

## DIAGNOSIS OF UNILATERAL SINGLE SYSTEM ECTOPIC URETER IN GIRLS IN THE ERA OF MAGNETIC RESONANCE UROGRAPHY (MRU)

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**Objective:** The ectopic ureter frequently drains an ectopic dysplastic or hypoplastic kidney. The present study aims at defining the role of MRU in establishing the diagnosis of this anomaly.

**Patients and Methods:** Between February 1996 and March 2000, 11 girls presented or were referred to our department for management of urinary incontinence. Their age at presentation ranged from 4-9 years (mean 6.5 years). Radiological work up included abdominal ultrasound (US), excretory urogram (IVU), voiding cystourethrography (VCUG), <sup>99m</sup>Tc-DMSA renal scan, enhanced spiral computed tomography (CT) and magnetic resonance urography (MRU).

**Results:** Ultrasound showed evidence of a solitary kidney with failure to visualize a contralateral kidney in 7/11 patients. In the remaining 4 patients (36.4%), US revealed a pelvic kidney in two and a pelvic cystic mass in another two patients. IVU revealed only one functioning renal unit in all cases. None of the patients showed vesicoureteral reflux on VCUG. On <sup>99m</sup>Tc-DMSA, a single

kidney was seen in 9/11 patients and ectopic pelvic kidneys with normal contralateral kidneys in 2/11 patients. The 7 patients, in whom US and <sup>99m</sup>Tc-DMSA scan had failed to localize the kidney, underwent CT scanning which visualized a pelvic hypoplastic kidney with a normal contralateral kidney in 2/7 patients. The remaining 5 patients underwent MRU that disclosed a normal kidney with a contralateral lumbar hypoplastic kidney in one and a pelvic ectopic kidney in four. The patients were managed by nephrectomy (n=9) and ureteroneocystostomy (n=2).

**Conclusions:** A single system ectopic ureter should be suspected in all girls with continuous urinary dribbling after the age of successful toilet training. With the inclusion of MRU into radiological workup, dysplastic or hypoplastic kidneys can be accurately localized. MRU is indicated for the diagnosis and for therapeutic planning in such cases.

**Key words:** urinary incontinence, ectopic ureter, MRU

### INTRODUCTION

The term "ectopic ureter" has universally been used to describe a ureter that terminates at the bladder neck distally in one of the mesonephric duct structures or that is incorporated into any of the nearby Mullerian duct structures such as the vagina, uterus and cervix in females<sup>1</sup>. A renal unit parenchyma drained by an ectopic ureter is difficult to locate and may be identified only by alternate imaging studies.<sup>2</sup> A single system ectopic ureter in girls is a rare anomaly and difficult to diagnose.<sup>3</sup> MRU is a relatively new imaging modality which seems

to have some advantages over existing modalities like multiplanar imaging capabilities and absence of ionizing radiation.<sup>4</sup>

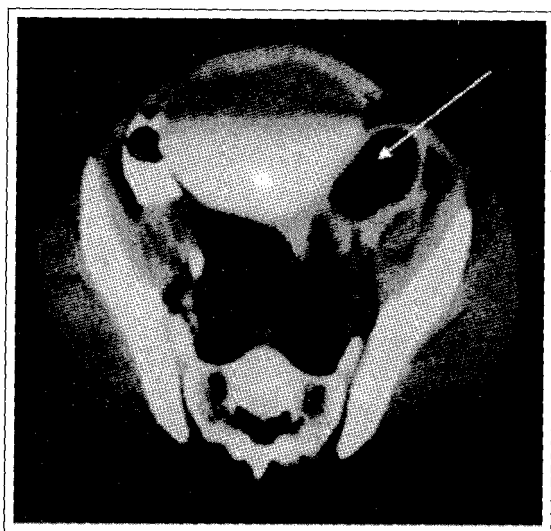
This study aims at defining the role of MRU in establishing the diagnosis of single system ectopic ureter.

### PATIENTS AND METHODS

Between February 1996 and March 2000, four girls presented and seven were referred (total: 11 girls) to the Urology Department of

**Table 1:** Data of Radiological Work-Up and Management of Renal Units (RU) Drained by a Single System Ectopic Ureter

No.	IVU	US	Tc-DMSA	CT	MRU	Management
1	non-visualized	pelvic RU	pelvic RU	not done	not done	ureteroneocystostomy
2	non-visualized	pelvic RU	pelvic RU	not done	not done	ureteroneocystostomy
3	non-visualized	pelvic cystic	absent	not done	not done	nephrectomy
4	non-visualized	pelvic cystic	absent	not done	not done	nephrectomy
5	non-visualized	absent	absent	pelvic hypoplastic	not done	nephrectomy
6	non-visualized	absent	absent	pelvic hypoplastic	not done	nephrectomy
7	non-visualized	absent	absent	absent	lumbar hypoplastic	nephrectomy
8	non-visualized	absent	absent	absent	pelvic hypoplastic	nephrectomy
9	non-visualized	absent	absent	absent	pelvic hypoplastic	nephrectomy
10	non-visualized	absent	absent	absent	pelvic hypoplastic	nephrectomy
11	non-visualized	absent	absent	absent	pelvic hypoplastic	nephrectomy



**Fig. 1:** Enhanced CT of the pelvis of a 5-year-old girl with ectopic ureter showing a hypoplastic pelvic left kidney (arrow)

Assiut University Hospital, Assiut, Egypt, for management of continuous urinary dribbling after the age of toilet training. The patients' age at presentation ranged from 4-9 years (mean 6.5). Radiological work up included abdominal ultrasound (US), excretory urogram (IVU), voiding cystourethrography (VCUG), <sup>99m</sup>Tc-DMSA renal scan, enhanced spiral computed tomography (CT) and magnetic resonance urography (MRU). CT was done for patients in

whom US, IVU and DMSA scan had failed to localize the kidney. MRU was resorted to in cases of failure of CT to localize the kidney. The preparation for MRU included four hours fasting (food and fluid) and intravenous hydration followed by 0.3-0.5 mg/kg IV furosemide 20 minutes prior to imaging. The procedure was carried out under general anesthesia. MRU (conventional sequence) was done using T1 and T2-weighted sequences. MRU was performed using heavy T2 weighted sequence images with a fat suppression pulse that allowed reduction in the signal intensity of retroperitoneal fat. Images were taken both in the coronal and sagittal planes of the abdomen and pelvis. The procedure took about 30 minutes. Surgical management was performed according to the imaging findings confirmed by surgical exploration data.

## RESULTS

US showed evidence of a solitary kidney with failure to visualize a contralateral kidney in 7/11 patients (63.6%). In the remaining four (36.4%), US revealed a pelvic cystic mass and pelvic kidney in two patients each. IVU showed a unilateral non-visualized renal unit in all cases. None of the patients showed vesicoureteral reflux on VCUG. On DMSA scan, a single kidney could be seen in 9/11 patients (81.8%) and ectopic pelvic kidneys with normal contralateral kidneys in the remaining two patients (18.7%). The seven patients in whom US



Fig. 2: MRU in a 6-year old girl with ectopic ureter showing a hypoplastic pelvic left kidney (<<<) and its draining ureter (arrow)

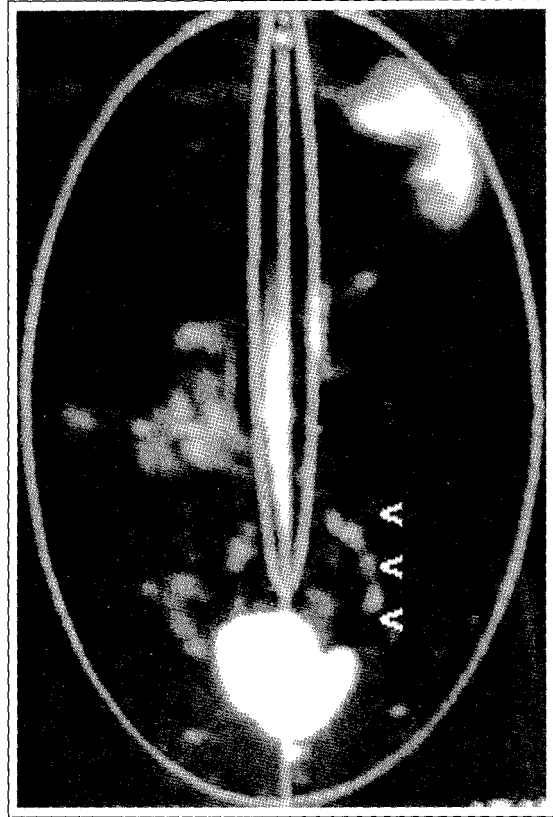


Fig. 3: MRU in a 9-year-old girl with ectopic ureter showing a hypoplastic pelvic left kidney (<<<)

and DMSA scan had failed to localize the suspected ectopic ureter underwent CT which visualized a pelvic hypoplastic kidney (Fig.1) with a normal contralateral kidney in two patients (28.6%). The remaining five patients underwent MRU that disclosed a normal kidney with a contralateral lumbar hypoplastic kidney in one and pelvic ectopic kidney in four (Fig.2, 3). MRU defined all the renal units and their draining ectopic ureters. It showed the ureters throughout their course and demonstrated that they actually ended ectopically (Table 1).

Examination under anesthesia revealed no visualized orifice of the ectopic ureter but a damp vestibule in all cases. On surgical exploration the site of the ectopic ureteric opening was found to be in the distal urethra and in the vestibule in six and five cases, respectively. The patients were managed by nephrectomy in nine cases, and by ureteroneocystostomy in two patients in whom the kidney could accumulate the radiotracer (Table 1). All patients

gained urinary continence and were symptom free after surgery.

### DISCUSSION

Ectopic ureters clinically appear between 2 and 12 times more commonly in females than in males<sup>5</sup>. In the female, the urethra and vestibule are the most common sites<sup>3</sup>. The ectopic ureter frequently drains an ectopic dysplastic or hypoplastic kidney. More than 80% of ectopic ureters in females drain a duplicated collecting system. A small percentage involves a solitary kidney.<sup>6</sup>

An ectopic ureter draining a single system renal unit is a rare anomaly, especially in females. The age at diagnosis ranges widely.<sup>5</sup> Incontinence of urine in girls with an otherwise normal voiding pattern after the age of toilet training is the classic symptom of an ectopic ureteral orifice.<sup>1</sup> It has been termed "paradoxi-

cal incontinence<sup>7</sup>. Physical examination may be helpful for the diagnosis of an ectopic ureter.<sup>3</sup>

Some radiological modalities like US, IVU<sup>8</sup>, DMSA scanning<sup>9,10</sup> and CT<sup>11</sup> may have a role in the diagnosis of an ectopic ureter draining a duplicated system renal unit. The results of the present study support the conclusion that a combination of life-long wetting and solitary kidney on IVU should draw the urologist's attention to a diagnosis of ectopic ureter.<sup>8</sup> A renal parenchyma associated with an ectopic ureter is often thinner than that of a normally draining lower pole.<sup>12</sup> The reduced renal function of a renal unit drained by an ectopic ureter may result from primary dysplasia, obstruction, vesicoureteral reflux or recurrent infection.<sup>13</sup> It is hard to accept the claims about the superiority of renal scintigraphy for the detection of either hypoplastic ectopic or poorly functioning kidneys as a fact based on a limited experience in few cases<sup>9,10</sup>. Handling and excretion of any renal radiotracer depends on numerous factors including the functional status of the kidneys<sup>14</sup>. MRU does not require a functioning kidney to depict its collecting system and ureter - small amounts of urine in the urinary tract are sufficient to be visualized by MRU.<sup>4</sup> In a study carried out by Tang et al., MRU was reported to be valuable in revealing non functioning urinary tract units that were invisible on excretory urography<sup>15</sup>.

MRU may possibly be used as a primary diagnostic method for ectopic ureters when they are not detected by standard imaging techniques, or as a non-invasive substitute for some interventional studies in infants. MRU has been reported to provide a high accuracy in the evaluation of suspected ureters of duplicated collecting systems.<sup>16</sup> Radiological work-up in the form of US, IVU, isotope scanning and CT are no reliable methods for the diagnosis of single system ectopic ureters draining poorly functioning renal units, such as hypoplastic or dysplastic kidneys. With the addition of MRU, all these dysplastic or hypoplastic kidneys can be accurately localized. At the same time, MRU defines either the normal or the ectopic termination of a draining ureter. In this issue, MRU is superior to CT. The consensus is that in cases of suspected ectopic ureters in the presence of solitary kidneys which are non-visualized by US, IVU or CT, MRU should be recommended. This radiological modality can provide an accurate plan for the management of ectopic ureters, bearing in

mind that it lacks function assessment. Radioisotope scintigraphy may be helpful for the functional evaluation of a renal unit drained by an ectopic ureter.

In conclusion, MRU can detect dysplastic or hypoplastic kidneys drained by a single system ectopic ureter. Being superior to other radiological modalities in this issue, it should be the gold standard non-invasive imaging modality for the diagnosis and planning of the management of this rare anomaly.

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## RESUME

### Le diagnostic de l'ectopie urétérale unilatérale chez les filles à l'ère de l'urographie par résonance magnétique (MRU)

**Objectifs:** Définir le rôle de l' MRU dans le diagnostic de l'ectopie urétérale unilatérale. **Patientes et Méthodes:** Entre février 1996 et mars 2000, 11 filles se sont présentées ou ont été référées à notre hôpital pour incontinence urinaire. Leur âge à la première consultation était de 4 à 9 ans (moyenne 6,5). Le bilan radiologique réalisé a inclu une ultrasonographie abdominale (USA), une urographie (IVU), une cysto-urethrographie (VCUG), une scintigraphie rénale à l'acide 99 m technetium-dimercaptosuccinique (<sup>99m</sup>Tc-DMSA), une TDM spiralée rehaussée (CT) et une urographie par résonance magnétique (MRU). **Résultats:** L'ultrason a montré un rein unique avec absence de visualisation du rein controlatéral chez 7 des 11 patientes. Chez les 4 patientes restantes (36.4%), l'échographie a révélé un rein pelvien dans deux cas et une masse kystique pelvienne chez deux patientes. L'UIV a montré un rein unique fonctionnel chez toutes les patientes. Aucun cas de reflux du vesico-urétéral n'a été mis en évidence sur les VCUG. 99m Tc-DMSA a révélé un rein unique chez 9 des 11 patientes et un rein ectopique pelvien avec le rein controlatéral normal chez 2/11 patientes. Les 7/11 patientes dans lesquelles l'USA et la scintigraphie au <sup>99m</sup>Tc-DMSA avaient manqué de localiser le rein ont subi une TDM qui a visualisé un rein hypoplasique pelvien avec un rein controlatéral normal chez 2 patientes. Les cinq patientes restantes ont subi une MRU qui a montré un rein normal avec un rein controlatéral hypoplastique lombaire dans un cas et un rein ectopique pelvien chez 4 patientes. Les malades ont subi une néphrectomie dans 9 cas et une urétéroecystostomie chez 2 patientes. **Conclusion:** Un uretère ectopique unilatéral devrait être suspecté chez toutes les filles avec incontinence urinaire continu après l'âge d'acquisition de la propreté. Avec l'avènement de la MRU tout rein dysplasique ou hypoplasique peut être localisé correctement. MRU est indiqué pour le diagnostic et pour l'indication de l'attitude thérapeutique de cette anomalie.

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