Case Report

Anterior Urethral Valves

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We studied the clinical presentation and management of four patients with anterior urethral valves; a rare cause of urethral obstruction in male children. One patient presented antenatally with oligohydramnios, bilateral hydronephrosis and bladder thickening suggestive of an infravesical obstruction. Two other patients presented postnatally at 1 and 2 years of age, respectively, with poor stream of urine since birth. The fourth patient presented at 9 years with frequency and dysuria. Diagnosis was established on either micturating cystourethrogram (MCU) (in 2) or on cystoscopy (in 2). All patients had cystoscopic ablation of the valves. One patient developed a postablation stricture that was resected with an end-to-end urethroplasty. He had an associated bilateral vesicoureteric junction (VUJ) obstruction for which a bilateral ureteric reimplantation was done at the same time. On long-term follow-up, all patients demonstrated a good stream of urine. The renal function is normal. Patients are continent and free of urinary infections. Anterior urethral valves are rare obstructive lesions in male children. The degree of obstruction is variable, and so they may present with mild micturition difficulty or severe obstruction with hydro-ureteronephrosis and renal impairment. Hence, it is important to evaluate the anterior urethra in any male child with suspected infravesical obstruction. The diagnosis is established by MCU or cystoscopy and the treatment is always surgical, either a transurethral ablation or an open resection. The long-term prognosis is good. [Asian J Surg 2006;29(3):165–9]

Key Words: anterior urethral valves, infravesical obstruction

Introduction

Anterior urethral valves are rare congenital anomalies of uncertain embryology.1–4 They must be considered in the differential diagnosis of lower urinary tract obstruction. We highlight the varied presentation depending on the age and the degree of obstruction and review the various treatment alternatives.

Materials and methods

We reviewed the medical records of four children who were treated for anterior urethral valves. The data were analysed with reference to age at presentation, clinical features, appearance on micturating cystourethrogram (MCU) and cystoscopy, and treatment and outcome (Table).

The first child presented antenatally with oligohydramnios, bilateral hydronephrosis and thickened bladder at 32 weeks of gestation. The child was born at 35 weeks and the birth weight was 2.9 kg. The child was active with good tone. Both the kidneys and the bladder were palpably enlarged. There was no evidence of pulmonary hypoplasia. The child underwent a cystoscopy on the first day with a size 7.5 French scope. Suprapubic pressure aided in the identification of obstructive crescentic leaflets at 7 and 8 o’clock within the glanular urethra. Further advancement of the scope was aided by the dilatation of the proximal urethra. The bladder was trabeculated. A bugbee electrode

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Table. Clinical features, treatment and outcome of the four children treated for anterior urethral valves

<table>
<thead>
<tr>
<th>No.</th>
<th>Age at presentation</th>
<th>Presentation</th>
<th>MCU</th>
<th>Cystoscopic appearance</th>
<th>Associated findings</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Antenatal USG at 32 wk</td>
<td>Antenatal USG – oligohydramnios, thick-walled bladder and bilateral hydronephrosis</td>
<td>Tight valves within the anterior urethra; dilatation of proximal urethra; no VUR</td>
<td>Crescentic valves in glanular urethra at 7 and 8 o’clock</td>
<td>IVU showed bilateral VUJ obstruction – subsequent bilateral reimplantation</td>
<td>TUR, repeat TUR for residual valves, open resection for tight stricture</td>
<td>Good stream of urine, no UTI, FU cystoscopy – urethra, bladder and ureteric orifices – normal FU IVU – mild clubbing, renal function normal</td>
</tr>
<tr>
<td>2</td>
<td>At birth</td>
<td>Poor stream of urine</td>
<td>Valves of anterior urethra; dilatation of posterior urethra; trabeculated bladder; no VUR</td>
<td>Concentric valves in bulbar urethra</td>
<td>Bilateral hydroureteronephrosis, thick-walled bladder, no VUR, no VUJ obstruction</td>
<td>Initial TU fulguration, check cystoscopy – TU division of residual valves, check cystoscopy – TU resection</td>
<td>Good stream of urine, no UTI, no straining FU USG – no hydroureteronephrosis, renal function normal</td>
</tr>
<tr>
<td>3</td>
<td>At birth</td>
<td>Poor stream of urine</td>
<td>No</td>
<td>Valves in anterior urethra</td>
<td>None</td>
<td>TUR</td>
<td>Good stream, no UTI, no straining, renal function normal</td>
</tr>
<tr>
<td>4</td>
<td>9 yr</td>
<td>Frequency and dysuria, stream – good</td>
<td>Dilated posterior urethra</td>
<td>Valves in anterior urethra</td>
<td>Bilateral grade 1 VUR</td>
<td>TUR</td>
<td>No frequency or dysuria</td>
</tr>
</tbody>
</table>

MCU = micturating cystourethrogram; USG = ultrasonography; VUR = vesicoureteric reflux; IVU = intravenous urogram; VUJ = vesicoureteric junction; TUR = transurethral resection; UTI = urinary tract infection; FU = follow-up; TU = transurethral.
was carefully advanced and pushed towards the valves using coagulation current. This achieved fulguration of the valves. Postoperative MCU did not show any residual valves or reflux. A renal diethylene tetraamine pentaacetic acid (DTPA) scan was done 1 month after ablation. This revealed bilateral hydroureruteronephrosis with delayed excretion (T1/2 right 789 minutes and left side 24 minutes). A check cystoscopy revealed residual valves that were incised to release the obstruction. Although fulguration was performed using a bugbee, for the purpose of incision we used a crochet-shaped hook that was withdrawn from the posterior to the anterior urethra so as to engage the valve leaflets in an antegrade manner and disrupt the valves. Application of an electric current to this hook then achieved a more substantial resection of the valves. It was decided to repeat the cystoscopy at a later date for completing the resection. However, the child defaulted and returned for follow-up only after about 18 months for straining during micturition. Imaging with an excretory urogram revealed persistent bilateral hydroureruteronephrosis due to bilateral vesicoureteric junction (VUJ) stenosis. Cystoscopy was repeated at that time, which revealed a tight fibrotic stricture about 12 mm within the external meatus. An open operation was done and an urethrotomy with excision of anterior urethral stricture was performed. The patient defaulted follow-up again and came back after 2 years. MCU revealed a recurrent stricture within the anterior urethra with proximal dilatation (Figure 1). A planned definitive open urethroplasty (short segment resection and end-to-end anastomosis) with bilateral ureteric reimplantation (Leadbetter) was performed. A temporary nephrostomy (Figure 2) had to be inserted on the right side for worsening hydronephrosis on the 2nd postoperative day. This was removed once nephrostanogram at 2 weeks showed good flow into the bladder. Following the final definitive open urethroplasty and ureteric reimplantation, the patient is asymptomatic and has a good stream of urine without any straining. He has undergone two check cystoscopies at 2-year intervals, which revealed a non-dilated urethra with a normal bladder and ureteric orifices. Intravenous urogram (IVU) after 2 years showed mild clubbing with prompt excretion of contrast at 5 minutes. The renal function is normal.

The second patient was seen and treated elsewhere since birth when he presented with postmicturition dribbling and a palpable bladder. Ultrasound demonstrated bilateral hydroureruteronephrosis and a thick-walled bladder. MCU showed a stenosis in the anterior urethra with proximal dilatation. There was no vesicoureteric reflux (VUR). This was treated with circumcision, meatotomy and urethral dilatation. Urethral dilatation was repeated monthly as part of a dilatation schedule. However, even after more than 2 years, the child continued to have straining at micturition. Repeat MCU was reported as a spasm of the anterior urethra with proximal dilatation and bladder trabeculations. Thereafter, the child was referred to us at the age of 3 years for his poor stream.
of urine. Ultrasound showed bilateral hydroureteronephrosis. MCU revealed a stricture-like obstruction within the anterior urethra with proximal dilatation. For the purpose of relieving the obstruction, a cystoscopy with fulguration of valves in the bulbar urethra was performed. Postoperative MCU after 1 month revealed a residual crescentic valve in the bulbous urethra. Since the patient had already undergone repeated manipulation of the urethra, it was decided to only incise the valve to prevent stricture formation from aggressive attempts at resection. Relief of obstruction was confirmed on table by visualizing a good stream of urine. Two follow-up cystoscopies were performed at yearly intervals and residual flaps were resected. The urethral calibre was noted to have normalized. Follow-up ultrasound revealed that the hydronephrosis was decreasing. The renal function is normal.

The third patient presented at the age of 2 years with complaints of poor stream of urine. He was circumcised at the age of 1 year for a phimosis that was thought to be the cause of his poor stream. An examination revealed that he had meatal stenosis and hence, a meatomomy was performed. After the meatomomy, a check cystoscopy was performed. This revealed crescentic valves within the anterior urethra without any proximal dilatation. The bladder was normal. The valves were cystoscopically resected and adequacy confirmed on table by visualizing a good stream of urine. Following this resection, he has been asymptomatic and has a good stream of urine.

The fourth patient is a 9-year-old boy who complained of increased frequency of micturition and primary nocturnal enuresis. He also had dysuria without any manifest urinary infection. Urine cultures and renal function tests were normal. Ultrasound showed a thickened bladder and normal kidneys. MCU revealed a dilated posterior urethra with bilateral grade one VUR. Dimercaptosuccinic acid (DMSA) scan did not show any renal scars. Cystoscopy (Figure 3) revealed valves within the anterior urethra that were resected at the same time. The bladder was normal. Postoperative follow-up revealed relief from frequency and dysuria.

Results

Dilatation of the upper tract in the first patient decreased on subsequent IVU after 2 years. There was prompt uptake and excretion of contrast. On long-term follow-up, the patients recovered well with good stream of urine and normal renal function. Patients are continent and free of urinary infections.

Discussion

Anterior urethral valves are rare congenital anomalies. Although the cause is uncertain, various theories have been proposed.\textsuperscript{1–4} It could be the result of an unsuccessful attempt at urethral duplication\textsuperscript{1,2} or misalignment between the proximal and distal urethras\textsuperscript{3,4} or congenital cystic dilatation of periurethral glands\textsuperscript{1} with flap valve formation or excess developmental tissue remnant.\textsuperscript{3} They may or may not be associated with a proximal diverticulum.\textsuperscript{5} They can cause a spectrum of disease ranging from only urethral dilatation to bilateral hydronephrosis with renal impairment.\textsuperscript{6} There may be severe obstruction presenting with either severe bilateral hydronephrosis or bladder rupture with urinary ascites and azotemia in neonates and infants.\textsuperscript{2,7} A lesser degree of obstruction would present with symptoms related to micturition such as dribbling or enuresis. MCU with good views of the glanular urethra is the investigation of choice.\textsuperscript{3,7} Cystoscopic identification is possible and this is aided by suprapubic pressure over the filled bladder.\textsuperscript{5} The valves appear as circumferential cusps or membranes.\textsuperscript{2} The treatment options are transurethral resection, open urethrotomy and excision of the valves or excision of a segment of the urethra bearing the valves and end-to-end anastomosis.\textsuperscript{2} Transurethral valve ablation.
is the treatment of choice.\textsuperscript{2} We treated all patients with primary transurethral resection to improve drainage. This was followed by ultrasound studies for upper tract dilatation, and check cystoscopies to detect any residual urethral dilatation and bladder trabeculation. Two of our patients underwent resection of residual valvular flaps at subsequent check cystoscopies. Open excision and urethroplasty are indicated in the presence of severe urethral dilatation and deformity. The complications of open repair are urinary extravasation, stricture and urethrocutanous fistula, whereas cystoscopic ablation is associated with stricture and persistent urethral dilatation.\textsuperscript{8} One management algorithm\textsuperscript{9} recommends a primary transurethral fulguration for a normal calibre undilated urethra, vesicostomy for poorly emptying tracts and a one- or two-stage urethroplasty if there is an associated diverticulum. Unlike with posterior urethral valves, the eventual outcome is good. Long-term follow-up does not show any residual radiological dilatation and the incidence of chronic renal failure is less than 5%.\textsuperscript{9} In summary, although rare, anterior urethral valves must be considered in the differential diagnosis of infravesical obstruction in male children. They are detected on either MCU or cystoscopy. The treatment is always surgical and has to be tailored according to the dilatation of the proximal urethra. The long-term prognosis is good.

References