ORIGINAL RESEARCH PAPER

General Surgery

KEY WORDS: urethral duplication, double stream of urine, complete type IIa1

COMPLETE DUPLICATION OF URETHRA: A RARE ANOMALY

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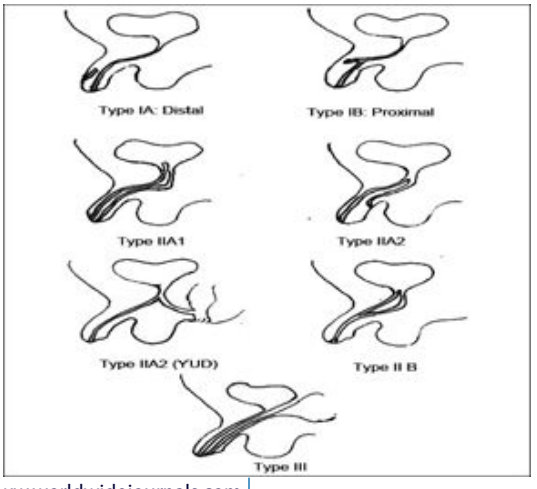
ABSTRACT

True duplication of male urethra whether in conjunction with single or double penis is a rare anomaly. Slightly less common are those accessory channels, by which their reasons of length are clearly distinguishable from small blind pits encountered on the glans penis, and around which controversy has raged as to whether be regarded as urethral homologues. Urethral duplication is a rare congenital anomaly, mainly affecting boys. Generally, the duplication develops on the saggital plane; the accessory urethra may run dorsally or ventrally to the orthotopic one. The embryology of this pathology is still controversial.
Diagnosis requires a high index of suspicion. Optimal knowledge of the duplication is significant for the managing surgeon in order to avoid complications. The overall outcome is good.

Introduction:
Urethral duplication is a rare congenital anomaly, with <200 cases reported worldwide; the condition is unique to males. Duplication of the urethra may be either in a sagittal (dorsal or ventral) or coronal plane relative to the orthotopic urethra. Urethras that duplicate coronally typically appear only with concomitant bladder duplication.

Several classifications have been proposed trying to distinguish between the different types of urethral duplication, and to define an appropriate plan of management. Based on the location of the external urinary meatii, Williams and Kenawi described epispadiac, hypospadiac, spindle, and collateral types. However, their classification has been criticized for lacking many of the anatomical details. A year later, Effmann and colleagues introduced their famous classification which was based on detailed radiological anatomy as seen in the patients urethrograms. The latter has been described as the most exhaustive classification and has gained widespread acceptance among radiologists and urologists as well.

Urethral duplication (UD) presents with varied clinical manifestations such as deformed penis, twin streams, urinary tract infection (UTI), urinary incontinence, serous discharge from sinus, outflow obstruction and associated anomalies. It may be complete or incomplete. The accessory urethra often presents as dorsal or ventral midline openings and rarely opens eccentrically (Coronal/collateral). Dorsally opening accessory urethra is the most common type.



Effmann's classification: Type IA: Incomplete distal UD Type IB: Incomplete proximal UD; Type IIA1 (Complete UD): Two-meatus non-communicating urethras arising independently from the bladder, Type IIA2 (Complete UD): Second channel arising from the first and exiting independently (including YUD), Type IIB: Complete UD bifurcate and rejoining at one meatus; Type III: UD as a component of caudal duplication.

Case report:
A 7 years old boy came to surgical OPD with chief complaint of thin stream of urine. There was no h/o burning micturition or trauma to penis. On examination, scrotum was normal and had fully descended testis. He had meatal stenosis for which he was being advised for meatotomy. Symphysis pubis was felt normal without any gap. His blood urea was 16mg/dl and creatinine was 0.7mg/dl. An ultrasonogram of abdomen revealed no genitourinary or any other anomaly. After meatotomy, patient symptoms were not relieved. It was noticed that patient was passing a very thin stream of urine. On opening there was a second meatal opening lateral and inferior to the urethra which had undergone meatotomy. Hence, suspicion of urethral duplication was made. On re-exploration, a very small stenotic urethral meatus was noticed lateral and inferior to main meatus, infant feeding tube number 5 could not be introduced in this meatus. Meatotomy of this meatus is done and catheter could freely go in the bladder when catheter was introduced in the earlier meatus it would not enter the bladder. Post operatively, the patient was passing urine with good stream through the second meatus. Post op dye study shows both urethral opening in the bladder. This confirmed type 2A1 urethral duplication.

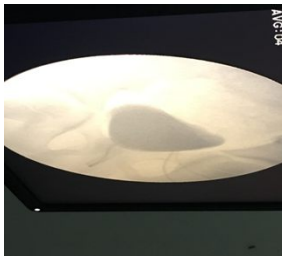


Figure 1: A dye study had been done showing two adjacent urethral opening going in continuity upto bladder.

Discussion:
Das and Brosman classified duplicated urethra into three types. Type I is a complete accessory urethra arising from a separate or confluent opening within the bladder and extending to an external orifice. Type II includes accessory urethras that arise from the

primary urethra and may or may not extend to a distal orifice. Type III arises from the bladder neck or prostatic urethra and opens onto the perineum. The main urethra may be atretic. Firlit classified duplication as a urethra that arises proximally from the bladder, bladder neck or duplicated bladder. Its distal course usually is dorsal to the main urethra. The complete form extends from the bladder to the glans. Most duplication occurs in the same sagittal plane, on top of the other. Less commonly, openings of UD lie collaterally (side by side) in the frontal plane. The classification of Williams and Kenawi includes epispadiac, hypospadiac, spindle and collateral types. According to Urakami et al, collateral urethral duplication in the frontal plane has been reported in only nine cases in the literature.

Embryonically, urethral duplication is not well understood and various hypotheses exist. Urethral duplication can be caused by the growth arrest of the urogenital sinus or abnormal Müllerian duct termination or misalignment of the termination of the cloacal membrane with genital tubercle. Depending on duplication type, patients may be asymptomatic. Symptoms include UTI, epididymitis, and incontinence. Diagnosis can be made using VCUG or RUG. Urodynamic study helps to confirm the position of the functional urethra to distinguish it from congenital urethroperineal fistula.

Radiological investigation includes voiding cystourethrography and retrograde urethrography (RUG) which should be carried out in lateral projections for visualization of the size, shape and position of the two channels. Effman et al showed that catheterization of a ventrally placed urethra was easier. IVU may demonstrate a wide symphysis pubis in case of epispadiac accessory urethra. Other associated anomalies are unilateral renal agenesis, ureteral duplication and a duplicated bladder. USG can demonstrate the exact length of any stricture segment or any extra luminal abnormal soft tissue or diverticulations. However, USG is user-dependent. The advantage is the lack of radiation exposure. MRI is an excellent investigation for the evaluation of duplicated urethras and the periurethral soft tissues. MRI can demonstrate with precision the sizes, shapes and positions of the two urethras as well as other associated genitourinary abnormalities. However, till recently MRI has only been used as an adjunct for the visualization of the urethra. Urethrocystoscopy is required in assessing the duplicated urethra especially when the imaging results are inconclusive. The danger in poor demonstration of tract is the possibility of inadequate and improper preparation for surgery leaving a residual tract or damage to adjoining tissues during surgery. Differential diagnoses include congenital anterior urethrocutneous fistula, post circumcision urethrocutneous fistula, lacuna magna (which is a problem especially in patients with hypospadias, when the lacuna lies distal to the meatus), urethral diverticulum and dilated Cowper's gland. Diagnosis of UD is based on genital examination and will be confirmed by VCUG or retrograde-urethrogram. Evaluation of the normal functioning of urethra is mandatory.

Indications for surgery include cosmesis, annoying symptoms such as a double stream, urinary incontinence, recurrent urinary tract infection, obstructive symptoms and associated genitourinary or other anomalies. Treatment of UD would depend on the type of urethral duplication and associated malformations. All efforts should be made to preserve the sphincter. The simplified treatment scheme proposed by Salle et al .

Treatment of urethral duplication should be individualized based on the anatomic variant and also clinical symptoms and severity of anomaly. Some patients do not require treatment if they are free from infection and incontinence. Simple accessory duplicated urethras may be fulgurated with a Bugbee™ (Gyrus ACMI, Southborough, MA) electrode and allowed to scar and close. Others need to be excised. If both urethra are functional and end adjacent to each other on the glans, then the septum between the two meatus can be excised to give a single urinary meatus. Injection of a sclerosing agent into the epispadic urethra has been described by Acimi and colleagues but it can cause fibrosis of the corpora cavernosa.

Conclusion:

Urethral duplication is a rare congenital anomaly presenting in various types and some with associated other malformations, and hence management of these patients must be individualized. The patient presented with type2A1 UD.

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