Pediatr Surg Int (1993) 8: 60–61

Pediatric Surgery International

Case report

Agenesis of the bladder: a rare clinical entity in a male child

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Accepted 18 November 1991

Abstract. Agenesis of the bladder, one of the rarest anomalies of the urinary bladder, was for the first time managed successfully by primary ileocecal functional bladder reconstruction at the Children's Hospital, Institute of Medical Sciences, Banaras Hindu University. The salient features along with a review of literature are presented.

Key words: Bladder agenesis – Primary reconstruction of bladder – Ileo-cecal bladder – Children

Introduction

Complete absence of the urinary bladder was found in most cases at postmortem examination in 7 out of 19,046 autopsies of anomalous genitourinary systems [1]. The first case report of bladder agenesis was published in 1654 [3]; since then 37 further cases have been reported [2].

Case report

A 2½-year-old male was admitted with a history of dribbling of urine since birth, intermittent high-grade fever, and failure to thrive for 1 year. At the time of hospitalization he was irritable, running a high fever (102–104°F), moderately dehydrated, pale, and malnourished. There was severe ammonia dermatitis over the perineum. On abdominal examination the kidneys and bladder were not palpable, while on rectal examination a distended posterior urethra was felt without any appreciable bladder. Except for coronal hypospadias, no other congenital anomalies could be detected.

Laboratory studies showed the hemoglobin to be 7.5 g%, blood urea 53.55 mmol/l, serum creatinine 0.25 mmol/l, and serum electrolytes within the normal range. Routine microscopy of the urine revealed numerous pus cells and culture showed *Escherichia coli* sensitive to amikacin sulphate. Ultrasonography revealed bilateral hydronephrosis but the urinary bladder could not be visualized. A micturating cystourethrogram revealed a small, dilated proximal urethra, bilateral gross reflux with the left kidney in the pelvis, and no evidence of a bladder. Dye had also entered the seminal vesicles (Fig. 1). After improvement of the renal status by resuscitative measures (catheterization, IV fluids, antibiotics) and IV pyelogram showed poor excretion of dye on both side with right-sided hydronephrotic changes and a small, pelvic left kidney.

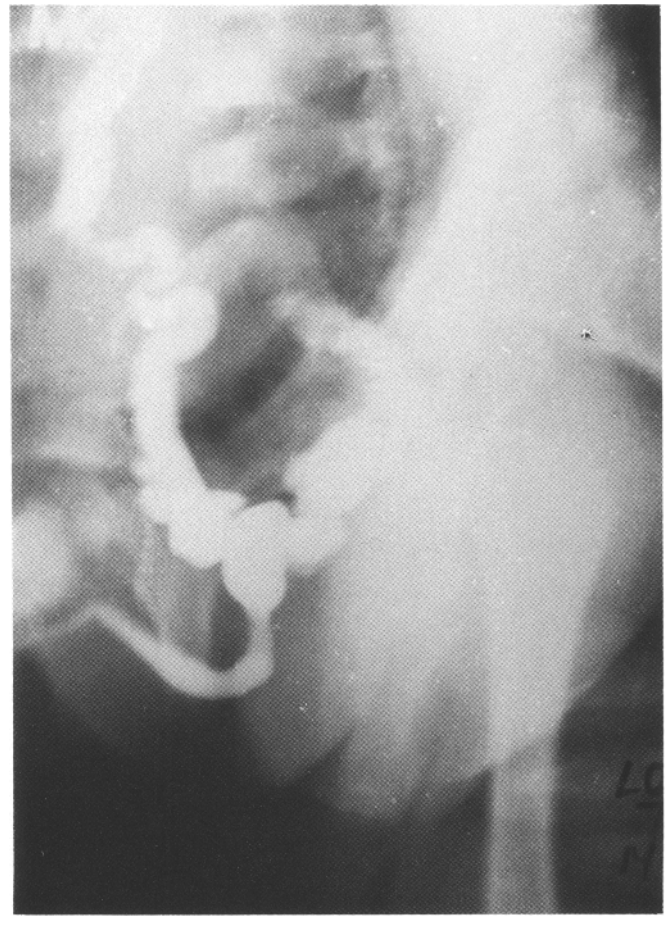

Fig. 1. Micturating cystourethrogram showing bladder agenesis and bilateral gross reflux

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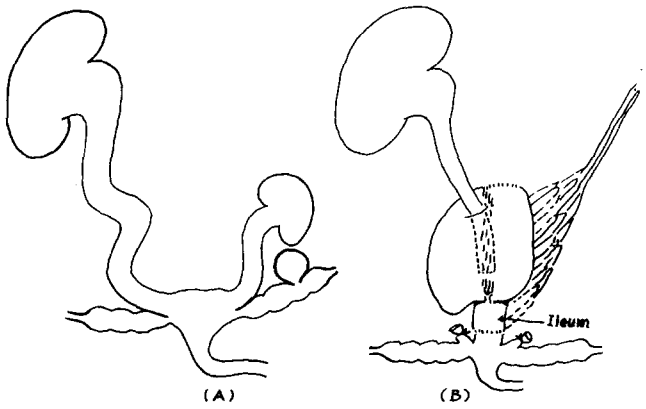

Fig. 2. Line diagram showing **A** peroperative findings and **B** operative procedure

At exploration via a lower midline incision, the urinary bladder could not be visualized. On opening the pelvic peritoneum, both ureters were dilated and tortuous and opened into the distended posterior urethra near the inferior border of the pubic symphysis. Both seminal vesicles were also distended; the left one contained a 1×1-cm cyst. The ejaculatory ducts on both sides opened into the lower ends of the ureters just proximal to their confluence with the urethra. The left kidney was dysplastic (2×1.5 cm) and lying at the pelvic brim.

Left nephroureterectomy was done, severing the ureter 0.5 cm above the site of insertion of the ejaculatory duct. The seminal vesicle cyst was also excised. The ileocecal valve was found to be competent and capable of holding 70 ml fluid. An isolated segment from the ileocecal region was used for construction of a functional bladder using the ileocecal valve as the bladder neck. The right ureter was also severed as described for the left side. After sacrificing the extra length of distal ureter, the latter was reimplanted into the cecum on the anterior taenia, forming a 3-cm-long submucosal tunnel (Leadbetter). The 2-cm ileal stump was then anastomosed to the top of the proximal urethra (Fig. 2). A silastic stent was left in the right ureter for 2 weeks.

The postoperative recovery was uneventful. Follow-up after 2 months showed a voiding volume of 80–100 ml with a dry period of $1\frac{1}{2}$ to 2 h. The child is thriving well, has gained weight, and the serum creatinine and urea are within the normal range.

Discussion

Up to now, 37 cases of congenital absence of the bladder have been reported in the literature, only 9 of which have been managed surgically. Only 1 was in a male patient [5]. In all the reported cases there was agenesis of the bladder and urethra and the ureters were found opening either into the vagina, the rectum, or a common cloaca [1, 3, 4]. The embryological explanations given for such anomalies were: (1) failure of mesonephric duct structures to form the trigone and posterior urethra [6]; and (2) failure of the urorectal septum to fulfill its function to partition the cloaca into an anterior bladder and posterior intestinal tract so that the ureters empty into the rectum or vagina [5].

The present case is unique because the entire urethra was well-formed and no fistula existed between the urorectal systems, in contrast to the other reported cases. It appears that the embryological aberration occurred after urorectal septum migration was complete and there was only failure of development of the anterior cloaca which led to absence of the bladder. This anomaly is frequently associated with the absence of one kidney [6], but in the present case there was a dysplastic, unascended left kidney. The previous 9 surgical cases were managed mostly by diversion, e.g., cutaneous ureterostomy, ileal conduit, or ureterosigmoidostomy [1–6]. We have provided a functional bladder of ileocecum; the patient is better accepted socially and psychologically because of the ability to pass urine per viam naturalis.

References

1. Campbell H (1951) Clinical pediatric urology. WB Saunders, Philadelphia, p 256
2. Duckett JW, Caldamone AA (1985) Agenesis and congenital hypoplasia of bladder. In: Kalalis PP, King LR and Belman AB (eds) Clinical pediatric urology, vol. 2. WB Saunders, Philadelphia, p 747
3. Gould GM, Pyle WL (1937) Anomalies of the bladder. In: Gloud GM, Pyle WL (eds) Anomalies and curiosities of medicine. Sydanham, New York, p 295
4. Graham SD (1972) Agenesis of bladder. J Urol 107: 660–661
5. Muecke EC (1986) In: Walsh PC, Gittes RF, Perlmutter AD, Stamey TA (eds) Campbell's urology, vol. 2. WB Saunders, Philadelphia, pp 1876–1877
6. Palmer JM, Russi MF (1969) Persistent urogenital sinus with absence of the bladder and urethra. J Urol 102: 590–594