

Y-type Urethral Duplication with Rectal Implantation of the Urethra in a Boy with Long-gap Esophageal Atresia: A Case Report

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ABSTRACT

Urethral duplication is a rare congenital anomaly of the lower urinary tract which includes a wide range of anatomical variations. Y-type urethral duplication, which represents 6–30% of all forms, describes the condition, in which a functional urethra is implanted in the rectum with dysplastic topic urethra within penis. We report a case of premature newborn boy with multiple congenital anomalies (long-gap esophageal atresia with tracheoesophageal fistula, atrial and ventricular septal defect) and II A2 Y-type of urethral duplication. Diagnosis and management as well as review of literature on this subject are presented.

Key words: Esophageal atresia, lower urinary tract anomalies, urethral duplication

INTRODUCTION

Urethral duplication is a rare congenital anomaly of the lower urinary tract characterized by the presence of two urethral channels with a wide range of anatomical variants. There are published only a limited number of case series in literature and about 300 cases are reported.^[1-10]

Effmann's classification of duplication of the urethra was published in 1976^[10] and is still the most widely used [Figure 1]. The Y-subtype represents 6–30% of all urethral duplications.^[11-14] A special form of Y-type duplication represents urethral duplication with rectal or perineal implantation of the urethra (type IIA2 according to Effmann's classification). Usually, single case reports and few case series were published.^[11-15]

We report a case of newborn male with multiple congenital anomalies and Y-type II a urethral duplication with rectal implantation.

- Type IA: Incomplete distal urethral duplication
- Type IB: Incomplete proximal urethral duplication
- Type IIA1: Complete urethral duplication (two non-communicating urethras arising independently from the bladder)
- Type IIA2: Complete urethral duplication (the second channel arises from the first and courses independently into a second meatus)
- Type IIA2 (YUD): Complete urethral duplication (the second channel arises from the first and opens within perineum or rectum)
- Type IIB: Complete urethral duplication with bifurcation and one meatus
- Type III: Urethral duplication as a component of partial or complete caudal duplication.

CASE REPORT

Premature newborn boy (34 Hbd) with multiple congenital anomalies including esophageal atresia, atrial and ventricular septal defect, and costal anomalies was admitted to our department. During esophageal surgery

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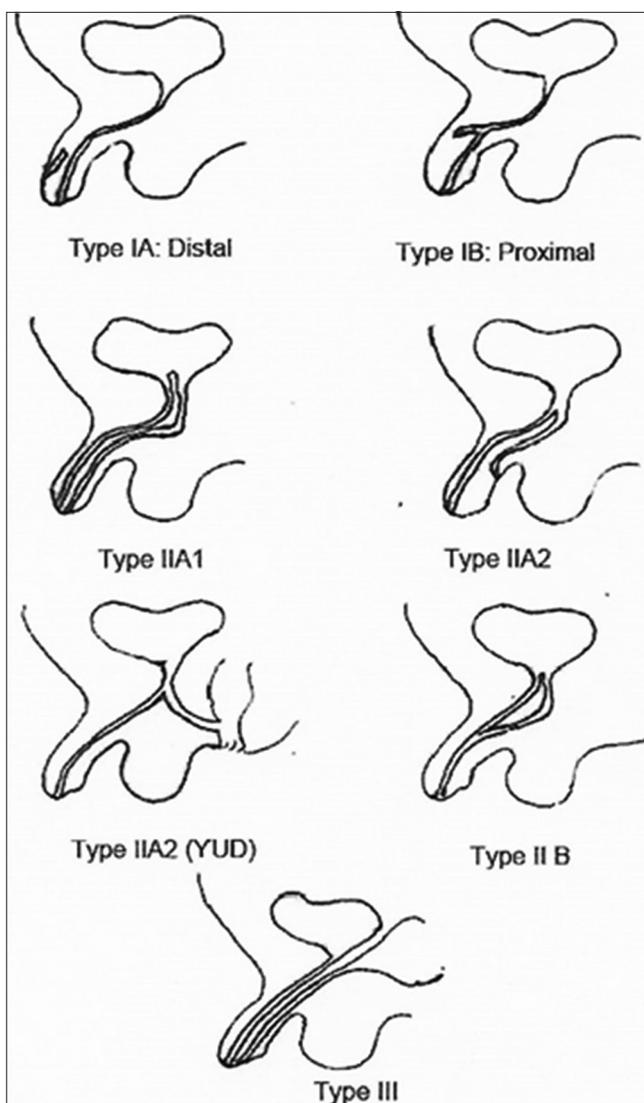


Figure 1: Urethral duplication according to Effmann's classification

intraoperatively, long-gap esophageal atresia with lower tracheoesophageal fistula was confirmed, and after ligation of fistula, gastrostomy was done as the first step of operative treatment. Before the operation, the boy could not be catheterized despite few attempts – on physical examination, small penis with normally placed external urethral meatus was present. Suprapubic cystostomy (Cystofix) was placed. Antegrade cystourethrography through suprapubic tube (Cystofix) revealed Y-type urethral duplication with narrow dorsal urethra within the penis and wide ventral urethra connected with the rectum [Figure 2].

Clinically, after the closure of cystostomy, the boy had urinary flow through the anus and only minimally dribbling through the penile urethra. Retrograde urethrography showed extremely narrow dorsal urethra and confirmed



Figure 2: Antegrade cystourethrography through suprapubic tube (Cystofix): Y-type urethral duplication with narrow dorsal urethra within the penis and wide ventral urethra connected with the rectum, contrast visible within rectum; normally shaped urinary bladder, no vesicoureteral reflux

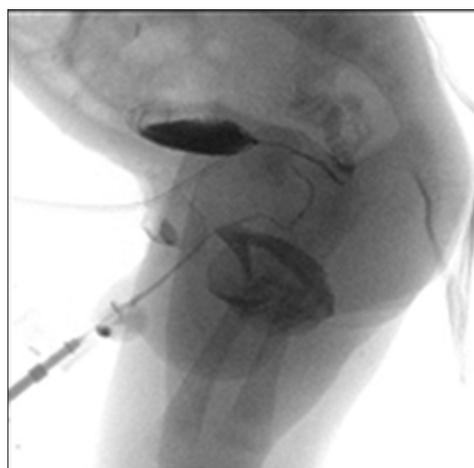


Figure 3: Retrograde urethrography: Extremely narrow dorsal urethra (within the penis shaft), ventral urethra opening into the rectum

the presence of Y-type IIA duplication with ventral urethra opening into the rectum [Figure 3]. No dilatation of upper urinary tract was visible on abdominal ultrasound. Transperitoneal ultrasound revealed abnormal structure of the penis, i.e., very short corpus spongiosum ending at the level of penile shaft and single corpus cavernosum within the penis [Figure 4].

Then, the boy underwent unsuccessful attempt of Fokker operation to approximate both parts of esophagus to perform primary anastomosis, and finally, upper pouch esophageal fistula was created and reconstruction of esophagus was planned at a later date. For those reasons, vesicostomy was performed to avoid potential urinary tract infections. In the future, anterior sagittal transrectal approach (ASTRA)

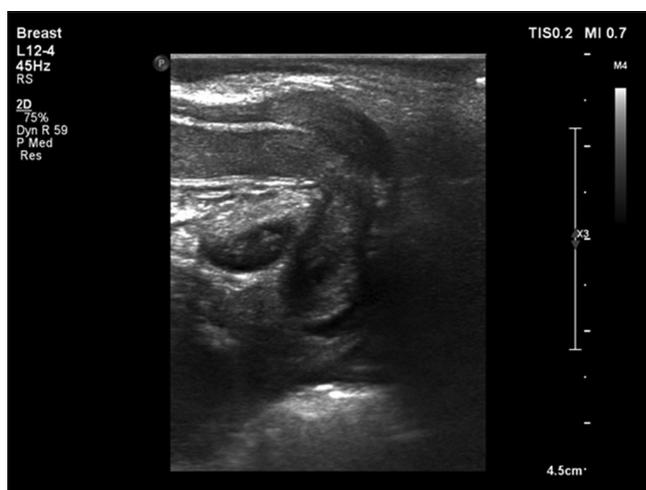


Figure 4: Transperitoneal ultrasound: Short corpus spongiosum ending at the level of penile shaft and single corpus cavernosum within the penis

procedure to separate the ventral, functioning urethra from the rectum leaving a definitive perineal urethrostomy, was planned as proposed by Macedo *et al.*^[14,16]

DISCUSSION

Y-type urethral duplication according to Effmann's classification is categorized as type IIA2 anomaly. In this condition, ventral urethra originates from the bladder neck or posterior urethra and is opened either in the perineum or within the rectum and dorsal dysplastic urethra opens within the glans. Both urethras typically are patent, but ventral urethra is the functional one with perineal urination or urination through the anus, while dorsal urethra produces mostly urinary dribbling.^[11-16] In older patients, the clinical presentation with recurrent urinary tract infections, epididymitis, and perineal abscess can be observed.^[11,12,17] In our patient, Y-type urethral duplication with rectal implantation of ventral urethra was diagnosed within newborn period.

In 2016, Meier and Latiff reported 28 cases collected from the literature of Y-type urethral duplication with normal dorsal urethra and small ventral fistula to perineal skin.^[18] This rare variant of Y-type duplication, also described as congenital posterior urethrocutaneous fistula, can be usually managed by removal of ventral urethra through perineal approach,^[19-21] and it is suggested to separate this anomaly from Y-type duplication with rectal implantation.^[22]

There is no definitive management strategy for Y-type urethral duplication. Basically, the management of patients with urethral duplication should be based on the identification of the functional urethra. Various operative techniques were proposed.^[11-14]

Reconstruction of penile urethra is more challenging option and usually needs multistage procedures and is prone to reoperations and failure.^[11] Recently, the ASTRA approach with repositioning of functioning, ventral urethra implanted in the rectum to perineal position as a perineal urethrostomy is strongly advocated. Macedo *et al.* do not recommend to perform great efforts to bring the functional urethra to the tip of the penis. Removal of dysplastic urethra following the ASTRA approach seems to be safe and feasible.^[14,16] We planned for our patient in whom also abnormal structure of the penis is present to perform the same procedure after restoring the continuity of the digestive tract.

We found in literature only one similar case of urethral duplication in a child with VATER association.^[23] VACTERL/VATER association is typically defined by the presence of at least three following congenital malformations: vertebral defects, anal atresia, esophageal atresia with tracheoesophageal fistula, cardiac defects, renal anomalies, and limb abnormalities.^[24] In our patient, this association was suspected due to the presence of esophageal atresia with tracheoesophageal fistula, ventricular septal defect, atrial septal defect, costal anomalies, and urethral duplication.

CONCLUSION

In any case of lack of capability of bladder catheterization resulting in suprapubic cystostomy, before removal of suprapubic tube, antegrade cystourethrography through the tube should be performed to delineate the anatomy of the lower urinary tract to exclude anatomical anomaly.

REFERENCES

1. Lima M, Destro F, Maffi M, Proietti DP, Ruggeri G. Practical and functional classification of the double urethra: A variable, complex and fascinating malformation observed in 20 patients. *J Pediatr Urol* 2017;13:42.e1-000000.
2. Lopes RI, Giron AM, Mello MF, Neto CM, Santos JD, Moscardi PR, *et al.* Urethral duplication type influences on the complications rate and number of surgical procedures. *Int Braz J Urol* 2017;43:1144-51.
3. AbouZeid AA, Safoury HS, Mohammad SA, El-Naggar O, Zaki AM, Hassan TA, *et al.* The double urethra: Revisiting the surgical classification. *Ther Adv Urol* 2015;7:76-84.
4. Onofre LS, Gomes AL, Leão JQ, Leão FG, Cruz TM, Carnevale J, *et al.* Urethral duplication a wide spectrum of anomalies. *J Pediatr Urol* 2013;9:1064-71.
5. Coleman RA, Winkle DC, Borzi PA. Urethral duplication: Cases of ventral and dorsal complete duplication and review of the literature. *J Pediatr Urol* 2010;6:188-91.
6. Arena S, Arena C, Scuderi MG, Sanges G, Arena F, Di Benedetto V, *et al.* Urethral duplication in males: Our experience in ten cases. *Pediatr Surg Int* 2007;23:789-94.
7. Salle JL, Sibai H, Rosenstein D, Brzezinski AE, Corcos J. Urethral duplication in the male: Review of 16 cases. *J Urol* 2000;163:1936-40.

8. Podesta ML, Medel R, Castera R, Ruarte AC. Urethral duplication in children: Surgical treatment and results. *J Urol* 1998;160:1830-3.
9. Woodhouse CR, Williams DI. Duplications of the lower urinary tract in children. *Br J Urol* 1979;51:481-7.
10. Effmann EL, Lebowitz RL, Colodny AH. Duplication of the urethra. *Radiology* 1976;119:179-85.
11. Lima M, Destro F, Di Salvo N, Gargano T, Ruggeri G. Fate of males with urethral "Y-duplication": 40-year long follow-up in 8 patients. *J Pediatr Surg* 2017;52:1335-9.
12. Singh S, Rawat J. Y-type urethral duplication in children: Management strategy at our center. *J Indian Assoc Pediatr Surg* 2013;18:100-4.
13. Haleblan G, Kraklau D, Wilcox D, Duffy P, Ransley P, Mushtaq I, *et al.* Y-type urethral duplication in the male. *BJU Int* 2006;97:597-602.
14. Macedo A Jr., Silva MI, Pompermaier JA, Ottoni SL, de Castro R, Leal da Cruz M, *et al.* The anterior sagittal transrectal approach (ASTRA) for cases associated with rectal implantation of the urethra: A retrospective review of six cases. *J Pediatr Urol* 2017;13:613.e1-000.
15. Kumaravel S, Senthilnathan R, Sankkarabarathi C, Bagdi RK, Soundararajan S, Prasad N, *et al.* Y-type urethral duplication: An unusual variant of a rare anomaly. *Pediatr Surg Int* 2004;20:866-8.
16. Macedo A Jr., Ottoni SL, Leal da Cruz M, Pompermaier JA, Silva MI, Liguori R, *et al.* Y-type urethral duplication with rectal implantation of the urethra: Which is the best approach? *J Pediatr Urol* 2018;14:79-80.
17. Arda IS, Hiçsönmez A. An unusual presentation of Y-type urethral duplication with perianal abscess: Case report. *J Pediatr Surg* 2002;37:1213-5.
18. Meier DE, Latiff A. Y-type congenital urethral duplication with normal dorsal urethra and small ventral fistula to perineal skin 28th reported case. *J Pediatr Surg Case Rep* 2016;8:37-9.
19. Wagner JR, Carr MC, Bauer SB, Colodny AH, Retik AB, Hendren WH, *et al.* Congenital posterior urethral perineal fistulae: A unique form of urethral duplication. *Urology* 1996;48:277-80.
20. Dayanc M, Irkilata HC, Kibar Y, Bozkurt Y, Basal S, Xhafa A, *et al.* Y-type urethral duplication presented with perineal fistula in a boy. *Ger Med Sci* 2010;8:Doc33.
21. Sánchez MM, Vellibre RM, Castelo JL, Arias MP, Sarmiento RC, Costa AR, *et al.* A new case of male Y-type urethral duplication and review of literature. *J Pediatr Surg* 2006;41:e69-71.
22. AbouZeid AA, Mohammad SA, Radwan NA, Safoury HS, El-Naggar O, Hay SA, *et al.* Y-type urethral duplication: A True variant of the anomaly or a misnomer? *Eur J Pediatr Surg* 2016;26:245-51.
23. Nerli RB, Ghagane SC, Dixit NS, Hiremath MB. Urethral duplication in a child with VATER association. *Urol Case Rep* 2019;23:29-31.
24. Solomon BD. VACTERL/VATER association. *Orphanet J Rare Dis* 2011;6:56.

How to cite this article: Warchol S, Glug W, Bombinski P, Dudek-Warchol T. Y-type Urethral Duplication with Rectal Implantation of the Urethra in a Boy with Long-gap Esophageal Atresia: A Case Report. *Clin Res Urol* 2019;2(1):1-4.