Urethral duplication with rectourethral fistula: Review of two cases

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INTRODUCTION

Urethral duplication is a rare congenital abnormality of genitourinary tract with frequent occurrence in males.¹² Among several forms of urethral duplications, the prevalence of Y-type duplication counts to only 6%–30%.¹³ According to a classification system proposed by Effman et al., which helps in determining all clinical aspects of urethral duplication, the Y-type duplication is a special form of Type II A2 urethral duplication. It has two variants, the most common form is characterized by a stenotic orthotopic (dorsal) urethra, and a functional accessory urethra (ventral), which opens into the perineum, anal canal, or rectum, while the unusual form has the dorsal urethra functional and the ventral one hypoplastic.⁴⁵

We present two cases of Y-type urethral duplication with one case showing unusual clinical features and experience of their management.

CASE REPORTS

Two cases with rectourethral fistula, that is, Type II A2 Y-type urethral duplication are presented in this case report.
Case 1

An 11-year-old boy presented with a complaint of voiding urine per rectum since birth with no urethral voiding. After a physical examination, external urethral meatus was found at normal site with bifid scrotum and dorsal hypoplastic urethra, even a 5Fr ureteric catheter was not negotiable proximal to the glans. Investigations showed normal blood chemistry, ultrasonography (USG), and excretory urography (EU). Magnetic resonance urogram demonstrated T2-weighted axial section and multiplanar reconstruction showed urethrorectal fistula. Micturating cystourethrogram (MCU) showed a large capacity bladder. Retrograde urethrogram (RGU) was not possible due to aplastic dorsal urethra. Antegrade cystourethroscopy showed single bladder neck, normal verumontanum, and the posterior urethra coursed just distal to the verumontanum as flushed saline came out from the anus. It was diagnosed as Type II A of Y-type urethral duplication; with dorsal urethra being completely hypoplastic, except glanular urethra, and the ventral urethra was communicating with the rectum. Suprapubic cystostomy was done and the bladder was inspected. Hey Groove dilator was introduced from the bladder neck which came out from the inside of the anal canal. The patent ventral urethra was mobilized from rectum to its proximal part up to the membranous urethra, and then an inner preputial pedicled tube was used for end-to-end anastomosis with mobilized part of the ventral urethra. Thus, a neourethra was brought out from perineum to penoscrotal junction. The rectum was closed in two layers [Figure 1]. Postoperatively, the neourethra got fibrosed and obliterated after 1 month. Thus, a perineoscrotal flap urethroplasty was done. The patient is present with stabilized stoma and waiting for the 2nd stage urethroplasty.

Case 2

A 14-year-old boy presented with a complaint of voiding urine per rectum since birth with no urethral voiding. Physical examination revealed the normal position of external urethral meatus, and urethral catheter could be coursed up to 12 cm. On investigation, the blood biochemistry was within normal range, and renal USG revealed the right kidney to be congenitally hypoplastic. EU revealed a normally excreting left kidney with nonvisualization of the right kidney. MCU revealed a large capacity bladder with delineation of small part of posterior urethra. Urethrocytoscopy revealed normal caliber anterior urethra up to mid bulb part, but proximal to this, there was a complete closure of urethral opening. Antegrade cystourethroscopy showed a single bladder neck with normal verumontanum and posterior urethra. On evaluation, he was diagnosed as Type II A2, Y-type duplication of urethra with hypoplastic dorsal urethra at proximal part and normal caliber of penile urethra. The ventral urethra was communicating with rectum. The ventral urethra was mobilized from the rectum and anastomosed to the patent distal part of dorsal urethra in an end-to-end fashion. Rectum was closed into two layers [Figure 2]. The patient voided urine well through urethra with a good stream, and no further intervention was required.

DISCUSSION

Many theories are proposed to explain the various types of urethral duplication anomalies, but none of them cover all its variants. Probably, the mechanism for embryologic development of Y-type duplication could be, the faulty closure of the urorectal membrane, diminished growth of
the dorsoinferior wall of the urogenital sinus, or a fistula formation in the dorsal margin of the urogenital sinus due to a vascular accident.[6]

Our first case is quite novel as it does not fit into any of the proposed classification systems. Even its embryological mechanism is difficult to explain. The Y-type duplications are often accompanied with other congenital anomalies related to the genitourinary or gastrointestinal system.[7] Even in this report, Case 1 had bifid scrotum; and in Case 2, there was a hypoplastic right kidney.

MCU with RGU is the investigation of choice for confirmation of diagnosis, but in our patients, RGU was not possible due to hypoplastic dorsal urethra and even MCU was also difficult due to delineated urethrorectal fistula with over distended bladder. Cystoscopy is necessary to visualize the verumontanum and other urethral characteristics.

A complete repair of Y-type urethral duplication is definitely one of the most challenging reconstructive procedures of the genitourinary tract. On evaluation, the urethra with a larger caliber and an intact sphincter around the verumontanum was considered worth preserving. In cases where there is not enough foreskin for reconstruction, buccal mucosa is the choice for urethroplasty.[3] Whereas another intervention for urethroplasty was using scrotal flaps as suggested by Williams and Bloomberg. If the caliber of the dorsal urethra is adequate (24–26F), it can be anastomosed with the ventral urethra.[8] In patients with Y-type urethral duplication, if the caliber and quality of the orthotopic urethra is adequate, and the perineal tract is small, perineal tract excision alone can be used for reconstruction.[9,10] In the above-discussed cases, the second case was treated with end-to-end urethroplasty, and good surgical outcome was observed. However, for the treatment in the first case, perineoscrotal flap urethroplasty was done after the initial failure of inner preputial flap surgery. No complication or complaints were seen; and at present, the patient is waiting for the 2nd stage of surgery.

CONCLUSION

The Y-type urethral duplication is a complex condition to manage as it depends on the anatomy of the anomaly. Single-stage repair is feasible if the dorsal distal urethra is well developed, but in case of atretic distal urethra with limited local tissues for reconstruction, two-stage urethroplasty using inner preputial flap or buccal mucosa is required. Overall, the outcome is favorable in both the cases. Although staging of the management seems to be time-consuming, good results can be achieved without any complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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