Disorders of the Bladder and Cloacal Anomaly

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KEYWORDS

• Urachal remnant • Patent urachus • Bladder exstrophy • Cloacal exstrophy
• Persistent cloaca

KEY POINTS

- Urachal anomalies are uncommon, rarely diagnosed prenatally, and most often suspected during the neonatal period when continuous or intermittent umbilical drainage is observed; they often are managed conservatively in children less than 6 months of age.
- Classic bladder exstrophy is a complex congenital anomaly with musculoskeletal abnormalities of the lower abdominal wall and pelvis and genitourinary abnormalities of the bladder, urethra, and external genitalia.
- Cloacal exstrophy is among the most severe congenital anomalies compatible with live birth, with features of bladder exstrophy present in combination with exstrophic bowel segments, imperforate anus, and, commonly, omphalocele and spina bifida.
- Persistent cloaca is a form of anorectal malformation that results in a confluence of rectum, vagina, and urethra into a common channel that extends to the perineum as a single opening.

INTRODUCTION

Interpretation of antenatal imaging consistent with urachal, bladder, and cloaca anomalies and optimal management of these problems are guided by an understanding of their embryologic origins.[1,2] The cloaca is an endoderm-lined primordial organ that is first apparent at the beginning of the second week of gestation. This structure represents a confluence of the primitive hindgut (dorsally) and the allantois (ventrally). Between the fourth and sixth weeks of gestation, the urorectal septum divides the endodermal cloaca into a ventral urogenital sinus and a dorsal rectum.[3,4] The cranial portion of the urogenital sinus is continuous with the allantois and develops into the bladder and pelvic urethra. During the second trimester, the allantoic duct and ventral cloaca involute as the bladder descends into the pelvis. This epithelialized

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fibromuscular tube obliterates into a thick fibrous cord, the urachus, which becomes the median umbilical ligament connecting the apex of the bladder with the umbilicus. Failure of the lumen of the allantois to completely obliterate results in a urachal remnant.[5] Exstrophy is thought to be secondary to the failure of the cloacal membrane to be reinforced by ingrowth of mesoderm. The cloacal membrane is subject to premature rupture, and, depending on the extent of the infraumbilical defect and the stage of development during which the rupture occurs, bladder exstrophy, cloacal exstrophy, or epispadias results.[6] The inciting event for persistent cloaca remains unclear but is thought to be secondary to the arrest of cloacal division by mesenchymal growth.[4]

URACHAL ANOMALIES

The urachus is an embryonic remnant derived from involution of the allantois; it is located in the preperitoneal space and connects the dome of the bladder to the anterior abdominal wall at the level of the umbilicus. The urachus is accompanied by umbilical arteries on either side, which undergo fibrosis and become the medial umbilical ligaments. Failure of the lumen of the allantois to completely obliterate results in a urachal remnant. Congenital urachal anomalies include patent urachus, urachal cyst, umbilical-urachus sinus, and vesicourachal diverticulum.[5,7] Patent urachus and urachal cyst account for most urachal anomalies, with patent urachus diagnosed most commonly in the neonatal period.[5,7–9] Although most often diagnosed postnatally, patent urachus can be detected on prenatal ultrasound or fetal magnetic resonance imaging (MRI). It is visualized as a cystic mass located at the base of the umbilical cord that communicates with the bladder.[10,11] Urachal cysts are more likely to present with infection or abdominal pain in later childhood or adulthood.[9,12] Patent urachus results from failure of the epithelial-lined urachal canal to obliterate, whereas urachal cysts have no communication with the bladder or umbilicus, although they can intermittently drain into either.[5] Given its location, a urachal remnant may be confused with other umbilical complications (ie, omphalitis, granulation tissue of the healing umbilical stump, or an infected umbilical vessel).[5]

Patient History/Physical Examination

- Patent urachus is suspected with umbilical drainage and abnormal-appearing umbilicus in the neonatal period; additional presentations include edematous umbilicus, granuloma, and delayed healing of the cord stump[13–15]
- Most common organisms cultured from umbilical drainage include *Staphylococcus aureus*, *Escherichia coli*, *Enterococcus*, *Citrobacter*, *Klebsiella,* and *Proteus*[8,9]
- Patent urachus must be distinguished from omphalitis, which typically presents as a superficial cellulitis and a patent omphalomesenteric duct remnant, which can present with feculent umbilical drainage[16,17]
- Associated anomalies causing in utero bladder outlet obstruction (ie, posterior urethral valves, prune belly syndrome, and urethral atresia) must be excluded; up to 14% of patients with patent urachus have postnatal confirmation of lower urinary tract obstruction[5]

Imaging/Additional Testing

- Diagnosis is confirmed by demonstration of a fluid-filled cystic structure on ultrasound scan extending from the dome of bladder to the umbilicus
- Passage of contrast at the umbilicus is seen on voiding cystourethrogram (**Fig. 1**); this is not always demonstrable