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Anterior Urethral Valve: A Rare But an Important Cause of Infravesical Urinary Tract Obstruction

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Authors' Contribution:

- A Study Design
- B Data Collection
- C Statistical Analysis
- D Data Interpretation
- E Manuscript Preparation
- Walluscript Frepara
- F Literature Search
- **G** Funds Collection

Summary

Background:

Urethral valves are infravesical congenital anomalies, with the posterior urethral valve (PUV) being the most prevalent one. Anterior urethral valve (AUV) is a rare but a well-known congenital anomaly. AUV and diverticula can cause severe obstruction, whose repercussions on the proximal urinary system can be important. Few cases have been described; both separately and in association with urethral diverticulum. The presentation of such a rare but important case led us to a report with highlighting its classic imaging features.

Case Report:

We present a case report of AUV with lower urinary tract symptoms in a 6-year-old boy with complaints of a poor stream of urine and strain to void. Unique findings were seen on Retrograde Urethrography (RGU) and Voiding Cysto-Urethrography (VCUG), i.e. linear incomplete filling defect in the penile urethra and associated mild dilatation of the anterior urethra ending in a smooth bulge. On cysto-urethroscopy the anterior urethral valve was confirmed and fulguration was done.

Conclusions:

Congenital anterior urethral valve is an uncommon but important cause of infravesical lower urinary tract obstruction that is more common in male urethra. It can occur as an isolated AUV or in association with diverticulum and VATER anomalies. Early diagnosis and management of this rare condition is very important to prevent further damage, infection and vesicoureteral reflux. AUV may be associated with other congenital anomalies of the urinary system; therefore a full evaluation of the urinary system is essential.

MeSH Keywords:

Cautery • Urethral Stricture • Urogenital Abnormalities • Urography • Urology • Vesico-Ureteral Reflux

Abbreviations:

PUV – posterior urethral valve; **AUV** – anterior urethral valve; **RGU** – retrograde urethrography; **VCUG** – voiding cysto-urethrography; **VUR** – vesico-ureteric reflux

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Background

Urethral valves are infravesical congenital anomalies with the posterior urethral valve (PUV) being the most prevalent one. Anterior urethral valve (AUV) is a rare but well-known congenital anomaly. AUV and diverticula can cause severe obstruction, whose repercussions on the proximal urinary system can be important. Few cases have been described; both separately and in association with the urethral diverticulum. The presentation of such a rare but important case led us to a report with highlighting its classic imaging features.

Case Report

A six-year-old male patient presented with complaints of dysuria and dribbling of urine. He had a history of poor urinary stream and straining during micturition. The symptoms had gradually increased in severity over the past 6 months. On examination, the abdomen was soft, the urinary bladder was palpable, and there was no swelling on the ventral aspect of the penis during voiding. Creatinine was 0.33 mg/dL and urea was 22.30 mg/dL. The patient underwent Retrograde Urethrography (RGU) and Voiding Cysto-Urethrography (VCUG). RGU showed a

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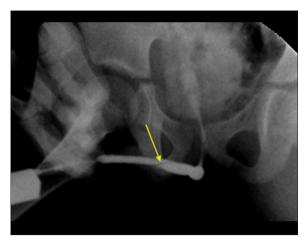


Figure 1. Anterior urethral valve: RGU shows linear filling defect in the penile urethra (yellow arrow).

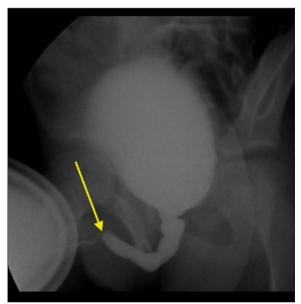


Figure 2. MCUG shows dilatation of the proximal urethra with a smooth bulging at the level of the anterior urethra (yellow arrow).

linear incomplete filling defect in the penile urethra arising from the ventral aspect of urethral wall (Figure 1). VCUG showed mild dilatation of the anterior urethra ending in a smooth bulge. The urinary bladder showed mild irregularity of the outline but there was no vesico-ureteric reflux (VUR) or urinary bladder diverticulum (Figure 2). Based on the classic imaging features, the anterior urethral valve was defined as a diagnosis. The patient underwent cystourethroscopy. It revealed valve-like leaflets of the penile urethra arising from the ventral aspect or urethral wall (Figure 3). In the same session, fulguration was performed with unipolar Bugbee (Figure 4) under general anesthesia.

Discussion

The anterior urethral valve is a rare congenital anomaly that causes lower urinary tract obstruction in children. It can occur as an isolated entity or in association with a proximal diverticulum; probably representing a spectrum of the disease [1].

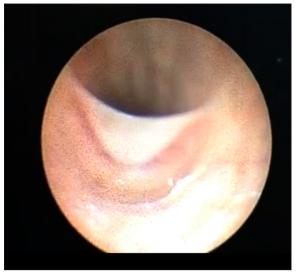


Figure 3. Anterior urethral valve: Valve-like leaflet in the penile urethra on cystourethroscopy.



Figure 4. After fulguration of the anterior urethral valve on cystourethroscopy.

Various proposed hypotheses include an abortive attempt at urethral duplication, failure of alignment between the proximal and distal urethra, imbalanced tissue growth in the developing urethra leading to a remnant of excess tissue acting as a valve, and congenital cystic dilatation of periurethral glands leading to a flap-like valve [2]. Forty percent of the AUVs are located in the bulbar urethra, 30% at the penoscrotal junction, and 30% in the pendulous urethra [3]. The clinical manifestation is highly variable and depends on age of the patient and degree of obstruction. It may present with severe obstruction and bilateral severe hydrouretero-nephrosis, end-stage renal disease, and even bladder rupture [4]. It is a posteriorly directed semilunar fold and arises from the anterior urethra. It can mimic an anterior urethral diverticulum, but the posterior lip is absent in the valve [5]. It can very rarely be associated with posterior urethral valve [6].

VCUG is the diagnostic modality of choice in the diagnosis of anterior urethral valve. It can reveal a dilated or

elongated posterior urethra, a dilatation of the anterior urethra, a thickened trabeculated bladder, a hypertrophied bladder neck, VUR, and urethral diverticula. The urethra appears dilated proximal to the valve and narrowed distal to it on VCUG. A valve may be demonstrated on RGU as a linear filling defect along the ventral wall, or it may show a dilated urethra ending in a smooth bulge or an abrupt change in the caliber of the dilated urethra on VCUG [7]. VCUG may also reveal an associated anomaly in addition to demonstrating the valve. One-third of cases have shown VUR and upper tract deterioration was present in one-half of cases [8]. Urethroscopy usually helps in confirming the diagnosis. The treatment includes the destruction of the valve by electrocautery or by a resecting hook [8].

Conclusions

Congenital anterior urethral valve is an uncommon but important cause of infravesical lower urinary tract obstruction that is more common in male urethra. It can occur as an isolated AUV or in association with diverticulum. Both of these entities probably represent the spectrum of the disease [1]. AUV is more common in the bulbar urethra (40%) while it occurs at the penile urethra in 30% of cases and at the penoscrotal junction in 30% of cases. Few cases have been described in the navicular fossa [2]. It has also been described in adolescents and adults [3] and in association with VATER anomalies. AUV may be associated with other congenital anomalies of the urinary system; therefore, a full evaluation of the urinary system is essential [4].

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