Urethral Duplication in the Setting of Posterior Urethral Valves

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Abstract

Urethral duplication is a genitourinary anomaly that may rarely present in conjunction with posterior urethral valves. The management algorithm of children affected by both these urologic conditions is still unknown, given the low incidence and the complexity of each of these conditions. We present our clinical experience with the management of a child with Y-type urethral duplication and posterior urethral valves.

ABBREVIATIONS

GU: Genitourinary; PUV: Posterior Urethral Valves; mg/dL: milligrams/deciliter; VACTERL: Vertebral defects, Anal atresia, Cardiac defects, Tracheo-Esophageal fistula, Renal anomalies, and Limb abnormalities

INTRODUCTION

Congenital structural abnormalities are detected in approximately 1% of pregnancies in the prenatal period, with approximately 20% of cases involving the genitourinary (GU) system, and the majority of these GU abnormalities being hydronephrosis [1]. Bilateral hydronephrosis, particularly in boys, is frequently associated with posterior urethral valves (PUV) and may present with other GU anomalies. One rare anomaly that has been reported in the literature to occur with PUV is urethral duplication. Typically diagnosed in early childhood, urethral duplication can present in both genders, but is more commonly seen in boys [2–5]. While there have been many case reports of urethral duplication in the literature, optimal management is still unclear, and can range from conservative management to surgical intervention, depending on the function of the duplicated urethra. In the small number of cases with urethral duplication and concurrent PUV, management is even more uncertain, given the evolving nature of PUV and risk for long-term bladder dysfunction and renal failure. As such, we report our experience with the management of a young boy with a combination of urethral duplication and PUV.

CASE PRESENTATION

A term male infant with prenatally-detected bilateral grade 4 hydronephrosis, distended bladder, and cystic dysplasia of the right kidney was transferred to our hospital from an outside facility shortly after birth. Creatinine was elevated at 0.98 milligrams/deciliter (mg/dL) 72 hours after birth. Genitourinary exam was significant for a distended abdomen, abnormally deficient prepuce, and two urethral meaty, one located in the orthotopic penile position and the other within the scrotal skin (Figure 1). The more ventral, ectopic meatus was only noted after unsuccessful attempts at catheterization of the glanular meatus by the neonatologist. The remainder of his physical exam was consistent with an otherwise well-developed term infant, and a cardiac murmur secondary to a ventricular septal defect that was managed conservatively.

Voiding cystourethrogram demonstrated a small trabeculated bladder, bilateral grade 5 reflux with renal cortical thinning (Right-Left), and a dilated posterior urethra consistent with posterior urethral valves (Figure 2). With an elevated baseline creatinine, the infant's bladder was initially managed with an indwelling catheter in the scrotal meatus in the first few days of life. Primary transurethral valve ablation was performed via the scrotal meatus after the first week of life, along with retrograde urethrogram (Figure 3), which confirmed the presence of type

Figure 1 (A) Relatively normal-appearing phallus with orthotopic meatus; (B) Ectopic scrotal meatus (arrow).
IIA2 (Y-type) complete urethral duplication, with a patent but stenotic penile urethra. The verumontanum and valves were identified to be proximal to the urethral bifurcation. Within a week after valve ablation, the bilateral hydronephrosis improved and his nadir creatinine reached 0.25 mg/dL. The infant was then managed with prophylactic antibiotics with spontaneous voiding predominantly via the scrotal meatus and occasional drips noted from the glanular meatus.

Despite conservative management, the infant developed recurrent febrile urinary tract infections with concurrent acute renal failure. Initial evaluation was consistent with residual valve scar tissue, for which he underwent repeat ablation. The child continued to void primarily via the scrotal meatus, with significant polyuria. He developed subsequent urinary tract infections, which were suspected to be related to obstruction from the duplicated urethra, as well as poor bladder compliance with concurrent polyuria/polydipsia, likely from valve bladder syndrome. As such, he was initiated on a regimen of clean intermittent catheterization and overnight drainage through the scrotal meatus and occasional drips noted from the glanular meatus.

DiscoN\ the combination of Y-type urethral duplication with PUV is an extremely rare occurrence [6-12]. Various authors have postulated that urethral duplication may result from ischemia and/or the abnormal development of the Mullerian duct or urogenital sinus during gestation [13-15] but the relation between these processes and the development of PUV is still unclear. Based upon clinical studies, various classification systems have been proposed, but the Effmann classification system (Figure 4) appears to be the most widely accepted [16]. In the case of Type IIA2 complete urethral duplication, the more ventral urethra is often considered to be the more functional urethra and likely containing the urethral sphincter, despite the dorsal urethra more often being in the orthotopic position [16]. Our patient appears to be a variant of this category, as the continence of both urethral channels is provided by a single sphincteric mechanism, located proximal to the bifurcation point between the two urethral channels.

The clinical presentation of coincident urethral duplication and PUV may occur along a spectrum, commonly including abnormal prenatal imaging, weak or double urinary stream, recurrent urinary tract infections, urinary obstruction, and/or incontinence [6-12]. Early management of these children is initially focused on optimizing urinary drainage shortly after diagnosis, which may range from urethral catheterization through the most patent tract to upper tract urinary diversion, followed by valve ablation when possible. Subsequent management of the duplicated urethra then varies, in type and timing of treatments, depending on functional and cosmetic considerations. With minimal symptoms, observation of the urethral duplication or dilation of the dorsal urethra to optimize patency have been reported [8,10,11]. However, most case studies involving urethral duplication indicate that the majority of these children eventually undergo reconstructive surgery, or ablation or excision of the nonfunctional urethra [2,4,7,8]. Yet, the long-term effects of this surgical intervention on the management of PUV, ectopic urethra affords the unique ability for easy intermittent self-catheterization in the setting of the valve bladder.

DISCUSSION

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and inversely, the impact of PUV on the management decision process for urethral duplication, are still unclear [17].

In our case study, despite early ablation of the PUV, our patient is still at high risk for long-term renal failure and voiding dysfunction, particularly given his dysplastic right kidney and valve bladder syndrome. Also, while the ectopic meatus provides facile catheterization for drainage, in the absence of catheterization, it is unclear if the urethral duplication itself is obstructive in nature. With a proximal urethral sphincter and patency of both urethral channels distal to the sphincter, surgical intervention may include excision of the ectopic urethra, excision of the orthotopic urethra with translocation of the ectopic urethra, or reconstruction of both tracts into a singular orthotopic urethra. Other than cosmesis, such reconstructive surgery may have implications on future toilet-training and sexual function. Given the evolving nature of his PUV, reconstruction to an orthotopic urethra may either improve or complicate the status of his renal function, and potentially impact his risk for end stage renal disease and renal transplantation. Thus, the optimal management of urethral duplication with concurrent PUV in this child is still unknown, and will likely rely on the child’s future renal health, as well as growth and development.

REFERENCES