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Prenatal versus Postnatal Diagnosis of a Distended Bladder and Hydronephrosis
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Lower Pole Ureteropelvic Junction Obstruction in a Duplex System With Antenatal Bilateral Hydronephrosis
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Urinoma Misdiagnosed as Simple Renal Cyst in Prenatal Ultrasound Leading to Delayed Diagnosis of Posterior Urethral Valves
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Bilateral Abdominoscrotal Hydroceles with Concurrent Lower Extremity Edema: Antenatal Presentation and Postnatal Management
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An Interesting Presentation of Synchronous Cystic Pulmonary and Renal Masses
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A Case of Migrated Vesicoamniotic Shunt Requiring Urgent Repair of Abdominal Wall Hernia
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Prenatal Diagnosis of a Congenital Mesoblastic Nephroma
Alexandra Colvin BS, Amanda F. Saltzman MD, Alonso Carrasco Jr. MD, Nicholas G. Cost MD
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An Interesting Case of an Aberrant Urachal Remnant
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Prenatal versus Postnatal Diagnosis of a Distended Bladder and Hydronephrosis

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Case Presentation

A boy born at 36 weeks and 4 days gestation was transferred to the neonatal intensive care unit (NICU) for evaluation. His mother had been seen prenatally by Urology due to bilateral hydroureronephrosis and a “keyhole” appearance of the bladder (Figures 1 and 2). Previous ultrasounds had also demonstrated oligohydramnios, but at the time of the urologic evaluation, the amniotic fluid level had normalized. The mother was counseled on potential posterior urethral valves (PUV) with plans for follow up after birth.

After transfer, catheter placement was attempted unsuccessfully by the NICU staff, and Urology was consulted. On exam, the baby’s abdomen was noted to have a wrinkled appearance consistent with prune belly syndrome, and leakage was noted from the umbilicus, consistent with patent urachus. His testicles were not palpable. Ultrasound demonstrated a decompressed bladder, bilateral ureterectasis without hydronephrosis, and bilateral renal dysplasia.

On his fourth day of life, the baby underwent cutaneous vesicostomy and peritoneal dialysis (PD) catheter placement. Urethral atresia was also confirmed; therefore, utilizing antegrade and retrograde approaches, a 5Fr feeding tube was navigated through the meatus, into the bladder, and out through the vesicostomy to use for planned P.A.D.U.A. (progressive augmentation by dilating the urethra anterior) procedure.

Over the next 6 weeks, the catheter was upsized to 14Fr. At discharge, it was removed, and his parents were performing clean intermittent catheterization (CIC) without difficulty.

Currently, the urethra remains patent, and he is maintained on prophylactic antibiotics, twice daily CIC, daily digital vesicostomy dilation, and PD. Orchiopexy, abdominoplasty, and genitourinary reconstruction are being postponed until patient is ready for renal transplantation to avoid disruption of PD at his nephrologist’s request.

This case acts as a reminder to consider the “zebras” as not all boys with prenatal findings suggestive of fetal lower urinary tract obstruction (LUTO) will have PUV.

Discussion

Fetal LUTO presents with several characteristic ultrasound features including bilateral hydronephrosis, a dilated bladder, and a dilated proximal urethra, all of which were seen in our patient1. The most common etiology for this constellation of findings is PUV1; however, as demonstrated here, ultrasound cannot distinguish this entity from other—rarer—diagnoses.

Our patient is a classic presentation of the rare findings of urethral atresia with phenotypic features similar to those seen in prune belly syndrome and with fetal survivability due to a patent urachus. Management of these boys’ urethras has changed very little since the P.A.D.U.A. procedure as initially described in 19882, and this technique worked very successfully in our patient.

Treatment of these children’s urethras, however, is only one part of the extensive care that they need. These boys suffer the effects of compromised renal and pulmonary development3. Each child inevitably has his own issues and needs; however, it is important to continue reporting these cases in the literature in order to create a shared experience among clinicians and generate a collective conversation about how best to manage them.

References

Case Presentation

We present one case among an institutional series of patients with lower pole ureteropelvic junction obstruction (UPJO) in duplicated collecting systems. The patient was seen at our institution at 2 months of age with a history of bilateral antenatal hydronephrosis. He was born full term and had chromosome 1 and 7 abnormalities, dysmorphic facies, hypotonia, and an ASD. His exam was also notable for a nonpalpable left undescended testis. Postnatal imaging reported bilateral pelviectasis. VCUG was normal. The patient was started on continuous antibiotic prophylaxis and referred to our institution.

An ultrasound at 2 months demonstrated a normal right kidney and a duplicated left kidney with upper pole grade 2 hydronephrosis and lower pole grade 4 hydronephrosis. Subsequent MAG-3 lasix renal scan demonstrated 100% contrast retention in the lower pole, whereas the upper pole had a T 1/2 of 4 minutes with 37% contrast retention. Function was unable to be accurately calculated.

An ultrasound at 4 months demonstrated progressively worsening left lower pole hydronephrosis with global parenchymal thinning (Figure 1).

At 4 months of age, the patient underwent a cystoscopy, left retrograde pyelogram, circumcision, laparoscopic left orchiectomy for an atrophic, abnormal left testis, and laparoscopic-assisted left lower pole dismembered pyeloplasty. The retrograde pyelogram was consistent with a partially duplicated left ureter (Fig. 2). Once the circumcision and laparoscopic left orchiectomy were completed, the left lower pole proximal ureter was identified laparoscopically. A small flank incision was then created, and the proximal ureteral segments and lower pole renal pelvis were brought up through the incision, isolated, and tagged. The atretic segment of the lower pole ureteropelvic junction was spatulated down into the common ureter and then anastomosed to the lower pole renal pelvis (Fig. 3). A Salle stent was placed, and subsequently removed at 10 days post-operatively.

An ultrasound 1 month later showed resolution of the patient’s upper pole hydronephrosis and modest improvement in his lower pole hydronephrosis. An ultrasound at 3 months post-operatively continued

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Lower Pole (continued from previous page)

to show complete resolution of the upper pole hydronephrosis, decreased lower pole hydronephrosis to SFU Gr 2, and interval growth of the left renal moiety.

Discussion

Though a rare entity, ureteropelvic junction obstruction in duplicated collecting systems is most commonly seen affecting the lower pole system with an upper pole reflecting a single infundibular segment (1). Presentation and evaluation are similar to those of patients found to have UPJO in single collecting systems. Options for surgical management, when indicated, include those seen in single systems, such as dismembered pyeloplasty or endoscopic pyelolithotomy of the affected segment. Alternatively, pyeloplasty with pyelopyelostomy to the renal pelvis or pyeloureterostomy to the proximal ureter of the unaffected segment or common ureter, as was done in our case, may be considered. (2)

References


Figure 3
Urinoma Misdiagnosed as Simple Renal Cyst in Prenatal Ultrasound Leading to Delayed Diagnosis of Posterior Urethral Valves

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Case Presentation

Posterior urethral valves are reportedly associated with urinoma in up to 8.5% of cases.¹ Herein we describe an atypical presentation of posterior urethral valves (PUV) with bilateral SFU grade I hydronephrosis and a “simple renal cyst” diagnosed in the third trimester prenatal ultrasound later found to be a sub capsular urinoma, resulting in a delayed diagnosis.

A 35-week old, 3140 g male neonate was born via spontaneous delivery in a community maternity unit. The pregnancy was unremarkable with the exception of the third trimester prenatal ultrasound (US) which revealed mild (SFU I/UTD 1) bilateral hydronephrosis (HN) and left sided simple renal cyst with a normal bladder. Recommendations were made for a referral to an outside pediatric urologist with 3-month follow-up US. US on the 1st day of life at a community hospital confirmed mild HN, left upper pole simple renal cyst, no ureteral dilation and a normal bladder. On day two of life, the neonate developed a febrile urinary tract infection (UTI) confirmed by catheter specimen and subsequently on day 8, care was transferred to the regional children’s hospital for PICC line insertion due to lack of access for intravenous antibiotics. Pediatric Urology was consulted and a repeat US was ordered which revealed left upper pole fluid collection suggestive of peri-renal urinoma, bilateral mild (SFU I/UTD 1) hydronephrosis and thickened bladder wall (Figure 1). Subsequent VCUG was ordered, which reviewed bilateral grade V VUR with tortuous ureters, hypertrophy of bladder neck, dilated posterior urethra and visible valves leaflets (Figure 2), confirming the diagnosis of PUV. Preoperative creatinine was 110 and endoscopic valve ablation with circumcision were performed without complications the following day. Post operative creatinine dropped to 67 at day 3 post-surgery, with complete resolution of the urinoma, as seen on Figure 3. The Patient had an uneventful recovery and was discharged home on post op day 14, on continuous antibiotic prophylaxis (trimethoprim) for bilateral grade V VUR.

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Uronoma Misdiagnosed (continued from previous page)

Discussion

As it sometimes happens in severe PUV cases, high bladder pressures may cause severe VUR with a pop-off mechanism that may eventually lead to rupture of the fornix and development of a sub capsular urinoma, with or without urinary ascites, phenomenon which is pathognomonic of PUV. This pop-off mechanism probably explains why the expected bladder wall trabeculations were not seen on VCUG and the right kidney was spared, showing only mild HN. In atypical presentations such as this, male neonates with an abnormal US and a confirmed UTI in the first few days of life should have a VCUG before hospital discharge to avoid missing the diagnosis of PUV, preventing the morbidity associated with delaying its diagnosis.

References


Figure 2

![Figure 2 Image]

Figure 3

![Figure 3 Image]
Bilateral Abdominoscrotal Hydroceles with Concurrent Lower Extremity Edema: Antenatal Presentation and Postnatal Management

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Case Presentation

The abdominoscrotal hydrocele is a relatively rare condition, defined as a palpable, fluid-filled mass with inguinoscrotal and abdominal components. Exceedingly more uncommon is the presence of concurrent lower extremity edema due to impaired venous return from extrinsic compression. Herein, we present a boy with large, bilateral abdominoscrotal hydroceles with concurrent lower extremity edema that were initially detected antenatally.

A three-month-old boy presented for evaluation of bilateral hydroceles, which were initially identified on antenatal ultrasound [Figure 1]. On examination, the infant was found to have large hydroceles (L>R) extending superiorly into the inguinal regions bilaterally, along with left lower extremity edema and petechiae due to impaired venous return [Figure 2]. Ultrasound demonstrated bilateral hydroceles with normal appearing testes. Due to the marked lower extremity edema, the patient was recommended to undergo surgery. The case began with diagnostic laparoscopy, confirming the collections were partially intra-abdominal and without evidence of a patent processus vaginalis on either side. The left-sided hydrocele was noted to be much larger both intra-abdominally and scrotally. Both hydroceles were drained, excised, and plicated through a combination of scrotal and inguinal incisions.

The patient tolerated the procedure well and was discharged home the day of surgery. There were no intra- or postoperative complications. Follow-up ultrasound at 3 months post-operatively revealed complete resolution of the hydroceles, with marked improvement of the left lower extremity edema [Figure 3].

Discussion

The prevalence of abdominoscrotal hydroceles is estimated to be approximately 3% of all hydroceles in children1. While simple hydroceles are commonly encountered and their surgical management is well described, the abdominoscrotal hydrocele presents a surgical challenge due to their large size and the difficulty in isolating delicate structures that are closely associated anatomically, including the vas deferens and splayed spermatic vessels. Our case demonstrates that both inguinal and scrotal approaches are safe and effective options.

A recent review article by Doudt, et al. highlighted the management of these cases and noted that the majority of the cases were approached inguinally with minor complication rates overall, regardless of the approach2. They noted that other approaches (scrotal and laparoscopic) were also observed to be effective.

Unique to our case is the initial presentation with antenatal detection, along with the presence of concurrent lower extremity edema due to impaired venous return. The left-sided hydrocele was noted to be much larger both intra-abdominally and scrotally. Both hydroceles were drained, excised, and plicated through a combination of scrotal and inguinal incisions.

The patient tolerated the procedure well and was discharged home the day of surgery. There were no intra- or postoperative complications. Follow-up ultrasound at 3 months post-operatively revealed complete resolution of the hydroceles, with marked improvement of the left lower extremity edema [Figure 3].

Discussion
An Interesting Presentation of Synchronous Cystic Pulmonary and Renal Masses

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**Case Presentation**

MH is a 7 month old male with a prenatal history of a left lung mass and multi-cystic right kidney. His left kidney was normal.

MH was born at 39 weeks gestation, through Cesarean section, which was performed due to a failure to progress after induction of labor. Mother’s prenatal history was unremarkable. The Apgar scores were 6, 7, and 9 at 1, 5, and 10 minutes, respectively. He was admitted to the NICU, and after a few days of supportive care, discharged home. On initial physical exam, MH’s right kidney was palpable. A renal ultrasound shortly after birth reaffirmed the multi-cystic right kidney, which resembled a multi-cystic dysplastic kidney (MCDK), but had an unusual amount of solid, normal appearing parenchyma, not consistent with diagnosis of MCDK. The initial chest radiograph demonstrated a left lung opacity and chest radiograph two months later demonstrated a cystic left lung lesion, favored to be a cystic pulmonary airway malformation (CPAM). The patient was asymptomatic.

At two months, a nuclear medicine renal scan demonstrated diminished, but significant function (31%) in the right kidney, without obstruction. This finding refuted the original diagnosis of MCDK, and made unilateral cystic disease or multilocular cystic nephroma more likely.

CT scan at 3 months of age demonstrated a 3.5 cm cystic left upper lobe lung mass. Multiple large cysts in the right kidney displayed mass effect, with normal enhancement of residual renal parenchyma, again not typical for MCDK. Because of the synchronous cystic pulmonary and renal masses, concern for a DICER 1 mutation was raised.

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Discussion

A palpable renal mass in the neonate is most often due to a congenital mesoblastic nephroma. However, this mass appeared on ultrasound to be most consistent with a variant of MCDK. Due to appreciable right renal function, other cystic masses were included in the differential, such as multilocular cystic nephroma or congenital mesoblastic nephroma. Concurrent cystic lesions in the lung and kidney should lead one to consider DICER1 mutation. The rare hereditary DICER1 mutation carries increased risk for patients to develop PPB, cystic nephromas of the kidney, ovarian Sertoli-Leydig tumors, bladder and cervical embryonal rhabdomyosarcomas, and other potential tumors [1, 2], all requiring long term follow up.

References


Synchronous Cystic (continued from previous page)

with the concern the lung mass could be a pleuropulmonary blastoma (PPB), which cannot be differentiated on imaging from CPAM. The cystic renal lesion could represent a multilocular cystic nephroma.

At three months of age, a thoracotomy and left upper lobectomy was performed without complication. Pathology demonstrated a CPAM without evidence of PPB and confirmatory testing was requested. At six months of age, a right nephrectomy was performed without complication. Pathology confirmed unilateral multifocal cystic dysplasia, but no evidence of tumor. Upon follow up, a 1 cm firm subcutaneous, non-mobile right scalp lesion was detected, concerning for a metastatic PPB. MRI of the lesion favored an epidermoid or dermoid cyst. This was excised by neurosurgery and pathology confirmed a dermoid cyst. Interestingly genetic testing revealed no evidence of the DICER1 mutation.

Figure 2

Figure 3
A Case of Migrated Vesicoamniotic Shunt Requiring Urgent Repair of Abdominal Wall Hernia

Courtney K. Rowe, MD, Paul A. Merguerian, MD, Kathleen Kieran, MD, Department of Urology, Seattle Children’s Hospital, Seattle, Washington

Case Presentation

35 y/o G2P1 female with singleton male pregnancy was noted to have echogenic kidneys and keyhole shaped bladder concern for bladder outlet obstruction on 14 week prenatal ultrasound. A vesicoamniotic shunt was placed at 18 weeks. Amniotic fluid levels were initially high, then decreased acutely in the third trimester at which point the shunt was no longer visualized in the abdomen (Figure 1). Since fluid levels were low-normal, the pregnancy was followed with weekly ultrasounds as the risk of early delivery was felt to be high.

The baby was delivered at 35 6/7 weeks due to decreasing amniotic fluid levels. Exam noted a 2cm left upper quadrant abdominal wall defect with herniated omentum; the distal coil of the shunt was unable to be visualized; however, abdominal X-ray showed two radio-opaque markers in the vicinity of the bladder (Figure 2). He had respiratory distress after birth and was found to have a small right pneumothorax and a tension left pneumothorax which was treated with a chest tube.

The baby was taken to the operating room 10 hours after birth; the herniated omentum was resected and the abdominal wall defect closed. Cystoscopy revealed Type III annular valves and the shunt migrated completely into the bladder. The shunt was removed intact endoscopically, which ablated the posterior urethral valves (Figure 3). The hernia tract appeared to be chronic and was successfully repaired via a small extension of the incision.

Voiding cystourethrogram on post-operative day 8 showed successful valve ablation with no extravasation from the prior shunt exit site. The baby’s creatinine was initially 0.8 mg/dL but increased to 6.1 mg/dL by 4 weeks of life, after which he was transitioned to peritoneal dialysis. He has had laparoscopic treatment of pyloric stenosis and a G-tube placed for poor PO intake. At this time, there are no residual pulmonary issues. He has hypertension and hypothyroidism well controlled with medication and no longer requires his G-tube. He will be considered for renal transplantation once his size and weight are appropriate.

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Migrated Vesicoamniotic Shunt (continued from previous page)

Discussion
We describe the rare complication of an abdominal wall hernia after fetal vesicoamniotic shunting. This patient had similar risk factors to prior case reports, including shunt placement above the umbilicus and increased intra-abdominal pressure due to the migrated shunt and resulting distended bladder.1,2

This case is notable for a few details in clinical management. The first is the successful pulmonary outcome despite shunt migration, which supports the decision to delay delivery as long as amniotic fluid levels remain normal or low-normal and the pregnancy is closely followed. Second, a large laparotomy incision is not necessary in the event of a migrated shunt with associated hernia as long as the shunt can be removed intact cystoscopically and the hernia repaired appropriately. This avoids a large peritoneotomy that would delay initiation of peritoneal dialysis. Finally, similar to prior reports, there was no evidence of any renal benefit from placement of the shunt in our patient, who will be dialysis- or renal transplant-dependent for life.3

References

Prenatal Diagnosis of a Congenital Mesoblastic Nephroma
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Introduction
Congenital mesoblastic nephroma (CMN) is the most common renal tumor in infancy, comprising 3% of all pediatric renal tumors.1 Although CMN typically behaves in a benign manner, it has rarely been reported to recur or metastasize.1 There are three histological subtypes of CMN, classic, cellular and mixed. We present a case of prenatally diagnosed CMN in a premature infant.

Case Presentation
A 61-day old female was referred to pediatric urology after a renal mass was detected on routine prenatal ultrasound. Medical history was significant for hypertension and prematurity (30 weeks). Surgical and family history were negative. Physical exam revealed a large, firm, non-tender right upper quadrant mass. Laboratory evaluation was significant for hemoglobin of 8.9 g/dL and creatinine of 0.21 mg/dL. The remainder of laboratory evaluation was normal. Computed tomography (CT) of the abdomen and pelvis revealed a heterogeneous 4.7 cm right renal mass (Fig. 1). Metastatic workup with chest CT was negative. After discussion at multi-disciplinary tumor board, the patient was enrolled in the COG tissue banking study, and taken to the operating room for a right nephrectomy and retroperitoneal lymph node dissection. Intraoperative frozen section revealed CMN. Operative time was 170 minutes. The hospital course was uneventful. Pathology (Fig. 2A, 2B) revealed a 5 cm classic CMN with tumor present on the renal vein adventitia margin but no luminal invasion (stage 3). Ten retroperitoneal lymph nodes were uninvolved. No adjuvant therapy is planned.

Discussion
CMN typically presents as an abdominal mass in a child less than 3 months of age. Approximately 15% of cases are found on prenatal ultrasound, with the majority of prenatally identified renal tumors being CMN.1,2 Patients occasionally present with vomiting, hematuria, anemia, hypercalcemia, or hypertension.2 These paraneoplastic syndromes can be due to parathyroid-like hormone, renin and prostaglandin secretion, and usually resolve after tumor resection.3 One-third of CMN cases display a classic histology pattern.2 Classic CMN is leiomyomatous with low mitotic activity, interlacing bundles of spindle cells, and infiltrative growth into the renal parenchyma and perinephric fat.1,3 On imaging, classic CMN characteristically is observed as a uniform soft tissue mass, with peripheral enhancement.3 Cellular and mixed histologies also exist and are associated with worse prognosis.

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Congenital Mesoblastic Nephroma (continued from previous page)

CMN is treated with radical nephrectomy, has an excellent prognosis, and overall survival is 95%.

Adjuvant chemotherapy is rarely required. Although initially described as benign, distant metastasis to lung, brain, heart, liver, or bone have been observed. Recurrence, which is more common with the cellular subtype and stage 3 tumors, tends to occur within 12 months, demonstrating the need for close follow-up for one year, although universal guidelines have not been established.

Conclusion

CMN is the most common pediatric renal tumor reported in infancy, typically considered benign, with an excellent prognosis. The treatment for CMN is nephrectomy, with close follow-up for the first year. Although general treatment and follow up is agreed upon, there is a need for consensus guidelines as cases that are stage 3 or of a cellular subtype can create difficulty in decision making about adjuvant therapy.

References

Figure 2. Pathologic specimen (A) photograph of the cut surface of the gross specimen (B) photograph of hematoxylin-eosin stain showing spindle shaped cells arranged in fascicles with low mitotic activity.
Case Presentation

A 2.5 year old presented to general surgery with yellow drainage from a cystic suprapubic lesion. Born at 36 weeks, he was otherwise healthy with normal prenatal ultrasounds. Physical examination was normal, except for a suprapubic epidermal inclusion cyst without drainage. At excision, the surgeon made an ellipse around it, but found with further dissection, it was part of a sinus tract. He dissected down towards the pubic bone, at which point he “tied it off like a sinus.” Pathology diagnosed the structure as a “benign cyst, most consistent with urachal cyst/remnant”. The general surgeon obtained a voiding cystourethrogram, which was normal, including no evidence of a patent urachus. An ultrasound, however, demonstrated a midline tubular structure anterior to the bladder (Figure 1).

The boy was lost to follow up, until he presented 15 months later to pediatric urology with “milky urine.” History and physical were notable for infrequent voiding and constipation. On review of his ultrasound images and pathology, there was concern for possible communication between the structure and the bladder, potentially as a source of debris. At surgery, cystourethroscopy was normal. The structure was isolated through a Pfannenstiel incision. It traveled as the urachus from the dome of the bladder into the pelvis, passing anterior to the bladder and pubis (Figure 2). The prior scar was excised, and the structure was removed in its entirety. Pathology returned urachal remnant (Figure 3). The patient did well after surgery, however, his milky urine persisted intermittently.

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Aberrant Urachal Remnant (continued from previous page)

Discussion

While aberrant urachal remnants have been described, case reports are exceedingly rare. Our patient’s area of drainage is more common of a congenital prepubic sinus (CPS). CPSes typically exit somewhere above the penis or pubic area. Their origin is more variable. In a 2001 study of 20 patients, most originated near the bladder, with only one opening into the bladder at the dome. This was considered a urachal remnant on pathology.1 A recent review of all reported cases found connections to the bladder in less than 5% of patients.2 Their course to the bladder also varies, with 61.5% of the tracts traveling over the pubic bone, 19.2% through it, and 19.2% under it.2

Numerous theories abound as to the embryologic origin of CPSes. The most common proclaims them a dorsal urethral duplication variant, with some immunohistochemical supporting evidence.1,3 Others hypothesize they are a minor form of a midline abdominal wall closure defect, primitive urogenital sinus fistula, or finally, remnant of the cloaca, where the umbilicophallic groove entraps a portion of cloacal membrane, allowing tubularization and tract formation.3 Whatever the cause, surgery is recommended, both to prevent recurrent symptoms and the possibility of later malignancy.2

We presented a urachal remnant with an unusual course from the dome of the bladder to the suprapubic region. These uncommon remnants may represent a variant of CPS, a term seeming to encompass many entities. With both urachal remnants and CPSes, surgical excision is curative.

References


Figure 3: Pathology slide demonstrating a lumen lined with urothelium (U), which in turn is surrounded by smooth muscle. There were areas with intestinal-type epithelium, including goblet cells (*), as well as inflammatory infiltrates.