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# Urethral duplication — A wide spectrum of anomalies

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**Abstract** *Objective:* Urethral duplications are rare lower urinary tract anomalies, with multiple anatomical variants described. This paper aims to separate this complex anomaly into different diseases, each with distinct clinical forms according to the disturbance during embryogenesis, yet noting a few similarities that may be helpful in their management. The classification system of urethral duplication is also discussed.
*Material and methods:* Twelve urethral duplication cases over a 14-year period were reviewed. Clinical presentation, the imaging studies used to ascertain anatomical details, type of urethral duplication and surgical correction used in the treatment of patients are presented.
*Results:* Nine patients had urethral duplication in the sagittal plane and three patients in the coronal plane. Of the patients with sagittal urethral duplication, 3 had pre pubic sinus, 3 had epispadiac urethral duplication, 1 had a dorsal urethral duplication deviated from the midline and 2 had hypospadiac urethral duplication. All the patients with coronal urethral duplication had associated bladder duplication. The surgical correction of the patients with sagittal urethral duplication included excision of the pre pubic sinus, excision of the duplicated urethra, and urethroplasty. Excision of the hemibladders' septum and closure of one bladder neck was the treatment for patients with coronal urethral duplication and bladder duplication.
*Conclusion:* Urethral duplication is a complex anomaly and the different manifestations probably have different embryological origins. Each group, sagittal or coronal, has a few similarities that may be helpful in their management, although every diagnosed case presents a unique anatomy and surgical treatment must be individualized.


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## Introduction

Urethral duplications are rare lower urinary tract anomalies. Several types of such anomalies have already been described in the literature. The most frequent anomaly occurs in the sagittal plane, in which case the duplicated urethra is in either the dorsal or ventral position in relation to the orthotopic urethra [1]. Usually the ventral urethra is the more functional of the two and contains the sphincteric mechanism. Duplications occurring in the coronal plane are quite rare and they are usually associated with bladder duplication [2].

The most used classification system of urethral duplication is that proposed by Effman. This classification system is based on radiological findings and divides urethral duplications into incomplete (type 1), complete (type 2) and coronal (type 3). Alterations during the different stages of embryogenesis of the urinary tract are responsible for each form of duplicate presentation. This paper aims to separate this complex anomaly into different diseases, each with distinct clinical forms according to the disturbance during embryogenesis, yet noting a few similarities that may be helpful in their management. To that end, urethral duplication cases over a 14-year period were reviewed. The classification system of urethral duplication is also discussed.

## Material and methods

Charts of patients diagnosed with urethral duplication were reviewed retrospectively. Ages at diagnosis, gender distribution and clinical presentation of the patients were analyzed.

The imaging studies used to ascertain anatomical details, the types of surgical correction and the follow-up are described.

## Results

Between 1997 and 2011, twelve patients, seven boys and five girls, with urethral duplication were treated (Table 1). Ages at diagnosis ranged from one day to one year and eleven months. The patients sought medical treatment either due to the diagnosis of genital anomaly after birth or due to mucous discharge from the accessory urethral orifice, double urinary stream, passing urine per anus along with every act of voiding, incontinence between normal voiding intervals, recurrent urinary tract infections. Duplication was an incidental finding during surgery in two patients.

All patients' evaluations included a voiding cystourethrogram to ascertain the anatomical details of the defect. In selected cases retrograde injection of contrast material into both channels in the anteroposterior and oblique position and urethrocystoscopy were done. Through those exams, we were able to establish the functional urethra, which may or may not have been in its orthotopic position, and the duplicated hypoplastic urethra. We also ascertained whether the duplicated urethra was in continuity with the functional urethra or with the bladder neck. In all patients in whom the duplicated urethra was in continuity with either the functional urethra or the bladder neck, the upper urinary tract was investigated by ultrasonography. In the patients with complex anomalies, either an intravenous pyelogram or magnetic resonance imaging (MRI) was also performed. Nine patients had urethral duplication in the sagittal plane and three patients in the coronal plane. Of the patients with sagittal urethral duplication, 3 had pre pubic sinus, 3 had epispadiac urethral duplication, 1 had a dorsal urethral duplication deviated from the midline and 2 had hypospadiac urethral duplication.

Three patients, one girl and two boys, had a blind ending duplicated urethra (pre pubic sinus). The duplicated urethra was located in the dorsal position in relation to the functional orthotopic urethra in all three patients. Although the patients sought treatment mainly for cosmetic reasons, they all had mucous discharge from the accessory orifice. In the boys, the accessory opening was located at the base of the penis (Fig. 1), and in the girl the accessory opening was located above the clitoris. All three patients underwent resection of the dorsal hypoplastic blind urethra (Fig. 2).

Three other patients, one girl and two boys, had a duplicated urethra in the dorsal position communicating with the lower urinary tract (epispadiac urethral duplication). Although none of them had associated classical bladder exstrophy, all showed characteristics of the bladder exstrophy—epispadias complex. The girl had pubic symphysis widening and a duplicated epispadiac urethra positioned above the bifid clitoris (Fig. 3); one of the boys had a bladder exstrophy variant with a duplicated bladder template, and the other was first diagnosed as having epispadias (Fig. 4). In these latter two patients, the urethral duplication was an incidental finding during surgery. The girl was diagnosed right after birth.

For the boy who was first diagnosed as having glandular epispadias, with mild dorsal penile curvature, the penis was degloved, and the dorsal duplicated urethra was dissected to below the pubic symphysis and completely resected near the bladder neck. This also corrected the mild dorsal penile curvature. The duplicated dorsal urethra in the girl was similarly dissected to below the pubic symphysis and completely resected near the bladder neck.

Except for the patient first diagnosed as having epispadias, these patients required another intervention to acquire continence. A Young-Dees-Leadbetter bladder neck reconstruction was performed in the boy, and in the girl a Mitchell bladder neck reconstruction. Both these patients had a wide open bladder neck identified during the procedure.

One patient had urethral duplication associated with a solitary kidney on the left side, with the ectopically implanted ureter and the ipsilateral deferent duct ending in a Mullerian remnant (Fig. 6). In this patient, the duplicated urethra was in the sagittal plane and dorsal to the orthotopic urethra, although it deviated from the midline (Fig. 5), running from the bladder neck to the glans. In addition to urine dribbling through the duplicated urethra, the patient complained of dysuria and straining. Excision of the hypoplastic urethra required a transpubic approach. The urethra was fully excised, and the left ectopic ureter was reimplanted. This patient's voiding dysfunction worsened in the postoperative period, and he required a transitory period of