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REVIEW ARTICLE

Congenital urogenital anomalies that are associated with the persistence of Gartner's duct: A review

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The embryogenesis and management of congenital urogenital anomalies that are associated with ureteric ectopia and the persistence of Gartner's duct are discussed. Ureteric ectopia with Gartner's duct cyst is caused by the failure of separation of the ureteric bud from the mesonephric duct, which leads to persistence of Gartner's duct, frequently with cystic dilation. Abnormal development of the ureter subsequently causes maldevelopment or absence of the ipsilateral kidney. The diagnosis and treatment of 2 adult women with congenital urethrovaginal fistula that was associated with unilateral single ectopic ureter, renal agenesis, and Gartner's duct anomaly are presented. Surgical repair of the urethrovaginal fistulae and removal of the Gartner's duct and cyst was performed transvaginally.

Clinicians who manage female incontinence must have a good understanding of congenital malformations of the urinary and genital tracts, the embryologic development of the urogenital systems, and their diagnosis and treatment. Ectopic ureters in the female patient frequently are not diagnosed until adulthood,[1] especially if the ectopic ureter is associated with severe renal dysplasia or agenesis and if childhood incontinence is not severe.

In this review, we describe the normal and abnormal embryologic development of the urogenital systems and discuss the diagnosis and treatment of 2 women, each of whom have a single ectopic ureter, renal agenesis, and persistent Gartner's duct abnormalities.

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Embryologic implications

Between 3 and 8 weeks of gestation, the urogenital systems develop from the intermediate mesoderm, which extends along each side of the dorsal body cavity of the embryo, and forms the urogenital ridge and mesonephric ducts. A pair of ureteric buds grows upwards from the distal mesonephric duct near its insertion into the cloaca and induces the overlying sacral intermediate mesoderm (metanephros) to develop into the primitive kidneys. The distal portions of the mesonephric ducts and attached ureteric buds migrate inferiorly to become incorporated into the posterior wall of the primitive bladder to become the definitive ureters, trigone, and bladder neck. The trigonal mesodermal tissue is covered by endoderm from the urogenital sinus. The paramesonephric ducts develop as an ingrowth of coelomic epithelium lateral to the mesonephric ducts and give rise to the fallopian tubes and uterus, with the epithelium of the vagina being derived from the endoderm of the

urogenital sinus and vaginal fibromuscular wall from the surrounding mesenchyme. The distal mesonephric ducts in the female are absorbed but may persist as vestigial remnants in the anterolateral vaginal wall down to the hymen (Gartner's duct cysts) and between the layers of the broad ligament (paraovarian cysts).

A ureter is defined as ectopic when it inserts into other than the superolateral portion of the trigone; possible sites of ectopic exit are the trigone, posterolateral wall of the urethra, anterolateral vagina, and vestibule and rarely the cervix and uterus. Ectopic ureters may also communicate indirectly with the urinary or genital tracts through a Gartner's duct cyst[2-5] and can be associated with single or duplex upper urinary tract system. Ureteric ectopia results from abnormal development of the mesonephric (wolffian) and paramesonephric (mullerian) ductal systems at 5 to 8 weeks of gestation. Gotoh and Koyanagi[4] proposed that ureteric ectopia with Gartner's duct cyst is caused by a failure of separation of the ureteric bud from the mesonephric duct that leads to persistence of Gartner's duct, frequently with cystic dilation. Abnormal development of the ureter subsequently causes maldevelopment or absence of the ipsilateral kidney.

Congenital malformations of the urinary tract are associated frequently with genital tract abnormalities, because interaction between the 2 ductal systems is necessary for normal growth. The finding of an anomaly in 1 system should alert the clinician to the possibility of an abnormality in the other. Thompson and Lynn[6] estimated that 35% of female patients with unilateral renal agenesis have partial or complete duplication of the genital tract. Semmens[7] found that renal agenesis was present in 43% of patients with uterus didelphys and that 10% of patients with genital tract abnormalities had an abnormal or ectopic kidney. The incidence of unilateral renal agenesis is approximately 1 in a 1000 autopsies.[2]

Case reports of Gartner's duct and cyst that are associated with a single ectopic ureter and renal agenesis are present mainly in the urologic literature.[2-5] Sheih et al[8] found 13 cases of cystic dilations in the pelvis that were associated with ipsilateral renal agenesis or dysplasia in a mass ultrasound-screening program for occult renal abnormalities of 280,000 school-aged children in Taiwan; 7 were young girls with Gartner's cysts. In another study, Shieh et al[9] documented 20 girls with renal dysgenesis and Gartner's duct cysts, one-half of which had coexisting mullerian duct anomalies. A small number of cases with Gartner's duct cyst in the literature had associated congenital vesicovaginal fistula[3,4,10]; to our knowledge, there have been no reported cases of an adult female with an urethrovaginal fistula with this combination of congenital abnormalities. However, the site of the fistula is described frequently as bladder neck and may well be similar to the cases described herein.

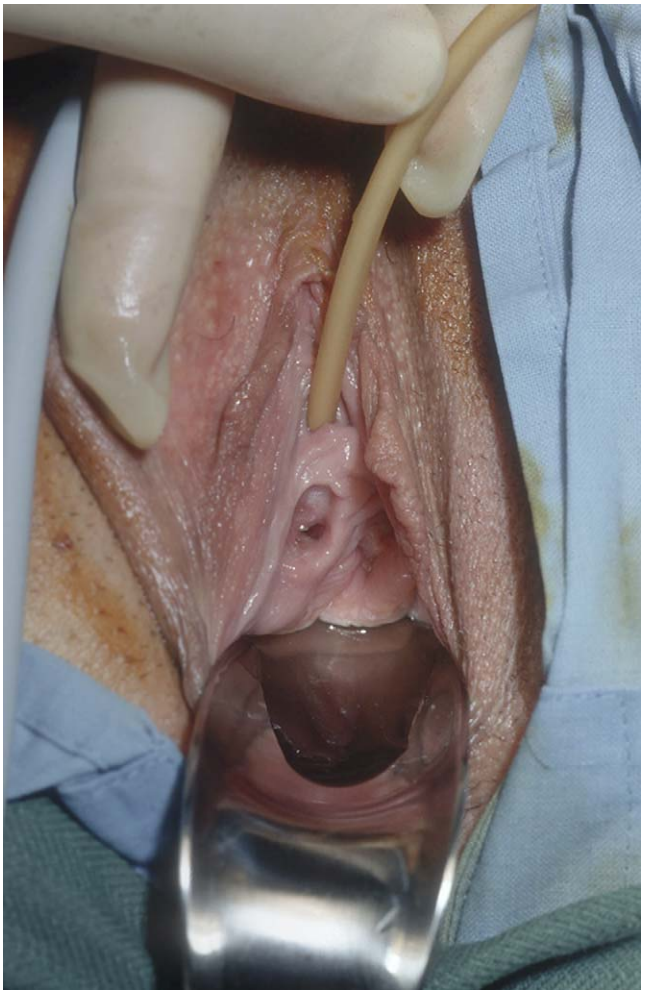

Figure 1 Ostium of ectopic ureter inferiolateral to external urethral meatus with an indwelling Foley catheter (case 1).

Case report

Case 1

A 32-year-old woman who was pregnant for the first time and who had known uterus didelphys, vaginal septum, and absent right kidney experienced urinary stress incontinence that was present during childhood but became more severe after her first pregnancy and uncomplicated vaginal delivery 4 years earlier. On examination, a fistulous opening was seen near the right of the external urethral meatus (Figure 1). Ultrasound examination confirmed the duplication of the uterus and cervix with bilateral normal ovaries. No kidney was visualized in the right renal fossa; the left kidney was 12 cm in length with no evidence of pelvicalyceal dilation. Intravenous pyelography showed rapid excretion from a normal left single renal pelvis, and computerized tomography was unable to locate any renal tissue on the right and confirmed the presence of mild left renal hypertrophy in urodynamic assessment. No detrusor