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CASE REPORT

Progressive Dilatation as a Successful Treatment for Y Duplication of Urethra

A case report

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ABSTRACT: Duplication of urethra is a rare congenital anomaly; a Y-shaped urethral duplication is the rarest variant and lacks a standardised treatment plan. We report a nine-year-old male patient with a Y-duplication of urethra diagnosed during neonatal age who presented to a tertiary care teaching hospital in India in 2018. The patient had undergone a vesicostomy on the seventh day of life for passing urine per anus and was lost to follow-up thereafter. At eight years of age, there was a failed attempt at disconnection of the duplicated urethral tract from the anus after colostomy. 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 was continent and asymptomatic.

Keywords: Urethral Duplication; Case Report; India.

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 the 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 the perineoanal region. There is no standard treatment described for this condition as it is rare. The progressive augmentation by dilating the urethra anterior (PADUA) technique has been successfully used for urethral hypoplasia, but promising results have not been described for its use in 'Y duplication' of urethra.[1] We present a case where the PADUA technique was successfully used to manage the Y duplication of urethra and discuss the possible reason for this successful outcome.

Case Report

A nine-year-old male patient presented to a tertiary care teaching hospital in India in 2018 with a cutaneous vesicostomy and right transverse loop colostomy done elsewhere. The patient was passing urine per anus at birth and was unable to pass urine per urethra, for which a vesicostomy was done on the 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 underwent a right transverse colostomy; an attempted surgical disconnection between the urethra and anal orifice through the perineal approach failed.

At the time of presentation, the patient was passing most of urine per vesicostomy but was occasionally passing 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 scan revealed a small ectopic kidney in the left lower abdomen, contributing to 23% of total renal function; 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 a 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 verumonatum. A standard 0.018 inch straight tip guidewire inserted through the fistulous opening in the posterior urethra was retrieved from the anal canal.

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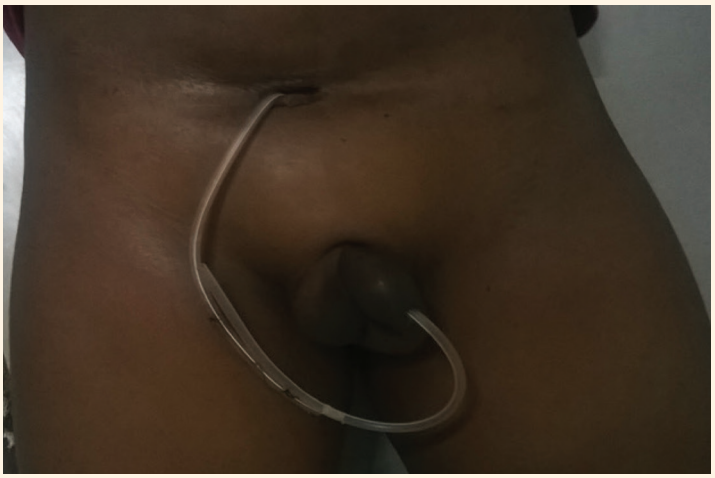

Figure 1: Photograph of a nine-year-old male patient showing a ring stent (arrow) of 16 Fr kept indwelling passing through orthotopic urethra and exiting through vesicostomy.

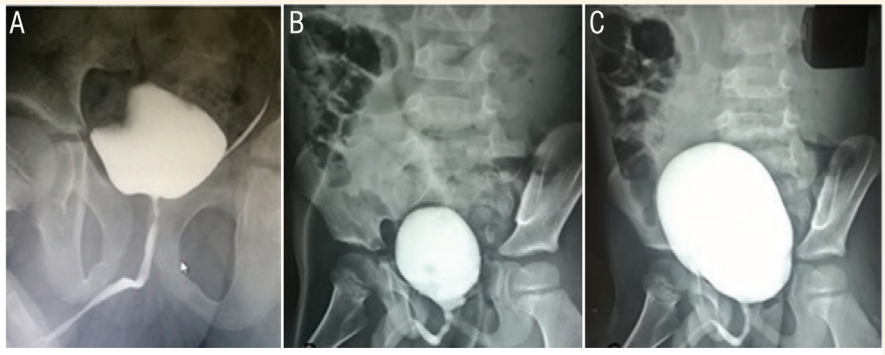

Figure 2: Cystourthrogram showing voiding (**A**) after blocking the vesicostomy, (**B**) immediately after closing the vesicostomy and (**C**) at six months follow-up.

There was a small pit at the tip of the glans resembling a stenotic meatus. A 0.018 inch 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 was performed which confirmed good caliber of the orthotopic urethra [Figure 2A].

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 10 days and on removal, the patient could pass urine per urethra. The patient was on clean intermittent self-catheterisation 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 and C]. Uroflowmetry done immediately after the vesicostomy closure had a flow rate of 5 mL/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 was 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 the 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, there was an associated left ectopic kidney with bilateral vesicoureteric reflux.

Being a rare condition there is no standardised method of treatment for this anomaly. Several surgeons have mobilised the perineal urethra to the scrotal skin level and laid open the orthotopic stenotic urethra.[3,4] At a second stage, a urethroplasty can be performed. 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.* tried to reinforce the urethra by strips of mucosa from the anterior anorectum.[6] Passerini-Glazel *et al.* successfully managed the orthotopic urethral hypoplasia using the PADUA technique wherein a slow progressive dilatation of the hypoplastic urethra was successful in 6 out of 8 cases.[1] In the present case, the same method of gradual progressive dilatation of the anterior urethra in the case of Y duplication of the urethra, was used. Some 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.* reported that the PADUA technique was ineffective in two cases of Y duplication of urethra as both the cases 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 can be summarised 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 is not forced.[1]

Conclusion

This case shows the successful management of a 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 a successful outcome. More cases of Y duplication of urethra should be managed by slow progressive dilatation of the hypoplastic orthotopic urethra before a definitive conclusion can be drawn.

AUTHORS' CONTRIBUTIONS

AP conceptualised 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.

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