

## Urachal remnants: from embryology to clinical practice

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**Abstract:** The urachus is a tubular structure that is apparent on the third week and connects the ventral cloaca to the yolk sac, as a progression from the allantois. Following the normal regression procedure, the urachus remains as the median umbilical ligament.

Urachal remnants are present in 1.03% of paediatric patients while in 92.5% of cases represent incidental findings. Urachal anomalies are classified in four types as patent urachus (50–52%), urachal sinus (15%), urachal cyst (30%) and urachal diverticulum (3–5%). Ultrasound scan is the most commonly performed diagnostic imaging study.

In case of symptomatic urachal remnants, surgical excision is indicated. Asymptomatic urachal remnants that are diagnosed at the neonatal period or early infancy should be watched up to 6 months of age, as they are likely to resolve. In persistent or symptomatic urachal remnants there is a risk of inflammation or even malignancy development, therefore we believe that there is indication for preventive surgical excision that may be performed either open or laparoscopically or by robot-assisted laparoscopy.

**Keywords:** patent urachus, urachal sinus, urachal cyst, urachal diverticulum, urachal remnants, bladder outlet obstruction, malignancy.

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### Introduction

Urachal remnants occur in 1.03% of children with a male to female ratio equal to 2:1 and in 92.5% of cases they represent incidental findings [1, 2]. Four types of urachal remnants exist [3, 4]. A patent urachus results when the urachus completely fails to obliterate. A urachal cyst develops if the urachus closes at both the umbilicus and the



bladder but remains patent between these two end points. A urachal sinus arises when the urachus closes at the bladder end but remains patent as a blind dilatation at the umbilical end. A vesico urachal diverticulum develops if the urachus closes at the umbilical end but the communication with the dome of the bladder remains.

The aim of the present short review article is to summarize already established and recent evidence, concerning the genesis, clinical manifestation, diagnostic approach, and treatment strategy of urachal remnants.

## Embryology

By the third week the cloacal region is present at the tail end of the human embryo. The cloaca is divided by the urorectal septum into a ventral portion urogenital sinus and a dorsal portion (develops into hindgut). The cloaca communicates with the yolk sac via the allantois. As the urinary bladder descends, its connection with the allantois, the urachus, undergoes gradual regression [5]. The urachus is in fact the progression of the allantois into a tubular structure, which becomes apparent around the third week and connects the ventral cloaca to the yolk sac [3].

This process is completed by the 26th–28th gestational week, although the exact mechanism remains unclear. It is believed that it may be due to either a low growth rate of the urachus compared to the fetal body, or a rapid fibrous proliferation of the urachal wall, or hyperplasia or apoptosis of the urachal mucosa causing obstruction of its lumen [6].

Following its expected obliteration, the urachus remains as the median umbilical ligament. However, in cases of incomplete regression, the urachus may persist as a sinus, cyst or diverticulum or may remain completely patent.

## Anatomy

The urachus extends between the apex of the bladder and the umbilicus. It lies between the transversalis fascia and the parietal peritoneum. Intraperitoneal location of the urachus is uncommon but may cause obstruction or entrapment of an intestinal loop between the anterior abdominal wall and the urachus leading to ischemia [7]. In some cases, the urachus may lie closer to one of the umbilical arteries and deviate from the midline to the right or left [8]. The urachus is 3–10 cm long and 8–10 mm wide.

The urachus consists of an outer muscle layer bridging with connective fibers, a medial submucosal layer that contains connective tissue and lymph vessels and an internal layer lined by epithelium that may be either transitional or columnar (70% and 30% respectively) [6, 9]. The proximal end (vesicular) of the urachus contains a small opening that is present in 10% of adults but may be present more often in

early childhood. Along the urachus small slit-like openings called Luschka enclosures may be seen.

Ectopic small or large bowel mucosa, squamous epithelium, muscle or nerve tissue can be found along the wall of the urachal remnants [10]. A patent urachus may present a lumen along the whole of its course to the bladder or may terminate as an atretic muscular remnant. Rarely the umbilical end of the urachus is positioned on the midline but is caudal to the umbilicus [2]. The size of a urachal cyst varies but is usually small. The most common location is the lower third of the urachus while the less common is the upper third [2].

### **Clinical presentation**

Apart from a patent urachus, urachal remnants are usually asymptomatic. However, there is a strong predisposition to infections by Gram positive or negative bacteria via the lymphatic, hematogenous or vesical route [11].

The urachal sinus may present with intermittent mucous discharge from its external opening, which is visible. In case of infection, the discharge becomes purulent [4, 9].

A urachal cyst may obliterate during the neonatal period or early infancy and become evident later in adult life due to development of bladder outlet obstruction [3]. The most common clinical presentation is infection. *Staphylococcus aureus* in more than half of the cases and *Escherichia coli* are usually the bacteria involved. An inflamed urachal cyst presents itself with localized, continuous lower abdominal pain and septic fever [3, 12–15]. If the diagnosis is delayed, especially in younger patients, the inflamed cyst may rupture towards the peritoneal cavity resulting in peritonitis [16]. When surgical management of a urachal cyst is delayed the possibility of recurrent infection is greater than 30% [13].

Older children may complain of feeling that their bladder is pulled towards the umbilicus during micturition. In neonates with patent urachus the umbilical cord is soaked in urine, which, due to lower specific gravity, is absorbed by Wharton's jelly and the cord becomes giant. Parental concern is usually caused by constant wetting at the umbilical area although the umbilical cord stump has fallen off. In 14–33% of patients with a patent urachus the underlying cause is a bladder outlet obstruction; in these cases, the urachus acts as a valve relief mechanism [17]. In some cases, a patent urachus may be asymptomatic and present in adult life due to development of bladder outlet obstruction [9].

The urachal diverticulum is usually asymptomatic. Clinical manifestations may be due to either an underlying pathology such as bladder outlet obstruction, or the effects of the diverticulum itself. Especially in the neonatal period posterior urethral valves and prune belly syndrome need to be ruled out [2]. A bladder diverticulum is associated with urine stasis and urinary infection, urolithiasis or paradoxical flow of urine

during micturition. The possibility of developing malignant neoplasm after adolescence should not be overlooked [18].

Urachal remnants that are undiagnosed or untreated at childhood may be responsible for malignancy in adult life [2, 4]. This type of malignancy accounts for less than 0.5% of all bladder malignancies and becomes evident in men in 65–70% of cases, aged 40–70 years [19, 20]. These neoplasms develop in 90% of cases at the junction of the urachus to the dome of the bladder, in 6% at the middle and in 4% at the umbilical end [20, 21]. Due to their extraperitoneal location they are asymptomatic at early stages and gradually extend both towards the umbilicus and the bladder wall. They metastasize via the blood or the lymphatic route, initially to the pelvic lymph nodes and soon to lungs, bones, liver and brain [22]. Histology types include adenocarcinoma, transitional or squamous or anaplastic cell carcinoma, yolk sac tumor, malignant histiocytoma and rhabdomyosarcoma [20, 21, 23, 24]. The most common type is adenocarcinoma, in 90% of cases, that results from transitional epithelium metaplasia to columnar epithelium [20, 25]. Moreover, 34% of bladder adenocarcinomas result from metaplasia of urachal remnants [20, 25].

Villous adenoma, a precancerous condition, and inflammatory pseudotumor have been described in retrospective studies on the pathology findings of resected urachal remnants [20, 21, 23–25]. Rarely, benign neoplasms, such as adenoma, fibroma, fibroadenoma or fibromyoma may arise from urachal remnants [20, 21, 23–25].

## Diagnosis

Differential diagnosis of an umbilical swelling includes omphalitis, umbilical granuloma, omphalomesenteric duct remnants and urachal remnants. It should be noted that urachal remnants are up to 10 times more common compared to omphalomesenteric duct remnants. Griffith *et al.*, describe an infant with patent both the urachus and the omphalomesenteric duct [26]. Kranbuhl *et al.*, report a girl with omphalomesenteric duct remnants and urachal remnants [27].

On examination a painful and firm swelling at the hypogastrium, along the midline or even laterally, may represent an inflamed urachal cyst [28]. The overlying skin up to the umbilicus may be erythematous. On inspection of the umbilicus a possible orifice of a fistulous tract must be noted. If present, as in cases of a patent urachus or a urachal sinus, the opening should be dilated with a fine probe and then catheterised so that a fistulogram can be performed. In case of a patent urachus, the visualized tract runs a course through the anterior abdominal wall to the bladder. The fistulous tract may reach the apex of the bladder or may terminate as an urachal cyst. In case the tract is blind ending, the diagnosis is urachal sinus.

A retrograde cystourethrogram should be part of the diagnostic work up to rule out bladder outlet obstruction. Furthermore, it may reveal a diverticulum at the dome

of the bladder or vesicoureteral reflux or it may indirectly show an urachal cyst as a smooth filling defect at the dome of a full bladder [29]. In 34% of cases a concomitant urinary disorder is diagnosed; in  $\frac{3}{4}$  it is vesicoureteral reflux [29, 30].

Ultrasound scan is the most commonly used imaging study in the diagnosis of urachal remnants. It is an accurate diagnostic modality for urachal cysts and urachal diverticula. Gleason *et al.*, in a retrospective study, report that in 663 out of 721 (92%) of patients with urachal remnants, the diagnosis was made by ultrasound [1]. In cases of a patent urachus ultrasound diagnosis may be achieved by longitude planes, in which a thickened tubular structure along the midline and below the umbilicus is shown. Ultrasound sonography is also helpful [6, 9, 31].

Computer tomography (CT) or magnetic resonance imaging (MRI) may be required in some cases. A patent urachus is visualized as a fluid filled tubular structure extending from the dome of the bladder anteriorly and superiorly towards the umbilicus [3]. It runs a course that is parallel to the medial umbilical ligaments, which derive from the occluded umbilical arteries [3].

In cases of urachal diverticulum, the ultrasound scan shows a thick-walled cyst that may be fluid filled and is located on and communicates with the dome of the bladder. On cystourethrogram, the diverticulum or its effects on the bladder may become evident. Finally, a urachal diverticulum may be an incidental finding on CT [32, 33].

A urachal cyst is diagnosed by ultrasound scan or CT and is shown as an intramural cystic structure along the middle or lower third on the midline of the anterior abdominal wall. Indications of an inflamed cyst include wall thickening or hyper echogenicity of its contents with debris [14, 32, 34–36]. Radiology-guided aspiration and culture and sensitivity testing may be required for an inflamed urachal cyst; in such cases definitive surgical treatment is postponed [12, 13, 34, 37].

Al-Hindawi *et al.* reported a case of an adult with a non-inflamed urachal cyst that showed eggshell calcification of its wall on imaging [28]. It is of note that eggshell calcifications on ultrasound or CT should raise suspicion of malignancy until otherwise proven. On the other hand, the presence of calcifications in a cystic or solid or mixed mass above the bladder may represent malignancy on the ground of urachal remnants [8, 38–41].

### Treatment strategies

Surgical excision is indicated for symptomatic urachal remnants [42]. According to a retrospective clinical study from Gleason *et al.*, 54 out of 721 patients (7.29%) with urachal remnants were symptomatic [1].

Management of asymptomatic urachal remnants remains under discussion. Issues yet to be addressed include the possibility of regression on one hand, and the potential

risks of complications on the other, mainly the possibility of developing malignancy [29]. Galati *et al.*, reported that 80% of patients with urachal remnants showed full resolution within the first six months of life and proposed that small urachal remnants diagnosed in the neonatal period or early infancy should be treated conservatively up to six months of age [30]. Naiditch *et al.* noted that 78 out of 103 patients with urachal remnants were symptomatic. Out of 19 patients treated with watchful waiting, 15 (78.9%) exhibited full resolution [43].

With regards to malignancy potential in cases of urachal remnants, the presence of epithelium may prove to be a significant point. It is well known that epithelium may undergo metaplasia and development of malignancy. This risk is limited when urachal remnants contain fibrous and muscle tissue only without epithelium. Copp *et al.*, tried to correlate histopathology with clinical course in 29 patients with urachal remnants, who were treated surgically [29]. According to clinical presentation, patients were grouped as asymptomatic — Group A (n = 5) and symptomatic — Group B (n = 24). On histopathology 3 out of 5 patients in Group A had epithelial tissue, as opposed to 17 out of 24 in Group B. This result however was not statistically significant ( $p = 0.63$ ) [29].

In cases of urachal remnants that remain without signs of regression but are asymptomatic, the risk of developing malignancy or inflammation still exists [10, 29, 30, 44]. Therefore, we believe that preventive excision is indicated [1].

Urachal remnants can be approached via the open or the laparoscopic route [44]. Fode *et al.*, performed robot-assisted laparoscopic en block resection in 9 patients with symptomatic urachal remnants. In 3 out of 9 patients the procedure was converted to open due to inadvertent damage to transversalis fascia while one patient experienced a spleen injury. That method is considered as promising, despite the small sample size [42].

Most authors report that their patients had an uneventful postoperative course without complications [10, 44]. On the other hand, Naiditch *et al.*, report that postoperative complications occurred in 5 out of 34 patients [43]. Data from larger series are required, in order to shed light on the complications incidence rates.

### Conflict of interest

None declared.

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