Urethral duplication in a child with VATER association

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Introduction

Urethral duplication (UD) is a rare anomaly with about 300 cases reported to date, usually seen in males and often associated with a number of anomalies involving other organs. Effmann's classification has described three types of urethral duplication. One out of three patients has an associated vesicoureteric reflux (VUR). In type I which is also the most common type, the urethra is partially duplicated and is almost always asymptomatic, requiring no further treatment. In type II, complete duplication of the urethra is observed. Type II urethral duplication is further classified as type IIA1 if both urethras arise from the separate bladder necks, type IIA2 (Y-type duplication) if one channel arises from the other, and type IIB if duplication with one meatus is observed. Type III urethral duplication comprises complete duplication of the urethra and bladder.

The exact embryogenesis of urethral duplication is not well understood. Symptoms vary depending on the type of duplication. Symptoms could include recurrent urinary tract infections (UTI), epididymitis, and urinary incontinence. Diagnosis is usually made on voiding cystourethrogram (VCUG) and/or retrograde urethrogram (RUG). We report a case of type IIA2 urethral duplication in a male child with multiple other anomalies.

Case report

A 14 year old male child presented to the Paediatric urology clinic with history of poor urinary flow and passing of urine from the undersurface of the penis since birth. He was earlier diagnosed to have tetralogy of Fallots, left sided grade V vesico-ureteric reflux (VUR) and mid penile hypospadias. He had undergone VSD (ventricular septal defect) closure and infundibular resection at the age of six years. At the age of 10 years, the child underwent left sided nephroureterectomy for non-functioning kidney with grade V VUR.

The child had recurrent episodes of fever and urinary tract infection after that. During the cardiac surgery, the child could not be catheterized and underwent suprapubic cystostomy. The child on examination had a normally placed dorsal urethral meatus at the centre the glans and a ventrally placed mid-penile hypospadiac meatus (Fig. 1a). The calibre of the dorsal urethra was small and could admit only 5 Fr infant feeding tube. Retrograde urethrogram/micturating cystourethrogram revealed a duplication of urethra type IIA2 (Fig. 1b). Serum creatinine was normal. Cystoscopy was done through the ventral hypospadiac meatus, and the calibre of the urethra appeared normal. A guide wire was passed through the dorsal meatus and it was seen emerging proximally at the site of proximal bulbular urethra.

Surgical procedure

It was decided to open the dorsal urethra and anastomose with the ventral urethra and at the same time repair the hypospadias. The child was having a permanent cardiac pacemaker and hence the settings were adjusted prior to surgery. Initially, under general anaesthesia and the child in lithotomy position, a 0.032 inch guide wire was passed through the dorsal meatus. A 10 Fr paediatric resectoscope was passed through the ventral meatus up to the site where the guide was seen emerging. Using a Collin's Knife the septum between the two urethras was incised till the penoscrotal junction. The common urethra appeared wide open. A 12 Fr urethral catheter was introduced through the ventral meatus.

An incision was made around the ventral meatus, the urethral plate and extended around the glans (Fig. 2a). The penis was degloved (Fig. 2b). An artificial erection was created to check for chordee. There was a minimal chordee of less than 30°. The chordee was corrected by dorsal plication sutures on both sides of midline. Through the ventral urethra an incision was made vertically into the dorsal urethra till the peno-scrotal junction to merge with the incision done endoscopically (Fig. 3a). The sides of the two urethras were anastomosed so as to create a single wide urethra (Fig. 3b). The hypospadias was repaired using the tubularized incised plate urethroplasty technique (Fig. 3c). The catheter was left in place for 10 days. The child voided well after catheter removal.

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Discussion

VACTERL association which is also termed as VATER association depending on the criteria used for diagnosis, is estimated to occur in 0.3–2.1 per 10,000 live births and includes at least several of the following defining component features: Vertebral anomalies, Anal atresia, Cardiac malformations, Tracheo-Esophageal fistula, Renal anomalies, and Limb abnormalities. Genitourinary (GU) anomalies are not
considered as one of the core component features of VACTERL association, however these malformations have been often reported. The child in this report had a number of major anomalies including genital urinary anomalies hence forming a part of the VATER association. Duplication of the urethra in the most extreme cases can be associated with complete duplication of the penis or urinary bladder. Urethral duplication may be sagittal or collateral. Sagittal duplication takes the form of two channels running one above the other in the sagittal plane, whereas in the collateral form, the two urethras run side by side. The child in our report had a sagittal duplication. Most urethral duplications occur in the sagittal plane within a single penis and most are incomplete. Usually in such cases the ventral urethra is the dominant one. The most common sagittal variety is an orthotopic principal urethral channel and an epispadiac accessory urethra lying dorsal to it. The child in our report had a dominant ventral urethra which had a hypospadiac opening.

One should have a detailed knowledge of urethral duplication as it is important when planning for any surgical procedure. Many children are usually asymptomatic and do not require any surgery. Indications for surgery are bothersome symptoms and cosmetic or functional deformity. Surgical reconstruction may vary from case to case. It may range from simple meatoplasty to complex staged urethroplasty, depending on the severity of case. A favourable outcome is achieved in most of cases after reconstructive surgery.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2018.11.018.

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