Case Report

Uncommon causes of anterior urethral diverticula in children: Two cases and review of literature

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Abstract

Anterior urethral diverticula are rare in children. Anterior urethral valves and associated diverticulum is the commonly discussed pathological entity in children. There is a lack of awareness among clinicians regarding less common presentations of anterior urethral diverticula in children; which can have a diverse involvement of the urinary tract. This report describes two uncommon presentations of anterior urethral diverticula in children, their diagnoses and management. A systematic differential diagnosis and review of anterior urethral diverticula in children is also presented.

Key Words: Child, congenital urethral diverticulum, cowper's duct syringocele, diagnosis, treatment

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INTRODUCTION

Anterior urethral valves were first described by Watts in 1906 as a cause of urethral obstruction and they are an uncommon cause of urethral obstruction in boys.[1] Anterior urethral valves and diverticula are rare in children.[2,3] Although less common than posterior urethral valves, the obstructive effects of anterior urethral valves and diverticula can be significant and can impact negatively on renal function.[3,4] Involvement of the urinary tract is diverse in anterior urethral valves and diverticula, ranging from mild urethral dilatation to bilateral hydroureteronephrosis with azotemia. Anterior urethral valves and associated diverticulum is the commonly discussed pathological entity in children.[1-3,5,6] Most of these cases are diagnosed on prenatal ultrasonography or in early life when they present with bilateral hydronephrosis and/or azotemia.[1,3]

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There is a lack of awareness among clinicians regarding other uncommon presentations of anterior urethral diverticula (AUD) in children which are not associated with anterior urethral valve. We describe two uncommon presentations of anterior urethral diverticula in children, their possible diagnoses, management and present a brief literature review on AUD.

CASE REPORTS

Case 1

A three year old boy with a background of Russell Silver Syndrome presented to us with a soft, non-tender lump (1.5 cm) with a bluish hue on the ventral side of the penis in the region of the penoscrotal junction which persisted after voiding. [Figure 1] The swelling was compressible and emptied on pressure with associated dribbling. Ultrasonography was performed to exclude renal anomalies. It showed that both kidneys were in orthotopic positions with no hydroureters or hydronephrosis. Micturating cystourethrogram (MCUG) revealed an AUD involving the mid portion of the penile urethra causing extrinsic compression of the urethra. [Figure 2] The urethra proximal to the diverticulum was dilated and there was bilateral grade 2-3 vesicoureteric reflux. The posterior urethra was normal. Cystourethroscopy confirmed a large diverticulum of the anterior urethra without a valve.

A provisional diagnosis of anterior urethral diverticulum was made. The patient underwent an open diverticular repair. A thin walled diverticulum was found extending into the subcutaneous plane. There was a wide communication with the native urethra. The diverticulum was excised and the defect was repaired in layers. He made uneventful recovery and the histology of the wall of the diverticulum revealed attenuated sparse smooth muscle with fibrosis.

Case 2

A 13-year old boy complained of poor urine stream and post void dribbling over the last few years with no documented urinary tract infections. He underwent a cystourethroscopy two years ago at another hospital that showed a narrowing in the anterior urethra and was dilated. He represented with worsening symptoms about a year later. The most recent ultrasonography demonstrated poor bladder emptying with a post-void residual of 250 ml at his first attempt. There was no hydronephrosis or hydroureter. A micturating cystourethrogram showed an urethral diverticulum in the area of the bulb causing extrinsic compression of the penile urethra. [Appendix 1] The urethra was otherwise normal and no vesicoureteric reflux was noted. He was subsequently referred to our service. He underwent a cystourethroscopy which confirmed a large urethral diverticulum with a narrow neck in the anterior urethra close to the bulb. [Figure 3] Urethroscopic examination of the diverticulum revealed a tiny communication in its posterior wall suggestive of a possible communication with the duct of the Cowper's gland. The rest of the urethra appeared unremarkable. The diverticulum was deroofed using a resectoscope knife with cutting current. The patient's symptoms resolved and a repeat urethroscopy 6 weeks later showed no narrowing of the urethra and a slight expansion of the urethra in the area of the diverticulum but was otherwise normal.

DISCUSSION

Congenital urethral diverticula and syringoceles of the Cowper's duct are important differential diagnoses of AUD in children. Clinicians need to be aware and maintain a high index of suspicion in order to enable a precise diagnosis.

Congenital urethral diverticula are seldom thought of as a differential diagnosis in children since the commonest reported differential diagnosis of a diverticulum in anterior urethra is an anterior urethral valve. This commonly presents in the antenatal or the neonatal period, when they cause severe obstructive uropathy. On the other hand, congenital urethral diverticula may present beyond infancy.[5] In that case their presentation may be limited to a ventral penile swelling, incontinence, and/or urinary tract infections.[1,3,5] It remains a matter of conjecture

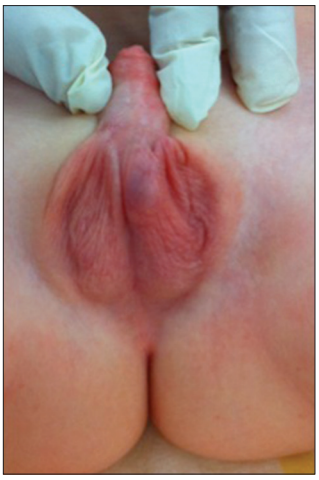


Figure 1: External appearance of a congenital anterior urethral diverticulum presenting as a swelling on the ventral side of the penis in a 3 year old. The swelling was compressible with associated dribbling (Case 1)

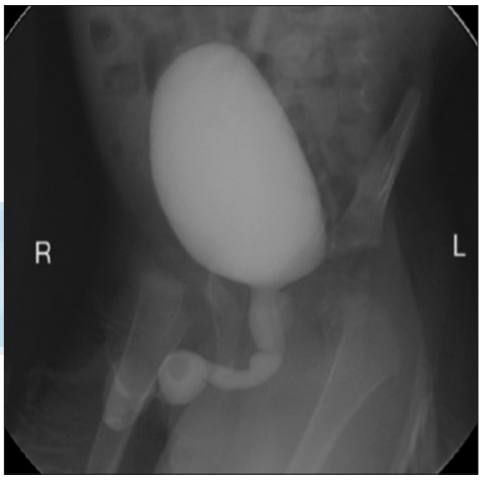
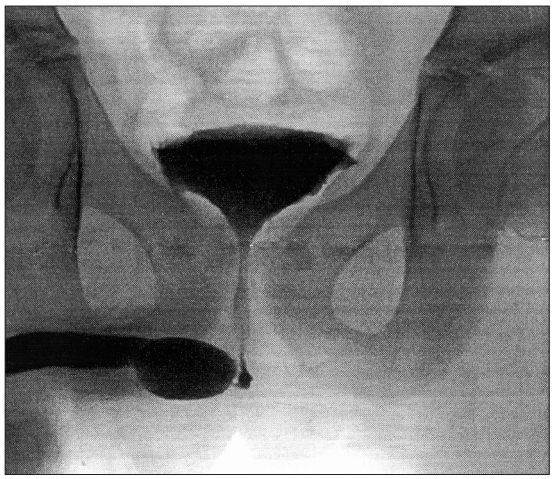


Figure 2: Micturating cystourethrogram showing congenital anterior urethral diverticulum with proximal urethral dilatation and bilateral VUR. Note the reflux of contrast into the prostatic utricle (Case 1)



Appendix 1: Micturating cystourethrogram showing diverticulum in the anterior urethra in the vicinity of the bulb causing extrinsic compression of the urethra suggestive of a syringocele of the Cowper's duct (Case 2)

whether the two represent two ends of the same continuum. Based on Firlit's classification of anterior urethral valve, the second type has a definitive diverticulum and we believe this is a type of AUD.[7] Our first case was atypical because the patient first presented at three years of age and with a perineal swelling.

Compared to anterior urethral valve, syringoceles are infrequently considered as diagnosis in children with AUD. Syringoceles is dilatations of the duct of Cowper's glands. Cowper's glands are two exocrine structures situated in the deep perineal pouch between fascial layers of the urogenital diaphragm.[8] Although more common in adulthood, McLellan *et al*. have reported that in their 40-year period in Boston there were nine children with AUD and in two of them continuity was demonstrated between the Cowper's duct and diverticulum.[1] They have been reported to cause lower urinary tract symptoms in children in the handful of reported cases.[8,9] Although, most paediatric syringocele cases are asymptomatic with signs appearing in adult age, clinicians must consider them as a differential diagnosis of AUD, especially in later childhood. Our second case highlights this point. It is unclear how long the valvular obstruction had been present but the history of an onset of voiding difficulties was only 2 years, suggesting an onset of obstruction in the teenage years. The possible causes of AUD in children have been summarised in Table 1.

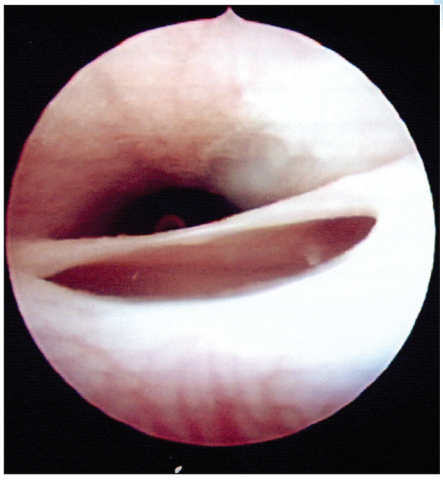


Figure 3: Endoscopic appearance of a "perforated syringocele".(Case 2)

Two possible mechanisms have been suggested for urethral obstruction due to AUD with/without a valve. [Figure 4] During voiding, the obstruction caused by the anterior urethral valve distends the urethral diverticulum just proximal to the valves. This phenomenon in its turn undermines and compresses the proximal urethra, resulting in further increased obstruction.[1,5] Others believe that the diverticulum is the primary lesion and that as the diverticulum fills with urine during voiding its distal lip is forced against the roof of the urethra, obstructing the flow of urine.[10] With progressive enlargement, the distal lip becomes more mobile, thus leading to a valvular obstruction to antegrade flow of urine. In case 2 it appears to have taken some years for the distal lip to form.

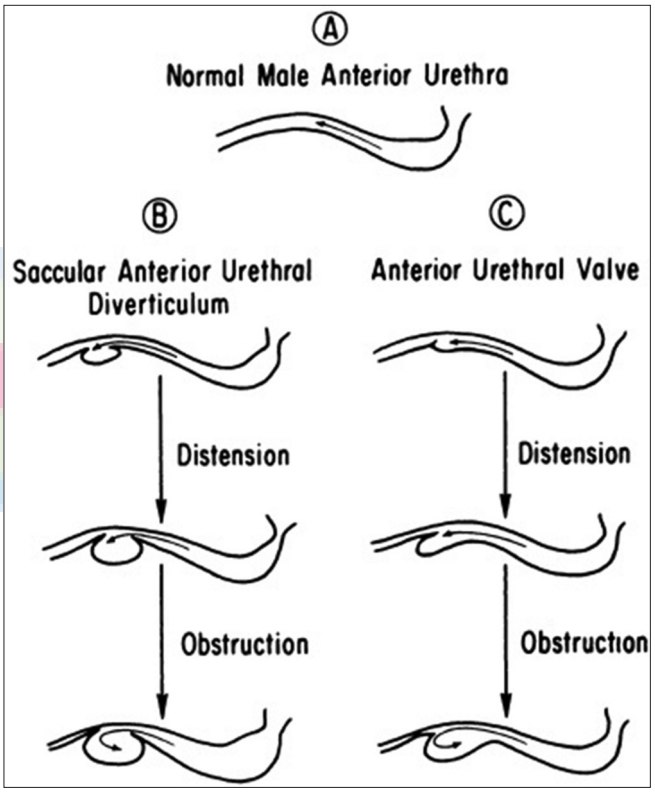


Figure 4: Two postulated mechanisms for explaining urinary obstruction caused due to anterior urethral diverticula in children. Reproduced with permission of Kirks DR, Grossman H. Congenital saccular anterior urethral diverticulum. Radiology. 1981;140 (2):367-72, Radiological Society of North America

Table 1: Causes of anterior urethral diverticula in children and their diagnostic characteristics

Condition	Common presentation	Findings
Anterior urethral valve and diverticulum	Antenatal hydronephrosis and hydroureter	Large diverticulum with distal lip, abnormal bladder, and bilateral hydronephrosis and hydroureter
Anterior urethral diverticulum (congenital)	Swelling, dribbling and urinary tract infections during early childhood	Moderate diverticulum in mid-portion of anterior urethra and mild proximal changes may be present
Syringocele of cowper's gland duct	Urinary tract infections and lower urinary tract symptoms in adolescents	Moderate to large diverticulum in the region of the bulb. cowper's gland duct may be visualized in continuity with the diverticulum
Iatrogenic anterior urethral diverticulum	Terminal dribbling and urinary tract infections	Previous hypospadias surgery

Table 2: Presentation and treatment of anterior urethral diverticula without anterior urethral valves in children: Summary of recently published cases

Authors	Year	Investigations	Presentation	Treatment
Kadian *et al*.[2]	2011	US, MCUG	12 year old with ventral penile swelling and poor urinary stream. Diverticulum in distal penile urethra	Open diverticulectomy
		US, MCUG	6 months old with swelling at the penoscrotal junction and poor urinary stream	Open diverticulectomy
Rawat *et al*.[12]	2009	US, MCUG, endoscopy	15 days old with ventral penile swelling and straining on micturition. No VUR	Open excision and urethral repair
			6 months old with swelling at penoscrotal region and straining on micturition. No VUR	Open excision and urethral repair
			4 years old with small ventral penile swelling and recurrent urinary tract infections. Mild hydroureteronephrosis. Right grade 2 VUR	Open excision and urethral repair
			1.5 years old with penile swelling and dribbling of urine. Thickened bladder on sonogram. No VUR	Open excision and urethral repair
Kibar *et al*.[13]	2007	US, MCUG	9 year old with poor urinary stream, hematuria, urine infections and penile ballooning. Sonogram showed bilateral hydroureteronephrosis. Bilateral grade 3 VUR	Open resection, diverticulectomy and urethroplasty due to size of the AUD
Arena *et al*.[7]		US, MCUG	5 boys age from 2 year old to 9 year old present with urinary tract infection or poor urinary stream or both. Diagnosed as type 2 anterior urethral valves based on Firlit's classification	Transurethral incision and electrofulguration
Howieson *et al*.[14]	2007	US, MCUG, endoscopy	3 week old with dribbling of urine and unwell. Sonogram showed a thick walled trabeculated bladder. Unsuccessful initial MCUG, hence cystogram later showed an anterior urethral diverticulum	Transurethral endoscopic incision
Kumar *et al*.[15]	2005	US, MCUG, endoscopy	15 day old with straining on micturition and passing urine in drops. Sonogram revealed bilateral hydroureteronephrosis and dilated bladder. Left grade 5 VUR, trbeculated bladder, suggestion of posterior urethral valve and faintly seen anterior urethra	Tranurethral endoscopic ablation of posterior urethral valve Post ablation VCUG detected AUD and had open repair of the AUD at 6 months of age

US: Ultrasonograpy, MCUG: Micturating cystourethrogram, VUR: Vesical ureteric reflux, AUD: Anterior urethral diverticulum

The diagnosis of uncommon causes of AUD in children requires a high degree of suspicion and an improved awareness of the variable presentations. In children with lower urinary tract symptoms, micturating cystourethrogram should be performed with careful attention to the urethral anatomy.[1,3,4] AUD are more accurately assessed in voiding films, however, in case of difficulty, retrograde urethrogram should be considered especially as narrowing due to diverticula can be differentiated from strictures on lateral views of voiding films. In congenital urethral diverticula, contrast material fills an oval out-pouching of the ventral aspect of the anterior urethra.[5] A congenital urethral diverticulum usually involves the mid-portion of the penile urethra. AUD associated with anterior urethral valve is not a true diverticulum because in AUD an acute angle is formed between the proximal part of dilated portion and the ventral floor but this acute angle is not present in anterior urethral valve.[2] On the other hand, a syringocele is located in the region of the bulb and contrast may delineate the duct of the Cowper's gland arising out of the posterior wall of the diverticulum. If necessary, retrograde urethrogram (with air contrast) may be used to obtain additional information especially when syringoceles are suspected, however they are rarely more useful than micturating cystourethrogram in children.[5,11]

Irrespective of the underlying aetiology, anterior urethral diverticula in children should be treated in order to prevent complications such as hydronephrosis and renal dysfuction secondary to urinary outflow obstruction and recurrent urinary tract infections.[1,2] A brief review of recently published data on anterior urethral diverticula in children is presented in [Table 2]. This table excludes publications which describe treatment of anterior urethral valve. As seen in [Table 2], the choice of treatment should be guided by the urethroscopic findings. Endoscopic cutting or deroofing should be attempted if the diverticulum has a distal lip which can be engaged and divided using a resectoscope knife or a hook as in case 2. Failing this, open diverticular repair should be considered as in case 1. Some authors choose open urethroplasty if the calibre of the urethra distal to the diverticulum is inadequate to permit safe instrumentation, or there is insufficient corpus spongiosum and periurethral tissue to avoid subcutaneous extravasation with the risk of a urethrocutaneous fistula.[1,3]

CONCLUSION

The uncommon causes of AUD in children include congenital urethral diverticulum and Cowper's duct syringocele. These can present as a lump, with lower urinary tract symptoms and/or low grade obstruction to urethra. The precise diagnosis requires a high index of suspicion and careful delineation of urethral anatomy. Treatment should be guided by urethroscopic findings.

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Commentary

Two rare pathologies of male urethra in children

The authors presented two patients with anterior urethral diverticulum and syringocele of the Cowper's duct managed by open diverticular repair and endoscopic unroofing with successfully, respectively. These anomalies are two different pathologies of the male urethra and rarely seen in children. Those patients often have nonspecific symptoms which contribute to the delay in diagnosis.

Most children with anterior urethral diverticulum have nonspecific urinary symptoms such as poor urinary stream, post-void dribbling, difficulty in micturition, urinary tract infection, enuresis or hematuria. Small diverticula may remain asymptomatic. Cystic swelling on the ventral surface of the penis and firm penile mass due to stone formation are most specific symptoms and signs of the urethral diverticula. Therefore, the first patient's symptom was not atypical. Some patients especially small children or neonates present with signs and symptoms of the severe urinary tract obstruction or obstructive uremia.[1] Older children may present with less severe symptoms.[2]

The patients with Cowper's syringocele may present with urinary tract infection, obstructive voiding symptoms, post-void dribbling, hematuria or dysuria. So the presentation of second patient was not atypical. Silveri *et al*. reported an infant with severe infravesical obstruction caused by ruptured Cowper's syringocele.[3] However majority of the patients with Cowper's syringocele remain asymptomatic in childhood period.

Anterior urethral diverticula and Cowper's syringocele should always be kept in mind during evaluation of a boy with lower urinary tract symptoms. In patients with urethral anomaly suspicion, initial imaging study should be ultrasound. Perineal and detailed urinary system ultrasounds reveal proximal urethral diverticula and changes of urinary bladder and upper urinary tract. The diagnosis of anterior urethral diverticulum or Cowper's syringocele is made by retrograde urethrography, voiding cystourethrography and cystourethroscopy. Second patient showed us that if radiological studies aren't completed properly or cystoscopy isn't performed carefully, misdiagnosis is always possible.

Cystourethroscopy is essential to confirm diagnosis and plan treatment. There are many treatment options based on the patient's condition. Small and asymptomatic lesions may be

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