

“CONGENITAL ANOMALIES OF URACHUS” EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE

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ABSTRACT

The urachus is developmentally the upper part of the bladder, both being derived from the ventral cloaca. This part of the bladder narrows more, but retains in miniature. It is then known as the urachus. The urachus shares in the post-partum descent of the bladder, and in the adult is a cone-shaped structure passing up from the bladder apex, and attached to the umbilicus only by adventitial bands derived from the umbilical arteries which it has pulled down in its descent. Like the bladder it has a peritoneal and an extra-peritoneal surface and has the transversalis fascia on its outer and the peritoneum on its inner surface. The lumen of the urachus remains patent throughout life, though it may be plugged in places by masses of epithelial cells which have peeled or come off in scales from the walls of its canal.

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Introduction

The fibrous remnant of the allantois, a channel between the bladder and the umbilicus is called urachus. In fetus, urine initially drains from urachus during first trimester of pregnancy. The channel of the urachus usually seals off and eradicates around the 12th week of gestation and what left is a small fibrous cord between the bladder and umbilicus called the median umbilical ligament.

Urachal anomalies have been described into four: Patent urachus, Urachal cyst, Urachal sinus, Vesicourachal diverticulum. Patent urachus also known as urachal fistula, Failure for the lumen of the urachus to be filled in leaves a patent urachus. The sign is leakage of urine through the umbilicus. Patent urachus needs to be surgically removed. Urachal cyst is a sac-like tissue that develops in the urachus. Urachus is a primitive structure that connects the umbilical cord to the bladder in the developing baby. Although it normally disappears prior to birth in some people, part of the urachus may remain in some people. Urachal cysts can develop at any age, but mostly affects older children and/or adults. Urachal cysts are not often associated with any signs or symptoms unless there are few complications such as infection. In these cases, symptoms may include abdominal pain, fever, pain with urination and hematuria. Treatment includes surgery to drain the cyst or remove the urachus. Urachal sinus is congenital anomalies of the urinary bladder caused by failure of obliteration of proximal or distal part of allantois, and the presentation of this anomaly is common in children and rare in adult. Vesicourachal diverticulum is the persistence of a segment of the urachus, presents as a protrusion at the vertex of the bladder. It may predispose to urolith formation [1].

The urachus, or median umbilical ligament, is a 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 and the allantois, which is a derivative of the yolk sac. 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 pre-surgical diagnosis is not easily made. Various abnormalities can be confusing unless one is familiar with the basic embryologic anatomy and imaging features of the sub-umbilical and pre-vesicle 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 [8].

Incidence

An urachal remnant occurs in approximately 1 in 5000 patients [9].

Ontogenesis for the normal development of urachus

The human allantois is thought to be originated from the posterior wall of the yolk sac, which extends into the connecting stalk forming a small diverticulum. The allantois initiates placental development, which is the site for Extra Embryonic Vasculogenesis. This occurs independently in the mesodermal wall of the allantois and contributes to the formation of the umbilical vessels. Allantoic vasculogenesis arise from the distal tip of the allantois. Distal tip of allantois contains mesoderm and both progresses distally, to fuse with the chorion and form the placental labyrinth. Proximally, progresses and connects with the dorsal aorta. The proximal portion of the urachus is the allantois and is continuous with the urinary bladder.

After birth, a median umbilical ligament is formed which is a fibrous stalk like structure. Median umbilical ligament extends from the urinary bladder to the umbilical region [5].

Ontogenesis for the abnormal development of urachus

The urachal abnormalities results from an incomplete closure of the allantois canal. There are four common abnormalities of urachus, patent urachus, urachal sinus, Urachal cyst, Vesicourachal diverticulum.

Patent urachus also known as urachal fistula, is a congenital disorder caused by the persistence of the allantois. Normally, the urachus seals to become the median umbilical ligament. Whereas in this case, the urachus remains open and urine drains from the bladder through an opening in the umbilicus. This occurs when the urachus did not seal off and there is a connection between the bladder and the umbilicus. Leakage of urine through the umbilicus is the main sign of patent urachus. A patent urachus can cause varying amounts of clear urine to leak at the umbilicus [3].

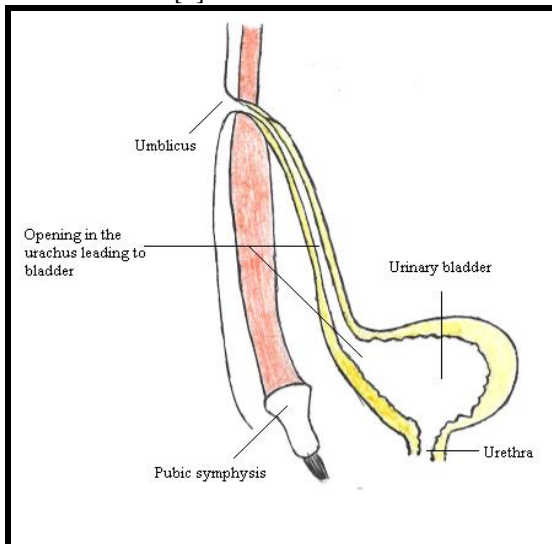


Fig 1. Diagrammatic representation of patent urachus.

Urachal sinus or umbilical-urachal sinus is congenital anomalies of the urinary bladder caused by failure of obliteration of proximal or distal part of allantois. This occurs when the urachus does not seal close to the umbilicus and leads to a blind ending tract from the umbilicus into the urachus to form a sinus. These can be asymptomatic or present with infection with abdominal pain and drainage of fluid [3].

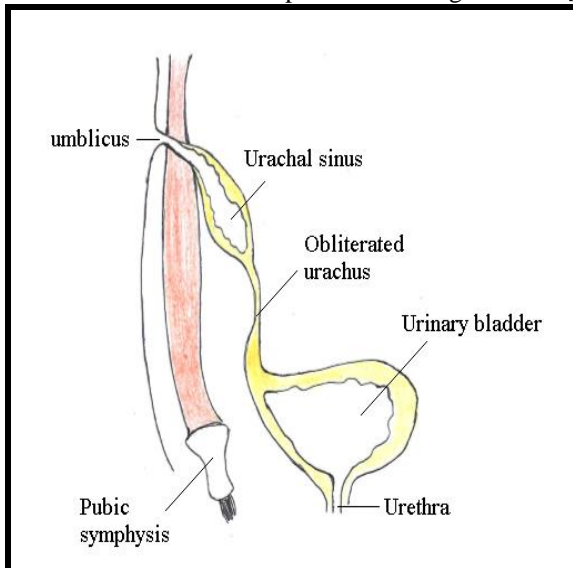


Fig 2. diagrammatic representation of urachal sinus.

Urachal cyst is a sinus remaining from the allantois during embryogenesis. It is a cyst formed in the remnants between the umbilicus and bladder. Urachal cyst is a type of cyst occurring in a determined portion of the urachus, presenting as an extra-peritoneal mass in the umbilical region. If infected, it is characterized by abdominal pain and fever. It may lead to peritonitis if there is any rupture or it may drain through the umbilicus [6].

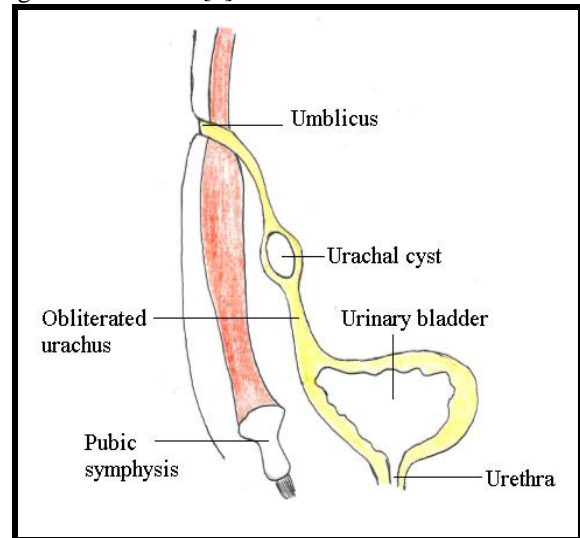


Fig-3: diagrammatic representation of urachal cyst.

Vesicourachal diverticulum this occurs when the urachus does not seal close to the bladder and leads to a blind ending tract from the bladder into the urachus called a diverticulum. These also can be asymptomatic or present with a urinary tract infection [24].

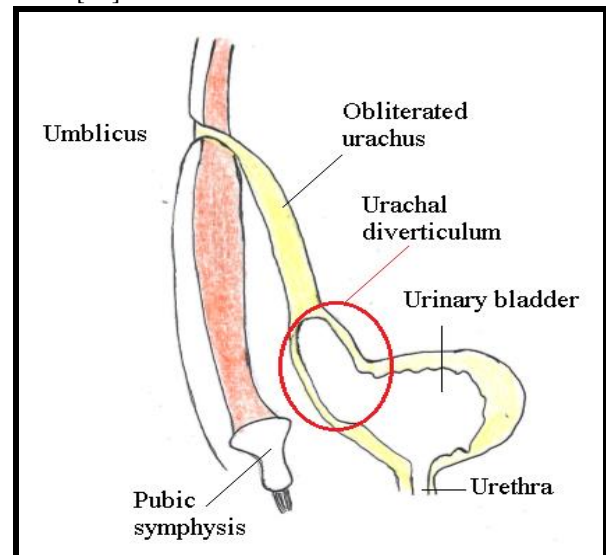


Fig 4. diagrammatic representation of vesicourachal diverticulum.

Discussion

Urachal anomalies are rare, and mostly found in early childhood. Four clinical urachal anomalies have been described: patent urachus, urachal cyst, urachal sinus, and vesicourachal diverticulum. Urachal sinus was the most common anomaly. It occurs in infancy or childhood. Sonography is the most accurate modality for diagnosis in these patients. The small fistula tract and sinus can be clearly seen from the ultrasound image. This may be because the pre-peritoneal space is free of interfering bowel gas. An extra-peritoneal excision is the treatment of choice for patients with urachal sinus. A patent urachus was the next common diagnosis in this anomaly [25].

It occurred most often in neonates with a prominent everted large umbilicus along with visible mucosa and a large fistula tract. Fistulography with the use of radiopaque contrast medium was helpful in the diagnosis. The concern that a patent urachus may be secondary to an infravesical urinary tract obstruction has justified the use of voiding cystourethrography with or without subsequent endoscopy to investigate this anomaly. Voiding cystourethrography was performed in all our cases to identify the fistula tract and, more importantly, to rule out the concomitant presence of bladder outlet obstruction or vesicoureteral reflux. In contrast to the study of Herbst, none of our patients had infravesical lesions. Therefore, it is unlikely that a persistent urachus was directly associated with the obstruction. Excision of the urachal tract to the bladder through an extraperitoneal approach with or without inclusion of the bladder cuff is the treatment of choice. Most urachal cysts in this series were infected, with pain, tenderness, erythema, and localized swelling in the infraumbilical region, when diagnosed. Sonography always revealed abscess formation, and an infected urachal cyst was thus suspected. Incision and drainage comprised the initial treatment in 3, and in only 1 patient did we need to remove the residual cyst remnant owing to persistent wound infection. *Staphylococcus aureus* was the most common bacterium cultured in this study. In conclusion, urachal abnormalities are rare. The optimal diagnostic imaging study depends on the clinical presentations. Our experience suggests that in young infants and neonates suspected of having a patent urachus, fistulography should be performed. The treatment of choice is excision of the patent urachus with or without inclusion of the bladder cuff. Any child who presents with a wet umbilicus should receive a sonographic examination to rule out the possible diagnosis of a urachal sinus. Surgical excision of the sinus is the goal of treatment. Nevertheless, most urachal cysts are asymptomatic until they become infected. Incision and drainage or delayed excisions with initial antibiotic therapy are the treatment of choice. Voiding cystourethrography does not seem necessary in view of the fact that none of the patients studied had an additional associated urinary tract anomaly.

Thirty-five children with anomalies of the urachus which have required surgical management have been encountered in this institution over a 20-year period. Of these 35, 19 were classified as patent urachus, 12 as urachal cyst, and 4 as urachal sinus. All cases were treated by excision or drainage. There was one death. Gastrointestinal and other genitourinary anomalies were commonly seen in these patients. Intravenous pyelography was helpful in identifying unsuspected associated genitourinary anomalies while cystography was not. Investigation of the gastrointestinal tract does not appear to be warranted in the absence of symptoms [12].

Conclusion

In humans, the allantois is very small and appears to be a rudimentary structure. Urachal anomaly is very rare and often ruled out in the early childhood. The allantois is essential for development to continue because its presence is required to initiate vascularization of the chorion [30].

The endodermal epithelium of the allantoic diverticulum originates from the most caudal limit of the yolk sac roof and grows into the adjacent body stalk mesoderm. Immediately, the allantoic vessels differentiate and grow distally to vascularize the chorionic mesoderm. A short time later, the allantoic region of the yolk sac roof will be incorporated into the body of the embryo to form the ventral wall or floor of the

hind gut. The development, the proximal or embryonic portion of the allantois is referred to as the urachus. Later the urachal portion of the allantois retains its ventral attachments to the urogenital sinus and extends cranially to the umbilicus. Eventually the urachus loses its lumen and becomes converted into fibrous cord like structure which persists in the adult as median umbilical ligament. Any rupture or disturbance in this process leads to the four characteristic anomalies [11].

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