Collateral Urethral Duplication in an Adult


Summary.- Objective: Congenital anomalies or malformations of the urethra that involve number are extremely rare and only 7 cases of trifurcation of the urethra and under 500 cases of urethral duplication have been described.

Methods: In the case we present, urethral duplication was diagnosed in adulthood, even though glans malformation with 2 urethral orifices was clearly evident and only 1 was functional.

Results: The condition is usually diagnosed in childhood due to the presence of 2 urethral meatus or from double stream if both are functional; at other times, the condition is diagnosed from complications that trigger infections or obstruction or if it is associated with more extensive malformations.

Conclusions: Urethral duplication is an extremely rare birth defect, and diagnosis in adulthood is even more uncommon. No single embryologic argument explains all the anatomic variants of urethral duplication. The treatment of the urethral duplication should be individualized according to the type of duplication and the clinical symptoms.

Keywords: Urethral duplication. Congenital malformations.

INTRODUCTION

Urethral duplication is a rare birth defect, and a PubMed search of the medical literature shows around 500 cases. The condition is seen primarily in males. The embryology, etiology, and pathogenesis of urethral duplication is not fully elucidated. It may be diagnosed at birth visually if there are two meatus–particularly if both are functional—or when the resulting symptoms or possible associated malformations are investigated. Treatment is
individualized and ranges from therapeutic abstention, as in our patient, to complicated surgeries.

CASE REPORT

A 48-year-old man, married and with 2 children, but with no relevant history reported 2 episodes of short-lasting voiding symptoms with fever in the past 2 years, consistent with episodes of acute prostatitis. The patient presented with symptoms of repeat prostatitis. The examination revealed 2 orifices in the glans in collateral position, which suggested the existence of 2 meatus, and a balanic morphology apparently corresponding to failed double glans development during the embryonic stage (Figure 1). The remainder of the physical examination, including the genital-perineal examination and digital rectal examination, were normal. When the patient was again asked about the clinical evidence, he confirmed that voiding and ejaculations were only present from the right meatus, although minimal secretion was occasionally observed from the left meatus. Neither the patient nor his partner found the abnormal anatomy unusual. During the visit, a Foley Ch 16 catheter was advanced through the right meatus up to the bladder without difficulty; the left meatus could only be catheterized with a Ch 6 catheter up to 5 or 6 cm.

The retrograde and voiding urethrography through the right meatus was normal. Retrograde urethrography combined with catheter in both orifices (Figure 2) showed a normal urethra for the right meatus, and a narrow, irregular path that appeared to end at the prostate or bladder base on the left, but was apparently not connected to the lumen of the right urethra or the bladder.

All other examinations, including intravenous urography, urosonography, and CT scans, were normal and no related malformation was observed.

The prostatitis was treated with antibiotics, and at the time of writing 4 years later, the patient was asymptomatic and had not required any other treatment.

DISCUSSION

The names used for this entity (urethral duplication or accessory urethra) are somewhat confusing because they include a wide variety of urethral malformations from the anatomic and clinical point of view, ranging from complete urethral duplication between bladder and glans to a short, narrow accessory urethral segment that is blind (2-4). Attempts have been made to systematize the description of the condition, mainly by two classifications proposed by Williams (1975) and Effman (1976). Williams based his classification on the hypospadiac or epispadiac termination of the duplicated urethral meatus and the plane of urethral separation. Effman uses the criteria for complete and incomplete duplication and for the urethral orifice sites (2-4). Most cases of urethral duplication are not equivalent, but can be included in 1 of these 2 categories. However, this is sometimes difficult, as in our case where 2 meatus are observed in a coronal view of the glans and a stenotic urethral channel appears to reach the prostate in a patient with no evidence of related malformations. The case could perhaps be included in Williams’s Type V abortive form (Table I).

Nevertheless, most authors essentially try to distinguish between urethral duplication and accessory urethra: the first term would refer to a urethra -according to the classification proposed by Ortolano5-(Figure 3) that originates in the urinary bladder, the neck of the urinary bladder, or the prostate and would be complete if it had its own external orifice, regardless of site (glans, penis body, or pelvic floor) and incomplete if it exits externally through the normal urethral channel. An accessory urethra originates in the anterior urethra and is complete if
it reaches the exterior or incomplete if the other end is blind. The term also refers to a channel with an external orifice, almost always ventral and blind that does not communicate with the urethra (4,5).

Urethral duplication is an extremely rare birth defect, and diagnosis in adulthood is even more uncommon (6-8). Although the current incidence is unknown, no more than 500 cases have been described (6). Most cases occur in boys; in girls it is even rarer and almost always seen in association with a double bladder. Moreover, urethral duplication involving diphallus or penile duplication (1 case per 5.5 million births) is extremely rare. The presence of urethral duplication may be associated with other malformations in the genitourinary tract (urethral valves, vesicoureteral reflux, renal agenesis, etc), gastrointestinal apparatus, or other organs.

No single embryologic argument explains all the anatomic variants of urethral duplication. Numerous hypotheses have been proposed, including intrauterine processes (e.g., ischemia, maternal hormonal abnormalities, environmental factors, therapeutic drugs) that would cause anomalies in the termination of the Müllerian ducts, defective growth of the urogenital sinus, etc. during organ differentiation. For the most common urethral duplication (dorsoventral or sagittal), the proposed explanations include abnormal continuity in the division process of the urorectal septum during bifurcation of the urethral groove, bifurcation of the urethral channel, delayed fusion of the genital tubercle, and partial failure in invagination of the lateral mesoderm between the endoderm and ectoderm layers of the cloacal membrane (2,4,7,8-12).

Collateral duplication, such as diphallus, could originate from the urogenital precursors or could arise when the lateral elements fuse to form midline structures and create a full organ; another explanation could be an error of septum creation, that would explain the high incidence of associated midline defects (3,7).

### Table I.

<table>
<thead>
<tr>
<th>Williams Classification</th>
<th>Effmann Classification</th>
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<tr>
<td><strong>Type I</strong></td>
<td><strong>Type I</strong></td>
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<tr>
<td>Epispadiac urethral duplication.</td>
<td>Incomplete urethral duplication.</td>
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<tr>
<td><strong>Complete:</strong> 2 channels independently arising from bladder.</td>
<td><strong>A. Distal:</strong> opens on dorsal or ventral penile surface, but does not communicate with urethra or bladder.</td>
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<td><strong>Incomplete:</strong> outlet of single channel from bladder, that divides distally.</td>
<td><strong>B. Proximal:</strong> arises from normal urethra and ends blindly in periurethral tissue.</td>
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<tr>
<td><strong>Abortive:</strong> blind penile sinus.</td>
<td><strong>Complete urethral duplication.</strong></td>
</tr>
<tr>
<td><strong>Type II</strong></td>
<td><strong>Type II</strong></td>
</tr>
<tr>
<td>Hypospadiac urethral duplication.</td>
<td><strong>A. With 2 meatus.</strong></td>
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<tr>
<td><strong>Complete:</strong> 2 noncommunicating channels from bladder until exit to exterior.</td>
<td>1. 2 noncommunicating urethras arising from bladder.</td>
</tr>
<tr>
<td><strong>Incomplete:</strong> urethra divides into 2 at level distal to bladder.</td>
<td>2. Duplicated segment arising from normal urethra.</td>
</tr>
<tr>
<td><strong>Abortive:</strong> blind sinus dorsal to urethra, with both hypospadiac meatus.</td>
<td><strong>B. With 1 meatus.</strong></td>
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<tr>
<td><strong>Type III</strong></td>
<td><strong>Type III</strong></td>
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<tr>
<td>Spindle urethra: urethra divides into 2 and unites into common channel distally.</td>
<td>Urethra duplication arising from bladder or posterior urethra and uniting into common channel distally.</td>
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<tr>
<td><strong>Type IV</strong></td>
<td><strong>Type III</strong></td>
</tr>
<tr>
<td>Y-duplication: with preanal or perineal accessory canal.</td>
<td>Urethral duplication as a component of partial or complete caudal duplication.</td>
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<tr>
<td><strong>Type V</strong></td>
<td><strong>Type III</strong></td>
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<tr>
<td>Collateral urethral duplication.</td>
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</table>
From the histologic point of view, urethral duplication can be defined as the juxtaposition of two channels comprised of smooth muscle walls lined with transitional epithelium, which excludes congenital urethral diverticula and urethral fistulae associated with an anorectal malformation (12). Most urethral duplications occur in the sagittal plane. Duplications in the coronal view are extremely rare and usually associated with midline malformations, double bladder, or complex caudal duplication (3,4,10). The embryologic explanation is just as unclear in this case.

The clinical manifestations vary considerably and will depend on the anatomic type of urethral malformation; the most common are double stream, obstructive symptoms, urinary infection, urinary incontinence, and penile curvature. Many cases are completely asymptomatic.

The condition is usually diagnosed in early childhood from symptoms or when an examination reveals 2 urethral meatus. Retrograde and voiding cystography is the most useful examination for diagnosing and classifying the condition. However, additional imaging studies (intravenous urography, sonography, etc) will complete the upper urinary tract examination, find any associated anomalies, and assess any obstruction accompanying duplication (11). The condition has been studied by combining urethrography with magnetic resonance

![Diagram](image-url)

**Figure 3.** A) complete urethral duplication. B) incomplete urethral duplication. C) variants of accessory urethra.
(MR) imaging which has advantages over conventional urethrography because the precise 3-dimensional topography of the anomalies and the related neighboring structures can be analyzed.

The treatment of the urethral duplication should be individualized according to the type of duplication and the clinical symptoms. In many patients, no treatment is required because there are few or no symptoms. Others (obstructions, incontinence, double stream in hypospadiac or epispidiac meatus) will require functional surgery or a cosmetic procedure and entail a wide variety of treatments (endoscopy, urethrourethrostomies, excision of the urethra duplication, complex urethroplasties, or urinary bypass). The true urethra with proper sphincter mechanism must be taken into consideration in the sagittal separation; it tends to be ventral and the verumontanum and external sphincter must be identified by cystourethroscopy before excision surgery is performed [7,10,11].

**CONCLUSIONS**

Urethral duplication is an extremely rare birth defect, and diagnosis in adulthood is even more uncommon. No single embryologic argument explains all the anatomic variants of urethral duplication. The clinical manifestations vary considerably and will depend on the anatomic type of urethral malformation. The treatment of the urethral duplication should be individualized according to the type of duplication and the clinical symptoms.

**REFERENCES AND RECOMMENDED READINGS**

*of special interest, **of outstanding interest*


