



Y-type urethral duplication: A case series of four cases with good outcome

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ABSTRACT

Urethral duplication is a rare congenital condition. This case series intends to strengthen the concept of Y-type urethral duplication (new anatomic variation of an accessory limb of duplicated urethra). We have conducted a retrospective analysis collecting information from four cases of type IIA-2Y variety of urethral duplication (Effmann classification), which is one of rarely found subtype with its embryogenesis, diagnosis and single stage repair with outcome. Four babies around 1- 6 months of age range (mean- 3.2 months), complaining of passing urine from a site other than penis which includes anal canal, perineal area. All babies were investigated for other associated anomaly and were planned for surgery. Three out of four cases had undergone excision of accessory urethra and urethral stent in ventral urethra in postoperative period for 8-10 days with good results. In one case, posterior sagittal anorectoplasty (PSARP) approach was used to delineate accessory urethra and end to end urethral anastomosis was done with better outcome. All babies are on regular follow up till now ranging 6months to three years. It is important to delineate the functional urethra before planning for intervention so as to address the problem.

Key Words: Urethral duplication, Y-type urethra, orthotopic, heterotopic urethra.

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Introduction

Y type urethral duplication is one of rarely found entity among all cases of urethral duplications. Different authors have named this anomaly as λ-type, H-type or type IIA 2 Y (Effmann et al classification special type) duplication indicating the confusing aspect of

this type of urethral duplication [1-3]. We would continue to denote it as Y type urethral duplication as it is commonly found in searched English literature. It is characterized by the presence of two limbs-penile limb (stenotic/patent urethral channel) and an ectopic limb (fistulous tract- rectal, anal and perineal).

A similar condition has been described in literature as a type in the female in which there is a congenital fistulous connection between the vestibule and the anal orifice (“H-type” anovestibular fistula) [4]. The embryologic

explanations for the etiology of duplications can be summarized as incomplete mesodermal union, abnormal Mullerian structures, and ischemic events in the embryogenesis and defects in the development of urogenital sinus [5,6]. Another proposed theory of Y duplications is impaired growth of the dorso inferior wall of the urogenital sinus [3,4].

In this article, we try to elaborate the nature of this rare condition by studying four cases which were managed over the past years, with a detailed analysis of their clinical and radiological picture.

Case reports

We retrospectively reviewed four cases of Y type urethral duplication. All available hospital records were evaluated for presentation, investigation, management and final outcome. One case of four cases was a 2-month-old man, and parents complained of passing urine from anal opening since birth without per urethra urination. Bowel habit was normal with good continence with no symptoms of urinary tract infection (UTI). Normal scrotal & penile anatomy. Hernial orifices were normal. Normal opening of urethral meatus with feeding tube 8 Fr negotiable to some extent (about 3cm). Both testes descended. On digital rectal examination, mild soddening of perianal skin without fissure in ano. Sphincter tone felt to be normal. All walls normal on digital palpation. Mildly fecal stained fluid seepage from anal canal to outside on removal of finger.

Ultrasound of urogenital system appeared normal with renal function test in normal limit. Preoperative voiding cystourethrogram (VCUG) couldn't be done due to failure of catheterization prior procedure. Under proper sedation, an opening could be cannulated in

anterior rectal wall with urine flow from it (Stenotic variety).



Fig. 1. A 2 months old baby with urethral duplication (stenotic penile limb and an ectopic perineal patent urethral channel).

Similarly rest three cases were addressed in outpatient department (OPD) by complaint of passing urine from both normal meatus and from a site in perineum or from anal canal in different proportion pertaining to continence and adequacy of urinary stream. All cases were posted for operation room (OR) after thorough workup and VCUG preoperatively if possible. Four male babies were included in the study with urethral Y-duplication from last 5 years medical record who underwent our evaluation. Their age at presentation ranged from 1 to 6 months (mean - 3.2 months). One baby had history of cutback anoplasty on day 2 of life. The baby of 4 month age had vesicoureteral reflux grade I. Rest two were having only urethral duplication as etiology. All three cases with double stream were planned for VCUG which delineated both tracts in one case and rest two case were having a normally delineated orthotopic urethra in voiding phase. Heterotopic urethra were latter cannulated and contrast pictures were taken. A characteristic posterior kinking in the urethral wall at the site

of origin of the accessory channel could be seen in lateral view films (Fig. 2).



Fig. 2. A contrast in lateral view films shows a characteristic posterior kinking in the urethral wall at the site of origin of the accessory channel.

This kink may reflect the presence of tension across urethroanal channel which was also noticed during the excision of the accessory channel.

A single-staged procedure was performed in all four patients, in stenotic variety, rectum was mobilized in PSARP fashion, and anterior rectal wall mobilization resulted in delineating the heterotopic urethra which was mobilized extending up to perineum (Fig. 3). Baby position was changed to lithotomy position with midline incision and mobilization of nonfunctional but patent distally orthotopic urethra and end to end urethral anastomosis over 10 Fr urethral stent by using 6-0 PGA suture (Fig. 4). 10 Fr stent was removed on post-op day 21 with baby voiding with a good stream and post-op VCUG was normal (Fig. 5). Urethral calibration was done and result was excellent.

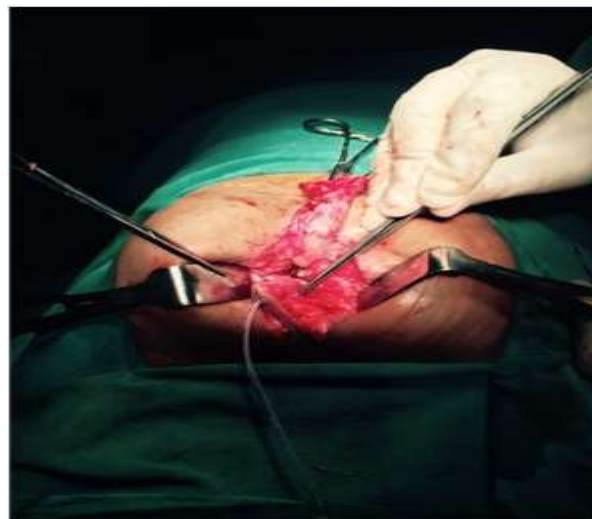


Fig. 3. Mobilization of rectum and heterotopic urethra in PSARP fashion.

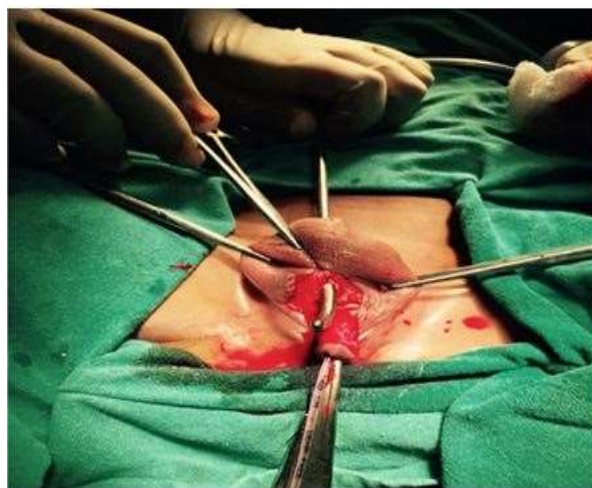


Fig. 4. Mobilization of nonfunctional but patent distally orthotopic urethra.



Fig. 5. Post-op day 21 with baby voiding with a good stream.

For rest three cases, babies were positioned in lithotomy position. Heterotopic urethra was cannulated with 7 or 8 Fr infant feeding tube (IFT) and orthotopic urethra was catheterized with 10 Fr IFT (Fig. 6). The fistula tract was circumscribed at the skin and then carefully dissected sharp and blunt mode outside the anal sphincter muscle complex up to the entry into the posterior urethra / prostatic urethra with easily palpable feeding tube for guidance (Fig. 7). The fistula tract was ligated near to the prostatic urethra taking care not to damage the prostate or urethra proper. Urethral stent was removed on post-operative day 10 and discharged.

On follow-up, all babies were doing well with good voiding stream. In one of three case, progressive dilatation of anterior urethra was done followed by excision of accessory urethra. All cases were calibrated with feeding tube on follow up.



Fig. 6. Heterotopic urethra was cannulated with 7 infant feeding tube and orthotopic urethra was catheterized with 10 Fr infant feeding tube.



Fig. 7. The orthotopic urethra carefully dissected up to the entry into the posterior urethra.

Discussion

Urethral duplications are rare pathology of the lower urogenital tract with a most around 300 cases reported in literature [7]. Effmann et al in 1976 classified urethral duplication which is extensively described in various article, so not described here in detail. All four cases were included in type IIA2 Y. It can be further divided into three subcategories in terms of orthotopic urethral patency, as described by Lima et al. These are: pure, steno atretic and abortive form. One of the case, stenotic variety in our study can be considered as steno-atretic form (25%) and rest three are pure form (75%) as compared to only 43% pure form in their study [2].

In 1996, Wagner showed the presence of transitional epithelium in accessory urethra as an element to define it [6]. Excised specimen in our cases shows both transitional epithelium and smooth muscles confirming it to be accessory urethra. Mane et al in 2009, [7] stated that out of total 8 cases of Y type

duplication, they have achieved correction only in 3 cases as single stage procedure (37.5%). In our study we have tried as single stage in all four cases with good results till now but final outcome depend on long term follow up. Moreover stenotic variety in our study could be possibly managed in single go due to distal patency of penile urethra and possible end to end anastomosis avoiding staged urethroplasty or use of buccal mucosa bladder mucosa free graft.

Ortolano and Nasrallah reported [8] a case of hypospadiac urethral duplication that was successfully treated with urethral dilation to a final calibre of 14 Fr on the ectopic ventral urethra, which is commonly more functional and anastomosed end to side to orthotopic urethra. We have adopted PADUA (progressive augmentation by dilatation urethra anterior) [2] for one case so as to dilate anterior urethra followed by just excising the ectopic urethra with good results.

Till now with continued follow up ranging 6 months to 3 years, our cases seems to have no complaint of poor stream, stasis as compared to 37.5% cases of stricture in study by Mane et al. [7].

Urethral duplication in which the ventral. Heterotopic opening into the perineum branches from the posterior prostatic urethra and is actually the smaller of the two channels while the orthotopic dorsal urethra is the larger and more functional channel. The most common, current terminology for this abnormality however, remains congenital posterior urethroperineal fistula (CUPF)" which is not described in original Effmann's classification. Three of our cases can be considered in this variety of CUPF. Less than 30 cases are described in literature [9,10], 28th reported case was by Meier DE, Latiff A in 2016 [9].

Lopes et al. [11] studies thirteen boys with urethral duplication and concluded that patients with incomplete duplication (type I A or IB) can totally be asymptomatic, with no need of surgical correction. Type IIA2 is the most complex form of duplication to correct and multiple procedures might be required because of the very hypoplastic orthotopic dorsal urethral tissue [12].

Vesicoureteral reflux, renal dysplasia, pyelo-ureteral duplicity, bladder wall thickening obstructive megaureter, multicystic kidney are associated pathology as described by Lima et al in their series [2], we have found no associated anomaly but more number of cases or more period of follow-up is necessary to affirm association with other anomalies.

In our practice, we believe that it is important to identify and retain the functional urethra [13, 14]. It is a mistake to discard the Heterotopic urethra (when functional) preserving the hypoplastic branch in the normal position or vice versa. The identification of the functional urethra has important surgical implications. This delicate concept was well enlightened by Stephens and Donnellan who recommended the use of the functional urethra in order to avoid sphincter damage and use of a dysgenetic branch [15].

Conclusion

The Y-duplication is a rare and special type of urethral duplication with peculiar anatomy, embryology and thus treatment options. The vital step in patient assessment is delineation of the functional urethra and associated anomalies, if any. The surgical correction can be successful in cases as a single stage. However, there are patients with complex associated anomalies, needing appropriate graft procedure for urethral reconstruction in a staged manner. In addition, surgical urologic

experience is critical, being the majority of the procedures delicate and complex. All these elements affect the long-term outcome. Counselling of parents about detailed plan of action with expected results and complication, need of multiple procedure for satisfactory aesthetic and functional results is of paramount importance.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Informed and written consent was taken from patient and her parents to publish this case report.

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