Y-Duplication of the Urethra: A Rare Case Report

A Kumar, N Goyal, S Trivedi, U Dwivedi, P Singh

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Abstract

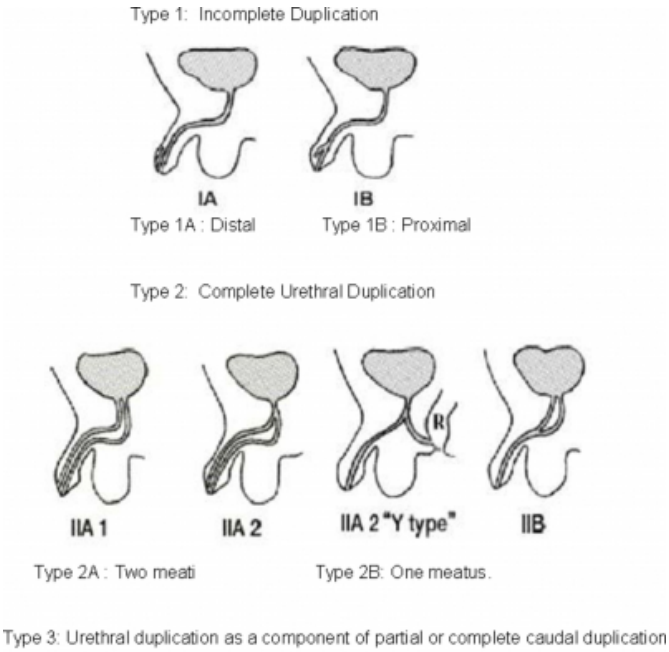
A 21-year-old male with complete urethral duplication presented with continent voiding per rectum and only thin stream of urine from the normal meatal site on straining. Patient had a normal external meatus and an ectopic urethral opening just inside the anal verge. Retrograde urethrogram confirmed Y-duplication of the urethra. He was managed successfully with a single stage urethroplasty under a covering colostomy. We report this case because of its extreme rarity.

INTRODUCTION

Urethral duplication is a rare congenital anomaly with a number of variable forms, probably representing different embryological defects. This anomaly is unique to males with few cases reported in females.[1] Most duplicate urethras are in the sagittal plane, occasionally they lay in the coronal plane (side by side) relative to the orthotopic urethra.[2] There are several classification schemes described for urethral duplication. Firlit[3] classified duplication as a urethra that arises proximally from the bladder, bladder neck or duplicated bladder. Its distal course is usually dorsal to the main urethra. The complete form extends from the bladder to the glans. The classification of Williams & Kenawi[4] includes epispadic, hypospadic, spindle and collateral type. The most widely used classification was established by Effman et al. (Figure-1).[5] This classification is functional and represents all clinical aspects of urethral duplication; however, it does not distinguish sagittal from coronal collateral duplication.

Figure 1

Figure 1: Effman's Classification



The Y subtype represents 6-30% of all urethral duplications.[6] The ventral or perineal urethra in this duplication is generally felt to be the most functional and contains the sphincter mechanism as well as verumontanum. The orthotopic urethra is typically poorly developed.[7] Some classify a subtype with larger orthotopic urethra and minute perineal branches as congenital perineal urethro-cutaneous fistulas.[8] Wagner et al.[7], however, classified all congenital

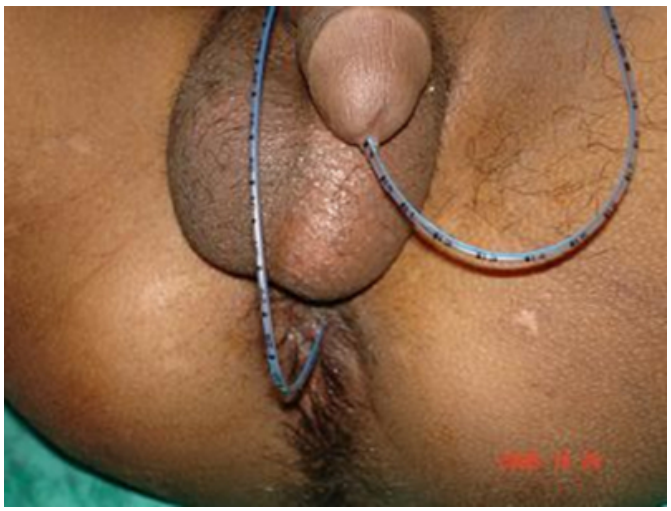
urethro-perineal fistulas as urethral duplications with a hypoplastic ventral urethra as they showed that these tracts, lined by transitional epithelium, occurred in the absence of infection or the predisposing condition for forming a fistula. We present a case of Y-urethral duplication and reviewed the literature.

CASE REPORT

A 21-year-old male presented with complaint of voiding per rectum since birth. He was passing a good amount of urine via the posterior tract but a thin stream of urine from the normal meatal site on straining. Physical examination revealed a well nourished adult with normal external genitalia. An ectopic urethral opening could be seen just inside the anal verge at the 12 o'clock position (Figure-2).

Figure 2

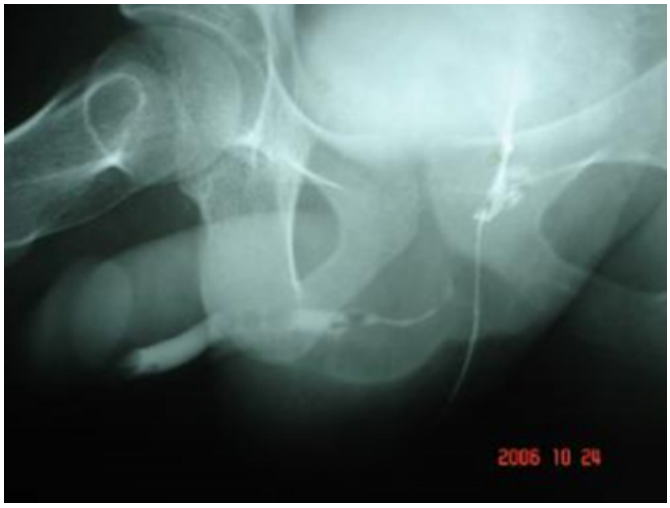
Figure 2: Accessory urethral meatus just proximal to the anal verge.



Blood chemistry was normal. Excretory urography revealed bilateral normally excreting kidneys. Retrograde urethrogram (Figure-3) & voiding cysto-urethrogram revealed a normally placed penile urethra with a stricture at the bulbar urethra and an accessory limb opening into the anal canal.

Figure 3

Figure 3: Retrograde urethrogram showing accessory perianal tract and stricture at the bulbar segment of the orthotopic urethra.



The patient was managed with single stage operation through a transverse (smiling) incision in the perineum. The accessory perianal branch was detached from the anal verge and mobilized towards the orthotopic urethra. The orthotopic urethra was mobilized down to the glans. The stricturous bulbar segment of the orthotopic urethra was excised and its proximal end was closed by oversewing with 3-0 chromic catgut. A neo-urethra was created with the accessory pre-anal tract and the penile urethra with a native tubed pedicled local skin flap (perineal skin) filling the gap between the two ends. This was done under covering sigmoid colostomy. The postoperative period was uneventful and the urethral catheter was removed after three weeks. The patient voided well with good stream.

DISCUSSION

Urethra duplication is a rare congenital anomaly with less than 200 cases reported.[6] Embryology of this condition is unclear. Casselman & Williams[9] stated 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 form with a dorsal epispadiac channel. Das & Brosmas[10] reported that abnormal termination of the mullarian duct was responsible for urethral duplication. Rica et al.[11] suggested that asymmetry in the closure of the urorectal septum results in an urethroperineal fistula. In spite of the numerous theories proposed to explain this anomaly, no single theory explains all the various types of anomalies.

As compared to the other forms of urethral duplications, Y-

type duplications are often associated with other severe congenital anomalies like imperforate anus, cloacal exstrophy, conjoined twins, early amnion rupture syndrome, prune belly syndrome and hand-foot-genital syndrome.[2]

The clinical presentation may vary. There should be a high index of suspicion in patients with anorectal anomalies. Signs & symptoms of presentation, aside from the observation of passing the urine from the perineum, include UTI, epididymitis and cystitis. Patients may present with urinary tract obstruction and in severe cases progress to chronic renal insufficiency.[2]

A physical examination is of paramount importance in the diagnosis of these children. The children's urinary stream must be carefully assessed to determine its location and character. Furthermore, careful observation for the presence of dribbling from the orthotopic or duplicated urethra is crucial.

Micturating cystourethrogram with retrograde urethrogram is the investigation of choice for confirmation of diagnosis. The upper tracts must be assessed as 80% of patients are associated with upper urinary tract abnormalities.[2]Occasionally, cystoscopy is necessary to visualize the verumontanum and other urethral characteristics. An acceptable repair of Y-type urethral duplication may be considered one of the most challenging reconstructive procedures on the lower urinary tract. After evaluating and identifying the Y-type duplication, the best urethra is that with the largest caliber, around the verumontanum with an intact sphincter. Several surgical techniques have been described to treat urethral duplication. Some prefer to use the apical urethra even when it is hypoplastic. Williams & Bloomberg proposed urethroplasty using scrotal flaps.[12] When no foreskin is available for repair, buccal mucosa is a reasonably good option for urethroplasty.[6] In a few cases, others described anastomosis of the ventral urethra to the dorsal urethra when the latter had adequate caliber.[12]In patients of Y-type duplication with a confirmed normal orthotopic urethra and a small perineal tract, perineal tract excision alone can be successful, taking care of the external sphincter and the neurovascular bundle.[7,8]

Knowledge of urethral duplication is important with respect to surgical procedure. Management of sagittal Y-urethral duplication involves complex staged urethroplasty but can be repaired in single stage by an experienced surgeon. Overall, the outcome is favorable in the majority of cases and urinary continence as well as excellent cosmesis is achieved.

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Author Information

Abhay Kumar, M.S.
Department of Urology, Institute of Medical Sciences, Banaras Hindu University

Neeraj Kumar Goyal, M.S.
Department of Urology, Institute of Medical Sciences, Banaras Hindu University

S. Trivedi, M.Ch.
Department of Urology, Institute of Medical Sciences, Banaras Hindu University

U. S. Dwivedi, M.Ch.
Department of Urology, Institute of Medical Sciences, Banaras Hindu University

P. B. Singh, M.Ch.
Department of Urology, Institute of Medical Sciences, Banaras Hindu University