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Case Series

Urology Section

Congenital Anterior Urethral Obstruction: A Case Series

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ABSTRACT

Congenital Anterior Urethral Diverticulum (CAUD) and Anterior Urethral Valve (AUV) are rare but well-described causes of urinary obstruction in male children. The clinical presentation can range from isolated poor urinary stream with normal bladder and upper tracts to severe obstructive uropathy with renal failure. In the present case series, six children with anterior urethral obstruction managed between June 2014 and June 2024 were retrospectively identified. Age at presentation, clinical features, investigations, management and long-term follow-up were studied. The age at presentation ranged from newborn to seven years. Severe disease, as evidenced by progressive renal failure, was present in 1 of the 6 children (16.6%). One child had undergone foetal intervention by valve ablation but had progressive renal failure on follow-up. Three children (50%) underwent diverticulum excision with urethroplasty, of which one child developed a postoperative urethrocutaneous fistula. One child with AUV had a dorsal urethral fold causing obstruction, unlike the ventral pathology observed in the other children. CAUD and AUV are rare yet important causes of obstructive uropathy. Careful assessment is crucial for making the correct diagnosis. Temporary diversion may be essential when cystoscopy or primary repair is not feasible or in cases of urosepsis.

Keywords: Anterior urethral diverticulum, Anterior urethral valve, Obstructive uropathy

INTRODUCTION

CAUD and AUV are rare causes of urinary obstruction in male children. They can occur anywhere in the anterior urethra, with approximately 40% in bulbar urethra, 30% in penoscrotal junction and 30% in penile urethra described in literature [1]. CAUD typically occurs as an outpouching on the ventral aspect of urethra through a defect in the corpus spongiosum, leaving a thin-walled urethra with a proximal and distal rim on either side of the defect. The distal end of the diverticulum acts as a flap, obstructing urinary outflow. AUV, however, does not have a proximal rim and the corpus spongiosum is intact [2,3]. Antenatal diagnosis, urinary tract infections, poor stream with dribbling and swelling on the ventral aspect of the penis are common presentations. It is rarely seen in association with posterior urethral valves [4,5]. The condition can result in all features of obstructive uropathy and demands careful examination for diagnosis and management [6].

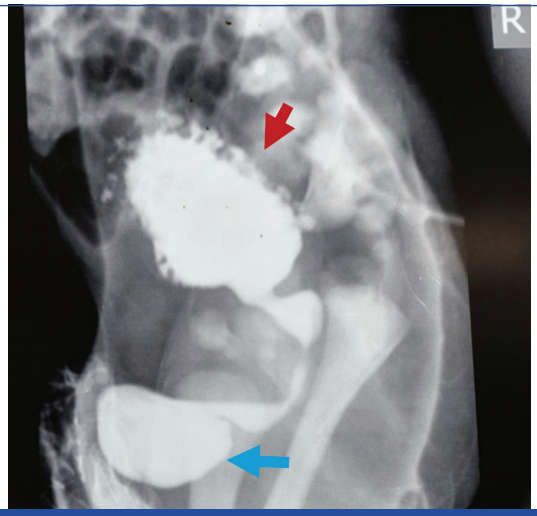
The case series was undertaken after approval from the Institutional Research Centre (IRC/2024/Protocol/208) and Institutional Ethics Committee (GMCKKD/RP 2024/IEC/229).

CASE SERIES

Case 1

A newborn baby boy presented to the outpatient department with dribbling of urine and poor urinary stream in the first 48 hours of life. There was no significant antenatal history. The baby had a palpable bladder. On genital examination, a swelling was noticed in the proximal penile urethra, with urine expressible on its compression [Table/Fig-1]. An Ultrasound (USG) of the abdomen was done which showed a thickened bladder wall with normal bilateral ureters and kidneys. A Micturating Cystourethrogram (MCU) demonstrated trabeculated bladder, absent Vesicoureteric Reflux (VUR) and an outpouching in the ventral urethra, suggestive of an anterior urethral diverticulum [Table/Fig-2]. Excision of the diverticulum with urethroplasty was done at two weeks of age [Table/Fig-3]. The child had a good urinary stream postoperatively and his bladder changes resolved over time.

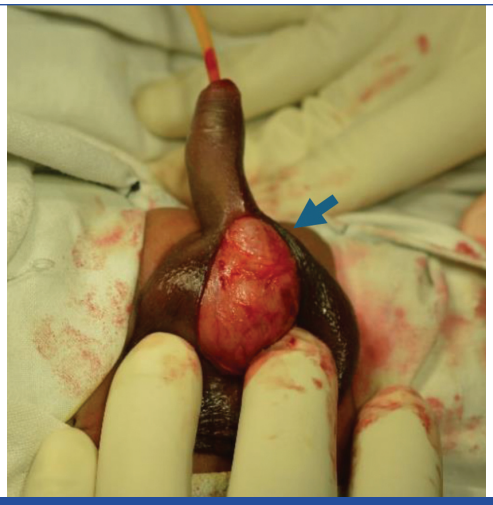
[Table/Fig-1]: Case 1: Clinical picture showing penoscrotal diverticulum (Blue arrow shows diverticulum).



[Table/Fig-2]: Case 1: MCU demonstrating the diverticulum and trabeculated bladder. (Blue arrow points to anterior urethral diverticulum, red arrow shows irregular bladder wall).

Case 2

A one-month-old male infant was referred to the Emergency Department with a vesicostomy, presenting with frank pus discharge



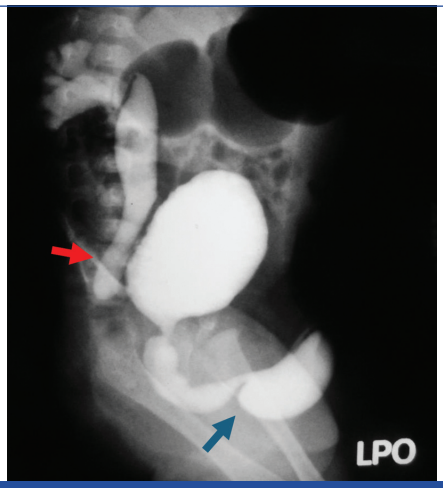
[Table/Fig-3]: Case 1: Intraoperative appearance of diverticulum. (Blue arrow points to anterior urethral diverticulum).

per urethra and decreased vesicostomy drainage. The child had undergone antenatal intervention for urethral obstruction at 30 weeks of gestation in the form of foetoscopic laser ablation of the valve. He was born at term and had undergone cystoscopy with MCU, AUV ablation and vesicostomy under general anaesthesia in the newborn period. At presentation, the child was febrile, with poorly draining vesicostomy and a 2×2 cm swelling on the ventral aspect of penis at the penoscrotal junction, with frank pus discharge on expression of the swelling.

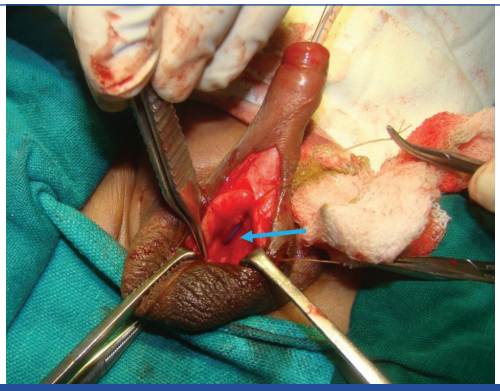
On evaluation, the child had elevated renal parameters. The MCU was reported as trabeculated small bladder with absent VUR. A Magnetic Resonance Urogram (MRU) done at a previous hospital showed bilateral hydroureteronephrosis, right dysplastic kidney and small left kidney with cystic changes and lost Corticomedullary Differentiation (CMD). A higher diversion was done as a salvage procedure, along with closure of the vesicostomy. The child improved clinically and is thriving well at three years of age, but has progressively rising creatinine levels. He is on renal supportive measures at present.

Case 3

A four-month-old boy presented with a poor urinary stream noticed from birth. He also had history of two episodes of febrile Urinary Tract Infection (UTI) at one month and three months, for which he was treated with antibiotics. His mother had noticed a swelling on the undersurface of the penis that increased in size while voiding. An USG was suggestive of right hydroureteronephrosis and a MCU showed right grade IV VUR with a trabeculated bladder and a saccular defect in the anterior urethra [Table/Fig-4]. Primary repair with excision of the diverticulum and urethroplasty was done [Table/Fig-5]. The child developed a urethrocutaneous fistula at the repair site, that was closed after six months. His VUR resolved and bladder changes reversed on follow-up.



[Table/Fig-4]: Case 3: MCU demonstrating diverticulum with reflux. (Blue arrow points to diverticulum, red arrow demonstrate grade V vesico-ureteric reflux).



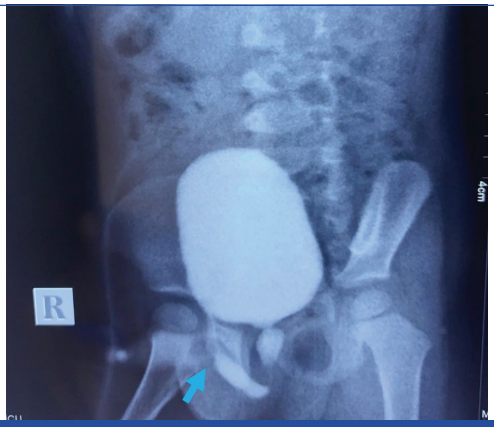
[Table/Fig-5]: Case 3: Intraoperative diverticulum picture. (Blue arrow shows the urethral catheter within the opened diverticulum).

Case 4

A one-and-a-half-year-old boy presented with history of thin urinary stream and straining on micturition that had been present from birth. His external genitalia appeared normal on examination. An USG showed normal bilateral kidneys and ureters, but catheter could not be passed for the MCU due to distal urethral obstruction. On cystoscopy, he had a membranous valve on the dorsal aspect of urethra at the level of corona, that was ablated with resectoscope. Postoperatively, child had good urinary stream on follow-up.

Case 5

A two-year-old boy was admitted with poor urinary stream noticed by the parents from birth. His antenatal scans were normal and he had no history of urinary tract infections. On clinical examination, there was no noticeable swelling in the external genitalia. His USG showed a trabeculated bladder with bilateral normal kidneys and ureters. The MCU was suggestive of an AUV [Table/Fig-6]. Cystoscopy showed a ventral valve at the level of proximal penile urethra. The valve was ablated at 6 o'clock position with resectoscope. Child had a good urinary stream on follow-up.



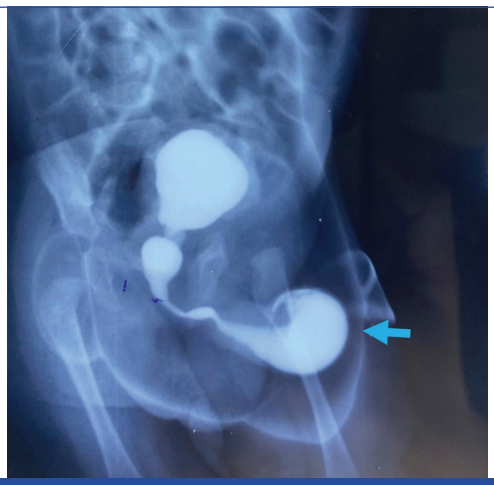
[Table/Fig-6]: Case 5: MCU with Anterior Urethral Valve (AUV). (Blue arrow points to the position of valve).

Case 6

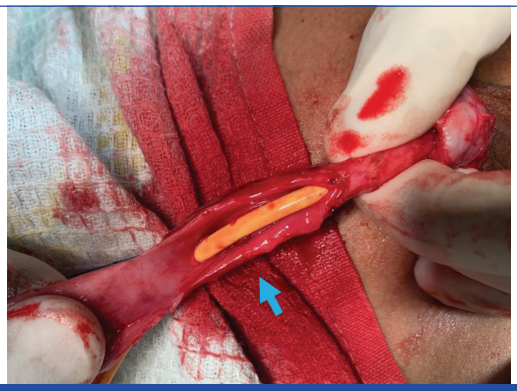
A seven-year-old boy presented with persistently poor urinary stream and dribbling. On examination, an obvious swelling was noticed on the ventral aspect of the distal penis and a catheter could be negotiated only on compression of the same. He had a history of undergoing cystoscopy and fulguration twice for posterior urethral valve. His MCU showed a large diverticulum on the distal penis, along with posterior urethral dilatation and absent VUR [Table/Fig-7]. A MRU was suggestive of bilateral hydroureteronephrosis. A nuclear renogram Dimercapto Succinic Acid (DMSA) showed normal kidneys without scarring. On cystoscopy, a large diverticulum, 0.5 cm proximal to the meatus, was seen with deficient corpus

spongiosum. Excision of the diverticulum with urethroplasty was done over a 10F Foley catheter [Table/Fig-8]. The child had a normal urinary stream post-procedure.

The summary of all the cases has been presented in [Table/Fig-9].



[Table/Fig-7]: Case 6: MCU demonstrating distal penile diverticulum (blue arrow). (Blue arrow shows opened diverticulum with catheter inside).



[Table/Fig-8]: Case 6: Intraoperative picture.

corpus spongiosum, epithelial nest sequestration after the closure of urethral folds, aborted urethral duplication and incomplete hypospadias [2,7-9]. Clinical presentations include antenatal diagnosis, poor urinary stream, swelling noted on the ventral penis, UTIs, urosepsis and renal failure. MCU is the investigation of choice in majority of cases, with USG, Retrograde Urethrogram (RGU), DMSA and uroflowmetry forming the complementary modalities in diagnosis and management [3-5,7]. Cystoscopy is the gold standard investigation for both diagnosis and treatment. AUV can be resected using laser, electrocautery, resecting hook, or cold knife [2,7]. Vesicostomy and perineal urethrostomy may serve as temporary diversions in children with urosepsis and renal failure or in preterm neonates where cystoscope cannot be negotiated [10]. Large diverticulum needs excision with reconstructive urethroplasty or plication [11,12].

The age at presentation in this series ranged from newborn to seven years. Severe disease, as evidenced by progressive renal failure, was present in 1 (case 2) of the 6 children (16.6%), which almost correlates with previous studies (18-20%) [4,7,11]. The antenatal intervention done in this child emphasises its feasibility; however, assessment of any clinical benefit would need a larger study. No prior case series have reported on children with antenatal intervention. Three children underwent excision of the diverticulum with urethroplasty and all of them had a fibrous wall forming the diverticulum, with deficient corpus spongiosum. One child with AUV (case 4) had a dorsal urethral fold causing obstruction, unlike the ventral pathology seen in the other children. This has been reported in only one previous study [4]. Two out of 6 children (33%) had distal penile obstruction, 1 out of 6 had penile obstruction (16%) and 3 out of 6 children (50%) had penoscrotal obstructing lesion. The complication noted in the study was a urethrocutaneous fistula in one child; however, this was repaired in a second surgery without issues. Urethrocutaneous fistula is the most common complication following diverticulum excision with urethroplasty [7,11]. The last child in the series underwent isolated fulguration of the posterior urethral valve twice without the diagnosis of his anterior urethral diverticulum. This re-emphasises the importance of careful clinical examination and detailed cystoscopy in diagnosing this rare condition [6]. Similar cases from the literature have been tabulated in [Table/Fig-10] [3,4,7,11,13,14].

Case No.	Age	Antenatal intervention	Presentation	Position of valve/diverticulum	Procedure	Follow-up
1	Newborn	No	Poor stream	Penoscrotal	Open diverticulectomy	Normal stream
2	1 month	Foetoscopic laser ablation of valve	On vesicostomy with urosepsis	Penoscrotal	Ureterostomy	S. Creatinine 4 mg/dL, on renal supportive therapy
3	4 months	No	Poor stream Febrile UTI	Penoscrotal	Open diverticulectomy	VUR resolved
4	1½ years	No	Poor stream	Distal penile	Valve ablation	Stream improved, MCU- normal
5	2 years	No	Poor stream	Penile	Valve ablation	Stream improved
6	7 years	No	Poor stream	Distal penile	Open diverticulectomy	Stream improved Normal appearing urethra on cystoscopy

[Table/Fig-9]: Patient characteristics.

DISCUSSION

The aetiopathogenesis of AUV and CAUD remains uncertain. Different proposed embryological hypotheses include cystic dilatation of urethral glands, mesenchymal defects in the development of the

Name of author	No. of cases	Children with upper tract involvement	Children with elevated renal parameters	Children with posterior urethral dilatation	AUV/CAUD	Intervention VA/EU/D	Outcome R/RF/E
Present study	6	50% (3/6)	16% (1/6)	16% (1/6)	3/4	3/3/1	5/1/-
Piplani R et al., [11]	7	14% (1/7)	-	42% (3/7)	3/4	3/4/-	7/-/-
Rawat J et al., [7]	14	14% (4/14)	7% (1/14)	-	1/12	1/12/-	12/-/1
Menon P and Rao KLN [4]	27	66% (18/27)	18% (5/27)	18%(5/27)	11/16	16/13/4	21/3/1
Jain P et al., [3]	8	NA	12% (1/8)	12% (1/8)	5/3	7/1/1	8/-/-
Cruz-Diaz O et al., [13]	11	72% (8/11)	27% (3/11)	-	10/1	7/-/4	9/2/-
Prakash J et al., [14]	7	57% (4/7)	28% (2/7)	-	3/4	3/4	7/1/-

[Table/Fig-10]: Comparison with previous published case series [3,4,7,11,13,14].
VA: Valve ablation; EU: Excision with urethroplasty; D: Diversion; R: Resolution; RF: Renal failure; E: Expired; NA: Not available; '-': Nil

CONCLUSION(S)

AUV and CAUD are rare but important causes of urinary obstruction in male children. Antenatal detection and intervention are feasible, but their role in changing outcomes need further evaluation. Early diagnosis and appropriate intervention can cause reversal of upper tract changes and improve outcomes. Excision of the diverticulum with urethroplasty is the treatment of choice for CAUD, as there is deficiency of corpus spongiosum, unlike in AUV.

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