

**Title:**

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

**Authors:**

Vasilis Stavrinos<sup>1,2</sup>, Paul Charlesworth<sup>1</sup>, Dan Wood<sup>2</sup>, Divyesh Desai<sup>1,2</sup>, Abraham Cherian<sup>1,2</sup>, Imran Mushtaq<sup>1</sup>, Peter Cuckow<sup>1,2</sup>, Naima Smeulders<sup>1,2</sup>

**Affiliation addresses:**

<sup>1</sup> Department of Paediatric Urology, Great Ormond Street Hospital NHS Foundation Trust, Great Ormond Street, London WC1N 3JH, UK.

<sup>2</sup>Departments of Paediatric and Adolescent Urology, University College London Hospitals NHS Foundation Trust, 235 Euston Road, London NW1 2BU, UK.

**Corresponding author:**

Miss Naima Smeulders

Department of Paediatric Urology, Great Ormond Street Hospital NHS Foundation Trust, Great Ormond Street, London WC1N 3JH, UK.

Tel +44 20 7405 9200 Fax +44 20 7813 8260

Email: [naima.smeulders@gosh.nhs.uk](mailto:naima.smeulders@gosh.nhs.uk)

# Isolated bilateral simplex ureteric ectopia: bladder capacity as an indicator of continence outcome.

## Extended Summary

### Introduction

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

### Objective

Potential pre-operative indicators for post-operative continence are explored in eight BSEU girls without cloacal, ano-rectal or spinal anomalies.

### Study Design

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

### Results

All underwent ureteric re-implantation at 0.5-5.5years, in five without BN surgery and in three with a Young-Dees-Leadbetter BN tightening. Of those without BN surgery at re-implantation, one is still less than 4years, three achieved satisfactory continence for their age, but one has had multiple procedures culminating in BN closure, ileocystoplasty and

Mitrofanoff. Amongst the BN-tightening group, one was in nappies at 4years, one had residual stress incontinence after two further BN injections and one proceeded to Artificial-Urinary-Sphincter after two BN injections (**Table**). Five patients had significant renal impairment.

Ureteric re-implantation	Dry		Incontinent		Further surgery
	Day + night	Day	Stress	In nappies	
<b>Without BN surgery</b>	68% → 125% (11) 30% → 50% (4)	125% → 125% (8)			Small → → 50% EBCA BNC, cystoplasty, Mitrofanoff – dry (20)
<b>With BN surgery</b>			“moderate” → 100% BNI - (20)	20% → 50% (4)	<20% → 50% AUS - dry (16)

**Table:** Continence outcomes for the 7 patients aged  $\geq 4$  yrs. Observed bladder volumes as a percentage of EBCA (expected bladder capacity for age). Arrow indicates transition from pre-operative %EBCA to post-operative %EBCA; age at last follow-up in brackets. BN: bladder neck. BNC: bladder neck closure. BNI: bladder neck injection. AUS: artificial urinary sphincter.

## Discussion

This study identifies pre-operative bladder capacity on non-invasive and intra-operative observations as an indicator of outcome. Three-quarters of children with at least 30%EBCA on pre-operative assessments achieved satisfactory continence after ureteric re-implantation alone when assessed at minimum of 4years. In contrast, those with cystoscopically small bladders or bladder volumes less than 20%EBCA continued to suffer from incontinence (despite bladder neck tightening at re-implantation) and underwent further procedures. Bladder capacity can also be correlated to continence outcomes in previously published series. Polyuria associated with renal impairment can exacerbate the challenge for continence.

## Conclusion

Pre-operative 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 re-implantation. Where continence remains elusive, patients should be counselled that a further BN injection is occasionally of value although more significant bladder neck procedures are required for most.

**Keywords:** bilateral ectopia, ectopic ureter, simplex system, urinary incontinence, bladder neck reconstruction.

## 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 (BSEU), 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, whilst 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].

BSEU pose a major therapeutic challenge. Although most series in the literature are small due to the rarity of the condition, ureteric re-implantation alone may not accomplish continence unless the defective bladder neck or small bladder capacity (if present) are also addressed. Wakhlu et al[6] reported poor continence outcomes for 3 of 5 BSEU patients who underwent re-implantation alone. Similar findings have been published by Heuser et al[7],

supporting the view that in certain patients bladder neck reconstruction or augmentation are important complementary procedures and their omission could lead to further surgery. In a survey by Podesta et al[8], 6 out of 7 girls without spinal anomalies were dry by day following bilateral ureteric re-implantation. However, 3 had also had a Young-Dees procedure. For others, continence is attained only after a multitude of procedures, culminating in bladder neck closure and cystoplasty in 4 of 6 patients in a study by Jayanthi et al[9] and urinary diversion in 3 of a series of 9 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 8 patients with bilateral simplex ureteric ectopia without cloacal, ano-rectal or spinal anomalies is reviewed for possible links between preoperative findings and post-operative continence.

## **Materials and methods**

With institutional approval, an electronic records search between 1985-2012 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 ano-rectal, cloacal or spinal anomalies were excluded from this series. Patients with anomalies that were unlikely to affect post-operative 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, micturating cystourethrogram, cystoscopic and intra-operative observations. The expected bladder capacity for age (EBCA) was calculated based on the formula  $30\text{ml} + (30\text{ml} \times \text{age in years})$  for children above 2 years of age[11] and  $38\text{ml} + (2.5\text{ml} \times \text{age in months})$  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, ano-rectal 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**. Abnormal antenatal ultrasound findings in 3 and neonatal renal impairment in 1 prompted early urological assessments after birth. All other cases were diagnosed between the ages of 3 months and 5 years, having presented primarily with recurrent urinary tract infections (3) and incontinence (1). Follow-up ranged from 19 months to 20 years: upper and lower urinary tract outcomes are tabulated (**Tables 2 and 3**).

### *Upper urinary tract outcomes*

At presentation, only 3 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 (cases 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 bladder neck (6/16 ureters), below the bladder neck (5/16 ureters) or within the distal urethra (5/16 ureters). In five cases (3,4,6,7,8) the bladder capacity was described as either small or moderate. The bladder neck was wider than anticipated in 3 patients (3,5,6) and the trigone was underdeveloped or absent in two (3,5). Micturating cystourethrography in 7 patients documented vesico-ureteric reflux on at least one side in all; additionally some inferences about bladder capacity and bladder neck competence were made in conjunction with other assessments.

All girls underwent ureteric re-implantation at 6 months to 5.6 years of age. In 5 cases this was without bladder neck (BN) surgery, but in 3 a Young-Dees-Leadbetter BN tightening was additionally performed. Amongst 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 (AUS) after two BN injections. Of those patients without BN surgery at re-implantation, one has had multiple procedures for continence culminating in BN closure, ileocystoplasty and Mitrofanoff at 20 years, one is still less than 4 years of age, the remaining three achieved continence to a satisfactory degree for their age (see **Table 3**).

The two groups differed in terms of their pre-operative bladder capacity (**Table – Extended Summary**). In patients undergoing BN surgery at re-implantation the documented cystoscopic, sonographic and/or urodynamic bladder volumes were considerably smaller than the expected bladder capacity for age (generally less than 20%) or the bladder



appeared small at cystoscopy. In contrast, the group that underwent re-implantation alone tended to have volumes at 30-100% of EBCA and moderate or normal bladder size on cystoscopy.

## **Discussion**

Experience with isolated bilateral simplex ectopic systems is limited and only a few series have been reported in the literature. Surgical management can be challenging and continence after ureteric re-implantation alone is not guaranteed. This study identifies pre-operative bladder capacity on non-invasive and intra-operative observations as an indicator of outcome. Three-quarter of children with at least 30% EBCA on pre-operative assessments achieved satisfactory continence after ureteric re-implantation alone when assessed at 4 years of age or more. In contrast, those with cystoscopically small bladders or bladder volumes less than 20% EBCA continued to suffer from incontinence (despite bladder neck tightening at re-implantation) and underwent further procedures.

BSEU is often associated with malformations that could potentially impede continence, particularly cloacal or spinal anomalies where anatomical or neurological continence mechanisms are directly undermined. Our series therefore excluded patients with cloacal/ano-rectal or spinal abnormalities. Other coexisting anomalies of the genital, gastrointestinal, nervous or cardiorespiratory systems were noted in patients with BSEU and have already been described in the literature[15-17], but such abnormalities were unlikely to affect continence outcomes in our series. As in previously published series significant renal impairment owing to renal dysplasia, persisting dilatation or UTI-associated scarring was observed in most of our patients. Polyuria associated with renal impairment can exacerbate the challenge for continence in this group of patients[8].

In simplex system ectopia where renal function is preserved, the surgical approach of choice is re-implantation of the distal ureter. If the upper tract is dysplastic and the renal unit is poorly functioning, a nephroureterectomy is indicated. In unilateral cases this can yield satisfactory results[1,13,14]. However, bilateral ectopia and its anatomical particularities described above pose a considerable challenge for the paediatric urologist. Post-operative outcomes are poor, although continence and control of recurrent infections can be achieved in selected cases[8,9].

Continence outcomes were appraised in seven patients aged at least four years, when maturation of neurological and anatomical continence mechanisms has mostly occurred. Continence success rates are similar to those previously reported in the literature[15], summarised in **Table 4**. In this cohort, daytime continence was achieved by ureteric re-implantation alone in three girls, by bladder neck surgery followed by bladder neck injections in a fourth, BN surgery, injections and AUS in a fifth, while one patient achieved continence only after bladder neck closure with augmentation cystoplasty. One girl remained incontinent at the age of 4 years.

Bladder capacity can also be correlated with continence outcomes in previously published series. Podesta *et al* published a series of seven patients with bilateral ureteric ectopia without spinal anomalies[8]. Five were offered ureteric re-implantation alone, of whom four were deemed to have had a good capacity bladder. Of these, three were dry day and night. Three patients were found to have a small bladder capacity. All were offered a Young-Dees bladder neck procedure (in two with and in one after ureteric re-implantation), achieving daytime dryness for all although two were enuretic (one due to polyuria).

A previous series from our institution more than 30 years ago documented a “fair” bladder in 5 and a “small” bladder in 4 of 9 patients with bilateral ureteric ectopia who survived infancy[10]. Those with a “fair” bladder achieved satisfactory continence in two, improved continence in two, but no improvement in one after ureteric re-implantation with bladder neck tightening. Those with a “small” bladder achieved continence by additional augmentation cystoplasty in one but the remainder proceeded to urinary diversion.

Since, Jayanthi *et al* have highlighted the multiple procedures endured by those who do not achieve continence after ureteric re-implantation alone[9]. Of the six patients with bilateral ureteric ectopia in the absence of spinal anomalies, continence was achieved only after bladder neck closure, augmentation cystoplasty and Mitrofanoff procedure in four while two were wet awaiting or had refused further surgery after previous bladder neck procedures. The authors suggest that bladder neck closure should be offered to those incontinent after a first bladder neck procedure. In our cohort, too, those with poor continence after initial surgery usually needed several further procedures to achieve continence, and indeed, one patient proceeded to bladder neck closure with augmentation cystoplasty.

Early identification of the prognostic factors for continence may improve optimal surgical decision-making. Heuser *et al* thought that incontinence in bilateral ureteric ectopia reflected a deficient bladder neck to an otherwise normal bladder[7]. A small bladder capacity does indeed appear to reflect bladder neck incompetence and, when detected pre- or intra- operatively, can indicate the need for BN tightening at re-implantation. The desirable outcome in bladder neck reconstructive surgery is a competent neck that allows micturition without a raised intravesical pressure. This balance is difficult to achieve[18]. Where it fails, not all patients opt for bladder neck closure and augmentation cystoplasty,

and bladder neck injections can be considered as secondary procedures with occasional success.

## **Conclusion**

Bilateral simplex ureteric ectopia is a rare and challenging condition associated with incontinence and chronic renal impairment. Pre-operative bladder capacity appears to be an indicator of inherent BN function and a thorough assessment of the urinary tract by cystoscopy, ultrasound, MCUG and functional imaging may guide the surgeon on the need for BN surgery at the time of ureteric re-implantation. Where continence remains elusive, patients should be counselled that a further BN injection can be considered although more significant bladder neck procedures including bladder neck closure are required for most.

## **Ethical Approval**

This work was conducted after gaining institutional approval.

## **Conflict of interest**

None.

## **Funding**

This research did not receive any specific grant from funding agencies in the public, commercial or not-for-profit sectors.

## **References**

- [1] Chowdhary SK, Lander A, Parashar K, Corkery JJ. Single-system ectopic ureter: a 15-year review. *Pediatr Surg Int* 2001;**17**:638–41.
- [2] Cox CE, Hutch JA. Bilateral single ectopic ureter: a report of 2 cases and review of the literature. *J Urol* 1966;**95**:493–7.
- [3] Johnston JH, Davenport TJ. The single ectopic ureter. *BJU Int* 1969;**41**:428–33.

- [4] Tanagho EA. Embryologic basis for lower ureteral anomalies: a hypothesis. *Urology* 1976;**7**:451–64.
- [5] Yan YJ, Feng ZX, Min ZH, Jin YX. Single-system ectopic ureters associated with renal dysplasia. *Pediatr Surg Int* 2004;**20**:851–4.
- [6] Wakhlu A, Dalela D, Tandon RK, Chandra H, Wakhlu AK. The single ectopic ureter. *BJU Int* 1998;**82**:246–51.
- [7] Heuser M, Zöller G, Seseke F, Zappel H, Ringert RH. Bladder dysfunction in children with bilateral single ectopic ureters. *J Pediatr Surg* 2002;**37**:E15.
- [8] Podestà E, Scarsi PL, Di Rovasenda E, Ferritti S, Magillo P, Dodero P. Vesical continence in bilateral ectopic single ureters. *J Urol* 2001;**165**:2363-5.
- [9] Jayanthi VR, Churchill BM, Houry AE, McLorie GA. Bilateral single ureteral ectopia: difficulty attaining continence using standard bladder neck repair. *J Urol* 1997;**158**:1933–6.
- [10] Williams DI, Lightwood RG. Bilateral single ectopic ureters. *BJU Int* 1972;**44**:267–73.
- [11] Hjälmås K. Urodynamics in normal infants and children. *Scand J Urol Nephrol* 1988; **S114**:20–7.
- [12] Holmdahl G, Hanson E, Hansom M, Hellström AL, Hjälmås K, Sillén U. Four-hour voiding observation in healthy infants. *J Urol* 1996;**156**: 1809–12.
- [13] Mathews R, Jeffs RD, Maizels M, Palmer LS, Docimo SG. Single system ureteral ectopia in boys associated with bladder outlet obstruction. *J Urol* 1999;**161**:1297–1300.
- [14] Choudhury RS, Chadha R, Bagga D, Puri A, Debnath PR. Spectrum of ectopic ureters in children. *Pediatr Surg Int* 2008;**24**:819–23.
- [15] Koyanagi T, Tsuji I, Orikasa S, Hirano T. Bilateral single ectopic ureter: report of a case. *Int Urol Nephrol* 1977;**9**:123–127.
- [16] O'Connor E, Peeraully R, Shepherd G, Shenoy M. Challenges in the Management of Bilateral Single-system Ectopic Ureters in Male Infants. *Urology* 2014;**83**:1373–7.
- [17] Ahmed S, Barker A. Single-system ectopic ureters: a review of 12 cases. *J Pediatr Surg* 1992;**27**:491–6.
- [18] Mollard P, Mouriquand PD, Buttint X. Urinary continence after reconstruction of classical bladder exstrophy (73 cases). *BJU Int* 1994;**73**:298-302.



Ureteric re-implantation	Dry		Incontinent		Further surgery
	Day + night	day	Stress	In nappies	
<b>Without BN surgery</b>	68% → 125% (11) 30% → 50% (4)	125% → 125% (8)			Small → → 50%EBCA BNC, cystoplasty, Mitrofanoff – dry (20)
<b>With BN surgery</b>			“moderate” → 100% BNI - (20)	20% → 50% (4)	<20% → 50% AUS - dry (16)

**Table Extended Summary:** Continence outcomes for the 7 patients aged  $\geq 4$  yrs. Observed bladder volumes as a percentage of EBCA (expected bladder capacity for age). Arrow indicates transition from pre-operative %EBCA to post-operative %EBCA; age at last follow-up in brackets. BN: bladder neck. BNC: bladder neck closure. BNI: bladder neck injection. AUS: artificial urinary sphincter.

**Table 1**

Pt	Current age (years)	Follow up		Presenting symptoms	Ureteric ectopia	Other anomalies	Procedures (age at surgery in brackets)
		Start (years)	Duration (years)				
1	3	0	1.6	Antenatal right hydronephrosis Oligohydramnios Recurrent UTI	R: urethra (dilated ureter coursing through bladder neck) L: BN	No	Right open nephro-ureterectomy (7 months) Left ureteric re-implantation (16 months)
2	5	0	4	Antenatal bilateral hydronephrosis Recurrent UTI	R: distal urethra, UO wide open L: just below BN	No	Cystoscopy and bilateral end ureterostomies (10 months) Bilateral cross-trigonal ureteric reimplantation (12 months) Revision of right vesico-ureterostomy (stenosis) (19 months)
3	5	0	4	Acute renal failure Failure to thrive Recurrent UTI	R: Urethra, distal half L: Below BN	Coarctation, choledochal cyst, biliary atresia, mild developmental delay, seizures	Bilateral cross-trigonal ureteric reimplantation and loose BN repair (15 months)
4	11	2	9	Recurrent UTI	R: BN, moderately open L: BN, wide open	No	Left laparoscopic nephrectomy and right ureteric re-implantation (3.5 years)
5	14	2	6	Recurrent UTI Incontinence	R: very low on underdeveloped trigone L: BN level	No	Right retroperitoneoscopic nephrectomy and left cross-trigonal ureteric reimplantation (5 years)
6	23	0	20	Antenatal dilated pelvic kidney Recurrent UTI Incontinence	R: right below BN level mid urethra L: BN level	Imperforate hymen	Left nephrectomy (5 months) Right ureteric reimplant and BN reconstruction (15 months) Left ureteric stumpectomy (4 years) BN injection (7 years)
7	24	5	11	Incontinence	R: BN, large L: distal urethra close to the sphincter	No	Bilateral ureteric reimplantation and BN tightening (5 years) BN injection (7 years), BN injection (13 years), BN injection (13 years), AUS (15 years)
8	31	0	20	Recurrent UTI Incontinence	R: urethra, below BN L: urethra, below BN	Small ASD, Episodic AF	Right pyeloplasty (4 months) Bilateral ureteric re-implantation (2 years) Right to left TUU (11 years), colposuspension (12 years) Right nephrectomy (13 years), ileocystoplasty & left ureterolysis (17 years), BN closure, redo Mitrofanoff (18 years), Left ureteric re-implantation including Boari flap (19 years)

**Table 1.** Summary of patient characteristics, follow up, presenting symptoms, site of ectopia, associated anomalies and surgical procedures. The procedures are in chronological order (top to bottom), with age at surgery in brackets. AUS: artificial urethral sphincter. BN: bladder neck. UO: ureteric orifice. UTI: urinary tract infections.



**Table 2**

Pt	Ultrasound	Functional imaging	Renal function
1	Right: gross hydronephrosis, Left: mild calyceal dilatation	Right: 10% Left 90%	CKD stage III Creatinine 53 (71) $\mu\text{mol/l}$
2	Bilateral gross hydroureteronephrosis with clubbed calyces and tortuous hydroureters Left: global cortical thinning	Right: 65% Left: 35% Bilateral renal scarring	CKD stage I Creatinine 20 (22) $\mu\text{mol/l}$
3	Bilateral hydroureteronephrosis Bilateral poor corticomedullary differentiation, increased echogenicity	Bilateral renal scarring	CKD stage III Creatinine 46 (227) $\mu\text{mol/l}$
4	Right: prominent upper pole calyx with cortical thinning, echogenic parenchyma, poor corticomedullary differentiation Left: hydroureteronephrosis, cortical thinning	Left: 9% -> 3%	CKD stage II GFR 74ml/min/1.73m <sup>2</sup> , Cr 53 $\mu\text{mol/l}$ at 3yrs Rising creatinine 180 (180) $\mu\text{mol/l}$ Hypertension
5	Right: small non-dilated kidney, echogenic parenchyma with cortical cysts Left: hydroureteronephrosis, normal echogenicity	Right: 9% Left: 91%	CKD stage I GFR 107ml/min/1.73m <sup>2</sup> , Cr 37 $\mu\text{mol/l}$ at 3yrs Creatinine 35 (44) $\mu\text{mol/l}$
6	Right: normal kidney, no hydronephrosis Left: small, echogenic pelvic kidney, dilated ureter	Left: non-functioning	CKD stage II GFR 81ml/min/1.73m <sup>2</sup> , Cr 52 $\mu\text{mol/l}$ at 5yrs GFR 68ml/min/1.73m <sup>2</sup> , Cr 75 $\mu\text{mol/l}$ at 16yrs Creatinine 76 (82) $\mu\text{mol/l}$ Hypertension
7	No hydronephrosis	Right: 75% Left: 25%	CKD stage I Creatinine 54 (96) $\mu\text{mol/l}$
8	Bilateral hydroureteronephrosis Left: echogenic parenchyma, cortical thinning, calyceal dilatation	Right: 10% Left: 90%	CKD stage II GFR 93ml/min/1.73m <sup>2</sup> , Cr 62 $\mu\text{mol/l}$ at 14yrs GFR 78ml/min/1.73m <sup>2</sup> , Cr 75 $\mu\text{mol/l}$ at 17yrs Creatinine 133 (292) $\mu\text{mol/l}$

**Table 2:** Upper urinary tract. Pre-operative ultrasound and functional imaging findings and renal outcomes: most recent serum creatinine is presented in the final column (maximum creatinine in brackets).

**Table 3**

Pt	Age at last FU (years)	Procedures	Preoperative findings			Postoperative findings	
			Cystoscopy	MCUG/IRC	Bladder volume	Bladder volume	Continence outcome
1	1.6	Right open nephro-ureterectomy (7 months) Left ureteric re-implantation (16 months)	No comments on bladder size	N/A	21 mL at 7 months (38% EBCA)	N/A	In nappies at 19 months
2	4	Cystoscopy and formation of bilateral end ureterostomies (10 months) Bilateral cross-trigonal ureteric reimplantation (12 months) Revision of right vesico-ureterostomy (stenosis) (19 months)	Good-sized bladder	Left VUR	14 mL at 3 months (30% EBCA)	50% EBCA 43ml voids at 2.5 years	Continent from age 3yrs
3	4	bilateral cross-trigonal ureteric reimplantation and loose BN repair (15 months)	Moderate-sized smooth bladder, no trigone, wide open BN	Competent BN Left VUR	14 mL at 1 year (20% EBCA)	47ml at 2.5 years (50% EBCA): 30min dry interval	In nappies at 4 years
4	11	Left laparoscopic nephrectomy and right ureteric re-implantation (3.5 years)	Smooth bladder	Competent BN Bilateral grade IV VUR	71ml at 2.5 years (68% EBCA)	190 mL voids at 8 yrs (70% EBCA) UDS at 11yrs: large capacity (450ml, 125%EBCA), stable compliant bladder	Continent
5	8	Right retroperitoneoscopic nephrectomy and left Cohen ureteric reimplantation (5 years)	No trigone, slightly gaping bladder neck	Left VUR, grade III	230ml void at 5yrs on BFA (125% EBCA)	Bladder capacity and emptying maintained	Continent day – Nappies at night
6	20	Left nephrectomy (5 months) Right ureteric reimplant and BN reconstruction (15 months) Left ureteric stumpectomy (4 years) BN injection (7 years)	Moderate-sized smooth bladder, wide open BN and patulous urethra	Left VUR	Not measured – “moderate” at cystoscopy	100% EBCA UDS at 8 years: instability resolved on anticholinergics, reasonable compliance, no leak; stress leak on UDS at 20 years	Occasional mild dampness
7	16	Bilateral ureteric reimplantation and BN tightening (5 years) BN injection (7 years), BN injection (13 years), BN injection (13 years) AUS (15 years)	Small-sized, smooth bladder	Right VUR, grade II	Empty on repeated ultrasounds	50% EBCA 188ml at 12 years; UDS at 10 years: small, stable, compliant bladder, urgency without leak	Continent after AUS
8	20	Right pyeloplasty (4 months) Bilateral ureteric re-implantation (2 years) Right to left TUU (11 years), colposuspension (12 years) Right nephrectomy (13 years), ileocystoplasty & left ureterolysis (17 years) BN closure, redo Mitrofanoff and insertion of stent (18 years) Left ureteric re-implantation including Boari flap (19 years)	Small capacity	Bilateral VUR	“Very small” at surgery	50% EBCA 220 mL at 14 years, daytime urgency and frequency, enuresis	Continent after BN closure

**Table 3:** Lower urinary tract preoperative findings and postoperative outcomes. The age and surgical procedures are repeated for clarity. The expected bladder capacity for age (EBCA) is calculated separately for children aged below 2 years and those above 2 years. Bladder capacity is expressed as a percentage of EBCA (measured volume/EBCA x 100). AUS: artificial urethral sphincter. BN: bladder neck. EBCA: expected bladder capacity for age. MCUG: micturating cystourethrogram. UO: ureteric orifice. UTI: urinary tract infections.

**Table 4**

Series	N (M/ F)	Re-implant alone	Re-implant +YDL BNR	Other
Williams & Lightwood 1992[10]	9 (3/6)		6 (5“fair”, 1“small” bladder) - 2 dry - 2 improved - 1 wet - 1 “small”-> diversion	3 (3“small” bladder) reimplant + BNR + cystoplasty - 1 dry - 2 -> diversion
Ahmed & Barker 1992[17]	3 (2/1)	3 dry		
Jayanthi et al 1997[9]	6 (1/5)	5 - 5 wet -> 4 BNC + cystoplasty		1 – BN sling + ureterocystoplasty - 1 wet
Wakhlu et al 1998[6]	5 (1/4)	5 - 1 dry (male) - 1 dry for 3hrs - 3 wet -> 1 diversion		
Podesta 2001[8]	7 (0/7)	5 (4“good” bladder) - 3 dry - 1 wet - 1 “small” -> BNR	2 (2“small” bladder) - 2 enuresis (1 polyuria)	
Heuser et al 2002[7]	2 (0/2)	2 -2 wet -> 1 AUS		
Choudhury et al 2008[1]	6	5 - wet (3 improved)		1 reimplant with ureterocystoplasty - improved
This series	8 (0/8)	5 (2“good”, 2“moderate”, 1“small” bladder) - 2 dry - 1 enuresis - 1 wet at 1.5yrs - 1 “small”-> ->BNC + cystoplasty	3 (3“small” bladder) - 1 wet at 5yrs - 1 ->BNI - 1 ->BNI->AUS	

**Table 4:** Summary of the main published series of patients with bilateral simplex ureteric ectopia without cloacal, anorectal or spinal anomalies. The first surgical procedures and post-operative outcomes are summarised in columns 3-5. Our series is summarized in the last row for comparison. AUS: Artificial urethral sphincter. BNR: bladder neck reconstruction. BNC: bladder neck closure. BNI: bladder neck injection.