A rare case of urethral triplication in association with tethered cord and vertebral anomalies

Jujju J. Kurian, Susan Jehangir Homi and Immanuel S. Karl

In this manuscript, we describe the case of a 5-year-old boy with Y triplication of the urethra in association with fatty filum terminale, tethered cord and vertebral anomalies. Unlike a Y duplication in which the penile meatus is hypoplastic, a Y triplication can have a penile meatus of adequate calibre, which can be used as a functional conduit. Most patients with urethral triplication have been found to have associated malformations. To our knowledge, this is the first reported case of urethral triplication seen in association with fatty filum terminale, tethered cord and vertebral anomalies. ***Ann Pediatr Surg*** **12:119–121 © 2016 Annals of Pediatric Surgery.**

Annals of Pediatric Surgery 2016, **12**:119–121

Keywords: rectus flap, tethered cord, urethral triplication, vertebral anomaly

Department of Paediatric Surgery, Christian Medical College, Vellore, Tamil Nadu, India

Correspondence to Jujju J. Kurian, MBBS, MS, MRCS, MCh, Department of Paediatric Surgery, Christian Medical College, Vellore, 632004 Tamil Nadu, India
Tel: +91 948 623 6433; fax: +91 416 223 2035;
e-mail: jujjujacobkurian@gmail.com

Received 4 November 2014 **accepted** 22 June 2015

Introduction

Urethral triplication is an extremely rare congenital anomaly of the lower urinary system. To date, only 11 cases have been reported in the English literature. Herein, we present the case of a 5-year-old boy with Y triplication of the urethra with concomitant fatty filum terminale, tethered cord and vertebral anomalies. To the best of our knowledge, such an association has not been reported.

Case report

A 5-year-old boy was referred to our department with complaints of passing urine per rectum since birth. A failed attempt to correct this anomaly had been made earlier elsewhere. He had recently undergone detethering of the spinal cord when he developed progressive lower limb weakness. On examination, the tip of the phallus revealed two urethral openings (Fig. 1). An 8-Fr catheter was inserted through the ventral meatus, whereas the dorsal opening was not catheterizable.

Voiding cystourethrography revealed a single urethra originating from the bladder neck, which split into three channels – two ending on the penis and one communicating with the rectum (Fig. 2). At surgery, a 7.5-Fr cystoscopy was passed through the ventral penile urethral meatus. It revealed an adequate-sized urethra up to the bladder neck and the opening of the Y fistula just distal to the verumontanum. A guidewire passed into this opening exited through the anus (Fig. 3).

An inverted U-shaped incision was made in the perineum. The dissection was carried out between the urethra and the anterior rectal wall, which led to the identification of the urethrorectal channel with the help of the guidewire. The channel was disconnected from the rectal wall and closed with 6-0 PDS sutures. The abdomen was then opened using a lower midline incision. The left rectus abdominis muscle was then dissected preserving the inferior blood supply. Further dissection from the abdominal side resulted in the opening up of the space between the rectum and the bladder/urethra. This left rectus abdominis muscle flap was then interposed between the urethra and the rectum. A 6-Fr catheter was passed through the ventral penile urethral meatus into the bladder and retained. A covering colostomy was performed.

The postoperative period was uneventful. The urethral catheter was removed after 2 weeks. The covering colostomy was closed after 3 months. At 18-month follow-up the child is voiding with a good urinary stream through the ventral penile meatus.

Discussion

Urethral triplication or trifurcation is a congenital anomaly characterized by the presence of accessory urethra originating from the bladder, bladder neck or urethra, and opening externally at any position on the penis, perineum or the anorectum. A tract is said to be complete when the accessory tract opens externally or onto the distal part of the normal urethra. It is considered incomplete when the accessory tract is blind ending [1].

Many theories have been proposed to explain the embryology of an accessory urethra. These include abnormal termination of the Mullerian duct, failure of growth of the urogenital sinus, defective fusion of the mesoderm in the midline and continued splitting of the urorectal septum [2]. However, none of the theories was able to adequately explain the complete spectrum of urethral triplication. In 2008, Van der Putte [3] suggested that urethral triplication may be due to an abnormal division of the urogenital sinus. Currently, this theory has been accepted widely to explain the embryology of urethral triplication.

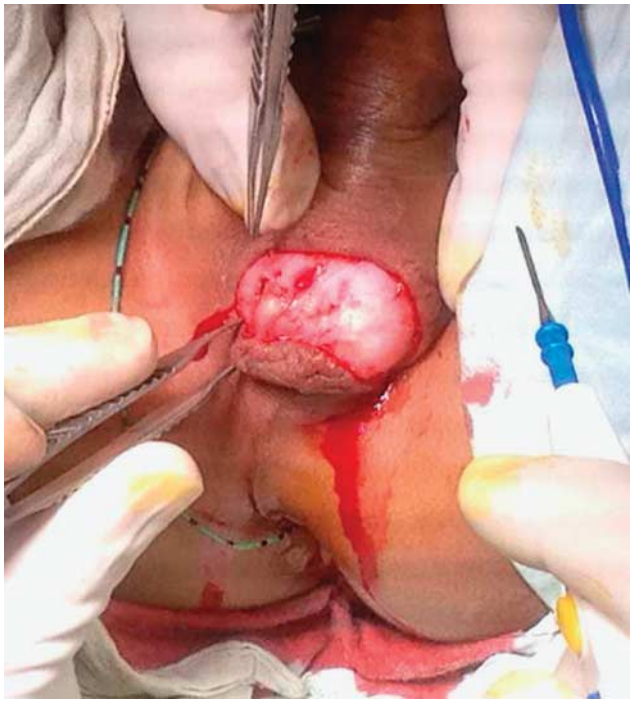
There are two broad categories of urethral triplication reported in the literature: three orifices ending on the penis or penoscrotum (anterior), or Y triplication with an anorectal or perineal opening. Clinical presentation depends on the anatomical subtype, which includes

1687-4137 © 2016 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000469366.65557.5e

Fig. 1

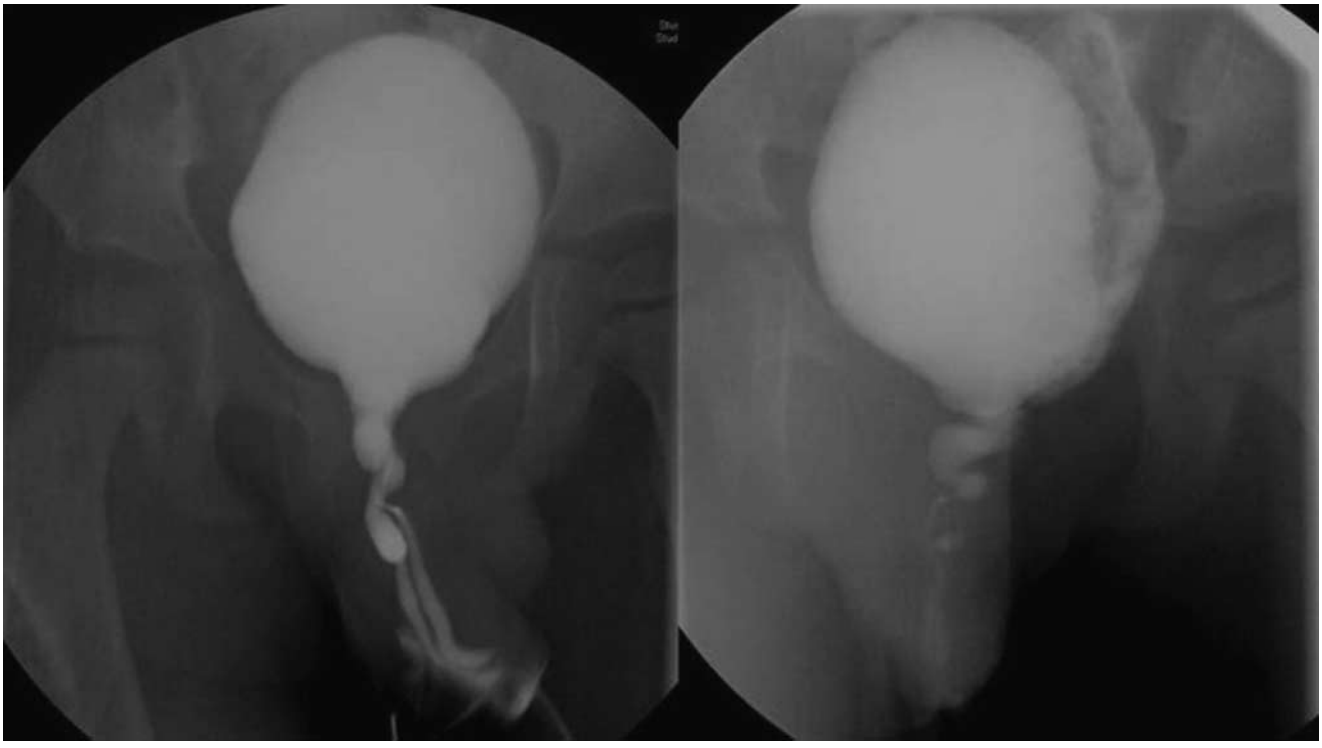
The tip of the phallus revealing two urethral openings

Fig. 3



Cannulated urethro rectal channel

Fig. 2



Two penile streams and rectum filling

urinary incontinence, urinary tract infection, multiple urinary streams, discharging sinuses, abnormal appearance or a primarily anal urinary stream as in our case. Radiological investigations such as voiding cystourethrogram and retrograde urethrogram are useful in delineating the anatomy. Urodynamic study may be helpful in

Table 1 Comparison among all urethral triplications in literature

References	Age of patient	Type	Size of urethra (Fr)			Associated anomalies
			Dorsal	Middle	Ventral	
Forgaard and Ansell [1]	22 years	Penile	Stenotic	16	Stenotic	Right pelvic kidney, left ureteral duplication, hemivertebrae, rib anomalies
Toguri *et al.* [4]	4 years	Y	Stenotic	8	–	
Zimmermann and Mildenberger [5]	28 days	Y	Stenotic	–	–	Anterior urethral diverticulum
Wirtshafter *et al.* [6]	46 years	Penile	3	3	26	Right dysplastic kidney, left duplex kidney
Schmeller and Schirmer [7]	15 years	Penile	10	8	18	Left solitary kidney, right undescended testes
Zattoni *et al.* [8]	48 years	Penile	Stenotic	Stenotic	Normal	Right renal agenesis, mild left hydronephrosis trabeculated bladder
Gülerçe *et al.* [9]	20 years	Penile	10	8	12	Left ureter duplication, right pelvic kidney
Hirselj *et al.* [10]	2 months	Penile	Less than 5	5	–	Tracheoesophageal fistula, vertebral anomaly, patent ductus arteriosus, horseshoe kidney
Kajbafzadeh *et al.* [2]	5 days	Y	3	3	–	Left multi cystic dysplastic kidney, anterior ectopic anus, bilateral vesico-ureteric reflux and ureterovasal reflux
Lin *et al.* [11]	17 months	Penile	5	3	8	No associated anomalies
Tourchi *et al.* [12]	6 years	Y	6	6	–	Caudal duplication
This study	5 years	Y	Less than 5	8	–	Fatty filum terminale, tethered cord, vertebral anomalies

determining the status of the bladder and the external sphincter in those presenting with incontinence.

Eleven cases reported in the literature are summarized to provide a comprehensive understanding of this rare lower urinary tract anomaly (Table 1) [1–2,5–12].

Determining the dominant functional urethra is a critical first step in the management of a patient with accessory urethra [12]. In the majority of cases of Y duplication of the urethra the ventral perineal meatus is dominant, whereas the dorsal penile meatus is hypoplastic. Hence, the ventral perineal meatus is preserved and lengthened by means of various methods to reach the glans penis. A previous study from our centre by Sinha *et al.* described the use of anterior rectal wall for posterior urethral lengthening [13]. However, in some instances of Y triplication the penile meatus may be of adequate calibre, which can serve as the functional urethra once the rectal opening is closed. Thus, determining the adequacy and functionality of the penile urethra in Y triplication can simplify and improve surgical outcome.

Conclusion

Urethral triplication is an extremely rare entity. Unlike a Y duplication in which the penile meatus is hypoplastic, a Y triplication can have a penile meatus of adequate calibre, which can be used as a functional conduit. Most patients with urethral triplication have been found to have associated malformations. To our knowledge, this is the first reported case of urethral triplication seen in association with fatty filum terminale, tethered cord and vertebral anomalies.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Forgaard DM, Ansell JS. Trifurcation of the anterior urethra: a case report. *J Urol* 1966; **95**:785–787.
2 Kajbafzadeh A-M, Taleb S, Montaser-Kouhsari L, Tanhaeivash R, Monajemzadeh M, Mehdizadeh M. Urethral triplication and urethrovasal reflux in 5-day-old male infant. *Urology* 2011; **78**:186–188.
3 Van der Putte SCJ. Duplication of Accessory Phallic Urethra (Urethra Triplication) in the Female, Signaling Mesenchymal Interruptions of the Cloacal Membrane as a Cause for Sagittal Urethral Duplications. *Pediatr Dev Pathol* 2008; **12**:487–492.
4 Toguri AG, Churchill BM, Rabinowitz R. Y-type urethral triplication. *J Urol* 1977; **118**:684.
5 Zimmermann H, Mildenberger H. Posterior urethral duplication and triplication in the male. *J Pediatr Surg* 1980; **15**:212–214.
6 Wirtshafter A, Carrion HM, Morillo G, Politano VA. Complete trifurcation of the urethra. *J Urol* 1980; **123**:431–432.
7 Schmeller NT, Schirmer HK. Trifurcation of the urethra: a case report. *J Urol* 1982; **127**:545–546.
8 Zattoni F, Gennari T, Rovasio A. Y-type urethral triplication. *Br J Urol* 1982; **54**:195.
9 Gülerçe Z, Nazli O, Killi R, Girgin C, Erhan O. Trifurcation of the urethra: a case report. *J Urol* 1992; **148**:403–404.
10 Hirselj DA, Lowe GK, Jayanthi VR. Urethral triplication with meatuses terminating on the penis: a rare case presenting with bifid urinary stream. *Pediatr Radiol* 2009; **39**:1227–1229.
11 Lin H, Shen X, Geng H, Xu M. Treatment and medical followup of a boy with urethral triplication. *Urology* 2012; **80**:214–215.
12 Tourchi A, Kajbafzadeh A-M, Khakpour M, Mohammadi Nejad P, Mousavian A-A, Kalantary M. Concomitant urethral triplication, bladder, and colon duplication. *Int Urol Nephrol* 2012; **44**:41–44.
13 Sinha S, Sen S, Chacko J, Thomas G, Karl S, Mathai J. Yduplication of the male urethra: use of anterior anorectal wall for posterior urethral lengthening. *Pediatr Surg Int* 2006; **22**:529–531.