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# Anterior urethral valve, a rare cause of bilateral vesicoureteral reflux in a two year old boy: A case report and a review of the literature

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***Abstract***

Anterior urethral valve (AUV) is a rare cause of obstructive uropathy in children. These are much rarer than posterior urethral valves. We report a case of a child with AUV and high grade bilateral vesicoureteric reflux. He was managed by transurethral laser ablation. The left sided reflux resolved after ablation. The right sided reflux persisted. The child is awaiting ureteric reimplantation.

***Keywords***

Anterior urethral valve; vesicoureteral reflux; obstructive uropathy; valve ablation; children.



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## Introduction

Anterior urethral valve (AUV) is an uncommon cause of obstructive uropathy in children and is 7–30 fold less common than a posterior urethral valve (PUV) [1,2]. The first description of this congenital anomaly was reported by Watts in 1906 and, the first report on AUV was published by Williams et al., in 1958 [3,4]. It has wide variety of clinical presentations, depending on the age of the patient and the degree of obstruction [5]. The diagnosis of AUV is difficult and can be missed; this situation may lead to a spectrum of complications ranging from simple urethral dilatation to bilateral hydroureteronephrosis resulting in end-stage renal disease (ESRD) [4,6]. Voiding cystourethrography (VCUG) is the imaging modality of choice, but endoscopic visualization is always required to confirm the diagnosis [5]. The definitive

management is transurethral fulguration of the valves, reserving vesicostomy or urinary diversion for those with persistent symptoms, or premature patients with very small urethral calibers [7]. Pre-operative azotemia, vesicoureteral reflux (VUR) and recurrent urinary tract infections (UTI) have been implicated as the prognostic factors [8]. We herein present a case of AUV that resulted in bilateral hydroureteronephrosis.

## Case report

A 2 year old boy was admitted with complaints of fever with chills, straining during micturition, poor stream and dysuria since 1 month. Past history suggested multiple episodes of UTI. He had previous history of fulguration of anterior urethral valves at one month of age and circumcision at seven months of age done at a private hospital. Blood hematological investigations and renal profile were normal (Serum creatinine was 0.97 md/dl). Urine was turbid and urine routine and microscopy showed plenty of pus cells per high power field. Abdominal ultrasound (USG) revealed bilateral moderate hydronephrosis and hydroureter, no calculi, large capacity bladder with no intrinsic lesion, post void residual volume was 180cc; the whole of the urethra was dilated. VCUG revealed dilated proximal anterior and posterior urethra, a large capacity bladder and bilateral grade 4-5 VUR [Fig. 1, 2].

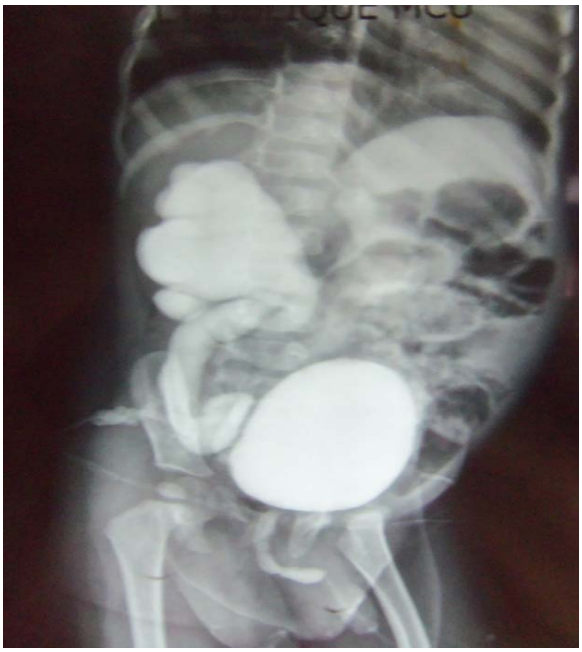

**Fig. 1.** Oblique film of pre-operative VCUG showing dilated posterior and proximal anterior urethra.

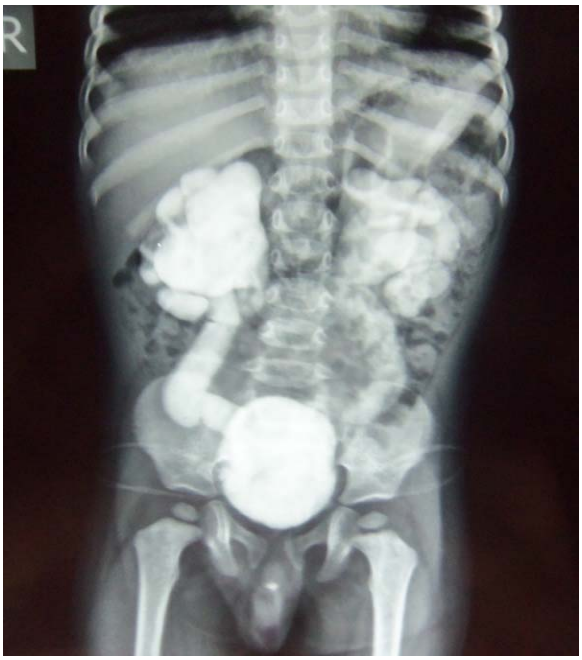

**Fig. 2.** The contrast-filled phase of pre-operative VCUG showing bilateral Grade IV-V VUR.

Cystoscopy was done and residual anterior urethral valves were found [Fig 3].

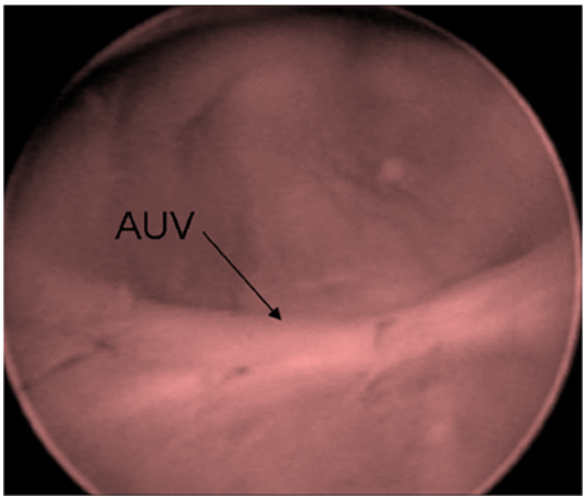


**Fig. 3.** Cystoscopic image showing AUV in the floor of urethra like a semilunar fold.

Laser fulguration of the residual valve was done. Patient was started on chemoprophylaxis for VUR and kept on regular follow-up. After one year, VCUG was showed right side grade 5 VUR. The left side VUR had resolved [Fig 4].

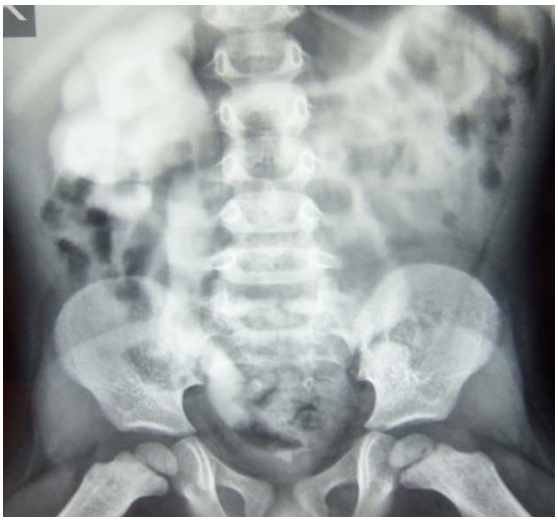

**Fig. 4.** VCUG done one year after AUV fulguration showing VUR only on Right side.

The child had recurrent breakthrough infections on chemoprophylaxis. Hence, ureteric reimplantation was planned.

## Discussion

An anterior urethral valve is a posteriorly directed semilunar fold arising from the floor of the anterior urethra and causing urethral obstruction during micturition [9]. The embryology of AUV remains controversial. However, the embryological mechanisms implicated in AUV are an abortive attempt at urethral duplication [10], and failure of alignment between the proximal and distal urethra [11]. For these lesions, no family pattern of inheritance has been detected. An AUV can be located anywhere in the anterior urethra as follows: bulbous 40%, penoscrotal junction 30% and penile 30% [12]. Associated malformations with AUV are rare, despite the many cases of an association between diverticula and prune belly syndrome [7].

The clinical presentation of AUV is highly variable, depending on age of patient and the degree of obstruction. Antenatally diagnosed patients usually present with bilateral hydronephrosis and in severe cases, they may present with megaureters and/or megacystis [13]. Antenatal diagnosis, when not made, unless suspected, diagnosis can be difficult. The most common symptoms

include difficulty in voiding, dribbling on micturition, incontinence, poor urinary stream and recurrent UTI, enuresis, spontaneous bladder rupture, bilateral severe hydroureteronephrosis, azotemia, urinary ascites and the most catastrophic of all, ESRD [14-16]. In one-third of patients, there is VUR, and in half of them, upper tract damage is present [17]. In our case, UTI and poor urinary stream were the most common symptoms.

VCUG is the diagnostic imaging modality of choice for AUV [11,13,16]. Typically, the anterior urethra appears dilated proximal to the valve, and narrowed distal to it. In VCUG study, it might represent as either a linear filling defect along the ventral wall, a dilated urethra ending in a smooth bulge or an abrupt change in the caliber of the dilated urethra [2,18]. VCUG also may identify an associated diverticulum, megacystis or any other associated abnormalities [14]. Additionally, in the newborn or infant male with suspected infravesical obstruction, the addition of transpenile voiding US can detect the infrequent AUV in the absence of the more common PUV [19]. In our case, he had bilateral VUR and AUV. Urethrocystoscopic examination usually confirms the diagnosis. However, this procedure was difficult to achieve in premature and young patients because retrograde flow during urethroscopy flattens the valve against the urethral wall [20]. Typically, the AUV might appear as a semilunar tissue flap (cusp like) or a circumferential “iris-like” membrane [16].

AUV can also be associated with an anterior urethral diverticulum, most often a proximal saccular type [21]. The embryological understanding of AUV and diverticula has been greatly discussed. The anterior urethral diverticulum develops from focally incomplete development of the corpus spongiosum or incomplete fusion of a segment of the urethral plate [22]. In imaging studies, whereas the proximal lip of a diverticulum forms an acute angle with the ventral floor of the urethra, in cases with AUV, the proximal part of the AUV forms an obtuse angle with the ventral floor [23]. AUV has been also reported to be associated with PUV [24]. Here, in our case, it was not along with the diverticulum.

The purpose of the treatment of AUV is to entirely excise the urethral valve and re-establish the continuity of the urethra. Treatment methods are either open surgical excision with reconstruction or transurethral ablation. Transurethral valve ablation can be cured with a laser, electrocautery or cold knife [13]. In centers where pediatric cystoscope is not available or in cases of very low birth weight neonates, a temporary

cutaneous vesicostomy may be performed [13]. Mizuno et al [25] performed prenatal intervention for a patient with severe AUV. Although the efficacy of this intervention on renal function awaits further evaluation, they believed that pulmonary maturation was successfully attained, and the exacerbation of renal function problems was prevented. The complications of open surgical repair can be urinary extravasation, stricture and urethrocutaneous fistula, whereas cystoscopic ablation is associated with urethral stricture requiring urethral dilatation [18]. Laser fulguration of the residual valve was done in our case.

The prognosis of patients with AUV is generally good. Prognostic factors suggested are pre-operative creatinine levels, VUR and UTI [13]. Patients with AUV with high pre-operative creatinine levels and associated with vesicoureteral reflux and urinary tract infections had a 25-fold increase in poor renal outcomes in a study by Routh et al [8,13]. Only low grade VUR (Grade I-II) have been found to show improvement after AUV fulguration in literature; the patients with higher grades of VUR (Grades III-V) required ureteral reimplantation [13]. Hence, as with other pediatric urological diseases, long term follow-up of these patients is essential as in our case.

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