African Journal of Urology
1110-5704

Vol. 12, No. 3, 2006
141-143

CONGENITAL URETHROPERINEAL FISTULA: REPORT OF 2 CASES AND ANALYSIS OF THE LITERATURE

H. BADAWY, M. YOUSSEF, A. SAAD, S. ORABI AND A. HANNO.
Section of Pediatric Urology, Department of Urology, Main University Hospital, University of Alexandria, Egypt.

INTRODUCTION

Urethral duplication in children has been described by Aristotle[1], and since then many classifications have been proposed to describe this rare anomaly both anatomically and functionally[2,3].The hypospadiac type of urethral duplication is a rare form of urethral duplication[4]. It is characterized by the presence of a functional ventral urethra opening in the perineum, anus or rectum[4]. Congenital urethroperineal fistula (CUPF) mimics the hypospadiac form of urethral duplication, but here the dorsal urethra is the functional one and the ventral urethra is just an accessory channel[4].

The treatment of hypospadiac urethral duplication aims at keeping the ventral functioning urethra intact and at mobilizing it to a normal position, while the treatment of CUPF simply consists of an excision of the ventral channel[4,5]. To ensure the right treatment and to avoid problems that will occur if the dominant urethra is not correctly identified it is mandatory to understand and recognize the two clinically similar but pathologically different, albeit rare, conditions.

We herein report on two cases of CUPF, their clinical presentation and management.

CASE REPORT

Two boys, 3 and 3.5 years old presented with an abnormal passage of urine from the perineum, together with micturition from a normal eutopic meatus. In addition, there was continuous perianal dribbling of urine shortly after the end of micturition. The younger child suffered from repeated febrile urinary tract infections.

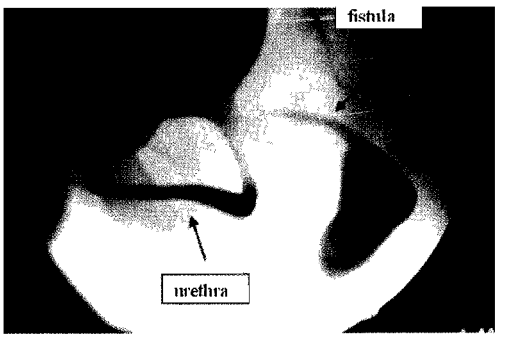

Fig.1: Voiding cystourethrogram of the 3-year-old child showing a tiny fistulous tract leading to a wide tract ending in the perineum again by a tiny fistulous tract

In both boys, a small opening about 1.5 cm anterior to the anal verge was identified, from which continuous dribbling of urine was noticed during micturition. Ultrasound of the upper urinary tract and bladder was completely normal. Voiding cystourethrography (VCUG) revealed a tiny fistulous tract communicating with the posterior urethra that opened in the perineal skin by a tiny fistulous tract, with an intervening dilated pouch between the two narrow segments (Fig.1)

In the younger boy, a grade II left vesicoureteral reflux was seen. Ascending urethro-

gram revealed a normal dorsal urethra and failed to opacify the fistulous tract, but the fistula was demonstrated by injection of contrast through the perineal orifice (fistulogram).

Cystourethroscopy revealed a normal dorsal urethra with a normal verumontanum and external urethral sphincter in both patients. Injection of methylene blue through the fistulous tract showed that the proximal orifice was connected to the upper part of the posterior surface of the posterior urethra just below the bladder neck.

In both boys, surgery was performed in the lithotomy position through an inverted perineal U-shaped incision. The fistula (with a lacrimal probe inside) was dissected up to its site of entry into the posterior urethra. The tract passed in close proximity to the rectum inferiorly, but deviated anteriorly and to one side (left in the first boy and right in the other) as it went cephalad to reach the floor of the prostatic urethra below the bladder neck . The fistulous tract was ligated and severed a few millimeters away from the posterior urethra. A 10 Fr urethral silicon catheter was placed for 7 days.

After 3 months, both children micturated normally with no evidence of recurrent fistula. VCUG revealed no residual fistulous tract (Fig. 2).

In both patients, histopathology of the fistulous tract revealed a normal transitional epithelium lining the entire surface of the fistulous tract except at the most distal part where it was lined by stratified squamous epithelium

DISCUSSION

Urethral duplication is a rare anomaly of the lower urinary tract encountered predominantly in male children[6].The Y-form of urethral duplication entails the presence of an accessory channel arising from the prostatic urethra and opening anywhere in the perineum, anal canal or rectum[2].

The main differences between the Y-form of urethral duplication and the CUPF is the

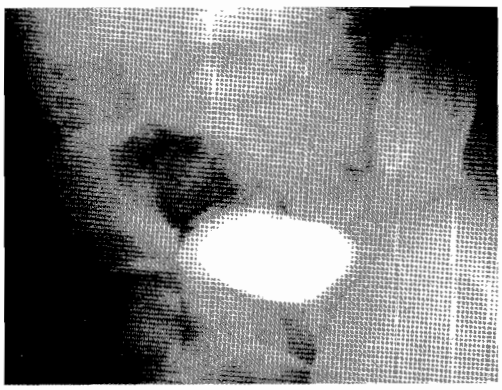

Fig. 2: Postoperative voiding cystourethrogram after 3 months showing absent fistula.

position of the functioning urethra and the urinary stream[4]. In the Y-duplication, the ventral urethra is the functional one, whereas in CUPF, the main urethra is the dorsal normally situated one[4].

Children with urethroperineal fistula usually present at an older age[7-9]. In the report of Bates and Lebowitz on four cases of CUPF, the children were 5,6 and 17 years old and only one child presented at the age of 7 months[4]. The two boys seen in our department were 3 and 3.5 years old.

Although in both our patients the diagnosis had already been established by imaging, cystourethroscopy was done to confirm the position of the functional urethra. Ultrasound of the upper tract revealed no anomalies in either of the two boys; however it has to be performed routinely owing to the possibility of associated upper tract anomalies[4,7]

The treatment of urethral duplication entails preservation of the dominant ventral urethra. The ventral urethra is mobilized and a preputial skin flap could be used as a tube to advance the meatus to its normal position. Alternatively, two-stage urethroplasty can be performed like a routine hypospadias repair[10]. On the other hand, the management of CUPF just consists of excision of the accessory channel [4,7-9].

An inverted U shaped perineal incision was done in both children because it gives more access to the perineum. A lacrimal probe inserted inside the fistula facilitated dissection and prevented injury to the rectum. Injury of the external sphincter during dissection was avoided by leaving the final few millimeters of the fistula attached to the prostatic urethra.

Recurrence of the fistula was previously reported by Bates and Lebowitz[4]. In the present report, the short stump that was left attached to the prostatic urethra was double ligated, and urethral diversion through an indwelling catheter for 7 days seemed sufficient to prevent recurrence.

Histopathologic examination of the excised fistulous tracts showed a transitional epithelium lining the whole lumen of the fistula denoting that it is originating from the lower urinary tract.

Whether CUPF is a special type of urethral duplication or a separate entity has not yet been determined clearly in the literature. Wagner et al.[7] reported three cases of CUPF and considered them a special type of urethral duplication, and this is reiterated by the report of Kumaravel et al.[9]. On the other hand, Bates and Lebowitz[4] in their report on four cases of CUPF considered this pathology a completely separate entity from the urethral duplication. This seems to be more plausible in view of the anatomical differences and the different surgical approach.

REFERENCES

1. Gross RE, Moore TC. Duplication of the urethra. Report of two cases and summary of the literature. Arch.Surg.1951;60:749-61.
2. Effmann EL, Lebowitz RL, Colodny AH. Duplication of the urethra.Radiology.1976 Apr;119(1)179-85.
3. Woodhouse CR, Williams DI. Duplications of the lower urinary tract in children.Br.J.Urol.1979 Dec;51(6)481-7.
4. Bates DG, Lebowitz RL. Congenital urethroperineal fistula.Radiology.1995 Feb;194(2)501-4.
5. Ortolano V, Nasrallah PF. Urethral duplication. J.Urol.1986 Oct;136(4)909-12.
6. Prasad N , Vivekanandhan KG, Ilangovan G, Prabakaran S. Duplication of the urethra.Pediatr. Surg.Int.1999 Jul;15(5-6)419-21.
7. Wagner JR, Carr MC, Bauer SB, Colodny AH, Retik AB, Hendren WH. Congenital posterior urethral perineal fistulae: a unique form of urethral duplication.Urology.1996 Aug;48(2)277-80.
8. Arda IS , Hicsonmez A. An unusual presentation of Y-type urethral duplication with perianal abscess: case report.J.Pediatr.Surg.2002 Aug;37(8)1213-5.
9. Kumaravel S, Senthilnathan R, Sankkarabarathi C, Bagdi RK, Soundararajan S , Prasad N. Y-type urethral duplication: an unusual variant of a rare anomaly.Pediatr.Surg.Int.2004 Dec;20(11-12)866-8.
10. Salle JL, Sibai H, Rosenstein D, Brzezinski AE, Corcos J. Urethral duplication in the male: review of 16 cases.J.Urol.2000 Jun;163(6)1936-40.

Editorial Comment:

The authors of this article must be commended on their fine work. The article gives a concise, yet complete clinical, radiological, histological and surgical description of their two index cases. They base their findings within the context of the small body of world literature on these fistulas and they usefully emphasize the hazards of failing to appreciate the clinical difference between these fistulas and the "Y-type" duplication.

Dr. John Lazarus
Red Cross Children's Hospital, Cape Town, South Africa

Corresponding Author:
Dr. Haytham BADAWY
Unit of Pediatric Urology,Department of Urology,
Faculty of Medicine, University of Alexandria, Egypt.
Email: hithamalmetwale@yahoo.com