# *Late presentation of posterior urethral valves*

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KHURSIGARA N, MCGUIRE BB, FLOOD H. Late presentation of posterior urethral valves. The Canadian Journal of Urology. 2011;18(3):5739-5741.

*Late presenting posterior urethral valves are very rare and often present ambiguously. The consequence of late detection can be profoundly detrimental, resulting in persistent voiding dysfunction and/or renal failure. We present an unusual case of a patient with posterior urethral valves who presented at the age of 28 years. We review the literature and discuss the clinical features, diagnosis, and treatment of this condition.*

**Key Words:** posterior urethral valves, adulthood, late diagnosis, outcome

## Introduction

Posterior urethral valves (PUV) is an extremely rare phenomenon that often goes undetected in adult males due to the insidious onset of symptoms. The consequences of late diagnosis include long term voiding dysfunction and/or renal failure. We report one case of late presenting PUV and highlight the importance of maintaining a high index of suspicion in these cases.

Accepted for publication November 2010

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## Case report

A 28-year-old male presented with a 2 month history of dysuria. Six years previously he underwent cystolitholapaxy at another institution. His biochemical analysis, urinalysis and ultrasound scan were normal on this admission. Uroflowmetry demonstrated a maximum flow rate of 23.7 mL/s, an average flow rate of 13.7 mL/s, and a postvoid residual of 135 mL. Micturating cystourethrogram (MCUG) revealed a dilated prostatic urethra and reflux into the seminal vesicles and Cowper's glands, Figure 1. The patient proceeded to cystoscopy which demonstrated proximal prostatic urethral dilatation, a full, trabeculated bladder, and posterior urethral valves, Figure 2. Valve ablation using an 8 Ch pediatric Sachse's urethrotome

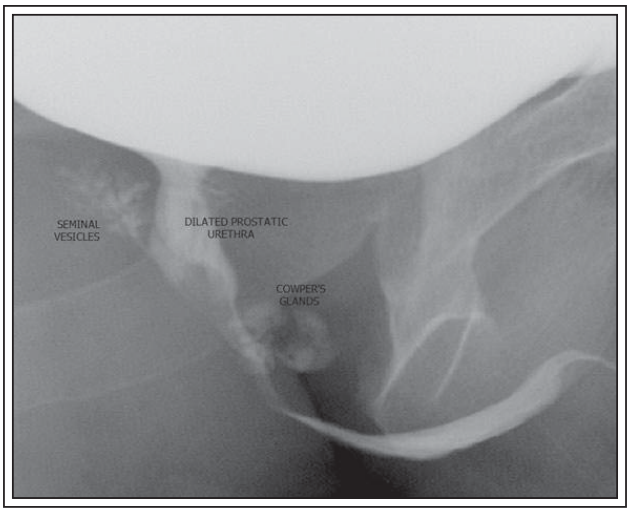


**Figure 1.** Micturating cystourethrogram demonstrating a dilated prostatic urethra, and reflux into the seminal vesicles and Cowper's glands.

with a valve hook was performed. One month postoperatively, the patient was asymptomatic, with a postvoid residual of 0 mL.

## Discussion of clinical features

PUV occurs exclusively in males and has a reported worldwide incidence of 1/5000 to 1/8000 live births.[1,2] Although very rare, it remains the most common cause of congenital lower urinary tract obstruction and the commonest obstructive uropathy leading to childhood renal failure.[3] Most cases are confirmed in utero with ultrasonographic evidence of bilateral hydroureteronephrosis, oligohydramnios and a persistently distended bladder.[4] Antenatal diagnosis allows for early intervention and a reduction of severe and chronic sequelae.

When PUV is diagnosed postnatally, the spectrum of clinical findings is broad, ranging from severe to mild forms.[3] Generally, the more severe the obstruction, the earlier the presentation. During infancy, severely affected males can present with renal failure, congestive heart failure and respiratory distress.[2] When detrusor contraction can overcome PUV obstruction, changes in the upper tract are minimal, allowing PUV to remain silent.[5] In these rare cases, the onset of symptoms is often ambiguous, thus protracting diagnosis until adolescence or adulthood. Voiding dysfunction, nocturnal enuresis, urinary frequency, urinary tract infection and gross hematuria, have been reported in males with late presenting PUV.[6] Our patient's elevated postvoid residual volume and previous history of bladder stones illustrate the subtle nature of symptoms in mild cases of obstruction. Rare symptoms such as postejaculatory dysuria,[7] retained ejaculation,[8] and perineal pain (secondary to refluxing into Cowper's glands)[9] have also been cited in the literature as being related to late presentation of PUV. A delay in diagnosing PUV is further compounded when concurrent comorbidities exist. Benign prostatic hypertrophy, urethral stenosis, prostatitis and urethritis are pathologies that can be potentially confounding when superimposed on late presenting PUV.[10] Delayed presentation of PUV has been estimated to account for only 10% of all PUV cases.[11] However, due to the indistinct and subtle nature of the clinical findings, some believe this to be an underestimation; Hendren proposed that these cases were not uncommon, but just infrequently identified.[12]

## Discussion of diagnosis and treatment

The diagnosis of PUV should be confirmed by micturating cystourethrogram and cystoscopy. The findings on MCUG often include signs of chronic outflow obstruction (trabeculation, diverticulae), a dilated posterior urethra and bilateral vesicoureteral reflux. There is often a clear demarcation between the hypertrophied body of the bladder neck and the dilated posterior urethra which results in the classic spinnaker-sail appearance on sagittal view. Cystoscopy can confirm the presence of the valves although this can often be overlooked without a strong index of suspicion.

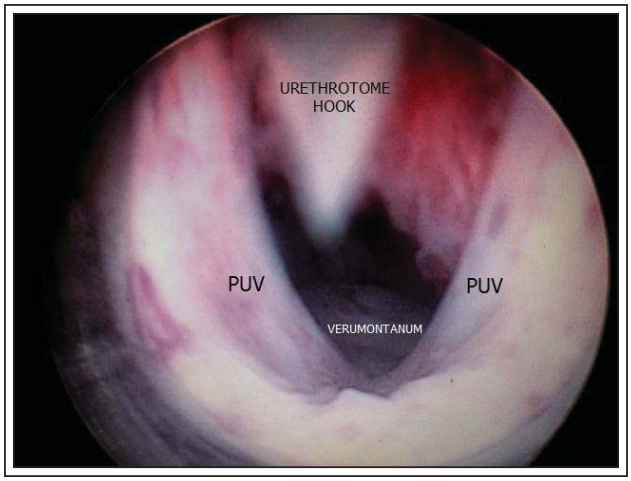


**Figure 2.** Intraoperative video still image demonstrating ablation of posterior urethral valves (PUV) with a Sachse urethrotome.

Most cases reported in the literature responded favorably to surgical valve ablation with a dramatic resolution of symptoms. Irreversible damage to the urinary tract was deemed likely in those with persistent bladder dysfunction post-operatively.[6] Late diagnosis of this rare condition carries a poor prognosis, up to 43% of patients with PUV may progress to end stage renal failure by the age of 30 years.[13] The importance of a high index of suspicion with early diagnosis and treatment is therefore critical.

## Conclusion

Maintaining a high index of suspicion leads to early diagnosis of the rare case of PUV in an adult male with lower urinary tract symptoms. Early detection and intervention is vital to minimizing the possible long term sequelae of permanent voiding dysfunction and/or renal failure. □

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