Imaging of the Urachus: Anomalies, Complications, and Mimics[1]

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Abbreviation: VCUG = voiding cystourethrography

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SA-CME LEARNING OBJECTIVES

After completing this journal-based SA-CME activity, participants will be able to:

- Explain the embryologic and anatomic considerations of the normal urachus.
- Identify and describe the spectrum of radiologic findings for each different type of urachal anomaly.
- Discuss the imaging features of potential complications and mimics of urachal abnormalities.

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Urachal anomalies are more common than previously thought, with more cases discovered incidentally, because of the increased use of cross-sectional imaging. Although an abnormal persistence of an embryologic communication between the bladder and the umbilicus is often recognized and managed in childhood, it may persist into adulthood, with a greater risk of morbidity. Congenital urachal anomalies that are detected early can benefit from an optimized management including surgical approach with a complete resection of the urachal remnant in cases when spontaneous resolution or medical management has failed. At imaging, the different types of urachal anomalies have a distinct appearance. A patent urachus is recognized as an elongated patent connection between the bladder and the umbilicus. An umbilical-urachal sinus is depicted as a blind focal dilatation at the umbilical end, whereas a vesicourachal diverticulum is a focal outpouching at the vesical end. Urachal cysts are visualized as midline fluid-filled sacs most frequently located near the bladder dome. Complications of urachal anomalies have nonspecific clinical findings and can mimic other abdominal and pelvic processes. Potential complications, such as infection and tumors, should be recognized early to ensure optimal management. Understanding of the embryonic development of the urachus is necessary for the radiologist to diagnose the wide variety of urachal disease.

Introduction

The urachus is a ductal remnant that arises embryologically, originating from the involution of the allantois and cloaca and extending between the bladder dome and the umbilicus. During normal gestational development, the urachus involutes and its lumen is obliterated, becoming the median umbilical ligament (1).

Congenital urachal anomalies result from a failure of the developing urachus to completely obliterate its lumen, and represent uncommon and often underdiagnosed entities that may be found incidentally at imaging or manifest clinically with the development of mostly nonspecific abdominal or urinary symptoms. Persistence of undetected urachal anomalies may cause abdominal and/or urinary complications in both pediatric and adult populations as a result of

TEACHING POINTS

- The urachus or median umbilical ligament represents the embryologic remnant of two embryologic structures: the cloaca and the allantois.
- The urachus extends from the anterosuperior surface of the bladder to the umbilicus and lies in the extraperitoneal space of Retzius (or retropubic space) between the transverse fascia and parietal peritoneum.
- Four types of urachal anomalies have been described, on the basis of the location of the abnormal residual patency along the urachal tract: patent urachus (sometimes referred to as urachal fistula), urachal cyst, umbilical-urachal sinus, and vesicourachal diverticulum.
- The recommended surgical approach for all urachal anomalies is complete excision of the urachal remnant.
- Calcifications are present in 70% of the cases and are mostly seen at the periphery of the urachal mass; however, they may also be central or a combination of both. The presence of calcifications in a midline soft-tissue mass along the course of the urachal tract is considered pathognomonic for the diagnosis of urachal adenocarcinoma.

chronic urinary stasis, inflammation, and infection, predisposing the patient to potential morbidity and malignancy (2). In addition, urachal anomalies can be confused with other abdominal and pelvic entities unless one is familiar with the embryologic development, anatomy, and imaging features of the different urachal anomalies and their potential complications.

The reported incidence of urachal anomalies is approximately one in 5000 population for adults, with a significantly lower rate of one in 150 000 population among infants (3,4). There is a higher prevalence in men than women (5,6). Most anomalies of the urachus are unexpected, being detected incidentally and more frequently with the increased use of cross-sectional imaging.

The objective of this article is to review the normal development, anatomy, and imaging appearance of the urachus. The imaging features of congenital urachal anomalies (ie, patent urachus, urachal cyst, umbilical-urachal sinus, and vesicourachal diverticulum), potential complications (most importantly infection and neoplasm), and mimics will also be discussed and illustrated.

Embryologic Development and Anatomy

The urachus or median umbilical ligament represents the embryologic remnant of two embryologic structures: the cloaca and the allantois (7). The allantois is an extraembryonic diverticulum that appears early in the 3rd week of gestational life located on the posterior aspect of the yolk sac and projects into the body stalk. The cloaca is an endoderm-lined structure, which is divided by the

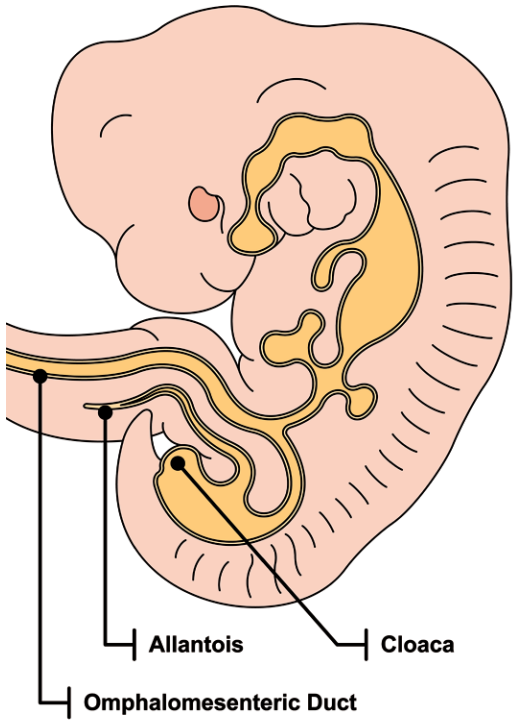
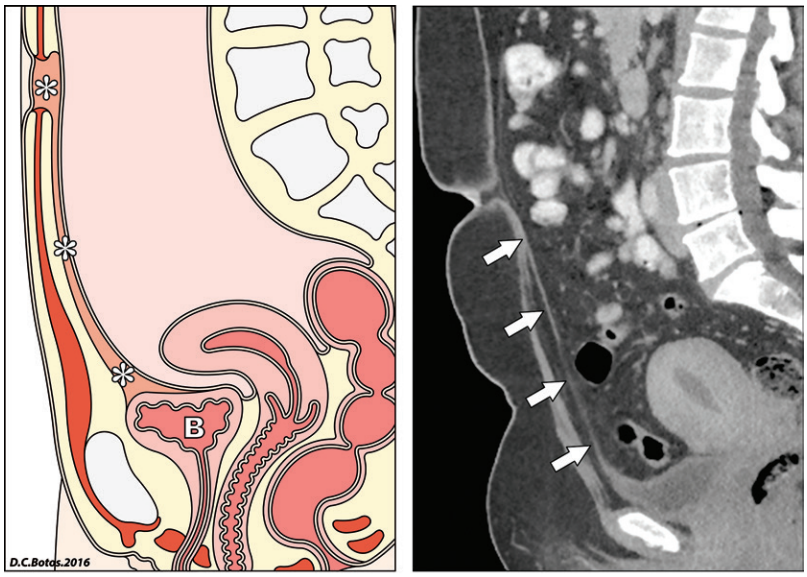


Figure 1. Illustration of a 4th-week embryo shows the cloaca as a blind-end portion of the hindgut and the allantois projecting into the body stalk.

urorectal septum between the 4th and 7th weeks of gestation, creating the urogenital sinus ventrally and the rectum dorsally (Fig 1). The inferior portion of the urogenital sinus later becomes the pelvic and penile urethra in males and the pelvic urethra and vaginal vestibule in females. The superior portion of the ventral urogenital sinus later becomes the bladder and its apex, which subsequently becomes the bladder dome, which is contiguous with the allantois (8–12).

Around the 4th to 5th gestational months, involution of the allantois and urogenital sinus occurs as the bladder descends into the pelvis, forming the urachus, which elongates and becomes a fibromuscular cordlike structure connecting the apex of the bladder to the umbilicus. Urachal remnants are present in almost 100% of infants at birth and regress due to fibrosis with age, eventually becoming the median umbilical ligament. Two-thirds of adolescents and one-third of 35-year-old adults show remnant urachal tissue, but it is seen in only 3% of the general population at autopsy (13–16). These remnants are rarely seen and are not considered pathologic, unless they fail to obliterate, retaining some degree of luminal patency (16).

The urachus extends from the anterosuperior surface of the bladder to the umbilicus and lies in the extraperitoneal space of Retzius (or retropubic space) between the transverse fascia



a. b.

Figure 2. **(a)** Illustration shows the anatomic location (*) of the urachus in the space of Retzius. *B* = bladder. **(b)** Sagittal contrast-enhanced computed tomographic (CT) image in a 42-year-old woman demonstrates the urachus as a cordlike structure extending between the bladder dome and the umbilicus (arrows).

and parietal peritoneum (Fig 2) (17). It is accompanied on both sides by the medial umbilical ligaments, which are the obliterated remnants of the umbilical arteries and may on occasion merge with the urachus, causing it to mildly deviate from the midline (9). The length of the urachus ranges from 3 to 10 cm and it generally has an approximate diameter of 8–10 mm (9,17). Histologically, it is usually composed of three layers: an epithelial lining that is usually transitional cell but may also be columnar, a middle submucosal layer of connective tissue, and an outermost muscular layer that is continuous with the detrusor muscle (14,18). However, remnants without an epithelial lining have also been described (15).

Classification and Imaging of Congenital Urachal Anomalies

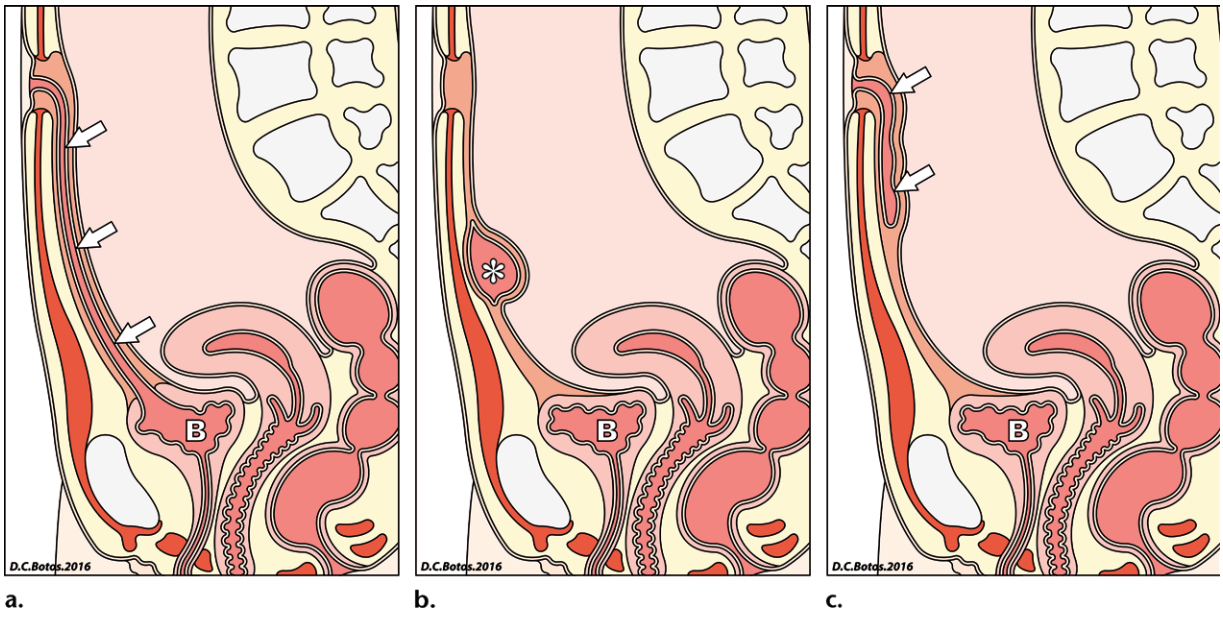
The true incidence of congenital urachal anomalies is uncertain. Before the routine use of cross-sectional imaging, 315 cases had been reported in the literature, collected between 1550 and 1970 (19). In the modern era of cross-sectional imaging, these diagnoses have become more prevalent, and asymptomatic cases are being more frequently diagnosed on the basis of incidental imaging findings.

Four types of urachal anomalies have been described, on the basis of the location of the abnormal residual patency along the urachal tract: patent urachus (sometimes referred to as urachal fistula), urachal cyst, umbilical-urachal sinus, and vesicourachal diverticulum (Fig 3). A fifth type has also been reported but is in fact a variant of the umbilical-urachal sinus, which is a sinus with intermittent drainage caused by almost complete obliteration of the umbilical portion of the sinus (16). The most common type of urachal anomaly is a patent urachus (accounting for 47%), followed by urachal cyst (30%), sinus (18%), and, least commonly, vesicourachal diverticulum (3%) (7,20–23). However, these rates of relative prevalence predate the advent of widespread imaging and may underestimate the prevalence of the asymptomatic types. A recent study of urachal anomalies in children found that urachal cysts are in fact the most common type, accounting for 69% of cases (24).

Imaging of suspected urachal anomalies can be performed with various modalities. The most commonly used imaging modality for initial screening is ultrasonography (US), because it is fast, readily available, and does not entail radiation exposure, which is especially important in children and young adults (25,26). The reported diagnostic accuracy of US for urachal anomalies exceeds 90%, according to some authors (27). Other imaging modalities, such as sinography, CT, and magnetic resonance (MR) imaging, are usually recommended if US fails to allow diagnosis of the anomaly (28).

Patent Urachus

Patent urachus, or urachal fistula, is characterized by the presence of a free and persistent communication between the bladder and the umbilicus, usually leading to urinary leakage from the umbilicus (19). Rarely, the vestigial lumen is extremely



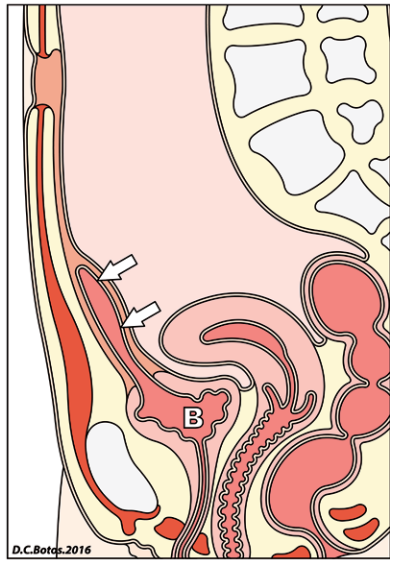


Figure 3. Illustrations of the four types of urachal anomalies. *B* = bladder. **(a)** Patent urachus (arrows). **(b)** Urachal cyst (*). **(c)** Umbilical-urachal sinus (arrows). **(d)** Vesicourachal diverticulum (arrows).

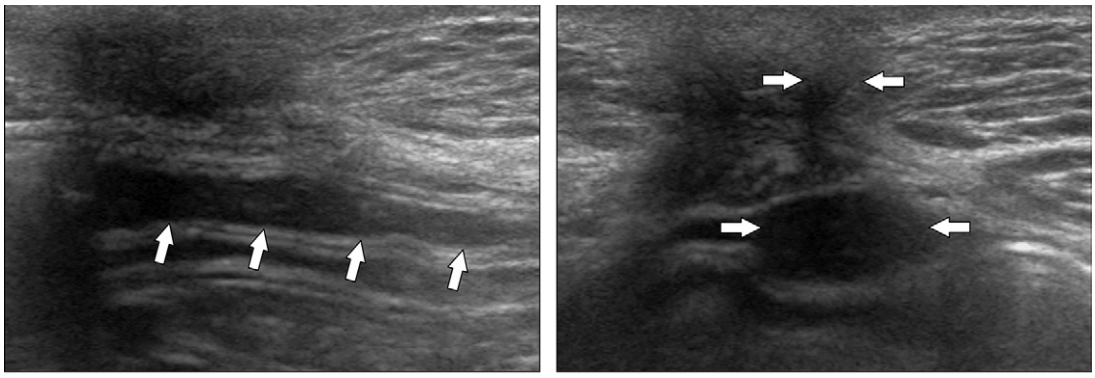
narrow and patients remain asymptomatic, presenting only when bladder obstruction causes bladder pressures to rise, leading to leakage (9). This anomaly is most frequently detected during the neonatal period by physical examination in which the presence of continuous urinary drainage from the umbilicus, and related abnormal appearances of the navel, including an edematous umbilicus, granulomas, and/or delayed cord stump healing, are seen (11,29,30). Prenatal diagnosis is usually not possible.

A definitive diagnosis of patent urachus can be made with imaging. Demonstration of a tubular structure with a hypoechoic wall and anechoic content extending from the bladder dome to the umbilicus at US is diagnostic (Fig 4). Confirmation of the suspected anomaly can also be accomplished at fluoroscopy with either a sinogram or voiding cystourethrography (VCUG) by demonstration of a contrast material–filled tract extending from the bladder to the umbilicus (11,14). Although fluoroscopy is usually not necessary for the diagnosis, the possible association with vesicoureteral reflux leads some authors to recommend VCUG for complete evaluation (27). Other imaging modalities such as CT and MR imaging can also be diagnostic and are usually performed in the setting of uncertainty or suspected complications (Fig 5).

Urachal Cyst

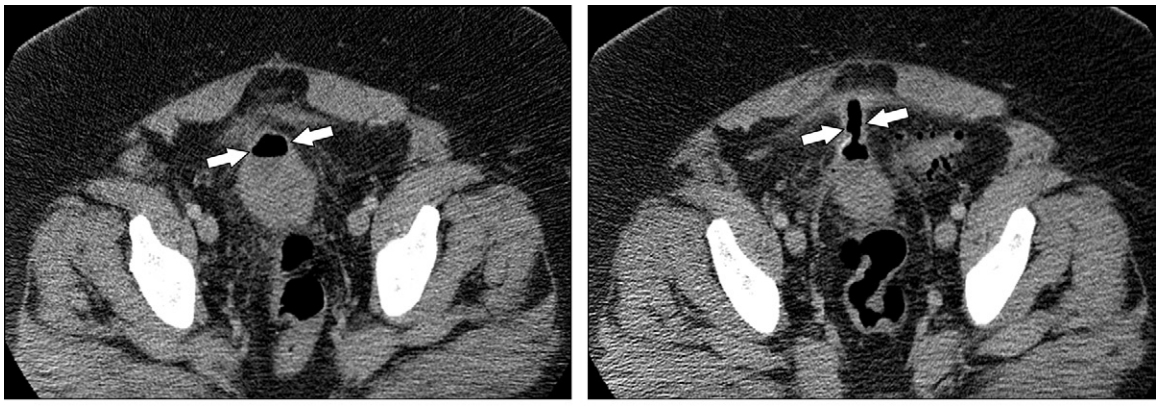
Urachal cysts develop when both the umbilical and bladder ends of the urachus are obliterated, but a focal segment remains patent somewhere along the course of the urachus, more commonly at the lower third of the urachal tract (9). Cysts are usually small and asymptomatic. The diagnosis is often made only when they become symptomatic in childhood or adulthood because of complications, mainly infection, or as an incidental finding at imaging performed for unrelated reasons (9,24).

At imaging, the diagnosis of an uncomplicated cyst can be made by demonstration of a midline homogeneous fluid-filled structure anywhere along the theoretical course of the urachus between the bladder and the umbilicus (9). This is usually achieved by visualization of the cyst with US or CT. MR imaging is also helpful, although usually performed for evaluation of unrelated disease, with the cyst diagnosed as an incidental finding (Fig 6).



a. b.

Figure 4. Patent urachus in a 32-year-old man with umbilical pain and a small purulent discharge through the navel. **(a)** Longitudinal US image shows a fluid-filled hypoechoic tubular structure (arrows) extending from above the bladder (not shown). **(b)** Transverse US image at the anterior abdominal wall shows the patent urachus as a fluid-filled collection (lower arrows), just beneath the level of the umbilicus (upper arrows point to umbilical shadow).



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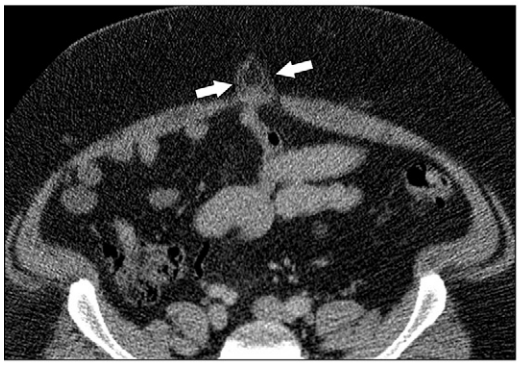


c.

Figure 5. Patent urachus in a 42-year-old man with a history of urine leakage from the umbilicus and persistent urinary infections. The patient presented with fever and a painful palpable suprapubic mass. Axial CT images **a–c** show a tubular structure (arrows) containing air extending from the bladder dome to the umbilicus with minimal enhancement after contrast material injection, most likely consistent with an infected patent urachus.

US, CT, and MR imaging reveal a thickened and fusiform blind dilatation of the urachus at the umbilical end with no communication to the bladder (Fig 8). Confirmation of the diagnosis can be obtained with sinography, confirming the lack of communication between the sinus tract and bladder.

Umbilical-Urachal Sinus

Umbilical-urachal sinus occurs when the umbilical end of the urachus fails to obliterate and persists as a fusiform outpouching structure just below the navel (23,31,32). This urachal anomaly represents a potential space in which the accumulation of cellular debris can favor the development of potential complications such as infection and less commonly stone formation (33). Periumbilical pain and tenderness with a periodic discharge through the navel and a wet umbilicus are usually reported in patients with an umbilical-urachal sinus (Fig 7) (1,34).

Vesicourachal Diverticulum

Vesicourachal diverticulum is the most rarely detected urachal anomaly, caused by an incomplete closure of the urachus at the bladder end, manifesting as an outpouching from the anterior bladder dome at the location of the urachal attachment. Urachal diverticula usually have a large opening and drain well into the bladder, decreasing the frequency of complications, and therefore usually remaining asymptomatic and discovered as incidental findings at imaging studies performed for other reasons (5,14).

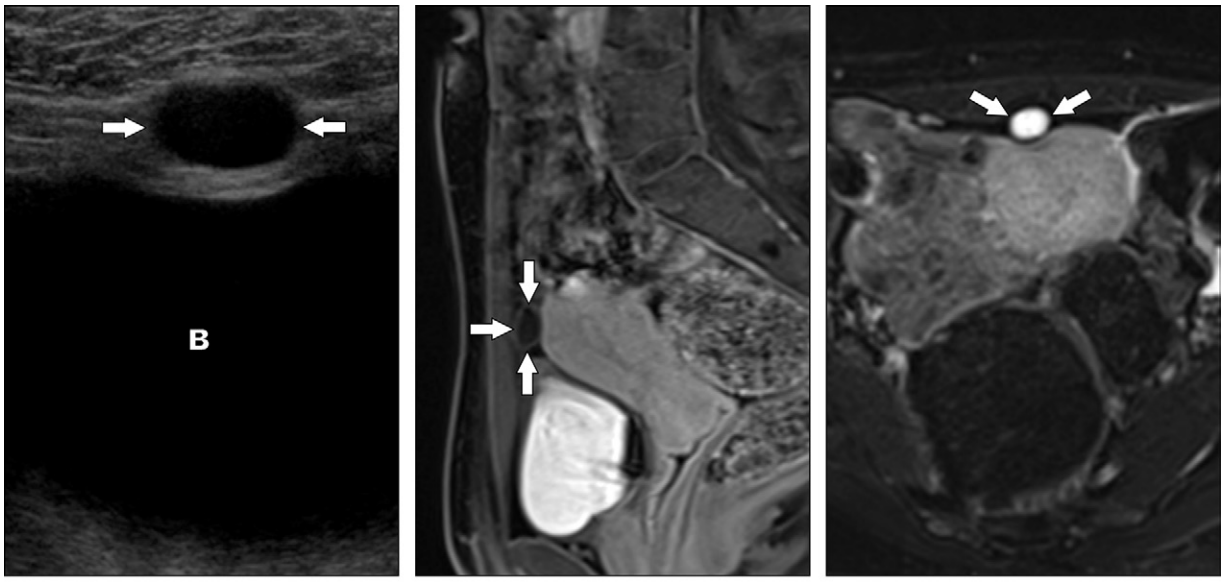


Figure 6. Urachal cyst in a 29-year-old woman with recurrent urinary tract infections. **(a)** Transverse US image shows a round anechoic structure (arrows) above the superior surface of the bladder *(B)*. **(b, c)** Sagittal gadolinium-enhanced T1-weighted **(b)** and axial T2-weighted **(c)** fat-suppressed MR images demonstrate a fluid-filled collection (arrows) at the midline lower abdomen immediately above the anterosuperior aspect of the bladder, consistent with a urachal cyst.

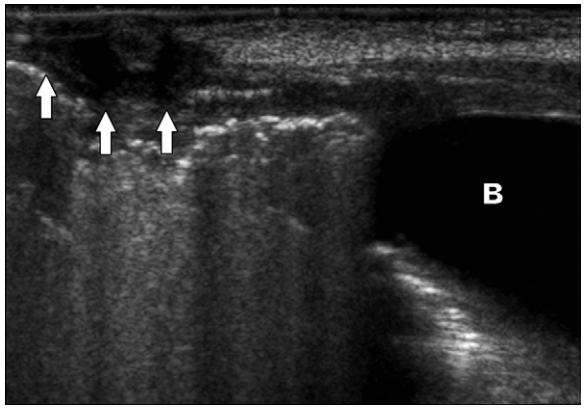


Figure 7. Umbilical-urachal sinus draining to the skin in a 32-day-old girl. Longitudinal US image shows a rounded fluid-filled hypoechoic structure (arrows) just below the navel with no visible communication to the bladder *(B)*. (Courtesy of Jeanne S. Chow, MD, Boston Children's Hospital, Boston, Mass.)

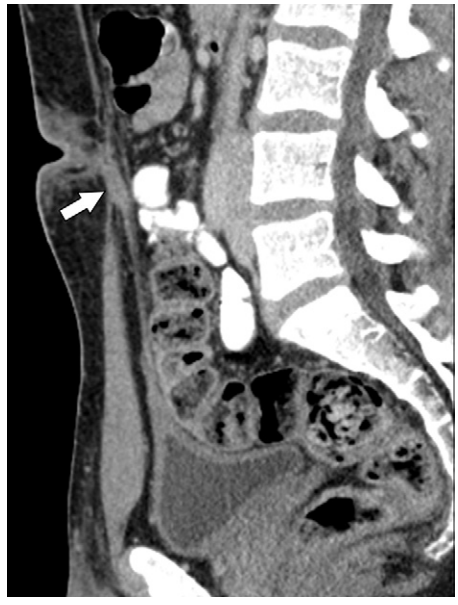
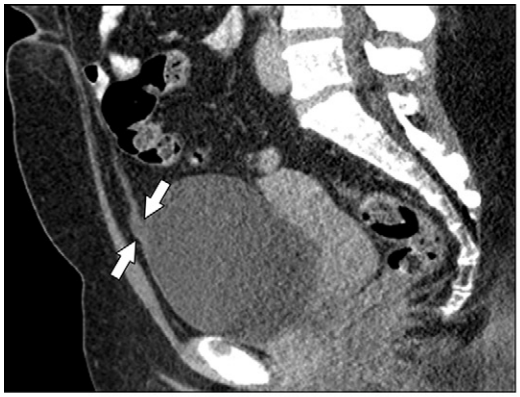


Figure 8. Umbilical-urachal sinus as an incidental finding at contrast-enhanced CT performed after trauma. Sagittal contrast-enhanced CT image shows a focal dilatation (arrow) of the urachus at the umbilical end with no communication to the bladder.

US, CT, and MR imaging can all demonstrate a midline focal protruding extension of the bladder arising from its dome with no communication to the umbilicus or the rest of the urachal tract (Fig 9) (35). This can also be identified at VCUG as a tubular contrast material–filled structure extending superiorly from the anterior bladder dome.

Management of Congenital Urachal Anomalies

The approach to patients with urachal anomalies has evolved over time and remains controversial (24). Although symptomatic patients have traditionally been treated surgically, given the pathophysiology of the disease, some authors now follow a treatment algorithm that is dependent on age and symptoms. With increasing reports of spontaneous resolution of urachal remnants with age, including soon after birth, symptomatic infants are now usually treated medically, and only in cases of recurrent symptoms or failure of spontaneous resolution are patients under 1 year of age treated surgically (27).



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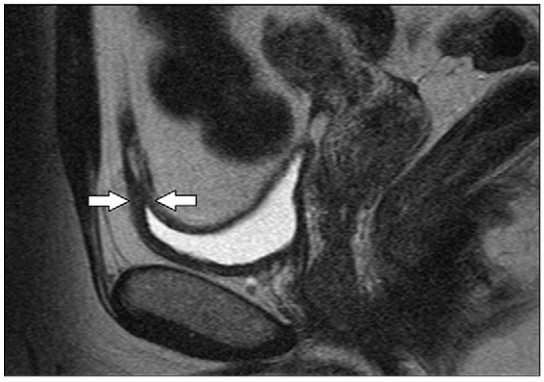

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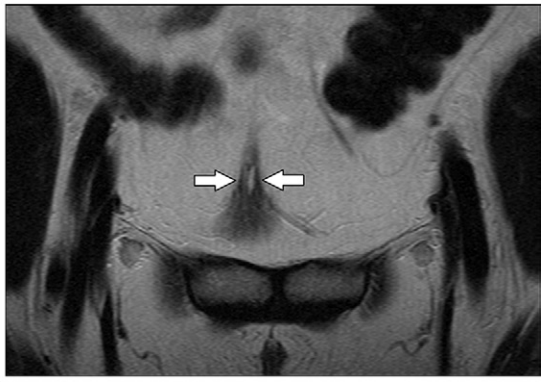


c.

Figure 9. Vesicourachal diverticulum in a 52-year-old woman with abdominal pain. Sagittal contrast-enhanced CT image **(a)** and sagittal **(b)** and coronal **(c)** T2-weighted MR images reveal a midline protruding extension (arrows) of the bladder at its apex with no communication to the umbilicus.

Follow-up US is recommended in these patients to provide evidence allowing confirmation of regression of the remnant. In older children, a more aggressive approach to symptomatic anomalies may be adopted, although some authors recommend a conservative approach regardless of age (24).

Surgical excision of asymptomatic urachal anomalies has been proposed to prevent complications, mostly to prevent the development of malignancy. It has been suggested that this approach in children is perhaps too aggressive because the development of malignancy in children's urachal remnants is extremely rare, with only relatively few case reports in the literature (27,36), and the tendency toward spontaneous regression. However, even in adults, some authors believe the benefits of prophylactic excision to be limited in the presence of an asymptomatic lesion (37).

The recommended surgical approach for all urachal anomalies is complete excision of the urachal remnant. Anomalies that extend to the bladder (patent urachus and vesicourachal diverticulum) require excision of the remnant along with placement of a bladder cuff to prevent long-term complications (26,38). This radical excision includes excision of the medial umbilical ligaments along with the urachal remnant and the adjacent peritoneum (3). The excision can be accomplished either by a traditional open surgical approach using an infraumbilical transverse or vertical midline incision, or by a laparoscopic approach, which has been proven to be safe and effective (mostly in adults), but has also been used in children as young as 5 months old (3,26,38). Robotically assisted techniques may also be used (1,4). In anomalies that do not extend to the bladder dome (umbilical-urachal sinus and urachal cysts) a segmentary bladder resection is not always considered necessary, although this approach remains somewhat controversial (39).

Complications of Urachal Remnants

Urachal anomalies may cause considerable morbidity if not detected and treated promptly. The potential development of complications depends on the existence of contributing factors, including age (given the presence of distinct complications for the pediatric versus the adult populations) and also on the type of urachal anomaly that is present.

Complications in infants include, most commonly, infection of the urachal remnant. Associated genitourinary conditions that may lead to

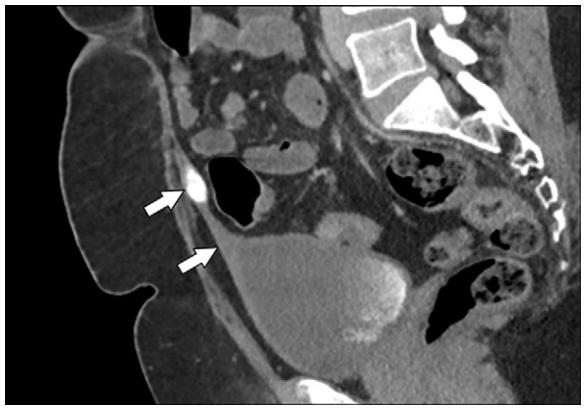


Figure 10. Sagittal contrast-enhanced CT image in a 43-year-old woman demonstrates a suspected vesicourachal diverticulum (bottom arrow) complicated by a calcification (top arrow).

potential complications most frequently include vesicoureteral reflux, but also include hypospadias, meatal stenosis, crossed renal ectopia, umbilical or inguinal hernias, anal atresia, cryptorchidism, omphalocele, and ureteropelvic obstruction (5,15,29). In the adult population, the most commonly encountered complication is also infection, as it is with the pediatric population, and the development of malignancy. These will be further discussed in detail.

Other reported complications include umbilical granulomas and persistent urinary tract infections. Calcifications or stone formation can also occur and are most commonly present in patients with urachal cysts or vesicourachal diverticula as a result of chronic urinary stasis associated with calcification of glandular epithelium in the cyst wall or calculus formation within the urachal remnant (Fig 10) (22,29,35). Progressive enlargement of a urachal cyst has also been reported, leading in some cases to rupture, bowel obstruction, or fistulization into adjacent bowel (24).

Infection

Infection represents the most common complication of urachal anomalies and may produce sometimes marked nonspecific symptoms, including abdominal pain and tenderness, fever, erythema, purulent urinary discharge, and occasionally a palpable mass. Infection routes for bacterial migration can be lymphatic, hematogenous, or by direct extension from the bladder; *Staphylococcus aureus* is the most commonly isolated organism, followed by *Escherichia coli*, *Enterococcus*, *Citrobacter*, *Klebsiella*, and *Proteus* (2,11,29,40). Although it is unusual, severe infection can result in the formation of complex fistulas and abscesses, with the attendant risk of potential intraperitoneal rupture causing peritonitis and sepsis (5,9,41).

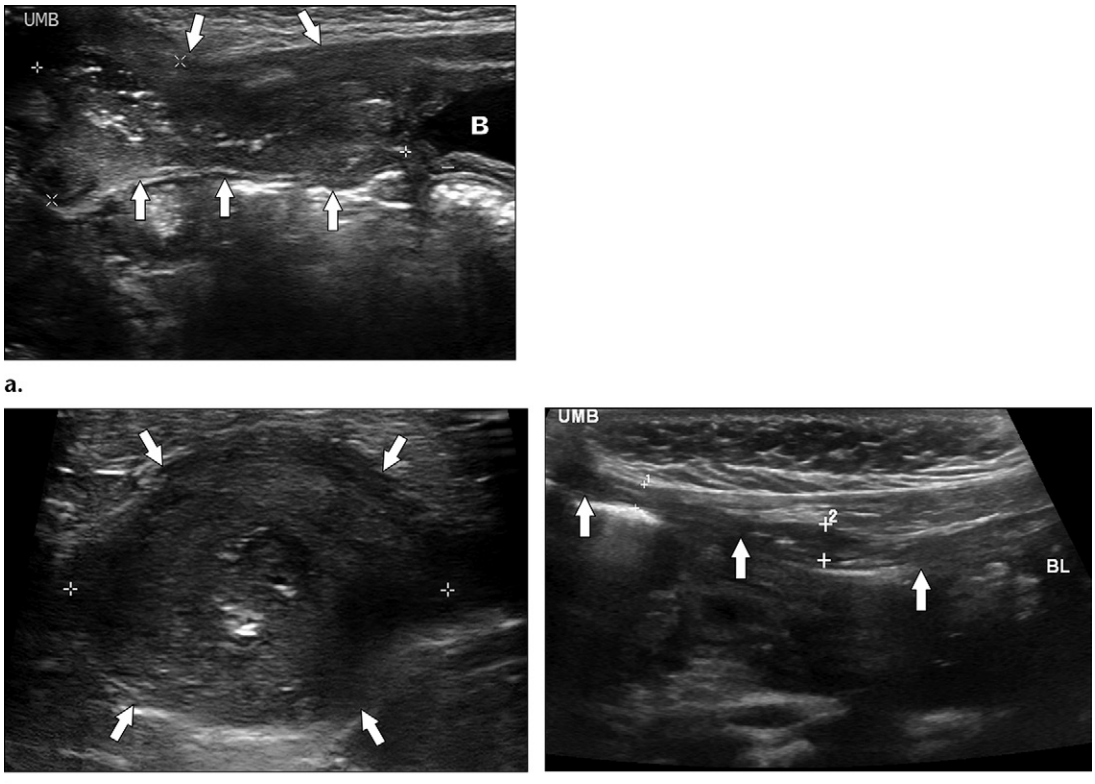
Imaging evaluation of infected urachal remnants on the basis of the type of anomaly can include cystography, sinography, cystoscopy, US, CT, and MR imaging (18). US is usually the first imaging tool to be used in patients with a suspected infection of urachal remnants. If US is not diagnostic, CT or MR imaging may be subsequently employed to help reach a definitive diagnosis, as well as to evaluate the relationship of the infected urachal anomaly with the surrounding tissue and adjacent organs (5,40,42). Imaging findings that should prompt consideration of infection in the setting of an acute clinical presentation include the presence of a urachal remnant with complex echogenicity at US (Fig 11) and heterogeneous attenuation with variable (usually increased) contrast enhancement at CT (Fig 12). MR imaging can also be used to aid in determining the extent of infection involving the bladder and other adjacent structures (18).

When patients present with an infected urachal anomaly, initial management should include broad-spectrum antibiotic therapy, if possible, even before culture results are available and initial percutaneous or surgical drainage is performed. After the infection has cleared, complete excision of the urachal remnant can be performed with open surgical, laparoscopic, or robotically assisted techniques to mitigate the 30% risk of infection recurrence and potential malignant degeneration (1,9,14,32).

Neoplasms

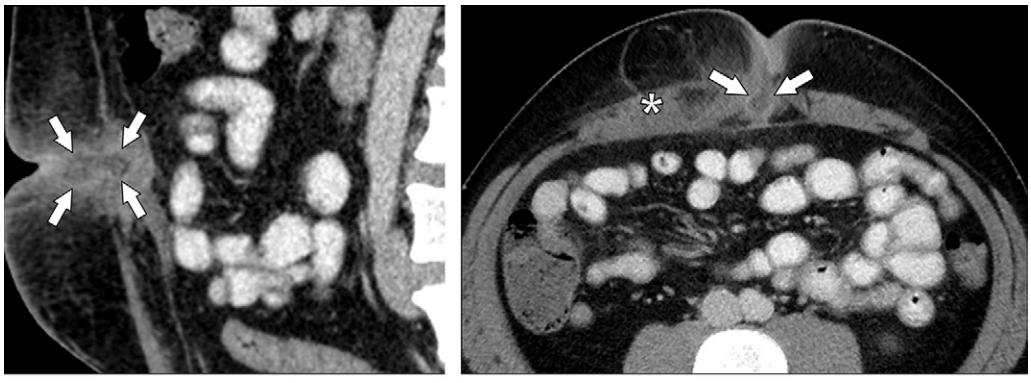
Benign.—Benign urachal tumors have been described in the literature only as case reports. They are extremely rare and can arise from any portion of the urachal tract, usually mimicking urachal malignancy. The most commonly described benign tumors of the urachus are adenomas and cystadenomas, followed by fibromas, fibromyomas, fibroadenomas, and hamartomas (13,43). Because they mimic malignancy, the diagnosis is usually made at histopathologic analysis after resection.

Malignant.—Malignant urachal neoplasms are also rare, accounting for less than 1% of all blad-



a. b. c.

Figure 11. Infected urachal remnant in an 18-day-old boy. **(a, b)** Longitudinal **(a)** and transverse **(b)** US images show enlargement (arrows) of a urachal remnant with complex echogenicity and thickened walls, suggesting infection. *B* = bladder. **(c)** Follow-up US image after antibiotic therapy shows the urachal remnant to be clear of infection, showing as a tubular hypoechoic structure (arrows) located between the umbilicus *(UMB)* and the bladder *(BL)*. (Courtesy of Jeanne S. Chow, MD, Boston Children's Hospital, Boston, Mass.)



a. b.

Figure 12. Infected umbilical-urachal sinus in a 36-year-old woman with periumbilical pain. **(a)** Sagittal contrast-enhanced CT image shows a hypoattenuating fluid collection with rim enhancement (arrows) at the location of an umbilical-urachal sinus. **(b)** Axial view shows enlargement (*) of the right rectus abdominis muscle with a fluid collection (arrows) and fat stranding, suggesting infection of the urachal remnant.

der cancers. They usually remain undiscovered for a long period of time and may be found incidentally at imaging or at an advanced stage when symptoms of local invasion or systemic spread have developed (43–46).

Although urachal remnants are lined by urothelium, 80% of urachal cancers are adenocarcinomas, including mucin-producing adenocarcinomas (69%) and mucin-negative adenocarcinomas (15%). The remaining 20% of urachal cancers are urothelial, squamous, and sarcomatoid neoplasms (43,44,46). This differs from the most common type of bladder cancer, which is typically urothelial. It is unclear why adenocarcinoma is the predominant type, and it has been hypothesized that chronic irritation is responsible for metaplasia of the transitional epithelium into columnar epithelium. It is also possible that some

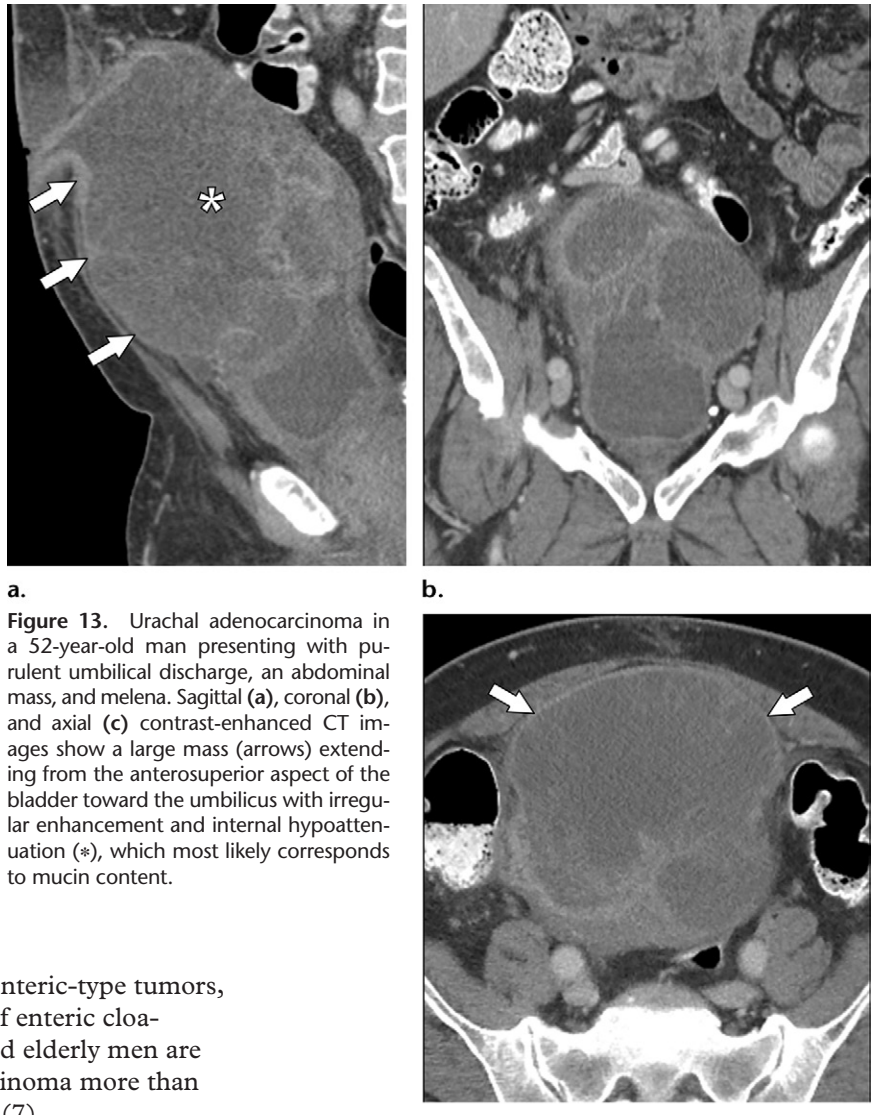
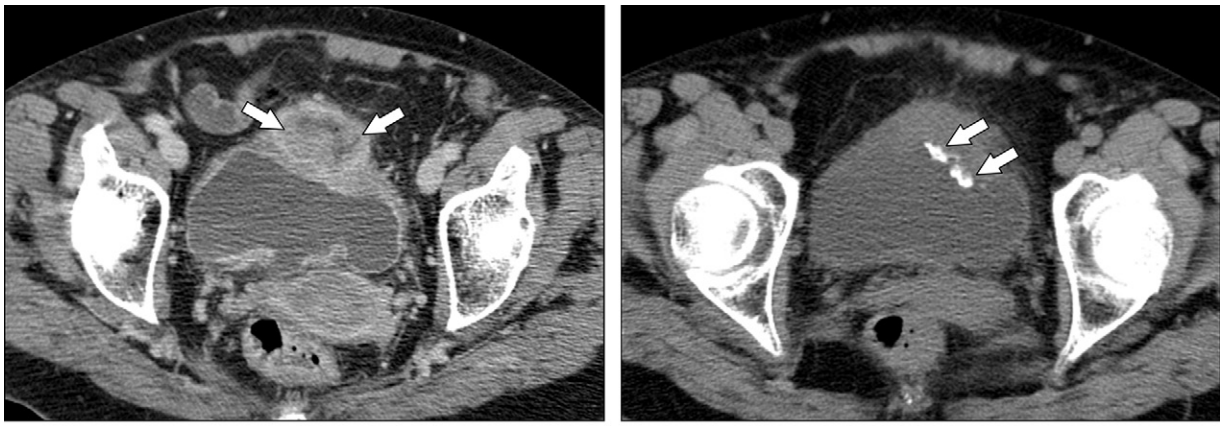


Figure 13. Urachal adenocarcinoma in a 52-year-old man presenting with purulent umbilical discharge, an abdominal mass, and melena. Sagittal **(a)**, coronal **(b)**, and axial **(c)** contrast-enhanced CT images show a large mass (arrows) extending from the anterosuperior aspect of the bladder toward the umbilicus with irregular enhancement and internal hypoattenuation (*), which most likely corresponds to mucin content.

adenocarcinomas, especially enteric-type tumors, originate in embryonic rests of enteric cloacal cells (46). Middle-aged and elderly men are affected by urachal adenocarcinoma more than twice as frequently as women (7).

Unfortunately, urachal adenocarcinomas are generally large and manifest with a more prominent extravesical component than do other nonurachal tumors of the bladder (44). Hematuria and a palpable suprapubic mass are the most commonly associated symptoms. Other manifestations can include abdominal pain, dysuria, mucosuria, purulent or hematic discharge from the umbilicus, and obstructive symptoms (47,48). Diagnostic evaluation for urachal carcinoma must include a detailed clinical history and physical examination, urinalysis with urinary cytology, and imaging, including CT and/or MR (49).

Approximately 90% of urachal carcinomas develop in the portion of the urachus adjacent to the bladder and gradually grow in the cranial direction (7,44). Cystography may reveal a filling defect in the bladder dome or the presence of extrinsic compression. US is often performed as the initial imaging modality and can provide a general impression of the lesion, including location and the demonstration of a midline soft-tissue mass with complex echogenicity and calcifications adjacent to the lower abdominal wall (32,43,44,48). Both CT and MR imaging can be used to confirm the US findings or as the first-line imaging modalities for the evaluation of local disease, tumor extension, and the presence of pelvic lymph node involvement or distant metastases. At CT, urachal carcinoma can be cystic, solid, or mixed. It usually appears as a midline mass adjacent to the bladder dome with extension into the space of Retzius, demonstrating heterogeneous low-attenuation components, which represent mucin content (Fig 13). Calcifications are present in 70% of cases and are mostly seen at the periphery of the urachal mass; however, they may also be central or a combination of both (50). The presence of calcifications in a midline soft-tissue mass along the course of the urachal tract is considered pathognomonic for the diagnosis of urachal adenocarcinoma (Fig 14) (32,43,44). At MR imaging, sagittal images may be the most helpful for evaluation of urachal neoplasms, as they will typically manifest as a



a. b.

Figure 14. Urachal adenocarcinoma in an 80-year-old woman who presented with irritative voiding symptoms. **(a)** Axial contrast-enhanced CT image shows an exophytic mass (arrows) arising from the anterior aspect of the bladder with central hypoattenuation and peripheral enhancement. **(b)** Calcifications (arrows) are identified within the urachal mass.

Staging Systems for Urachal Carcinomas

Stage	Mayo Clinic (Ashley et al [45])	Ontario (Pinthus et al [54])
I	Tumor confined to urachus and/or bladder	Tumor confined to urachus
II	Tumor extension beyond urachus and/or bladder	Tumor confined to bladder
III	Tumor infiltrating regional lymph nodes	Tumor invading surrounding fat
IV	Tumor infiltrating nonregional lymph nodes or distant sites	Tumor extending into peritoneum, abdominal wall, and adjacent organs
Node-metastasis involvement	...	Local lymph node involvement and distant metastases considered separately

midline mass with focal areas of heterogeneous high signal intensity on T2-weighted images that are suspicious for mucin content (44,51). However, high signal intensity on T2-weighted images can also resemble other conditions such as fluid collections or necrosis; mucinous carcinomas show enhancement of glandular tissue following contrast injection, distinguishing it from fluid or necrosis (52). Solid components of the tumor are isointense to soft tissue on T1-weighted images and enhance following the administration of intravenous gadolinium contrast material (44).

Several clinical staging systems for urachal carcinomas have been established and are somewhat controversial, with none formally validated. Sheldon et al (53) proposed the first staging system in 1984, which has been replaced by the Mayo Clinic staging system proposed by Ashley et al (45) and the Ontario staging system proposed by Pinthus et al (54), both in 2006 (Table). The initial system of Sheldon et al (53) did not take into account the development of urachal malignancy along the whole urachal tract and considered the presence of regional lymph nodes to be distant metastases (36). However, owing to the limited number of cases in both the Mayo and Ontario systems and the lack of accurate classification for the local invasion patterns of urachal neoplasms, none of these clinical staging systems have yet been validated (49,55).

The prognosis of urachal adenocarcinoma is usually poor due to late presentation and advanced stage with local invasion. The 5-year survival rate for urachal adenocarcinoma is variable and has been reported to be approximately 49% after treatment (49,56,57), mostly related to clinical staging at the time of presentation rather than the histologic subtype, location, or resectability (29,32,44). Local recurrence is often seen within 2 years of surgery (43,49). Metastatic urachal cancers are considered lethal, with reports of greater than 90% of patients dying of the disease in about 1 year (45,49,56). Metastases appear initially at the pelvic lymph nodes and later occur in the bladder (as surface implants), bowel, lungs, liver, brain, and/or bones (58).

The only curative treatment is surgical resection, which may achieve long-term survival if the

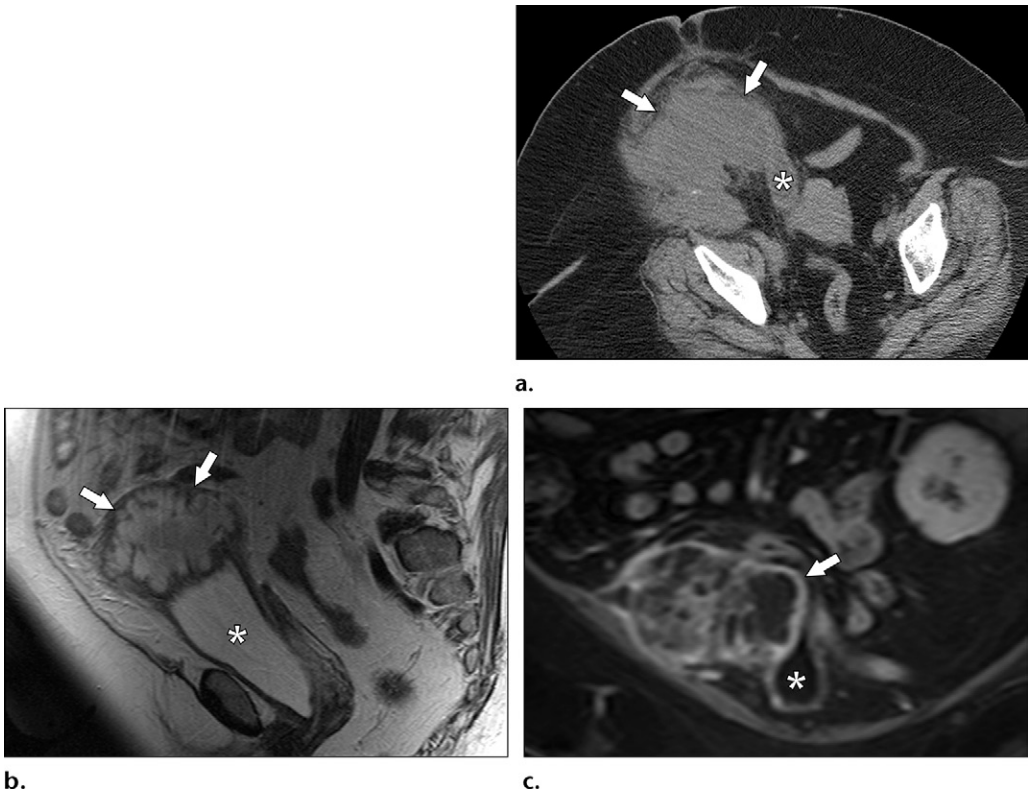


Figure 15. Right pelvic mass in a 66-year-old woman with a history of breast cancer, lower abdominal pain, and fever. ∗ = bladder. **(a)** Axial CT image shows a large soft-tissue mass (arrows) in the right lower quadrant that abuts and distorts the right superior aspect of the bladder. **(b, c)** Sagittal T2-weighted MR image **(b)** shows this lesion as a cystic and solid mass (arrows), which appears to be tethered to the bladder dome with multiple thickened enhancing septa on a coronal postcontrast image **(c)**. The differential diagnosis included an ovarian mass and an infected urachal cyst or malignancy. The lesion was resected and histopathologic analysis demonstrated the presence of pelvic malacoplakia.

tumor has been detected promptly. An aggressive surgical approach is warranted, including cystectomy and en bloc resection of the urachal mass, urachal tract, umbilicus, posterior rectus fascia, and pelvic lymph nodes, to avoid synchronous tumors along the urachal tract. Although an open surgical approach is typically favored, minimally invasive techniques could be appropriate in some instances, as they may be associated with less surgical morbidity, fewer operative complications, and a faster recovery (57,59).

Differential Diagnoses and Mimics

Urachal remnants, when symptomatic, can mimic numerous abdominal and pelvic diseases that may be clinically indistinguishable due to the nonspecific symptoms that urachal anomalies are often associated with. In addition, other entities can also mimic urachal anomalies at radiologic evaluation (Fig 15). Differential diagnosis of urachal anomalies can be narrowed by proper assessment of lesion location and morphology, specific radiologic findings, patient demographics, and clinical history (Fig 16).

Patent urachus and umbilical-urachal sinus may have similar clinical presentations to that of persistence of the omphalomesenteric duct. They may manifest as umbilical discharge, although the patent omphalomesenteric duct can be distinguished from urachal anomalies by demonstration of a communication between the anomalous tract and the bowel. Other navel pathologic conditions, such as umbilical granulomas, omphalitis, infected umbilical vessel, and granulation of a healing umbilical stump, can also be confused with urachal anomalies (11,19,30,34). The correct diagnosis can be confirmed initially by using cystography or fistulography to demonstrate a patent tubular structure extending from the bladder dome to the umbilicus in cases of a patent urachus, or the presence of a blind-ending focal dilatation at the umbilical end for an umbilical-urachal sinus (29,32). Rarely, other inflammatory diseases, including endometriosis involving the dome of the bladder, can mimic urachal lesions. US, CT, and MR imaging can also provide support for the diagnosis and should be performed in cases of doubt or suspected complications.

Urachal cysts generally are asymptomatic unless they attain a large size or become infected and in that setting can develop symptoms that may be

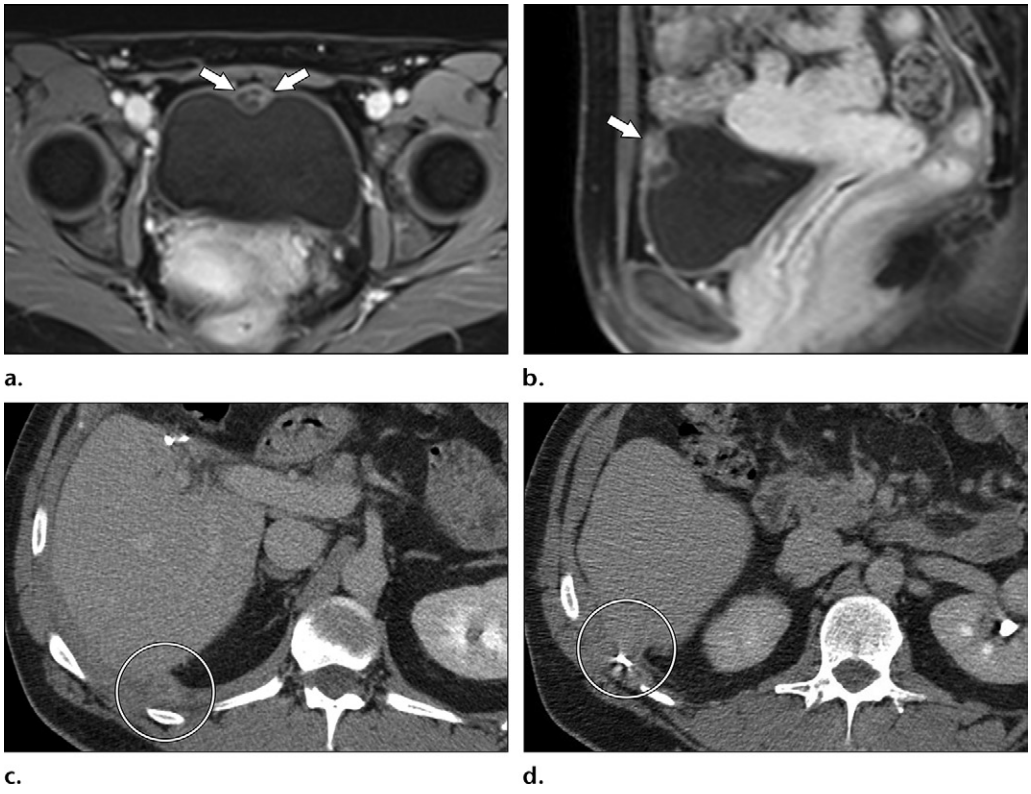


Figure 16. Mimic of a complicated urachal remnant in a 49-year-old woman with kidney stones. **(a, b)** Axial **(a)** and sagittal **(b)** gadolinium-enhanced fat-suppressed T1-weighted MR images reveal a midline loculated lesion (arrows in **a**) in the anterosuperior bladder wall with associated peripheral enhancement (arrow in **b**), raising the question of a complicated urachal remnant. The lesion proved to be an unsuspected inflamed focus of a dropped gallstone following cholecystectomy and not an infected urachal remnant or malignancy. **(c, d)** Axial CT images obtained 2 years earlier show the dropped gallstone (circle in **c**) and clip (circle in **d**).

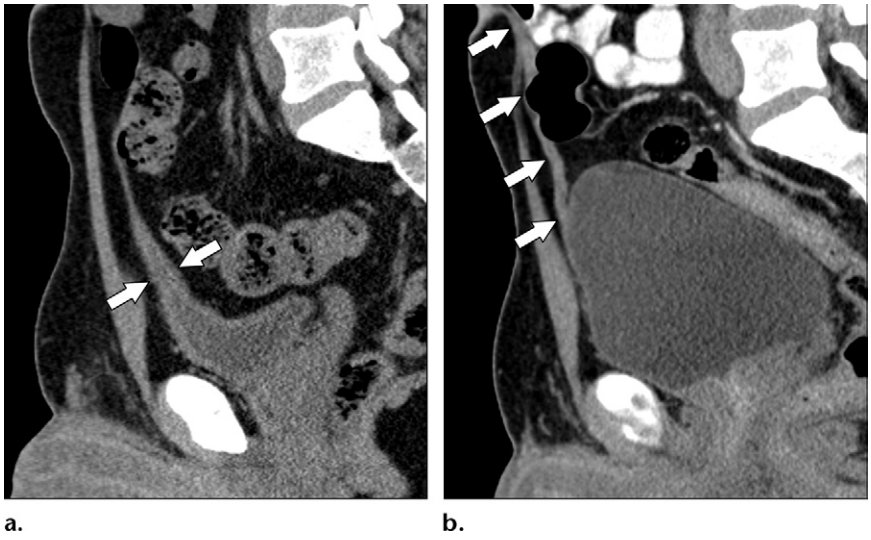


Figure 17. Vesicourachal diverticulum mimic in a 38-year-old man with right flank pain and a history of kidney stones. **(a)** Sagittal contrast-enhanced CT image shows a focal dilatation (arrows) of the urachus at the bladder end, raising the question of a vesicourachal diverticulum. **(b)** Follow-up CT image with optimal distention of the bladder shows an actual tubular connection (arrows) between the bladder dome and the umbilicus, most likely consistent with a patent urachus or urachal remnant rather than a diverticulum. As the patient was asymptomatic, no further workup was performed.

mistaken for those of other intra-abdominal and pelvic inflammatory processes, such as urinary tract infections, acute appendicitis, ovarian torsion, or complicated Meckel diverticulum (4,19,42). US, CT, or MR imaging can be helpful in differential diagnosis, providing better information regarding the exact anatomic location of the cyst and its relationship to both the bladder and the umbilicus.

Vesicourachal diverticulum can be mistaken for a bladder diverticulum, postsurgical collection, and other types of urachal remnants (Fig 17). US should show a vesicourachal diverticulum as an extraluminal outpouching sac at the bladder dome without any communication with the umbilical end further superiorly. If US findings are not definitive, CT and MR imaging can

be performed to help confirm the diagnosis (35). Sagittal reformatted multidetector CT images or sagittal images obtained at MR imaging may be helpful to better show the relationship with the bladder.

Urachal adenocarcinomas can be confused with other entities, including benign urachal tumors, nonurachal carcinomas of the bladder, and other adenocarcinomas from different organs such as the prostate, colon, and rectum (which may involve the bladder). Distinguishing these entities is important as they require different management strategies (49). Infection of a urachal anomaly may also mimic urachal adenocarcinoma, and the diagnosis is often challenging at imaging.

Conclusion

Urachal anomalies in children and adults may, if not detected and appropriately treated, cause considerable morbidity and potentially mortality. Knowledge and understanding of the normal embryonal development of the urachus and its morphologic characteristics is important to identify and diagnose these abnormalities.

Clinical diagnosis is often delayed because of nonspecific symptoms or the lack of symptoms. US is usually the modality of choice for initial assessment, especially in children, with CT and MR imaging both serving as robust modalities for further characterization and evaluation of complications, mainly infection and the development of malignancy. Early detection of urachal anomalies can help optimize an appropriate surgical approach if required and reduce the risk of subsequent development of malignancy.

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