



Urethral Duplications in Children – A Case Series

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Abstract

Background: Urethral duplications are rare congenital anomalies with multiple anatomical variants. They mostly occur in the sagittal plane and can be associated with other congenital urogenital malformations. According to Effmann urethral duplications are classified into incomplete (type I), complete (type II), and complete associated with caudal duplication (type III).

Methods: We hereby describe three cases of urethral duplications and their respective treatment.

Results: Two cases were type I duplications. One patient was treated by complete resection of the blind ending secondary urethra, whereas no treatment was performed in the second patient. The third case was a IIA-2 Y-subtype duplication that needed complex reconstruction.

Conclusions: There is no need for surgical management in asymptomatic patients with type I urethral duplications. Type IIA-2 Y-subtype duplications are extremely rare, and their treatment must be individualized according to anatomical and physiological features.

Keywords: Urethral duplication; Y-duplication; Urethral anomaly

Introduction

Urethral duplications are seldomly seen urinary tract variants consisting of aberrant partial or complete development of an additional urethra [1]. They may be associated with different urogenital abnormalities, including bladder duplication and bladder exstrophy [2]. The etiology of this disease is not well understood. In the majority of cases, urethral duplications occur in the sagittal plane with the accessory urethra located dorsally to the orthotopic urethra [3]. The most commonly applied classification according to Effmann et al. divides urethral duplications into incomplete duplications (type I), complete duplications (type II), and duplications as a component of partial or complete caudal duplication (type III) [4]. Types I and II are further subdivided (Table 2) [5]. Whereas Type I duplications are typically clinically silent without urinary discharge or infections of the accessory urethra, complete duplications may be the cause for urinary incontinence and repeat urinary tract infections.

The aim of this case series is to describe the clinical presentation and respective treatment of this rare disease in three male children suffering from different types of urethral duplication.

Materials and Methods

We hereby present three male patients born between September 2008 and November 2013 with urethral duplications (Table 1). Thereof, two children had type IA duplications according to Effmann et al [4]. In one child, the ectopic urethra was successfully excised, whereas there was no treatment in the other child who also had Down syndrome. The third child presented with IIA-2 Y-subtype urethral duplication which consisted of a dystopic urethral orifice into the anus and a dysplastic dorsal urethra. A three-staged procedure was performed to restore the anatomy. All children were treated at two pediatric surgery centers in Switzerland (the University Hospital of Bern and the Cantonal Hospital of Aarau).

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Figure 1: Type 1A urethral duplication. Unrepaired blind ending secondary orifice in the axial plane lateral to the orthotopic urethra.

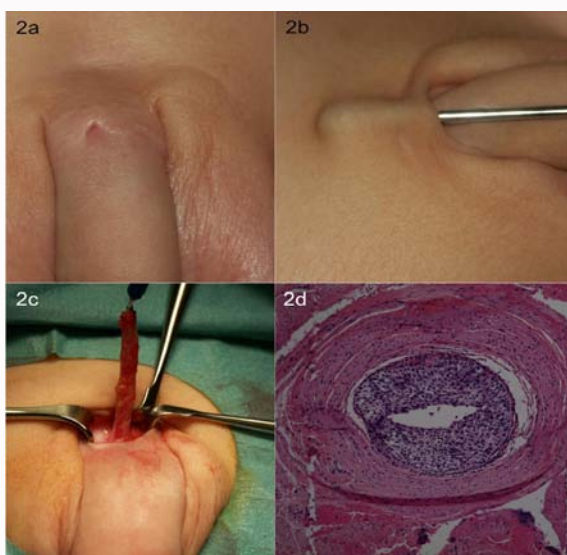


Figure 2: Type 1A urethral duplication. (A) Preoperative image of accessory pore originating 2 cm proximal to the orthotopic urethra. (B) Sounding of pore and marking of secondary urethra with betadine injection. (C) Complete accessory urethral resection (D) Histology showing fistula tract with urothelium and low-grade chronic inflammation.

Results

Case 1

Case 1 describes a boy with Down syndrome who suffered from spontaneously resolving Myelodysplastic Syndrome (MDS) in the neonatal period. A secondary urethral orifice was found incidentally by the child's paediatric oncologist at 5 years old (Figure 1). No urinary tract or local infections were noted. Ultrasound showed an unaffected bladder without wall thickening, as well as proportionately sized and normally structured kidneys and ureters. Micturating cystourethrogram demonstrated an anatomically positioned penile urethra arising from the common bladder neck. No vesicoureteral reflux was noted, and the bladder could be fully emptied. The secondary orifice was blind ending without connecting to the primary urethra or the bladder, corresponding to Type IA (Table 2). Because it was a short, incomplete and hence asymptomatic duplication, surgical excision was not performed in accordance with the child's mother.

Case 2

Case 2 describes a boy with a blind-ending accessory urethral

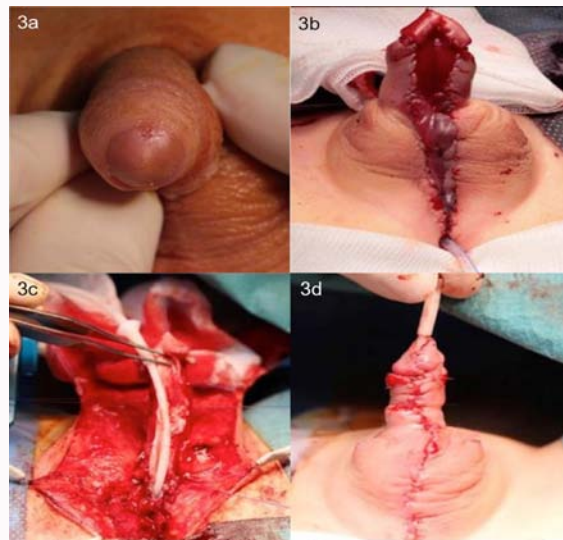


Figure 3: Urethral duplication IIA-2 Y-subtype repair. (A) Preoperative picture of hypoplastic, nonfunctional dorsal glandular meatus. (B) Resection of hypoplastic distal urethra and urethral bed substitution using a pedicle preputial flap, according to Bracka et al [7]. (C) Tubularization of graft, insertion of urethral stent, and correction of scrotum bifidum with scrotal transposition (D). Completed urethroplasty.

orifice incidentally discovered at an inguinal hernia repair follow-up when he was 10 months old (Figure 2A). The boy was asymptomatic, without discharge of urine from the accessory orifice, and otherwise healthy. Ultrasound revealed a blind ending secondary urethra 2 cm proximal to the orifice, along with unaffected intra and retroperitoneal organs. Eleven months after first recognition, complete circumcision due to symptomatic phimosis in combination with excision of the secondary urethra was performed. The pore was sounded (Figure 2B), and betadine solution was injected to mark the course of the secondary urethra. The pore was closely cut down to the orthotopic urethra and resected completely (Figure 2C). The operation was performed in an outpatient setting. Histologic examination confirmed a fistula tract with urothelium and low-grade chronic inflammation (Figure 2D). Postoperative follow-up 1.5 months after excision showed complete wound healing without relapse.

Case 3

Case 3 presented as a newborn boy with severe pyelonephritis. The patient passed urine rectally with only a few drops from the dorsal glandular meatus (Figure 3A), concomitant with IIA-2 Y-subtype urethral duplication. Further diagnostic work-up (renal ultrasound, micturating cystourethrogram, Tc-99m dimercaptosuccinic acid renal scintigraphy, and magnetic resonance urography) showed right-sided grade 4 vesicoureteral reflux with 10% function of the right kidney. Unilateral right-sided loop cutaneous ureterostomy [6] was performed without complication at 3 months old allowing retrograde urination from the bladder, and the child was discharged 4 days later after an uneventful post-operative course.

Separation and mobilization of the ventral urethra was performed at 14 months old using an anterior sagittal trans-anorectal approach. The hypoplastic distal urethra was resected, and urethral bed substitution using a pedicle preputial flap (first stage of the two-stage repair according to Bracka [7]) was performed (Figure 3B). Surgery was uneventful. At 1.5 years old, the second stage of urethral reconstruction, consisting of tubularization of the graft, was done

Table 1: Patient characteristics and management.

Case	Sex	Presenting age (months)	Recognition	UTIs	Imaging	Effmann type	Associated diseases	Therapy	No. of surgeries	Age at 1 st Surgery (month)
1	male	60	Pediatric oncologist (follow-up after MDS of the neonatal period)	No	Ultrasound, VCUG	IA	Down syndrome, MDS (of the neonatal period)	No	0	-
2	male	10	Pediatric surgeon (follow-up after inguinal hernia repair)	No	Ultrasound	IA	No	Complete excision	1	21
3	male	0 (at birth)	midwife	Yes	Ultrasound, VCUG, renal scintigraphy	IIA-2Y	Right-sided kidney failure, crossed dystopia of the kidney (left to right)	Three-staged procedure	3+2	3

UTI = Urinary Tract Infection, MDS = Myelodysplastic Syndrome and VCUG = Voiding Cystourethrogram

Table 2: Effmann classification of urethral duplications.

Type I	Incomplete duplication
Type IA	Distal incomplete duplication
Type IB	Proximal incomplete duplication
Type II	Complete duplication
Type IIA	Complete duplication, 2 meatuses
Type IIA-1	Complete duplication, 2 meatuses, 2 non-communicating urethras arising independently from the bladder
Type IIA-2	Complete duplication, 2 meatuses, second channel arising from the first urethra
Type IIA-2 'Y-subtype'	Complete duplication, 2 meatuses, second channel arising from the first urethra and courses into a second meatus that may open posteriorly into the rectum
Type IIB	Complete duplication, 1 meatus
Type III	Urethral duplication as a component of complete or partial caudal duplication

together with right-sided nephroureterectomy due to worsening of right-sided kidney function to less than 10% (Figure 3C and 3D). The scrotum bifidum was also corrected with scrotal transposition, a urethral stent was inserted, and a suprapubic catheter was placed.

The patient was anuric during the immediate postoperative period, so emergent abdominal ultrasound was performed. This showed an intraperitoneal urinoma together with pyelectasis of the remaining left kidney. Emergent reoperation revealed a left-sided ureteral injury by virtue of crossed dystopia of the kidney (left to right) that was not diagnosed before this operation. The ureter was mobilized to the left side, and ureteroneocystostomy using the psoas hitch ureteral reimplantation technique [8] was undertaken. Voiding cystourethrogram performed 18 days after the second stage of urethral reconstruction showed a small leakage at the middle section of the penile urethra. The suprapubic catheter was hence left in place. Repeat voiding cystourethrogram one week thereafter showed no fistula and a normal sized urethra, so the suprapubic catheter was removed. The subsequent five-month follow-up showed good cosmetic and functional results without any complications.

Discussion

Urethral duplication reflects a very rare urogenital malformation that occurs more frequently in males than females. Only a few more than 325 cases have been reported in the literature to date [9]. They typically occur in the sagittal plane [10] and consist of squamous epithelia surrounded by smooth muscle identical to the orthotopic urethra. The aetiology of urethral duplication is still not clear and is attributed to altered embryogenesis. The most commonly quoted hypothesis is the one of Patten and Barry, which purports a misalignment of the lateral folds of the genital tubercle and the

ventral end of the cloacal membrane. Other authors describe urethral duplications as due to Mullerian duct termination and growth arrest of the urogenital sinus [10]. However, no single theory is able to explain all different subtypes of urethral duplication. Different anatomical variants of urethral duplication led to several different classification systems [11], with that proposed by Effmann and colleagues being the most used [3]. Effmann divides urethral duplications into three different types (Table 2). In a series of pediatric urology case reports, the relative frequency of type I, type II, and type III duplications was 25%, 63%, and 12%, respectively [9]. In the present case series, two type IA cases with blind ending accessory urethras and one IIA-2 Y-subtype are shown.

Most blind-ending accessory urethras are clinically asymptomatic and rarely associated with other anomalies. If symptoms exist, they may include sinus tract infection and mucus discharge. One child with type IA duplication in the present study was otherwise healthy, while the other child had Down syndrome. Both duplications were incidentally noted by health professionals during physical examination. There's no need for therapeutic management in asymptomatic patients apart from cosmetic issues. Accordingly, the child with type IA duplication and symptomatic phimosis was treated by complete excision of the blind ending urethra, whereas the other child with Down syndrome was not treated.

In contrast to type I duplications, type II and type III duplications are associated with clinical symptoms including a doubled urinary stream, urinary incontinence, recurrent urinary tract infections, bladder outflow tract obstruction, and passing urine per anus with voiding [3,9,12-14]. Type II and III duplications occur along with more severe congenital anomalies. A variant of type IIA-2 duplication,

referred to as the 'IIA-2 Y-subtype', describes a secondary urethra coursing into a second meatus that opens posteriorly into the rectum/perineum [1]. This type is extremely rare with less than 20 cases being reported until the year 2000 and is associated with hypospadias, epispadias, cleft palate, congenital heart disease, tracheoesophageal fistula, and imperforate anus [9,10,14]. Type III, which describes complete or partial caudal urethral duplication, occurs most often with bladder and/or penis duplication, as well as the afore-mentioned abnormalities [3].

These duplications (type II and III) are hence typically diagnosed directly at birth [15]. Correspondingly, the child presenting with the IIA-2 Y-subtype duplication in this study showed repeated urinary tract infections, as well as higher grade vesicoureteral reflux with reduced kidney function. Surgical treatment was necessary to relieve the kidney and to restore anatomy. A three-step procedure was thereby chosen consisting of (i) unilateral loop cutaneous ureterostomy at 3 months old, (ii) separation and mobilization of the ventral urethra at 14 months old, and (iii) urethral reconstruction at 18 months old.

Conclusion

In general, there is no need for therapeutic management in asymptomatic patients with type I urethral duplications apart from cosmetic issues. Type IIA-2 Y-subtype duplications are extremely rare, and their treatment must be individualized according to anatomical and physiological features. Vesicoureteral reflux and repeated urinary tract infections are common co-findings of this subtype of urethral duplication.

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