

Case Reports

Complete Duplication of Male Urethra: Case Report and Literature Review

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Abstract

Introduction: Urethral duplication is a rare congenital anatomical anomaly. It can often be associated with other organ system anomalies. **Methods:** We report a case of a male child with VACTERAL association and complete duplication of the urethra. The associated congenital anomalies and the various different urethral imaging techniques were also described. **Results:** The child underwent surgical repair of the duplicated urethra and resection of the associated anterior megalo-urethra at age 6. Early post op follow up showed excellent cosmetic and functional outcome. **Discussions:** We reviewed the literature regarding the embryology of the male urethra. Classification of the different anatomical variants of urethral duplication and their proposed aetiologies were also discussed.

Key words

Complete duplication; Male urethra

Introduction

Double urethra is a rare congenital urogenital anomaly. Aetiology is unknown. It is often associated with other organ system anomalies, in particular of the gastrointestinal tract. Herein, we describe a male child with complete duplication of the urethra and the subsequent surgical corrective procedure. In addition, we discuss the recent literature in the classification and management of urethral duplication.

Methods

A full term baby boy was born with unremarkable antenatal history. He was diagnosed to have VACTERAL

association shortly after birth. He suffered from absent bilateral radii, 13 pairs of ribs, lower sacrum hemi-vertebra, large ventricular-septal defect, low lying cord at L4 level with fatty film at S3 and anorectal malformation. For his urological system, he had small right kidney, ectopic insertion of left ureter to the proximal urethra, urethral duplication and anterior megalourethra. Anorectal-plasty, cardiac corrective surgery and multiple orthopaedics operations were performed in his early childhood.

The child had normal intelligence and was able to walk unaided with normal gait. He had independent activities of daily living and did not suffer from urinary or bowel incontinence. He was instructed to gently squeeze the redundant urethra after voiding to express the residual urine, other than this, he had no overt urinary symptoms and no recurrent urinary tract infection.

Magnetic resonance urogram (MRU) and DMSA scan of the kidneys showed no significant vesico-ureteric reflux, partial duplication of the left collecting system and a left : right split renal function of 60% : 30%.

Urodynamic study showed plateau shaped voiding curve, Qmax of 9.7 ml/s, normal bladder emptying efficiency, normal bladder compliance and absent of detrusor over-activity during filling phase.

The definitive urological correction was postponed to age 6 after discussion with his parents as he already had

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multiple operations since birth. Furthermore, given his relatively lack of symptoms and the complexity of the corrective operations, we also agreed with the scheduled elective reconstruction of the urethra when the continence and bladder function were more certain urodynamically.

Results

Examination under anesthesia, rigid cystoscopy and micturating cysto-urethro-gram (MCUG) (Figure 1) confirmed complete duplication of urethra in a coronal plane, with a small urethral opening at the dorsal side of the glans permitting a 24 gauge angiocatheter cannulation only and a normal sized urethral opening at the ventral side of the glans (at the level of the coronal region) permitting a 8 French Vygon catheter cannulation. The ventral urethral was also associated with a urethral diverticulum from coronal level to mid penile level.

After safeguarding the urethral opening with cannulation, the penis was degloved to the base. The ventral urethral diverticulum was slit opened at the midline, joining the dorsal duplicated urethra, which was also opened at the midline down to the mid penile region. Urethroplasty was done with 7-O PDS continuous subcuticular and excess skin was resected (Figure 2). The 8 French Vygon urethral stent was kept for 14 days post operatively to ensure adequate urine drainage in addition to another 10 French single lumen supra-pubic catheter. The patient was able to void spontaneously after the removal of the urethral stent and was discharge home 14 days after the operation. Subsequent follow up voiding curve showed normal bell shaped curve

and complete bladder emptying with no residual urine in the urethra.

Discussions

Complete duplication of the urethra is very rare. It is more common in male and the most commonly accepted classification is the Effman's classification which is as follows.¹

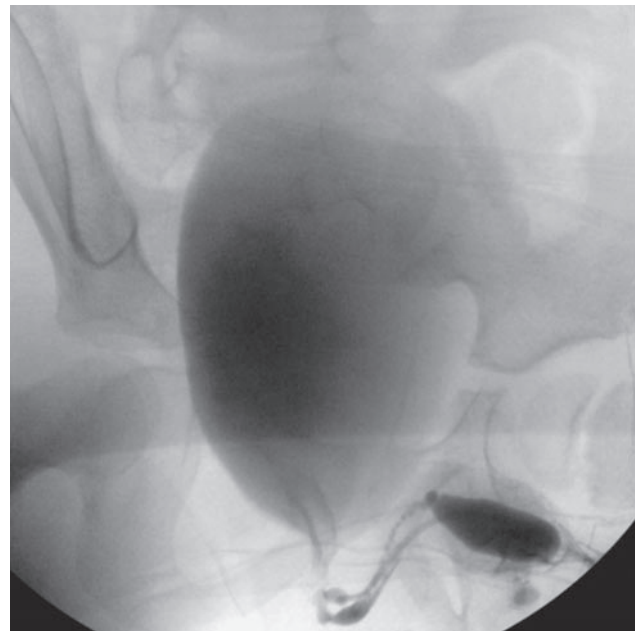


Figure 1 MCUG showing the complete urethral duplication with the anterior meglo-urethra.

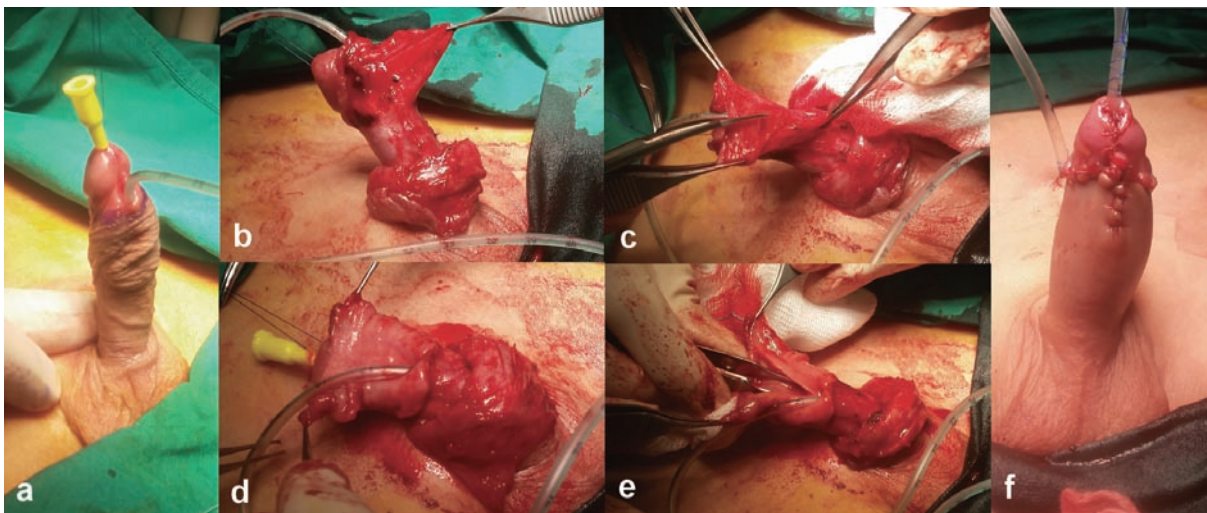


Figure 2 Intra-operative photographs showing the anatomy of the duplicated urethra and the surgical repair.

Type 1: incomplete urethral duplication

Type 2: complete urethral duplication

Type 3: urethral duplication as a component of partial or complete caudal duplication

According to the Effman's classification, our patient's duplicated urethra belongs to type 2.

By studying cloacal and urethral development in mouse embryos, Hynes and Fraher² described the formation of urethra as a separation from the phallic skin with a formation of plate-like structure, and this theory was later supported by the scanning electron microscopic photographs in staged rat embryos.³ Various different theories have been proposed to explain the unusual congenital anomaly of urethral duplication, Pippi Salle, et al,⁴ suggested the misalignment of the termination of the cloacal membrane with the genital tubercle as the cause. However, no single theory seems to explain all the multiple anatomical variants described in the literature, some suggested that the many different manifestations of urethral duplication is probably due to the different embryological origins,⁵ explaining each distinct clinical forms according to the different disturbances during embryogenesis.

Urethral duplication can sometimes also be associated with bladder duplication.⁶ Complete duplication of bladder and urethra is more commonly associated with anomalies of other organ system including gastrointestinal anomalies and vertebral anomalies. Campbell proposed splitting of the vesico-urethral anlage as the cause, suggesting that whether the split occurs before or after division of the cloaca by the urorectal septum determines if rectal anomalies will be present.⁶ While there may be overlapping features among genital duplication and the urorectal malformation sequence, no genetic basis has been identified.⁷

Patients with urethral duplication are usually asymptomatic, except for a urinary double stream and cosmetic problems. However, it has also been reported to be detected in the prenatal ultrasound findings of a male foetus,⁸ and it can also present as recurrent urinary tract infection in an adolescent boy⁹ and as bladder outlet obstruction in an adult male.¹⁰

While some patients may opt for conservative management for the complete duplication of urethra; as it can sometimes only gives rise to relatively mild symptoms such as double urinary stream, our patient opted for early urethral reconstruction as the pre-op voiding curve showed

mild obstructive pattern, which may potentially be due to the narrowed urethral opening at the dorsal side. Furthermore, the associated anterior megalourethra may also posed as an infection risk due to urine stasis and it was cosmetically unappealing.

Complete urethral duplication, although extremely rare, remains as one of the most complex urological congenital anomalies. We presented a baby boy with VACTERAL association and complete duplication of the urethra. Various different modalities of imaging, including MRU and MCUG have also been discussed in the work up of this uncommon clinical entity. A complete knowledge of the anatomy of the anomaly helps surgeon to form an individualised management plan for each patient.

Declaration of Interest

None.

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