Total reconstruction of the urethra in boy with urethral duplication

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ABSTRACT

Introduction: Urethral duplication is a very rare congenital abnormality. Its clinical presentation varies according to its anatomical variant. It can be localized from the bladder neck to the distal part of the urethra and it can be complete or incomplete. It is most common at the sagittal plane with a ventral and dorsal urethra, the ventral urethra being the more functional. Management is generally complex and will depend on the type of urethral duplication. The case of a boy with Effmann classification Type II-AY urethral duplication is presented here.

Objective: To demonstrate total reconstruction of the urethra with oral mucosa graft and transverse dorsal preputial flap (Duckett technique) as definitive treatment method in patients with urethral duplication.

Clinical case: In a single surgical operation two approaches were carried out. First, anterior sagittal approach was used to dissect the urethra at the level of the prostatic urethra, isolating the ventral, or functional, urethra and anastomosing a six centimeter tubular oral mucosa graft up to the base of the penis distally at the midline. This was then followed by supine decubitus approach, degloving the prepuce and exposing the axis of the penis at its base. A seven centimeter tubular transverse dorsal preputial flap was proximally

RESUMEN

Introducción: La duplicación de uretra es una anomalía congénita muy rara; su presentación clínica varía de acuerdo a su anatomía variante, pudiéndose localizar desde el cuello vesical hasta la parte distal de la uretra, puede ser completa e incompleta y es más común en el plano sagital con una uretra ventral y dorsal, siendo más funcional la parte ventral. Generalmente el manejo es complejo y dependerá del tipo de duplicación uretral. Se presenta el caso de un niño con duplicación uretral tipo II-AY según la clasificación de Effmann.

Objetivo: Demostrar la reconstrucción total de uretra en pacientes con duplicación uretral como método definitivo en estos tipos de pacientes con injerto de mucosa oral y toma de colgajo dorsal transverso prepucial; técnica de Duckett.

Caso clínico: Se operó por dos vías en el mismo acto quirúrgico, por vía sagital anterior para abordaje y disección de la uretra a nivel de la uretra prostática aislando la uretra ventral o funcional y anastomosando un injerto tubular de mucosa oral de seis centímetros de largo hasta la base del pene en la línea media distalmente, acto seguido abordaje en posición decúbito supino, degloviendo el prepucio y exponiendo el eje del pene hasta su base y mediante colgajo dorsal transverso...
anastomosed to the oral mucosa graft up to the external urethral neomeatus at the tip of the glans. A 12 Fr urethral splint was left in place for twenty-one days. The urethral splint was removed on the twenty-first postoperative day, and micturition test was satisfactory. Cystotomy was left in place and removed after six months. Patient presently is without cystotomy catheter and has transurethral micturition. Vesicoureteral reflux was corrected and he continues to be under out-patient surveillance.

**Keywords:** Total reconstruction of the urethra, urethral duplication, Mexico.

**INTRODUCTION**

Urethral duplication is a very rare congenital alteration and its anomalies are very complex. The variations with which it presents are thought to be due to different embryological causes of uncertain etiology. Its management does not always require surgery. Surgical cases can be carried out by advancing the ventral urethra within the perineum followed by urethroplasty. 1,2 Around 188 cases have been reported in the literature and this condition tends to be more frequent in males. 3 Casselman and Williams proposed that embryologically, urethral duplication should be due to a partial failure or irregularity in lateral mesoderm penetration over the ectoderm and endoderm layers of the cloacal membrane at the midline, with a dorsal epispadic canal. 4 Das and Brosman stated abnormal termination of the Mullerian duct as being responsible for urethral duplication 5 and Rice suggested asymmetry in urorectal septum closure, resulting in urethroperineal fistula. 6 Effman has classified urethral duplication into 3 types:

- **Type I:** incomplete urethral duplication (A: distal / B: proximal)
- **Type II:** complete urethral duplication (A1: two different urethras that do not connect with one another upon exiting the bladder and open as 2 urethral meatuses / A2: a second urethra exits that derives from the first urethral canal with a different urethral meatus / A2-Y: two openings of the urethral meatus at different planes, B1: two urethras exit the bladder joining at the posterior urethra and exiting as a single urethral meatus.
- **Type III:** urethral duplication with partial component or complete caudal duplication. 7

Patient is an 11-year-old boy with Type II-AY urethral duplication with bilateral grade III vesicoureteral reflux. Oral mucosa graft was placed 8 anastomosing its proximal portion to the prostatic urethra and its distal portion to the proximal end of the preputial dorsal flap up to the glans in a single surgery. Bilateral Hendren-type reimplantation was carried out 3 months after surgery.

**CASE PRESENTATION**

Patient is an 11-year-old boy with past medical history of multiple hypospadias surgeries, and cystotomy and diversion at 3 months of age. Patient presented with micturition through the perineum from a perineal meatus at the midline and in front of the rectum (Figure 1). Evaluation included micturition cystourethrogram that revealed double urethra at sagittal plane divided at the prostatic urethra. Dorsal urethra had distal extension up to external urethral meatus with hypoplasia. Ventral urethra went from prostatic urethra downwards in the perineum and in front of the rectum and was the functional one. Patient also presented with bilateral grade III vesicoureteral reflux (Figures 2 and 3).

**Cystoscopy:** permeable perineal ventral urethra that extended up to the prostatic urethra and continued up to the bladder neck and bladder. The 8 Fr cystoscope sheath did not pass through the hypoplastic dorsal urethra.

**Excretory urography:** moderate dilatation of the bilateral pyelocaliceal system.
Laboratory work-up: full blood count with Hb 12.3 m/dL, Ht 34.2%, leukocytes 6250/mL, platelets 320,000/mL. Blood chemistry: glucose 70 mg/dL, creatinine 0.8 mg/dL, urea 30.2 mg/dL, sodium: 142 mEq/mL, and potassium: 3.5mEq/mL.

Micturition cystography: double urethra in sagittal plane divided at the prostatic urethra: dorsal urethra with distal extension up to external urethral meatus with hypoplasia and ventral urethra from the prostatic urethra downwards in the perineum and in front of the rectum; this was the functional urethra. In addition bilateral grade III vesicoureteral reflux was revealed with no apparent bone alterations in the sacrum.

Treatment sequence: A 6 cm free oral mucosa graft was taken and was tubularized, leaving it prepared for anastomosis (Figures 3 and 4). With patient placed in jack-knife position graft anastomosis to prostatic urethra was carried out, adding transverse pedunculated preputial flap to complete the neourethra (Figures 5 and 6). Three weeks after surgery, and with no transurethral catheter, adequate transurethral flow was observed (Figure 7).
CONCLUSIONS

Urethral duplication is a very rare and complex anomaly and the existing anatomical complexity represents a great challenge in regard to surgical management. Presented here is the case of a boy with Type II A-Y urethral duplication with ventral urethra extending from prostatic urethra to perineum that was successfully corrected with total urethral reconstruction with 6 cm oral mucosa graft and 7 cm preputial flap. The present complex case is very rare and was successfully managed with the use of oral mucosa graft resulting in an ideal alternative for these types of cases.

BIBLIOGRAPHY