Case Report

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Urethral duplication in a 12-year-old child

Faustin Félicien Mouafo Tambo, Jacques Birraux[1], Ambroise Wonkam[2], Zacharie Sando[3], Yvonne Walburga Joko[2], Pierre-Yves Mure[4], Claude Le Coultre[1], Gervais Ondobo Andze, Maurice Aurélien Sosso[5]

ABSTRACT

Urethral duplication is a rare congenital malformation affecting mainly boys. The authors report a case in a Cameroonian child who was diagnosed and managed at the Gynaeco-Obstetric and Paediatric Hospital, Yaounde. The malformation was characterized by the presence of an incontinent epispadic urethra and a normal apical urethra. We describe the difficulties faced in the management of this disorder in a developing country.

Key words: Child, duplication, urethra

INTRODUCTION

Duplication of the urethra is a rare malformation[1–7] with important anatomical polymorphisms. Different classifications have been proposed. Until 2009, fewer than 400 cases have been described in literature.[3,4,8,9] In sub-Saharan Africa its prevalence is unknown, and very few cases have been reported. We describe a new case diagnosed and managed at the GOPHY (Cameroon). To the best of our knowledge, this is the first documented case of a urethral duplication operated in Yaounde. In this case report we present our experience in managing this congenital malformation in the context of a developing country.

Department of Surgery and Subspecialties, Paediatrics Surgical Unit, Yaounde Gynaeco-Obstetric and Paediatrics Hospital (YGOPH), Yaounde-Cameroon, [1]Department of Paediatrics Surgery, Paediatrics Surgical Unit, Children Hospital, 6 street Willy-Donzé, 1211 Geneva-Switerland, [2]Department of Morbid Anatomy and Morphologic Sciences, Genetic unit, [3]Department of Morbid Anatomy and Morphologic Sciences, Pathology Unit, Yaounde Gynaeco-Obstetric and Paediatrics Hospital (YGOPH), Yaounde-Cameroon, [4]Department of Paediatrics Surgery, Paediatrics Surgical Unit, Université Claude Bernard – Lyon I, GHE-HFME, 253 BD Pinel, 69500, France, [5]Department of Surgery and Subspecialties, Faculty of Medicine and Biomedical Sciences, University of Yaounde I, Cameroon.

Address for correspondence:
Dr Faustin Félicien Mouafo Tambo,
Department of Surgery and Subspecialities, BP4362 Yaounde, Cameroon
E-mail: faustintambo@yahoo.fr

CASE REPORT

The child, KLMG, 12 years old, the first child in a family and living in poor socioeconomic conditions, was seen during a specialised consultation for children with malformations of the external genitalia at the GOPHY. This boy presented with continuous urine leakage through the dorsal urethra since the age of 2 months when he was circumcised. His birth weight was 2500 g. At 3 months he had seizures and had been treated with antiepileptic drugs but had, as sequelae, psychomotor developmental delay, failure to thrive, and left hemiparesis.

Physical examination revealed a child with left hemiparesis, an incontent anus and scoliosis. The external genitalia were well developed. The penis had a normal apical meatus, through which urine occasionally passed, and an incontinent epispadic urethral meatus through which urination mostly occurred [Figures 1 and 2].

The paraclinical evaluation revealed 46,XY karyotype. The voiding cystourethrogram [Figures 3-5] showed a complete urethral duplication, with insertion of the epispadiac urethra at the level of the bladder neck. Renal ultrasound revealed bilateral hydronephrosis. This was of concern to us since the patient was incontinent. Radiography of the pelvis revealed a major pubic disjunction and agenesis of the coccyx. [Figure 4].

Surgical intervention began with endoscopy. Using a paediatric cystoscope we were able to visualise the entire length of the epispadic urethra, right up to its insertion at the bladder neck; the normal urethra, which was of good calibre; and the bladder neck, which looked normal. The bladder appeared normal, with normally positioned ureteral openings, though the right ureteral orifice was dilated.

The two urethras were catheterised with size 6 and size 10 probes, respectively [Figure 6]. Then, a subcoronal

incision was made and complete stripping of the layers of the penis was done in order to permit safe dissection. The dorsal neurovascular bundle was isolated [Figure 7]. A circumferential incision was done around the epispadic urethral meatus and the latter dissected through the glans and then up to the base of the penis [Figure 8]. The epispadic meatus was dissected through the glans, up to the base of the penis, and was sutured at this point using PDS 7-0 absorbable stitches.

The postoperative course was uneventful, and the transurethral catheter was removed on the 10th day. The urinary incontinence completely stopped following surgery. Pathological examination revealed a tubular structure with a lumen bordered by well-differentiated squamous epithelium resting on a fibrous chorion,



Figure 1: The normal apical urethral meatus



Figure 2: Micturition through the epispadic urethral meatus



Figure 3: VCUG lateral view

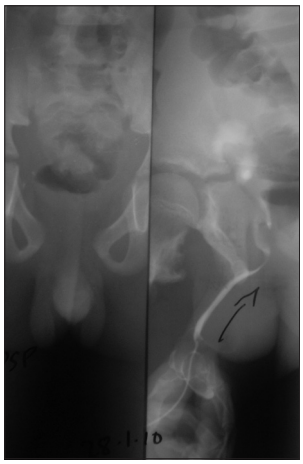


Figure 4: Pubic disjunction (left) and VCUG (right)

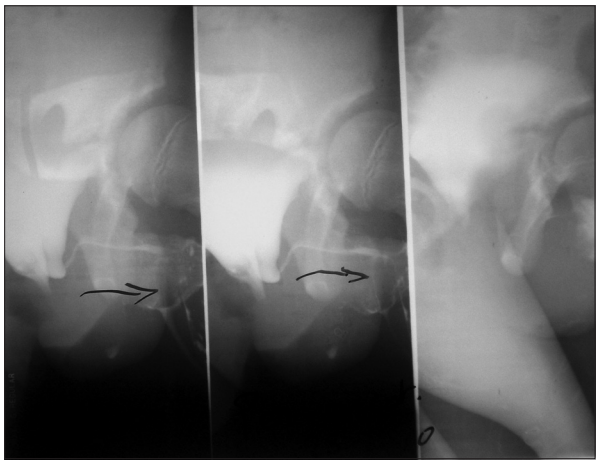


Figure 5: VCUG lateral view

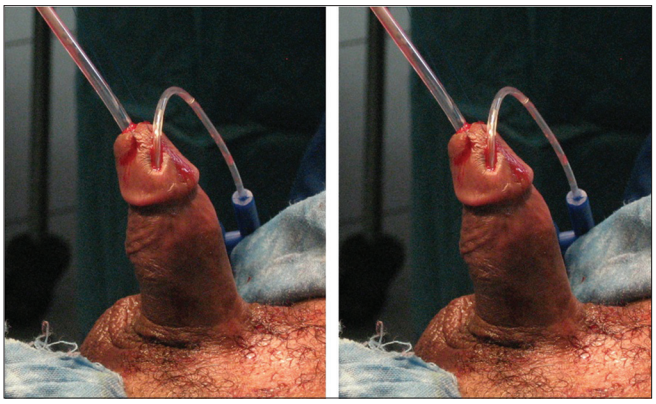


Figure 6: The catheterisation of both urethras



Figure 8: Circumferential incison with exposure of the base of the penis

consistent with a urethra.

DISCUSSION

Urethral duplication, first described by Aristotle,[1] is one of the rarest malformations of the urinary tract. It most often affects boys, as in this reported case. Only 35 cases have been described in girls.[5] Its clinical presentation is varied, and many classifications have been proposed, of which the most commonly used is that of Effmann[6] [Figure 9]. The Effmann classification is anatomical, but it does not permit differentiation of the sagittal from the lateral forms of this duplication.

Our patient had features consistent with an Effmann type IIA10,[6] with two urethras that were completely separate, from their origin at the bladder up to the distal meatus. However, there is a complex association of malformations in this case, with a incontinent anus, agenesis of coccyx, and disjunction of the symphysis pubis, similar to what is described in the epispadias-bladder exstrophy complex by Pippi Salle.[4] The age of the child is also remarkable when compared to the 29-month average observed in Salle's study.[4] This is partly explained by the family's poor socioeconomic conditions. However, the patient's neurological status

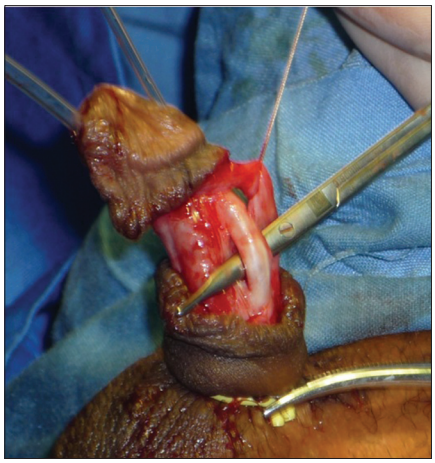


Figure 7: Identification and isolation of the neurovascular bundle

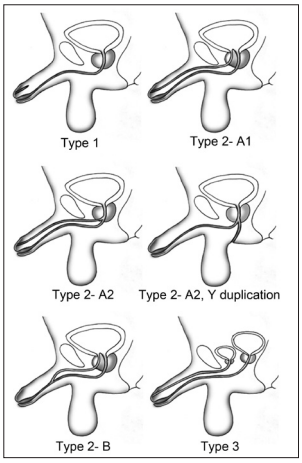


Figure 9: The Effmann classification

also certainly contributed to the neglect of the classic symptoms of incontinence; the incontinence was likely accepted as part of the neurological problem without any attempt being made to exclude potentially curable causes.

Diagnosis of this condition is based on simple clinical examination of the penis and is confirmed by a voiding cystourethrography, which will permit a description of its anatomical type. The ultrasound looks for abnormalities classically associated with urethral duplication, and comorbidity as upper urinary tract dilation as observed in our case. Most authors are of the opinion that surgery is necessary, especially when the urethral duplication is symptomatic. We started our surgical intervention with a cystoscopy, which confirmed that the apical urethra was normal and also showed the presence of the veru montanum. The excision was then performed using a single incision, with complete stripping of the penis. Pshramis,[10] on the other hand, has proposed a double approach: perineal

and pre-pubic.

The injection of a sclerosing agent into the epispadic urethra, as described by Acimi,[11] seems dangerous to us because of the risk of fibrosis of the corpus cavernosa.

A favourable postoperative course is described classically, as was also observed in our case despite the various difficulties faced in our setting.

CONCLUSION

Urethral duplication is a rare disorder A cystography is crucial for determining the anatomical type and hence its classification. Surgery is indicated when the child is symptomatic, and the postoperative course is uncomplicated in most cases. The paediatric surgeon must always remember that the accessory urethra is usually situated dorsal to the functional urethra regardless of the position of the meatus.

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REFERENCES

1. Arena S, Arena C, Scuderi MG, Sanges G, Arena F, Di Benedetto V. Urethral duplication in males:our experience in ten cases. Pediatr Surg Int 2007;23:789-94.
2. Bieganski A, Launay S, Biserte J, Robert Y. Urethral duplication type II in a male child: A case report. J Radiol 2002;83:650-2.
3. Tillig B, Moritz RP, Muller W, Jaeger HD. Duplications of the male urethra two cases reports. Eur J Pediatr 1999;9:189-92.
4. Pippi Salle JL, Sibai H, Rosenstein D, Brzezinski AE, Corcos J. Urethral duplication in the male: Review of 16 cases. J Urol 2000;163:1936-40.
5. Cost NG, Lucas SM, Baker LA, Wilcox DT. Two girls with urethral duplication. Urology 2008;72:800-2.
6. Effmann EL, Lebowitz RL, colodny AH: Duplication of the urethra. Radiology 1976;119:179-85.
7. Kumar A, Goyal NK, Trivedi S, Dwivedi US, Singh PB. Y-duplication of the urethra: A rare case report. Internet J Surg 2007;11.
8. Ouattara Z, Keita M, Doumbia D, Mari*ko A,* Tembely A, Ouattara K. Duplication de l'urètre chez le garçon à propos d'un cas. Mali Med 2005;3;37-9.
9. Rane SB, Obiadah A, Dhende NP, Arlikar J, Achrya H, Thakur A, *et al*. Urethral duplication in children: Our experience of eight cases. J Pediatr Urol 2009;5:363-7.
10. Pshramis KE, Colodny AH, Lebowitz RL, Retik AB, Bauer SB. Complete patent duplications of the urethra. J Urol 1986;136:63-7.
11. Acimi S, Haouari H, Malah N. Duplication de l'urètre chez le garcon. Prog Urol 2003;13:698-9.

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