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Our experience on urethral duplication in children: 4 case reports and literature review

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ABSTRACT

Urethral duplication is a rare lower urinary tract anomaly, which can present with a variety of clinical manifestations. Here, we report four cases of urethral duplication and review the literature to report the diagnostic approach and treatments used for this condition. The cases demonstrated the variety of clinical symptoms and anatomy considerations of this particular malformation. Surgical treatment must be individualized according to the anatomical finds.

Key Words: Urethra; duplication; urinary tract anomaly; children

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Introduction

The urethral duplication is a rare lower urinary tract anomaly first described by Aristotle [1]. It can present a variety of clinical manifestations such as deformed penis, twin streams, urinary tract infection, urinary incontinence or flow obstruction and other associated anomalies [2]. Surgical management is determined by urethral and bladder neck anatomy as well as the presence of symptoms [3]. This article aims to report four cases of urethral duplication, discussing their particular presentations and management.

Case 1

A ten years-old boy started with urinary loss by the perineal region during every micturition, one year before the medical appointment. The patient denied any prior urinary complains. On physical examination, he had a small orifice on the perineal midline, 1 cm anterior to the anus. The voiding cystourethrogram demonstrated a duplicated pathway from the penile urethra to the perineal region [**Fig. 1A**]. The patient was submitted to a single stage repair by surgical resection of the duplicated path from the perineal region, all the way up to the urethra

[Fig. 1B]. The histopathological exam confirmed the urethral tissue. At 1 year follow-up he was asymptomatic.

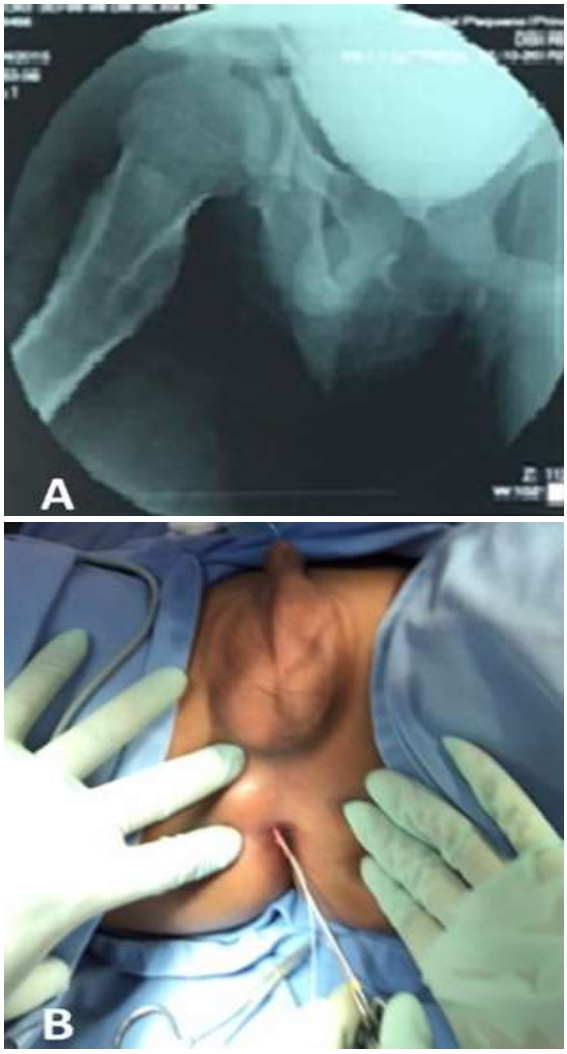

Fig. 1. **(A)** Voiding cystogram demonstrating the duplicated urethra. **(B)** Surgical resection.

Case 2

A 5 years-old boy, referred as hypospadias, presented no urinary complains. Physical exam detected distal hypospadias, no ventral chordee. Surgical correction was proposed. During the surgical procedure the urethral catheterization stopped at the middle shaft. Urethral dilatation was tried without success. After a very careful inspection, a small orifice was detected in the ventral midline. Catheterization was possible with a 6fr tube that progressed to the bladder. An incomplete urethral duplication, with 3 cm length, was characterized [Fig. 2A]. The correction was performed with Koff technique, the posterior wall of the ventral urethra was opened and both urethras were communicated [Fig. 2B]. The follow-up was uneventful.

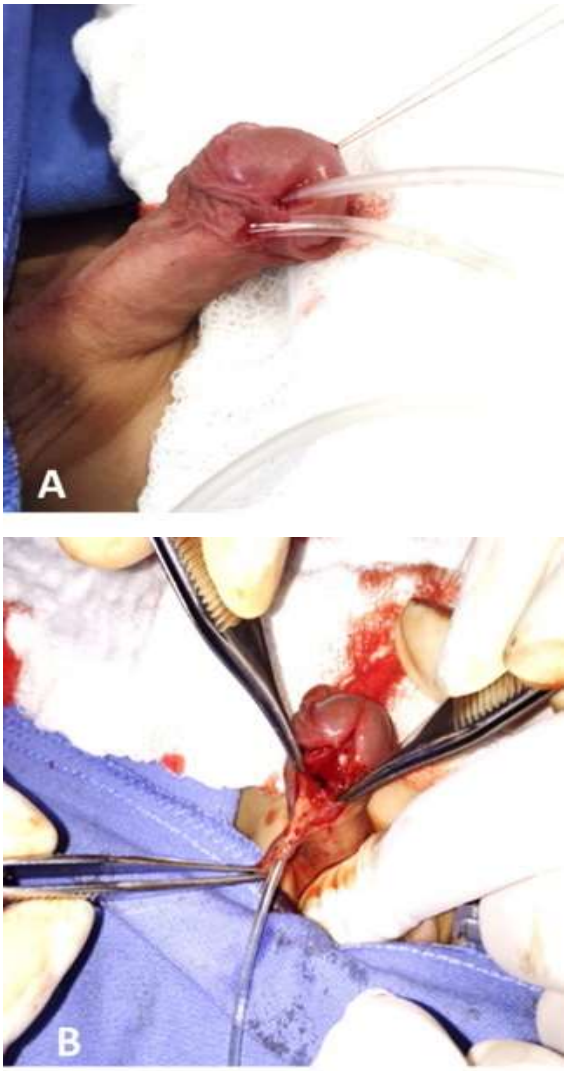

Fig. 2. (A) Urethral duplication. **(B)** Ventral urethra posterior wall opening.

Case 3

A 4 years-old boy presented with rectal urine discharge. During cystourethrogram attempt, a very thin urethra in the dorsal aspect of the

penis was identified. After contrast injection into the bladder, the ectopic urethra was detected to end into the rectum [**Fig. 3A**]. Surgical correction was performed in two steps: at first the urethra was mobilized from the rectum into the perineum in association with a colostomy. Six months later, a free graft urethral tube of 10 cm was used to reconstruct the urethra from the perineum to the glans [**Fig. 3B**]. An anastomotic stricture required multiple dilatations. After a 3 years follow-up the patient present no complains.

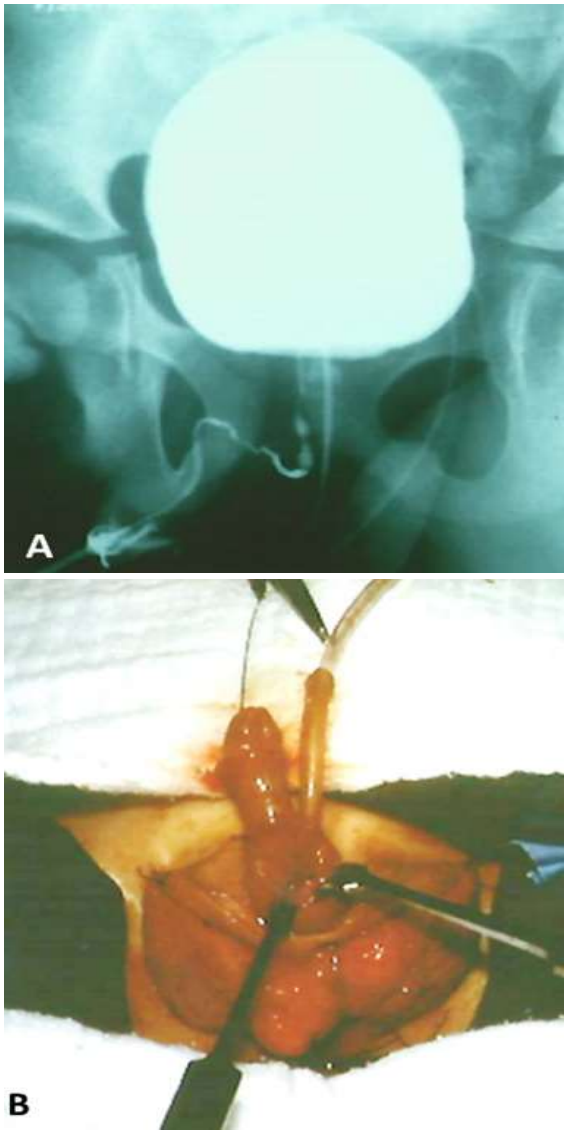

Fig. 3. (A) Cystourethrogram, urethral duplication. **(B)** Surgical correction.

Case 4

A 1 month-old patient was referred for congenital hydronephrosis and hypospadias without any previous urinary symptom. During the first clinical exam the presence of two urethral meatuses were detected on the tip of the glans and another in the perineum [**Fig. 4A**]. An urethrocistogram showed two regular caliber urethras, one topic and another extending to the perineum. Both presented a common posterior dilated urethra, suggesting posterior urethral valve [**Fig. 4B**]. Endoscopic valve ablation was performed uneventfully. After an 8 months follow-up period the patient is asymptomatic and waiting for surgical correction, possibly with ventral urethral resection.

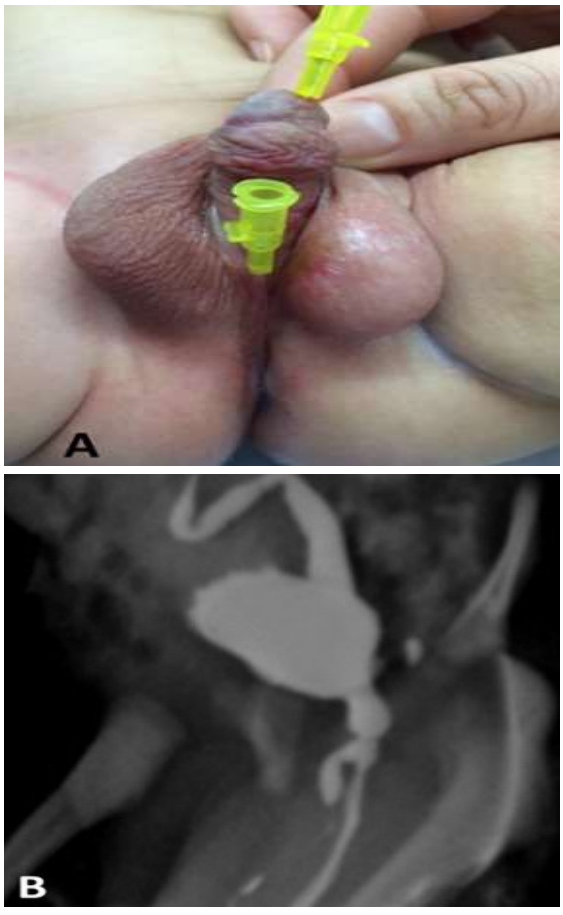

Fig. 4. (A) Urethral duplication. **(B)** Cystourethrogram.

Discussion

Urethral duplication is a rare anomaly with about 300 cases reported in the literature [1]. The embryogenesis of urethral duplication is not well understood and various hypothesis exist. The commonly accepted hypothesis for complete urethral duplication results from an abnormal relationship between the lateral folds of the genital tubercule and the ventral end of the cloacal membrane, proposed by Patten and Barry [3].

The diagnosis is usually made by voiding cystourethrogram, demonstrating two channels [4]. The most used classification was proposed by Effman as showed in Figure 5. Type 1, blind accessory channel is the most common [4]. Classification of urethral duplication according to Effmann et al. is as follow:

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

In our series we were able to investigate and operate different types of this particular malformation.

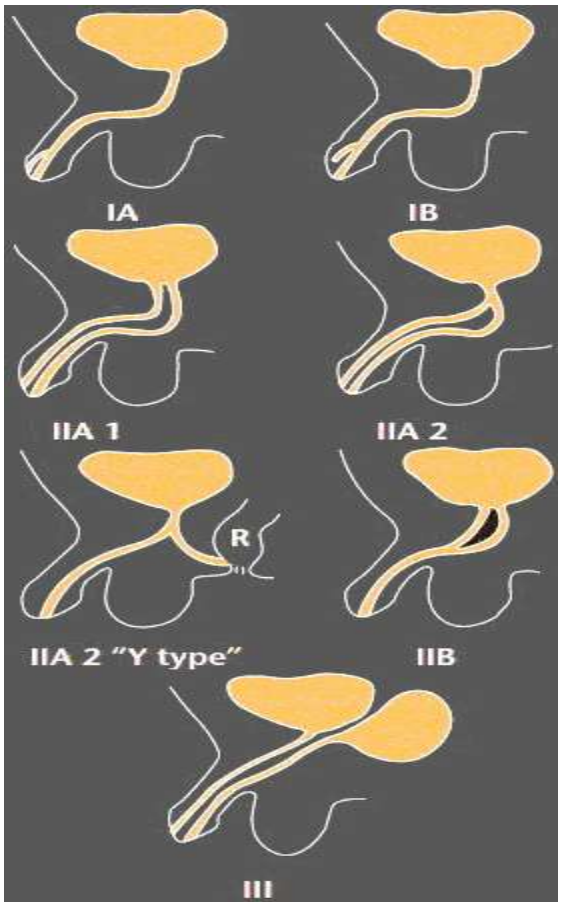

Fig. 5. Effman's Classification.

Besides the identification of the malformation itself, it is important to recognize and preserve the functional urethra, it is a serious mistake to discard the functional ectopic urethra, preserving the orthotopic one [5].

Type I is the most common and simple to repair. The patients may be assyntomatic, but most cases present mucous discharge and urinary infection. The treatment involves the excision of the accessory urethra [2]. In type II duplication, the dorsal urethra is usually hipoplastic. The functional urethra usually opens to the perineum. In a few cases the orthotopic urethra is the functional one [4].

The surgical repair of duplication is challenging. The key points for successful

correction are identification of the functional urethra, usually presenting normal verumontanum and an intact sphincter [6,7]. Despite the variety of malformation and surgical procedures performed, the only complication detected in our series was urethral anastomotic stricture.

Conclusion

Urethral duplication is a rare and complex congenital anomaly. Careful investigation is necessary to define the functional urethra and guide the best surgical approach.

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