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Patent Urachus

Authors:

Swayam P. Nirujogi, MD
Rotating Intern, Sanford Medical Center, Bemidji, Minnesota

Giridhar Guntreddi, MD, MD FAAP
Hospitalist, Sanford Medical Center, Bemidji, Minnesota

Jayasree Vasudevan Nair, MD FAAP
Hospitalist, Sanford Medical Center, Bemidji, Minnesota

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A newborn boy, born to a 21-year-old gravida 2, para 2 mother with a history of tobacco use in our buprenorphine treatment program, was admitted to a level 2 nursery for continuous monitoring and drug screening.

Urine drug screening test results from the patient and his mother were positive for buprenorphine. Finnegan Neonatal Abstinence scores were progressively worsening, so the patient was started on morphine. On day 11 of life, the patient had a fever with a maximum temperature of 38.4°C, and he became fussier than usual. On physical examination, he was noted to have umbilical discharge (**Figure 1**).

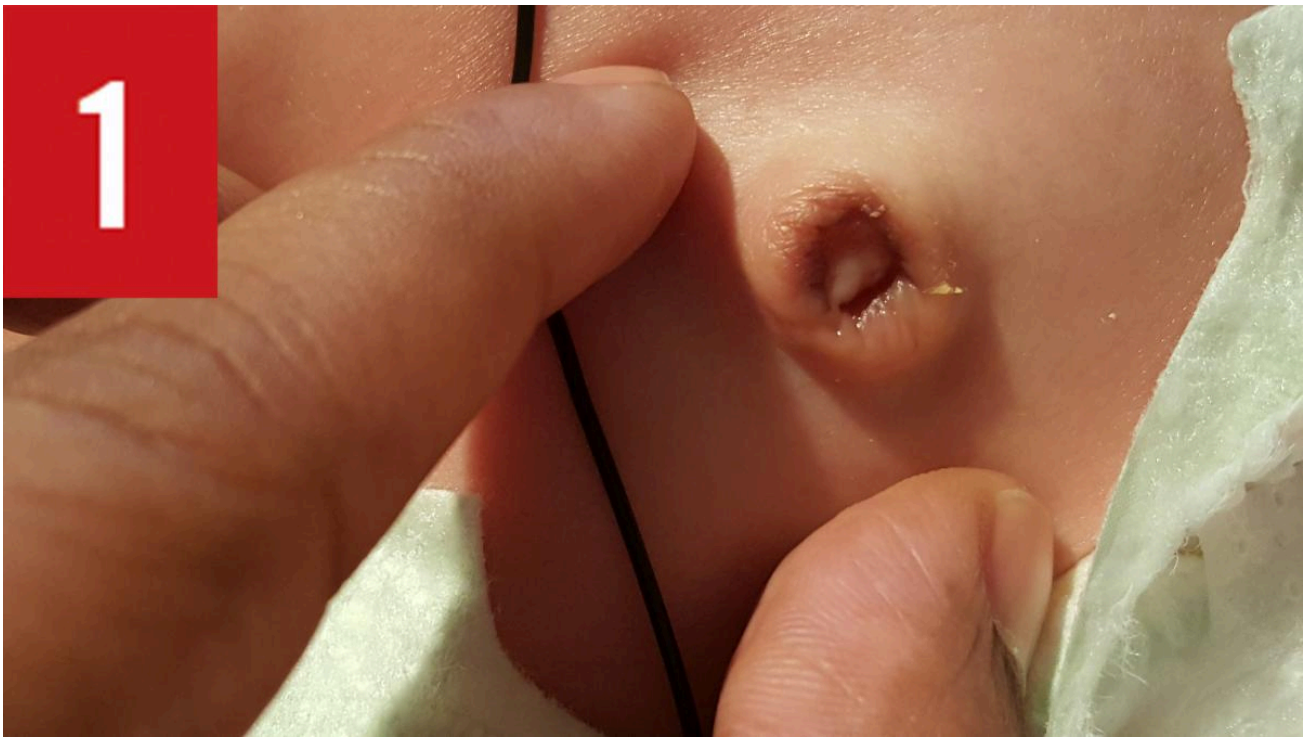

Figure 1. *Umbilical discharge.*

Laboratory tests. A workup for sepsis was performed, as well as a complete blood cell count, a C-reactive protein level, blood culture, urinalysis and culture, and umbilical discharge culture. The patient was started on intravenous ampicillin and gentamicin pending culture results. Subsequently, urine culture showed 40,000 CFU/mL of *Escherichia coli* and 10,000 CFU/mL of *Enterococcus faecalis*. Umbilical discharge culture showed rare group B streptococci (*Streptococcus agalactiae*) and rare coagulase-negative staphylococci, both of which were pansensitive to all antibiotics. Because the patient had a urinary tract infection (UTI) in the presence of umbilical drainage and the presence of *E faecalis*, an embryologic defect was suspected. Therefore, ultrasonography of the umbilicus and kidneys and bladder was performed to delineate the anatomical defect (**Figure 2**).

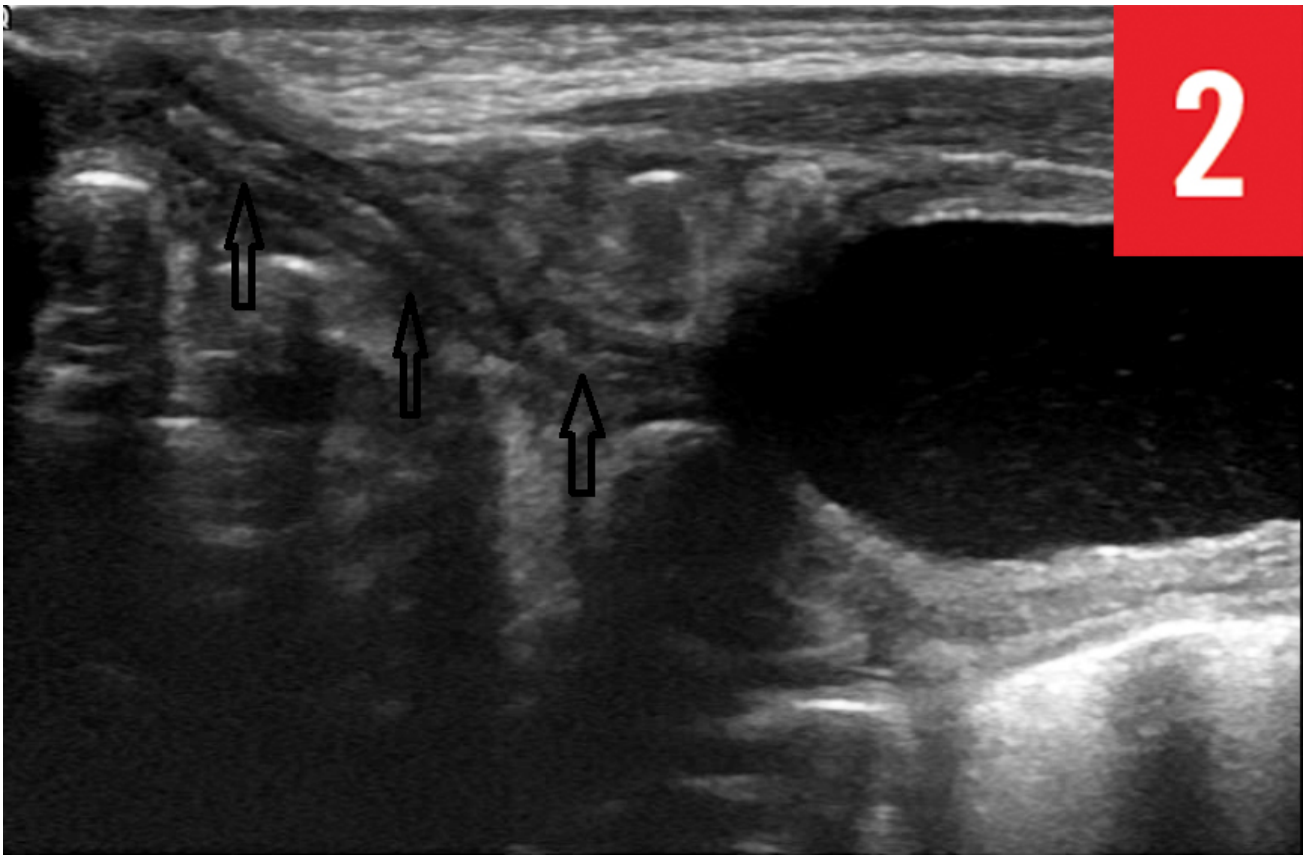

Figure 2. *Ultrasonogram of patient's abdomen and bladder. The arrows show the tubular connection from the bladder to the patient's umbilicus, suggestive of patent urachus.*

Diagnosis. Considering the physical examination findings, the ultrasonography findings, and the patient's presentation, a diagnosis of patent urachus was made. The ultrasonogram of the abdomen showed hypoechoic bands extending from the bladder dome to the umbilicus, suggestive of a patent urachus anomaly.

Discussion. Patent urachus is a rare disorder with an estimated prevalence of 1 to 2 cases per 100,000 deliveries. The urachus is the intra-abdominal remnant of the embryologic allantois, which is seen approximately 16 days after conception as a diverticulum from the caudal wall of the yolk sac.

An estimated 2% of the population has a urachal anomaly. The most common types of are patent urachus, which is a communication between the umbilicus and bladder; urachal sinus, which is when the umbilical end is open but there is no communication with the bladder; urachal diverticulum, which is when a cap forms on the dome of the bladder; urachal cyst, which is when the central part of the tract is patent and fills with fluid; and atretic urachal remnant or urachal chorda, which is when the entire tract persists as a cord.

Patent urachus accounts for approximately 10% to 15% of urachal anomalies. Urachal anomalies are associated with recurrent UTIs, sepsis, and neoplastic transformation such as

urachal adenocarcinoma, which is seen in late adult life. Patent urachus frequently coexists with congenital lower urinary tract obstruction and is associated with posterior urethral valves (PUVs) in approximately one-third of cases.[1]

The urachus is an embryologic canal that connects the superior pole of the developing bladder to the umbilicus. In the fourth week of embryogenesis, the flat trilaminar embryonic disc folds and becomes a cylindrical C-shaped fetus, which narrows the opening of the yolk sac to the embryo. This narrowed opening contains umbilical vessels, the urachus, and the omphalomesenteric duct. The omphalomesenteric duct connects the yolk sac to the developing gut. At the same time, the allantois—a diverticulum of the caudal hindgut—forms bands and becomes the urachus. Both the omphalomesenteric duct and the urachus involute. Occasionally, this obliterative process is incomplete, leading to a persistent urachal remnant.[2]

The clinical presentation of patent urachus varies. It most commonly presents with umbilical discharge and abnormal appearance of the umbilicus. However, it can also present as a prominent everted large umbilicus along with visible mucosa and a large fistula tract. Fistulography with the use of radiopaque contrast medium can be helpful in the diagnosis. An umbilical cystic mass, which diminishes in size over the course of pregnancy, is a suggestive prenatal sonographic finding for urachal anomalies.[3] Infected urachal cysts may present with fever, UTI, abdominal pain, an intra-abdominal mass, or peritonitis. Ultrasonography is the preferred initial diagnostic modality, because it can quickly differentiate between urachal anomalies, an umbilical hernia, and local abscess. If the anatomy is unclear, computed tomography may be helpful.[4] A voiding cystourethrogram is not necessary unless there are concerns about obstruction such as from PUVs.

Treatment. Owing to the rarity of this condition, the literature mainly consists of case reports. Few case reports of nonoperative management of urachal remnants have been published. Although obliteration of the urachus was originally thought to be a prenatal occurrence, recent literature suggests that this process may occur postnatally, as well. In 1998, Zieger and colleagues performed ultrasonography on 102 asymptomatic infants and found a morphologic variant of a urachus in all of them. Infants who underwent a second ultrasonography scan 3 to 5 months later (n = 70) showed spontaneous involution of the urachus. The researchers concluded that involution of the urachus is not complete at birth, and that infants with urachal anomalies may be observed in expectation of spontaneous resolution.[5]

In 2003, Ueno and colleagues reported one of the largest series of urachal remnants, 20 symptomatic cases and 36 cases incidentally diagnosed by way of ultrasonography.[6] The researchers concluded that surgical intervention should be avoided in all children younger than 1 year, and that surgical resection of patent urachal remnants should be restricted to children

older than 1 year with multiple clinical episodes.[6] Uninfected urachal cysts require surgical excision due to the risk of developing infection.

The optimal management of infected urachal cysts is unclear. A review of the literature suggests a 2-stage approach—drainage and antibiotics followed by excision. Few case reports suggest that infected urachal cysts obliterate with time after drainage. One case report of patent urachus with PUVs shows that initial urine diversion with a catheter and subsequent PUV repair led to spontaneous closure of the urachus.[7]

The most common surgical therapy for patent urachus is complete excision, with or without excision of the bladder dome. Low complication rates have been reported, with the most common complication being wound infection. Recently, laparoscopic excision has been described as an alternative therapeutic approach.

Outcome of the case. Our patient's case was followed by a pediatric urologist. Because there is no abscess or acute abdomen, the size of the urachus will continued to be monitored with ultrasonography every 3 months.

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