



Congenital Urachal Anomalies: Embryology, Key Imaging Considerations and Management

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Abstract

Congenital urachal anomalies occur when the urachus, an embryologic structure arising from the allantois and urogenital sinus, fail to obliterate. The urachus is a tubular structure extending from the dome of the urinary bladder to the umbilicus. Partial or incomplete obliteration leads to various anomalies including patent urachus, umbilical-urachal sinus, and urachal cyst and vesicourachal diverticulum. These anomalies are uncommon and usually asymptomatic but can present with non-specific abdominal signs and symptoms. Here we describe the embryological and anatomical features of the different urachal anomalies as well as the relevant imaging findings to aid diagnosis and management.

Keywords: Urachal anomalies; Patent urachus; Urachus cyst; Umbilical-urachal sinus; Vesicourachal diverticulum

Introduction

Urachal anomalies arise when there is a partial or complete failure of the urachus to obliterate: The urachus is an embryological structure formed by the allantois and urogenital sinus. In the first month of gestation, the role of the urachus is to facilitate the removal of waste products through the placenta via the umbilical cord [1]. It normally obliterates by the time of birth, and failure to do so can lead to various urachal anomalies. These can present with various clinical problems in childhood. Although urachal anomalies are relatively uncommon with a reported incidence of 1/5000 adults, the increased use of cross-sectional imaging has led to increased detection of urachal anomalies [2], thus familiarity with the underlying anatomical, embryological and imaging findings are important for diagnosis and management.

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Embryology

The urachus is an embryonic remnant which becomes obliterated during fetal life forming a fibrous cord-like structure (urachus remnant) or median umbilical ligament [3]. Embryologically, the urachus develops from two structures called the allantois and the cloaca; the allantois appears on day 16 as a finger-like outpouching from the caudal wall of the yolk sac. The allantois remains a transient extraembryonic portion of the urachus. Anteriorly, the cloaca is divided into two structures by the second week of gestation, including the primitive urogenital sinus posteriorly, which is the precursor of the fetal bladder. This is contiguous with the allantois ventrally and the cranioventral end of the bladder opens into the allantois at the level of the umbilicus. By the fifth month of gestation, the bladder descends into the pelvis, and the apical portion (the urachus) stretches and gradually narrows with eventual obliteration of the lumen by fibrous proliferation. In one-third of adults, it may still be visible at microscopic examination as a structure communicating with the bladder lumen, but is considered closed in terms of function by the latter half of fetal life [1,4].

Anatomy

Anatomically, the urachus is seen as a tubular structure extending from the anterior dome of the urinary bladder to the umbilicus. It is retroperic and lies in the extraperitoneal space of Retzius, between the transverse fascia and the parietal peritoneum. It is accompanied on both sides by the medial umbilical ligaments which are obliterated remnants of the umbilical arteries. It is normally between 3 cm to 10 cm in length [1,4]. Histologically, it is composed of three layers. In approximately 70% of cases, the innermost layer is transitional epithelium, and in the remaining cases columnar cells are seen. The middle layer is formed of connective tissue and the outer layer

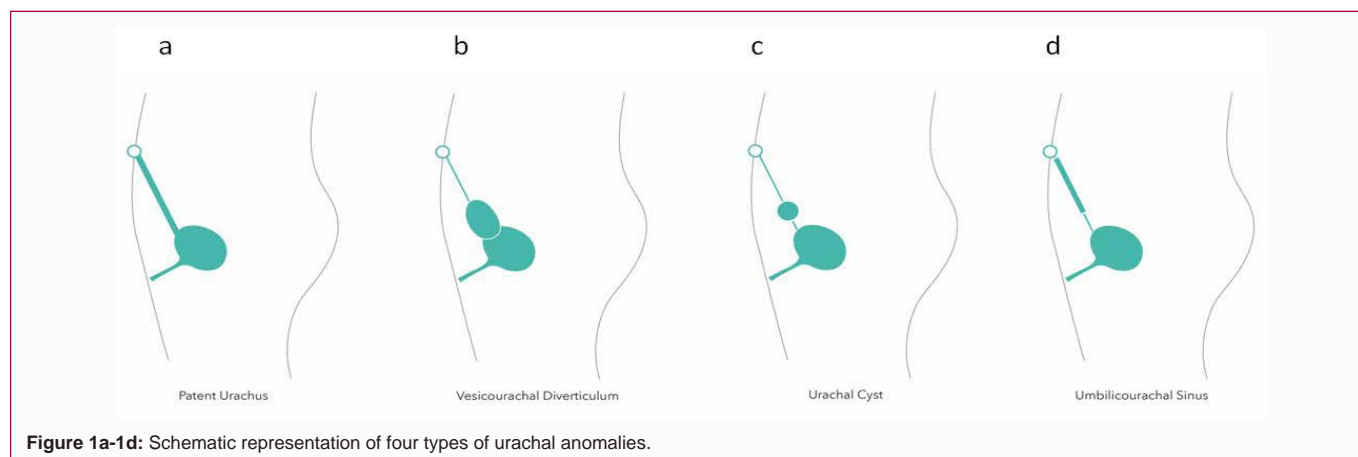


Figure 1a-1d: Schematic representation of four types of urachal anomalies.

is continuous with the detrusor muscle [2]. Incomplete involution results in urachal anomalies, which can be divided into four different groups including patent urachus (urachal fistula), umbilical-urachal sinus, and vesicourachal diverticulum and urachal cyst. The latter three are usually asymptomatic and may close at birth [1].

A patent urachus accounts for 47% of urachal anomalies [2], and occurs in 3 in 1,000,000 live births [1]. It is the complete failure of involution and is characterized by a persistent communication between the urinary bladder and the umbilicus. It is usually detected during the neonatal period due to urinary leakage from the umbilicus, and abnormal appearances of the umbilicus including edema and delayed cord stump healing [5]. A urachal sinus results when the umbilical end of the urachus fails to obliterate; the resulting blind-ending dilatation from the umbilical end, just deep to the abdominal wall, has no communication with the bladder. The sinus opening into the umbilicus can cause periumbilical pain, periodic discharge, especially in children where they can present with cloudy, serious or bloody fluid discharge. Most are asymptomatic however and discovered incidentally [1,2]. A urachal cyst is defined as the persistence of a focal portion, usually the lower third, of the urachus; it does not communicate with either the bladder or the umbilicus [1,2,6]. They are usually small and asymptomatic, unless they become infected, but can vary considerably in size [1,4,6]. A vesicourachal diverticulum develops when the umbilical end of the urachus completely obliterates without obliteration of the vesical end; this manifests as an outpouching from the anterior bladder dome [2,4]. These are also usually asymptomatic presenting as an incidental finding. As the diverticula usually have a large opening with good drainage into the bladder, there is reduced frequency of complications, such as, infection and intra-urachal stone formation, which are more commonly seen in the adult population [1,4]. This anomaly is usually seen in patients with chronic bladder outlet obstruction [4]. A schematic representation of the four urachal anomalies are depicted in Figure 1.

Imaging features of urachal anomalies

A diagnosis of a patent urachus can usually be made from the clinical history and ultrasound alone [5,7]. Sonographic features include a tubular structure with a hypoechoic wall and anechoic content extending from the bladder dome to the umbilicus (Figure 2a, 2b). Further complete evaluation can be done with fluoroscopy, either as a fistulogram or cystogram (Figure 2c, 2d), where the contrast delineates the tract extending from the urinary bladder to the umbilicus [4]. A patent urachus can act as a “pop off” valve for

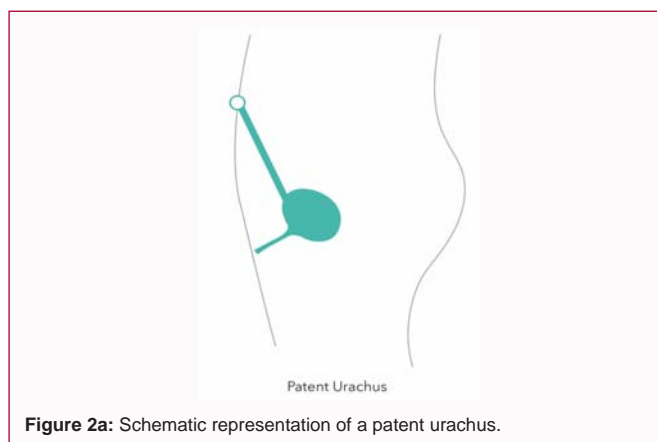


Figure 2a: Schematic representation of a patent urachus.

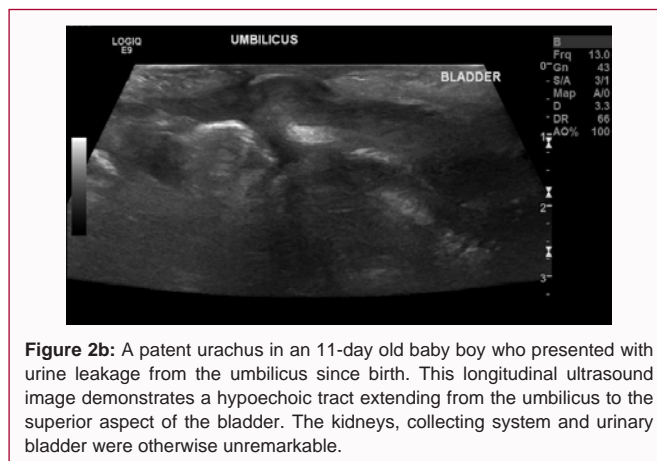


Figure 2b: A patent urachus in an 11-day old baby boy who presented with urine leakage from the umbilicus since birth. This longitudinal ultrasound image demonstrates a hypoechoic tract extending from the umbilicus to the superior aspect of the bladder. The kidneys, collecting system and urinary bladder were otherwise unremarkable.

congenital causes of bladder outlet obstruction (such as posterior urethral valves and urethral atresia which are known associations), and thereby provide some protection to the upper renal tract [8,9].

Similar to a patent urachus, ultrasound of an umbilical-urachal sinus will demonstrate a fusiform, thickened tubular blind-ending structure in the midline below the umbilicus with no communication to the bladder (Figures 3a-3d). Further characterisation can be done with a fistulogram, where contrast injected into the sinus does not track into the urinary bladder [2].

Ultrasound is the imaging modality of choice for urachal cysts and demonstrates the anechoic fluid cavity in the midline of the lower abdominal wall along the theoretical course of the urachus (Figures

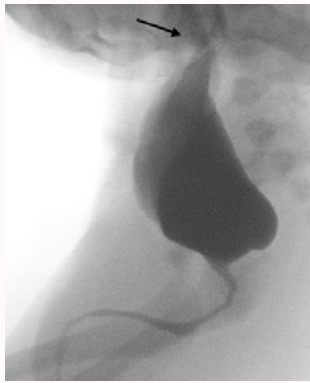


Figure 2c: An anteroposterior contrast study with retrograde injection of contrast into the orifice of the channel at the umbilical end (arrow).

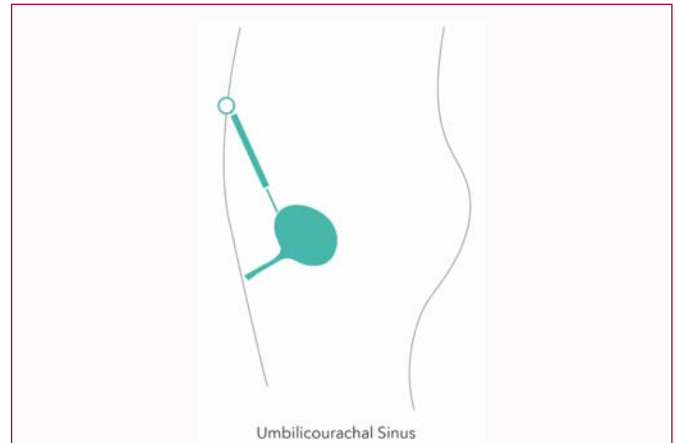


Figure 3a: Schematic representation of an umbilical-urachal sinus.

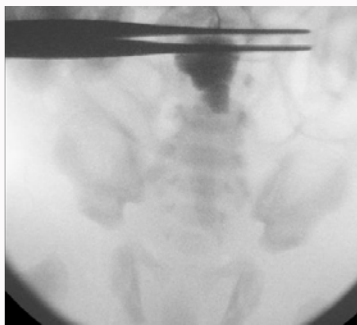


Figure 2d: A delayed oblique image then demonstrates a tract extending from the umbilicus to the anterosuperior bladder. The urethra is normal. The baby underwent surgical treatment and was discharged home two days post-surgery with urinary catheter and followed up with repeat normal urinary tract ultrasound and resolution of symptoms.



Figure 3b, 3c: A three-week-old baby girl presented to the emergency department with fever, erythema and purulent discharge from the umbilicus. A longitudinal grey scale ultrasound image b) demonstrates a focal echogenic fluid collection with a tract extending inferiorly towards but not contiguous with the urinary bladder. The color Doppler image c) demonstrates increased vascularity. A diagnosis of an infected umbilical-urachal sinus was made and the patient was treated with antibiotics.



Figure 3d: A follow-up abdominal ultrasound following completion of antibiotic treatment was performed. This longitudinal image demonstrates complete resolution of the umbilical-urachal remnant.

4a-4c). CT or MR may be required to assess for complications or to exclude alternative diagnoses, particularly in the adult population. It typically demonstrates a homogenous fluid attenuating lesion posterior to the abdominal wall on CT, and it is of high signal on T2-weighted MR sequences. It may mimic a soft tissue mass and the most important imaging differential is that of adenocarcinoma. Urachal neoplasms are rare, accounting for <1% of bladder cancers, and they usually present with advanced disease, local invasion or systemic spread. Urachal adenocarcinomas are generally large cystic or solid (or mixed) tumors with extravescal components and ultrasound demonstrates complex echogenicity and calcification. On CT, there is usually a midline mass adjacent to the bladder dome with heterogenous low attenuation components, representing mucin content. Peripheral calcification is present in 70%; calcification in a midline soft tissue mass along the urachal tract is considered pathognomonic for urachal adenocarcinoma [2]. Of note, urachal cysts predispose to adenocarcinoma due to prolonged urinary stasis and associated infection [4].

The vesicourachal diverticulum on ultrasound will demonstrate an extraluminal fluid-filled sac protruding from the bladder without communication to the umbilicus (Figures 5a-5c) [4]. On CT and MR, a midline cystic lesion is seen but the differentiating feature from a urachal cyst is clear communication with the bladder and its anterosuperior location.

Complications

Common complications include infection of the urachal

remnant, presenting with a range of non-specific symptoms including abdominal pain, fever, erythema, purulent urinary discharge and

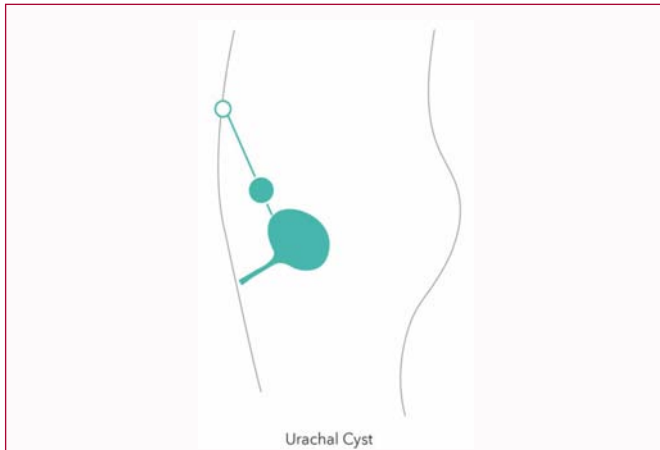


Figure 4a: Schematic representation of a urachal cyst.

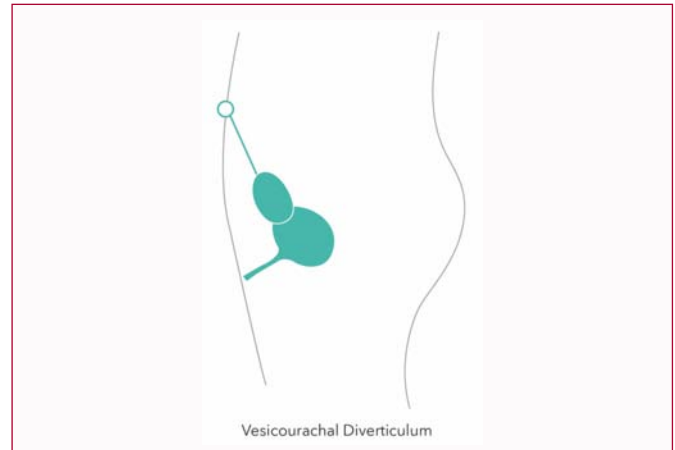


Figure 5a: Schematic representation of vesicourachal diverticulum.



Figure 4b: Sagittal reconstructed CT urogram in a 68-year-old patient presenting with hematuria. There is a collection of fluid in the midline of the anterior abdominal wall, between the umbilicus and pubis (arrows). There is some adjacent fat stranding in keeping with an infected urachal cyst. The patient was treated for a urinary tract infection with oral antibiotics.

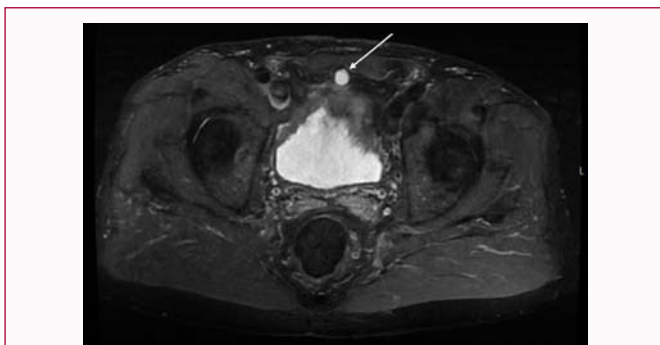


Figure 4c: An incidental finding of a urachal cyst on a MR pelvis performed for a 59-year-old female patient with hip pain. Axial T2-weighted MR image shows a well-defined T2 hyperintense cyst (arrow) abutting the anterior and superior surface of the bladder.



Figure 5b, 5c: A 62-year-old patient presented with right sided loin to groin pain, increased urinary frequency and dysuria. The axial (b) and sagittal (c) reconstructed CT urogram images demonstrate a midline tubular structure arising from the anterosuperior segment of the dome of the bladder. This does not communicate with the umbilicus. Note the surrounding fat stranding related to infection of the vesicourachal diverticulum. There are multiple punctate calcific foci within the diverticulum (arrow) indicative of evolving stone formation within this structure.

occasionally a palpable mass. The route of infection may be through lymphatics, hematogenous or *via* the urinary bladder [1]. Severe infection, although unusual, can result in complex fistulas and abscesses, and rupture with resultant peritonitis and sepsis [1,4].

Ultrasound is usually used as first line imaging in patients with suspected urachal remnant infection, but CT and MR imaging may be useful if ultrasound is non-diagnostic to delineate the anatomy or alternative diagnosis. Imaging features indicative of infection include the presence of urachal remnant with complex echogenicity and thickened walls on ultrasound or a heterogeneous appearance and

higher than water attenuation seen on contrast-enhanced CT [4]. An important differential diagnosis to an infected urachal remnant is urachal carcinoma, although this is extremely rare and is usually seen in the adult population.

Management

Currently, there is no uniform consensus for the management of urachal anomalies; most of the literature advocates surgical removal of the urachal remnant in both the pediatric and adult population regardless of symptoms to avoid future complications. However, the literature is scarce concerning the therapeutic value of prophylactic surgery, thus clinical cases need to be treated on an individual

basis. With infected urachus remnants and symptomatic patients, management usually involves antibiotic therapy and drainage [2]. Following resolution of infection, surgical excision is often required to reduce recurrence of infection and potential malignant degeneration [1]. Surgical resection typically involves exploration of the umbilicus *via* an infra-umbilical approach, which can be extended transversely if required or directly through the umbilicus itself. Resection can also be done laparoscopically, and it is important to delineate all structures within the umbilicus prior to excision of the urachal remnant. Care should be taken to ensure sound closure of the bladder dome, usually in two layers. Post-operatively, patients usually have an indwelling urinary catheter for 5 to 7 days with antibiotics prophylaxis. Post-operative histological assessment of the excised remnant is vital. For urachal malignancy, imaging features alone are not diagnostic and cannot reliably distinguish between tumor and infection, thus biopsy and/or fluid aspiration would be beneficial for diagnosis and treatment planning [10]. Definite management will depend on staging and degree of tumor differentiation.

Conclusion

Urachal anomalies are rare and may go unrecognized. There has been an increased frequency in detecting these anomalies with the increased use of imaging. It is therefore important to appreciate the embryological and anatomical features of these anomalies to understand the potential presenting signs and symptoms and complications that can occur. Ultrasound is the primary imaging modality for initial investigation, characterisation and diagnosis. Early detection of these anomalies is important to aid management and reduce risk of complications.

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