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Incontinence due to an infrasphincteric ectopic ureter: why the delay in diagnosis and what the radiologist can do about it

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Abstract *Purpose.* To determine (1) the reasons for the frequently long delay in the diagnosis of an infrasphincteric ectopic ureter in girls, and (2) what role the radiologist can play in decreasing the delay. *Materials and methods.* Twelve girls were referred to our hospital from June 1994 until April 1997 for evaluation of constant urinary dribbling and/or vaginal discharge. Available imaging studies, radiology reports, and clinic notes were reviewed. *Results.* Mean age at the time of diagnosis was 6 years 7 months (range 2 years 10 months to 11 years 11 months). Mean delay until diagnosis after presentation was 2 years 5 months. Excluding the one girl whose ectopic ureter was diagnosed while she was still in diapers, mean age at the time of the first parental "complaint" was 4 years 9 months. The significance of the classic history of constant urinary dribbling was not recognized by physicians in 7 girls for 4 months to 7 years 10 months after presentation. Physical exam was not meticulously performed, as the ectopic orifice was visible in 8 of 12 girls. Imaging studies were ineffectively utilized: no imaging was done (for 2 years in 2 girls), inappropriate studies were done (ultrasound and voiding cystourethrography) and were misleading, studies were called normal when they were not (ultrasound and excretory urography), or perinatal imaging led to the incorrect assumption of a congenitally absent kidney in one girl and a multicystic dysplastic kidney in another. Excretory urography (EU) was diagnostic in all 10 girls with a duplex kidney, and computed tomography (CT) was supportive in 2 with a dysplastic kidney. CT was an adjunct in 3 girls; a Tc-99m-dimercaptosuccinic acid (DMSA) scan was needed in 2. *Conclusion.* The classic history of constant urinary dribbling in a successfully toilet-trained girl should immediately lead to an imaging search for the portion of kidney (or entire kidney) drained by an infrasphincteric ectopic ureter. EU should usually be the first imaging performed and is often the only imaging study needed.

Introduction

A constant dribble of urine, every day and every night for as long as anyone can remember, in a girl who has been successfully toilet-trained is the characteristic story of a young girl who has a ureter that drains ectopically outside the bladder and below the sphincter [1–5]. This ectopic ureter usually carries urine from the upper pole of a duplex kidney but occasionally drains a kidney with a single collecting system [1, 2, 6]. The suspected diagnosis is usually easily confirmed by excretory urography (EU), which also shows the affected side or sides and assesses the function of the abnormal moiety or kidney. Unfortunately, young girls are still being treated

with drugs, psychotherapy, behavioral modification, and bladder control exercises, sometimes for years, before the correct diagnosis is made and surgery finally brings relief. This paper is a reminder that when a young, successfully toilet-trained girl presents for imaging with constant day and night wetness, an ectopic ureter must be the primary diagnostic consideration. An excretory urogram should be the first and will usually be the only imaging test she will need.

Materials and methods

From June 1, 1994 to April 20, 1997, 12 girls were referred to our hospital for evaluation of urinary dribbling and/or vaginal discharge. The available imaging studies, radiology reports, and clinic notes from other hospitals were reviewed along with our hospital's medical records and imaging studies. The parents of two girls with incomplete records were contacted by telephone to supply additional information.

Results

The 12 girls ranged in age from 2 years 10 months to 11 years 11 months; the mean age was 6 years 7 months. The mean time from either their first urinary imaging evaluation or the first parental complaint to the time of diagnosis was 2 years 5 months (range 1 week to 8 years 1 month). In six girls, the time to diagnosis was less than four months; in the other six, the mean time to diagnosis was 4 years 9 months.

In 11 girls, the history was of constant urinary dribbling despite normal toilet training. In one of these 11, the history was obscured by neglect, sexual abuse, and residence in foster homes. The urinary dribbling was not evaluated until she developed vaginal discharge associated with a urinary tract infection at age 6 years 3 months. The 12th patient, age 2 years 10 months, presented with vaginal discharge. She was in diapers and so her urinary dribbling had not yet been recognized.

Physical examination showed the orifice of the ectopic ureter in eight girls. It was just inferior and lateral to the ipsilateral side of the urethral meatus in seven of these eight. In the remaining four girls, the ureter inserted higher in the vagina and was not visible on physical exam.

In ten of the girls, the ectopic ureter drained the upper pole of a duplex kidney, five on the left and five on the right. A normal duplex kidney was present on the other side in six of these ten. The 11th girl had a small dysplastic ectopic right kidney with a single collecting system and a ureter that drained into the vagina. The 12th girl was followed for what was believed prenatally to be a multicystic dysplastic kidney. Later, this dysplastic kidney with a single collecting system, lying in the lower part of the left renal fossa, was found to be functioning and draining urine into the vagina.

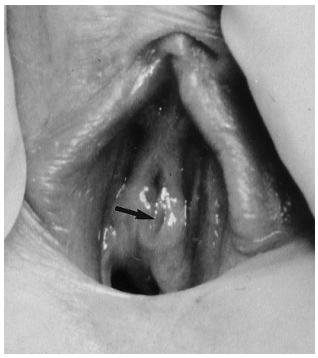

Fig. 1 The ectopic orifice (of the upper pole ureter of the left duplex kidney) is just inferior and slightly lateral *(arrow)* to the urethral meatus

The reasons for the delay in diagnosis were many. First, parents and caregivers were often slow to realize that their child's incontinence was abnormal. Excluding the one girl whose ectopic ureter was discovered while she was still in diapers, the mean age of the child at the time of the first parental complaint was 4 years 9 months (range 3 years 1 month to 7 years 2 months). Second, the classic history of constant urinary dribbling, which was present in all of the girls except for the one who was still in diapers, was not recognized as significant by the pediatrician or family doctor in seven girls for between 4 months and almost 8 years. Three of these girls had an almost two-year delay because no imaging was done after the initial complaint, but once imaging was done, the diagnosis was quickly made. Third, the ectopic orifice was not recognized on physical examination. Of the eight girls with visible ectopic orifices (Fig. 1), their mean age at diagnosis was 6 years 8 months. Fourth, and of most importance to radiologists, imaging studies were done but were misinterpreted. The most common imaging-related reason for delay in diagnosis was calling an abnormal ultrasound (US) or EU normal. In two girls who were thought to have a solitary functioning kidney, the possibility that another kidney was present and might be responsible for the wetting was never entertained.

Imaging studies

Ultrasonography. Ultrasonography was performed in nine patients prior to EU. The upper pole calyx or calyces drained by the ectopic ureter were dilated in three of nine; in another, the proximal ureter was dilated. The diagnosis was correctly suggested in three of these four.

In the other five, either the ultrasound exam was normal or only subtle abnormalities were present and were not seen or were incorrectly identified (Fig. 2). One US showed normal bilateral duplex kidneys. Two girls had a small duplex kidney (not recognized as such) with an eccentric sinus (the lower pole sinus), and a larger normal contralateral duplex kidney. Another girl had a

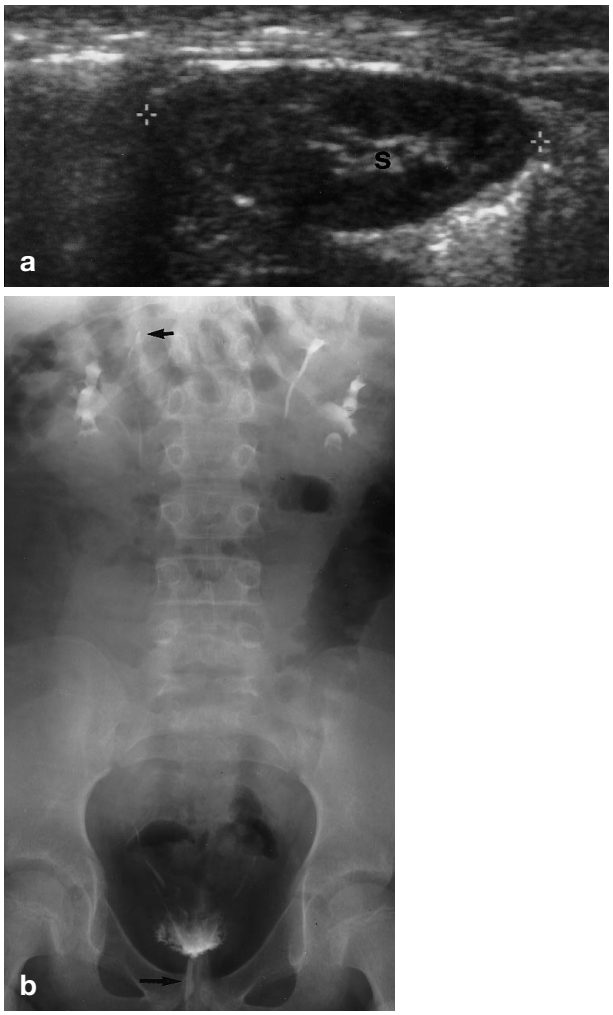

Fig. 2a, b Abnormal US called normal; therefore, EU not done. Two-year-old girl hospitalized for pyelonephritis. **a** Ultrasound showed a right kidney that was normal except for an eccentric sinus *(s)*. There was a normal left duplex kidney. VCUG showed grade 2 reflux on the left into a bifid ureter. Several more ultrasound exams were done and were called normal. Reflux resolved after three years. It was noticed that she constantly dribbled urine, but this was attributed to "voiding dysfunction related to vesicoureteric reflux" even after the reflux had resolved. Treatment of wetting included behavioral modification and prescription drugs from age 3 years 10 months until age 9 years 6 months, when a pediatric urologist was consulted. **b** EU showed a normal left duplex kidney. There was a poorly functioning right upper pole with a non-dilated upper pole calyx *(upper arrow)* and ureter. This ureter could be seen to drain below the base of the bladder *(lower arrow)*. She underwent right upper-to-lower ureteroureterostomy and has been dry ever since

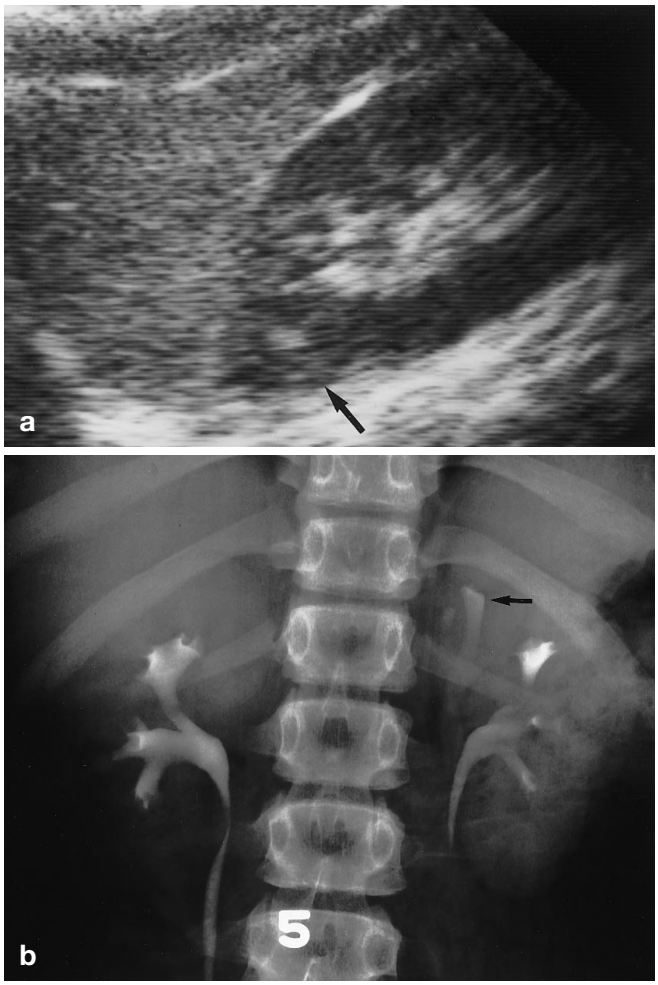

Fig. 3a, b Abnormal US called normal. This girl presented at age 4 years with constant dribbling of urine in the day and bedwetting at night. **a** US was interpreted incorrectly as normal with "no evidence of a duplex kidney." In retrospect, the small left upper pole *(arrow)*, visible on each image, may have been mistaken for the adrenal gland. Perineal exercises were prescribed. She was followed for eight years and was always wet. **b** At age 12, an EU revealed a duplex left kidney with good function of the upper pole *(arrow)*. Physical examination showed the ectopic ureteral orifice just to the left of the urethral meatus. She had left common-sheath reimplantation and has been dry, for the first time in her life, ever since

very small upper pole, perhaps misinterpreted as a normal adrenal gland (Fig. 3). A fifth had a small nondilated upper pole, misinterpreted as an adrenal mass (Fig. 4).

Excretory urography. In only one girl was EU the first imaging study done. In the others, it was the second to the ninth study. The excretory urogram was abnormal and suggestive of an ectopic ureter by either direct or indirect signs in all. Although well described in the radiology literature [2, 3, 7], these signs were not always appreciated. EU was called normal in only one girl (Fig. 5); the abnormal axis and the increased distance from the lower pole ureter to the spine were not recognized.

Contrast was seen in the upper pole collecting system on the affected side in all ten who had a duplex kidney.

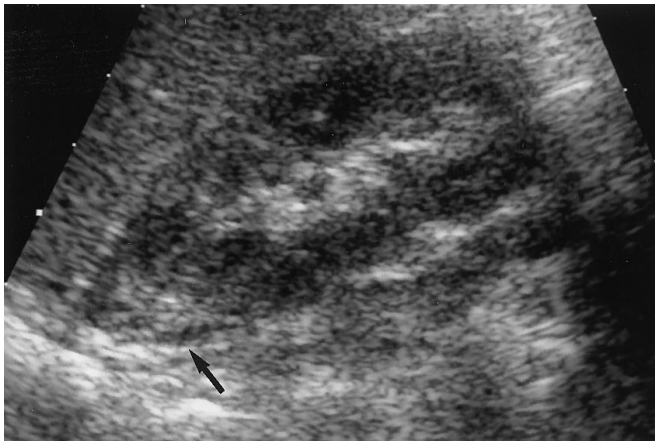

Fig. 4 Upper pole called adrenal mass. US in 2 year, 10-month-old girl with vaginal discharge shows right duplex kidney that was mistakenly called a normal single-system kidney. The upper pole of the kidney, drained by an ectopic ureter, was mistakenly called an adrenal mass *(arrow)*. This led to a CT scan (not shown) that correctly identified the upper pole of the right duplex kidney

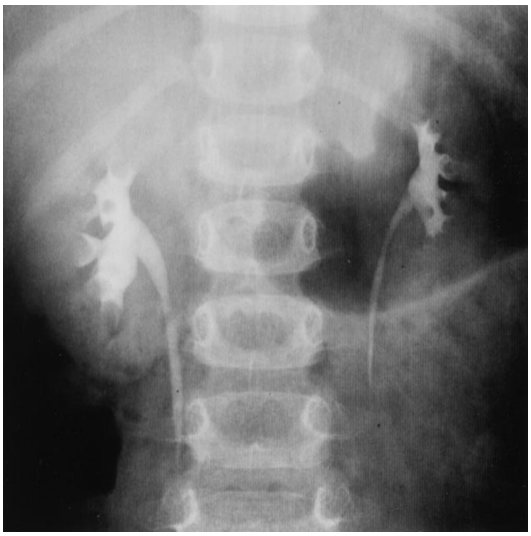

Fig. 5 Abnormal EU called normal. At age 3 years 3 months, this girl was evaluated for "persistent urine dripping after normal bladder training." EU performed elsewhere was incorrectly interpreted as normal. The left lateral deviation of the left lower pole ureter relative to the normal position of the contralateral single ureter, and the vertical orientation of the well-opacified lower calyces were not recognized. VCUG done at the same time was indeed normal. One month later, US at our hospital showed a normal right kidney and a duplex left kidney with a dysplastic upper pole and a dilated ureter that inserted into the vagina. This ureter was reimplanted into the bladder and the girl was dry for the first time in her life

In two, it was very faint. In one of these, a CT scan was performed to confirm the presence of the dysplastic upper pole (Fig. 6).

In six girls, a normal contralateral duplex kidney was seen. In those six, on the abnormal side, contrast was seen in a small upper pole collecting system surrounded by a relatively thin rim of renal tissue, compared to the normal contralateral upper pole.

The ureter that drained the lower pole of the affected side was displaced laterally when compared to the single contralateral ureter in three of the four girls without bilateral duplication. In the six with a normal contralateral duplex kidney, the two lower pole ureters were equally distant from the spine. In the seven with very tiny upper poles, the axis of the lower pole calyces was more vertically oriented than normal (Fig. 5).

In two of the 12 girls, contrast was actually seen in the ectopic ureter below the base of the urinary bladder (Fig. 2). In one of these, contrast was also faintly seen in the vagina on a pre-void film.

A hypertrophied "solitary" kidney was seen in two girls; the contralateral dysplastic kidney was not urographically evident.

Discussion

The diagnosis of an extravesical infrasphincteric ectopic ureter in girls who are *always* wet, despite normal toilet training and normal voiding, is still often delayed, sometimes for years. The history is usually classic and should lead to a relentless visual search for the ectopic ureteral orifice and imaging search for the upper pole of the kidney (or entire kidney) that it drains. The diagnosis can usually made by history and physical exam, and confirmed with EU. EU also shows the side or sides affected and reveals the size and degree of function of the culprit, whether it be the upper pole of the duplex kidney or a kidney with a single collecting system.

Contrast-enhanced computed tomography may help when EU is normal or nearly so and an occult upper pole needs to be located or confirmed (Fig. 6) [3, 8]. A Tc-99m-dimercaptosuccinic acid (DMSA) scan is usually needed to identify an occult, ectopic, small dysplastic kidney with a single collecting system (Fig. 7 a) [9, 10], and then a focused, contrast-enhanced CT examination is used to locate the kidney more precisely before surgery (Fig. 7 b).

In ureteral duplication, the underlying abnormality is the development of a second ureteral bud, cephalad to the normal bud, from the mesonephric (Wolffian) duct. This ureter subsequently inserts lower than normal at the corner of the bladder trigone [4, 11]. A ureter that inserts anywhere else is considered ectopic. However, the ectopic orifice in girls who are always wet is extravesical and infrasphincteric. Only in females does the ectopic orifice insert below the urethral sphincter. Girls with infrasphincteric ureteral orifices are incontinent of urine as long as the upper pole (or kidney) makes urine.

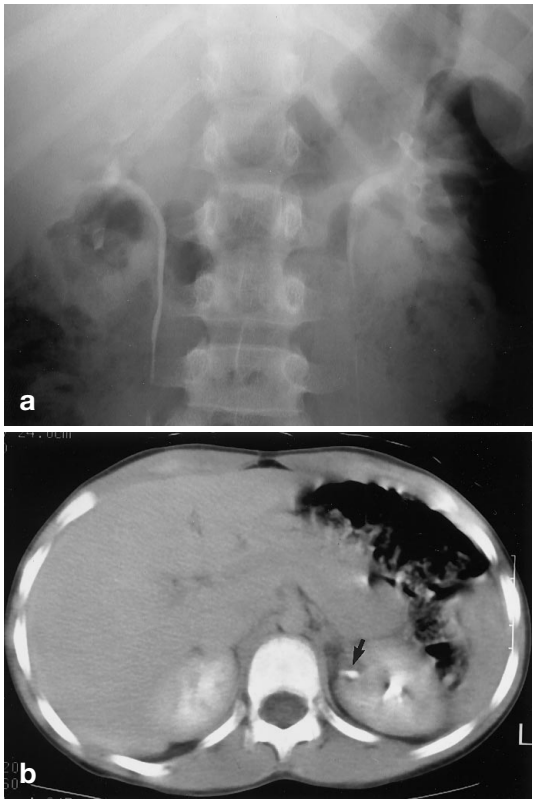

Fig. 6 a, b Grossly delayed referral. Nine-year-old girl was always wet despite normal toilet training. Pediatrician was not told until she was seven. Urine cultures all negative. Referred to pediatric urologist at age nine. **a** EU was finally done and shows subtle signs of left duplex kidney with dysplastic upper pole. The left ureter is further from the spine than the right. **b** CT scan confirmed presence of ureter draining poorly functioning left upper pole *(arrow)*. Left upper pole nephrectomy was performed and she was dry for the first time in her life

In this series, the ectopic orifice was just beneath and ipsilateral to the urethral meatus in seven girls and was in the vagina in five. The frequency of a visible perineal orifice was higher in these girls than has been previously described [4].

Usually, a ureter with an ectopic orifice drains the upper pole of a duplicated renal collecting system. Less often, it drains a kidney with a single collecting system [2, 4]. Mackie and Stephens showed that the more ectopic the ureteral orifice, the more dysplastic the upper pole it drains [12]. Therefore, the infrasphincteric ectopic ureter is usually associated with a small, dysplastic renal unit. These dysplastic nephrons lack concentrating ability; they are therefore difficult to see with EU unless a diligent search is done.

The infrasphincteric ectopic orifice, especially if it inserts into the vagina, is occasionally stenotic [5, 13]. In this unusual situation, the stenosis may lead to dilatation of the ureter and renal collecting system, thus making recognition easier on US. However, US cannot exclude the presence of an ectopic ureter. It is diagnostic only if a duplex kidney with an abnormal dysplastic upper pole with a dilated collecting system and a dilated ureter below the bladder are identified. This was seen in only two of the patients. A variable amount of upper pole pelvicalyceal dilatation in a duplex kidney is suggestive but not diagnostic [13]. The upper pole cortex may be thin, or if dysplastic it may be hyperechoic with or without small intraparenchymal cysts [13]. The lower pole collecting system may be dilated from vesicoureteric reflux. A normal duplex kidney in the presence of the symptoms described herein suggests that the other side is the culprit; this was the case in the six girls in this series whose ectopic ureter drained an upper pole. More subtle findings include an eccentric central echo complex (actually the lower pole sinus) (Fig. 2 a) and the pseudo-adrenal sign, where the tiny upper pole resembles the normal adrenal gland (Fig. 4).

Since US can be completely normal in girls with incontinence due to an ectopic ureter, or the abnormality can be very subtle, US is not recommended as the first imaging test. Furthermore, whether US is normal or abnormal, EU is still needed to evaluate ipsilateral upper pole function and to look for a contralateral occult duplication, since bilateral infrasphincteric ectopic ureters are possible [2]. (No bilateral ectopic ureters were seen in this series.) When a small, usually ectopic, dysplastic kidney with an ectopic ureteral orifice is present, it is usually impossible to find with ultrasound or EU, even though its presence is inferred from the combination of a classic history and a single normal kidney shown by US or EU. A DMSA scan is usually needed for general localization prior to a focused, contrast-enhanced CT study (Fig. 7).

Excretory urography should be used to visualize or infer drainage of an upper moiety or single-system kidney by an ectopic orifice [2, 3, 14]. The key to detection is to concentrate on the kidneys, not the ureters. The urographic findings in a duplex kidney with a dysplastic upper pole drained by an ectopic ureter are well described. There is often increased distance from the spine to the opacified lower pole pelvis or ureter (Fig. 5). The upper pole ureter is usually medial to the lower pole ureter and, if dilated, may displace the lower pole ureter far laterally [14]. However, even if the caliber of the upper pole ureter is normal, the lower pole ureter will usually be farther from the spine than normal. If the upper pole ureter is located anterior to the lower pole ureter, however, the distance from the spine to the lower pole ureter will be normal. This is not the usual situation [3]. The lower pole calyces are normally opacified with contrast, while the upper pole drained by the ectopic ureter may be quite small and have poor concentrating ability. Normal calyces may not be visible; instead, just a faint tubular structure may be seen (Fig. 9). Nephroto-

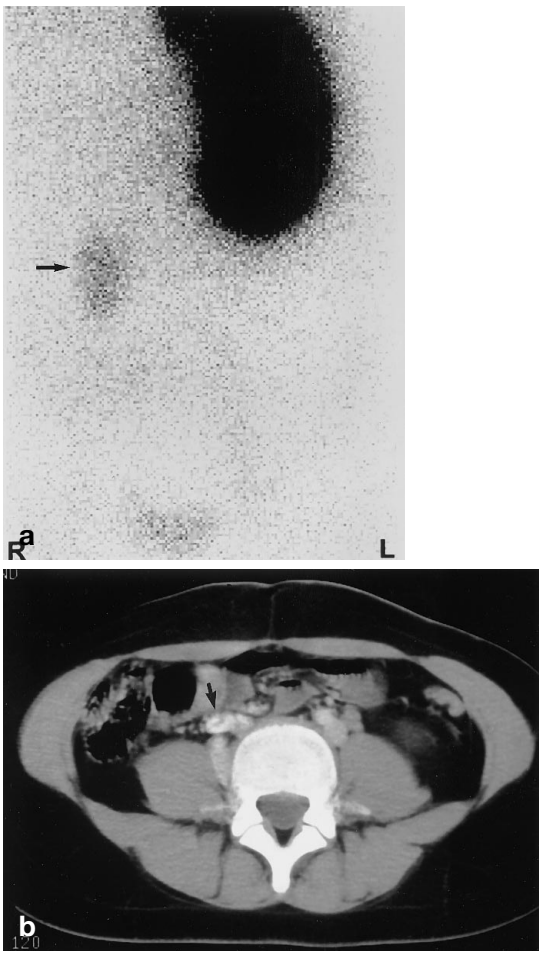

Fig. 7a,b Poorly functioning single system kidney and absence of ureteral orifice in the bladder mistaken for renal agenesis. This girl was diagnosed shortly after birth as having left-sided primary megaureter on EU that was done for evaluation of urosepsis. The right kidney was not seen. There was no right ureteral orifice at cystocopy and right renal agenesis was diagnosed. This incorrect deduction led her physicians to misinterpret her many years of constant day and night wetness. She underwent pharmacologic treatment, perineal exercises, and behavior modification until the age of 8 years 2 months when, in desperation, she was referred to a pediatric urologist. A Tc-99m-dimercaptosuccinic acid (DMSA) scan (**a**) and an enhanced CT scan (**b**) demonstrated the small, poorly functioning dysplastic ectopic right kidney *(arrows)* that had a single collecting system draining into the vagina. This kidney was resected; postoperatively, she was dry after a lifetime of wetting

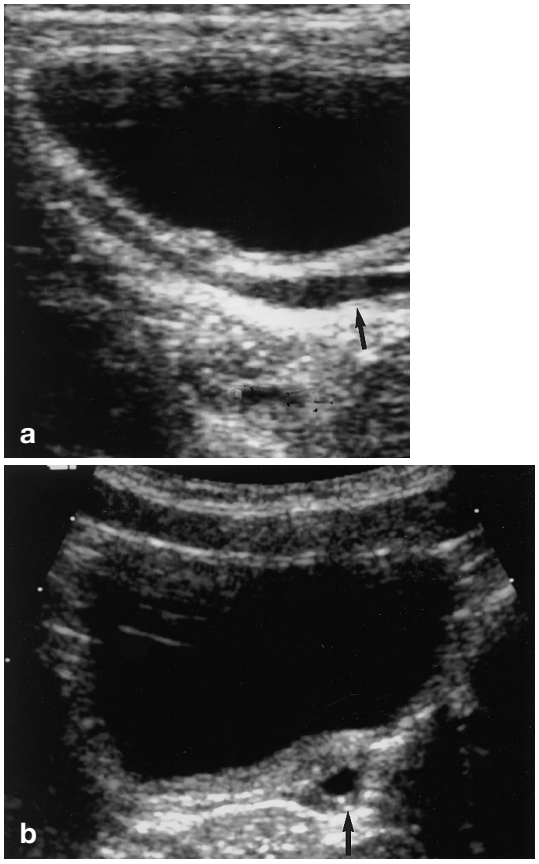

Fig. 8a,b Poorly functioning single-system kidney mistaken for multicystic dysplastic kidney (MCDK). This girl had been followed for a presumed left MCDK, discovered on a prenatal ultrasound examination. EU and VCUG were done at three months of age. A single right kidney was identified; there was nonvisualization of the left kidney. An ectopic ureteral orifice was not a concern at that time, as she was still in diapers. Ultrasonography was performed at ages 7 months, 2 years, and 3 years 4 months. US, done at age 2, demonstrated a 2.7 cm small dysplastic kidney presumed to be multicystic and nonfunctioning, on the left and compensatory hypertrophy on the right. A dilated left ureter was present but it was not recognized. A left kidney was not found on subsequent US examinations. She underwent delayed toilet training at the age of 4 years 6 months. Over the next 6 months, her mother noticed that her underwear was always damp and told her pediatrician. He thought it was probably not significant but recommended that they tell the radiologist during her next scheduled US. That study (**a**, longitudinal and **b**, transverse) at age 5 years 4 months demonstrated a dilated left ureter that ended below the bladder *(arrows)*. The left kidney was not seen. A DMSA scan and contrast-enhanced CT (not shown) revealed the small, dysplastic left kidney, which was removed. Postoperatively, she was dry for the first time in her life

mography may help with direct visualization of this unit. A normal duplex kidney on EU, just as on US, is a clue that the other kidney is the culprit. A normal contralateral duplex kidney was seen in six of the ten patients with ectopic upper pole ureters who were studied, and in 43 % in the series of 41 children reported by Mandell et al. [2].

When EU is normal, which is rare, the classic history should be enough to warrant contrast-enhanced CT to find the dysplastic upper pole that is almost certainly present [3, 8].

Nine of the girls in this series had the correct diagnosis made by EU. EU was, however, on average the third imaging study performed (range 2nd–9th). One pa-

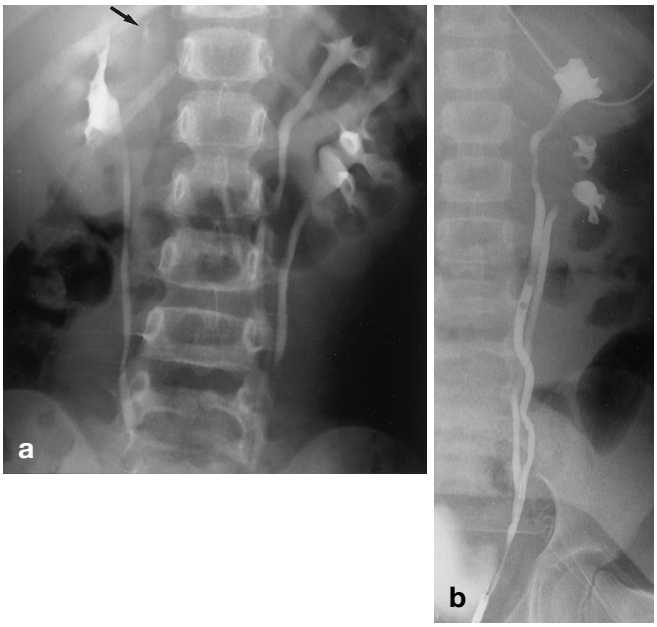

Fig. 9 a, b The occasional need for retrograde ureterography. **a** EU in girl with continuous urinary dribbling shows a normal left duplex kidney and an abnormal right duplex kidney with a tiny, nondilated upper pole *(arrow)*. The right lower pole ureter is the same distance from the spine as the left lower pole ureter. **b** Left retrograde ureterogram. During cystoscopy, a single right ureteral orifice was identified, the orifice draining the right lower pole. At the left corner of the trigone, there was only one orifice. To be absolutely sure that there were was not also an ectopic ureteral orifice on the left, a catheter was inserted into the orifice at the left hemitrigone and contrast injected. It shows that there is a left bifid ureter, thus excluding the presence of bilateral ectopic upper pole orifices

tient's first imaging study after 2 years of evaluation without imaging was a very suggestive EU, and the occult upper pole was confirmed by CT (Fig. 6). The combination of DMSA scan and contrast-enhanced CT was necessary in two girls with a small dysplastic kidney draining through a single ectopic ureteral orifice (Fig. 7).

Voiding cystourethrography (VCUG) has no role in the diagnosis of an infrasphincteric ectopic ureteral orifice. Cystoscopy and retrograde ureterography are usually not necessary. They are helpful when the patient has bilateral duplex kidneys and the sites of insertion of

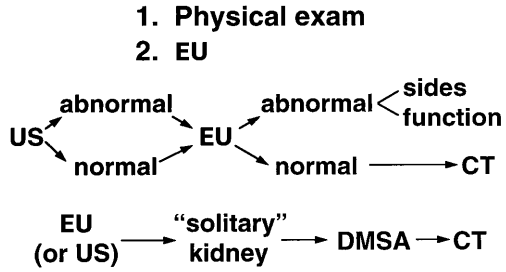

Fig. 10 Algorithm for imaging evaluation of girls with continuous urinary dribbling

the upper pole ureters is unknown, since occasionally there are bilateral infrasphincteric ectopic ureters (Fig. 9).

In conclusion, when a young girl presents with constant urinary dribbling, every day and every night as long as anyone can remember despite normal toilet training, the diagnosis of infrasphincteric ectopic ureter is very likely. The history of constant wetting in an otherwise normal girl is the key to the diagnosis. The ectopic ureteral orifice is often visible just ipsilateral and inferior to the normal urethral meatus, as was true in seven of 12 girls.

Delays in diagnosis occurred when (1) parents failed to recognize or admit that constant dampness or wetting was a significant problem, (2) the classic history was not recognized to be the hallmark of ureteral ectopia, (3) a careful search for the ectopic orifice was not performed, or (4) imaging studies were misinterpreted.

Ultrasound examination is not recommended in these patients; if it is normal, EU is needed and if it is abnormal, EU is still needed. If US is interpreted as normal, this may delay the search for the ectopic ureteral orifice, as was true in seven girls who had a delay in diagnosis of one month to nearly seven years before the diagnostic EU was done.

Given the appropriate history, the imaging approach is straightforward (Fig. 10): if EU is abnormal, proceed with surgery (only rarely is a retrograde ureterogram needed); if EU is normal, proceed to CT imaging to find the occult dysplastic upper pole; if EU shows only a solitary kidney, a DMSA scan to find the ectopic, dysplastic, single-system kidney on the other side is followed by a focused contrast-enhanced CT study for more precise preoperative planning.

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