Agenesis of the bladder with solitary renal dysplasia: management of a challenging condition

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NAZIF O, MACNEILY AE. Agenesis of the bladder with solitary renal dysplasia: management of a challenging condition. The Canadian Journal of Urology. 2004;11(2):2220-2222.

Bladder agenesis is an extremely rare congenital anomaly. We report a case of bladder agenesis in a newborn girl with a prolapsing ectopic ureter and solitary dysplastic kidney. The classification of this disorder and the management of this patient are reviewed.

Key Words: bladder agenesis, oligohydramnios, ectopic ureter, neobladder, Mitrofanoff

Introduction

Bladder agenesis is an extremely rare congenital anomaly.[1] Including this patient, there are 49[2-4] recorded cases in the English literature. Most cases of bladder agenesis are associated with severe anomalies and in-utero death, with only 17 live birth cases reported, all except one of which are female.[2-6] There is little information in the literature describing the embryology, etiology, and management of this condition.We present a case of a newborn female with bladder agenesis and her management.

Accepted for publication February 2004

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Patient presentation

At the 18 week prenatal ultrasound, a female fetus was diagnosed with oligohydramnios and a solitary dysplastic left kidney. The fetus received an amniotic infusion and was subsequently delivered at 41 weeks gestational age without complication by a healthy 33-year-old primigravid mother. Birth weight was 3820 gm and APGAR scores were 7 at 1 minute and 8 at 5 minutes. There were no complications of oligohydramnios including pulmonic, orthopedic, facial, or neurologic abnormalities. Physical examination of the introitus revealed a small

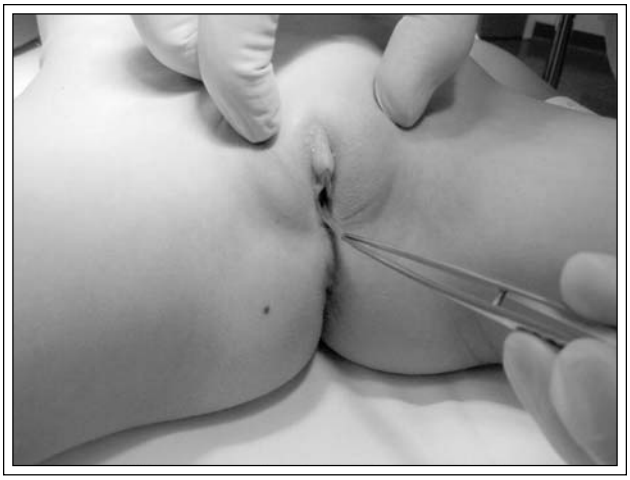

Figure 1. Perineum at 3 years and 7 months old. Forceps identify the ectopic ureter at the introitus and external to the vagina.

interlabial mass, presumably the ureter Figure 1. Upon application of pressure to the abdomen, urine was expressed from this structure.

Two ultrasounds done by the age of 6 days old confirmed the presence of an echogenic dysplastic kidney in the left renal fossa with multiple cortical cysts.

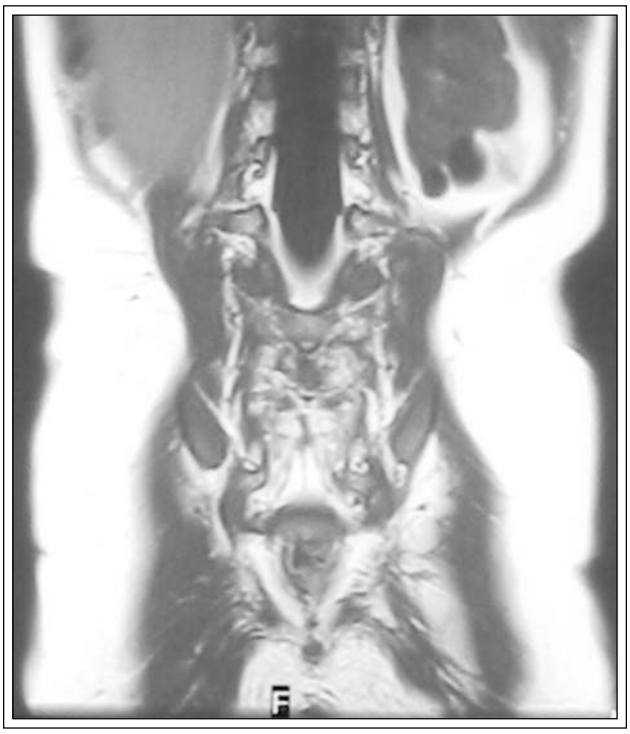

Figure 2. Magnetic Resonance image of retroperitoneum. Note dysplastic kidney in left renal fossa. No kidney is seen in right renal fossa or ectopically.

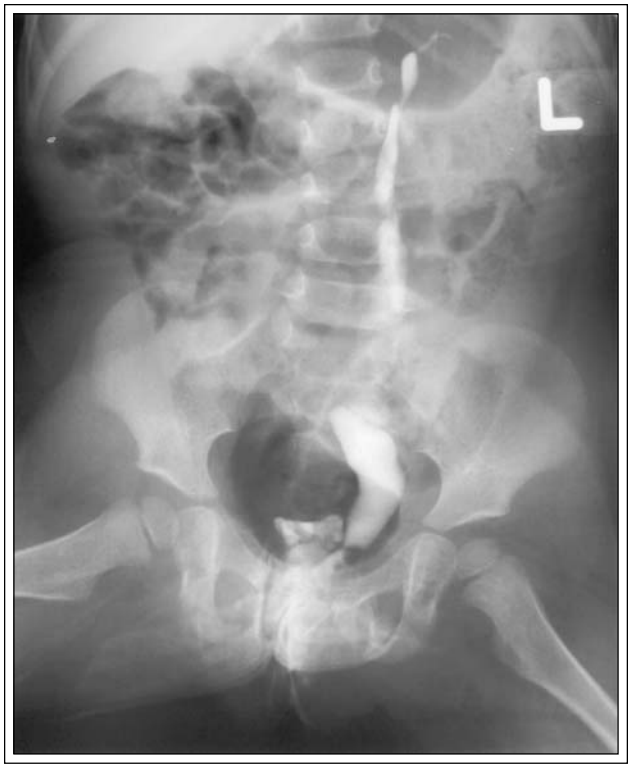

Figure 3. Retrograde pyelogram at age 2 years old. A ureteral catheter is identified within the tortuous and dilated left ectopic ureter. The left kidney collecting system is not well opacified. Contrast material delineates the vagina. No bladder is seen.

The right kidney was not identified. A small amount of fluid was seen in a pelvic structure presumed to be the vagina and no bladder was identified.

At 9 days old, creatinine was 228 umol/L (normal 10 – 90), urea 10.5 mmol/L (normal 1.8 – 8.2), and potassium 5.6 mmol/L (normal 3.5 – 5.5). Daily prophylactic amoxicillin was instituted to protect against urosepsis.

Magnetic resonance imaging revealed a solitary dysplastic left kidney with multiple cysts Figure 2. A voiding cystourethrogram was attempted, but failed because there was no identifiable urethral opening.

At 2 years of age, cystoscopy and retrograde pyelography were undertaken. A retrograde pyelogram was performed via the prolapsing ureter exiting the perineum. No urethral orifice, cervix or bladder was evident Figure 3.

Discussion

Embryology

The embryology of bladder agenesis is largely unknown. Krull[7] suggested that bladder agenesis may

be related to secondary atrophy of the anterior division of the cloaca. Malposition of mesonephric ducts and ureters results in a lack of distention and accumulation of urine in the bladder. With ectopic ureters, renal function may be preserved.

The defect associated with bladder agenesis is severe and is often associated with multiple anomalies including urologic, neurologic, and orthopedic deficiencies.

Classification

Metoki et al[1] developed a classification system for bladder agenesis divided into five types dependant on the location where the ureter(s) implant.

1. remnant cloacal
2. rectal
3. urogenital sinus
4. vaginal
5. ectopic

Our case is classified as the vaginal type.

Management

There is little described in the literature regarding management of this condition. In surviving cases, urinary diversion is recommended by ureterosigmoidostomy, an external stoma, or neobladder formation.[1,4,6]

Our case was complicated by presentation with a solitary dysplastic kidney. Her renal function was stable initially after birth, but worsened after urinary tract infections. The child was constantly wet with continuous dribbling. By two hand half years of age, she was failing to thrive and a gastrostomy tube was placed to increase caloric intake. Somatotropin growth hormone resulted in an improvement in growth. In light of worsening renal function and her failure to thrive, it became clear that she would require renal transplantation.

Prior to renal transplantation, urological reconstructive options were discussed with the family including ileal conduit, intestinal neobladder, or temporary diversion via a cutaneous ureterostomy. We felt that a neobladder with a catheterizable stoma would be the best solution for long term protection of a graft and optimal socialization.

At age 3 years and 7 months old, the patient underwent pretransplant urinary tract reconstruction. The following operations were performed on the patient: cystoscopy, laparotomy, ileo-cecal neobladder formation, and catheterizable Mitrofanoff stoma. At the time of cystoscopy, she was identified to have a shallow blind ending vagina with termination of the ureter at the vestibule. Again, there was no evidence of urethra, bladder, or cervix. Laparotomy revealed blind-ending fallopian tubes, absent uterus, and two cystic ovaries. A hemodialysis catheter was placed at the time of surgery in anticipation of renal decompensation in the perioperative period. The patient responded favorably to the surgery and did not require perioperative dialysis.

The patient was transplanted pre-emptively with a living related renal graft from her father at age 4 years and 4 months old. The graft was placed extraperitonealy on the recipient's right and anastomosed to the great vessels, posterior to the neobladder. The post operative course was uneventful, and discharge creatinine was 26 umol/L (normal 10 - 60).

Since continent diversion, the parents have been catheterizing the stoma every 4 hours, and the patient is dry. Nocturnally, she sleeps for 8 hours while remaining dry.

At 5 years old, she developed a closed loop small bowel volvulus resulting from adhesions. This necessitated resection of 50 cm of small bowel. Aside from this complication, she continues to thrive since transplantation. □

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