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CASE REPORT

Urethral duplication with bilateral megaureter and bladder outlet obstruction: unusual case managed by PADUA technique

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Introduction

Urethral duplication is a rare congenital anomaly of the lower urinary tract. The most used classification system was suggested by Effmann [1] in 1976, based on radiological findings. The duplication most commonly occurs in the sagittal plane with one urethra located ventrally and the other dorsally [2]. Treatment depends on the type of duplication and comorbidities [3].

We describe an unusual presentation of complete hypoplastic type 2 A-2 urethral duplication according to Effmann with bilateral obstructive megaureter and prolonged bladder outlet obstruction, which was managed through Progressive Augmentation by Dilating the Urethra Anterior (PADUA).

Case report

A 2-year-old boy with prenatal diagnosis of bilateral hydronephrosis presented recurrent febrile urinary tract infections (UTIs). Physical examination revealed double urethral meatus, one orthotopic and one coronal hypospadic (Figure 1(a,b)). Weak urinary stream from the hypospadic meatus and dripping from the orthotopic one was observed with abdominal press. Ultrasonography (US) showed bilateral hydronephrosis with severe left megaureter, thickened bladder wall and bilateral paraureteral diverticula. Voiding cystourethrogram (VCUG) was not performed due to impossibility to insert urinary catheter. ^{99m}Tc-MAG3 renal scan revealed symmetric split renal function with obstructive pattern.

Cystourethroscopy showed hypoplastic ventral penile urethra and semi-atretic dorsal urethra ending on the glans tip. Obstruction of the bladder neck with absence of prostatic bundle in the orthotopic urethra was observed. The two urethras ended in a single obstructed bladder opening. Dilation of the bladder neck was performed and both urethras were catheterized (Figure 1(c)). Therefore, VCUG and retrograde urethrography confirmed the diagnosis of type 2 A-2 urethral duplication according to Effmann classification [1] (Figure 2).

Five months later, due to breakthrough febrile UTIs, the patient underwent open transvesical resection of bilateral paraureteral diverticula, advancement of right ureter, tapering and reimplantation of left ureter, 'Y-V' widening bladder neck plasty, and calibration of the ventral urethra. The dorsal urethra ending on the glans tip was left untouched.

Postoperatively, the patient developed prolonged bladder retention, requiring suprapubic urinary diversion for 6 months. Progressive augmentation by gentle catheterization of the ventral urethra (PADUA) was performed [4], using a 3 F ureteral catheter as a guide to introduce an open tip 8 F Foley catheter passing into the bladder. The Foley catheter was left in place for approximately 8 weeks and replaced with larger ones at 2-month intervals until satisfactory caliber of urethra was achieved (12 F). Endoscopic section of the bladder neck was performed 6 months after the reconstructive surgery. One week later, the supravesical diversion was clamped, spontaneous voiding was obtained, and the urinary diversion was removed. Post-operative VCUG showed normal capacity bladder, with regular walls and no diverticula, and normal profile of the ventral urethra at voiding (Figure 3).

To date, the child is asymptomatic, voiding through the hypospadic meatus with dry intervals, and with the orthotopic meatus functionally obliterated. The ventral urethra has maintained good caliber at 1-year follow-up and US showed recovery of the hydronephrosis and ureteral dilation, decrease of the detrusor hypertrophy with no significant post-voiding residual urine. The hypospadias repair has been postponed.

Discussion

Our case had uncommon clinical presentation. At time of diagnosis, the urethral duplication was associated with severe bladder neck obstruction, bilateral obstructive megaureter, and bilateral paraureteral diverticula. The dorsal orthotopic urethra was semi-atretic whereas the ventral urethra, which is usually of normal size and function, was severely hypoplastic. Surgical correction required bilateral ureteral reimplantation and widening bladder neck plasty, but it was complicated by prolonged bladder retention requiring suprapubic urinary diversion.

A similar case of urethral duplication presenting with chronic bladder retention, left scrotal transposition and left

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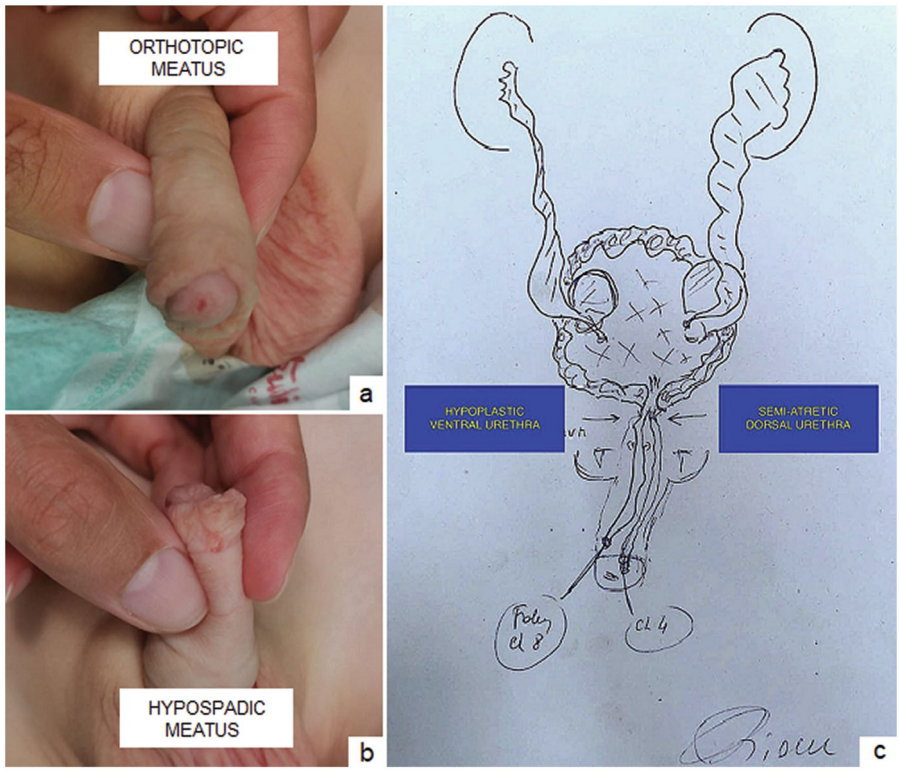

Figure 1. Patient anatomy: (a) narrow orthotopic meatus of the dorsal urethra; (b) coronal hypospadic meatus of the ventral urethra; (c) urethral duplication with semi-atretic dorsal urethra and hypoplastic ventral urethra; bilateral obstructed megaureter; bilateral paraureteral diverticula; bladder neck obstruction.

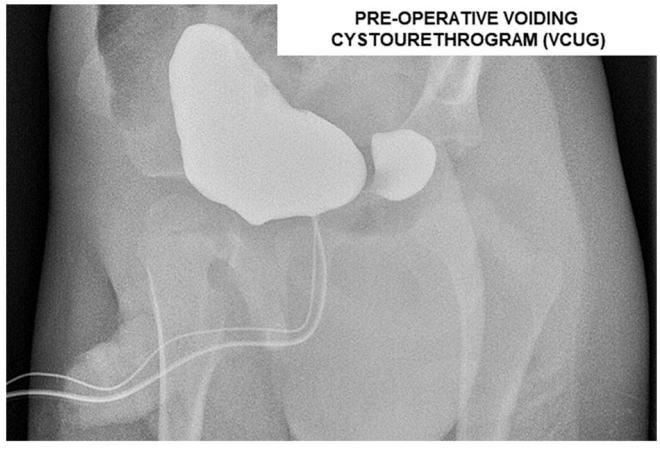

Figure 2. Pre-operative VCUG and retrograde urethrography confirmed the diagnosis of urethral duplication type 2 A-2 according to Effmann.

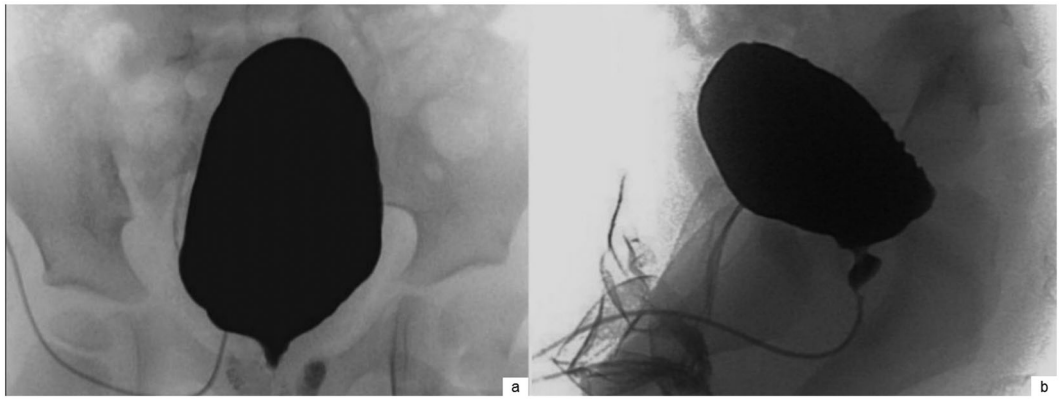

Figure 3. Post-operative VCUG through suprapubic catheter showed: (a) normal capacity bladder, with regular walls and no diverticula; (b) normal profile of ventral urethra at voiding phase.

renal agenesis was already described in the pediatric literature [5]. Both urethras were not functioning in such patient and supravesical Mitrofanoff outlet channel was needed to allow clean intermittent catheterization and solve the chronic urinary retention.

Conversely, we decided to manage our patient using a less invasive treatment strategy. We adopted the PADUA procedure, described by Passerini-Glazel et al. in 1988 [4] for treatment of severe urethral hypoplasia. Alternative treatment options would have been staged urethroplasty or 1-stage urethroplasty using free graft (buccal mucosa or preputial skin) [2,3]. However, these alternatives are not without complications, and may necessitate multiple and/or prolonged hospitalization. We preferred to not treat the dorsal obstructed urethra and adopt delicate progressive dilation of the ventral hypoplastic urethra.

In our experience, the PADUA procedure resulted in significant improvement of urethral caliber and progressive functionalization of the urethra with no long-term complications. The hypospadias reconstruction has been postponed to better assess the long-term outcome of the obstructed urethral duplication repair.

Author contributions

Maria Escolino: conceptualization; manuscript writing; manuscript editing.
Paolo Caione: conceptualization; manuscript editing; final supervision.
Mariapina Cerulo: manuscript editing; final supervision.
Benedetta Lepore: data collection; manuscript editing.
Annalisa Chiodi: data collection; manuscript editing.
Rachele Borgogni: data collection; manuscript writing.
Ciro Esposito: conceptualization; manuscript editing; final supervision.

Disclosure statement

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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