Y-type urethral duplication presented with perineal fistula in a boy

Y-Typ-Doppelung der Urethra mit einer perinealen Fistel bei einem Jungen

Abstract

Urethral duplication is a rare congenital anomaly of the lower urinary system and has varied presentation. According to the Effmann classification, type IIA2-Y urethral duplication is charcterized by the duplicated urethra originating from the bladder neck and opening into either the rectum or the perineum. The accessory urethra is normal and functional and the normally positioned dorsal urethra is hypoplastic and stenotic in unusual form of Y-type duplication. We present a new case with unusual form of Y-type duplication and discuss its treatment.

Keywords: congenital abnormalities, fistula, duplication, urethra, perineum

Zusammenfassung

Die Doppelung des Harnleiters ist eine seltene angeborene Störung der ableitenden Harnwege und hat unterschiedliche Einordnungen erfahren. Beim Typ II A-Y (Klassifikation von Effman) beginnt die Doppelung der Urethra am Blasenhals und mündet entweder im Rektum oder am Perineum. Der zusätzliche Harnleiter ist normal ausgebildet und funktional. Der normal positionierte dorsale Harnleiter ist hypoplastisch und in einer ungewöhnlichen Y-Form stenosiert. Hier wird ein neuer Fall von ungewöhnlicher Harnleiterdoppelung vom Y-Typ vorgestellt und die zugehörige Therapie diskutiert.

Schlüsselwörter: angeborene Fehlbildungen, Fistel, Doppelung, Harnleiter

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Introduction

Urethral duplication is a rare congenital anomaly of lower urinary system. The clinical presentation differ according to the anatomical variant and there are a variety of types and several classification systems [1], [2]. Y-type duplication is a special form of type IIA2 urethral duplication, two urethras originating from the bladder neck. It has two variants; the usual form of Y-type duplication is characterised by a stenotic orthotopic (dorsal) urethra and a more functional accessory (ventral) urethra, which opens into the perineum, perianal region or the anal canal [3]. The unusual form includes a normal dorsal urethra and hypoplastic ventral urethra. According to this classification, our patient presents an unusual form of urethral duplication. We reported a 15 year old boy with unusual form of Y-type urethral duplication and reviewed the literature.

Case presentation

A 15 year old boy presented with perineal dripping during micturation and had recurrent urinary infection since childhood. The penis and meatal opening were normal but he had a 1x1 mm fistula in perineal area in physical examination.

A wide investigation of radiology was made including renal sonography, intravenous pyelography, voiding cystourethrography, fistulography and retrograde urethrography. Renal sonography and intravenous pyelography were normal. The fistulography and retrograde urethrography showed a Y-shaped urethra, the second urethra was thin and coursing from the posterior urethra to the perineum (Figure 1a+b). During cystourethroscopy methylene blue was injected into the perianal fistula and the flow was seen in the prostatic urethra (Figure 1c–e).

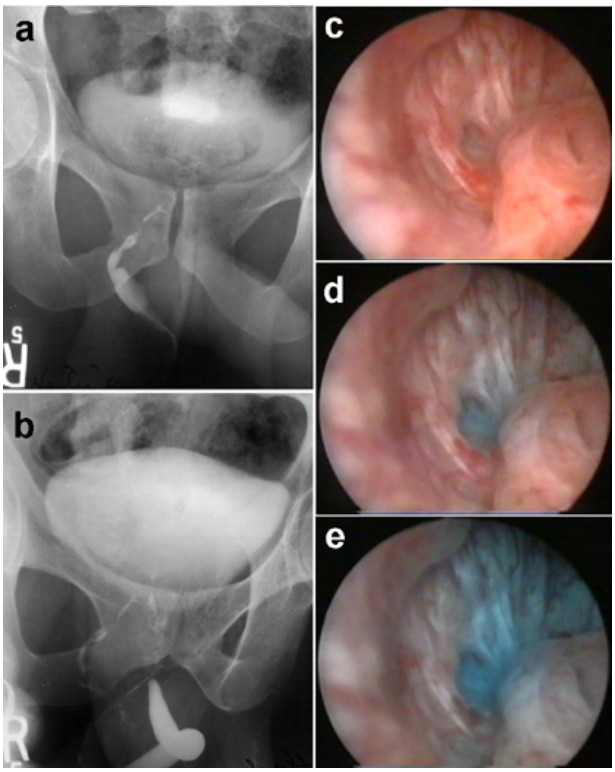

Figure 1: (a) Appearance of accessory urethra on fistulography and (b) on voiding cystography; (c, d, e) methylene blue flow, which was injected into the perianal fistula, was seen in the prostatic urethra during cystourethroscopy

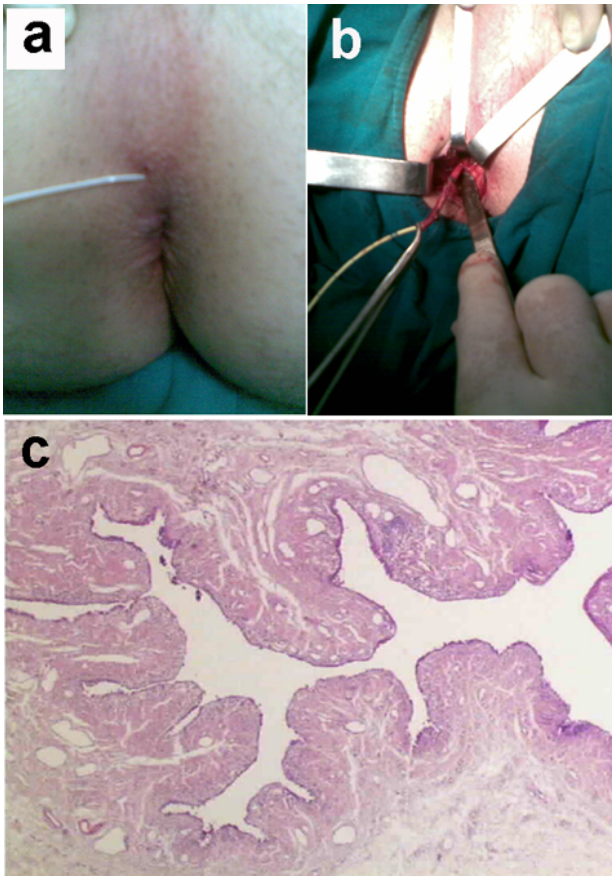

Figure 2: (a) Urethral opening of accessory urethra, (b) dissection and exclusion of accessory urethra, (c) pathologic examination of accessory urethra

Y-type urethral duplication was diagnosed and surgery approach was decided. After inserting an urethral catheter into the meatus of the accessory urethra, it was dissected through perineal incision until the posterior side of the prostatic urethra. The accessory urethra was ligated with a 3\0 vicryl and carried out (Figure 2a+b).
Histopathologic examination of the excised tract demonstrated a lining of transitional cell epithelium (Figure 2c), thus confirming that it was urethral duplication rather than urethral fistula. The patient was normal and asymptomatic at one year follow-up.

Discussion

The embryology of urethral duplication is unclear because there are probably different causes for the various types of anomalies, and numerous theories have been postulated [1], [4]. On the one side, two explanations have been offered for embryological development of complete duplication of bladder and urethra: (i) excessive constriction between the urogenital and vesicourethral portions of the ventral cloaca, and (ii) a supernumerary cloacal septum that indents the epithelial wall of the bladder [5]. On the other side, possible mechanisms of embryologic development of Y-type duplication are faulty closure of the urorectal membrane, impaired growth of the dorsoinferior wall of the urogenital sinus or fistula formation in the dorsal margin of the urogenital sinus due to possible vascular accident [6]. There appears to be a misalignment of sorts between the termination of the cloacal membrane and its relationship with the developing genital tubercle and urogenital sinus [7].
Y-type duplication may be associated with disorders of midline development and other congenital anomalies such as cloacal exstrophy, conjoined twins, early amnion rupture syndrome, and hand-foot genital syndrome. A relationship between urethral duplication and disorders of midline development has been found such as VATER (vertebral defect, anus imperforate, tracheoesophageal fistula, radial and renal dysplasia) and VACTERL complex (vertebral, anal, cardiac, tracheal, esophageal, renal, limb etc) [8], [9]. We did not find any other anomalies in our patient with detailed evaluation before beginning the treatment process.
Effmann classification includes all clinical aspects of urethral duplication and it is more useful for the surgeon in helping him make the decision for surgery (Figure 3). According to Effman classification, Y-type urethral duplication (TypeAII 2) defines two urethras originating from a common bladder neck and states that the accessory urethra opens into the area perineum to rectum. Y-type urethral duplication usually has a more functional ventral channel and a hypoplastic or stenotic dorsal (orthotopic) channel. Hence, some researchers/scholars suggest that this type of duplication exists only when the ventral urethra is the functional urethra [10]. However, when the ventral urethra is hypoplastic, the anomaly is classified as a congenital urethroperineal fistula [10]. On the other

hand, Wagner et al. accepted all congenital urethroperineal fistulas as urethral duplications [11]. These researchers/scholars believe that the presence of transitional cell epithelium lining the tract, as we observe in the case of our patient, is very supportive of a true urethral duplication.

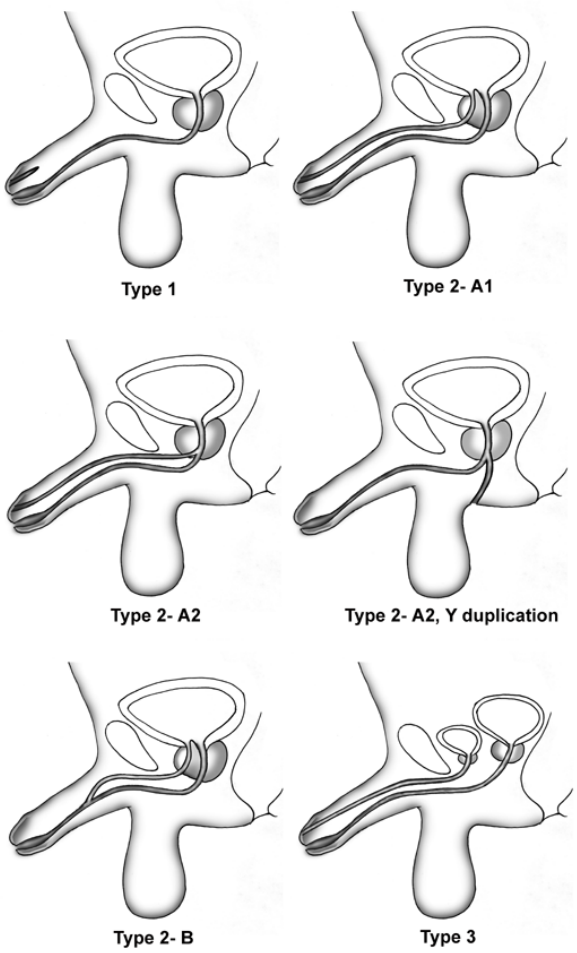

Figure 3: Classification of urethral duplication proposed by Effmann et al. [1]

Clinical significance of urethral duplication is various. Most cases of urethral duplication with genetic anomalies are diagnosed during infancy. Urinary symptoms show a wide range, such as double stream, incontinence, urinary tract infection, or outflow obstruction. In most cases, patients with unusual form of urethral duplication present no symptoms, excluding the occasional double stream and urinary tract infection. Our patient was not only suffering from recurrent urinary infections, but also experiencing dribbling of urine in perineal area since birth.

Diagnosis of Y-type urethral duplication is made with voiding cystourethrography, retrograde urethrography and fistulography. Urethrocystoscopy should be performed to confirm the radiographic findings [2], [3], [4], [12]. Imaging and viewing of the lower urinary tract is used to find out the other lower urinary tract abnormalities such as posterior urethral valve, bladder neck obstruction and megalourethra, which might be associated with Y-type urthral duplication [12], [13], [14], [15]. Upper urinary tract should be carefully examined with renal ultrasonography and intravenous urography, because urethral duplication may be associated with upper urinary tract anomalies such as solitary kidney, hydronephrosis, multicystic dysplastic kidney and obstructive megaureter [12], [16]. The fistulography and retrograde urethrography showed a Y-shaped urethra. The second urethra was thin and coursing from the posterior urethra to the perineum. Renal sonography and intravenous pyelography were normal in our patient.

The treatment for duplicate urethra depends on the patient's symptoms and the type of anomaly. Y-type duplications with hypoplastic dorsal urethra and stenotic meatus obviously require more complex intervention than other kinds of repair [4], [17]. In cases in which the dorsal urethra is the normal channel, as observed in our patient, excision of the accessory anterior channel is the final surgical treatment and can be successful [10], [11]. The alternative methods of sclerosis or fulguration of the accessory channel also have been reported [18]. Sclerosis is associated with risks of corporal thrombosis, fibrosis, impotency, and incontinence; hence, it should be avoided. In our case we excised the accessory urethra.

In conclusion, an unusual Y-type urethral duplication should be suspected in any patient who has a history of recurrent urinary infection attacks and recurrent perineal dripping during micturation. We suggest that a simple resection of the ventral channel is curative for Y-type duplication with a normal orthotopic urethra.

Notes

Competing interests

The authors declare that they have no competing interests.

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Please cite as
Dayanc M, Irkilata HC, Kibar Y, Bozkurt Y, Basal S, Xhafa A. Y-type urethral duplication presented with perineal fistula in a boy. GMS Ger Med Sci. 2010;8:Doc33.
DOI: 10.3205/000122, URN: urn:nbn:de:0183-0001229

This article is freely available from
http://www.egms.de/en/journals/gms/2010-8/000122.shtml

Received: *2010-04-21*
Revised: *2010-10-04*
Published: *2010-11-29*