

Urethral duplication: a rare cause of recurrent urinary infection

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Abstract

Urethral duplication is quite a rare congenital anomaly with ill-defined aetiology. Patients often present with penile deformity, recurrent urinary infections, urinary incontinence, serosal discharge from the accessory urethra, and difficulty in urinating. Urethral duplication is most commonly grouped according to the Efmann classification. It has 3 main types as Type I, II, and III. There is no consensus on its therapy. There are non-surgical solutions, including follow-up without therapy, as well as many surgical options, including urethral reconstruction. Anatomical urethra and the external sphincter should absolutely be delineated when a surgery is contemplated. We herein report the case of a two-year-old male patient referred to our clinic with recurrent urinary infection, bilateral hydronephrosis and difficulty in urinating who was diagnosed with urethral duplication. We discuss our findings with review of the relevant literature.

Keywords: Urethral anomalies, Urethral duplication, Congenital.

Introduction

Urethral duplication (UD) or double urethra is quite a rare anomaly with ill-defined aetiology. Various theories have been put forward to explain its aetiology. Among them are abnormal closure of the Mullerian canal, incomplete mesodermal closure, ischaemic injury during embryogenesis, and abnormalities during development of urogenital sinus.¹ Patients often present with penile deformity, recurrent urinary infections, urinary incontinence, serosal discharge from the accessory urethra, and difficulty in urinating.²

We herein report a two-year-old male patient with recurrent urinary infection and difficulty in urinating who was diagnosed with urethral duplication during operation.

Case Report

A two-year-old male patient being followed up at the paediatric nephrology department was referred to our

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clinic with recurrent urinary infection, bilateral hydronephrosis and globus vesicalis. His genital examination was not remarkable, except for phimosis and globus vesicalis. Urinary bladder was emptied with an 8F feeding catheter and a urinary sample was sent for analysis. Laboratory examinations revealed a normal complete blood count (CBC) and creatinine level. Urinalysis showed positive leucocyte and nitrite. Escherichia Coli proliferation of 10⁵ COL/ML was observed in urinary culture. The patient was treated with antibiotics guided by antibiogram, and a urinary computerised tomography (CT) scan without contrast was ordered to evaluate the state of hydronephrosis and to evaluate the evidence of urinary stone. It showed dilatation in the collection system of both kidneys and ureters, with the right-sided dilatation being more pronounced. There was no evidence of urinary stone. It also revealed diffused thickening of urinary bladder wall. A cystoscopy was recommended. A trans-urethral cystoscopy examination was performed under general and caudal anaesthesia in October 2013. No urethral pathology was detected, but the bladder neck was high and there was an increased trabeculation in the bladder. Then, the preputium was separated from mucosa with the help of a clamp. During this intervention, a second meatus was observed at the sagittal axis, immediately above the urethral meatus (Figure-1A). A 0.35mm guide-wire was sent from the mouth of the sinus. The guide advanced through a lumen. The sinus was then catheterised with a 5F ureteral catheter. A retrograde urethrography was taken to visualise both urethral tracts. In urethrography, the contrast material passed into urinary bladder through both normal and accessory urethral tracts (Figure-1B). Upon these findings, the patient was diagnosed with complete UD, with both urethra opening into the same urinary bladder. Ten days after the operation a magnetic resonance imaging (MRI) was performed to better visualise the urogenital pathology. This examination visualised the passage of the contrast material along the dorsum of the penis when given from the accessory urethra (Figure-1C). The patient relatives were offered excision of the ectopic urethra. However, they declined it and, therefore, it was decided to put the patient on nitrofurantoin prophylaxis. The patient had 3 urinary tract infections with one-month intervals. Thus, the patient

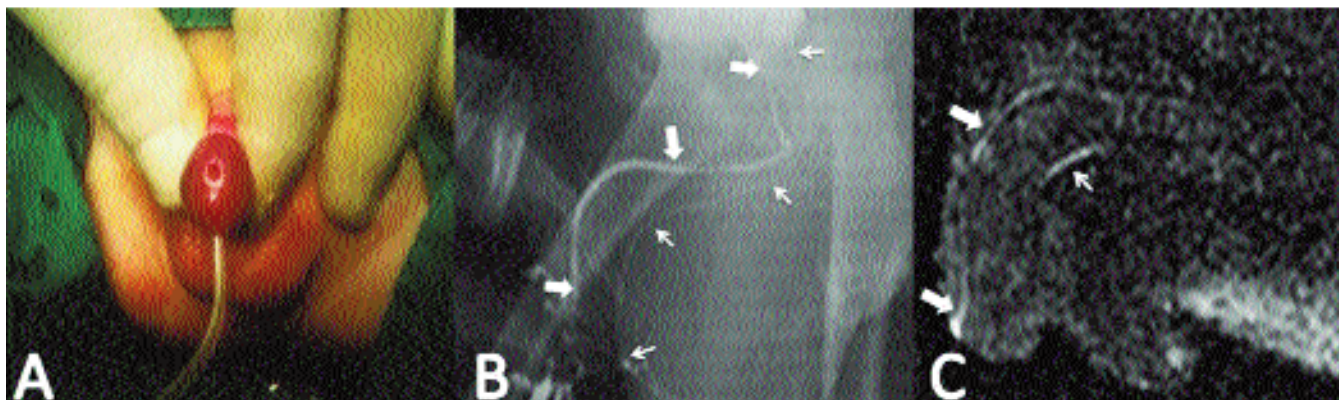
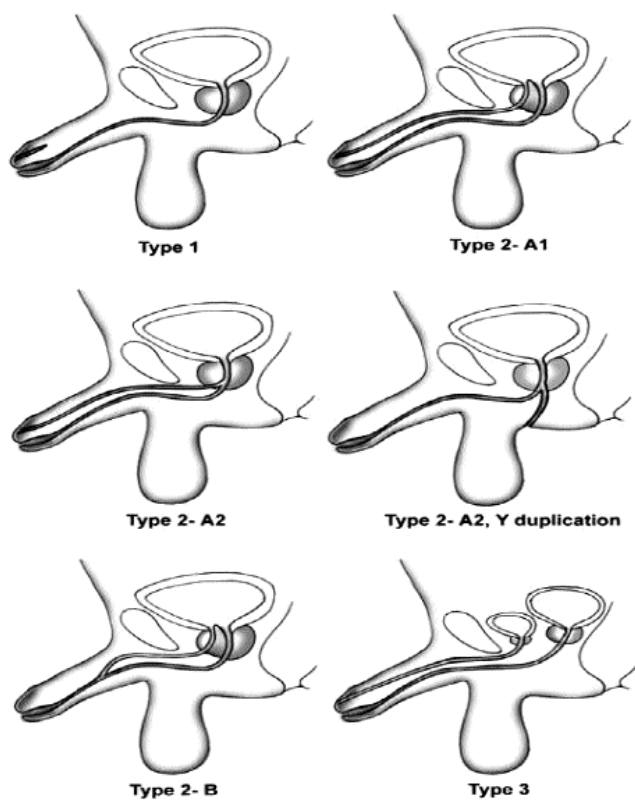


Figure-1: The anatomical urethra and the accessory urethra are seen on the photo (A). On sagittal urethrography imaging (B), the contrast agent is passing from both the accessory (thick white arrow) and normal (thin white arrow) urethra into the bladder. On sagittal fat saturated T2-weighted imaging (C), normal urethra (A-B, thin white arrow) opens anatomically, but the accessory urethra opens onto the dorsum of the glans penis (A, thick white arrow).



Type I: Blind-ending accessory urethra.
Type II: Completely patent accessory urethra.
IIA1: Two non-communicating urethra arising independently from the bladder
IIA2: A second channel arising independently into a second meatus (Y duplication)
IIB: Two urethra arising from the bladder or posterior urethra and uniting to form a common channel distally.
Type III: Accessory urethra arising from duplicated or septated bladders.

Figure-2: Illustration of Effmann classification.

relatives were re-offered surgery but they declined, and therefore, the patient underwent circumcision.

Discussion

UD is a rare urinary system anomaly in male and female children. So far, about 300 cases have been reported in literature.³

Various classification methods have been proposed for a better understanding of UD. Currently, the most commonly used classification system is the one proposed by Effmann.⁴ According to Effman classification, urethral duplications are divided into three main groups, as Type I, II, and III (Figure-2). Type IA is the most common type. Our case was considered as Effmann Type II A1.

This anomaly may be accompanied by various pathological conditions, including hypospadias, epispadias, anterior urethral valve, lacuna manga, prostatic urethral polyps, megalo-urethra, syringocele (dilated Cowper gland), and congenital urethral fistula.⁵ Detailed patient history and physical examination are important parts of the diagnostic process. Despite this, UD diagnosis can be made during circumcision and/or hypospadias surgery.

Imaging tests should definitely be used for confirming the diagnosis. The main diagnostic imaging procedures include voiding cystourethrography, intravenous pyelography, ultrasonography (USG), retrograde urethrography, and MRI. Intravenous pyelography allows visualisation of bony structures, renal agenesis, and upper urinary system.⁶ USG is helpful in differential diagnosis of complete duplication of urethra, and evaluation of large diverticles, ovarian cysts and surrounding soft tissues.⁷ A

prenatal diagnosis of urethral duplication is possible by evaluating foetal micturation with Doppler ultrasonography.⁸ Anatomical course, shape, diameter and relationship with urinary bladder of both urethra can be readily assessed with urethrography. UD is a complex anomaly and different manifestations probably have different embryological origins. MRI and CT scan help in diagnosing the accompanying genitourinary and gastrointestinal abnormalities (including solitary kidney, Mullerian anomalies, duplicated colons and double anuses etc).⁹ MRI provides excellent information on urethral width and length, and structure of periurethral soft tissues.¹⁰ By this way, other genitourinary and gastrointestinal anomalies can be detected and surgical planning may be better accomplished.

There is no consensus on the treatment of UD. Some authors recommend no therapy for asymptomatic cases.¹¹ However, incontinence, obstructive symptoms, double micturation, recurrent infection and cosmetic concerns are among indications for surgery.¹² Treatment options for UD include follow-up without a specific therapy, urethral dilatation, perineal urethrostomy, urethrourethrostomy, and surgical operations such as urethroplasty surgery using buccal mucosal or tubularized grafts.¹³ In cases where reconstructive surgery is planned, a cystoscopy should definitely be performed to assess functional urethra and sphincteric mechanism. With the help of imaging tests, the relationship of the accessory urethra with genitourinary system should be delineated and a treatment plan be based on the anatomical type of the duplication. Our patient could not be diagnosed despite cystoscopy. A physical examination is very important in the diagnosis of this age of children. In patients with serious phimosis, anaesthesia may be required to avoid pain and agitation while evaluating the accompanied anomalies. The accessory urethra could be visualised with genital examination and imaging modalities after peeling off the preputium from the surrounding tissues under anaesthesia. Our patient

underwent circumcision after recurrent bouts of urinary tract infections. However, the decision for circumcision should be reviewed in urethroplasty operations where the surgeon intends to use the preputium.

Conclusion

UDs are among rare causes of urinary retention and recurrent urinary infection. Patient's relatives should be exhaustively informed of the treatment plan. Urologists should keep in mind the fact that correct diagnosis may require anaesthesia during the physical examination of patients with phimosis.

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