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**Type II A2 duplication of urethra in an 8-month-old male child:
A case report**

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Abstract Urethral duplication is a rare congenital anomaly, usually found with multiple anatomical variants. In this article was presented a case of urethral duplication in an 8-month-old male child. The malformation was characterized by the presence of continent hypospadiac and normal apical urethra. Retrograde urethrogram through both urethral tracts simultaneously revealed the malformation as Effmann Type II A-2. The accessory ventral urethra was excised without complication.

Key words Urethral duplication; Effman classification; accessory urethra; congenital anomaly; reconstructive surgery.

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INTRODUCTION

Urethral duplication is a rare congenital malformation and is more frequently found

in males. Approximately 325 cases have been reported in the literature to date. Although a number of theories have been proposed to describe the embryologic development of urethral duplication, the aetiology and mechanism of this disorder is unknown. Thus, there are various types of

classification. Williams and Kenawi [1] classify the urethral duplications according to the location of the ectopic urethra compared to the normal urethra. Effman et al. [2] identified a more detailed classification into three types associated with subclassifications. The clinical presentation differs according to the anatomical variant present. We present a case of Type II A-2 urethral duplication in an 8-year-old boy and the literature review.

CASE REPORT

An 8-month-old male child presented with complaint of a double stream of urine since birth. Both urethras were continent, and there was no complaint of a burning sensation or dribbling of urine during micturition. External genitalia were well developed. Physical examination revealed a normal, healthy child with no other associated congenital abnormality. There was a normal meatus at the apex of the glans and a secondary meatus on the ventral aspect of the glans. A voiding cystourethrogram showed a double urethra as far the membranous urethra. Renal ultrasonography revealed both kidneys to be normal. Retrograde urethrogram through both tracts simultaneously confirmed a second channel arising from the primary urethra distal to its junction with the bladder and coursing

independently to a second meatus (Effmann Type II A-2). Surgical treatment began with endoscopy to view the length of the urethra accessory and the normal urethra. The accessory ventral urethra was excised without complication (Fig. 1A, B).

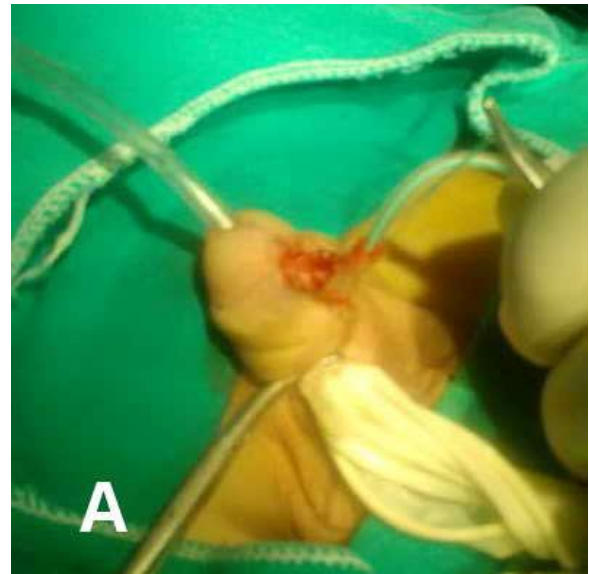


Fig. 1A, B. The catheterisation of both urethras and the secondary meatus over the ventral aspect of the glans.

DISCUSSION

First described by Aristotle, urethral duplication is a rare congenital anomaly and is often associated with other abnormalities in the genitourinary tract, heart, bowel, and bones [3–5]. Arena et al. [6] found that in 60% of cases is accompanied by genitourinary malformation such as ureteropelvic junction obstruction, extrarotation of the penis, vesicoureteral reflux, renal ectopia, renal agenesis, or posterior urethral valves. There are serious intestinal anomalies as combined oesophageal-duodenal atresia or malrotation of the gut in one out of ten cases. There was no additional abnormality in our patient.

Various theories have been proposed to explain the embryological development of urethral duplication, such as incomplete mesodermal fusion, abnormal Mullerian ducts, ischemic events in the embryogenesis, and defects in the development of the urogenital sinus [1,4,7–9]. Casselman and Williams [10] 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 [7] stated that abnormal termination of the Mullerian duct was responsible for the development of urethral duplication. Rica et

al. [11] reported that asymmetry in the closure of the urorectal septum results in an urethra-perineal fistula.

Gross and Moore classified urethral duplications as a complete second passage from the bladder to the dorsum of the penis or as an accessory pathway that ends blindly on the dorsal or ventral surface [12]. However, the classification of Effmann et al. [2] is the most commonly accepted in this area (Fig. 2).

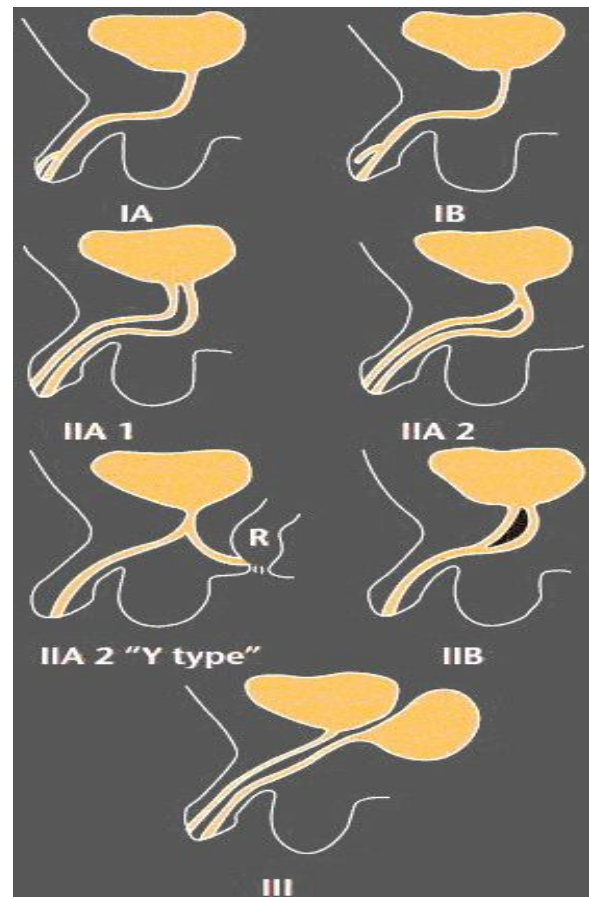


Fig. 2. Classification of urethral duplication according to Effmann et al. Type I (A,B): Blind ending, incomplete urethral duplication or accessory

urethra. Type II A1: Complete patent urethral duplication with the second channel arising independently from the bladder. Type II A2: Complete patent urethral duplication with the second channel arising from the first and courses into a second meatus. Type II A2 (Y Type): Complete patent urethral duplication with the second channel arising from the first and courses into a second meatus located in the perineum. Type II B: Complete patent urethral duplication with the second channel arising independently from the bladder but joining the first and coursing into one meatus. Type III: Urethral duplication as a component of partial or complete caudal duplication

This classification is the more functional, representing all clinical types of urethral duplication. 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 [13]. According to this classification, our case belonged to Type 2 A-2; the two urethras join below the external sphincter, and there is no incontinence.

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. Ultrasound may be helpful in the diagnosis of other associated anomalies. The first step in the management of urethral duplication is to recognize the

normal-functioning urethra; in fact, the true urethra is that with the largest calibre, a normal verumontanum, and an intact sphincter [14]. Hence, cystoscopy applies prior to surgical intervention to confirm the apical urethra and also shows the presence of the verumontanum [15]. However, many of the patients had a very long segment of atretic orthotopic urethra that did not allow the passage of the smallest cystoscope or an endoscopic guidewire, thus precluding the possibility of using the progressive augmentation by dilatation of urethra anterior technique [16]. Additionally, if it is found to be stenotic, the duplicated urethra should be used for reconstruction [17].

Treatment of urethral duplication should be individualized based on the anatomic types and also clinical findings and severity of the accompanying anomaly. Many patients are asymptomatic and do not require any surgery. Surgical treatment is needed for correction of cosmetic deformity, such as epispadiac meatus, double stream, and perineal urine leakage. If the two urethral orifices are too close to each other on the glans, then the septum between the two meatus can be excised to give a single urinary meatus. Other than this, most procedures involve excision of the accessory urethra with reconstruction of the normal urethra [18,19]. In our case, the hypospadiac

accessory urethra was excised completely. Single-stage repair of this type is feasible where the accessory ventral urethra is mobilized from the normal urethra in the membranous region. The long-term outcome in this patient was very good.

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