

CLINICAL CASE

URETHRAL DUPLICATION – A RARE CASE

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Abstract

Urethral duplication is a very rare congenital anomaly that has scarcely been reported. The embryologic origin of this condition is being debated by a few theories but none of them can fully explain it. There is a large spectrum of anatomical variations and this generated a number of classifications. While some of them described the urethral duplication in sagittal plane, others in the coronal plane. The prevailing classification used nowadays is the Effmann's classification. We present a rare case of a 5 years and 11 months old male patient having urethral duplication. Using the previously mentioned classification, we categorized his condition as type IA. The surgical treatment of this condition was performed (removal of the accessory urethra) in order to prevent complications like recurrent local infections and impaired micturition and also for esthetical issues.

Keywords: *urethral duplication, accessory urethra***Introduction**

Urethral duplication is a very rare congenital anomaly with about 300 reported cases and it is more frequent in males [6]. This condition is often associated with genitourinary and gastrointestinal anomalies.

Concerning the embryologic origin of the urethral duplication there are a few theories, but having in mind all the types of this condition, none of them can fully explain it. For example, one theory considers that this anomaly is caused by an abnormal Müllerian duct termination [3], while others state that a misalignment of the termination of the cloacal membrane with the genital tubercle [5] is the cause. On the other hand, another theory testifies that an asymmetry in the closure of the urorectal septum is the cause of a urethral-perineal fistula [3].

Case presentation

We present the case of a 5 years and 11 months old, male patient who was admitted to the hospital due to an abnormal orifice on the midline, in the pubic area, at the base of the penis. This respective abnormality was present since birth. Moreover, during his childhood the patient reported a few episodes of local swelling and emission of a yellow fluid through it. These episodes had a spontaneous resolution.

Clinical examination showed a 1 mm diameter orifice at the base of the penis, on the midline, covered by a skin fold. The urethral meatus opened normally at the tip of the glans, the penis was normal and regarding the testes, both of them were present inside the scrotum.



Figure 1- Opening of the accessory urethra on the midline, on the dorsal side of the penis



Figure 2. Opening of the accessory urethra

At that time, the probable diagnosis was urethral duplication. The patient underwent a full workup (blood and imaging tests). The abdominal ultrasound showed both kidneys were of a normal size, shape and position, there were no dilation of pyelocaliceal system, the urinary bladder had normal shape and dimensions and the other abdominal organs had normal ultrasound aspect. The IVU indicated both secretion and excretion present at 7 minutes and normal aspect of pyelocaliceal systems and ureters. A VCUG was performed (anteroposterior and lateral view) indicating no VUR and no post-micturition residual urine. Contrast agent was injected during VCUG through the accessory urethral meatus. The meatus followed the main urethra and stopped near the pubic symphysis. Having in mind the Effmann classification and the imaging findings, we decided to categorize our patient's urethral duplication as type IA.



Figure 3. VCUG – bladder and main urethra

Without undergoing surgery, the anomaly can lead to recurrent local infections which can affect the main urethra and the micturition. Both this aspect and the esthetic issue indicated the need for surgery. Before the surgical procedure, we performed a urethrocystoscopy which revealed there was no communication at the anterior urinary bladder wall, at the bladder neck or the anterior wall of the urethra and the posterior urethra was normal.

Before the operation, the patient was properly rehydrated. The surgical intervention was performed under general anesthesia with orotracheal intubation. A urinary catheter was placed in the main urethra as well. The operation started with placing a CH6 urinary catheter in the accessory urethra and injecting methylene blue into it. After making an incision around the small orifice at the base of the penis, we dissected the accessory urethra away from the surrounding tissue. At 2 cm away from the distal end, the fistula started narrowing, changed direction, heading towards the pubic symphysis and became very adherent to the penile dorsal vessels. This required a very careful

dissection[1]. At 4 cm away from the distal end, the accessory urethra was situated above the pubic symphysis and it had no lumen. At this stage we decided to double ligate the urethra and remove it. A subcutaneous drain was placed before closing and histological examination was required for the removed accessory urethra.



Figure 4. Dissected accessory urethra

Postoperative evolution of the patient was good. The urinary catheter was removed at 3 days after the surgery and the patient was discharged 2 days later, in a good state.

The result of the histological examination of the removed accessory urethra showed there was urothelial tissue, confirming the diagnosis of urethral duplication.

Discussions

Urethral duplication is an extremely rare congenital anomaly with less than 300 cases reported to date [7-10] and its diagnosis is a real challenge. This anomaly is more frequently seen in males, only 11.6% cases being reported in females [2].

There is a wide spectrum of anatomical variations of urethral duplication and this generated a number of classifications. The most known and used nowadays is Effmann’s classification [4]. Three categories have been differentiated, as shown in the table below.

Type	Details
Type I - Incomplete urethral duplication	I A. Blind-ending distal-accessory urethra communicating with the body of the penis, but not with the main urethra or bladder.
	I B. Blind-ending proximal-accessory urethra arising from the main urethra but ending in the periurethral tissues.
Type II – Complete urethral duplication with functional accessory urethra. There are two subtypes: A (two meatuses) and B (one meatus).	II A1. The two non-communicating urethras emerge from different origin points in the bladder.
	II A2. The accessory urethra originates from the main one and terminates in its own meatus, located either on the penis or the perineal region (Y-form variant).
	II B. Even though the two urethras have different origin points, they unite distally into a common channel.
Type III	Involves previously existing duplicated or septated bladders, each being continued by its own urethra.

Table 1. Effmann’s classification of urethral duplication [4]

Conclusions

Urethral duplication is a very rare congenital anomaly. The need for a unique, personalised surgical approach stems from the fact that each reported case had its own anatomical variant and embriological origins.

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