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Urethral duplication in girls: Three cases associating an accessory epispadiac urethra and a main hypospadiac urethra

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Summary

Introduction
Urethral duplication is extremely rare in girls, with less than 40 cases reported so far. Most of them present as a prepubic sinus. Literature is scare regarding aetiology, classification and management in other forms. This study presents three cases of sagittal urethral duplication in girls presenting a main hypospadiac urethra and an accessory epispadiac urethra.

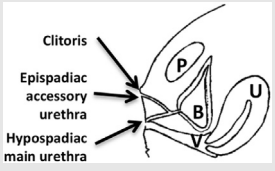
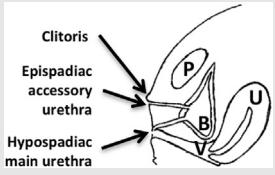
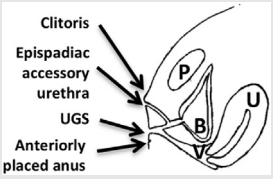
Patients and methods
Medical records were retrospectively reviewed of three girls with urethral duplication managed over a 30-year period at a single institution. Circumstances of diagnosis, management and outcomes were analysed.

Results
The oldest case presented as a neonatal retrovesical mass with an accessory clitoral stream, whereas the two more recent cases presented with antenatal hydrocolpos and bilateral ureterohydronephrosis. Cases 1 and 3 had an incomplete duplication, while Case 2 had a complete form. In Case 3, the duplication was associated with a urogenital sinus and an anteriorly placed anus. Management involved resection of the epispadiac accessory urethra to achieve continence, with dilatation and/or mobilisation of the hypospadiac one. All girls are now aged >5 years old and are continent, and one is old enough to have normal menstruation. Renal function is normal in all. The summary table presents the schematic anatomical description as shown on micturating cystourethrogram and endoscopy, as well as the management for each patient.

Discussion
Step-by-step management is necessary in urethral duplication. The neonatal emergency is to release the urinary tract compression by evacuating urinary retention or hydrocolpos. Later in infancy, decision has to be taken regarding the urethras. If the resection of the epispadiac accessory urethra seems acceptable to achieve continence, the attitude towards the hypospadiac channel is more controversial and should be individualised. Embryologic and aetiopathogenic pathways are still missing to uniformly characterise the malformation.

Conclusion
Paediatric urologists should remember that there is a wide spectrum of urethral duplication in girls, and that various presentations exist beside the more classic prepubic sinus.

Table Presentation summary of the cases.

	Patient 1	Patient 2	Patient 3
Anatomic description			
Treatment	- Endoscopic vaginal dilatation - Resection of accessory urethra	- Vaginal dilatation - Resection of accessory urethra - Partial urogenital sinus mobilisation	- Drainage of hydrocolpos through a catheter placed in the Urogenital sinus - Currently awaiting treatment

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Introduction

Urethral duplication is a rare malformation, especially in girls, with less than 40 cases reported in the literature so far. The most frequent forms develop in the sagittal plane, and several classifications exist for boys [1–3]. The classification established by Stephens in 1983 was adapted for girls in 2014 [4], and distinguishes a prepubic sinus from a dorsal accessory urethra in the epispadiac position. In the latter case, the duplication can be complete if the accessory urethra emerges directly from the bladder, or incomplete in cases of a Y-division of the two urethras after their emergence from the bladder. This report presents the clinical outcomes of three cases of urethral duplication in girls where the dorsal accessory urethra was associated with a ventrally displaced main channel. It discusses clinical presentation, management and long-term outcomes of this rare anomaly.

Case reports

Case 1

In 1979, a newborn girl presented with a retrovesical cystic mass associated with bilateral ureterohydronephrosis. There were no associated anomalies, particularly of the genital or digestive tract. Vaginography showed stenosis of the lower part of the vagina, and a vaginoscopy was performed during the first days of life, which confirmed the diagnosis of incomplete stenosis of the lower part of the vagina. Dilatation was performed with Hegar dilators to release the hydrocolpos and hydronephrosis. Micturating cystourethrogram (MCUG) showed an incomplete urethral duplication with an accessory dorsally placed urethra (Fig. 1A and B). There was no disjunction of the pubic symphysis and the spine was normal. Endoscopic evaluation was performed at 3 years of age, showing good vaginal calibre. Urethrocystoscopy showed a ventrally positioned main urethra ending in the hymen, like in female hypospadias. When the bladder was compressed, an accessory stream was visible at the base of a bifid clitoris (Fig. 1C).

To avoid the risk of stenosis of the main urethra, the accessory channel was not completely excised, but a clitoroplasty, as described by Randolph [5], was performed to burry it and prevent it from leaking. The main urethra was left in its hypospadiac position.

After 14 years of follow-up, renal function remained stable, despite renal hypotrophy (−2 SD), the patient was continent per urethra and had normal menstruation.

Case 2

In 2002, a female foetus had an ultrasonographic antenatal diagnosis of hydrocolpos and bilateral hydronephrosis. There was no associated anomaly and the karyotype was 46, XX. At birth, hydrocolpos was confirmed by ultrasound, and drained through a catheter to decompress the urinary tract and decrease hydronephrosis. At day 8 of life, hydronephrosis persisted and examination under general anaesthesia found a narrow hypospadiac urethra, which was dilated. Subsequently, a CT scan with delayed films was performed, showing an abnormal communication between the posterior wall of the bladder neck and the anterior vaginal wall. Micturition cystourethrogram showed a complete urethral duplication (Fig. 2A), with the accessory urethra arising directly from the bladder and ending at the level of the clitoris, which was normal and non-bifid (Fig. 2). No accessory stream was clinically detectable. At 15 months of age, the accessory channel was resected until its origin above the bladder neck (Fig. 2B and C). There was no bladder neck around this accessory urethra. At the same time, the hypospadiac urethra was slightly mobilised anteriorly to place it just above the hymen with a Y-V plasty.

During childhood, the patient presented with episodes of urinary retention and febrile UTIs. At 30 months of age, partial urogenital sinus mobilisation, as described by Rink [6], was performed to bring the hypospadiac narrowed urethra to the perineum and enlarge it. The patient is now 8 years old. She is continent per urethra and has not had any further UTI or urinary retention. Her renal function is normal.

Case 3

In 2014, a female foetus was diagnosed with transient ascites at 12 weeks gestation. The karyotype was 46, XX. The second trimester ultrasound showed hydrocolpos in a duplicated genital tract and bilateral hydronephrosis with resolution of the ascites. Foetal magnetic resonance imaging (MRI) was performed to better define the anatomy; it showed complete duplication of the genital tract, bilateral

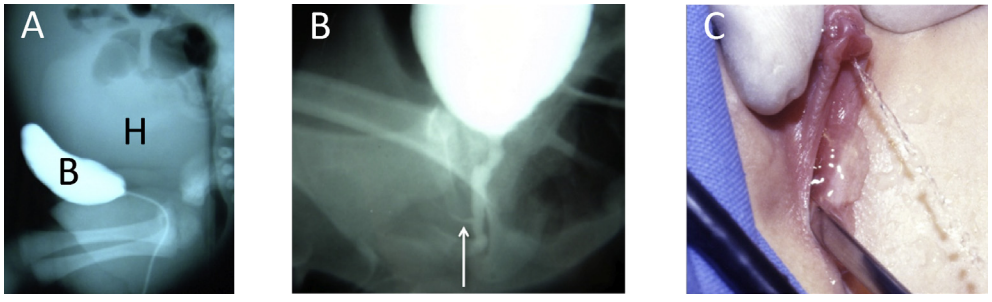

Figure 1 Case 1. A. Neonatal MCUG showing a large hydrocolpos (H) behind the bladder (B). B. Pre-operative MCUG showing an incomplete urethral duplication with a main hypospadiac urethra and an accessory epispadiac urethra (arrow). C. Per-operative clinical photograph showing the thin accessory stream below the clitoris when compressing the bladder.