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The single ectopic ureter

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Objectives To correlate renal function with the site of the ectopic orifice in patients with a single ectopic ureter and to evaluate the role of ureteric reimplantation in the preservation of renal function.

Patients and methods Forty-four patients (41 female, age 1.5 months to 20 years) with a single ectopic ureter have been managed in our institution in the last 21 years. The classical symptom of continuous wetting with intermittent normal micturition was reported in most of the female patients. The investigative evaluation included intravenous urography (IVU), cysto-urethroscopy, vaginoscopy with retrograde ureteric catheterization, micturating cysto-urethrography (MCU) and ultrasonography. Diuretic renography was carried out in four patients after it became available in 1992. Renal function was assessed in relation to urinary tract anomalies and with outcome after ureteric re-implantation.

Results Thirty-eight patients (two males) had a unilateral ectopic ureter; the ectopic orifice was vaginal in 12, vestibular in 11, urethral in nine, at the bladder neck in two, the seminal vesicle in one and undetermined in three. Twenty-one patients had renal and/or ureteric abnormalities, with reflux detected on MCU in three ureters. Associated anomalies included hypospadias (two, one female), skeletal anomalies (two), anorectal malformations (three), cryptorchidism (two), and unilateral cystic ovary (one). Two patients had preoperative hypertension. In 15 patients, renal function was considered sufficient to justify ureteric reimplantation, 14 of whom regained continence. One girl had suprapubic leakage from the bladder and died during secondary nephroureterectomy. Another girl had persistent incontinence; she was found to have contralateral duplex ureters with a vestibular ectopic orifice and was cured after upper polar hemi-nephroureterectomy. IVU and renography carried out in two patients each within 4 weeks of surgery showed a moderate improvement in renal function. Eight patients reported for follow-up after ureteric reimplantation (mean duration 11 months); none had hypertension or urinary infection. Twenty-three patients with rudimentary kidneys underwent nephroureterectomy. Histopathological examination of the excised kidneys showed moderate to severe dysplasia with chronic pyelonephritis. Six patients (one male) had bilateral single ectopic ureters, with normal renal function in the five females. Unilateral reimplantation in the boy resolved the symptoms; one girl died before surgery and the other four underwent bilateral ureteric reimplantation, after which one was dry for up to 3 h while the other three were incontinent, one of whom subsequently underwent urinary diversion.

Conclusions There was no clear correlation of renal function with the site of the ectopic ureteric orifice, as most of the patients with a vaginal ectopic ureter had sufficient renal function to justify renal preservation. Ureteric reimplantation preserved renal function, although the improvement after surgery was determined by the degree of renal dysplasia.

Keywords Single ectopic ureter, renal anomaly, reimplantation, outcome

Introduction

An ectopic ureter opening outside the bladder causes continuous urinary dribbling. In the Western hemisphere, most ectopic ureters are one moiety of a duplex system, but in South East Asia, single ectopic ureters are more common. The pathology and clinical features of single ectopic ureter have been well described [1–3] but this condition continues to cause diagnostic confusion and sometimes therapeutic delay [4,5]. In a retrospective study, we determined the association of renal function with the site of the ectopic ureteric orifice, evaluated the efficacy of the investigations used for diagnosis and assessed the role of ureteric reimplantation in the preservation of renal function.

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Patients and methods

The review comprised 44 patients (41 female and three male, age 1.5 months to 20 years); 28 patients presented within the first 3 years of life and the rest afterwards (Table 1). All patients were examined under anaesthesia

Table 1 Age and sex distribution of the 44 patients with ectopic ureter; number of bilateral cases in parentheses

	Age (years)		
Sex	*< 1*	*1–3*	*> 3*
Male	1	–	2 (1)
Female	11 (3)	16 (1)	14 (1)
Total	12	16	16

and by IVU and cysto-urethroscopy. Vaginoscopy was performed in 19 girls, with retrograde catheterization of the ectopic ureteric orifice (Fig. 1). Micturating cysto-urethrography (MCU) was carried out in six patients (one male with bilateral ectopic orifices), vaginography in four (Fig. 2), abdominal ultrasonography in five, and DTPA renography (after it became available in 1992) in four patients before and two after ureteric reimplantation. Percutaneous antegrade pyelography under ultrasonographic control was used to confirm ectopic ureter in the seminal vesicle in one patient. Forty-three of the patients (the exception being one girl with bilateral ectopic orifices) underwent surgery. If the involved renal unit had salvageable function on IVU/DTPA renography, the ureter was reimplanted, and in cases where the kidney was not visualized, nephroureterectomy was carried out. The outcome of surgery and morbidity were recorded, and the patients asked to report for regular follow-up.

Fig. 1. Retrograde catheterization and contrast study of a vaginal ectopic ureter.

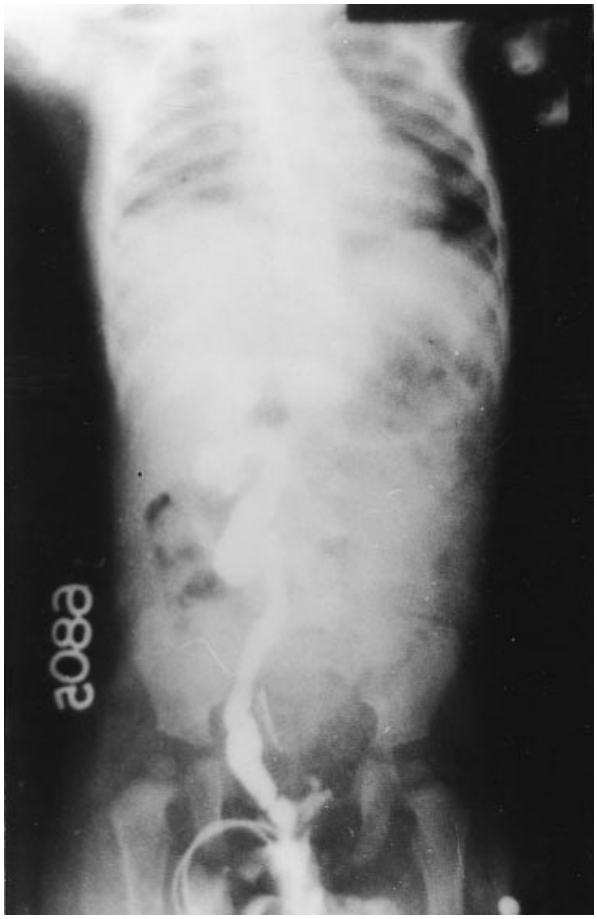

Fig. 2. Ectopic ureter detected by reflux of dye during vaginography.

Results

The patients were categorized into those with a unilateral (38, group I) and those with bilateral single ectopic ureters (six, group II). In group I, 28 girls had a history of continuous wetting since birth, with intermittent normal micturition. In four girls this symptom was noticed after surgery for anovestibular fistula, rectal atresia, common cloaca and female hypospadias, respectively. The parents attributed it to surgical interference, but complained about it when the girls became older and found the continual wetting a nuisance. The mothers were questioned in detail about this symptom; they uniformly reported that on finding a wet patch on the child's pants (usually cotton and coloured) the mother

would place the baby between her feet and the child would then sometimes void with a normal stream. Six patients (two male) presented with recurrent urinary infections. The distribution of the unilateral ectopic orifice is shown in Table 2; 20 were detected during preoperative investigations, 15 at operation by antegrade intubation and the remaining three could not be located at all.

Thirteen patients had bilaterally functioning kidneys on IVU (Fig. 3). Additional findings included hydroureteronephrosis on the ectopic side (six patients) and on the normal side (one), an ectopic kidney (two), ureteric diverticulum (on the ectopic side) and a ureterocele (on the normal side) in one patient each. In 25 patients, the involved kidney was not visualized on IVU. MCU showed reflux in two ureters (one ectopic and one contralateral normal ureter); in one girl, the catheter directly entered the ectopic ureteric orifice in the urethra and injection of contrast medium delineated the ectopic ureter. Abdominal ultrasonography identified a retrovesical dilated segment of ureter with hydronephrosis in the oldest male patient; percutaneous antegrade pyelography confirmed the diagnosis of ectopic ureter terminating in the seminal vesicle (Fig. 4). Urine culture showed *Escherichia coli* in two and *Streptococcus faecalis* in one patient. Associated anomalies in these patients included hypospadias in two (one female), thoracolumbar scoliosis (two), lumbarization of S1 (one), anovestibular fistula, rectal atresia and common cloaca (one patient each), cryptorchidism (two) and unilateral cystic ovary (one). Two patients had preoperative hypertension.

All children in group I were surgically explored: the findings included ipsilateral hydronephrosis (seven patients), rudimentary kidneys (21), ectopic kidney (two), pyonephrosis (one) and horseshoe kidney with ectopic orifice of the right ureter to the urethra in a patient who had undergone previous surgery for common cloaca. The ureteric abnormalities in 19 patients (17 ipsilateral and one contralateral ureter) included dilatation in 17, ureteric diverticulum in one and contralateral ureterocele in one.

Fig. 3. IVU showing renal function in the left kidney with a vaginal ectopic ureter.

Table 2 Sites of the ectopic orifice and the management in patients with unilateral ectopia

	Side			*Management*	
Site	*Right*	*Left*	*Total*	*Reimplanted*	*NU*
Vagina	7	5	12	8	4
Vestibule	6	5	11	1	10
Urethra	4	5	9	5	4
Bladder neck	1	1	2	1	1
Seminal vesicle	–	1	1	–	1
Unknown	–	3	3	–	3
Total	17	20	37	15	22

NU, nephroureterectomy.

Twenty-three patients underwent nephroureterectomy, with the ureter ligated as distally as possible. All the excised kidneys were 'nubbins' of dysplastic tissue (in one instance the rudimentary right half of a horseshoe kidney was removed) and the renal vessels were difficult to differentiate. All these children were relieved of their symptoms after operation. Histopathological examination of 16 excised kidneys showed moderate to severe dysplasia with chronic pyelonephritis.

Fifteen patients underwent combined extravesical and intravesical ureteric reimplantation; 13 had functioning kidneys on IVU and two had salvageable renal function on DTPA renography. In one child the contralateral ureter was also reimplanted after excision of a ureterocele. After surgery, 13 patients were relieved of their symptoms (four required urinary antiseptics for up to 6 weeks to treat UTIs). One girl had wound dehiscence and persistent urinary leakage from the bladder after

Fig. 4. Antegrade pyelography showing a left seminal vesicle ectopic orifice.

ureteric reimplantation; she subsequently underwent nephroureterectomy but died during the second operation. Another girl had persistent symptoms after left ureteric reimplantation; she was found to have a contralateral duplex system with the upper pole ureter terminating in the vestibule. She was cured after right upper pole heminephroureterectomy.

Among the six patients in group II with bilateral ectopic orifices (Table 3), five girls presented with urinary incontinence; the single boy presented with recurrent urinary infection. IVU showed bilaterally functioning kidneys in all these patients, with an ectopic orifice and axial rotation of the right kidney in the boy. MCU in this boy showed reflux into an ectopic ureteric orifice in a posterior urethral diverticulum. On cystoscopy, all patients had no trigone and a wide bladder neck; the two females also had a lax urethra. The urethral ectopic orifice was reimplanted in the boy, with excision of the urethral diverticulum; the other ectopic ureter proximal to the bladder neck was not displaced. Of the girls, one died before surgery and the other four underwent bilateral ureteric reimplantation. One child was partially relieved of her symptoms while the others were still incontinent. Bladder neck repair was declined by the relatives.

Table 3 Sites of ectopic orifice and management in bilateral ectopic ureters

Patient no.	*Right*	*Left*	*Management*
1	Vagina	Urethra	Bilat reimplant
2	Bladder neck	Urethra	Bilat reimplant, subsequent rectal bladder
3 (boy)	Above bladder neck	Urethra	Left reimplant
4	Urethra	Vestibule	Bilat reimplant
5	Vagina	Vestibule	Bilat reimplant
6	Vagina	Vestibule	Died before surgery

After nephroureterectomy, 13 children were followed for up to 5 years; all had normal bladder function. Of those undergoing reimplantation, eight were followed for up to 2 years; none had hypertension or urinary infection. IVU in two of these patients showed acceptable renal function and DTPA renography in two others showed a slight improvement. Among the patients with bilateral ectopic orifices, one subsequently underwent construction of a rectal bladder and another three girls remain only partially dry during the day and wet at night, but have refused further surgery.

Discussion

Overall, the ectopic ureter is commoner in males, but the single ectopic ureter shows racial differences; in contrast to reports from the Western hemisphere [2,3], the present series showed a marked preponderance in females, similar to that reported from Japan [6]. Most of the females had the classical symptom of continuous wetting with intermittent normal micturition [5]. We believe that this symptom complex is characteristic of the single ectopic ureter in girls and should cause no doubts about unilateral renal agenesis [4,6]. The other racial difference seems to be in the overall higher incidence of ureteric ectopia in a single system in India, for reasons unknown. Thus, during the period of this study (30 years) only two cases of ureteric ectopia were found in a duplex system in girls. However, it is likely that some boys might have been undetected because the diagnosis was not suspected.

Several investigations, including videolaparoscopy, have been used to locate the site of the ectopic orifice and the involved kidney by previous investigators [6–12]. We feel that IVU, cysto-urethroscopy and vaginal endoscopy with retrograde catheterization should be carried out in every patient suspected of having an ectopic ureter. Any kidney which does not have sufficient

function to be visualized on IVU will not excrete indigocarmine or phenazopyridine, and any ureteric orifice too small to be visualized on endoscopy will not allow reflux on MCU and vaginography. If the ipsilateral ureteric orifice is not detected in the trigone by cystoscopy, radionuclide scanning should be performed to identify the involved kidney and quantitatively assess its function [11–14]. When endoscopy, IVU and renography fail to locate the ectopic ureteric orifice/involved kidney, exploration is indicated; with this approach, any undue therapeutic delay will be avoided.

Of the present patients with a vaginal ectopic orifice, three-quarters had adequate renal function on IVU and/or renography [3,5,15], and were treated by ureteric reimplantation (Table 2). This incidence of renal function in vaginal ectopic ureter is higher than reported from the Western hemisphere [16] and closer to that reported from Japan [6]. Most patients with urethral and one with a vestibular ectopic orifice also had sufficient renal function for visualization on IVU. Thus the level of renal function in patients with a single ectopic ureter does not seem to correlate with the site of the ectopic orifice, probably because the renal dysplasia in those with a vaginal ectopic orifice is not proportional to the degree of displacement of the ectopic orifice [13]. Each kidney should therefore be assessed, despite the uniform presence of embryological abnormalities associated with the ectopic ureter.

The patients undergoing ureteric reimplantation in the present series had improved renal function and none developed hypertension or urinary infection. The degree of improvement in renal function was not as marked as that reported by Ahmed and Barker [17], because all their patients undergoing ureteric reimplantation had a urethral ectopic orifice with less associated renal dysplasia.

Ultrasound-guided antegrade pyelography was used to confirm the presence of an ectopic ureter terminating in the seminal vesicle in one patient; this investigation is more convenient than some of the other tests reported for this particular type of ectopic ureter [18].

The occurrence of both single and duplex ectopic orifices in the same patient is unique. To the best of our knowledge, this is the only such patient reported in the English literature. Unfortunately, the condition was not initially recognized, leading to multiple operations and a long period of investigation and treatment for this girl; however, she was finally cured of her problem.

Bilateral ureteric ectopic orifice is more challenging, especially in females where the absence of both ureteric orifices from the bladder leads to a small bladder and a poor chance of postoperative continence [17]. One of the present patients required urinary diversion after reimplantation and three others are still incontinent. Recent papers have reported better results in the treatment of the difficult problem of bilateral single ectopic orifices. The peculiar feature in the present patients with bilateral single ectopic ureters was the uniformly asymmetric sites of the ectopic orifice, which is at marked variance from previous reports [19–22]. Males usually have both ureteric orifices above the verumontanum and females in the urethra.

The present results show that renal function in patients with a single ectopic ureter does not correlate with the site of the ectopic ureteric orifice; definitive management must therefore be based on quantitative renal function, best assessed by renography. Extensive investigation to locate an elusive ureteric orifice can cause therapeutic delay that can be obviated by flank exploration. Ureteric reimplantation is of benefit when the involved kidney has $>15\%$ of overall function.

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