Bladder Agenesis and Associated Pelvic Arterial Anomaly in 2 Female Pediatric Patients

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Bladder agenesis is an extremely rare congenital anomaly of the genitourinary tract. Two female patients with known diagnoses of bladder agenesis presented for prerenal transplant evaluation and neobladder creation. Similar unique pelvic arterial malformations were identified through preoperative imaging and intraoperative examination. With these similar findings, it could be proposed that such anatomical variants are products of the same insult or involve a causal relationship, with vascular aberrancies potentially provoking pelvic organ maldevelopment. We advocate detailed mapping of the pelvic arterial tree preoperatively in bladder agenesis cases to prevent arterial injury and to safely delineate anatomy for appropriate transplant placement.

CASE 1
An 8-year-old female with a history of repaired Tetralogy of Fallot with severe pulmonary stenosis presented for prerenal transplant evaluation. Urologic history included bladder agenesis with a multicystic dysplastic left kidney and ectopic right pelvic kidney. She had a normal anorectum with total urinary incontinence. At presentation, the patient was in the second percentile for weight (19.8 kg), with a creatinine clearance of 9.20 mL/min/1.73 m², blood urea nitrogen 52 mg/dL and serum creatinine 3.5 mg/dL. Pelvic magnetic resonance imaging (Fig. 1) identified a partially fluid-filled vagina, free fluid in the area where the bladder should have been located, and a 4-mm diameter aberrant artery originating from the right internal iliac artery, extending anteriorly to cross the midline, traversing this fluid-filled region, and terminating at the left external iliac artery. Absence of the left common iliac artery was also identified. The patient subsequently underwent a left laparoscopic nephroureterectomy with the creation of Charleston neobladder. The anomalous artery was identified intraoperatively and not injured. Living donor renal transplant was successfully performed 3 months later, with the renal artery anastomosed to the infrarenal aorta and the vein anastomosed to the inferior vena cava without complications. She has a functioning renal transplant and continent neobladder 6 years postoperatively.

CASE 2
A 6-year-old female with a history of cloacal variant with ambiguous genitalia, imperforate anus, bladder and vaginal agenesis, and bilateral renal dysplasia was referred for creation of a neobladder in support of a renal transplant. She had been maintained on peritoneal dialysis since shortly after birth. She was taken to the operating room for Studer ileal neobladder creation, bilateral open nephroureterectomies, Malone antegrade continence enema, and Monti ileovesicostomy. Early in the operative course, the urachus was taken down, revealing a large 1-cm diameter pulsatile artery traversing where the bladder should have been. This aberrant major artery, located in essentially the same location as in Case 1, had not been identified on previous preoperative imaging studies as it was partially obscured by the dysplastic pelvic kidney (Fig. 2). Extreme attention was required during the operative procedure to protect this large variant artery, as the aortic bifurcation was anterior, in the expected field of the bladder (Fig. 3). The patient is being made renal transplant-ready 21 months after neobladder creation and remains on peritoneal dialysis.

DISCUSSION

Complete bladder agenesis is an extremely rare congenital anomaly of the genitourinary tract that is usually not
compatible with life. The reported incidence rate is approximately 1:600,000, with preponderance for the female gender (female:male ratio 30:1).\textsuperscript{1,2} Ninety percent of viable children born with this malformation are female, as ectopic drainage of urine from the ureters into various Müllnerian structures such as the uterus, vaginal wall, or vestibule conserves renal function and urine excretion for intrauterine lung development.\textsuperscript{3} Male patients only maintain urinary drainage through either the rectum or patent urachus.\textsuperscript{4} There are at least 60 bladder agenesis cases reported worldwide in the medical literature, with most recent data showing only 24 living patients. Our current report now adds 2 additional cases to this rare population.\textsuperscript{5,6}

The embryological maldevelopment resulting in this condition is complex and presumed to occur during weeks 5 through 7 of human embryogenesis when the cloaca is divided into the urogenital sinus and anorectal canal. Palmer and Russi proposed that after this division, the mesonephric ducts and ureter fail to incorporate in the bladder wall, leading to subsequent atrophy of the urogenital sinus, as the bladder is unable to acquire urine and distend. The posterior division involving the anorectal canal normally develops without complications in most cases, but hindgut abnormalities have been noted.\textsuperscript{7} Furthermore, there is a high incidence of additional anomalies associated with this diagnosis, which often involve other urogenital organs, skeletal system, and nervous system. As with the condition itself, such associations are often not compatible with life and lead to further morbidity and mortality. Findings in the literature include, but are not

Figure 1. Axial (A) and sagittal (B) T2 magnetic resonance images of Case 1 demonstrate large aberrant artery positioned anteriorly in pelvis. Coronal stacked maximum intensity projection images of Case 1 demonstrate absence of left common iliac artery (C).

Figure 2. T2-weighted turbo spin echo image of Case 2 with aorta displaced to right with central dysplastic left kidney. Aortic bifurcation is noted at inferior margin of left kidney.
limited to, renal agenesis, renal ectopy, multicystic dysplastic kidneys, genital ambiguity, duplicate or bicornuate uterus, scoliosis, spina bifida, and anencephaly. Both of our reported patients displayed a variation of these at presentation, leading to further complications and additional considerations in the treatment courses.8 Of particular interest with our 2 cases are the similar major arterial branches found incidentally in both patients. Others have noted major pelvic arterial anomalies in cases of bladder agenesis.9 Dykes et al has proposed that abnormal aorta and iliac artery findings in children with urogenital abnormalities (ie, cloacal malformations, bladder agenesis, urogenital sinus, and neuropathic bladder) stem from the abnormal persistence of ventral roots of the umbilical arteries during embryogenesis at weeks 3 and 4 of human development.9-11 In normal development, these umbilical arteries regress into the internal iliac artery and if this regression is interrupted, variations in anatomy can emerge.12 Although no definitive link between these 2 systems has been fully established in the literature, one could propose that such anatomical variants are products of the same insult or involve a causal relationship, with vascular aberrancies potentially provoking pelvic organ maldevelopment. It remains unknown whether the bladder agenesis or the arterial anomaly is the causal primary embryological defect.

Urinary reconstruction for bladder agenesis has been accomplished by a variety of continent or incontinent urinary diversions, with adjunctive procedures as needed (ie, transplant). We would advocate detailed mapping of the pelvic arterial tree preoperatively in bladder agenesis cases for 2 major reasons. First, accidental arterial injury and resultant hemorrhage could result in loss of blood supply to lower limbs and organs. Second, delineation of these arterial malformations are necessary for transplant planning, as the additional arterial load of a renal transplant could put the patient at risk for leg claudication if for example 1 iliac artery serves as the sole arterial supply for both legs and the transplant. Locating the transplant off the distal aorta rather than the functioning iliac artery might be the safer option, as performed in Case 1.

References