SUBMITTED 27 MAY 21
REVISION REQ. 25 JUL 21; REVISION RECD. 12 AUG 21
ACCEPTED 9 SEP 21
ONLINE-FIRST: SEPTEMBER 2021
DOI: https://doi.org/10.18295/squmj.9.2021.138

Progressive Dilatation as a Successful Treatment for Y duplication of Urethra

A case report

***Akash Pati, Subrat K. Sahoo, Bikasha B. Tripathy**

Department of Pediatric Surgery, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India
**Corresponding Author's e-mail: patiakash.ap@gmail.com*

Abstract

Duplication of urethra is a rare congenital anomaly that has been reported in case reports and case series. A Y-shaped urethral duplication is the rarest variant as per the classification suggested and hence lacks a standardized treatment option. We report a case of Y-duplication of urethra diagnosed during neonatal age and presented to us at nine years of age. The patient had undegone a vesicostomy at seventh day of life for passing urine per anus and was lost to follow up thereafter. An attempt at disconnection of the duplicated urethral tract to anus after colostomy, at eight years of age had failed. The patient was managed successfully by progressive dilatation of the orthotopic urethra, which required multiple stages, followed by separation of the urethra from the rectum. At three years follow up the patient is continent and asymptomatic.

Keywords: Y duplication of the urethra; PADUA technique; Rectourinary fistula; Urethral duplication.

Introduction

Duplication of the urethra is a rare congenital anomaly of the lower urinary tract, occurring more commonly in males. Multiple variants of duplication anomalies have been described for lower urinary tract. Duplication of the urethra can occur along with the bladder or in isolation. When it

affects the urethra alone, it usually occurs in the sagittal plane on a single phallus. Several types of anatomic variations in the duplicated urethra have been identified. The accessory urethra may be incomplete or complete, opening in the midline at normotopic, episapdaic, hypospadiac or perineoanal region. There is no standard treatment described for the condition as it is rare. The progressive augmentation by dilating the urethra anterior (PADUA) technique was used successfully for urethral hypoplasia,[1] but promising results have not been described for its use in "Y duplication" of urethra. We describe a case, where the PADUA technique has been used to successfully manage the Y duplication of urethra and have discussed the possible reason for our successful outcome.

Case Report

A nine-year-old male child presented to us with a cutaneous vesicostomy, and right transverse loop colostomy done elsewhere. The child was passing urine per anus at birth, and was unable to pass urine per urethra, for which a vesicostomy was done at seventh day of life. An ultrasonogram of the abdomen done during neonatal age was suggestive of left ectopic kidney in midline, sub umbilical position and healthy right kidney and bladder. Following the vesicostomy the patient was lost to follow up. At eight years of age, the patient again presented to a surgeon and had underwent a right transverse colostomy. As per the records, an attempted surgical disconnection between the urethra and anal orifice through the perineal approach had failed.

At the time of presentation to us, the patient was passing most of the urine per vesicostomy and was occasionally dribbling drops of urine per anus. A small pit-like opening was present on the tip of the glans. The rest of the genitalia and abdominal examination was unremarkable. On digital rectal examination, anal opening of the fistula was palpable anteriorly just inside the anal verge. A functional nuclear imaging revealed a small ectopic kidney in the left lower abdomen, contributing to 23% of total renal function, and there was no morphological or functional abnormality in the right kidney. A voiding cystourethrogram done after blocking the vesicostomy demonstrated a very thin caliber anterior urethra and a normal posterior urethra. The urinary bladder was smooth in outline and of small volume with bilateral grade two vesicoureteric reflux. During micturition, the dye was flowing into the rectum. The distal colostogram was unremarkable.

Cystoscopy performed through the vesicostomy revealed a small capacity bladder with a smooth wall. The left ureteric orifice was ectopic, opening medially on the trigone while the right ureteric orifice was orthotopic. Bladder neck was regular, and scope could be negotiated through the bladder neck into the posterior urethra. There was a fistulous opening seen in the posterior wall of the posterior urethra distal to the veru. A standard 0.018" straight tip guidewire inserted through the fistulous opening in the posterior urethra was retrieved from the anal canal.

There was a small pit at the tip of the glans resembling a stenotic meatus. A 0.018" straight flexible tip guidewire introduced through this meatal pit and was retrieved at the stoma of the vesicostomy under guidance of a cystoscope inserted through the vesicostomy. A well lubricated three French gauge (Fr) ureteral catheter was inserted over the guidewire smoothly and was retrieved through the vesicostomy. The catheter was left indwelling, with a plan for weekly dilatation. Every week, after adequate lubrication, a progressively larger catheter was passed gently over a guidewire and left indwelling as a ring stent. This mode of progressive dilation continued for several weeks until a dilatation of 16 Fr was achieved. [Figure 1]. A voiding cystourethrogram after blocking the vesicostomy [Figure 2 A] was performed which confirmed good caliber of the orthotopic urethra.

The fistulous connection between the posterior urethra and anal canal was closed via a perineal approach. The perineal body muscles were interposed between the anterior wall of the rectum and the urethra to prevent a recurrence. The colostomy and vesicostomy were reversed after four weeks. Postoperatively, the indwelling bladder catheter was kept for ten days and on removal, the patient could pass urine per urethra. The patient was on clean intermittent self-catheterization and a weekly calibration schedule until six months following vesicostomy closure.

A good urethral caliber was seen on voiding cystourethrogram, performed after closing the vesicostomy and at six months follow up. [Figure 2B&C]. Uroflowmetry done immediately after the vesicostomy closure had a flow rate of 5ml/sec which increased to 12 ml/sec at six months follow-up and there was no post void residue. At three years follow up, the patient is continent

with a normal voiding stream. The parents of the patient provided informed consent for publication of this case.

Discussion

Y duplication of the urethra occurs when the prostatic urethra splits into two channels with one coursing to the glans, and the other more functional one extends ventrally to the perineal area near the anus. Less than 50 cases of Y duplication of urethra have been described in English literature.[2] The orthotopic urethra is characteristically stenotic. This form of aberration is usually associated with other congenital anomalies in the genitourinary and gastrointestinal tract. In the present case described, there was an associated left ectopic kidney with bilateral vesicoureteric reflux.

Being a rare condition there is no standardized method of treatment for the anomaly. Several surgeons have mobilised the perineal urethra to the scrotal skin level and laid open the orthotopic stenoptic urethra [3,4]. At a second stage they have performed a urethroplasty. Singh and Rawat have also performed a similar method, but they have completely excised the orthotopic urethra and have tried to reconstruct the neourethra from locally available healthy tissue.[5] Sinha et al have tried to reinforce the urethra by strips of mucosa from the anterior anorectum.[6] Passerini – Glazel et al have successfully managed the orthotopic urethral hypopalasia by PADUA technique.[1] The authors have reported a slow progressive dilatation of the hypoplastic urethra to be successful in 6 out of 8 cases. We have employed the same method of gradual progressive dilatation of anterior urethra in the case of Y duplication of the urethra. Some authors do not agree with the PADUA technique of dilatation, as repeated trauma leads to stricture formation and the hypoplastic urethra is not usable.[5] Lima et al have reported, PADUA technique, to be ineffective in two cases of Y duplication of urethra as both the cases have developed urethral stenosis.[7]

The urethra is a distensible structure and probably remains hypoplastic because of the presence of a low-pressure channel to the rectum. As a result, the anterior urethra is not challenged by the voiding pressure and urinary stream and remains hypoplastic in cases with Y duplication. The cause of failure of the PADUA technique, as mentioned by different authors can be summarized

as multiple injuries to the hypoplastic urethra resulting in stricture. A gentle progressive dilatation is likely to have less trauma on the orthotopic urethra. Hence, a smooth progressive dilatation will give a successful result without a stricture of the urethra. The procedure of dilatation is such that the catheter is large enough to fill the lumen of the urethra but not forced.[1]

Conclusion

We could successfully manage the patient using the PADUA technique described for urethral hypoplasia. The patient had a good urinary stream from the orthotopic meatus and had no post void residue. Gentle progression of the dilatation was the key to success. However, more cases of Y duplication of urethra should be managed by slow progressive dilatation of the hypoplastic orthotopic urethra before any definite conclusion is formulated.

Authors' Contribution

AP conceptualized the report. AP, SKS and BBT were involved in data collection and acquisition, preparation of the original manuscript and revision of the manuscript. All authors approved the final version of the manuscript.

References

1. Passerini-Glazel G, Araguna F, Chiozza L, Artibani W, Rabinowitz R, Firlit CF. (1998)The P.A.D.U.A. (Progressive Augmentation by Dilating the Urethra Anterior) Procedure for the Treatment of Severe Urethral Hypoplasia. Journal of Urology. 140(5 Part 2):1247–9.
2. Wagner JR, Carr MC, Bauer SB, et al. Congenital posterior urethral perineal fistulae: a unique form of urethral duplication. Urology 1996; 48:277 - 80.
3. Podesta Miguel L., Medel Ricardo, Castera Roberto, Duarte Adolfo C. (1998)Urethral duplication in children: surgical treatment and results. Journal of Urology. 160(5):1830–3.
4. Wakhlu A, Wakhlu AK. (1998)Management of H-type urethral duplication. Journal of Pediatric Surgery. 33(8):1238–42.
5. Singh S, Rawat J. Y-type urethral duplication in children: Management strategy at our center.(2013) J Indian Assoc Pediatr Surg. 18(3):100–4.

6. Sinha S, Sen S, Chacko J, Thomas G, Karl S, Mathai J. (2006)Y-duplication of the male urethra: use of anterior anorectal wall for posterior urethral lengthening. Ped Surgery Int. 22(6):529–31.
7. M. Lima, F. Destro, M. Maffi, D. Persichetti Proietti, G. Ruggeri. (2017)Practical and functional classification of the double urethra: A variable, complex and fascinating malformation observed in 20 patients. Journal of Pediatric Urology, 13(1): 42.e1-42.e7

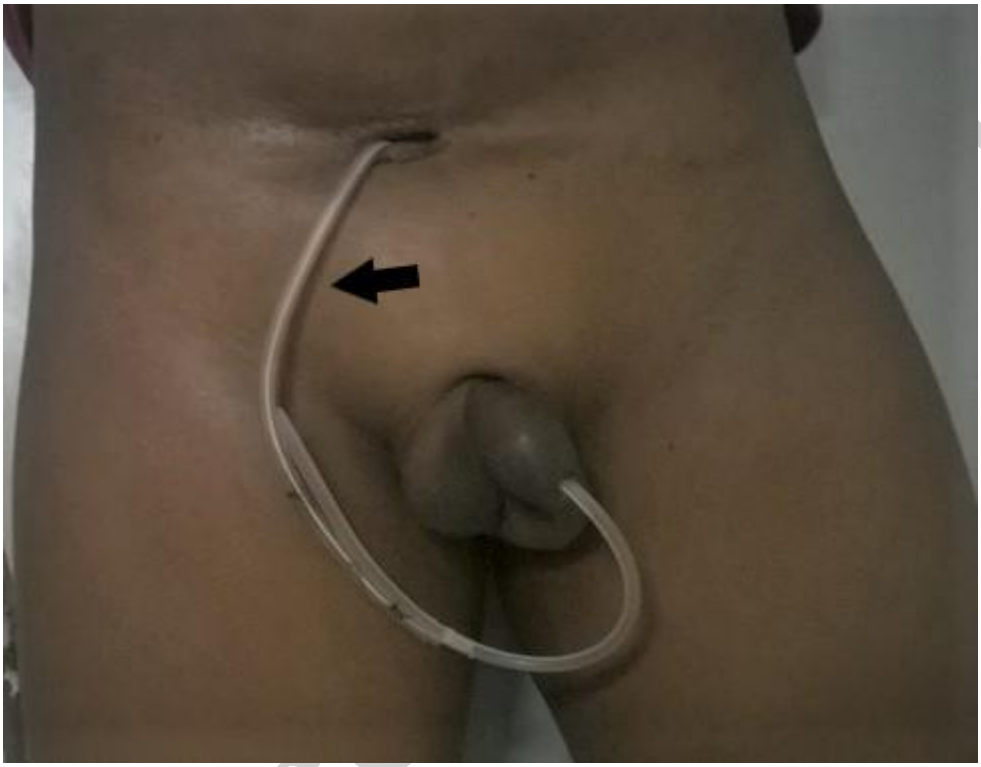

Figure1: Ring stent (Black Arrow) of 16 Fr kept indwelling passing through orthotopic urethra and exiting through vesicostomy.

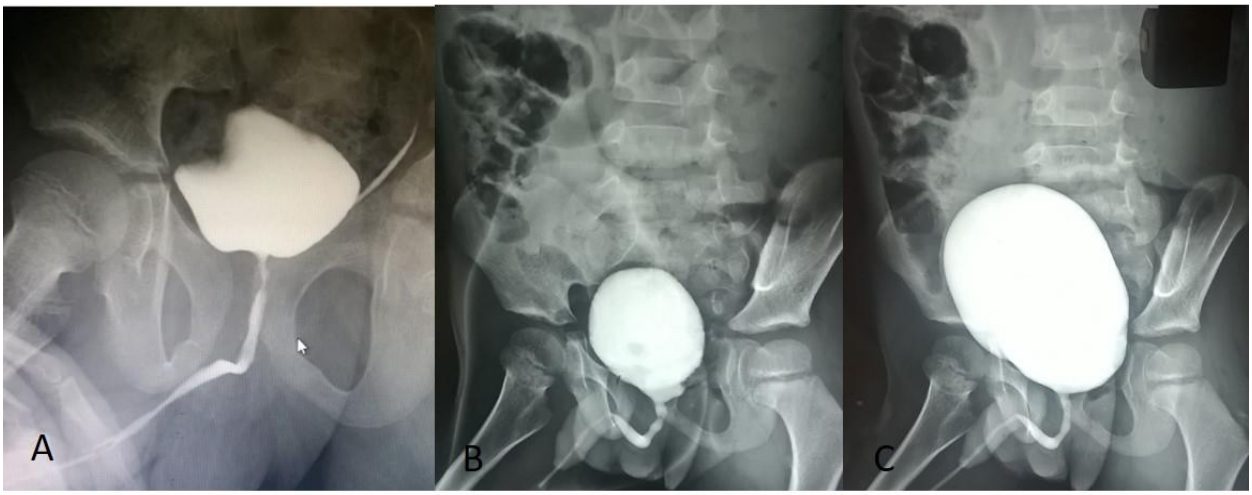

Figure 2: Voiding cystourthrogram done after blocking the vesicostomy (A), immediately after closing the vesicostomy(B) and at 6 months follow up (C).