Type IIA2 urethral duplication: report of an unusual case

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Objective **This report describes a rare case of type IIA2 sagittal urethral duplication.**

Summary background data **The presentation, investigation, and management of this rare anomaly are briefly discussed.**

Methods **A 3½-year-old boy presented with urinary obstruction and recurrent urinary tract infection due to a stenosed dorsal urethra and segmental stenosis of the dominant ventral urethra. The child also had left-sided vesicoureteric reflux. Staged surgical management consisted of an initial vesicostomy followed by serial dilatation of the ventral urethral stricture, left ureteric reimplantation, and a 2-cm long distal urethrourethrostomy between the dorsal urethra, opening at the tip of the penis, and the ventral urethra, which had a hypospadic opening at the base of the glans.**

Results **The functional and cosmetic outcomes were satisfactory.**

Conclusion **The management needs to be individualized as best suited for the patient.** ***Ann Pediatr Surg*** **11:55–58**

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Keywords: urethra, urethral duplication, urethral stricture

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Introduction

Duplication of the urethra is a rare anomaly that ranges from blind accessory channels to a true duplication with/without complete duplication of the urinary bladder [1]. The most commonly used classification of urethral duplications was described by Effmann *et al.* [2]. In this report, we describe a rare case of sagittal urethral duplication (Effmann type IIA2) in a 3½-year-old boy who presented with urinary obstruction due to a segmental stricture in the dominant, normal caliber ventral urethra, grade 5 left-sided vesicoureteric reflux (VUR), and a history of recurrent urinary tract infection (UTI) with right-sided epididymo-orchitis. Staged surgery for management of the various components of the anomaly was successful [3].

Case report

A 3½-year-old boy presented with straining during micturition and passage of urine from two openings, one near the tip of the penis and another on the undersurface of the penis. There was a history of three episodes of febrile UTI in the past, two of these being associated with painful enlargement of the right testis. Urine culture of past two episodes of UTI showed *Escherichia coli* and was treated as per the sensitivity report with amoxicillin clavulanic acid. On examination, there were two urethral openings. The dorsal urethral meatus was narrow in caliber and opened at the tip of the glans, whereas the ventral meatus, which was normal in caliber, opened in a hypospadiac position at the base of the glans (Fig. 1). There was no chordee and the prepuce was intact. The child was passing urine in a poor stream from the ventral meatus and only in drops from the dorsal meatus. There was no dribbling of urine between acts of micturition. He also had a very firm, mildly tender right testis.

Biochemical renal function tests were normal. Abdominal ultrasonography (US) showed left-sided hydroureteronephrosis with a distended thick-walled bladder. US showed the right testis to be enlarged with increased vascularity suggestive of epididymo-orchitis. A retrograde urethrogram showed a sagittal duplication of urethra, with the narrow dorsal urethra originating from the ventral urethra in the prostatic region (Fig. 2). There was a short-segment narrowing of the ventral urethra in its bulbomembranous part, and the urethra proximal to the narrowing was dilated, elongated, and tortuous until the bladder neck (Fig. 3). The urinary bladder was large with an irregular outline, and there was left-sided grade 5 VUR. Examination under anesthesia showed a very narrow caliber dorsal urethra allowing only the stylet of an 8-Fr Foley's catheter to be passed. Endoscopy confirmed narrowing of the ventral urethra starting 5 cm proximal to the meatus. A Bloksom's cutaneous vesicostomy was performed.

Subsequently, a ^{99m}Tc DMSA scan showed thinning of the cortex of the left kidney with multiple cortical scars and a function of 30.04%. The right kidney showed normal function and excretion. Six months later, an antegrade cystourethroscopy was performed through the vesicostomy. A small guide wire passed through the dorsal meatus was seen to enter the common prostatic urethra just distal to the verumontanum. The bladder neck was competent, whereas the proximal ventral urethra was dilated. The stricture in the ventral urethra was sequentially dilated with urethral dilators up to 8-Fr size. A urethral bougie was passed into the bladder and a 1-0

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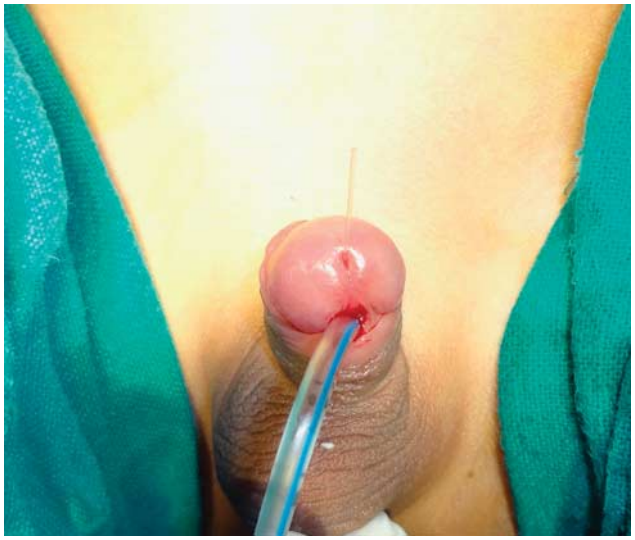
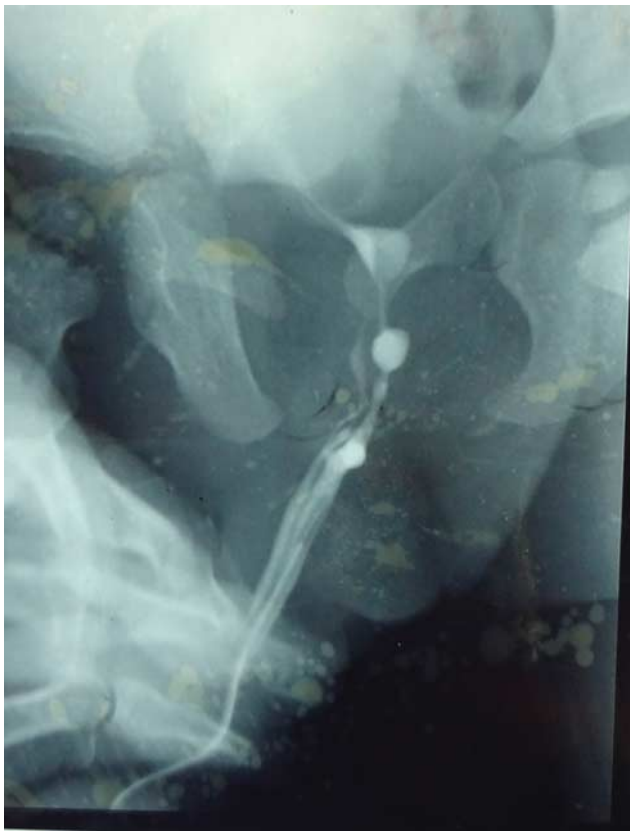
Fig. 1



Clinical picture of the urethral anomaly, showing two orifices in the glans penis: dorsal urethral meatus at the tip with a 24-G venflon and ventral urethral meatus in a hypospadiac position at the base of glans with a 8-Fr infant feeding tube. Note a thick septum separating both the orifices.

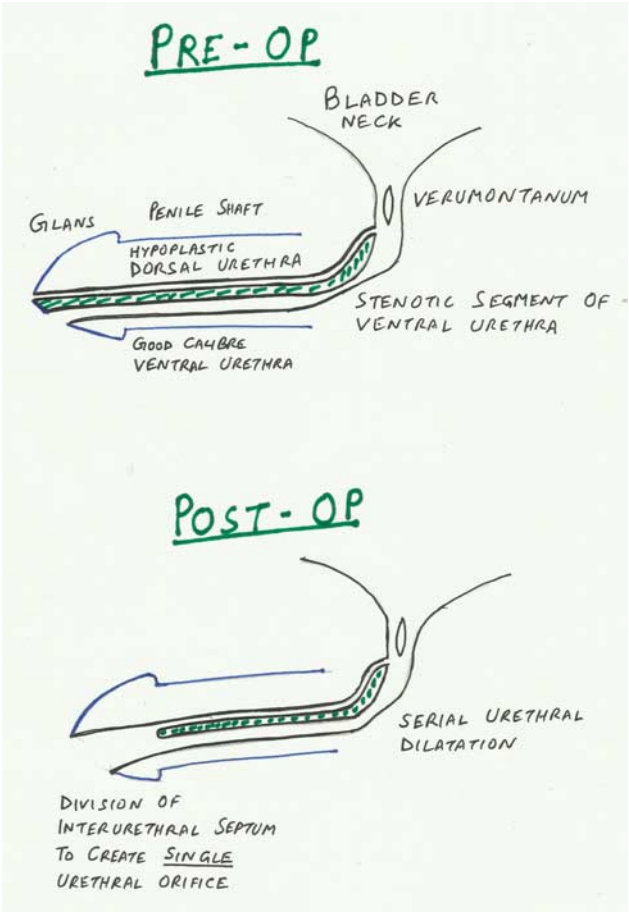
Fig. 2



Retrograde urethrogram showing a sagittal duplication of urethra with the narrow dorsal urethra originating from the ventral urethra in the prostatic region.

Prolene (Ethicon Inc., Baddi, Haryana, India) thread was tied to its tip and taken out through the external ventral meatus to be kept for future urethral dilatations by the

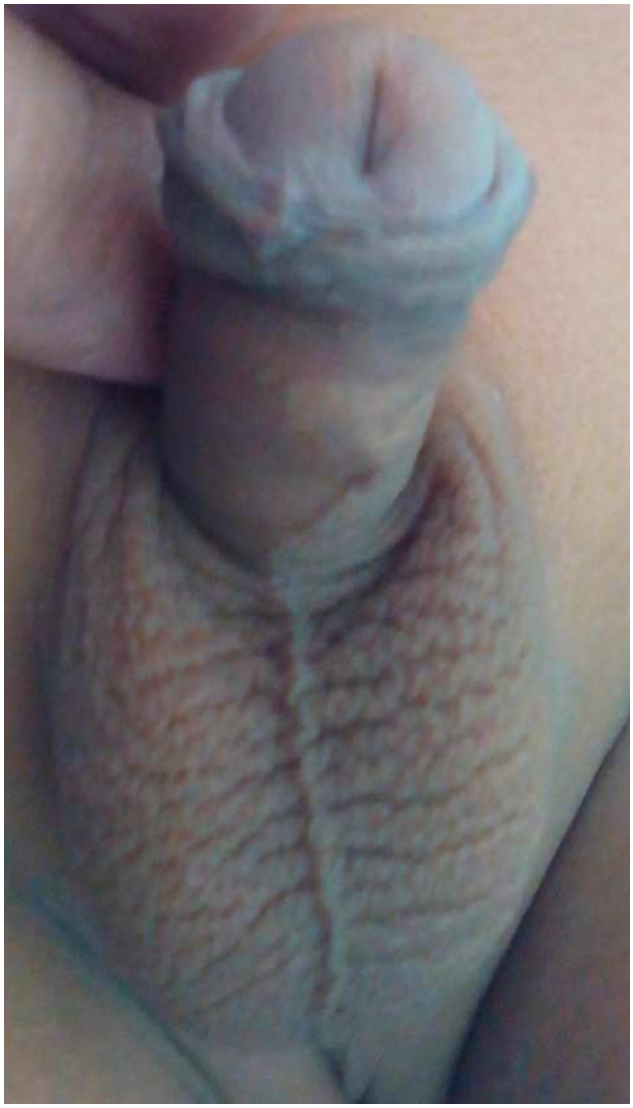
Fig. 3



Schematic diagram showing the anatomy at the first presentation and after division of the interurethral septum.

rail–road technique. Subsequently, at 3-week intervals, urethral dilatations were performed, using the rail–road technique by tying urethral catheters to the thread and increasing the size of the catheters until the ventral urethra could easily be dilated to size 10 Fr. Thereafter, without the use of the in-situ thread, urethral dilation with Clutton's bougies until size 10/13 Fr could easily be performed. Three months later, using a Pfannenstiel incision, the vesicostomy was mobilized. The bladder was still of large capacity. The left ureter was mobilized and reimplanted in a transtrigonal manner. Postoperatively, the urinary stream from the ventral urethra was good in caliber. During the initial visits, the child complained of nocturnal incontinence, but this diminished gradually over a period of 3 months. There were no new episodes of UTI. Three months following the ureteric reimplantation, the septum between the two urethras was divided proximally for around 2 cm by crushing the intervening tissue between the blades of an artery forceps before its division by a scissors. Following this, the child passes urine in a good single stream through the single neomeatus at the base of the glans (Figs 3 and 4). The neomeatus as well as the ventral urethra were calibrated easily using a 10-Fr infant feeding tube on the first few OPD visits. At 1-year follow-up, both the parents and the child are satisfied with the functional and cosmetic

Fig. 4



Appearance of the neomeatus following division of the interurethral septum.

outcome. We have counseled the parents for the need for follow-up with a repeat micturating cystourethrogram (MCU) and a DTPA renal scan. After relief from symptoms, they seem a bit reluctant due to the unpleasant experience while performing MCU. They are in the long waiting list for DTPA renal scan in resource-limited healthcare system.

Discussion

Urethral duplication is rare with less than 200 cases of the various forms of the anomaly being described in the literature. In 1975, Williams and Kenawi [3] divided urethral duplications into sagittal and collateral categories. The more common sagittal categories were classified into the epispadic, hypospadic, spindle urethra, and Y-duplication groups, with the epispadic and hypospadic urethral duplications being further subdivided into complete, incomplete, and abortive varieties [3]. The more commonly used and comprehensive classification, described by Effmann *et al.* [2], classified urethral duplications into: type I, blind, incomplete urethral duplication; type IIA, complete patent urethral duplication with two meatus (type IIA1, two noncommunicating urethras arising independently from the bladder, and type IIA2, a second channel arising from the first and coursing independently to a second meatus); type IIB, complete patent urethral duplication with one meatus (two urethras arising from the bladder or posterior urethra and uniting to form a common distal channel); and type III, urethral duplication as a component of partial or complete caudal duplication. In type IIA2 duplications, the ventral channel is the more normal or dominant one and passes through the prostate and sphincter mechanism [4,5]. As also seen in our patient, in patients in whom the dorsal accessory channel and its meatus are positioned normally, the ventral channel opens in a hypospadic position anywhere from the penoscrotal junction to the glans [2,5].

Most patients with patent urethral duplication are asymptomatic or complain only of a double urinary stream, although incontinence, dysuria, and infection are occasionally noted [2]. Obstruction has been described when the dorsal urethra has a narrow distal segment or a stenotic meatus that causes obstruction and distension of the proximal portion, with resultant external compression and secondary obstruction of the more normal ventral urethra [4]. Our patient, however, presented with a double stream with obstruction to both streams, as the ventral urethra, which was otherwise of good caliber, had a segmental stricture in the bulbar region. Pippi Salle *et al.* [6] have also reported a case of type IIA2 duplication with stenosis of the ventral hypospadic urethra, recurrent UTI, and left-sided VUR, in whom, as in our patient, an initial vesicostomy was performed. Ortolano and Nasrallah [7] described a somewhat similar case of type IIA2 urethral duplication with urinary obstruction, due to stenosis of both urethral channels, in whom a preliminary vesicostomy was required. Mane *et al.* [8] also reported performing vesicostomy as an initial procedure in a case of type IIA2 urethral duplication. Our patient had recurrent episodes of UTI with epididymo-orchitis. This may be a consequence of the dilated posterior urethra proximal to the obstruction, allowing reflux of infected urine into the vas deferens. A similar finding has been reported earlier in the literature [9].

Radiological evaluation should show both urethral channels fully, and for this a MCU with oblique films is the primary study [4]. A retrograde urethrogram with injection of contrast medium into both channels simultaneously may be necessary, if the accessory channel is very narrow or the child is unable to void [4,10]. As in our patient, cystourethroscopy may be necessary to visualize the verumontanum and other urethral characteristics [2].

Associated upper urinary tract anomalies are uncommon but include ectopic kidney, megaureter, unilateral or bilateral VUR, and hydronephrosis [11–14]. Therefore, an abdominal US should be performed in all patients [4,6,7], whereas an intravenous urogram may be considered for

those with more complex findings [6] or where the US shows abnormal findings.

The treatment of urethral duplication depends on the anatomy of the duplication and on its clinical manifestations [4]. Patients with mild symptoms may not require treatment, but surgery should be considered for disturbing symptoms such as an annoying double stream, urinary obstruction or incontinence, or a severe cosmetic defect [4].

The anatomy of the duplication should be defined and the functional urethra identified before any corrective surgical intervention. A variety of surgical procedures have been described for managing urethral duplications. Complete excision of the accessory urethra by a penile or a combined penile and retropubic approach [14,15] is the most definite procedure [4,6], although it involves extensive and time-taking dissection from the corpora with which it is intimately attached. Ligation, fulguration, or sclerosis of the accessory channel have also been described [16], but are not so popular. Pippi Salle *et al.* [6] cautioned against using the accessory urethra because it is hypoplastic and the risk for inadequate urine flow is high. Instead, a urethroplasty should be performed in continuity to hypospadiac but good caliber ventral urethra, choosing from various techniques of hypospadias repair. However, if the dorsal urethra, although nondominant, is of good caliber, a side-to-side urethrourethrostomy may be performed [8,17]. In our patient, the distal urethral opening was at the tip of the penis, and division of the distal septum between the urethras (distal urethrourethrostomy) resulted in a single, adequate meatus at the base of the glans. As the dorsal urethral channel was continent, this procedure avoided extensive dissection of complete excision and is equally effective. Such a relatively simple procedure has been described earlier [4,7] and can result in a satisfactory cosmetic and functional outcome.

An extremely unusual finding in our patient was a segmental stenosis of the ventral urethral channel in the bulbar area, necessitating a preliminary vesicostomy. In contrast to the somewhat similar case reported by Ortolano and Nasrallah [7] in whom both urethral channels were severely hypoplastic, in our patient, the ventral urethra was otherwise of adequate caliber. The stricture was successfully managed by initial retrograde and later prograde dilatation using progressively increasing sizes of urethral catheters and dilators. Ortolano and Nasrallah [7] also performed progressive dilatation of the ventral channel for 4 months with silicone tubing, resulting in a satisfactory outcome. Pippi Salle *et al.* [6] performed an initial vesicostomy in a boy with an apical and a hypospadic meatus with left-sided VUR, and followed it up with an onlay-flap urethroplasty with ureteric reimplantation.

Conclusion

Our report illustrates the fact that management of type IIA2 urethral duplication needs to be individualized, as urinary obstruction may not be because of the stenosed dorsal urethral channel but due to an abnormality in the ventral urethra. An initial vesicostomy may be a useful temporizing procedure in patients presenting with urinary obstruction, as delineation of the exact anomalous anatomy may require detailed assessment of imaging studies. Excision of the dorsal urethral channel is not mandatory in all symptomatic patients, especially when it opens close to the tip of the penis, and its distal portion can be anastomosed to the terminal ventral urethra for a satisfactory functional outcome.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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