

# Urachal Remnant Diseases: Spectrum of CT and US Findings<sup>1</sup>

## CME FEATURE

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## LEARNING OBJECTIVES FOR TEST 5

After reading this article and taking the test, the reader will be able to:

- Describe the basic embryologic and anatomic features of the urachus.
- Identify the four types of congenital urachal anomalies and their CT and US features.
- Recognize the manifestations of an infected urachal remnant versus urachal carcinoma at CT and US.

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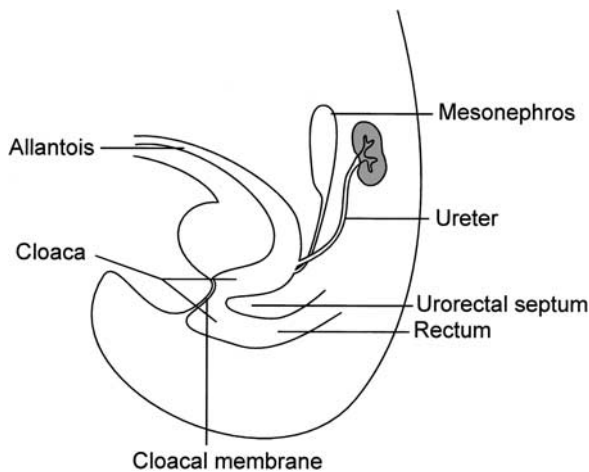
Computed tomography (CT) and ultrasonography (US) are ideally suited for demonstrating urachal remnant diseases. A patent urachus is demonstrated at longitudinal US and occasionally at CT as a tubular connection between the anterosuperior aspect of the bladder and the umbilicus. An umbilical-urachal sinus manifests at US as a thickened tubular structure along the midline below the umbilicus. A vesicourachal diverticulum is usually discovered incidentally at axial CT, appearing as a midline cystic lesion just above the anterosuperior aspect of the bladder. At US, it manifests as an extraluminally protruding, fluid-filled sac that does not communicate with the umbilicus. Urachal cysts manifest at both modalities as a noncommunicating, fluid-filled cavity in the midline lower abdominal wall located just beneath the umbilicus or above the bladder. Both infected urachal cysts and urachal carcinomas commonly display increased echogenicity at US and thick-walled cystic or mixed attenuation at CT, making it difficult to differentiate between them. Percutaneous needle biopsy or fluid aspiration is usually needed for diagnosis and therapeutic planning. Nevertheless, CT and US can help identify most disease entities originating from the urachal remnant in the anterior abdominal wall. Understanding the anatomy and the imaging features of urachal remnant diseases is essential for correct diagnosis and proper management.

**Index terms:** Fetus, abnormalities, 839.1462, 839.1463, 839.1491 • Fetus, genitourinary system, 839.1462, 839.1463, 839.1491 • Fetus, growth and development, 839.1462, 839.1463, 839.1491 • Urachus, 839.20, 839.31, 839.324

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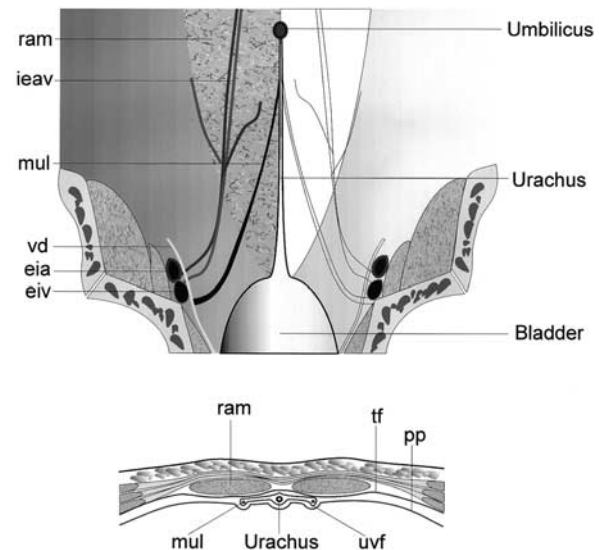


**Figure 1.** Drawing illustrates the continuity of the ventral cloaca and the allantois in an 8-week-old fetus.

### Introduction

The urachus, or median umbilical ligament, is a midline tubular structure that extends upward from the anterior dome of the bladder toward the umbilicus. It is a vestigial remnant of at least two embryonic structures: the cloaca, which is the cephalic extension of the urogenital sinus (a precursor of the fetal bladder), and the allantois, which is a derivative of the yolk sac (1). The tubular urachus normally involutes before birth, remaining as a fibrous band with no known function. However, persistence of an embryonic urachal remnant can give rise to various clinical problems, not only in infants and children but also in adults. Because urachal remnant diseases are uncommon and manifest with nonspecific abdominal or urinary signs and symptoms, definitive presurgical diagnosis is not easily made. Various abnormalities can be confusing unless one is familiar with the basic embryologic anatomy and imaging features of the subumbilical and prevesical region. Because computed tomography (CT) and ultrasonography (US) display cross-sectional images and the urachus in the anterior abdominal wall is located away from interfering intestinal structures, these modalities are ideally suited for demonstrating urachal anomalies (2–5).

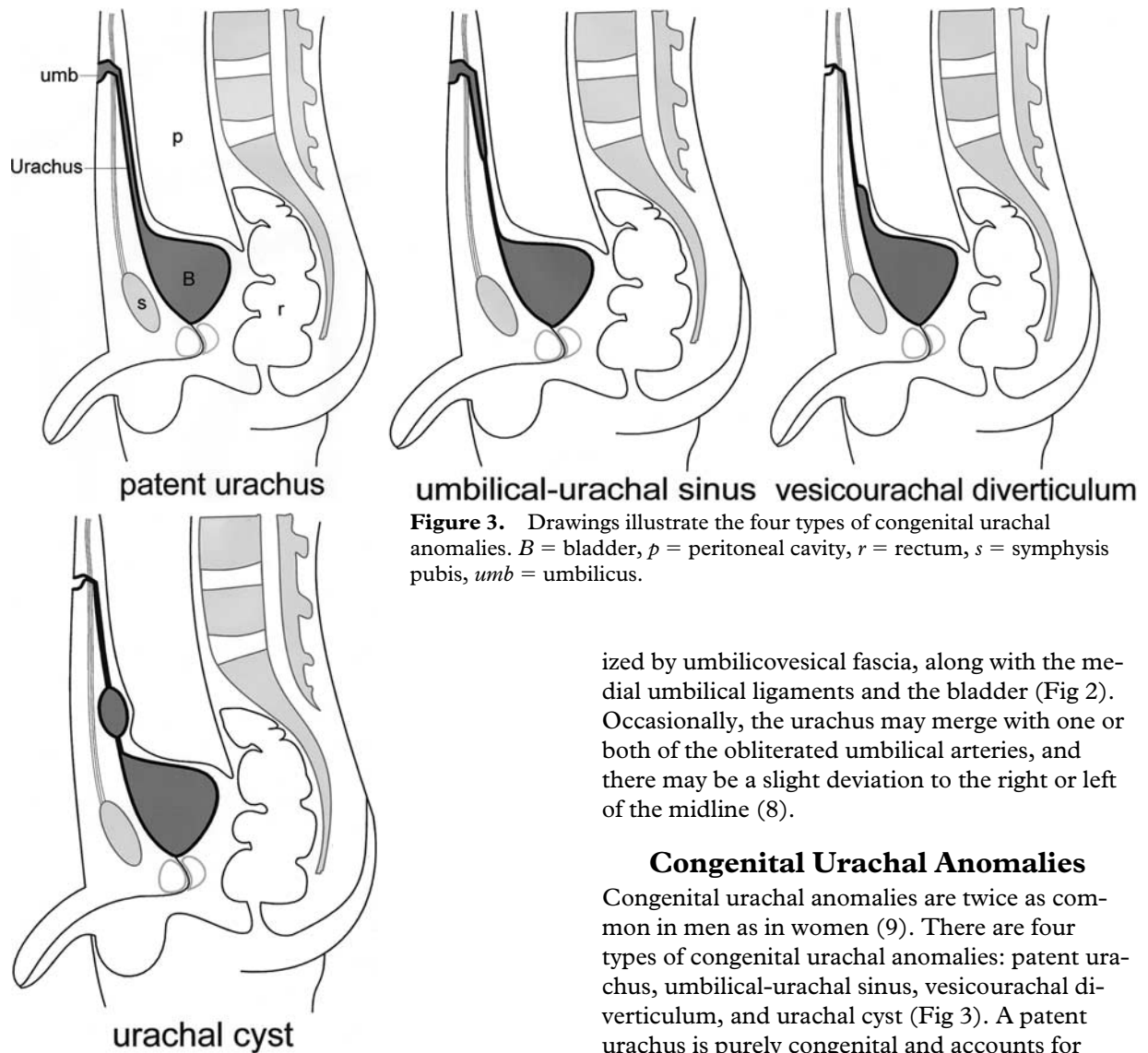
In this article, we review the embryologic and anatomic features of the urachus. We also discuss and illustrate the spectrum of CT and US findings in both congenital urachal anomalies (patent urachus, umbilical-urachal sinus, vesicourachal diverticulum, urachal cyst) and acquired urachal remnant diseases (infection, neoplasm).



**Figure 2.** Drawings of the lower anterior abdominal wall as seen from inside the peritoneal cavity (top) and in the transverse plane (bottom) show the urachus extending from the dome of the bladder to the umbilicus along with the medial umbilical ligaments (obliterated umbilical arteries) (*mul*), which lie within the perivesical space between the transverse fascia (*tf*) and the parietal peritoneum (*pp*) and are surrounded by the umbilicovesical fascia (*uvf*). *eia* = external iliac artery, *eiv* = external iliac vein, *ieav* = inferior epigastric artery and vein, *ram* = rectus abdominus muscle, *vd* = vas deferens.

### Embryology and Normal Anatomy

The allantois appears on about day 16 as a tiny, finger-like outpouching from the caudal wall of the yolk sac (1). The bladder develops from the ventral portion of the expanded terminal part of the hindgut, the cloaca, which is contiguous with the allantois ventrally (Fig 1). The cranioventral end of the bladder opens into the allantois at the level of the umbilicus; thus, the bladder initially extends all the way to the umbilicus. By the 4th or 5th month of gestation, the bladder descends into the pelvis and its apical portion progressively narrows to a small, epithelialized fibromuscular strand, the urachus (1). In late embryonic and fetal life and early postnatal life, the urachal portion, which is still microscopic, fails to grow; thus, its lumen remains narrow and is usually obliterated by fibrous proliferation. In one-third of adults, it may be visible at microscopic examination as a structure communicating with the lumen of the bladder; however, in terms of function it can be considered closed by the latter half of fetal life (6).



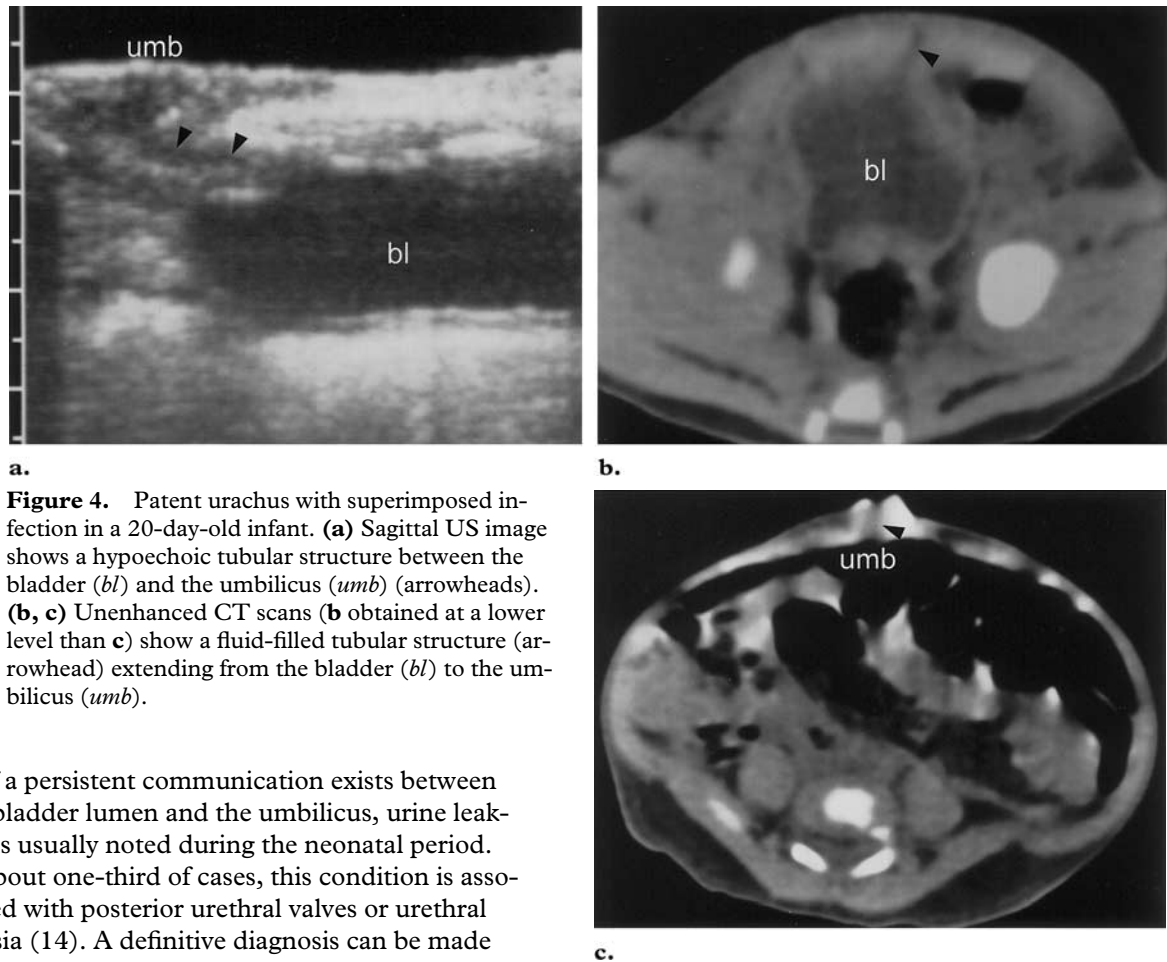
**Figure 3.** Drawings illustrate the four types of congenital urachal anomalies. *B* = bladder, *p* = peritoneal cavity, *r* = rectum, *s* = symphysis pubis, *umb* = umbilicus.

ized by umbilicovesical fascia, along with the medial umbilical ligaments and the bladder (Fig 2). Occasionally, the urachus may merge with one or both of the obliterated umbilical arteries, and there may be a slight deviation to the right or left of the midline (8).

### **Congenital Urachal Anomalies**

Congenital urachal anomalies are twice as common in men as in women (9). There are four types of congenital urachal anomalies: patent urachus, umbilical-urachal sinus, vesicourachal diverticulum, and urachal cyst (Fig 3). A patent urachus is purely congenital and accounts for about 50% of all cases of congenital anomalies (10). An umbilical-urachal sinus (representing about 15% of cases), vesicourachal diverticulum (about 3%–5%), or urachal cyst (about 30%) may close normally after birth but then reopen in association with pathologic conditions that are often categorized as acquired diseases (7,10–13). The majority of patients with urachal abnormalities (except those with a patent urachus) are asymptomatic. However, they may become symptomatic if these abnormalities are associated with infection.

The urachus varies from 3 to 10 cm in length and from 8 to 10 mm in diameter. It is a three-layered tubular structure, the innermost layer being lined with transitional epithelium in 70% of cases and with columnar epithelium in 30%. The structure is surrounded by connective tissue and an outermost muscular layer in continuity with the detrusor muscle (6,7). Along its path from the bladder to the umbilicus, the urachus lies between the transverse fascia and the parietal peritoneum contained in the pyramidal, retropubic, preperitoneal perivesical space compartmental-

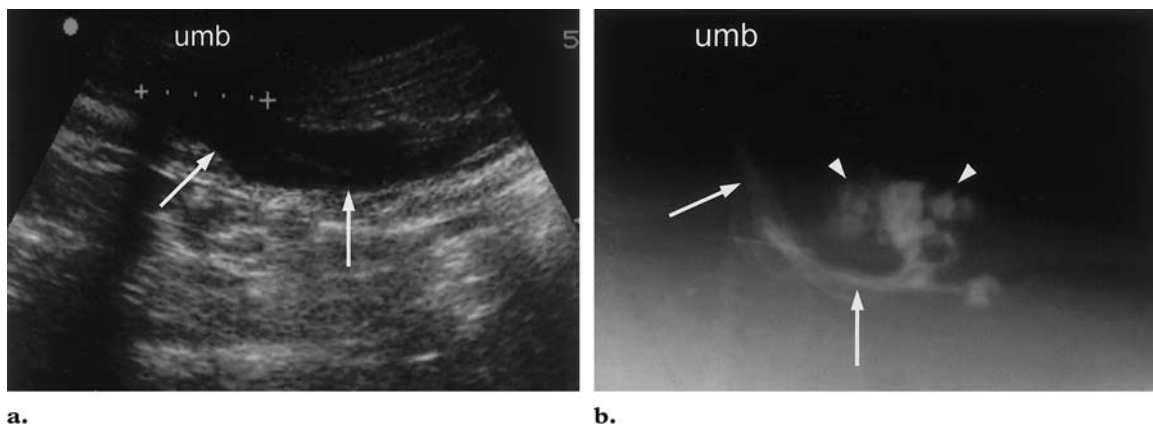


**Figure 4.** Patent urachus with superimposed infection in a 20-day-old infant. **(a)** Sagittal US image shows a hypoechoic tubular structure between the bladder (*bl*) and the umbilicus (*umb*) (arrowheads). **(b, c)** Unenhanced CT scans (**b** obtained at a lower level than **c**) show a fluid-filled tubular structure (arrowhead) extending from the bladder (*bl*) to the umbilicus (*umb*).

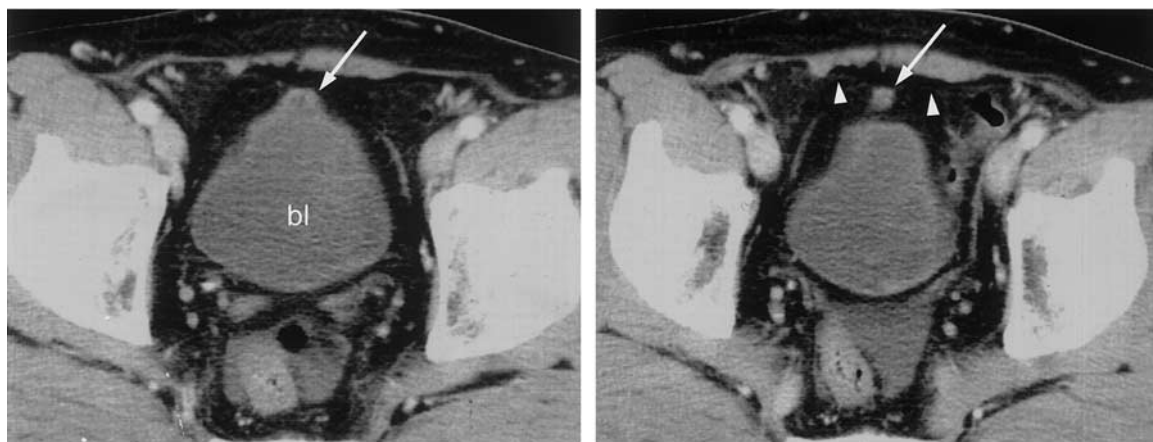
If a persistent communication exists between the bladder lumen and the umbilicus, urine leakage is usually noted during the neonatal period. In about one-third of cases, this condition is associated with posterior urethral valves or urethral atresia (14). A definitive diagnosis can be made with sinography or cystography (15,16). Patent urachus as a tubular connection between the anterosuperior aspect of the bladder and the umbilicus is demonstrated at longitudinal US and occasionally at CT performed in infants during the bladder-filling stage (Fig 4). Some patients with patent urachus are asymptomatic, and sometimes an acquired obstructive lesion of the lower urinary tract may result in umbilical-urinary fistulas (7).

Umbilical-urachal sinus consists of blind dilatation of the urachus at the umbilical end. A small opening into the umbilicus is generally present and may result in periodic discharge (7,10). A thickened tubular structure along the midline below the umbilicus can be visualized at US (Fig 5a). It is usually associated with an infection of the urachal remnant and confirmed at sinography (Fig 5b).

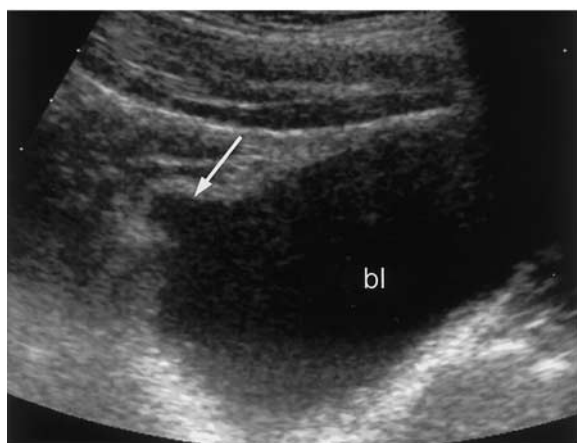
In vesicourachal diverticulum, the urachus communicates only with the bladder dome. This condition results when the vesical end of the urachus fails to close. Vesicourachal diverticulum is asymptomatic in most cases and is usually discovered incidentally at axial CT performed for unrelated reasons, appearing as a midline cystic lesion just above the anterosuperior aspect of the bladder (Fig 6a, 6b) (17). US readily demonstrates an extraluminally protruding, fluid-filled sac that does not communicate with the umbilicus (Fig 6c). This lesion tends to be found in patients with chronic bladder outlet obstruction and may be complicated by urinary tract infection, intraurachal stone formation, and an increased prevalence of carcinoma after puberty (18). In infants, vesicourachal diverticulum is commonly accompanied by prune-belly syndrome (9).



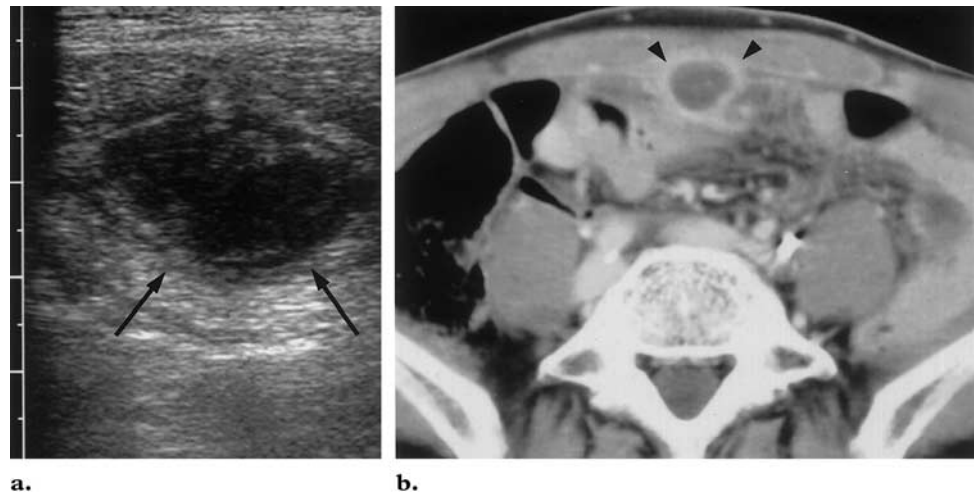
**a.** **b.**  
**Figure 5.** Infected umbilical-urachal sinus in a 19-year-old man. **(a)** Sagittal US image shows a hypoechoic tubular structure (arrows) extending from the umbilicus (*umb*) (cursors) just beneath the anterior abdominal wall. The caudal end of this tubular structure is obliterated. **(b)** Cross-table lateral sinogram shows a blind sinus tract (arrows) with no communication with the bladder. The complex caudal end of the sinus tract (arrowheads) suggests superimposed infection and fistula formation through the anterior abdominal wall. *umb* = umbilicus.



**a.** **b.**  
**Figure 6.** Vesicourachal diverticulum as an incidental finding in a 58-year-old man. **(a, b)** Axial CT scans (**a** obtained at a lower level than **b**) demonstrate a small, anterosuperior outpouching (arrow) representing a urachal diverticulum arising from the apex of the bladder (*bl*). The umbilicovesical fascia (arrowheads in **b**) allows localization of the urachus in the extraperitoneal perivesical space. **(c)** Sagittal US image shows a localized hypoechoic outpouching (arrow) communicating with the uppermost portion of the bladder (*bl*), thereby helping confirm the CT findings.



**c.**



**Figure 7.** Infected urachal cyst in a 55-year-old man. **(a)** Transverse US image shows a complex cystic lesion lying along the course of the urachus midway between the bladder and the umbilicus (arrows). The lesion demonstrates mixed internal echogenicity, a thick outer wall, and a shaggy inner margin. **(b)** Contrast material-enhanced CT scan shows a strongly enhancing, thick-walled cystic lesion with perilesional infiltration just beneath the abdominal wall in the midline (arrowheads).

A urachal cyst develops if the urachus closes at both the umbilicus and the bladder but remains patent between these two endpoints. It occurs primarily in the lower one-third of the urachus and less frequently in the upper one-third (19). Urachal cysts are usually small but vary considerably in size (9). They become symptomatic when they enlarge but are sometimes found as incidental masses during routine examination (20). CT or US shows a fluid-filled cavity in the midline lower abdominal wall (12,16,21–23). Eggshell calcification of the cyst wall is rarely reported (24). As with other urachal anomalies, infection is the most common complication of urachal cyst, and the majority of cysts are infected at the time of diagnosis (12,19,21,25,26). Superinfected urachal cyst manifests as wall thickening and demonstrates an attenuation higher than that of water at CT and soft-tissue components and mixed echogenicity at US (Fig 7).

### Acquired Urachal Remnant Diseases

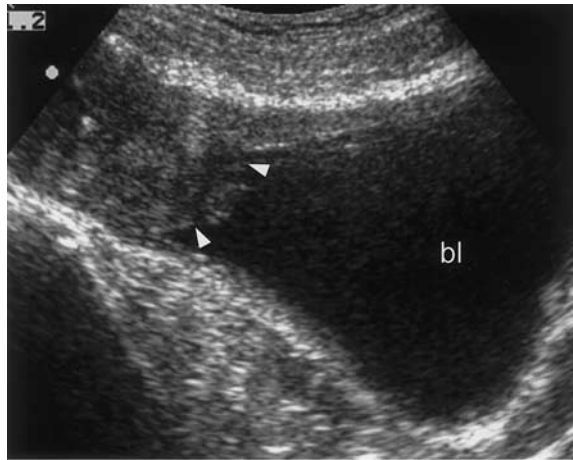
#### Infection

Urachal tract remnants that abnormally remain patent are often subject to infection. Furthermore, these infected remnants are frequently confused with a wide spectrum of midline intraab-

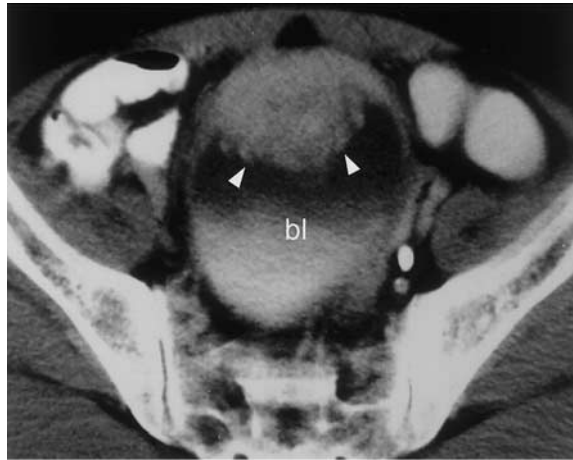
dominal or pelvic inflammatory disorders at clinical examination and with malignant tumors at imaging (12,19,26). The route of infection may be lymphatic, hematogenous, or vesical, and a wide variety of gram-positive and gram-negative micro-organisms have been cultured from infected urachal remnants (27). Depending on the variations in the patency of the urachal lumen, the drainage of infected fluid along the urachus can determine the acquired forms of umbilical-urachal sinus (drainage through the umbilicus), vesicourachal diverticulum (drainage through the bladder), or patent urachus or alternating sinus (drainage in both directions) (9). Rarely, spontaneous rupture of an infected cyst into the abdominal cavity leads to localized or generalized peritonitis (28). Complex echogenicity at US and inhomogeneous attenuation with variable contrast enhancement in and around the disease process at CT make it difficult to differentiate an infected urachal remnant from urachal carcinoma (Fig 8). In the majority of cases, percutaneous needle biopsy or fluid aspiration is mandatory for diagnosis and therapeutic planning (12,19,25, 29). Total removal of the cyst wall is essential because there is a 30% reinfection rate (25) and carcinoma may develop in an unresected or incompletely resected urachal remnant (9,10).

#### Tumors

Benign urachal neoplasms including adenomas, fibromas, fibroadenomas, fibromyomas, and hamartomas are extremely rare; however, they are

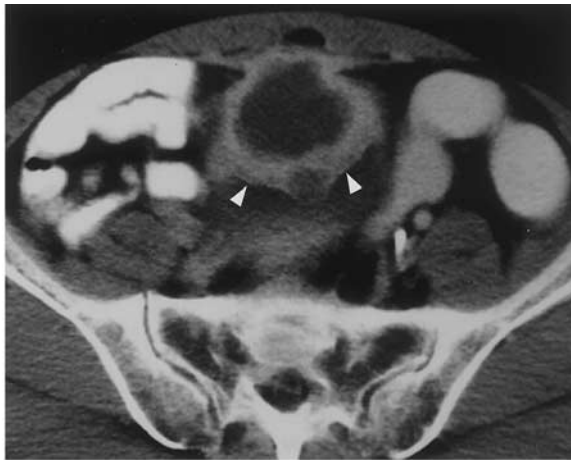


a.



c.

important in that they mimic urachal malignancy (30–32). Malignant urachal neoplasms are also rare, representing less than 0.5% of all bladder cancers (33). Although the normal urachus is most commonly lined by the transitional epithelium, urachal carcinoma predominantly manifests as adenocarcinoma (90% of cases), probably due to the metaplasia of the urachal mucosa into columnar epithelium followed by malignant transformation; conversely, 34% of bladder adenocarcinomas are of urachal origin (33,34). At histologic analysis, mucin production is found in up to 75% of cases. A minority of urachal carcinomas



b.

**Figure 8.** Organized abscess in the urachal remnant in a 53-year-old man. **(a)** Sagittal US image shows a mass with mixed echogenicity (arrowheads) contiguous with the dome of the bladder (*bl*). **(b, c)** Contrast-enhanced CT scans (**c** obtained at a lower level than **b**) demonstrate a strongly enhancing mass (arrowheads) with internal fluid attenuation involving the ventrocranial portion of the bladder (*bl*). Adjacent bladder wall thickening is also seen. Preoperative differentiation of an infected urachal remnant from urachal carcinoma was not possible. An organized abscess was confirmed at surgery.

demonstrate transitional, squamous, or anaplastic cell characteristics (33,35–37). These tumors are most commonly seen in patients 40–70 years of age, two-thirds of whom are men (38).

Urachal tumors are typically silent because of their extraperitoneal location; consequently, the majority of patients exhibit local invasion or metastatic disease at presentation (33,39). Ninety percent of urachal carcinomas arise in the juxtavesical portion of the urachus and extend superiorly toward the umbilicus and inferiorly through the bladder wall (Fig 9). Urachal carcinomas may be confused with primary tumors of the bladder dome; unlike vesical tumors, however, urachal



**a.**  
**Figure 9.** Mucinous adenocarcinoma arising from the urachal remnant in a 38-year-old woman.

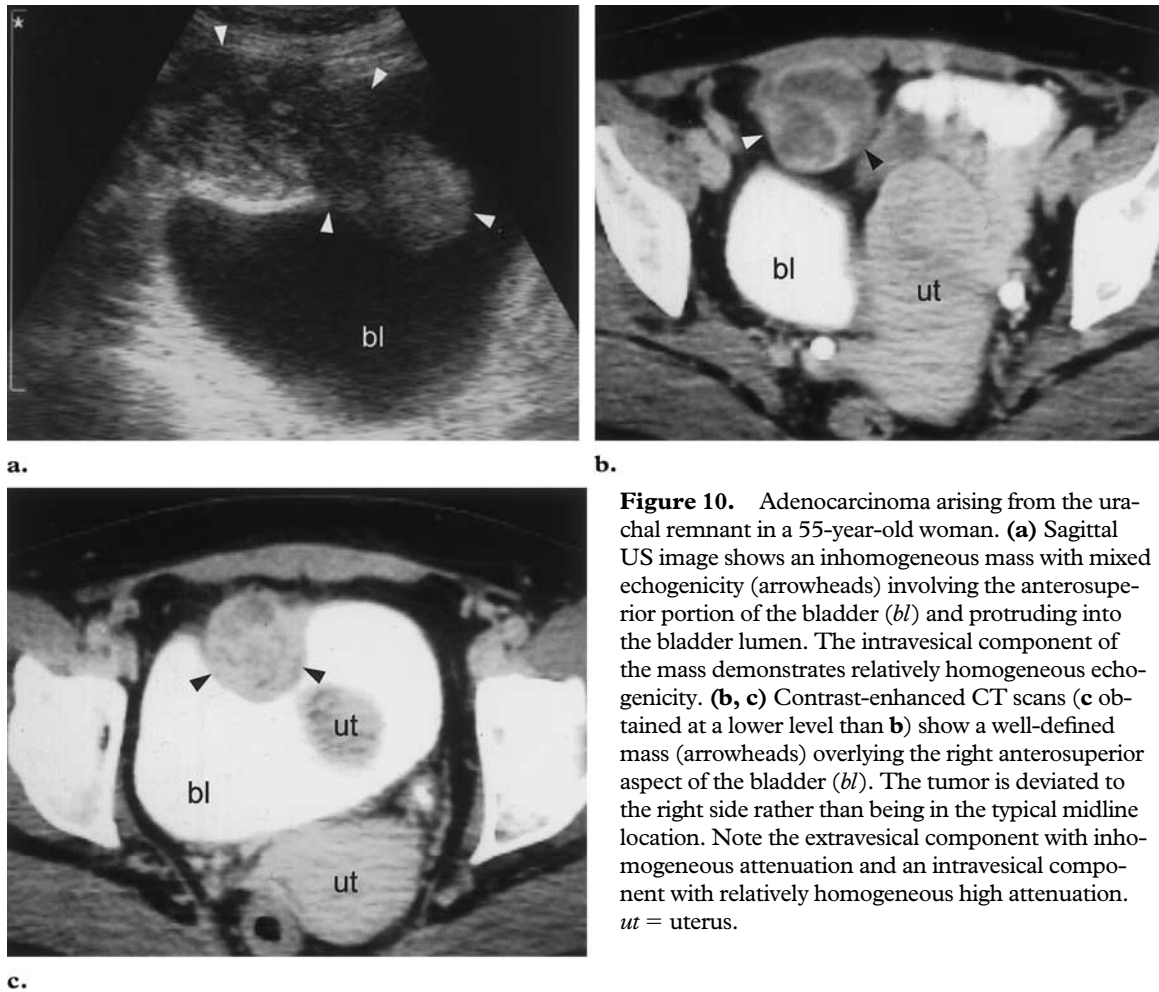
(a) Sagittal US image shows a mass with mixed echogenicity and foci of increased echogenicity (arrowheads) involving the dome of the bladder (*bl*). (b) Contrast-enhanced CT scan shows a low-attenuation mass with thin, curvilinear calcifications peripherally (arrowheads). *ut* = uterus. (c) CT scan obtained 2 cm lower than **b** shows a solid, high-attenuation mass with a smooth posterior border (arrowheads) contiguous with the dome of the bladder (*bl*). Small, punctate calcifications are also seen in the center of the lesion.

tumors have a propensity to grow in the perivesical space toward the umbilicus (Figs 9, 10). A primary bladder carcinoma arising in the mucosa of the bladder apex will usually manifest with less of an extravesical component than is seen in urachal cancer. The minority of urachal carcinomas are located in the middle of the urachus (6% of cases) or near the umbilical end (4%) (35). Some tumors deviate toward the right or left paramedian portion from the midline (Fig 10); this is because, during embryologic development, the urachus occasionally deviates from the midline to merge with one of the obliterated umbilical arteries (8,9). The direction of tumor growth or local invasion influences the deviation from the midline in such cases.

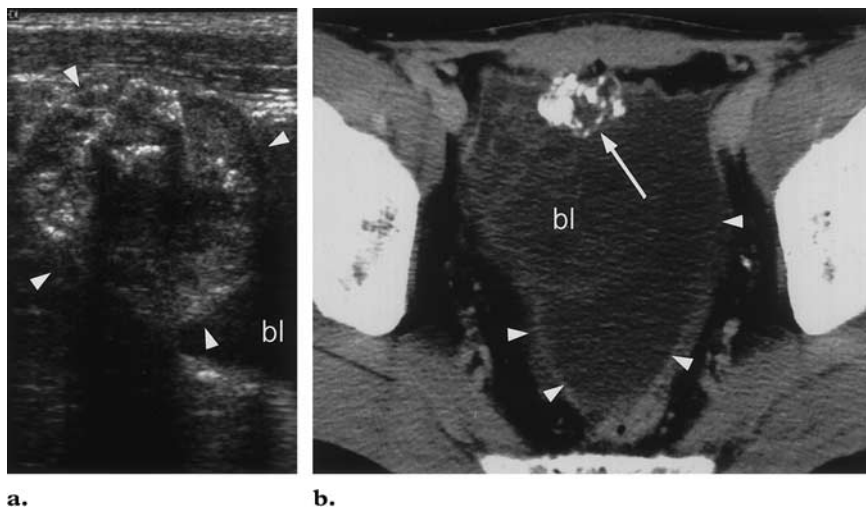
As with some other mucinous adenocarcinomas of the abdominal organs, urachal carcinomas may produce typical psammomatous calcifications that are well depicted at CT (5,9). Calcification occurs in 50%–70% of cases and may be punctate, stippled, or curvilinear and peripheral (Figs 9, 11) (8,40–42). Calcifications in a midline supravescical mass are considered nearly diag-

nostic for urachal carcinoma. At CT, urachal carcinoma may be solid, cystic, or a combination of the two. Low-attenuation components are seen in 60% of cases, reflecting the mucin content (Figs 9, 10) (8). As with other tumors in the walls of cysts in other locations, it may be impossible to distinguish a true cystic urachal carcinoma from a carcinoma arising in the wall of a urachal cyst (41). The presence of an unencapsulated caudal part involving a portion of the bladder wall and of an often cystic encapsulated supravescical portion are considered highly characteristic (Figs 9, 10) (42). US allows localization of the tumor and detection of highly echogenic calcifications as well as of the solid components of the tumor in the anterior abdominal wall without any obscuration by intraabdominal organs including air-filled bowel (Figs 9–11). The mucin-containing cystic portion of the tumor demonstrates increased echogenicity rather than an anechoic pattern, and there may be a discrepancy between CT and US

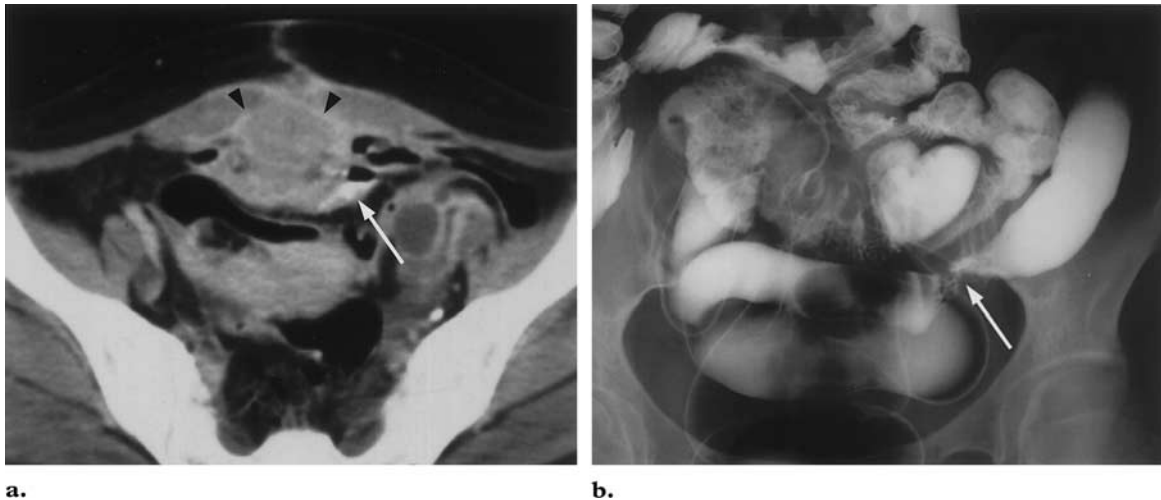




**Figure 10.** Adenocarcinoma arising from the urachal remnant in a 55-year-old woman. **(a)** Sagittal US image shows an inhomogeneous mass with mixed echogenicity (arrowheads) involving the anterosuperior portion of the bladder (*bl*) and protruding into the bladder lumen. The intravesical component of the mass demonstrates relatively homogeneous echogenicity. **(b, c)** Contrast-enhanced CT scans (**c** obtained at a lower level than **b**) show a well-defined mass (arrowheads) overlying the right anterosuperior aspect of the bladder (*bl*). The tumor is deviated to the right side rather than being in the typical midline location. Note the extravesical component with inhomogeneous attenuation and an intravesical component with relatively homogeneous high attenuation. *ut* = uterus.



**Figure 11.** Mucinous adenocarcinoma arising from the urachal remnant in a 40-year-old man. **(a)** Sagittal US image shows a soft-tissue mass with multiple thick flecks of calcification (arrowheads) accompanied by posterior shadowing. **(b)** Unenhanced CT scan shows a densely calcified mass (arrow) projecting into the dome of the bladder (*bl*). A large amount of peritoneal fluid is seen surrounding the urinary bladder (arrowheads), a finding that suggests carcinomatosis.



**Figure 12.** Recurrent tumor arising from the urachal remnant in a 38-year-old woman who had undergone surgical resection of adenocarcinoma. **(a)** Contrast-enhanced CT scan shows a solid mass just beneath the linear alba (arrowheads). Note the fat plane obliteration between the anterior abdominal wall and the intraperitoneal bowel loops (arrow). **(b)** Image from a double-contrast colon study shows a stricture of the proximal sigmoid colon (arrow).

findings (Fig 9). As has been mentioned, it is difficult to distinguish infected urachal remnants from urachal carcinoma. The presence of hematuria, mural nodularity, and calcification at CT, along with a lack of adjacent inflammatory change, can be helpful as distinguishing features in some cases (43–45).

The prognosis in urachal tumor is related to the stage and degree of differentiation, although it is generally poor because the tumor arises in a clinically silent area and is discovered only after it has extended into the bladder lumen or manifests with symptoms related to its large size or extension into adjacent organs. CT may demonstrate both intra- and extravesical components and the effect of the tumor on surrounding structures (Figs 11, 12). A discrete perilesional spiculation or fat stranding is suggestive of, but not specific for, tumor infiltration because many cases of infection have demonstrated perilesional inflammatory spread overlapping peritumoral stranding at CT. CT is not accurate in identifying microscopic invasion of fat and bladder mucosa (8). Metastases occur initially in the pelvic lymph nodes, followed by systemic metastases to the lung, brain, liver, and bone (39). Local invasion and systemic metastases result in a 5-year survival rate of 6.5%–15% (38).

## Conclusions

Many of the features of urachal remnant diseases, including congenital lesions with or without superimposed infection and tumors, are well displayed at sagittal US. CT helps confirm the US findings and discloses the nature and local extension of the disease as well as any systemic metastases. Nevertheless, the CT and US findings in infected urachal cysts mimic those in urachal carcinoma. Because of the lack of specificity of CT and US in the differential diagnosis of solid urachal masses, a definitive pathologic diagnosis is required to optimize the surgical approach and preclude unnecessary radical surgery. Understanding the anatomy and the imaging features of urachal remnant diseases, along with the typical locations and distributions of these diseases, is essential for correct diagnosis and proper management.

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