsequent US showed sonographic features of pyelonephritis and pyonephrosis. Dilated ureters, thought to be related to the upper pole moieties of bilateral duplex systems, were seen inserting ectopically below the bladder base (Fig. 1). He was commenced on intravenous cefotaxime for urosepsis, and his symptoms settled with medical management.

A micturating cystourethrogram (MCUG) revealed a normal urethra, a bladder with a smooth outline, and no evidence of VUR during filling or micturition.

Seven weeks after the resolution of the acute infection, a magnetic resonance imaging urogram (MRU) was

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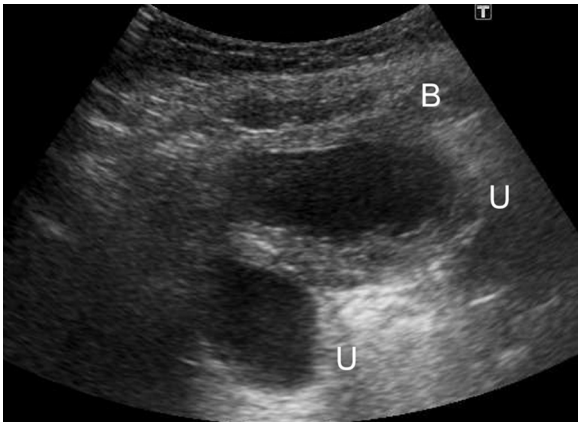

Figure 1. Renal tract ultrasound showing empty bladder (B) and dilated ureters (U) extending below the bladder base suggestive of ectopic insertion bilaterally.

performed under general anesthesia (Fig. 2). This clearly showed bilateral single pelvicalyceal systems with dilated clubbed calyces and bilateral ectopically inserting megaureters. A dimercaptosuccinic acid (DMSA) renogram showed that renal function was split almost equally between the 2 kidneys (right 53%; left 47%).

Cystoscopy and retrograde studies showed 2 dilated, tortuous ectopic ureters inserting below the bladder neck. No ureteric orifices were seen within the bladder.

At age 7 months, he underwent cystourethroscopy and bilateral reimplantation of the ectopic ureters. Access to the bladder was through a Pfannenstiel incision and extraperitoneal dissection. Both ureters were >15 mm in diameter and opened just distal to the bladder neck. They were divided as inferiorly as possible and were reimplanted bilaterally without tapering using the Politano-Leadbetter technique. Bilateral 4.7F blue ureteric stents (Bard Angiomed, Germany) and a 10F suprapubic catheter (SPC) were inserted, with separate exit sites on the skin.

Twelve days after his operation, the ureteric stents were removed. This required general anesthetic and cystourethroscopy, as they had become tangled around the balloon of the SPC. The SPC was clamped and removed after a further 10 days, once he had voided successfully per urethra.

Renal US performed 3 months after the operation showed reduced pelvicalyceal and ureteric dilatation. He has had several simple UTIs and so has been started on prophylactic trimethoprim, otherwise is doing well. He will be reviewed annually, with renal tract US and blood tests at ages 2 and 5 years.

Case 2

A term male infant with antenatally detected bilateral hydronephrosis was referred for a pediatric urology opinion. At birth, he was able to spontaneously pass urine but had a palpable bladder, masses in both flanks, and an initial creatinine of 5.6 mg/dL.

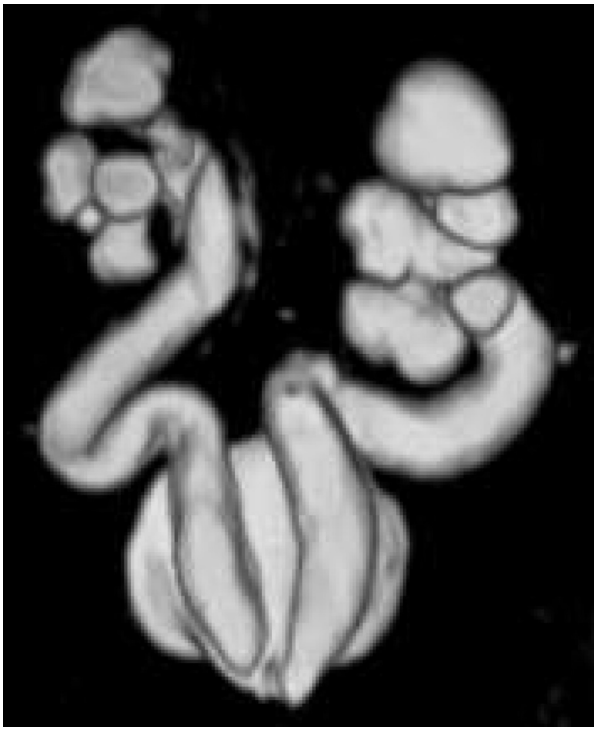

Figure 2. Magnetic resonance imaging reconstruction showing bilateral single pelvicalyceal systems with dilated clubbed calyces and dilated tortuous ureters inserting ectopically below the bladder neck.

He had a renal US and MCUG on day 3 of life. The US showed left hydronephrosis with a dilated, thick walled left ureter, which inserted below the bladder base. There was significant right-sided pelvicalyceal dilatation, with less ureteric dilatation. The MCUG confirmed a dilated ectopic left ureter and showed absent filling on the right side, but no evidence of posterior urethral valves. At age 2 weeks, a right nephrostomy was inserted to drain the dilated right renal pelvis. (Fig. 3) A right pyeloplasty was performed 6 weeks later for pelviureteric junction obstruction.

The ectopically inserted left ureter caused a bladder outlet obstruction, and so a SPC was inserted. Subsequent cystourethroscopy confirmed the presence of bilateral ectopic ureters draining into the posterior urethra (Fig. 4).

After these initial procedures, he developed recurrent UTIs, and so at age 7 months underwent bilateral ureteric reimplantation by the Cohen technique. MCUG 6 weeks postoperatively demonstrated no evidence of VUR and a normal urethra.

He was followed up 3 months after the operation, every 6 months for 3 years, and then annually. He has had no further UTIs, is growing well, and was successfully toilet trained. Blood tests have remained normal (most recent urea 4.8 mmol/L and creatinine 3.0 mg/dL).

US 6 months after the operation showed mild, bilateral dilatation of the renal pelvis and calyces, which has improved on subsequent scans. The left kidney showed