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Isolated bilateral simplex ureteric ectopia: Bladder capacity as an indicator of continence outcome

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Summary

Introduction
Isolated bilateral simplex ectopic ureters (BSEUs) are rare but pose a therapeutic challenge: ureteric reimplantation alone does not accomplish continence in all. Identifying the patients needing additional procedures for continence early could prevent multiple operations.

Objective
Potential preoperative indicators for postoperative continence are explored in eight BSEU girls without cloacal, anorectal, or spinal anomalies.

Study design
With institutional approval, all patients with BSEU between 1985 and 2012 were retrospectively reviewed. Cystoscopy determined the site of ureteric ectopia (6 of 16 at the bladder neck [BN], 5 of 16 below the BN, and 5 of 16 in the distal urethra). Bladders were assessed by a combination of ultrasound, urodynamics, micturating cystourethrogram, cystoscopic, and intraoperative observations. Expected bladder capacity for age (EBCA) was calculated by 30 ml + (30 ml × age in years) or 38 ml + (2.5 ml × age in months) for children greater or less than 2 years, respectively. Continence outcomes were appraised at a minimum of 4 years. The small number of patients precludes credible statistical analysis and therefore raw data are presented.

Results
Patients underwent cross-trigonal ureteric reimplantation at 1–5.5 years, in five without BN surgery and in three with a Young–Dees–Leadbetter BN tightening. Of those without BN surgery at reimplantation, four achieved satisfactory continence for their age, but one has had multiple procedures culminating in BN closure, ileocystoplasty, and Mitrofanoff. Among the BN-tightening group, one was in nappies at 4 years, one had residual stress incontinence after two further BN injections, and one proceeded to artificial urinary sphincter after two BN injections. Five patients had significant renal impairment.

Discussion
Patients with satisfactory continence after reimplantation alone and those needing further procedures tended to differ in their preoperative observations of bladder capacity and apparent BN competence. This study suggests preoperative observations of an empty bladder on serial ultrasound and/or a wide-open BN with small or even moderate bladder capacity at cystoscopy to indicate the need for BN surgery. In contrast, children with bladder filling to at least 30% of expected bladder capacity for age on preoperative ultrasound or apposition of the BN at cystoscopy may achieve satisfactory continence after ureteric reimplantation alone. Bladder capacity as an indicator of BN competence can also be correlated to continence outcomes in previously published series. Polyuria associated with renal impairment can exacerbate the challenge for continence.

Conclusion
Preoperative bladder capacity appears to be an indicator of inherent BN function and a thorough assessment of the urinary tract by cystoscopy, ultrasound, micturating cystourethrogram, and functional imaging may guide the surgeon on the need for BN surgery at the time of ureteric reimplantation. Where continence remains elusive, patients should be counselled that a further BN injection is occasionally of value although more significant BN procedures are required for most.

Table Continence outcomes for individual patients at their last FU. Cystoscopic observations regarding bladder size/BN competence and observed bladder volumes prior to cross-trigonal ureteric reimplantation with or without BN tightening.

Continence outcome	Patient	Age at FU	Pre-ureteric reimplantation cystoscopic finding bladder filling on US		Postoperative bladder function assessment
After ureteric reimplantation alone					
Continent	1	4 y	Not recorded	21 ml at 7 mo (36% EBCA)	100 ml at 4 y (67% EBCA)
	2	6 y	Good capacity	14 ml at 3 mo (30% EBCA)	281 ml at 6 y (130% EBCA)
	4	11 y	Not recorded	71 ml at 2.5 y (68% EBCA)	450 ml at 11 y (125% EBCA)
Enuretic	5	8 y	BN apposition	230 ml at 5 y (125% EBCA)	Capacity maintained
Proceeding to BNC cystoplasty	8	20 y	Small capacity BN wide open	Not recorded	220 ml at 14 y (50% EBCA) after colposuspension
Reimplantation with BN tightening					
In nappies	3	4 y	Moderate size BN wide open	14 ml at 12 mo (20% EBCA)	47 ml at 2.5 y (50% EBCA)
Proceeding to BNI	6	20 y	Moderate size BN wide open	Not recorded	Urodynamics: at 8 y 100% EBCA and at 20 y stress leak
Proceeding to AUS	7	16 y	Small capacity	Empty on serial US at 5 y	188 ml at 12 y (50% EBCA) after BNI

The EBCA is calculated for children aged below 2 years (38 ml + 2.5 ml × age in months) and those above 2 years (30 ml + 30 ml × age in years). Bladder filling is expressed as a percentage of EBCA (measured volume/EBCA × 100). AUS = artificial urinary sphincter; BN = bladder neck; BNC = bladder neck closure; BNI = bladder neck injection; EBCA = expected bladder capacity for age; FU = follow-up; mo = months; US = ultrasound; y = years.

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Introduction

Ureteric ectopia is characterised by an abnormal ureteric insertion into the lower urinary tract. Although the precise prevalence is uncertain, it is estimated that 80% of ectopic ureters are associated with ureteric duplication [1]. The remaining 20% consist of simplex or non-duplicated ectopic ureters, almost exclusively unilateral. At the extreme end of the spectrum are bilateral simplex ectopic ureters (BSEUs), a very rare entity with only a few cases reported in the literature without co-existing urogenital sinus or cloacal anomalies.

The proposed embryological origin of the simplex ectopic ureter has been previously described [2,3]. The ureteric bud forms towards the end of the fourth week as an outgrowth from the mesonephric duct. The caudal end of the duct is progressively absorbed into the urogenital sinus and becomes the trigone precursor, and the ureteric bud diverges from the duct and begins a cranial ascension. It is postulated that a more cranial origin of the ureteric bud results in delayed separation from the mesonephric duct, limiting the temporal window for ascent. As a consequence, the ureteric orifice assumes an abnormally low position and the trigone precursor is poorly formed [4]. In boys, the orifice of the simplex ectopic ureter is usually proximal to the external urethral sphincter. Therefore, common presentations include urinary tract infections, urgency or pain. In girls, the insertion of the ectopic ureter is usually distal to the sphincter and incontinence is the main presenting complaint. In ureteric ectopia, the kidneys are commonly dysplastic [5].

BSEUs pose a major therapeutic challenge. Although most series in the literature are small because of the rarity of the condition, ureteric reimplantation alone may not accomplish continence unless the defective bladder neck (BN) or small bladder capacity (if present) are also addressed. Wakhlu et al. [6] reported poor continence outcomes for three of five BSEU patients who underwent reimplantation alone. Similar findings have been published by Heuser et al. [7], supporting the view that in certain patients BN reconstruction or augmentation are important complementary procedures and their omission could lead to further surgery. In a survey by Podestà et al. [8], six out of seven girls without spinal anomalies were dry by day following bilateral ureteric reimplantation. However, three had also had a Young—Dees procedure. For others, continence is attained only after a multitude of procedures, culminating in BN closure and cystoplasty in four of six patients in a study by Jayanthi et al. [9] and urinary diversion in three of a series of nine patients by Williams and Lightwood [10].

It follows that early identification of the patient subset needing additional procedures for continence could diminish the need for multiple repeat surgeries. The experience of a single institution involving eight patients with bilateral simplex ureteric ectopia without cloacal, anorectal, or spinal anomalies is reviewed for possible links between preoperative findings and postoperative continence.

Materials and methods

With institutional approval, an electronic records search between 1985 and 2012 for the term "ectopic ureter" was performed. Retrieved records were retrospectively evaluated for co-existing anomalies and patient demographic information. Patients with duplex kidneys or co-existing anorectal, cloacal, or spinal anomalies were excluded from this series. Patients with anomalies that were unlikely to affect postoperative continence outcomes (such as congenital heart defects, imperforate hymen, biliary atresia, and epilepsy) were included.

Patient records were reviewed for clinical details and findings from ultrasound, micturating cystourethrogram (MCUG), nuclear scintigraphy, and cystoscopy were obtained. Overall renal function was assessed by serial serum creatinine and glomerular filtration rate (GFR). The upper tract was assessed by ultrasound for renal size, corticomedullary differentiation, parenchymal echogenicity, and cortical thickness, as well as pelvicalyceal and/or ureteric dilatation. All sonographic findings were corroborated with functional imaging. The site of insertion of the ureteric ectopia was determined by cystoscopy. Bladders were assessed by a combination of ultrasound, urodynamics, MCUG, cystoscopic, and intraoperative observations. The expected bladder capacity for age (EBCA) was calculated based on the formula 30 ml + (30 ml × age in years) for children above 2 years of age [11] and 38 ml + (2.5 ml × age in months) for children less than 2 years [12]. Continence outcomes were appraised at the age of at least 4 years, when maturation of neurological and anatomical continence mechanisms has mostly occurred. The small number of patients precludes valid formal statistical significance testing and therefore raw data is presented.

Results

Eight girls were diagnosed with bilateral simplex ureteric ectopia without cloacal, anorectal, or spinal anomalies between 1985 and 2012. A comprehensive summary of patient demographics, clinical presentation, associated anomalies, follow-up, and surgical procedures performed can be found in Table 1. Antenatal hydronephrosis in three and neonatal renal impairment in one prompted early urological assessments after birth. All other cases were diagnosed between the ages of 3 months and 5 years, having presented with recurrent urinary tract infections (3) and incontinence (1). Follow-up ranged from 4 to 20 years: upper and lower urinary tract outcomes are tabulated (Tables 2 and 3).

Upper urinary tract outcomes

At presentation, only three of 16 kidneys were non-dilated with normal cortico-medullary differentiation. Five renal units were removed for poor function. Five patients had significant renal function impairment (patients 1, 3, 4, 6, 8): three chronic kidney disease (CKD) stage II and two CKD stage III. In a sixth case (patient 7), there was a transient rise in creatinine after surgery, which resolved spontaneously.

Lower urinary tract outcomes

At cystoscopy, the site of ectopia was at the BN (6/16 ureters), below the BN (5/16 ureters), or within the distal urethra (5/16 ureters). In five cases (3, 4, 6, 7, 8), the bladder

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