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Case Series

Congenital anterior urethral valves and diverticula: a rare disease with good prognosis

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ABSTRACT

Congenital anterior urethral valves (AUV) are rare congenital anomalies causing lower urinary tract obstruction in children. It can occur as an isolated entity or in association with proximal diverticula. We present our experience of congenital AUV. We did a retrospective review of the medical records From September 2017 to August 2018 to analyze the clinical presentation, investigations, treatment, and outcome of 7 cases of AUV. Two patients were diagnosed as isolated AUV while five presented with associated diverticula. The age of presentation ranged from 6 months to 3 years. Weak voiding stream, swelling on the ventral aspect of the penile urethra and dribbling were the most common symptoms. Renal function was found to be deranged in two patients (28%). Hydro-ureteronephrosis was present in two boys (28%). Post-void residual volume was >20 ml (mean 55 ml) in all children. Transurethral fulguration was carried out on isolated AUV. Open resection and reconstruction were performed in patients with AUV and proximal diverticula. Surgical outcome was successful in all patients except for occurrence of surgical site infection in one patient. We conclude that in isolated AUV transurethral ablation is the treatment of choice. Primary excision and repair are preferred if a diverticulum has formed. Eventual outcomes of AUV are good if irreversible changes have not been established.

Keywords: Anterior urethral valve, Good prognosis, Fulguration, Excision

INTRODUCTION

Congenital anterior urethral valves (AUVs) are rare congenital anomalies causing lower urinary tract obstruction in children.[1] They can occur as an isolated entity or in association with the proximal diverticulum.[2] The valve may be the cause of proximal dilatation that eventually results in a saccular or bulbar dilatation known as an anterior urethral diverticulum (AUD).[3]

The original description of AUV was given by Williams et al, in 1958 and, since then, several cases have been described, separately as well as with proximal urethral diverticula.[4,5] There is a defect in the corpus spongiosum, leaving a thin-walled urethra leading to diverticulum. The distal edge of the diverticulum forms a flap that obstructs the flow of the urine.[6,7] AUD may be found anywhere in the anterior urethra with almost equal incidence at bulbar, penoscotal and penile locations.[1,8] The clinical presentation of AUVs is highly variable, and condition may be overlooked, leading spectrum of complications ranging from simple urethral dilatation to bilateral hydro-ureteronephrosis (HDUN) with renal impairment.[4]

These complications depend on the age of the patient at the time of diagnosis and the degree of obstruction.[3] Different treatment modalities, i.e. endoscopic or open interventions with variable outcomes, are available.[5] The present discussion highlights the clinical approach in identifying this condition and its treatment options.

CASE SERIES

During the 1year period between September 2017 and August 2018, seven boys were diagnosed with AUV. A retrospective review of the medical records was performed to analyze the clinical presentation, investigations, treatment, and outcome of these patients.

All patients were evaluated by history, clinical examination, and investigations including ultrasonography of whole abdomen, serum urea, creatinine and voiding cystourethrography (VCUG). The initial diagnosis was made on the basis of clinical picture (Figure 1a) followed by VCUG (Figure 1b) findings (wide-mouthed diverticulum at the penoscrotal junction) and confirmed by urethrocystoscopy (cusp-like valve or associated outpouching diverticulum (Figure 2a and 2b).

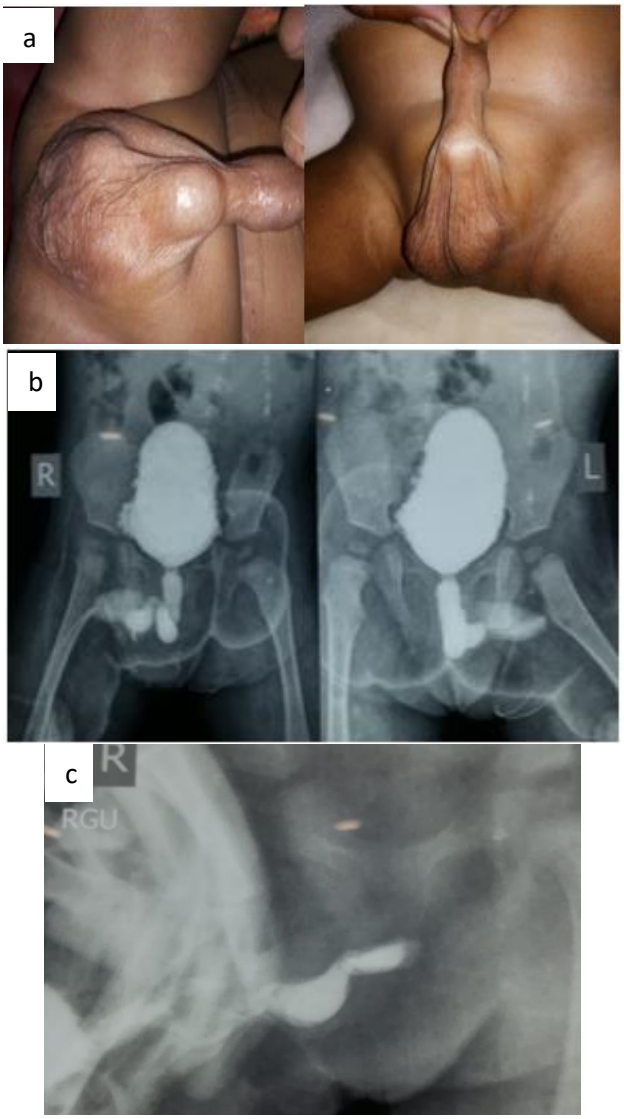

Figure 1: (a) Pre operative pictures depicting swelling on the ventral aspect of the penile urethra; b) VCUG depicting wide-mouthed diverticulum at the penoscrotal junction; (c) RGU depicting diverticulum at the penoscrotal junction

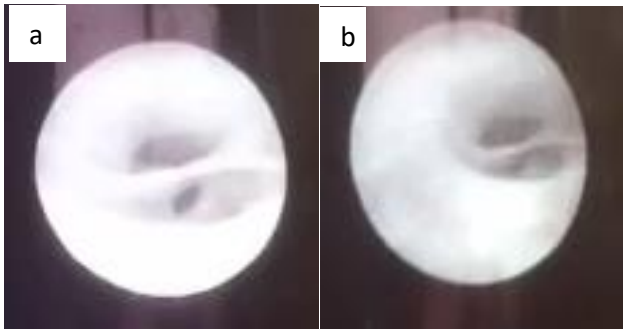

Figure 2 :(a) Urethroscopy picture showing anterior urethral valve; (b) urethroscopy picture showing anterior urethral valve and outpouching on the ventral wall (diverticula).

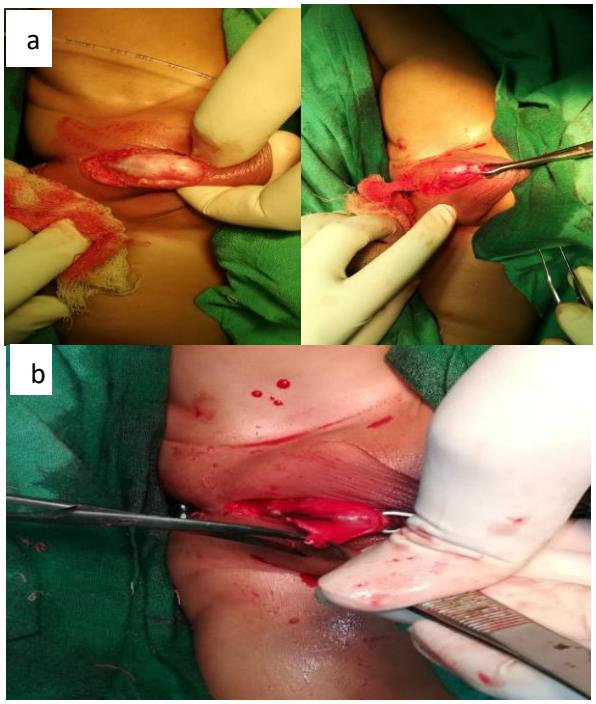

Figure 3: (a) Intraoperative picture (perineal incision); (b) intraoperative picture of tailoring the urethra.

In patients with isolated AUV, transurethral endoscopic fulguration (bipolar cautery) of the valve was carried out using a endoscope (6fr paediatric scope).[5,6] In boys with AUV associated with visible diverticulum, open surgical management was carried out. Postoperatively, all patients were reviewed in the outpatient department by history, clinical examination, uroflowmetry (if feasible) and post-void residual volume (PVRU) estimation at 3 months, and 3 monthly thereafter. Early results, course and complications were evaluated.[5]

Out of the 7 cases, 2 presented as congenital AUV alone, while 5 presented with associated diverticula. The age of presentation ranged from 6 months to 3 years. Weak voiding stream, swelling on the ventral aspect of the penile urethra and dribbling were the most common

symptoms. Renal function was found to be deranged in two patients (28%). Hydro-ureteronephrosis was present in two boys (28%). Post-void residual volume was >20 ml (mean 55 ml) in all children. On cystourethroscopy, cusp-like valves in the anterior urethra were seen in all children (Figure 2a) and associated diverticula were seen in 5 children (Figure 2b). Most common site was the bulbar urethra (57%). In two patients with isolated

AUV (type-1 AUV), transurethral endoscopic fulguration of the valves was carried out.[3] Five patients who had AUV associated with diverticula, (type-4 AUV) underwent open diverticulectomy and primary reconstruction (Figure 3).[3] In which the diverticulum was approached through midline perineal incision and urethra was tailored over 6fr infant feeding tube (IFT) catheter after the excision of the diverticulum, and perineal wound was closed in 3 layers. Postoperatively, one out of 5 patient who underwent open diverticulectomy and repair developed surgical site infection with urinary leakage from completely opened up wound with blocked catheter. He was then taken for secondary suturing of wound over a corrugated drain. He was discharged on day 5 and rest of the post op course was normal.

Postoperatively, on 3 months follow up visit all patients had normal urination without any swelling in the penile urethra and good urinary flow. Uroflowmetry and VCUG done were normal in all cases. On mean follow-up 9 months (range 3-12 months) all patients reported normal urinary stream and minimal PVRU, and disappearance of hydro-ureteronephrosis (HDUN) in both the patients who had it preoperatively.

Table 1: Patient data.

Age	Presenting feature	Site	VCUG	Renal function and USG	Final diagnosis	Treatment
10 months	Dribbling of urine	Bulbar	Bilateral grade v reflux with Hydroureteronephrosis and dialated anterior urethra	Grossly deranged renal function with bilateral hydroureteronephrosis	AUV	Endoscpic ablation
6 months	Swelling on ventral surface of penis	mid penile	Dialated penile urethra with no reflux	Normal	AUD	Open excision and repair
1.5 years	Recurrent UTI	Bulbar	No reflux with dialated anterior urethra	Normal	AUV	Endoscpic ablation
3 years	Swelling on ventral surface of penis	Bulbar	No reflux with dialated anterior urethra	Normal	AUD	Open excision and repair
2 years	Dribbling of urine	Penoscrotal	Dialated penile urethra with no reflux	Normal renal function with thickened UB	AUD	Open excision and repair
2.5 years	Swelling on ventral surface of penis	bulbar	No reflux with dialated anterior urethra	Normal	AUD	Open excision and repair
8 months	Dribbling of urine	mid penile	No reflux with flap like valve in anterior urethra	Normal	AUD	Open excision and repair
12 months	Recurrent UTI	penoscrotal	Bilateral grade 2 reflux with Hydroureteronephrosis and dialated anterior urethra	deranged renal function with bilateral hydroureteronephrosis	AUD	Open excision and repair

DISCUSSION

Congenital anterior urethral valve/diverticulum (AUV/AUD) is a rare condition. The first description of the condition was by reported by Watts in 1906, and the total number of descriptions in the literature is small.[1,9]

The exact embryological origin is not known with various proposed etiological mechanisms including, congenital cystic dilation of periurethral glands, resulting in a flap-like valve, an abortive attempt at urethral duplication, failure of alignment between the proximal and distal urethra imbalanced tissue growth in the developing

urethra resulting in excessive tissue remnant acting as a valve.[10-12] Various studies have shown that AUV may be found anywhere in the anterior urethra with almost equal incidence at bulbar, penoscotal and penile locations while we noticed that 57% were present in the bulbar urethra with penoscrotal locations (28%) and 1 case of mid penile urethra (14%).[1,8]

Obstructive changes accompanying AUVs have been classified by Firlit, who divided them into four basic types depending on the degree of urethral dilatation, presence of diverticulum and the grade of upper tract dilatation.[3] There had been studies claiming for AUV to have familial inheritance. But in our series no such association was present.[12-14]

AUD may present itself at any age, from infant to adult with common presenting complaints include difficulty in voiding, dribbling on micturition, poor urinary stream or recurrent UTI. In the neonates it can be life threatening by causing severe obstruction leading to severe HDUN and azotemia with urinary ascites. Older children with minimal obstruction may present with enuresis, postvoid dribbling or failure to thrive.[4] In the newborn and infant, infectious symptoms predominate, while voiding issues are main symptom in the older child.[15]

A child suffering from AUV will give typical history if poor urinary stream since birth and will mostly also complaints of some cystic swelling at the penile urethra, which when compressed leads to dribbling out of urine from the external meatus, and deflation of the swelling. Dribbling and diminished urinary stream were the most common symptoms of children in our series.[3,16,17] VCUG is the diagnostic investigation of choice for AUVs.[12,18] It shows dilatation of the anterior urethra (Saccular dilatation or uniform urethral dilation) with a small flap-like valve was observed in patients (Figures 1b).

VCUG may demonstrate an associated diverticulum, VUR or any other associated anomaly. Secondary vesico-ureteral reflux (VUR) has been reported in one-third of cases and upper tract changes in 50%.[8,19] Fewer than 5% of patients with AUV progress to renal failure.[16] In our study, VUR was present in 1 patient (14%) and HDUN was present in 3 patients (42%), and these patients presented at a later age. Two patients in our series had deranged renal functions on presentation, which got normalized after intervention.

Cystourethroscopy usually confirms the diagnosis.[2] It is both diagnostic as well as therapeutic. An outpouching from the ventral wall of the urethra having a proximal and distal rim may be seen in cases having simultaneous diverticula in cases of AUV. Various methods of treatment include open urethrotomy and excision of the valve, segmental uretherectomy of the valve-bearing area along with a primary end- to- end anastomosis and transurethral resection of the valves.[20] Transurethral endoscopic resection/fulguration is a minimal invasive and preferred method for isolated AUV. Success rate by cystoscopic fulguration is variable. Five of the 14 patients treated by cystourethroscopy required secondary surgical procedures in one of the large series reported.[21-23] In large diverticula the surrounding supportive tissue is deficient and a poorly draining cavity is likely to be left after transurethral treatment, so open diverticulectomy and primary repair are recommended.[16] It is believed that open primary reconstruction is better as the complete exposure of the diverticulum is possible and the surgical repair offers treatment in a single stage.[24]

The most used surgical option is open excision of the valve/diverticulum with urethral repair. This can be accomplished as a single-stage procedure except in patients with deranged renal function, azotaemia and sepsis. In such instances, one should divert the urinary passage either by prolonged catheter drainage or doing a perineal urethrostomy just proximal to the valve/diverticulum or suprapubic urinary diversion. Complications of Open excision may be extravasation of urine, stricture formation, and urethrocutaneous fistula. Whereas future formation of urethral stricture might be a complication of cystoscopic ablation.[25]

We performed open surgical excision and single stage repair in 5 of our patients (Figures 3) and 2 case with isolated valve underwent endoscopic fulguration of valve. As opposed to posterior urethral valves, the eventual outcome of AUV is good, long-term follow up does not show any residual radiological dilatation and the incidence of chronic renal failure is less than 5%.[16] We followed our patients for an average of 9 months, and all are doing well with renal function unaffected.

Our study had limitations in form of shorter follow up and small number of patients.

CONCLUSION

AUV is a rare diagnosis and high index of suspicion is essential for its diagnosis. It must be considered as the differential diagnosis of lower urinary tract obstruction. These patients if not excessively delayed for treatment are otherwise well in terms of their general condition as opposed to the patients of posterior urethral valves. The diagnosis is easily established by VCUG and severity is revealed by a USG. Endoscopic ablation provides a satisfactory result in isolated AUV and open repair (primary excision and reconstruction) is preferable if a diverticulum has formed. Eventual outcomes are good if irreversible changes have not been established.

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