Congenital anterior urethral valves and diverticula: Diagnosis and management in six cases

Jiledar Rawat, Tanvir Roshan Khan, Sarita Singh, Madhukar Maletha, Shivnarain Kureel

ABSTRACT

Background: Anterior urethral valves (AUVs) are rare congenital anomalies causing lower urinary tract obstruction in children. Although they are referred to as valves, these obstructive structures often occur in the form of a diverticulum. The urethra in these cases shows saccular or bulbar dilatation known as anterior urethral diverticulum (AUD). They typically occur where there is a defect in the corpus spongiosum, leaving a thin-walled urethra. This segment of the urethra balloons out during voiding, simulating a mass that is sometimes visible along the ventral wall of the penis. The swelling is fluctuant and urine dribbles from the meatus on compression. The present study highlights the clinical approach in identifying the condition and its treatment options, especially for those, presenting with urethral diverticula. **Materials and Methods:** We have studied children with congenital anterior urethral valves and diverticula. Six patients of AUVs with diverticula were admitted during the period of 2000-2007 and were prospectively evaluated. The mean age of presentation was 16 months (15 days to 4 years). Straining at micturition and a palpable penile swelling were the most common presenting features. The diagnosis was established by voiding cystourethrogram (VCUG) and supported by ultrasonography (USG). All patients were treated with single-stage open surgical excision except one who died preoperatively due to urosepsis. Initial lay opening of the penoscrotal urethra and delayed repair were done in one patient. **Results:** The surgical outcome was successful in all but one patient, who died of delayed presentation with severe back pressure changes, urinary ascitis and urosepsis. On long-term follow-up, all patients demonstrated good stream of urine. The renal functions were normal and the patients had no evidence of urinary infections. **Conclusion:** We propose that, the patients of AUVs, if not excessively delayed for treatment are otherwise well in terms of general condition as opposed to patients of posterior urethral valves. The diagnosis is easily established by VCUG and the severity is revealed by a sonogram. Open surgical excision is the method of choice for patients with a urethral diverticulum; however, cystoscopic fulguration is also feasible in selected patients. The outcome is excellent with minimal morbidity and mortality.

Key words: Anterior urethral valves, congenital, open surgery, urethral diverticulum

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Department of Pediatric Surgery, CSM Medical University (Upgraded KGMC), Lucknow, Uttar Pradesh, India

Address for correspondence:
Dr. J. D. Rawat,
Department of Pediatric Surgery,
C. S. M. Medical University (Upgraded KGMC),
Lucknow - 226 003, Uttar Pradesh, India.
E-mail: doctrk@gmail.com

INTRODUCTION

Congenital anterior urethral valves (AUVs) are an uncommon condition, causing lower urinary tract obstruction in male urethra. They can occur as an isolated entity or in association with the proximal diverticulum, both of which probably represent the spectrum of disease.[1] It is suggested that the valves cause proximal dilatation that eventually results in a saccular diverticulum.[2] The diverticula typically occur where there is a defect in the corpus spongiosum, leaving a thin-walled urethra. The distal edge of the diverticulum forms a flap that obstructs the flow of the urine. Commonest in the penoscrotal region, approximately one-third is seen in the penile urethra.[3] The clinical presentation of AUVs is highly variable, depending on the age of the patient and the degree of obstruction. Unless suspected, the diagnosis at times may be difficult. The present discussion highlights the clinical approach in identifying this condition and its treatment options.

MATERIALS AND METHODS

Six patients with varied presentations were prospectively evaluated during the period of 2000-2007. The mean age of presentation was 16 months (15 days to 4 years). The patients presented with dribbling of urine, poor stream and ventral penile swelling. The swelling became more prominent while the patient was in the act of micturition. These patients were difficult to catheterize as the catheter got coiled in the anterior urethra, as described elsewhere.[5] There were no associated anomalies in any of the patients. The serum creatinine levels were normal in all patients but one,

who had neonatal presentation with severe bilateral hydrouteronephrosis. Voiding cystourethrogram (VCUG) was diagnostic in all cases and confirmed the dilated urethra, proximal to the valve and narrow distal to it, or by a dilated urethra, ending in a smooth bulge. An urethrogram suggestive of meatal stricture in the presence of normal meatus was suggestive of a valve in fossa navicularis. In addition to defining the lesion in the urethra, VCUG also showed an associated diverticulum, and Vesico-ureteric reflux (VUR). Initial endoscopy was done in all cases preoperatively, to confirm the diagnosis which showed out-pouching on the ventral wall. All patients were operated by open surgical excision by a single surgeon. The operative procedure included an initial degloving of the penis up to its base by a circumferential sub-coronal incision. The urethra was tailored over an appropriate-sized catheter after the excision of the diverticulum, and reinforcement was done by the local dartos tissue. Excessive tissue was excised and the edges approximated. Initial proximal urethrostomy and delayed urethral repair were done in one patient because of the proximally located diverticulum (bulbar urethra). The clinical presentation, investigatory findings and surgical treatments of the patients are summarized in Table 1.

RESULTS

The surgical outcome was successful in all patients except one, who died of severe back pressure changes, urinary ascitis and urosepsis. Postoperatively, urethral obstruction was relieved in all cases, as indicated by a good urinary stream. All patients remained symptom free and had normal renal functions at follow-up ranging up to 3 years. Serum creatinine levels were normal in follow-up and patients were continent and free of infections.Ultrasonography (USG) has showed a complete disappearance of hydrouteronephrosis within a period of 3-6 months. Only one patient had the evidence of urethral stenosis that was presented to us about 1 month back only, and is being planned for cystoscopy.

DISCUSSION

Congenital anterior urethral valve/diverticulum (AUV/AUD) is a rare condition capable of producing significant obstructive uropathy, if not recognized in time or left untreated. The first description of the condition was by reported by Watts in 1906,[4] and the total number of descriptions in the literature is small. The embryological origin remains uncertain; various proposed aetiological mechanisms include an abortive attempt at urethral duplication;[6] failure of alignment between the proximal and distal urethra;[7] imbalanced tissue growth in the developing urethra resulting in excessive tissue remnant acting as a valve[7] and congenital cystic dilation of periurethral glands, resulting in a flap-like valve.[5] AUVs present with a spectrum of variable severity. Obstructive changes accompanying AUVs have been classified by Firlit,[2] as a continuity of pathology, who divided them into four basic types depending on the degree of urethral dilatation, presence of a diverticulum and the grade of upper tract dilatation.

Table 1: Presentation and management of the patients

Age	Presenting feature	Site	Voiding cystourethrogram (VCUG)	Renal function/ sonogram	Final diagnosis	Treatment
1 month	Severe uraemia with neonatal ascitis and shock	Navicular fossa	Bilateral grade V reflux with hydroureteronephrosis and dilated urethra up to the navicular fossa [Figure 3]	Grossly deranged renal parameters with bilateral hydroureteronephrosis	AUV	Conservative and supportive
15 days	Swelling at under surface of penis with straining at micturition [Figure 3]	Penoscrotal	Dilated penile urethra with no reflux	Normal	AUD	Open excision with urethral repair
6 months	Swelling in the penoscrotal area with straining at micturition	Penoscrotal	No reflux with a valve-like flap in penile urethra	Normal	AUD	Open excision with urethral repair
1.5 years	Recurrent urinary tract infection (UTI) with palpable bladder	Bulbar urethra	No reflux with dilated anterior urethra	Normal	AUV	Perineal urethrostomy with delayed urethroplasty
4 years	Recurrent UTI with a small visible swelling along the ventral penis	Penoscrotal	Right grade II reflux with trabeculated bladder and saccular dilation of the anterior urethra	Normal renal parameters with mild hydroureteronephrosis	AUD	Open excision with urethral repair [Figures 4 and 5]
1.5 years	Penile swelling with dribbling of urine	Mid-penile region	No reflux with saccular dilation of the anterior urethra	Normal renal parameters with a thickened bladder on a sonogram	AUD	Open excision with urethral repair



Figure 1: Dilatation of the anterior urethra proximal to the valve

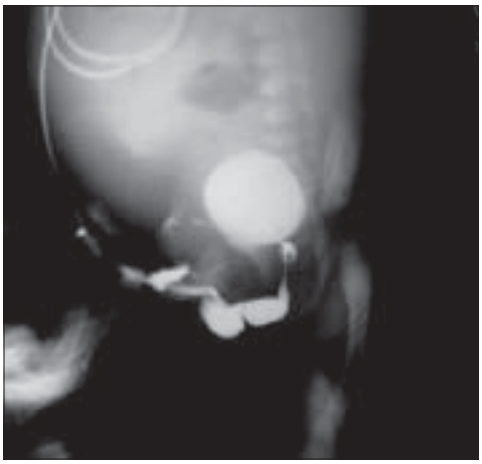


Figure 2: VCUG showing the anterior urethral diverticulum

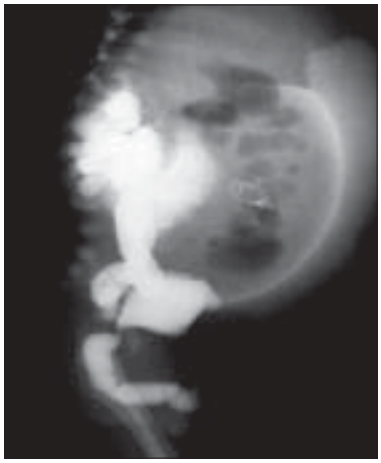


Figure 3: VCUG showing urethral dilatation with VUR and hydronephrosis

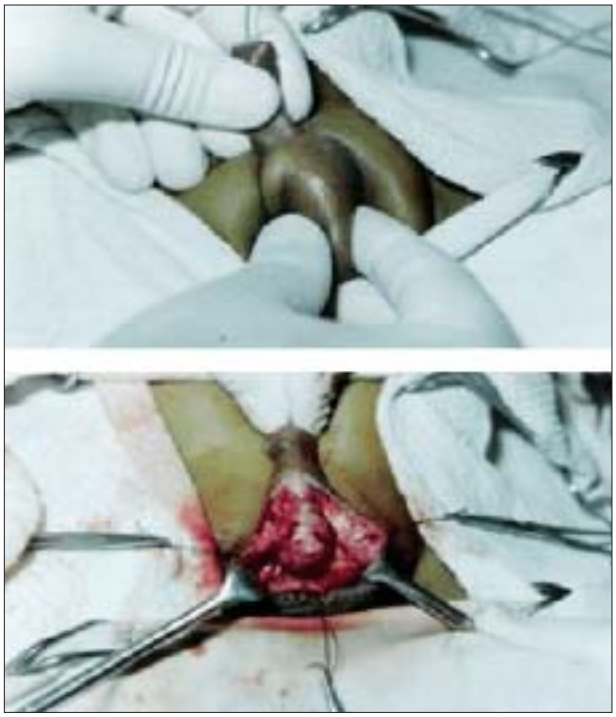


Figure 4: Preoperative and peroperative photograph of the diverticulum



Figure 5: Excision of the diverticulum done

The common presenting complaints include difficulty in voiding, dribbling on micturition, poor urinary stream or recurrent UTI. In the neonatal period or in infancy, AUVs may cause severe obstruction resulting in severe hydrouteronephrosis and azotaemia with urinary ascitis, and usually carries a bad prognosis. The same was true with one of our cases. Older children with minimal obstruction may present with enuresis, postvoid dribbling or failure to thrive.

Patients presenting with recurrent urinary tract infections and straining at micturition are first ordered a USG by the attending clinician. It is only after seeing the back pressure changes a suspicion arises and VCUG is ordered. At times, AUVs may escape diagnosis until adulthood. VCUG is the diagnostic investigation of

choice for AUVs and shows dilatation of the anterior urethra in various forms. Saccular dilatation and uniform urethral dilation with a small flap-like valve were the common types observed in our patients [Figures 1 and 2]. In addition to defining a lesion in the urethra, VCUG may show an associated diverticulum, VUR or any other associated anomaly [Figure 3].

Various methods of treatment include open urethrotomy and excision of the valve,[8] segmental uretherectomy of the valve-bearing area along with a primary end-to-end anastomosis[9] and transurethral resection of the valves. Open excision may result in complications like extravasation of urine, stricture formation, and urethrocutaneous fistula,[10] fortunately not seen in any of our patients. Presence of a valve-like finding will make the option of cystoscopic fulguration feasible. In this manner, urethroscopy can be diagnostic as well as therapeutic. 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.[11] 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.[12]

The most commonly 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 can give a chance of prolonged catheter drainage and wait for the general condition to improve. Another feasible option is doing a perineal urethrostomy just proximal to the valve/diverticulum with delayed excision and repair. Suprapubic urinary diversion is another viable alternative. Few of the series reviewed used this as an initial treatment option for patients presenting with gross pyuria.[12] We performed open surgical excision in all the patients of our series [Figures 3 and 4], after excising the excess tissue, as open primary reconstruction was better providing a complete exposure of the diverticulum and the surgical repair offered treatment in a single stage. The surgical outcome was successful and was comparable to the reported literature.

Long-term follow-up in the patients is also good with milder grades of reflux resolved within 1-2 years of treatment. The back pressure changes to the upper tracts also underwent rapid resolution. A recurrence after surgical correction is not known.

CONCLUSION

Although rare, AUVs must be considered in 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. Transurethral valve ablation with the help of a paediatric resectoscope is feasible in early cases; however, open excision and urethroplasty are indicated in the presence of severe urethral dilatation and deformity. The long-term prognosis is excellent.

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