

## Intermittent Urinary Incontinence Secondary to Inverted-Y Ureteral Duplication With Perianal Ectopia



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Inverted-Y ureteral duplication is one of the rarest anomalies of ureteral branching, which has been found to be more prevalent in females.<sup>2</sup> Females commonly present with urinary incontinence often associated with distal limb ectopia to the bladder neck, uterus, or vagina.<sup>2</sup> We present the case of a 7-year-old female with intermittent urinary incontinence who was found to have an inverted-Y ureteral duplication with perianal ectopia. We highlight the role of magnetic resonance urography in the evaluation of females with urinary incontinence and a normal renal/bladder ultrasound who are refractory to behavioral therapy. *UROLOGY* 127: 124–126, 2019. © 2019 Elsevier Inc.

Ureteral duplication is a common condition occurring in 0.8% of the population.<sup>1</sup> In most cases, ureteral duplication anomalies are associated with a duplicated collecting system. In females with complete duplication anomalies, upper pole ureteral ectopia may present with constant urinary incontinence or vaginal discharge.<sup>1</sup> Inverted-Y ureteral duplication is one of the rarest anomalies of ureteral branching with less than 40 cases reported in the literature.<sup>2</sup> The distal limb has been reported to drain into the bladder neck, uterus, vagina, epididymis, seminal vesicles, as well as having an associated ureterocele and ureteric stone.<sup>2-13</sup> However, we believe this is the first reported case of an inverted-Y duplication anomaly with perianal ectopia presenting with intermittent urinary incontinence.

Her mother had recently taken her to a local emergency room for evaluation of fluid leaking from a pinpoint opening lateral to her anus on the right side (Fig. 1). During the course of her visit, she was examined twice, initially there was no drainage identified, however, with relaxation of her pelvic floor muscles there was drainage noted. An MRI urogram was obtained which demonstrated a distal bifurcation of her right ureter with the anteromedial branch inserting normally into the bladder and the posterior branch extending toward the perianal soft tissue (Fig. 2A). A cystoscopy and right retrograde study confirmed the MRI urogram findings (Fig. 2B). She underwent laparoscopic removal of the pelvic segment of the ectopic ureteral branch via ligation with endoloop and has remained dry since surgery (Fig. 3).

### CASE DISCUSSION

A 7-year-old female with a past medical history significant for attention deficit disorder and recurrent urinary tract infections presented with her adoptive mother for further evaluation of persistent intermittent urinary incontinence. Prior to presentation, previous workup included a normal renal ultrasound and voiding cystourethrogram. Despite being placed on a bowel and bladder regimen, her urinary incontinence continued. At the time of presentation, she had regular voiding habits but noted that she had to wear a pad and was damp every day. She was on polyethylene glycol and reported regular bowel habits.

### DISCUSSION

The ureteral bud, the precursor of the ureter, arises from the caudal portion of the Wolffian (mesonephric) duct. The cranial portion of the ureteric bud meets the metanephric blastema and reciprocal induction results in development of the kidney from the metanephric blastema and the collecting system from the ureteric bud. Caudally, the mesonephric duct (with the ureteric bud) are incorporated into the cloaca as it forms the trigone.<sup>1</sup> Ureteral abnormalities arise related to alterations in the bud number, position, or time of development. Inverted-Y ureteral duplication is a rare form of ureteral duplication, first described in 1913.<sup>2,5</sup> Less than 40 cases have been described, primarily in females.<sup>2-13</sup> The development of inverted-Y duplication anomalies is unclear. It may be related to the development of 2 ureteric buds that fuse proximally resulting in a single ureteric bud joining the metanephric blastema.<sup>2,6</sup> The proximal limb inserts into the bladder while the distal limb has been reported to end

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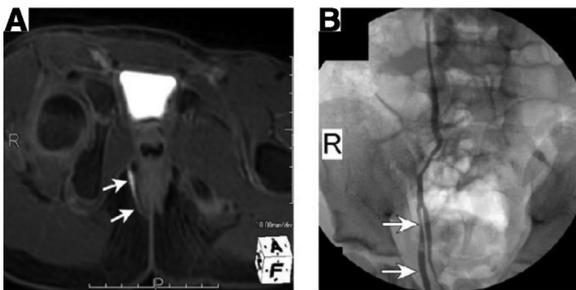
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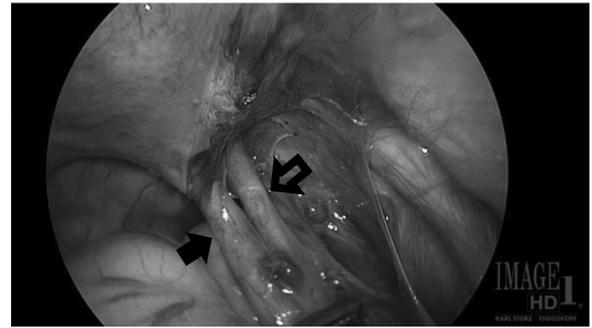
**Figure 1.** Urine leakage from right-sided ureteral perianal opening.

in a ureterocele, atresia, stasis with a calculus and ectopic insertion into the vagina, seminal vesicles, epididymis, uterus, and bladder neck.<sup>2-13</sup> The location of insertion of the distal limb results in associated symptoms.

Most ureteral anomalies are detected on prenatal ultrasound. The presence of a duplicated collecting system on ultrasound coupled with a history of persistent urinary incontinence will often confirm the diagnosis of an ectopic



**Figure 2.** (A) Axial-oblique multiplanar reformatted T1-weighted MR image obtained 50 minutes after the administration of IV Gadolinium shows a portion of the anomalous tract (arrows) coursing into the perineum. (B) Fluoroscopic spot obtained following injection of contrast into the right ureterovesical junction shows retrograde filling of a normal caliber right mid and proximal ureter with no evidence of mid or proximal duplication. At the junction of the mid and distal third of the ureter, near the pelvic inlet, inverted-Y duplication (arrows) of the distal ureter is seen.



**Figure 3.** Laparoscopic visualization of the inverted-Y duplication with the normal ureter inserting into the bladder (solid arrow) and the ectopic ureter (open arrow).

insertion or ectopic ureter. However, in the absence of a suspicious ultrasound and a history of intermittent urinary incontinence, ureteral ectopia may be missed. The rarity of inverted-Y duplication anomalies leads to delays in diagnosis.<sup>2,14</sup> Shiruma et al reported a case of persistent urinary incontinence after nephrectomy related to an inverted-Y duplication with ectopic ureteral insertion into the vagina.<sup>3</sup> In our case, the child was treated for several years for voiding dysfunction due to the intermittent nature of her urinary incontinence and a normal renal/bladder ultrasound. In this case, given the absence of a duplicated upper urinary tract, the lack of ureteral dilatation, and the lack of an ureterocele, the anomaly was not initially detected. Careful inspection of her perineum with pelvic floor muscle relaxation identified the ectopic orifice and drainage. Magnetic resonance imaging has been shown to more clearly demonstrate the anatomy of the renal parenchyma, renal collecting system, and the ureter and ureteral orifice compared to renal ultrasound and intravenous urography.<sup>15</sup> In this case, the MR urogram clearly identified the inverted-Y duplication anomaly as well as the distal limb extending to the perineum. We postulate that the distal limb passed through the pelvic floor muscles as her urinary incontinence occurred only with relaxation of her pelvic floor muscles. Although rare, inverted-Y duplication anomalies should be considered in females with persistent urinary incontinence not responding to standard behavioral regimens. MRI urogram is able to confirm the diagnosis and is useful in preoperative planning. A laparoscopic approach provided a cosmetically appealing result and resulted in minimal postoperative pain.

## References

1. Peters CA, Mendelsohn C. Ectopic ureter, ureterocele, and ureteral anomalies. In: Wein AJ, Kavoussi LR, Partin AW, et al. eds. *Campbell-Walsh Urology*. 11th ed. 4, Philadelphia: Saunders Elsevier; 2016:3075-3101.
2. Upadhyay J, Zuckerman JM, Shekarriz B. Minimally invasive surgical approach to excision of symptomatic ectopic inverted-Y ureteral duplications. *J Pediatr Urol*. 2013;9:e114-e116. <https://doi.org/10.1016/j.jpuro.2012.10.015>. Epub 2012 Nov 7.
3. Shimura H, Mitsui T, Aoki T, Kamiyama M, Yamagishi T, Takeda M. Persistent urinary incontinence after nephrectomy: a case of

- inverted-Y ureteral duplication with ectopic ureteral insertion into the vagina. *Urol Case Rep*. 2016;9:58–61. <https://doi.org/10.1016/j.eucr.2016.09.001>. Published online 2016 Oct 7.
4. Regan TC, Dejter Jr. SW. Inverted Y duplication of the ureter with associated ureterocele and bladder outlet obstruction. *J Urol*. 1996;155:642.
  5. Hajji F, Hammoune N, Miloudi M, Belasri S. Unusual variant of inverted Y ureteral duplication with an ipsilateral seminal vesicle cyst and renal dysgenesis. *Ann R Coll Surg Engl*. 2017;99:e19–e21. <https://doi.org/10.1308/rcsann.2016.0281>. Published online 2017 Jan.
  6. Schenkman NS, Fernandez EB, Wind G, Irby P. Inverted Y duplication of the ureter associated with a distal limb ectopic to the seminal vesicle. *J Urol*. 1994;152:946–947.
  7. Ecke M, Klatte D. Inverted Y-ureteral duplication with an uterine ectopy as cause of ureteric enuresis. *Urol Int*. 1989;44:116–118. <https://doi.org/10.1159/000281484>.
  8. Klauber GT, Reid EC. Inverted Y reduplication of the ureter. *J Urol*. 1972;107:362–364.
  9. Imatsu A, Iwama H, Katayama T, Sotoma T. A case of inverted Y ureter duplication with a blind-ending branch. *Jap J Clin Urol*. 1975;29:264.
  10. Mondet F, Ravery V, Roux CH, Hoffmann P, Delmas V, Boccon Gibod L. Partial ureteral duplication in an inverted Y with epididymal ureteric ectopia and intrasinus ureteral junction. *Surg Radiol Anat*. 1998;20:135–138.
  11. Liu L, Yang J, Zhu L, et al. Crossed-fused renal ectopia associated with inverted Y ureteral duplication, ectopic ureter, and bicornuate uteruses. *Urology*. 2010;75:1175–1177. <https://doi.org/10.1016/j.urology.2010.02.037>. Epub 2010 Mar 27.
  12. Mourad MS. Inverted Y ureteral duplication with a large ureterocele and draining a bifid renal pelvis in a man. *J Urol*. 1997;157:1353.
  13. Suzuki S, Tsujimura S, Sugiura H. Inverted Y ureteral duplication with a ureteral stone in atretic segment. *J Urol*. 1977;117:248–250.
  14. Kreissl MC, Lorenz R, Ohnheiser G, Darge K. Dystopic dysplastic kidney with ectopic ureter: improved localization by fusion of MR urography and (99m)Tc-DMSA SPECT datasets. *Pediatr Radiol*. 2008;38:241–244. <https://doi.org/10.1007/s00247-007-0660-5>. Epub 2007 Nov 20.
  15. Adeb M, Darge K, Dillman JR, Carr M, Epelman M. Magnetic resonance urography in evaluation of duplicated renal collecting systems. *Magn Reson Imaging Clin N Am*. 2013;21:717–730. <https://doi.org/10.1016/j.mric.2013.04.002>. Epub 2013 Aug 9.