

Ureteral Ectopia in Complete Duplicated System Treated with Ureteropyelostomy

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ABSTRACT Ureteral ectopia in girls may cause incontinence when it opens to the distal region of the external sphincter. Herein, we aimed to discuss the diagnosis and treatment of a case treated with ureteropyelostomy. The female patient with antenatally diagnosed left hydronephrosis had periurethral dripping when she was evaluated in the neonatal period. Continuous urine drainage from the periurethral area was observed in the examination under general anesthesia, but ectopic ureter opening was not seen. At two years of age, ultrasonography and renal scintigraphy showed similar findings and dripping was also present. Ureteropyelostomy and upper pole ureterectomy were performed with left anterior subcostal incision. Surgical treatment in patients who have duplex collecting system with ectopic ureteric opening varies according to the function of the renal unit drained by the ectopic ureter. Ureteropyelostomy and distal ureteral excision are appropriate treatment options in patients with functional upper pole.

Keywords: Ureter; duplication; urinary incontinence; ureteropyelostomy

The caudal opening of the ureteral orifice beyond the standart location of the bladder trigone is called ectopic ureter.¹ More than 80% of patients with ectopic ureter also have a complete duplex system.² The fact that affected individuals are asymptomatic often prevents them from being diagnosed with ectopic ureter. In females, the ectopic ureter may present with urinary incontinence. In both sexes, it may be diagnosed in the antenatal period while it may present with urinary tract infection or urinary obstruction in the postnatal period.

In male patients, the most common ectopic ureter orifice site is posterior urethra (50%). Other opening sites contain seminal vesicle in about one-

third of the patients, vas deferens, prostate, bladder neck, and epididymis. Bladder and upper urethra (33%), vaginal vestibule between the urethra and vaginal opening (33%), vagina (25%), cervix and uterus (<5%) are the most common terminal sites of female ureteral ectopy.³ In this study, we aimed to discuss the diagnosis and treatment of a case with ureteral ectopia in complete duplicated system treated with ureteropyelostomy.

CASE REPORT

The newborn female patient with antenatally diagnosed left hydronephrosis was admitted for postnatal follow-up. On inspection, continuous urine dribbling

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from the periurethral area was determined. The genital examination was normal and an ectopic opening ureteric orifice could not be seen. Urinary system ultrasonography revealed normal right kidney, a complete double ureteral system in the left kidney, and hydroureteronephrosis in the ureter draining the upper pole. With ultrasound in the left kidney, the lower pole parenchymal thickness was 15 mm, the anterior-posterior diameter of the pelvis was 3 mm and the degree of hydronephrosis was Society of Fetal Urology (SFU) Grade-1. Upper pole parenchymal thickness was 10 mm, pelvic anterior-posterior diameter was 6 mm and hydronephrosis degree was SFU Grade-2. Voiding cystourethrography (VCUG) was normal. Mercaptoacetyltriglycine (MAG3) dynamic renal scintigraphy showed functional upper and lower poles of the left kidney (Figure 1), and stasis with totally wash out after diuretic administration in the left upper pole and ureter. Differential renal function was 52% on the left and 48% on the right side. In the examination performed under general anesthesia, continuous urine dribbling was observed from the periurethral area, but no ectopic ureter opening was observed. On cystoscopy, the bladder was normal and ureteral orifices were in normal location bilaterally. Cystoscopy and vaginoscopy revealed no ectopic ureteral openings in the urethra, bladder or vagina. Left retrograde pyelography revealed only the left lower pole collecting system, and this collecting system and ureter were normal (Figure 2).

At the age of two years, repeated urinary system ultrasonography and MAG3 dynamic renal scintigraphy revealed similar findings, and the patient still had complaint of urine dribbling from periurethral area. Considering that the patient's condition is thought to be resolved before toilet training, ureteropyelostomy and ureterectomy of the dilated and tortuous upper



FIGURE 1: Renal involvement in the 3rd, 4th and 5th minutes of nuclear isotope administration (Left kidney with upper and lower poles on the left side, right kidney in the right side).



FIGURE 2: Left lower pole collecting system (Retrograd pyelography).

pole ureter as far down as could be reached were performed with the left anterior subcostal incision (Figure 3). Three Fr ureter stent was inserted cystoscopically to the lower pole ureter and it was placed to the upper pole pelvis during ureteropyelostomy. Early postoperative period was uneventful and she was discharged on the second postoperative day. Ureteral stent (proximal part was left out of the urethra) was removed in outpatient clinic on the postoperative 7th day. The patient had toilet training, while she was full continent and asymptomatic at the end of postoperative first and third year.

DISCUSSION

Embryologically, ureters thrive from mesonephric ducts at the fourth gestational week. The ureteral transcription fully occurs when the metanephric blastema is stimulated by two different ureteric buds that emerge from the mesonephric ducts. With respect to the Weigert-Meyer law, the lower pole ureter opens more laterally and cranially, and the upper pole ureter opens more caudally and medially as in our patient.^{4,5}

In the case series published in the United States and Great Britain, the majority of ectopic ureters are associated with a duplex kidney, ranging from 75 to

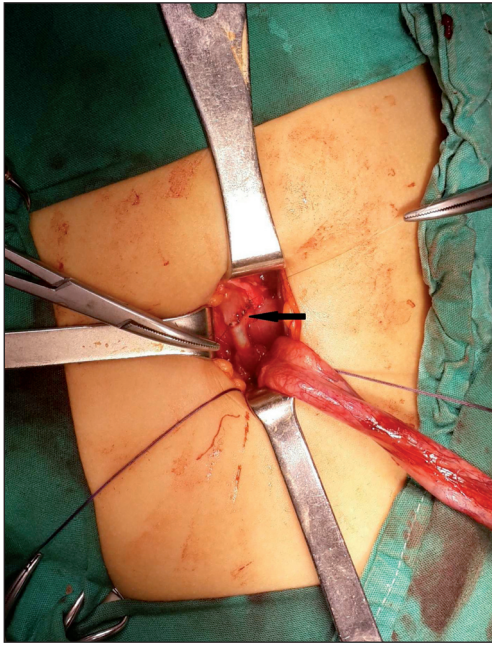


FIGURE 3: Ureteropyelostomy and dilated upper pole ureter before excision (The ureteropyelostomy site is marked with black arrow).

90 percent. In these reports, ectopic ureters in duplex kidneys are seen 8-9 times more common in females than in males.^{6,7} In contrast, ectopic ureters with single renal system are more common in India and Japan. In India, 75 percent of ectopic ureters were associated with a single kidney system. In this series, there was a strong female predominance.⁸ However, in reports from the United States and Great Britain, single-system ectopia is either equally or more likely to occur in males than in females.⁷ This may be related to genetic differences between populations.

The ectopic ureteral orifice is every time above the external sphincter in male patients. Therefore, male patients with ectopic ureter have no urinary incontinence. But typically, they present with hydronephrosis or urinary tract infection. In contrast, due to the ectopic ureters may get over the external sphincter, females sometimes present with a history of urinary incontinence. The incontinence is typically characterized by continuous dripping or moisture requiring the use of a pad. Since the upper pole drained by the ectopic ureter is generally dysplastic or low-functional, the volume of urine ectopically drained is also in low volume.² In our patient, the upper pole drained by the ectopic ureter was functional, and

there was constant wetting in the diaper and moisture in the vaginal area.

Ultrasonography is the first diagnostic method to detect possible hydronephrosis associated with ectopic orifice. Because of associated anomalies such as ureterocele or vesicoureteral reflux, VCUg should be seen in patients with double collecting system. Although rarely done, intravenous pyelography could be seen in patients with suspected double collecting system. However, the ureter draining the dysplastic or dysfunctional kidney may not be seen in intravenous pyelography.² In the case of diagnostic challenge with suspicion of ureteral ectopia, magnetic resonance urography is the ideal diagnostic method currently used. Magnetic resonance urography is able to indicate the dilated system, ectopic ureter and its extravesical opening and give detailed information about the malformation.⁹

Evaluation under anesthesia, cystourethroscopy and vaginoscopy should be used in the diagnosis, however endoscopic assessment is not always able to reveal ectopic orifice. Plaire et al. showed that 58 percent of the ureters ectopically open to the vagina, vestibulum or bladder neck may be assigned.⁷ In our patient, urine dribbling was seen but the ectopic opening of the ureter was not determined and ureteral catheterization could not be done.

Most of the ectopic ureters in a double collecting system are related with a dysplastic upper pole. Extraction of this part throughout with the proximal ureter is commonly curative. When the upper pole is well functional, the ectopic ureter is reimplanted into the bladder or anastomosed to the normal pole ureter as ureteroureterostomy or lower pole pelvis as ureteropyelostomy. In our patient, ureteropyelostomy was selected as a surgical preference to avoid ureteroureteral reflux (Yo-yo reflux).

In conclusion, the ectopic ureter should be kept in mind in case of continuous urine dribbling. Surgical treatment in patients who have duplex collecting system with ectopic ureteric opening varies according to the function of the renal unit drained by the ectopic ureter. Ureteropyelostomy and distal ureteral excision are ideal treatment options in patients with functional upper pole.

Informed Consent

Due to the fact that our study was a case report, ethics committee approval was not required. The permission to use the patient data in later studies was obtained at hospital admission.

Source of Finance

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or mem-

bers of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Serdar Moraloğlu, Sabri Cansaran, Oktav Bosnali, Ayşenur Celayir; **Design:** Serdar Moraloğlu, Sabri Cansaran, Oktav Bosnali, Ayşenur Celayir; **Control/Supervision:** Serdar Moraloğlu, Ayşenur Celayir; **Data Collection and/or Processing:** Sabri Cansaran, Serdar Moraloğlu; **Analysis and/or Interpretation:** Serdar Moraloğlu, Sabri Cansaran, Oktav Bosnali; **Literature Review:** Sabri Cansaran; **Writing the Article:** Serdar Moraloğlu, Sabri Cansaran, Oktav Bosnali; **Critical Review:** Ayşenur Celayir, Serdar Moraloğlu.

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