Type IA urethral duplication: A case report

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ABSTRACT

Background: Urethral duplication is a rare congenital malformation mainly affecting men and boys. Although a number of theories have been proposed to describe this condition, the actual mechanism of this disorder is still not clear. The most frequent anomaly occurs in the sagittal plane, in which the duplicated urethra is in either dorsal or ventral position in relation to the orthotopic urethra. The therapeutic management of these conditions is complicated and depends on the presence of symptoms as well as the type of anomaly.

Case Description: We present a case of urethral duplication in a 2 years old male child. The malformation was characterised by the presence of the meatus in the dorsal penoscrotal and accompanied by the sign of infection in it. Urethrography showed contrast stopped filling 2 cm from the meatus in the dorsal penoscrotal with a total dead-end impression as an accessory urethra and there was no abnormalities in the principal urethra looked from the contrast filled the penile urethra, bulbous urethra, prostatic urethra, and bladder. That was the form of Effmann Type IA urethral duplication. The dorsal accessory urethra was excised entirely without complication and he remains symptom-free eight months after surgery.

Conclusion: In summary, urethral duplication is a rare clinical condition. It has many variants that were classified by Effman. Management depends on the anatomy of the duplication involved and the patient's clinical symptoms.

INTRODUCTION

The urethral duplication, also known as double urethra, is a rare anomaly occurring in pediatric cases, this deformity is most often found in male children while for female children it is very rare, with a previous literature recording no more than 40 cases of double urethra found in female children.1,2 The clinical entity comprised of a wide spectrum with various anatomical variations. Thus, there are several classifications used in defining each case of double urethra or urethral duplication, such as the Effmann classification system.3 It can be complete or incomplete, the majority of cases occur in the sagittal plane, further divided into dorsal and ventral duplication according to the position of the ectopic urethra. Collateral duplication without bladder duplication may also be found without accompanying bladder duplication. The relative rarity of the case combined with the wide spectrum of manifestation creates some difficulties in pinpointing the exact pathophysiological mechanism. Several literatures had suggested that there were multiple mechanisms that may cause urethral duplication, each possibly related with different types of manifestation.1

Individually tailored management is required due to the spectrum above of clinical manifestations in cases of urethral duplication.1,4 The general consensus is, presently, to treat only symptomatic patients. The treatment aims to preserve renal function, achieve continence, and achieve the best possible cosmetic outcome. The ventral part of the urethra is always the functioning urethra, complete with sphincters and the urinary bladder. Several methods may be used in obliterating the accessory channels, such as injection of sclerosing agent or other ablative interventions. The techniques, however, are slowly being abandoned due to the risk of corporeal thrombosis and fibrosis. Reconstruction by combining the ventral and accessory urethra parts may be utilized, although it carries a potential risk for meatal stenosis and obliteration of dorsal meatus. The current golden standard is surgical excision of dorsal accessory urethra. The procedure itself needs to be done delicately in order to avoid damage to the external sphincter and neurovascular bundle.1

CASE REPORT

A 2-years-old male patient was referred to urology polyclinic at Fatmawati General Hospital with the chief complaint there was whitish discharge from a hole in the dorsal penoscrotal. That condition has been realised by the parents since the last one month, but they have noticed the small hole in the dorsal penoscrotal since the patient was 3-months-old. Previously they took the patients
to a paediatrician because the patients were fussy when urinating and looked reddish around the hole then stated there was inflammation. There was no difficulty in urinating. Good emission of urine, not branched, and there was no urine comes out of a hole in the dorsal penoscrotal. There were no abnormalities in growth and development or other congenital abnormalities. From the physical examination we found a normal external urethral meatus on the tip of the glans penis (Figure 1a) and the second meatus with a diameter of 2 mm in the dorsal penoscrotal (Figure 1b). The two testicles were found normal.

During urethrography and urethroscopy examination, it was obtained a meatus from the dorsal penoscrotal as long as 2 cm, a pinpoint hole and debris appeared, and the wire could not be inserted. Contrast stopped filling 2 cm from the meatus in the skin, with a total dead-end impression (Figure 2a). 7 Fr ureteroscopy sheath was inserted through the external urethral meatus on the tip of the glans penis; urethra, verumontanum, bladder neck and bladder were found normal. There were no holes along the urethra; the right and left ureteral opening were identified. Contrast filled the penile urethra, bulbous urethra, prostatic urethra, and bladder. There was no stenosis and there were no tracts to another place (Figure 2b).

Patients had no other complaint besides a whitish discharge from a meatus in the dorsal penoscrotal which is then followed by the presence of pain and redness around it. The patient was then planned for exploration and excision of the tract from the meatus in the dorsal penoscrotal.

**Surgical Technique**

Patient was positioned under general anaesthesia. The aseptic and antiseptic procedure was conducted at surgical site and the surrounding area. Evaluation of genitalia external was carried out; the external urethral meatus was located at the tip of glans penis, two testicles were found normal, and there was a second meatus at the dorsal penoscrotal.

Mattress stitch on glans penis using suturing thread prolene 4-0. Foley catheter 8 Fr was inserted at the opening of external urethral meatus at the tip of glans penis. Circumglandular incision was made following marker and penile degloving was conducted, identification of the dorsal tract (Figure 3a). The dorsal tract appears to the symphysis (Figure 3b). Excision of the dorsal tract and released up to the symphysis, obliteration of the tissue appeared (Figure 4). The tissue was sent to be
examined into the anatomical pathology laboratory. Raw surface was the covered using the byars flap technique with PDS 5-0 interrupted (Figure 5).

RESULT
The results of examination from anatomical pathology of the tissue showed a picture of the urethra with a chronic inflammatory reaction and no signs of malignancy appeared. Ten days after surgery (Figure 6), patient came back to our urology polyclinic. Physical examination showed good surgical wound healing, there was no discharge from the surgical wound in the dorsal penoscrotal was found as before and there were no problems with voiding.

DISCUSSION
Double urethra or urethral duplication is a congenital disorder which rarely happens. Approximately 150 – 300 cases have been reported to date and is more frequently found in male children, with a previous literature recording no more than 40 cases of double urethra found in female children. This disorder is often associated with other abnormalities in the genitourinary tract, heart, bowel, and bones. Arena et al., found that in 60% of cases is accompanied by genitourinary malformation such as ureteropelvic junction obstruction, extra rotation of the penis, vesicoureteral reflux, renal ectopia, renal agenesis, or posterior urethral valves. There was no additional abnormality in our patient.

Various theories have been proposed to explain the embryological development of urethral duplication, including disorders of mesoderm development, abnormal Mullerian ducts, ischemia during embryogenesis, and defects in the development of the urogenital sinus. Casselman and Williams suggested that a partial failure or an irregularity of the ingrowth of the lateral mesoderm between the ectodermal and endodermal layers of the cloacal membrane in the midline accounts for the forms with a dorsal epispadias channel. Das and Brosman stated that abnormal termination of the Mullerian duct was responsible for the development of urethral duplication. Rica et al., reported that asymmetry in the closure of the urorectal septum results in an urethra-perineal fistula.

There were a few anatomical variations of urethral duplication. Duplication may occur at bladder neck area or only affecting the distal part of the urethra. This disorder commonly occurs on the sagittal section, with urethra on the dorsal part and ventral of penis. Ventral urethra is usually more functional as it has sphincter and verumontanum mechanisms. Urethral duplication rarely occurs on the coronal section. There are many classifications used to categorise urethral duplication. Gross and Moore classified urethral duplication as a complete second passage originating from the bladder to the dorsum of the penis and or as an accessory pathway which is unfunctional that ends blindly on the dorsal or ventral surface. Williams and Kenawi categorised urethral duplication based on the ectopic location of the urethra compared to normal urethra. However, the most commonly used and accepted in this area recently is the Effmann classification. Effman categorised urethral duplication into three types. Type I is composed of incomplete with blind-ending and/or accessory urethra, if the accessory urethra opens onto the dorsal or ventral surface of the penis without communicating with principal urethra or bladder it is classified as type IA and classified as type IB when the accessory urethra emerges from principal urethra and terminates blindly within the periurethral tissue. Type II duplication is complete patent urethral duplication with four subtypes, namely type IIA 1 when the second channel of the urethra arising independently from the bladder, type IIA 2 if the second channel arising from the first urethra and courses into a second meatus, when the second meatus located in the perineum it is classified as
type IIA “Y”, and type IIB with the second channel arising independently from the bladder but joining the first and coursing into one meatus. Type III is when urethral duplication as a component of partial or complete caudal duplication, usually associated with bladder duplication.16

This classification is more functional, representing all clinical types of urethral duplication.17 Eiffmann type IIA 2”Y” is the most common type reported of urethral duplication18–20 and usually associated with severe congenital anomalies like imperforate anus, cloacal atrophy, conjoined twins, early amnion rupture syndrome, prune belly syndrome and hand-foot genital syndrome.21 According to this classification, our patient belonged to type IA. Although this classification is considered to be the most exhaustive, it does not distinguish sagittal from coronal duplication and does not recognise female form. Lima et al.12 suggested a new classification, the proposed classification scheme is based on the required surgical approach, this classification is more complete and includes coronal and sagittal pieces and also divided between men and women.

Diagnosis of urethral duplication is based on simple physical examination of the penis and is confirmed by a voiding cystourethrography or retrograde urethrogram to identification the presence of urethral duplication and its anatomical type.22 Ultrasound may be helpful in the diagnosis of other associated anomalies.23,24 Upper urinary tract abnormalities are found in up to 80% of patients and are diagnosed with intravenous urography or ultrasonography.3 Clinical signs and symptoms of urethral duplication are diverse. Patients may complain of intermittent discharge from the accessory urethra having two emission sites of urine, incontinence, recurrent urinary tract infections, obstructive symptoms, or no symptoms at all.25,26 In our patient there were no complaints experienced before and the until there was sign of infection in the accessory urethra with sign and symptoms in the form of whitish discharge from meatus of accessory urethra.

The first step in the management of urethral duplication is to recognise the normal-functioning urethra; in fact, the true urethra is that with the largest calibre, a normal verumontanum, and an intact sphincter. Urethroscopy can be conducted to evaluate the diameter and the presence of verumontanum.23 Treatment of urethral duplication should be individualised based on the anatomic types and also clinical findings and severity of the accompanying anomaly. Many patients are asymptomatic and do not require any surgery. There is no clear consensus yet regarding the management of urethral duplication, some experts recommended that surgical management is unnecessary in asymptomatic patients. Surgical treatment is needed for reconstructing deformities, such as dorsal meatus, to fix the emission sites of urine, incontinence, recurrent urinary tract infections, obstructive symptoms as well as to mend urinary leakage on the perineal area.23,27 Surgical management can be conducted with various techniques, consisting of one step to two steps of surgery, with simple coagulation or reconstructive surgery, depending the type of urethral duplication. If the two urethral orifices are too close to each other on the glans, then the septum between the two meatus can be excised to give a single urinary meatus. Other than this, most procedures involve excision of the accessory urethra with reconstruction of the normal urethra.18,28 The simplified treatment scheme proposed by Salle et al. based on the type of urethral duplication from Eiffmann classification. In the cases of type IA urethral duplicationspectrum above they proposed meatoectomy or excresis of the accessory urethra. Urethro-urethrostomy can be done with or without urethroplasty for the type IIA 1 while for the type IIA 2 the treatment is urethroplasty. They proposed to excresis the non-functional bladder for type III urethral duplication in sagittal plane and excresis bladder septum or hemibladder plus reimplantation of the urethra in the contralateral bladder zone for the type III urethral duplication in coronal plane without diphallia and if accompanied by diphallia penoplasty and glans plasty is needed. No surgical intervention is needed for cases of urethral duplication type IB and IIB.29

In our case the accessory urethra was excised completely, we did that procedure in accordance with the treatment scheme proposed by Salle et al. because in this Type I A urethral duplication the accessory urethra was blind-ending in distal location and was not connected to the principal urethra or bladder. The outcome of this patient was very good.

CONCLUSION

Urethral duplication is generally an uncommon congenital abnormality in urological practice presenting in various types and some with associated other malformation, which the aetiology and mechanism are unknown. Signs and clinical symptoms of these disorders vary; hence management of the patients must be individualised. Surgical treatment is needed for reconstructing deformities. The patient presented with Type IA urethral duplication. He remains symptom-free eight months after complete excision of the accessory urethra.
CONFLICT OF INTEREST
The authors affirm no conflict of interest in this study.

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AUTHOR CONTRIBUTIONS
Y.I.H made assessment of the case, performed the operation, followed up the patient, gifted knowledge and supervised the case; M.A collected the data, followed up the patient and wrote the manuscript.

REFERENCES