Ectopic Ureters and Ureteroceles in Dogs: Presentation, Cause, and Diagnosis*

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ABSTRACT:
Ectopic ureters are the most common congenital cause of urinary incontinence in dogs. Dogs with ectopic ureters usually present with urinary incontinence and are predominantly young females. Ectopic ureter is thought to occur because of disruption in normal embryogenesis and is commonly associated with other abnormalities of the urogenital tract. Diagnosis is usually made with contrast radiography and ultrasonography.

Although ectopic ureter is reportedly a rare condition in dogs, it is the most common congenital cause of urinary incontinence.¹ Ectopic ureter results from termination of one or both ureters at a site other than the trigone of the bladder.² Ureteroceles are cystic dilations of the intravesical submucosal portion of the distal ureter.³,⁴ They can be entirely within the bladder (intravesical or orthotopic ureteroceles) or in an abnormal position in association with an ectopic ureter (ectopic ureterocele).⁵,⁶

Other congenital anomalies capable of causing incontinence include congenital urethral sphincter incompetence and patent urachus.⁷,⁸ Congenital anomalies exhibit a wide range of anatomic variation and can occur alone or, more commonly, in conjunction with other abnormalities.¹,⁹ The high incidence of additional abnormalities justifies a thorough preoperative evaluation. Diagnostic tests available to determine the cause(s) of urinary incontinence include blood tests, urinalysis and urine culture, plain and contrast radiography, ultrasonography, cystoscopy, urodynamic measurements, computed tomography (CT), surgical exploration, or a combination of these tests. Surgery is the treatment of choice for ectopic ureters and ectopic ureteroceles.³,¹⁰ Persistent urinary incontinence is a common complication in dogs after surgical repair.²,⁶

This article focuses on the presentation, cause, and diagnosis of ectopic ureters and ureteroceles in dogs. Ureteroceles are discussed because of their similarities to ectopic ureters in presentation, diagnosis, and treatment.

SIGNALMENT, HISTORY, AND PHYSICAL EXAMINATION
Ectopic ureters are typically diagnosed in young female dogs (median age: 10 months) but can present at any age and in males.¹ Males tend to present later, with a median age of 24 months.¹ Females make up 89% to 95% of dogs diagnosed with ectopic ureters.¹,¹¹ This may be attributable to the ability to more readily identify urinary incontinence in female dogs.¹ It has

* A companion article on treatment appears on p. 311.
been suggested that the longer urethra of males is better able to oppose the distal flow of urine, which allows retrograde filling of the bladder. A similar pattern has been identified in humans, with about 90% of patients presenting before puberty and an incidence in females that is four times higher than that in males. Bilateral disease makes up 32% to 92% of canine ectopic ureter cases.

In 1984, a large North American study of 217 cases revealed seven breeds (i.e., Siberian husky, Newfoundland, bulldog, West Highland white terrier, fox terrier, miniature and toy poodle) to be at increased risk of ectopic ureter. In 1995, analysis of 175 cases from the United Kingdom found three breeds (i.e., Labrador retriever, golden retriever, Skye terrier) to be overrepresented. Based on these studies, familial genetics likely play a role in the cause of ectopic ureter.

Intermittent or continuous urinary incontinence since birth or weaning is the most frequently reported clinical sign in patients with ectopic ureter. Most dogs also display normal voiding patterns. Physical examination findings are often within normal limits, with the exception of moist or urine-stained hair in the perineal region. Urine scalding may cause secondary dermatitis, and owners may report frequent licking of the vulval or preputial area. Some dogs have vulvovaginitis, a vulvovaginal stricture, or a persistent hymen that can be detected digitally or with vaginoscopy. Dogs often have a history of recurrent urinary tract infections (UTIs), and some animals may have responded (partially or fully) to pharmacologic management of urethral sphincter mechanism incompetence.

Ectopic ureteroceles tend to present with urinary incontinence, and patients with ureteroceles often have a history similar to those with ectopic ureters. Patients with intravesical ureteroceles can have a variable presentation, including incontinence, dysuria, hematuria, chronic UTI, and complete or partial urinary obstruction; however, intravesical ureteroceles are often incidental discoveries with no associated signs. Many other differentials for urinary incontinence require consideration, including UTI, behavioral disorders, cystic or urethral calculi, neurogenic disorders, acquired urethral sphincter incompetence, en-
Partial urinary tract obstruction should also be considered because it may cause paradoxic incontinence.

**EMBRYOLOGY AND PATHOPHYSIOLOGY**

Understanding the embryologic development of the lower urinary tract helps explain the variability and complexity of the anomalies encountered in dogs.

**Normal Development**

Vertebrates have one of three distinct excretory organs: the pronephros, mesonephros, or metanephros. Some primitive fish still use the pronephros, amphibians appear to have replaced it with the mesonephros, and mammals and birds have developed the metanephros. During urogenital development, the pronephros, mesonephros, and metanephros appear in succession, and parts of each may be retained in a developing embryo. In mammals, only the duct of the pronephros is retained as the mesonephric duct. The mesonephros, which is temporarily active in canine fetuses, then becomes vestigial in females, and the mesonephric duct is retained as the deferent duct in males.

The metanephric duct, which is destined to become the ureter, is derived from a bud of the distal mesonephric duct close to the cloaca. Therefore, the mesonephric and metanephric ducts share a common excretory duct and opening when the bladder first forms (Figure 1). The metanephric duct continues to grow toward the metanephros, the tissue that forms the kidney. As the bladder grows, the common duct is absorbed and the mesonephric and metanephric ducts acquire individual openings (Figure 1). With further growth, the mesonephric ducts are displaced caudally and open on a prominence on the dorsal urethral wall while the ureteral openings remain in the bladder (Figure 1).

**Ectopic Ureters**

If the metanephric duct originates more cranially than normal on the mesonephric duct, the metanephric duct will not reach and establish an individual opening into the bladder. The metanephric duct is then carried caudally with the mesonephric duct to open in the urethra of females or the deferent duct or urethra of males.
in the bladder neck or urethra of females or the deferent duct or urethra of males\textsuperscript{19} (Figure 2). Most cases appear to terminate in the urethra of females and males.\textsuperscript{1} The reason for a ureter opening into the vagina, cervix, or uterus is less clear because these structures originate from the müllerian duct.\textsuperscript{19} Proposed explanations center on mesonephric structures linking into the müllerian duct system.\textsuperscript{19} Urinary incontinence may result because of the ectopic position of the ureteral orifice and/or disruption of the smooth muscle layer of the proximal urethral sphincter mechanism by the submucosal ureteral tunnel\textsuperscript{9} (Figure 2).

In contrast to dogs, 80% of humans with ectopic ureters and ureteroceles have a duplex kidney system in which the anterior and posterior portions of the kidneys are drained by two independent metanephric ducts.\textsuperscript{12} In humans with a duplex kidney system and ectopic ureter, the more cranial of the two metanephric ducts fails to reach the bladder.\textsuperscript{18} The cranial metanephric duct is subsequently carried caudally with the mesonephric duct into an ectopic position.\textsuperscript{18} In humans, the associated renal unit is often dysplastic and provides little if any function. This is thought to be a consequence of recurrent UTIs or abnormal interplay between the metanephric duct and developing renal tissue.\textsuperscript{12}

**Ureteroceles**

The embryologic origin of ureteroceles remains unknown.\textsuperscript{6} Proposed mechanisms include a stenotic ureteral opening, delayed fusion of the metanephric duct and urogenital sinus, arrested myogenesis of the distal ureter, and congenital weakness of the ureteral connective tissue.\textsuperscript{3}

**DIAGNOSIS**

**Clinical Pathology and Microbiology**

A complete blood cell count, serum chemistry profile, and urinalysis (with specific gravity, microbial culture, and sensitivity) should be conducted.\textsuperscript{16} In cases of ectopic ureter and ureterocele, results of the hematology and serum biochemistry evaluations are generally within normal limits.\textsuperscript{6} The exception is the patient with associated abnormalities of the upper urinary tract that has diminished renal function.\textsuperscript{8} Renal insufficiency may be present because of chronic pyelonephritis, obstructive uropathy, or concurrent congenital abnormalities.\textsuperscript{16} Concomitant UTIs are a common problem and have a reported incidence of 64% in cases of ectopic ureter.\textsuperscript{13,16} Obtaining a cystocentesis sample for urinalysis and culture may be difficult because of the small bladder size sometimes associated with continuous urinary incontinence.\textsuperscript{8} In these cases, we have had good success with ultrasound-guided cystocentesis. An alternative approach is to use a free-catch urine sample for urinalysis and specific gravity and to obtain a cystocentesis sample for culture at surgery.\textsuperscript{8}

**Radiography**

Traditionally, diagnosis of congenital anomalies in the lower urinary tract has focused on plain and contrast radiographic techniques, including IV urography, positive-contrast cystography, double-contrast cystography, retrograde urethrography, vaginocystography, and fluoroscopy.\textsuperscript{2,6,8,9,13,20}

Plain abdominal radiography can be used to assess the size, shape, and location of the kidneys and bladder and to help identify radiodense calculi.\textsuperscript{8}

IV urography has long been the method of choice in diagnosing ectopic ureters.\textsuperscript{2,8,9,12,21} The technique appears to have much greater sensitivity when combined with pneumocystography.\textsuperscript{9} Preparation for IV urography is an involved process: Dogs must be fasted for 24 hours, receive an enema, and have a normal hydration status.\textsuperscript{22} IV urography aids in identifying unilateral versus bilateral disease as well as the location, size, and morphology of the ureter and ureterovesicular junction.\textsuperscript{8} The technique can also be used to evaluate the upper urinary tract.\textsuperscript{8} In general, the renal pelvis and pelvic recesses in dogs do not exceed 1 to 2 mm in diameter, and the proximal ureter in dogs does not exceed 2 to 3 mm in diameter.\textsuperscript{22} The normal contrast-enhanced ureterovesicular junction in dogs produces a “J” or “hook” shape on a radiograph.\textsuperscript{9} An alteration (from a “J” shape to a straight line) in the angle that the ureter forms with the bladder is highly suggestive of ectopic ureter, even when the exact site of termination is not seen? (Figure 3). Lateral and ventrodorsal radiographic views are necessary to identify the terminal segment of nondilated ureters in the pelvic region.\textsuperscript{8} The accuracy of using ureterovesicular junction shape to diagnose ectopic ureter is 76%, with a sensitivity of 83%.\textsuperscript{9} Not visualizing a ureteral segment is usually normal because of waves of peristalsis.\textsuperscript{22} However, the segment of the ureter should be visualized at some time in the sequence of radiographs.\textsuperscript{22} This problem may be overcome with fluoroscopy, which can provide a continuous radiographic image of the ureterovesicular junction. Bladder size and position are difficult to evaluate because of lack of control of bladder distention when using IV urography and the tendency toward overdistention when using pneumocys-
Urography.\textsuperscript{1,8} Superimposition of surrounding structures may prevent visualization of the ureters.\textsuperscript{2,21} Retrograde filling of the bladder can occur from displaced ureters, which may obscure specific identification of the ureteral orifice.\textsuperscript{8} Therefore, diagnosis of ureteral ectopia often remains elusive after an IV urogram.\textsuperscript{2,8,9,13,21}

Retrograde urethrogram has limited value because of the presence of the catheter within the urethral lumen, which may obscure or obstruct a displaced ureteral orifice.\textsuperscript{8} However, retrograde vaginocystography is performed without a catheter in the urethral lumen and has been shown to be very useful in locating and evaluating the terminal orifice of the ectopic ureter, the entire length of the urethra, and the vaginal contour.\textsuperscript{8}

Contrast radiography allows correct prediction of the location of an ectopic ureter in 62\% to 77\% of cases; therefore, whenever available, adjunctive diagnostic modalities should be considered.\textsuperscript{9,10}

Additional abnormalities are detected in 70\% to 94\% of dogs undergoing ectopic ureter investigation.\textsuperscript{1,9} Abnormalities are most common in the kidneys, urinary collecting system, and ureters.\textsuperscript{9} Hydronephrosis is the most commonly reported abnormality associated with ectopic ureters and ureteroceles.\textsuperscript{1,4,9} Other abnormalities include absent, nonvisualized, small, or irregular kidneys and renal pelvis dilation due to pyelonephritis or hydronephrosis.\textsuperscript{9,23}

Investigating a suspected ureterocele requires the same radiographic protocol already outlined.\textsuperscript{3–6} Excretory urography is the preferred method of diagnosis, with vaginocystography being less reliable.\textsuperscript{3} Ureteroceles can be diagnosed with excretory urography as either a positive “cobra-head” dilation within the bladder or a negative filling defect in a cystogram related to impaired renal function.\textsuperscript{3} A functional classification system has been proposed for ureteroceles:\textsuperscript{5}

- Grade 1—No concurrent ureteral or renal disease
- Grade 2—Unilateral ureteral or renal disease
- Grade 3—Bilateral ureteral or renal disease

The system may help provide a prognosis for ureterocele cases (grade 1 has a more favorable prognosis than grade 3), lead to improved organization and reporting of future cases, and allow better comparisons between treatment success in human and canine ureteroceles.\textsuperscript{5} In our opinion, however, the small number of published canine ureterocele cases currently limits the usefulness of this grading system in veterinary medicine.

**Ultrasonography**

Ultrasonography is a practical and useful diagnostic test for ectopic ureter in dogs.\textsuperscript{20} In experienced hands, contrast radiography and ultrasonography have closely correlated results and similar sensitivities.\textsuperscript{20} Ultrasonographic diagnostic features include absence of normal urine flow from the ureter into the bladder (ureteral jet) or the ability to trace a distended ectopic ureter distally to the urethra\textsuperscript{21} (Figure 4). The small diameter of a normal ureter prevents visualization with ultrasonography.\textsuperscript{21} Ureteroceles can be identified with ultrasonography as smooth, thin-walled cystic structures that project into the bladder lumen or occur within the bladder wall.\textsuperscript{21,24,25} (Figure 5).
Ultrasonography has benefits in investigating the upper urinary tract.\textsuperscript{24,26} It provides information on the size, shape, and internal architecture of the kidneys.\textsuperscript{24} Doppler ultrasonography provides additional functional information by measuring renal blood flow parameters.\textsuperscript{24}

However, ultrasonography has limitations. Overlying bone may obscure imaging of an intrapelvic bladder neck, and ureteral jets are not always visible in normal dogs or those with ureteral infection or obstruction.\textsuperscript{24} Despite certain technical difficulties, ultrasonography has accuracy comparable with contrast radiography; however, ultrasonography takes less time and avoids the need for multiple radiographs or image intensification to identify the termination of the ureters.\textsuperscript{20} Nevertheless, for a full assessment, ultrasonography must be used in combination with appropriate radiographic procedures.\textsuperscript{24}

Computed Tomography
Contrast-enhanced CT may be a superior imaging procedure in identifying ectopic ureters in dogs.\textsuperscript{24} Because CT is not affected by superimposition, it allows better visualization of the ureterovesical junction than does traditional IV urography.\textsuperscript{21} Patient preparation is also minimized.\textsuperscript{21} The ability to conduct two-dimensional multiplanar and three-dimensional graphic reconstructions facilitates both accurate diagnosis and surgical planning.\textsuperscript{21} However, images of the ureterovesical junction are still limited by peristaltic contractions; therefore, multiple scans of the ureterovesical junction may be required.\textsuperscript{21} With operator experience, contrast-enhanced CT should reduce total procedure time and x-ray exposure compared with IV urography and multiple positional radiographs.\textsuperscript{21} Clinical use of this technology in veterinary medicine is still at an early stage, but initial results are promising.\textsuperscript{21}

Cystoscopy
In certain referral institutions, endoscopic examination of the vagina and lower urinary tract has become a routine part of the diagnostic approach in patients with clinical signs of lower urinary tract disease.\textsuperscript{27} Cystoscopy should be performed using general anesthesia and is appropriate in adult dogs and puppies heavier than 6.6 lb (3 kg).\textsuperscript{8} Rigid human adult and pediatric cystoscopes can be used in female dogs, whereas males require flexible cystoscopes.\textsuperscript{27} Use of cystoscopy has dramatically improved the diagnosis and classification of ectopic ureters and associated congenital abnormalities of the ureteral orifices, bladder, urethra, and vagina.\textsuperscript{8} It is indicated to confirm questionable lesions found following radiographic and ultrasonographic imaging of the lower urinary tract, especially when surgical exploration cannot be justified.\textsuperscript{27} Cystoscopy can be used to identify the specific morphology of the terminal segment of the ureter, location of the ureteral orifice, and flow through the opening.\textsuperscript{27} Use of cystoscopy can also provide a general appreciation of other abnormalities in the wall or mucosa of the bladder.\textsuperscript{27} A retrospective study of 25
female dogs presented for ectopic ureter revealed a 100% correlation between the results of the cystoscopic examination and the findings at surgery. Concurrent vestibular abnormalities (e.g., paramesonephric septal remnants, hymenal remnants, vestibulovaginal stenosis) were noted in all of the cystoscopic cases reviewed. Potential complications of the procedure include trauma and UTI (Figure 6).

Urodynamic Evaluation

Urethral pressure profiles provide a depiction of intrarethral pressures generated along the length of the urethra. The profiles are measured by using catheter-tip pressure transducers or a fluid-perfusion technique. Pressure profiles can be used to diagnose urethral incompetence. Urethral pressure profilometry requires specialized equipment that must be standardized by each user; however, it is not usually required for routine diagnostic investigations, and its use is limited to referral institutions. A study in which dogs with ectopic ureter were urodynamically assessed correctly predicted continence following surgery in eight of nine dogs, suggesting the potential to detect concurrent functional abnormalities of the bladder and urethra and the likely success of surgical correction. The usefulness of urethral pressure profilometry in cases of ectopic ureters has been questioned. Technically, an ectopic ureter may accidentally be catheterized, and when instruments are used to guide the catheter, they may interfere with resting values. The profile is also hard to interpret with the mechanical interference of the ectopic ureter passing across the proximal urethra. With surgical removal of this tissue, urethral function may change postoperatively.

CONCLUSION

Ectopic ureter is rare in dogs but is the most common congenital anomaly that causes urinary incontinence. The disease is thought to arise because of disruption in normal embryogenesis and is commonly associated with other anomalies within the urogenital tract. Most clinical cases involve young female dogs with a history of urinary incontinence, although wide variation has been documented. Ultrasonographic examination of the kidneys and ureters combined with cystoscopic examination of the lower urinary tract probably provides the most comprehensive and accurate assessment of suspected ectopic ureter. However, the diagnostic protocols should be tailored to the expertise and diagnostic equipment available to the clinician. Plain and contrast radiography are probably the most accessible diagnostic tools available to general practitioners. However, the discord between radiographic and surgical findings, even in the hands of board-certified radiologists, should be appreciated.

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REFERENCES

ARTICLE #4 CE TEST

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1. What is the most common congenital anomaly of the urinary tract that causes urinary incontinence in dogs?
   a. ureterocele
   b. patent urachus
   c. cystic diverticulum
   d. ectopic ureter
   e. congenital urethral sphincter mechanism incompetence

2. Ectopic ureter is most commonly diagnosed in
   a. young male dogs.
   b. young female dogs.
   c. young dogs and equally in both sexes.
   d. old male dogs.
   e. old female dogs.

3. Patients with ectopic ureter may present with a history of
   a. perivulvar or preputial dermatitis.
   b. vulvovaginitis.
   c. urinary incontinence.
   d. ascending UTI.
   e. any of the above

4. The metanephric duct (ureter) buds from the
   a. mesonephric duct.
   b. metanephric kidney.
   c. cloaca.
   d. urinary bladder.
   e. mesonephric kidney.

5. Canine ectopic ureters are thought to most commonly arise because of
   a. a second, more cranial metanephric duct.
   b. incomplete absorption of the common mesonephric and metanephric ducts.
   c. a stenotic ureteral opening.
   d. congenital weakness of urethral connective tissue.
   e. failure of the mesonephric duct to caudally displace in the bladder wall.

6. UTIs have reportedly occurred concurrently in ___% of ectopic ureter cases.
   a. 10
   b. 42
   c. 64
   d. 71
   e. 89

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7. The longstanding imaging method of choice to diagnose ectopic ureters and ureteroceles is
   a. ultrasonography.
   b. IV urography.
   c. vaginocystography.
   d. retrograde urethrography.
   e. contrast-enhanced CT.

8. Contrast radiography allows correct prediction of ectopic ureter in ________ of cases.
   a. 42% to 67%  
   b. 62% to 77%  
   c. 70% to 94%  
   d. 89% to 95%  
   e. none of the above

9. Ultrasonography of patients with ectopic ureter allows visualization of
   a. ureteral jets.
   b. distended ureters.
   c. ureteroceles.
   d. the internal architecture of the kidneys.
   e. all of the above

10. Cystoscopy has been shown to have a ____% correlation with surgical findings.
    a. 90  
    b. 92  
    c. 93  
    d. 95  
    e. 100