Double Anterior Urethral Valve Causing Obstructive Uropathy

Akshay Kalavant[1*], Shreesha Nayak[2], Anil B Halgeri[3], Prashant K Zulpi[4]

[1]Associate Professor, Department of Pediatric surgery, Shri Dharmasthala Manjunatheshwara University

[2] Junior Resident, Shri Dharmasthala Manjunatheshwara University

[3]Senior consultant, Shri Dharmasthala Manjunatheshwara University

[4]Assistant professor, Shri Dharmasthala Manjunatheshwara University

***Address for Corresponder:** Dr Akshay Kalavant, [2]Associate Professor, Department of Pediatric surgery, Shri Dharmasthala Manjunatheshwara University (Email: abkalavant2000@yahoo.co.in)

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Abstract

Posterior urethral valve is common congenital cause of lower urinary tract obstruction leading to chronic renal damage. Anterior urethral valve although 7 times less common compared to the former but can lead to similar outcome. There are various case reports suggesting concomitant anterior and posterior urethral obstruction. We are reporting a 2-year-old boy with anterior urethral diverticulum in two places. A high index of suspicion and a thorough micturating cystourethrogram is a key in successful management of the condition.

Keywords

- Double urethral valve
- posterior urethral valve
- anterior urethral valve
- double urethral obstruction
- obstructive uropathy

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Introduction

Posterior urethral valve and less commonly the anterior urethral valves are common cause of the congenital urethral obstruction. The condition may be missed if the clinical features are subtle. The prognosis may be adverse if identified late due to ongoing renal damage. There are sufficient documented cases of double urethral obstruction in the literature caused due to both concurrent anterior and posterior urethral valve. Double anterior urethral valve is a rare entity. There is just once another incidence documented of this kind in the English literature.[1]

Case Presentation

A two-year-old boy was referred to pediatric surgery unit who was being evaluated for grade 3 protein energy malnutrition with the findings of raised renal functional test parameters (blood urea 61 mg/dl and serum creatinine 1.9 mg/dl) associated with mild hydro uretero-nephrosis with loss of cortico-medullary differentiation of both the renal system on abdominal ultrasonography. On enquiring the parents, they did not give any history of obstructive urinary symptoms. On inspection the stream was seen to be thinner than usual. Micturating cystourethrogram showed normal bladder capacity; outline was mildly irregular and no vesicoureteric reflux. Urethral dilations were noted in two region- one in the bulbar urethra and another in the proximal penile urethra.(**Figure 1**) On cystoscopic examination- there was a partially ruptured membrane (probably cathetrization ruptures iris shaped valve) in the proximal penile urethra, through which the 8 Fr compact cysto-urethroscope could be negotiated.(**Figure 2**) There was another obstruction in the form of membrane noted in the bulbar urethra where the tip of the scope was negotiated through its upper margin to enter the bladder(**Figure 3**). Trigone was not well formed. Both the ureteric openings were normal in shape but were placed low in the bladder. Verumontanum was not well developed. Fulgration of the distal valve was done easily. The proximal membrane was thick and could be fulgrated using reasonably high cutting current. Catheter was placed for 10days. Urinary stream improved. The urinary creatinine level dropped to 1.1 mg/dl. The patient is under the follow up since last one year. The patient had to undergo urethral dilatation 3 times in first 6 months and has good urinary stream since

last 6 months. His serum creatine is 1.2 mg/dl.

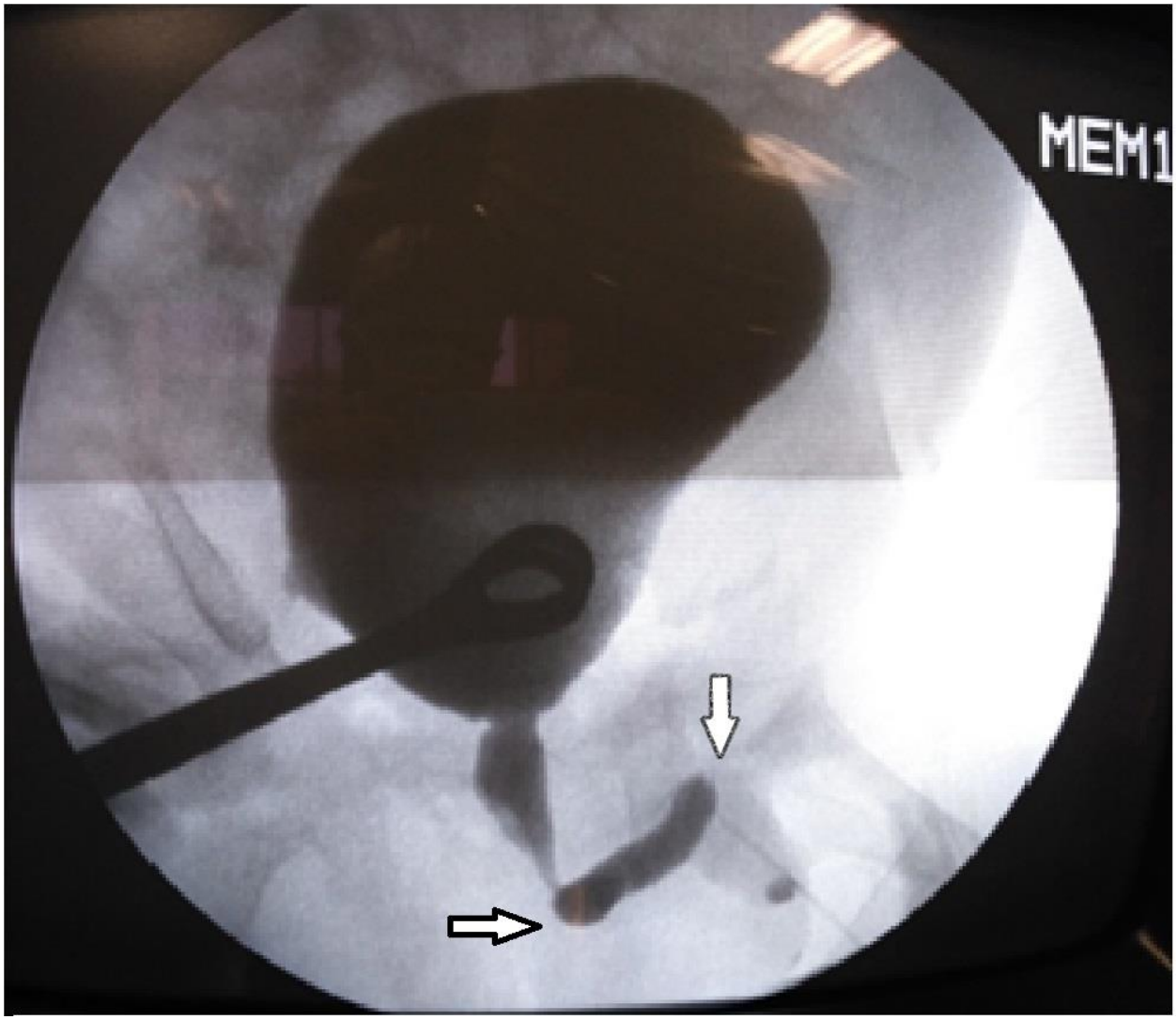

Figure 1: Micturating cystourethrogram showing constriction at two sites. Proximal anterior urethral valve (horizontal arrow) and distal anterior urethral valve (Vertical arrow) are shown in the diagram

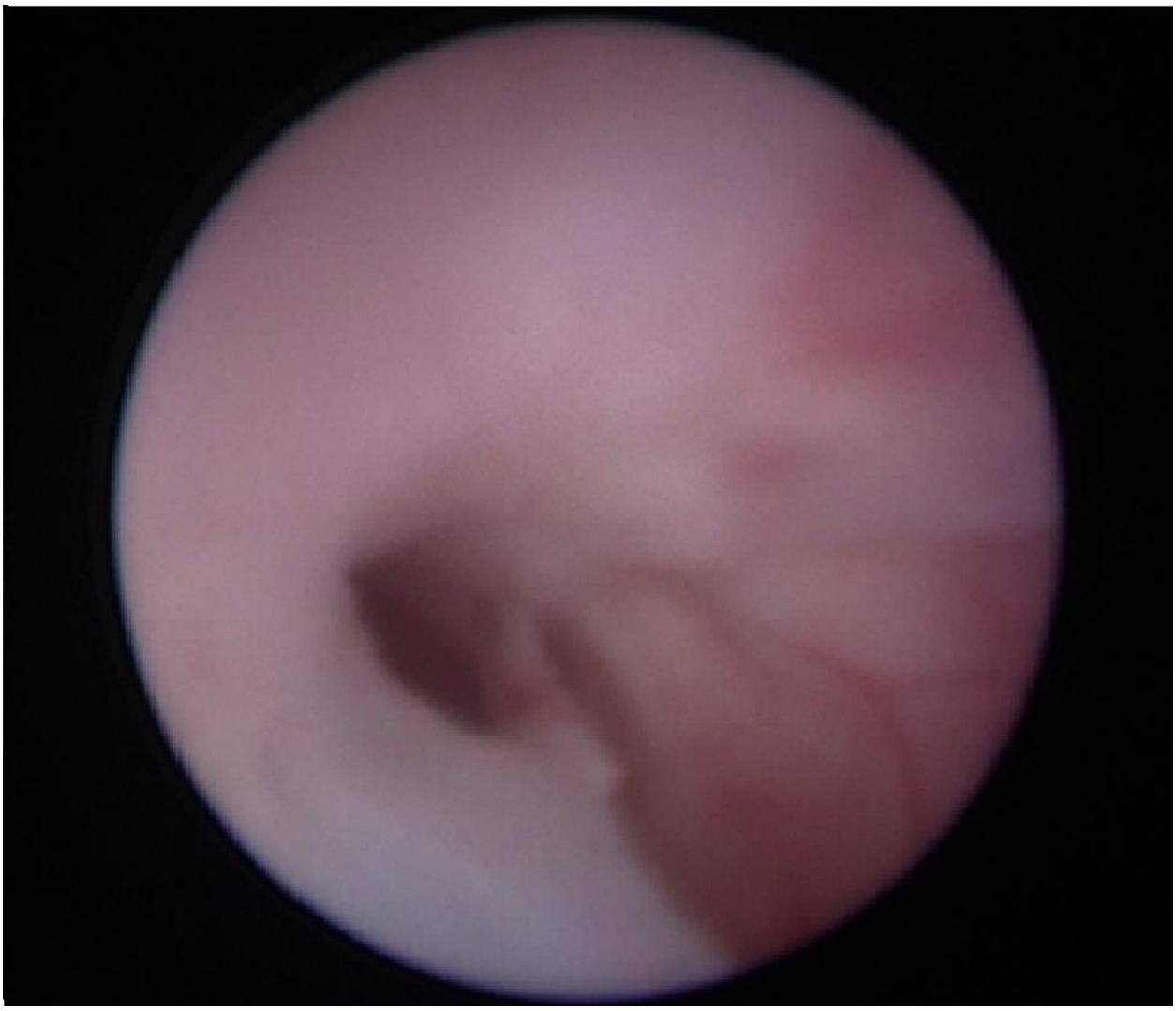

Figure 2: Cystoscopic view of distal urethral valve.

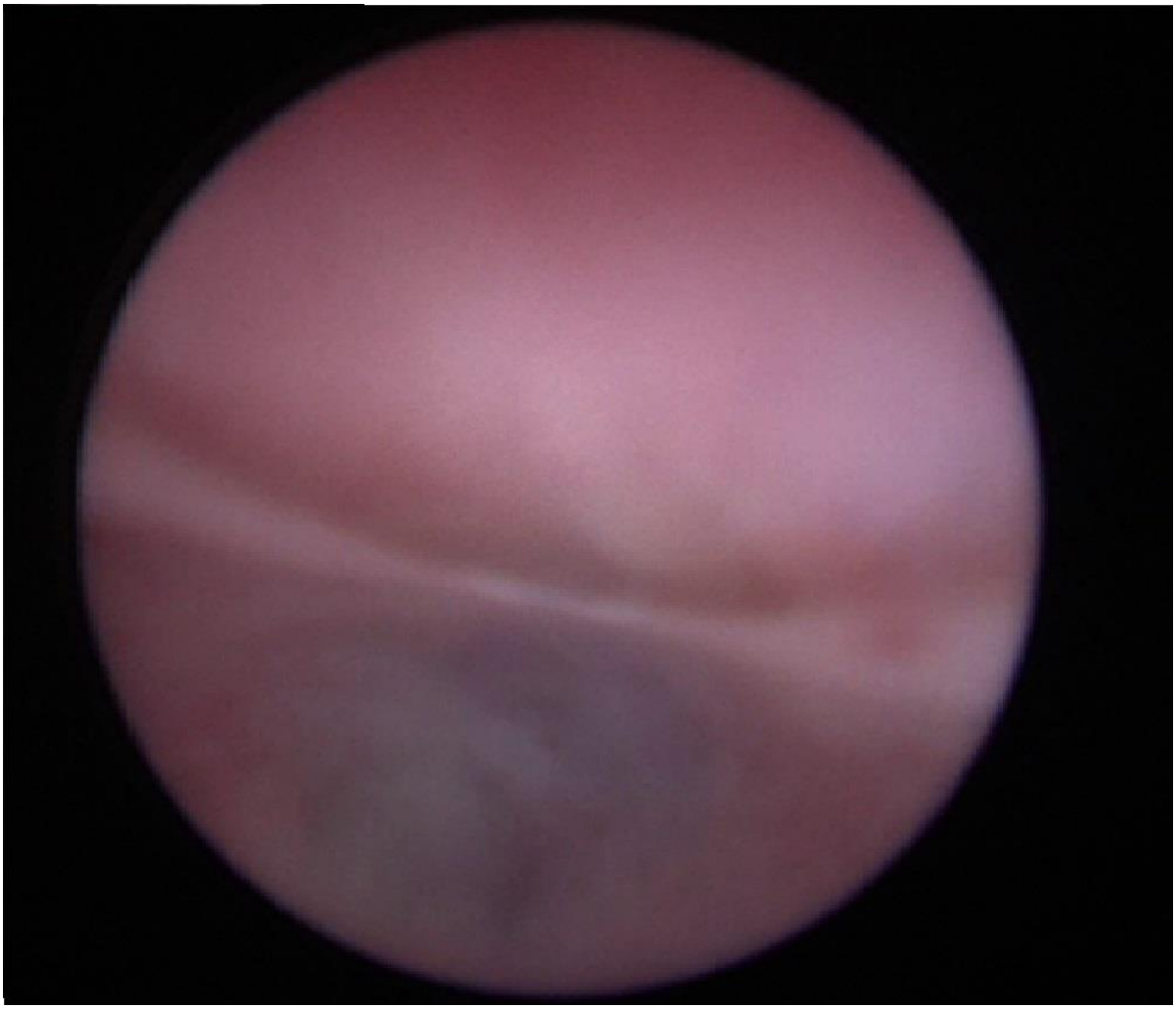

Figure 3: Cystoscopic view of proximal urethral valve. Opening noted at the superior part of the membrane.

Conclusion

Congenital lower urinary tract obstruction is one of the important causes of morbidity and chronic kidney disease. Posterior urethral valve is commonest among them.[2] Anterior urethral valve is seven times less common compared to the former. Anterior urethral valve has been well documented in the literature. One third of the diagnosis is made in antenatal period. Half of the valves are noted in bulbar urethra and rest in the penile urethra. The spectrum of presentation of anterior urethral valve is highly varied. It ranges from subtle urinary obstruction in isolated anterior urethral valve, may be associated with anterior urethral diverticulum and in severe cases may present as bladder dysfunction associated upper tract changes leading to end-stage renal disease.[3] This helps us in determining the mode of treatment required. Many authors consider that anterior urethral valve is always secondary to the anterior urethral diverticulum.[4] Whether valve led to diverticulum or vice versa is also controversial. Few authors have made distinction between valves from diverticulum and have suggested their independent existence.[5] About the one third of the conditions associated with the anterior urethral diverticulum.[3] Diverticulum tends to make an acute angle with the alignment of the urethra whereas dilatation proximal to the valves tend to be in obtuse angle in alignment to the urethra.[6] Our patient didn't have any diverticulum. The prognosis is variable and depends up on the extent of obstructive uropathy that has caused renal damage. Prognosis is seen to be good compared to posterior urethral valves as per some authors. End stage renal disease is seen in less than 5% of the patients.[7] Chronic bladder changes in more than 50% patients were noted in one study in which four out of 11 patients required urinary diversion procedures and 3 among them developed end-stage renal disease.[8] In similar study, all 6 patient except one among 9 cases were associated vesicoureteric reflux carrying risk of renal damage. Compared to the posterior urethral valves many patients with anterior urethral valves required repeat interventions are more likely to suffer chronic renal damage (22%). Urethral diverticulum was noted in only 2.[9] Our patient had mild bladder changes as noted in the MCUG and symptoms although Urodynamic study is awaited. Anterior urethral valves are the semilunar membranous structure arising from the floor of the urethral wall directed

posteriorly and sometimes presents with anterior urethral diverticulum. The shape of the valves may appear iris like or cusp like in cystoscopy. There are various classifications of AUV. There are sufficient evidences to suggest anterior urethral valve could be associated with posterior urethra valve as suggested by 20 documented cases.[10] The embryological origin of the posterior urethral valve and anterior urethral valve are different. Anterior urethral valve and anterior urethral diverticulum is thought to be to the flaw in the union of the glandular and penile urethra. Other theories include incomplete formation of corpus spongiosum and congenital cystic dilatation of periurethral gland.[11] Whereas another author suggests that anterior urethral valves could also develop as abortive process in urethral duplication and incomplete hypospadias.[10] Karnak et al is of the view that valves are developed due to abnormal alignment between the urethral folds in the proximal urethral and distal part of developing anterior urethra leading to subsequent imbalance in tissue growth. Similar valves may be noted in the ventral surface of distal urethra known as valve of Guerine.[5] Bulbar urethra (52%) is the most common site of the development of AUV followed by mid penile urethra (30%). Treatment conssists of trans-urethral fulgration for valves and open urethroplasty for large diverticulum.

There is only one mention of double anterior urethral valve in the English literature [1], which like us had iris like diaphragm distally and cusp like fold in the proximal obstruction. The condition is difficult to identify in the retrograde urethrogram or cystoscopy. The AUV is likely to be missed due to its rarity, hence the micturating cystourethrogram along with the complete urethral demonstration is necessary to identify the condition. MCUG is the diagnostic method of choice. Trans-penile ultrasonography also has been one of the diagnostic modalities.[12]

Ethical Consideration

Written consent for participation was obtained from the parent or guardian of the participant in the study**.** This study was approved by Shri Dharmasthala Manjunatheshwara University.

Acknowledgment

Not applicable

Conflict of interests

There is no conflict of interes

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