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A rare case of congenital Y-type urethral duplication

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Abstract Duplication of urethra is a rare congenital anomaly. We report a case of Y-type of urethral duplication with the accessory urethra arising from posterior urethra and opening in the perineum. The orthotopic urethra was normal. The accessory urethral tract was cored, transfixed and divided. At 1 year of follow-up, the patient has no urinary complaints.

Keywords Congenital; duplication; urethra; child.

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INTRODUCTION

Urethral duplication is a rare congenital malformation and only 325 cases have been reported in the literature to date [1,2]. It is more frequently seen in males and is often associated with genitourinary and gastrointestinal anomalies [1]. The clinical presentations vary from being asymptomatic to urinary tract infections (UTI), double stream or incontinence according to the anatomical variant present [1,2]. Although a number of theories have been proposed to describe the embryologic development of urethral duplication, the etiology and mechanism of this disorder is unknown [2].

We describe a 4 year old boy who presented with urinary dribbling from the external opening of the accessory urethra in the perineum.

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CASE REPORT

A 4 year-old boy presented with complains of dribbling of urine from a small opening in the perineum since birth (Fig. 1). He passed urine in a good stream from the normal urethral meatus. His bowel habits were normal and he had no history of passing fluid per anum. His physical examination was normal except for the small opening at the base of scrotum through which he passed drops of urine. The penile shaft and the anal opening were normal.

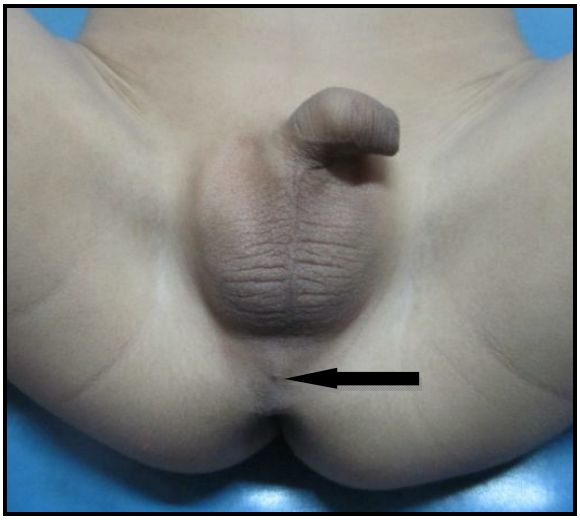

Fig. 1. External opening of the accessory urethra at the perineum (Arrow head).

Voiding cystourethrography (VCUG) revealed a fistulous tract from the prostatic urethra to the external perineal opening (Fig. 2). The native urethra was of normal caliber. There was no vesicoureteral reflux (VUR) and the bladder was normal.

On cystoscopy, the orthotopic urethra was normal and an internal opening was seen on the lateral wall of the prostatic urethra proximal to verumontanum (Fig. 3).

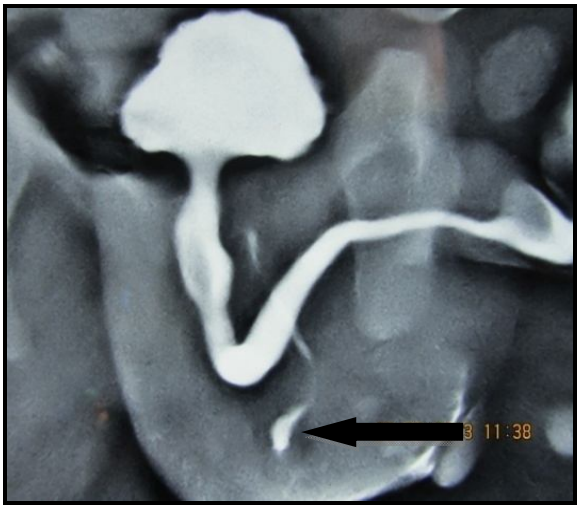

Fig. 2. Voiding cystourethrography showing a fistulous tract from the prostatic part of urethra to the external perineal opening (Arrow head).

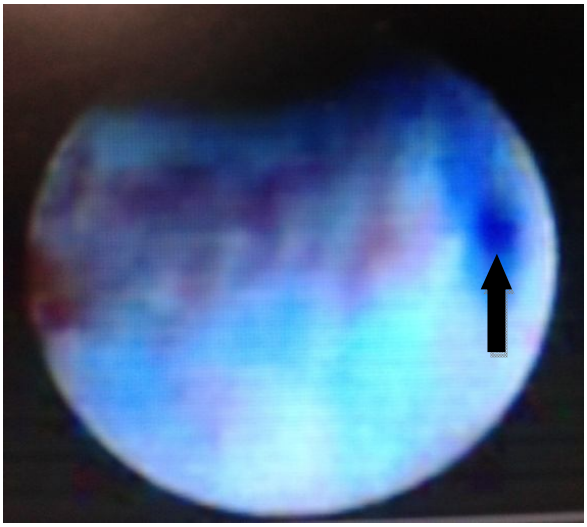

Fig. 3. Cystoscopy showing internal opening of the accessory urethra (Arrow head).

Intraoperatively, the patient was catheterized, an elliptical incision was taken around the external opening & the fistula tract was cored out upto the prostatic urethra, transfixed and divided (Fig. 4).

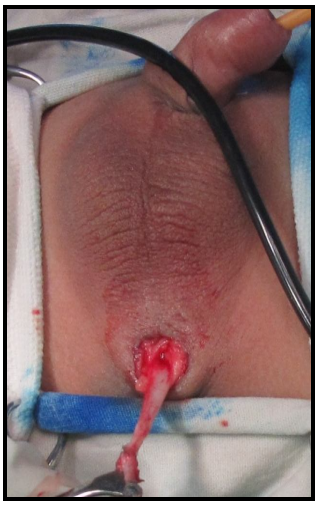

Fig. 4. Intraoperative image showing the cored out accesory urethra.

Histopathology showed transitional epithelium lining the tract. At one year of follow-up, repeat VCUG is normal and the patient has no urinary complaints.

DISCUSSION

Duplication of urethra is a rare congenital anomaly and is usually associated with other abnormalities in the genitourinary tract, heart, bowel, and bones [2-5]. It was first described by Aristotle and Vesalius [6,7]. Effmann et al. [2,3,8] has classified urethral duplication in to the following 3 types: Type I (25%): Blind-ending accessory urethra (incomplete urethral duplication). IA: Distal-duplicated urethras opening on the dorsal or ventral surface of the penis but not communicating with the urethra or bladder (the most common type). IB. Proximal-accessory urethra opening from the urethral channel but ending blindly in the periurethral tissues (rare). Type II (63%): Completely patent accessory urethra. It is divided into 2 parts: A (2 meatus) and B (1 meatus). Type II A1: Two non-communicating urethras arising independently from the bladder. Type II A2: Second channel arising from the first and coursing independently into a second meatus (Y-type). Type IIB: Two urethras arising from the bladder or posterior urethra and uniting into a common channel distally. Type III (12%): Accessory urethras arising from duplicated or septated bladders. This classification is the more functional, representing all clinical types of urethral duplication [2]. The most common type of urethral duplication is the Y type with a perineal or rectal fistula associated with stenosis of the anterior portion of the normally situated urethra [9].

One-third of patients with urethral duplication are associated with VUR [1]. Most urethral duplications found in the same sagittal plane and can be divided into either dorsal or ventral [10]. Some rare types of urethral duplication may occur in the same horizontal plane, and the urethra lies next to one another, one left and one right.

Additionally, the horizontal type may be associated with duplicated phallus or complete bladder duplication [10].

A dorsal urethral duplication occurs when there is a normal urinary meatus, dorsal chordee of the penis and a second epispadiac meatus along the shaft of the penis. The foreskin of the penis may be incomplete dorsally and the epispadiac meatus can also be found anywhere along the shaft of the penis [11]. Ventral urethral duplications are extremely rare and may also be complete or incomplete with a blind-ending urethra. There may be normally located urethra on the glans and a second along the ventral aspect of the penis and even throughout the perineum [10]. The calibration of the dorsal urethra may be of normal or narrow. The ventral urethra is considered the more normal because it generally passes out of the bladder neck and sphincter [11,12]. The "Y" duplication is formed when the prostatic urethra splits into two channels with one extending to the glans and the more functional ventral one coursing to the perineum adjacent the rectum [2,10]. In our case, the dorsal orthotopic urethra was normal and the ventral duplicated urethra was stenotic. This anomaly is however classified as a congenital urethroperineal fistula [13]. On the other hand, Wagner et al accepted all congenital urethroperineal fistulas as urethral duplications [13,14] because of the presence of transitional epithelium lining the tract as was in our case.

Various theories have been proposed to explain the embryological development of urethral duplication, but etiology and mechanism of this anomaly is not well understood. The male external genitalia occur from the cloacal folds, labioscrotal swelling, urogenital sinus and preputial swelling. Additionally, it is also composed of all 3 germ layers together with timely interaction of sonic hedgehog signals, fibroblast growth factors, Hox genes, bone morphogenetic proteins signals and androgens [3]. Casselman and Williams [15] suggested that a partial failure or an irregularity of the ingrowth of the lateral mesoderm between the ectodermal and endodermal layers of the cloacal membrane in the midline accounts for the forms with a dorsal epispadiac channel. Das and Brosman [16] stated that abnormal termination of the Mullerian duct was responsible for the development of urethral duplication. Currently, the most widely accepted theory for the cause of complete urethral duplication is that of Patten and Barry, and is supposed to result from an abnormal relationship between the lateral folds of the genital tubercle and the ventral end of the cloacal membrane [3,17,18]. But this theory

does not explain all subtypes of urethral duplication [3].

No genetic study for urethral duplication per se has been reported but its association with some disorders of midline development like VACTERL complex (vertebral anomalies, ventricular septal defect, anal atresia, trachea-oesophageal fistula, radial dysplasia, renal anomalies, cleft lip/palate) and Pallister Hall syndrome have favoured genetic linkage to the abnormalities of GLI3 gene on chromosome 7p13 as it interacts with the Sonic Hedgehog gene (Shh). These genes are responsible for early embryonic development that takes place between the fourth and sixth weeks of gestation [19].

Y-type urethral duplications differ from other forms of urethral duplications in that there are often other associated severe congenital malformations like cloacal exstrophy, conjoined twins, early amnion rupture syndrome, limb x body wall disruption, Prune Belly Syndrome and hand-foot-genital syndrome [19]. Urethral duplication is also strongly associated with anorectal malformations. Of all the congenital complete urethral duplications, the incidence of Y-type urethral duplications is about 30%, as reported by various published series [19].

Depending upon the type of duplication, the patients may be asymptomatic or present with symptoms of UTI, epididymitis or incontinence [1]. Incomplete ventral accessory urethra often presents as a preputial sinus or swelling which are usually either epidermoid or mucoid cysts [3]. Diagnosis of urethral duplication is based on simple physical examination of the penis and is confirmed by a voiding cystourethrography or retrograde urethrogram, which will allow identification of its anatomical type [2]. It can also be differentiated from urethral diverticulum by voiding pattern or VCUG [3]. Our patient was classified as type II A2, i.e., Y-type because the accessory urethra was arising from the posterior urethra and was opening into the perineum and was stenotic. The orthotopic urethra was normal in caliber.

The primary aim of surgical repair of Y-type urethral duplication is to preserve normal functioning orthotopic urethra with intact verumontanum, good caliber and intact sphincter [20]. Treatment of urethral duplication should be individualized based on the anatomic types and also clinical findings and severity of the accompanying anomaly [2]. The progressive augmentation by dilating the urethra anterior (PADUA) procedure is done with soft catheter for the treatment of severe ventral urethral hypoplasia in cases of prune belly syndrome, urethral duplication and patent urachus

[3,20]. Gradual dilatation of orthotopic urethra keeps the urethral patency and maintains the normal urethral development without morbidity of surgical procedures of urethrotomy or urethroplasty. Preputial island flap, buccal/bladder mucosal graft or combined grafts are used in a single or staged urethroplasty in cases of atretic penile orthotopic urethra and perineal duplication [3,20]. In our case, an elliptical incision was taken around the external opening & the fistula tract was cored out, transfixed and divided. The normal orthotopic urethra was left intact.

Ventral Y- urethral duplication is a rare congenital anomaly, often found associated with other anomalies. It has varied clinical presentations, therefore good history and clinical examination is essential. Diagnosis is confirmed by radiological and endoscopic studies. Surgical management needs to be individualized depending on the anatomy of the native and duplicated urethra.

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