Concomitant Anterior and Posterior Urethral Valves: A Comprehensive Review of Literature

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Posterior urethral valves (PUVs) are the most common cause of infravesical obstruction in children, whereas anterior urethral valves and/or diverticula (AUV/D) are less commonly encountered. Concomitant AUV/D and PUV is believed to be very rare and may be missed on the initial evaluation. In this review, we provide brief information on embryology of AUV/D and PUV to explain the concurrent presence of these anomalies. We also summarize the information on published cases of concomitant AUV/D and PUV in the English literature with a special focus on diagnosis and the importance of both voiding cystourethrography and careful urethrocystoscopy. UROLOGY 86: 151–157, 2015. © 2015 Elsevier Inc.

Posterior urethral valves (PUVs), with an incidence of 1 in 5000 to 8000 male births, are the most common cause of congenital lower urinary tract obstruction.[1] Less commonly encountered are obstructive anomalies of the anterior urethra with anterior urethral valves (AUV) and/or diverticula (AUV/D) at the top of the list. Although 10-30 times less prevalent than PUV, these conditions may be similarly devastating and need prompt diagnosis and treatment in early stages.[2]

A more challenging but less known phenomenon is double urethral obstruction by concomitant anterior and posterior valves. A deeper view on concurrent presence of these conditions may shed some light on the largely unknown etiology and embryology of both AUV/D and PUV. However, our understanding of concomitant AUV/D and PUV is limited to few case reports, and little is known about the presentation, diagnosis, and management of these patients. This limited knowledge together with the assumed rarity of this condition, at least in the published literature, may lead to underdiagnosing of concomitant AUV/D and PUV in children.

We hereby review the literature on concomitant AUV/D and PUV to summarize the clinical presentation, diagnosis, and treatment of this condition with a focus on pitfalls in initial workup. We also review some of the embryologic explanations behind AUV, anterior urethral diverticulum (AUD), and PUV in an attempt to explain the concurrence of these conditions based on the current evidence.

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METHODS

We reviewed the literature by searching the MEDLINE (PubMed) electronic database and Google Scholar through November 2014. We used various combinations of text words such as "anterior urethral valve," "anterior urethral diverticula," "posterior urethral valve," "double obstruction," and "infravesical obstruction" to identify relevant studies. We also used the "related articles" function in MEDLINE to find related studies. Articles were screened based on titles and abstracts, and relevant articles were selected for a full review. The reference list of related articles was also screened to find additional articles. Online-only articles or those not indexed in MEDLINE were also included. Non-English publications were excluded, unless an informative abstract was provided in English.

ANTERIOR URETHRAL VALVE VS ANTERIOR URETHRAL DIVERTICULUM

Which came first, AUV or AUD? Little consensus exists on the use of AUV and AUD terminology in the literature that leads to remarkable inconsistency in different reports. One theory attempts to explain this controversial subject by proposing that a spongy defect in urethra leads to formation of AUD, and the anterior lip sometimes acts as a valve when the diverticulum is filled.[3-5] This would also explain the situations that AUD is present without any valve effect. However, another theory sees AUV as the initial insult causing obstruction and turbulence in the proximal urethra leading to urethral bulging and diverticulum formation.[6] This theory would better account for AUVs without a diverticulum.[7,8] It can be suggested that an early obstructive AUV may affect the normal differentiation of proximal spongy tissues and explain the hallmark defects seen in AUD. A dilated or ruptured Cowper cyst should be differentiated with AUV and AUD, although this condition may share some of the characteristics of AUD and an early obstructive insult may be the underlying cause for its dilation.

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In 1978, Firlit et al[9] provided a classification for AUV that categorized diverticula as a subtype of AUV with the same pathology. In this classification, type 1 roughly represents AUV without diverticula and AUV types 2-4 represent AUD with or without changes in the upper tract.[9] The classification however does not cover AUVs that cause upper tract anomalies in absence of an AUD.[8] However, some other authors clearly differentiate between AUV and AUD and believe that these are distinct entities.[10,11] As proponents of this theory, Brueziere and Guerrieri[10] presented their clinical and surgical experience in 14 cases and concluded that AUD develops outside of the corpus spongiosum and should be treated by open surgical repair, whereas in AUV, the distention is covered by corpus spongiosum and should be treated endoscopically. However, what the authors referred to as "outside of corpus spongiosum" may simply represent incomplete spongy tissue formation with fibrotic and rudimentary remnants similar to pathologic findings of Dorairajan.[12] Considering these inconsistencies, we used AUV/D terminology in this review to represent AUV and/or AUD, unless otherwise specified.

AUD represents a ventral saccular dilation of the urethra with defects in the corpus spongiosum as its hallmark. The size of the diverticulum is variable based on the degree of tissue defect and probably duration of obstruction and pressure overload on the adjacent tissues. In pathology, a thin epithelial lining covers the diverticulum that is surrounded by a fibrotic and rudimentary spongy tissue.[12] The proximal corpus spongiosum may also show degrees of abnormality and fibrosis.[12] The distal lip of AUD may act as a "flap-valve" or have a "gate-effect" obstructing the urine flow during urination.[13] This probably happens when the diverticulum fills with urine and the pressure on the distal flap pushes it against the urethral roof to occlude the urethra.[2,14,15] AUV, if not associated with AUD, mostly represents a "cusp-like" or "iris-like" mucosal remnant in the ventral aspect of the urethra, with no or minimal bulging proximal to it. However, one can argue at what level of dilation a proximal bulging fulfills the criteria to be called a diverticulum, if presence of a spongy defect is not evident or assessable. These anomalies occur anywhere in the anterior urethra, most commonly at the penoscrotal junction and also the penile or bulbar urethra.

EMBRYOLOGY

Normal Development of External Genitalia in Male

Despite the recent advances in molecular aspects of embryology, our knowledge about the exact mechanisms in the development of urogenital tract is still limited. Complex interactions between androgen-independent, androgen-dependent, and hormonal and environmental factors play a role in external genitalia differentiation.[16] During early ambisexual (indifferent) phases, urorectal septum develops toward the cloacal membrane, urogenital and labioscrotal folds appear, and the primary perineal body and genital tubercle form mostly in a hormone-independent manner.[16] Under the effect of Y chromosome and androgens, masculine differentiation of external genitalia continues. Urethral plate and mesenchymal phallic tissue abut the genital tubercle and elongate caudally to form the phallus. The urethral plate canalizes to form the urethral groove (from endothelial origin), whereas urogenital folds (from mesothelial origin) migrate medially and meet at the midline to complete the cylindrical structure of the phallus and penile urethra from behind forward. The mesenchymal urogenital folds later differentiate into vascular erectile tissues (corpus spongiosum and corpora cavernosa). The cranial part of urogenital sinus also forms the membranous and prostatic urethra. Early in development, the Wolffian (mesonephric) ducts insert into the cloaca anteriorly as solid structures but soon they undergo canalization and migrate posteriorly and cranially to finally lie at the level of future verumontanum.[13] This gradual migration leaves "snail-track" marks in the urethra toward verumontanum that are absent in total agenesis of Wolffian ducts.[13,17] Most caudally, the cells grow to form glans penis, and canalization begins at the summit of glans by ectodermal ingrowth to meet the urethra formed by the urethral plate proximal to fossa navicularis. However, the formation of this part is more controversial and some disputed the ectodermal origin and proposed that the proximal urethral plate is also responsible for formation of the urethra in fossa navicularis by endodermal-mesenchymal interaction and endodermal differentiation.[18,19]

Embryology of Anterior Urethral Valve and/or Diverticula

Several theories are proposed considering embryologic origin of AUV/D. An early transient obstruction in the anterior urethra (eg, stenosis or preputial narrowing) was first proposed by Watts in 1906 (cited in the study by Williams and Retik[14]). Others also proposed that incomplete closure of urogenital folds or a failed attempt for urethral duplication might be the cause.[2,14] In 1886, Kaufman attributed AUD to a junctional obstruction between glandular and penile urethra (cited in Williams and Retik[14]). Similarly, Stephens in 1993[13] found obstructed glandular urethra in autopsies of 2 patients with megalourethra and proposed that delayed canalization of glandular urethra might be the main culprit. However, Williams and Retik[14] questioned most of these theories and advocated those proposed by Suter and Johnson that congenital dilation of periurethral glands or cysts that open into the ventral urethra is the underlying cause of AUD. Similarly, McLellan et al[15] supposed that rupture of Cowper cyst or syringocele leads to AUD formation. Although a promising theory, one should be skeptical that why the glands were prone to dilation in the first place and if any preceding event (eg, a transient obstruction) led to incomplete spongy tissue formation around the gland. Additionally, this theory does not fully explain the valves in more distal parts and also isolated