Ectopic ureter as a rare cause of urinary incontinence in a young lady.

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ABSTRACT
We report a case of a 16 year old female with urinary incontinence since birth due to left sided duplex ureter, with the upper pole draining into the posterior vaginal wall. The patient was managed with end to side proximal ureteroureterostomy into the normal ipsilateral ureter of the lower pole.

KEY WORDS: urinary incontinence, duplex ureter, ectopic ureter

INTRODUCTION
Pyeloureteral duplication or duplex system is one of the most common anomalies of the upper urinary tract. Ectopic ureter is defined as any ureter, single or duplex, that does not open in the trigonal region of the bladder. More than 80% of ectopic ureters are associated with complete duplex. In females, the ectopic ureteric opening may be located anywhere from bladder neck to perineum with urethra, vagina, and vestibule being the commonest sites of entry. An ectopic ureteric opening distal to the external sphincter or into the Mullerian system may be associated with incontinence, which can cause major social stigma for the patient.

CASE REPORT
A 16 year old girl presented with history of persistent dribbling of urine per vagina since birth. The patient was on diapers on a regular basis for persistent wetting; however she also had a normal voiding pattern. The patient had normal menstrual cycle. There was no history of surgery, trauma or tuberculosis. She had normal bowel habit and normal gait. Clinical examination revealed a normal urethral meatus with pooling of urine at the introitus without any obvious congenital anomaly. The spinal examination showed no abnormality.

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Hematological and biochemical parameters were
within normal limits. Urine culture showed no growth. Ultrasonogram revealed bilateral duplex system. CT urogram showed bilateral normal excreting kidneys, with partial duplex collecting system on the right side and complete duplex collecting system on the left side with ureteric opening of upper pole to the posterior wall of the vagina (figures 1-3). Micturating cystourethrogram showed a normal bladder outline, with no reflux or ureterocele (figure 4).

On vaginoscopy, no ureteric opening could be identified; however, continuous dribbling of urine was seen from the posterior wall of vagina. Cystoscopy findings were normal and a double J stent was inserted to left lower pole ureter. Upper tract was explored through a left flank incision and two ureters were separately identified on ipsilateral side (figure 5).

The proximal parts of both the ureters were dissected separately and end to side ureteroureterostomy of
upper pole ureter to lower pole ureter was performed with a double J stent in the lower pole ureter (figure 6). The distal stump of upper pole ureter was tied and left alone without any further dissection which could injure the common sheath of the two moieties at the distal end.

The postoperative recovery was uneventful and gauze piece placed at the vagina was seen dry within 8 hours, after which she was dry all the time. She was discharged on 4th postoperative day. Her double J stent was removed cystoscopically after 6 weeks. On follow up after 3 months, she remained dry, continent and with normal voiding pattern.

DISCUSSION

As most ectopic ureters are asymptomatic, the true incidence remains unknown. They are more common in the females with ratio of 6:1 and in females more than 80% of the ectopic ureters drain duplicated systems, whereas majority of ectopic ureters drain single systems in males. In females, the ectopic ureter may enter anywhere from the bladder neck to the perineum and into the vagina, uterus, and even rectum. In males, the ectopic ureter always enters the urogenital system above the external sphincter or pelvic floor, and usually into the Wolffian structures, including vas deferens, seminal vesicles, or ejaculatory duct.

The duplication of the ureters results from the existence of 2 separate ureteral buds arising from the mesonephric duct. The ureteral bud closest to the urogenital sinus, the lower pole ureter, will be absorbed into the urinary tract first and might be more cranial and lateral than normal. The upper pole ureter will remain associated with the mesonephric duct longer, and its orifice will be caudal and medial to that of the lower pole. This anatomic relationship between the upper and lower pole ureters, achieved by rotation and migration, is determined by the Weigert-Meyer law. Mackie and Stephens showed that the greater the distance of the origin of the ureteral bud from the middle section of the mesonephric duct, the more abnormal will be the induced renal segment.

Ectopic ureters may be asymptomatic in childhood and remain undiagnosed until adulthood. As in our case, the classic presentation of an extravesical, infra-sphincteric ectopic ureteral orifice in a girl is a lifelong history of persistent enuresis or dribbling associated with otherwise normal voiding habits. The diagnosis may be made on investigations for recurrent urinary tract infection. The other variable presentations in neonates may be abdominal mass due to massive hydrenephrosis and failure to thrive. In males, the ectopic orifice is never distal to the urethral sphincter but may be incorporated into the mesonephric duct derivatives. Therefore, recurrent epididymitis or a seminal vesicle mass may be the initial finding.

The majority of ureteroceles and ectopic ureters are detected through prenatal ultrasound imaging, even if the specific diagnosis is not made. Ultrasound can be useful in identifying a hydrenephrotic collecting system and dilated ureter at the level of the bladder. But, in most instances, the diagnosis of an ectopic ureter is confirmed by intravenous urogram, preferably a CT urogram. The usual radiographic finding is a nonvisualized or poorly functioning upper pole of a duplex system that may be hydrenephrotic. The upper pole displaces the lower pole downward, producing the classic “drooping lily” sign. In this case, as the upper pole was unobstructed and functioning, this sign was not seen.

In cases where an ectopic ureter is suspected MRI may be a good tool. A study by Anand Krishnan and Laurene S Baskin demonstrated the value of MRI in clarifying the diagnosis of ectopic ureter when conventional imaging is equivocal but clinical suspicion remains high. A micturating cystourethrogram should be performed to assess reflux into the ectopic ureter, as well as into the lower pole. Also, if there is concern about the salvageability of the upper pole, isotopic renal scanning should be considered. Our case did not demonstrate reflux on micturating cystogram and the upper pole was salvageable on isotopic scan.

The goals of therapy are preservation of renal function; elimination of infection, obstruction, and reflux; and maintenance of urinary continence. For an ectopic ureter, this can mean common sheath reimplantation or ureteroureterostomy, either low or proximal near the renal pelvis. Upper pole removal using a partial nephrectomy or heminephrectomy of a duplex system is typically the preferred treatment.
when there is clearly no function in the upper pole and if there is concern about how effective a drainage procedure may be because of massive dilation\(^5\). Pyeouterostomy is preferred by some surgeons to take care of the “Yo-Yo” effect\(^13\), which is seen with the distal ureteroureterostomy. Currently, the laparoscopic approach is more frequently used\(^14\). Ureteroureterostomy is a good treatment modality for managing an ectopic ureterocele or ectopic ureter and preserving upper pole kidney function, but a low ureteroureterostomy can be associated with a Yo-Yo effect. The complete excision of residual stump is not necessary unless the stump has proven vesicoureteric reflux. However, the development of urinary tract infection at the residual stump is a concern. Yong Seung Lee et al\(^{15}\), concluded that upper pole ureter diameter was correlated with the development of a urinary tract infection at the residual stump.

The ultimate prognosis after ureteroureterostomy in patients with duplicated ureter with ureteral ectopia is excellent. Our patient has been seen on a regular follow up and has been totally continent, with normal voiding pattern.

**CONCLUSION**

A high index of suspicion is required for an ectopic ureter in girls with persistent incontinence and a normal voiding pattern following toilet training. Appropriate imaging studies should be obtained and carefully interpreted. An ectopic ureter should be surgically reconstructed depending upon the degree of renal function and the presence of vesicouterteral reflux.

**REFERENCES**


