Journal of Pediatric Surgery xxx (2017) xxx–xxx

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Urodynamic performance in boys with Y-type urethral duplication☆,☆☆,★,★★

Ye Zhang, Yanchao Qu, Lili Jiao, Weiping Zhang *, Ning Sun, Jun Tian, Minglei Li, Hongcheng Song

Department of Urology, Beijing Children's Hospital Affiliated to Capital Medical University, Beijing 100045, China

ARTICLE INFO

Article history:
Received 11 April 2017
Received in revised form 24 May 2017
Accepted 1 June 2017
Available online xxxx

Key words:
Urethral duplication
Y-type
Urodynamics

ABSTRACT

Purpose: The aim of this study was to elucidate the urodynamic features of patients with Y-type urethral duplication.
Methods: Patients with Y-type urethral duplication were retrospectively analyzed. Clinical presentation, urodynamic findings, surgical methods, and treatment outcomes were reviewed.
Results: From 2014 to 2016, six boys were diagnosed with Y-type urethral duplication at our institution. All patients underwent urodynamic testing. Urodynamic testing in patient 1 and 2 revealed detrusor pressure as 100 cmH2O and 88 cmH2O in the voiding stage, while urinary flow rate were 0 ml/s and 2.8 ml/s with volume of residual urine as 300 ml and 110 ml respectively, which consistent with the typical urodynamic of lower urinary tract obstruction. Patient 1, 3, 4 and 6 showed impaired bladder compliance as 7.5 ml/H2O, 12 ml/H2O, 6 ml/H2O and 6 ml/H2O respectively. Patient 5 and 6 also showed maximum urethral pressure as 110 cmH2O and 125 cmH2O with maximum urethral closure pressure as 103 cmH2O and 110 cmH2O respectively in the resting state.
Conclusions: Y-type urethral duplication is one potential cause of lower urinary tract obstruction, as seen in the abnormal urodynamic findings in our patients. Further studies are needed to elucidate the characteristics of this rare condition and determine optimal surgical management.
Type of study: Retrospective case series.
Level of evidence: Level 4 observational study without controls.

Urethral duplication is a rare congenital anomaly of the lower urinary system. There are several types of urethral duplication and various classification systems based on anatomical differences [1]. The most widely used classification is reported by Effmann [2], which is based on detailed radiological anatomy as seen on the urethrogram. According to Effman, Y-type urethral duplication (YUD) is a particular form of type IIA2 urethral duplication, with roughly 50 cases reported in the English literature [3] and 30 in Chinese. YUD is characterized by a stenotic dorsal urethra and a more functional ventral urethra, which opens at the perineum, perianal region, or in the anal canal. We present six cases of YUD, along with urodynamic findings, to elucidate the clinical characteristics of the condition and our management experiences.

1. Materials and methods

This study was approved by the internal review board of Beijing Children's Hospital.

We treated six boys with YUD between 2014 and 2016. Two of six patients urinated primarily through an external anal or perineal orifice and the others urinated primarily through the glanular meatus.

Clinical features, urodynamic findings, and case management details are shown in Table 1.

Abbreviations: YUD, Y-type urethra duplication; VCUG, voiding cysto-urethrogram; IVU, intravenous urography; PADUA, progressive augmentation by dilating urethra anterior; LUTO, lower urinary tract obstruction; VUR, vesicoureteral reflux; UTIs, urinary tract infections; MUP, maximum urethral pressure; MUCP, maximum urethral closure pressure; Pdet, detrusor pressure; Qmax, maximum flow rate; Qave, average flow rate; PVR, post-voiding residual urine; ml, milliliter; cmH_2O, centimeters H_2O; ml/H_2O, milliliters per centimeters H_2O; ml/s, milliliters per second.

☆ Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

☆☆ Informed consent: Informed consent was obtained from all individual participants included in the study.

★ Funding: This study was funded by The Beijing Hospital Authority "DengFeng" Talent Training Plan (DFL20151102), The Beijing Hospital Authority "Yangfan": Pediatric Urology (201709), and Capital Development Fund of Health (2014-2-2093).

★★ The authors declare that there is no conflict of interests regarding the publication of this paper. All authors have received grants from Beijing Children's Hospital.

* Corresponding author at: Department of Urology, Beijing Children's Hospital Affiliated to Capital Medical University, Beijing 100045, China. Tel.: +86 13370115012; fax: +86 10 59718700.

E-mail addresses: zhangye81@hotmail.com (Y. Zhang), qych_326@163.com (Y. Qu), 13691098857@163.com (L. Jiao), zhangwp2016@126.com (W. Zhang), sn-sunning@tom.com (N. Sun), tianjundr@163.com (J. Tian), dr.limінglei@foxmail.com (M. Li), songhch1975@126.com (H. Song).

http://dx.doi.org/10.1016/j.jpedsurg.2017.06.001
0022-3468/

Table 1
Clinical data of patients with Y-type urethral duplication.

Case	Age at diagnosis	Functional urethra	Primary surgery	Note	Associated malformation	Urodynamic results	Management
1	2 years	Penile urethra	Ligation and excision of VU	Clean intermittent self-catheterization for 10 years after surgery, dysuria, no urethrostenosis found through urethrocystoscopy	Unilateral renal malrotation combined hypoplasia	Compliance 7.5 ml/H2O, Pdet 100 cmH_2O,Qmax 0 ml/s, PVR 300 ml	Terazosin taken orally
2	22 months	Penile urethra	Urethra dilatation, ligation and excision of VU	Recurrent UTIs, YUD confirmed by VCUG and IVU	Grade III bilateral VUR	Compliance 34 ml/H2O, detrusor stable (filling stage), Qmax 2.8 ml/s, Pdet.Qmax 88 cmH_2O (voiding stage), PVR 110 ml	Progressive urethra dilatation, terazosin taken orally
3	16 months	Ventral urethra	Two-stage urethroplasty,	Surgery repair of urethral fistula and urethra dilatation	Anorectal malformation and polycystic renal dysplasia	Compliance 12 ml/H2O, detrusor stable (filling stage), bell-shaped uroflowmetry curve, Qmax 13 ml/s, Qave 8 ml/s	untreated
4	17 months	Ventral urethra	Two-stage urethroplasty	Surgery repair of urethral fistula and urethra dilatation	None	Compliance 6 ml/H2O, detrusor stable (filling stage), bell-shaped uroflowmetry curve, Qmax 11 ml/s, Qave 8 ml/s	Untreated
5	9 years	Penile urethra	Bilateral ureteric re-implantation, waiting for further operation for YUD	UTI, anal leakage of urine while voiding for 1 year, YUD confirmed by VCUG and ultrasound	Grade II–III bilateral VUR, duplex kidney	Compliance 16 ml/H2O, detrusor stable(filling stage), Qmax 11 ml/s, Qave 6.2 ml/s, Pdet.Qmax 47 cmH_2O, no PVR, MUP110 cmH_2O, MUCP 103 cmH_2O (resting state)	Untreated
6	2 years	Penile urethra	Ligation and excision of VU	Perineum leakage of urine while voiding since birth, YUD confirmed by VCUG and ultrasound	None	Compliance 6 ml/H2O, detrusor stable (filling stage), Qmax 6 ml/s, Qave 1.7 ml/s, Pdet.Qmax 39 cmH_2O, no PVR, MUP125 cmH_2O, MUCP 110 cmH_2O (resting state)	Untreated

All patients were diagnosed based on symptoms, physical examination, voiding cysto-urethrogram (VCUG), intravenous urography (IVU), and cystourethroscopy (Fig. 1). Renal ultrasonography and imaging of the spine and heart were performed to detect associated malformations [4,5].

Associated congenital malformations in our patients included anorectal malformation, renal mal-rotation with hypoplasia, polycystic renal dysplasia, duplex kidney, Y-type duplication of ureter, and bilateral vesicoureteral reflux.

Patient 1 was referred to our department with dysuria and involuntary urinary dribbling after excision of a dysplastic ventral urethra ten years ago at another hospital. Patient 2 experienced recurrent urinary tract infections and anal leakage of urine while voiding. Diagnostic testing revealed an abnormal urinary meatus located on the anterior wall of the anal canal at the level of the dentate line. Accessory ventral urethra was excised after progressive urethral dilatation. Vesicoureteral reflux and hydronephrosis went into spontaneous remission after surgery.

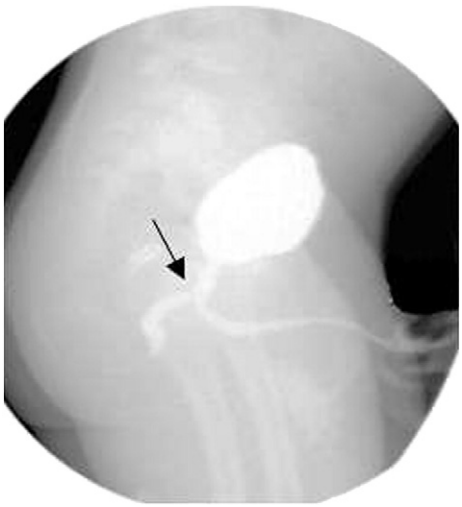

Fig. 1. Type IIA2 Y-type urethral duplication. Voiding cysto-urethrogram shows the ventral urethra coursing from the posterior urethra to the anal canal (arrow).

Patient 3 and 4 most urinated through their ventral urethra. VCUG revealed a Y-shaped urethra with a broad channel coursing from the posterior urethra to the anal canal. Both patients underwent two-stage urethroplasty. Patient 5 experienced urinary tract infection and anal leakage of urine while voiding with grade III bilateral vesicoureteral reflux. This patient underwent bilateral ureteric re-implantation firstly and waited for the further operation for urethral duplication. Patient 6 had a pinpoint-sized hole in the perineum, with urine leakage while voiding. After the hypoplastic ventral urethra was excised, he micturated normal with no uncomfortable feelings.

The complications in our patients were similar to those reported in literatures after urethroplasty [6]. Urethral fistula and urethrostenosis were diagnosed in patient 3 and 4, respectively, at 2-year-follow-up, no meatal stenosis or urethral diverticulum occurred. After surgical repair of the urethral fistula and urethra dilatation, all patients had a normally positioned glanular meatus with no skin chordee and all voided with a good stream.

2. Results

All patients underwent urodynamic testing during treatment (Table 1), the filling rate was between 5 ml and 8 ml per minute.

In patient 1, the detrusor pressure (Pdet) before detrusor contraction was 40 cmH_2O and the cystometric capacity was 330 ml. The bladder compliance was 7.5 ml/H_2O. Although the Pdet was higher than 100 cmH_2O during voiding, the patient failed to void and the residual urine volume was 300 ml (Fig. 2).

In patient 2, the compliance remained stable before detrusor contraction and the cystometric capacity was 170 ml. The bladder compliance was 34 ml/H_2O. During the voiding stage, the Pdet.Qmax was 88 cmH_2O and the Qmax was 2.8 ml/s. Post-void residual urine volume was 110 ml.

These two patients both had an unblocked orthotopic urethra confirmed with cystourethroscopy. With progressive urethral dilatation and oral-taken terazosin, voiding dysfunction in these patients improved during follow-up, as the urodynamic results showed that the Pdet at voiding stage dropped and the post-void residual urine decreased to less than 10 ml in both patients.

Please cite this article as: Zhang Y, et al, Urodynamic performance in boys with Y-type urethral duplication, J Pediatr Surg (2017), http://dx.doi.org/10.1016/j.jpedsurg.2017.06.001