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Congenital anterior urethral diverticulum in children: case series and review of the literature

Rajat Piplani[1*], Samir K. Acharya[2] and Deepak Bagga[2*]

Abstract

Background Congenital anterior urethral diverticulum (CAUD) is a rare condition in children. This condition can present at any age; however, it is more commonly identified in infants and older children. The patient may present with difficulty in micturition, dribbling of urine, poor urinary stream, or urinary tract infection. Children with large anterior urethral diverticulum with poor spongiosal support may also complain of cystic ventral penile swelling during micturition.

Methods In this series, we report seven such cases of congenital anterior urethral diverticulum treated over a period of 12 years (2008–2020). All cases presented to the Pediatric Surgery Department with dysuria, dribbling of urine, recurrent urinary tract infection, and/or fluctuant ventral penile swelling. They were further evaluated with retrograde urethrography with micturating cysto-urethrogram and cysto-urethroscopy.

Results Three cases had anterior urethral valves that were managed by cystoscopic fulguration of valves. Surprisingly, all these three cases had concomitant posterior urethral valves, while the other four cases presented with relatively larger and saccular anterior urethral diverticulum required excision of the diverticulum and primary urethral reconstruction.

Conclusions The cases with congenital anterior urethral diverticulum secondary to anterior urethral valves may also be associated with posterior urethral valves as seen in our case series. Surgeons should be aware of this association, and both valves should be fulgurated in the same sitting. A larger and saccular anterior urethral diverticulum requires excision and urethroplasty as a definitive procedure.

Keywords Urethral diverticulum, Congenital, Anterior urethral valves, Posterior urethral valves, Cysto-urethroscopy

Background

Congenital anterior urethral diverticulum (CAUD) is an uncommon condition in children. It is a rare cause of lower urinary tract obstruction and has been classified into the saccular variety and globular variety. It can be located anywhere on the anterior urethra but commonly seen at the distal bulbar and proximal penile urethra. The cause-and-effect relationship between the anterior urethral diverticulum (AUD) and anterior urethral valve (AUV) is still debatable. AUVs are congenital semilunar obstructing folds arising from

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the ventral floor of the anterior urethra. It can occur as an isolated entity or in association with a proximal diverticulum, probably representing a spectrum of the disease [1]. Some authors do not distinguish between AUD and AUV, while others believe that these are different entities. The diverticulum that is associated with AUV is not a true diverticulum because in AUD, an acute angle is formed between the proximal part of dilated portion and the ventral floor whereas this acute angle is not present in AUV. Congenital anterior urethral diverticulum may present itself at any age, from infant to adult [2]. The diverticulum and valve cause obstruction of the male urethra, and patient complaints of dysuria, dribbling of urine, recurrent urinary tract infection, or a fluctuant ventral penile swelling [3]. The treatment options in AUD include endoscopic division of the distal lip of the diverticulum [4], excision of the diverticulum with primary repair [5], marsupialization with staged urethroplasty, or even suprapubic diversion followed by definitive repair [6].

Methods.

This combined study was conducted at the Department of Pediatric Surgery at Safdarjang hospital, New Delhi, and All India Institute of Medical Sciences, Rishikesh, and a total of seven cases of congenital anterior urethral diverticulum were included over a period of 12 years (2008–2020). The study was exempted from the need to review by the Institutional review committees in view of retrospective data analysis. All cases presented with a history of dysuria, dribbling of urine with recurrent urinary tract infection, and/or fluctuant ventral penile swelling. These children with ages ranging from 3 months to 10 years were further evaluated with retrograde urethrography (RGU) with micturating cysto-urethrogram (MCUG) and cysto-urethroscopy. Furthermore, a definitive surgery was planned according to the radiological and cystoscopic findings, the presence or absence of urethral valves, and the size of the urethral diverticulum.

Results

Out of the total seven cases, three cases were managed by cystoscopic fulguration of the valves as all of them had concomitant posterior urethral valves (PUV) along with anterior urethral valves. The remaining four cases with relatively large anterior urethral diverticulum required excision of the diverticulum and primary urethral reconstruction. Only one child (case no. 4) had back pressure changes on MCUG with left-sided vesico-ureteric reflux (VUR) associated secondary to PUV (Table 1). Case no. 7 had urethra-cutaneous fistula and underwent repair after six months. However, all cases recovered well with a good urinary stream. On follow-up of these cases, a post-operative MCUG was done in all patients at three months post-surgery.

Discussion

Congenital anterior urethral diverticulum (CAUD) is a rare cause of bladder outlet obstruction in children. It is usually located between the bulbous and the mid-penile area. It is infrequent for an anterior urethral diverticulum to be in the distal urethra near the coronal level, as present in case numbers 5 and 7 of our series. Two varieties of CAUD have been described radiologically: saccular and globular [6]. However, the embryology of AUD still remains uncertain, and various proposed hypotheses have been studied. These include a development defect of the corpus spongiosum, cystic dilatation of the urethral glands, and sequestration of an epithelial nest after the closure of the urethral folds. With a lack of a corpus spongiosum, a urethral dilatation in this region may develop into a diverticulum [7]. Suter proposed the theory that a diverticulum of the urethra develops because of epidermal pockets communicating with the ventral urethral wall. As the anterior urethral tube forms, the urethral groove may leave behind epithelial cells that form a congenital cyst. Cysts in this region developing a communication with the urethra could lead to diverticulum formation as a result of the spontaneous rupture of the cyst into the urethral lumen [8].

Table 1 Back pressure changes on MCUG with left-sided vesico-ureteric reflux (VUR) associated secondary to PUV

S.No. (age)	CAUD on RGU	CAUD on cystoscopy	AUV	PUV	Associated VUR	Surgical procedure
Case 1 (7 years)	+	+	+	+	–	Cystoscopic valve fulguration/resection
Case 2 (10 years)	+	+	+	+	–	Cystoscopic valve fulguration/resection
Case 3 (3 years)	+	+	–	–	–	Excision of diverticulum and reconstructive urethroplasty
Case 4 (3 months)	+	+	+	+	+	Cystoscopic valve fulguration/resection
Case 5 (9 years)	+	+	–	–	–	Excision of diverticulum and reconstructive urethroplasty
Case 6 (4 years)	+	+	–	–	–	Excision of diverticulum and reconstructive urethroplasty
Case 7 (5 years)	+	+	–	–	–	Excision of diverticulum and reconstructive urethroplasty

Most of these children with AUD present with difficulty in micturition, dribbling of urine, poor urinary stream, or urinary tract infection. The clinical presentation varies and depends on the age and the degree of obstruction caused by the associated valves and diverticulum. A careful history will reveal that the child never had a good urinary stream since birth, and a tell-tale sign is a cystic swelling at the penile urethra [9]. On examination, in an uninfected anterior urethral diverticulum without complications, the mass is usually unattached to the overlying skin, non-tender, and mobile laterally. While the urine will be seen dribbling out of the external meatus on compression, the swelling tends to reduce in size. The diagnosis of AUD is typically made by retrograde urethrogram [Fig. 1] or MCUG [Fig. 2]. The presence of a penile or penoscrotal mass clinically and the proximal lip radiologically which is seen as an arcuate filling defect should readily distinguish the diverticulum from the valve [4]. In addition, the proximal lip forms an acute angle with the normal caliber proximal urethra in AUD, whilst in AUV, it forms an obtuse angle [6]. MCUG has an additional advantage of indicating proximal changes like megacystis, VUR, or any other associated anomaly. VUR has been reported in 20% of patients with AUD [10]. Ultrasonography (USG) has a role that compliments the contrast studies to diagnose the condition. It has the advantage of evaluating the upper tracts as well. Moreover, voiding USG has been found to be an alternative to contrast studies in making a diagnosis of AUD [11]. Cysto-urethroscopy is both diagnostic and therapeutic [Figs. 3 and 4]. A diverticulum typically appears as an outpouching from the ventral wall of the urethra and has a proximal and distal rim. In our series, we did cysto-urethroscopy in all the patients including children with CAUD. In one of the significant findings, we observed in these patients during urethroscopy is that a true anterior urethral diverticulum lightens up like a bulb through the defect in the spongiosa that

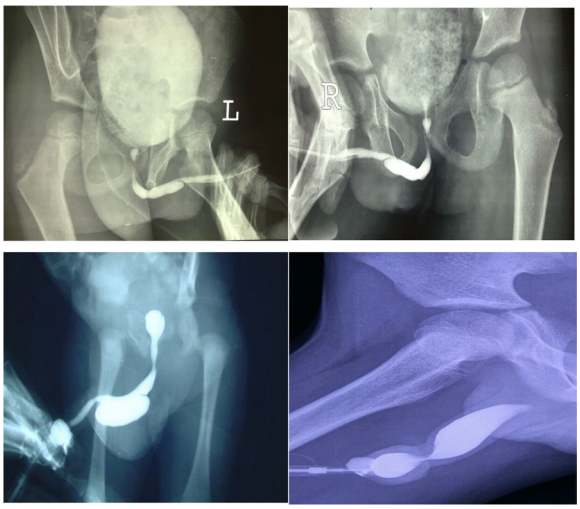

Fig. 1 RGU. **a** Case 1 with AUV. **b** Case 2 with AUV and small anterior urethral diverticulum. **c** Case 3 with large saccular anterior urethral diverticulum at the distal and mid urethra. **d** Case 5 with diffuse anterior urethral diverticulum

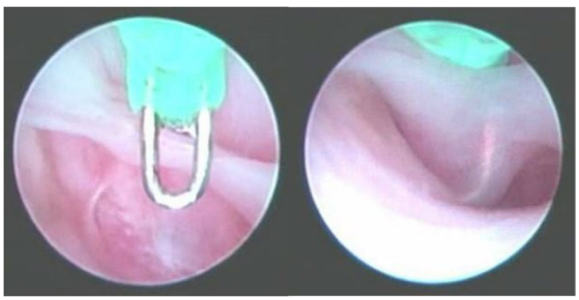

Fig. 3 Cystoscopic view of the anterior urethral valve (case 2) and resection of the valves done

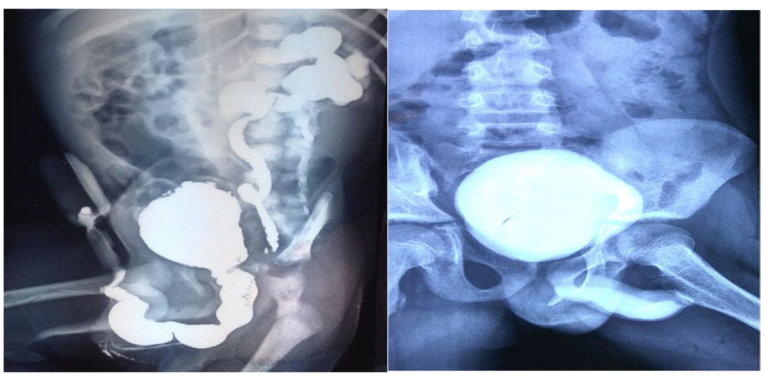

Fig. 2 MCUG. **a** Case 4 with AUV and PUV with dilated posterior urethra and left-sided VUR. **b** Case 5 with diffuse diverticular formation of the anterior urethra

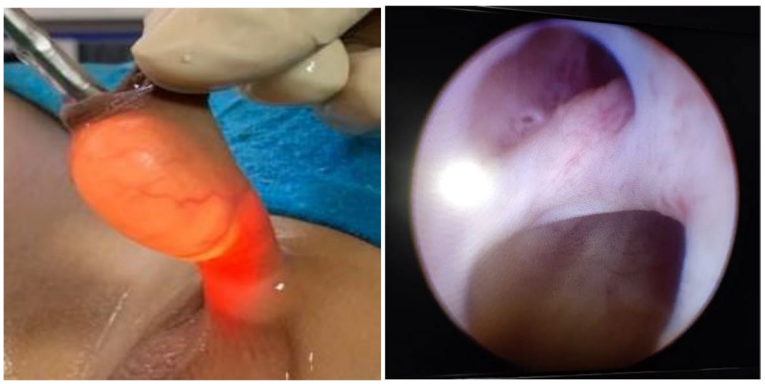

Fig. 4 Cysto-urethroscopy (case 7) showing "orange bulb" sign with large ventral urethral diverticula

typically depicts like an "orange bulb" [Fig. 4]. Moreover, cystoscopy should be done in all patients with obstructed symptoms and upper tract changes.

Diverticula can be described as saccular or diffuse. The saccular type is a localized protrusion from the urethral lumen into the ventral wall of the anterior urethra, and the diffuse type is a generalized dilatation of the entire anterior urethra. A congenital saccular diverticulum may produce anterior urethral obstruction by a valve-like mechanism of its distal lip, which obscures the urethral lumen during filling [12, 13]. The presence of a penile or penoscrotal mass clinically and radiologically, which is seen as an accurate filling defect, distinguishes the diverticulum from the valve. Also, the proximal lip of the diverticulum forms an acute angle with the rest of the urethra, while the anterior valve forms the obtuse angle [14, 15]. The primary differential diagnostic considerations include AUV, dilated Cowper's gland ducts, and posttraumatic diverticulum [14]. In dilated Cowper's gland ducts, a tubular channel is seen in the ventral surface of the bulbous urethra which it parallels. Its termination is in the urogenital diaphragm [14, 15].

Despite being on the spectrum of mesenchymal defects, megalourethra and anterior urethral diverticula are not the same and require different management strategies. Megalourethra are characterized by being primarily non-obstructive, with the well-established associated deficiency of the corpus spongiosum (scaphoid variant) or deficiency of both the corpus spongiosum and one or both corpora cavernosa (fusiform variant). These are rarely seen as an isolated defect and are usually associated with other congenital pathologies of the paediatric urinary tract (e.g., Prune-Belly syndrome) or other systems [16]. Their mode of presentation is usually evident by birth related to the evident swelling of the phallus along with dribbling and repeated infection of the stagnant urethral urine. Given their non-obstructive nature, the MCUG is not associated with any proximal problem in the urinary tract as a direct consequence of its presence. The treatment entails a careful reduction of urethroplasty followed in the long term by penile prosthesis insertion for those with severe corporeal deficiency, whereas AUVs are known to be primarily obstructive in nature and not associated with corporeal deficiency. They cause impedance to urinary flow across the urethra by their distal shelving semilunar lip that collects urine flowing antegradely to the diverticulum which in turn expands and obstructs the urethral lumen. Their mode of presentation is related to their obstructive nature across a spectrum involving antenatal pelvic dilatation, antenatal urinoma due to a ruptured bladder or calyceal fornix, or postnatally by repeated urinary tract infections, poor stream, dysuria, deteriorating bladder function, and a worsening upper tract later in childhood. The mainstay of treatment involves endoscopic de-roofing or incision nullifying its obstructive effect with very rarely a need to resort to surgical excision as an upfront treatment.

The treatment of AUD depends on the degree of obstruction, size of the diverticulum, and upper urinary tract changes. Transurethral resection (TUR) with a pediatric resectoscope is the treatment of choice for small, well-supported diverticula [Fig. 3] wherein the distal obstructing lip is resected [4]. But in the larger and saccular variety of anterior urethral diverticula, as also in our cases, open diverticulectomy and primary repair are recommended [Fig. 5]. This approach may provide a more uniform caliber of the urethra, but there is a risk of urethra-cutaneous fistula formation [17]. The technique of making a triangular flap which is fitted into the distal lip, and double breasting of the urethral suture line has also been described in the literature [5]. Some authors have also advocated the plication of the redundant diverticular

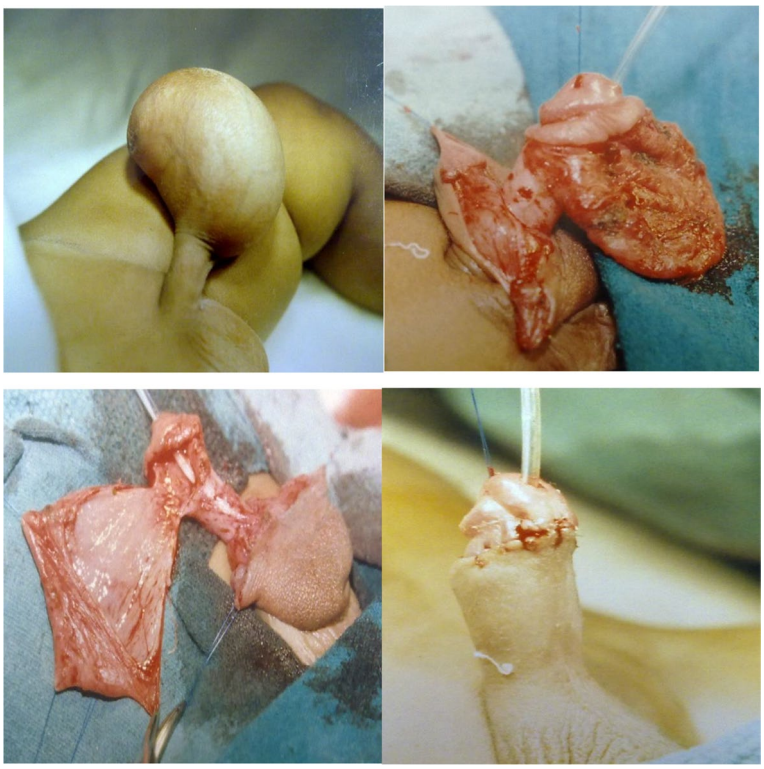

Fig. 5 Case 3. a Preoperative—showing large saccular anterior urethral diverticulum. b Intraoperative—separation of the diverticulum from the surrounding skin. c Intraoperative—excision of the diverticulum. d Postoperative—after urethroplasty

wall with good results [18]. Furthermore, where there are back-pressure changes of the upper tracts with deranged renal function, urinary diversion either by marsupialization of the diverticulum or even suprapubic cystostomy or vesicostomy [4, 6, 19] are better options. Moreover, the prognosis in these patients will further depend on the status of their upper tracts.

CAUD has been associated with various urologic and non-urologic anomalies like vesico-ureteral reflux, anterior urethral valve, penile torsion, patent ductus arteriosus, and polydactyly. However, its association with the posterior urethral valve is a rare occurrence with very few cases that have been previously reported [20, 21]. Although the presence of persistent urinary obstruction after posterior valve ablation usually suggests incomplete valve ablation, it is also good to bear in mind the rare obstructive defects in the anterior urethra as a differential diagnosis in these patients [20]. Concomitant PUV and AUD cases have characteristic micturating cystourethrography (MCUG) features of dilated posterior urethra, non-dilated membranous urethra, and dilated bulbar urethra while retrograde urethrography better delineates the valve along with distal urethra dimensions [21].

Conclusions

A history of poor urinary stream and dribbling, recurrent UTI, and a ventral penile or penoscrotal swelling or mass on examination strongly suggest the diagnosis of CAUD. Micturating cysto-urethrogram and retrograde urethrogram with full-length visualization of the urethra will confirm the diagnosis. Both anterior and posterior urethral valves can co-exist but can be missed on initial assessment. A meticulous cysto-urethroscopy should follow for confirming the diagnosis and endoscopic ablation or resection of the valves for AUV and small but well-supported diverticula. However, in a large saccular and diffuse CAUD, excision of the diverticulum and reconstructive urethroplasty is required.

Abbreviations

CAUD	Congenital anterior urethral diverticulum
AUD	Anterior urethral diverticulum
AUV	Anterior urethral valves
PUV	Posterior urethral valves
RGU	Retrograde urethrography
MCUG	Micturating cysto-urethrography
VUR	Vesico-ureteric reflux
TUR	Trans-urethral resection
USG	Ultrasonography

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None

Authors' contributions
The authors have read and approved the final manuscript. RP drafted the manuscript, contributed to the study design and interpretation of the data, and assisted and operated the cases. SKA revised the manuscript and assisted in the study design and data compilation. DB operated the cases and edited and concluded the study.

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Availability of data and materials
The datasets with more images and patient data are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Written informed consent was obtained from the patient's legal guardian for the publication of the case and any accompanying image.

Competing interests
The authors declare that they have no competing interests.

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