CASE REPORT

Anterior urethral valves

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Abstract

Objective: to discuss clinical signs, diagnostic tools and therapeutics of anterior urethral valves, an obstructive anomaly of the urinary system in males.

Description: signs of urinary tract obstruction were identified on pre-natal ultrasound in two male fetuses and the diagnosis of anterior urethral valves was made through post-natal evaluation. As an initial treatment, vesicostomy was performed in both patients. Later, the valves were fulgurated using an endoscopic procedure. During the follow-up period both patients presented normal renal function.

Comments: anterior urethral valves are a rare form of urethral anomaly that must be ruled out in boys with pre-natal ultrasound indicating infravesical obstruction. Vesicostomy used as an initial treatment rather than transurethral fulguration may prevent potential complications that can occur due to the small size of the neonatal urethra.


Introduction

Urethral valves are infravesical obstructive congenital anomalies that are more common in boys, and posterior urethral valve (PUV) is the most prevalent. The cases of anterior urethral valves (AUV) are still considered quite rare.1

The recognition and the original report on anterior urethral valves are attributed to D.I. Williams in 19582 and, since then, several cases have been described, both separately and in association with proximal urethral diverticula.3 In terms of distribution, these valves are more common in the bulbar urethra (40%), equally occurring both in the penile urethra and in the penoscrotal junction (30%). Some cases have been described in the navicular fossa, and mistaken for urethral meatal stenosis.4 Its occurrence has also been described in adolescents and adults,5,6 as well as in the VATER association.6

This report describes two cases of children with anterior urethral valve, emphasizing its clinical and therapeutic aspects.

Case description

Case 1

Pregnant woman submitted to prenatal ultrasonography in the 36th week of gestation, whose result suggested the presence of a fetus with PUV. Due to the worsening of bilateral hydronephrosis in a subsequent examination, a C-section was recommended.
The child was born with 3,240g, 47 cm and Apgar score of 9/10; there was a moderate amount of amniotic fluid with meconium (+++/4). The child presented dripping urination, with no urine jet and bulging at the base of the penis was observed during voiding. The value of serum creatinine on the third day of life was 1.0 mg/dL. Renal ultrasonography (US) at the time showed pronounced hydronephrosis on the right kidney, and a less intense one on the left kidney, with normal echogenicity of the parenchyma on this side, and a thick-walled bladder. Dilation of the urethra was also observed. The subsequent micturating cystourethrography (MCUG) confirmed the aspect of strained bladder (trabeculation of the bladder wall and pseudodiverticula) and showed the concomitant presence of vesicoureteral reflux on the right side, suggesting pyelourethral duplication on this side. Furthermore, the exam clearly showed a large urethral diverticulum, indicating an anterior urethral valve. (Figure 1)

After radiological examination, the infant developed septicemia, and urinary tract infection and pyonephrosis were diagnosed on the right side. The patient was submitted to exploratory lumbotomy and right upper pole nephrectomy with ureterectomy, followed by vesicostomy. The patient presented a good outcome in the postoperative period.

After 28 months a transurethral fulguration of the anterior urethral valve was carried out concomitantly with the closure of the vesicostomy; resection of the urethral diverticulum was not performed. The patient evolved without clinical intercurrent diseases, presenting a good urinary stream. The control micturating cystourethrography showed normalization of the lower urinary tract with spontaneous reduction of the urethral diverticulum (Figure 2), with discrete pyelocaliceal dilation at US. The serum levels of urea and creatinine were normal.

Case 2

The child was naturally delivered at 34 weeks of gestation, with an Apgar score of 9/10, weight of 2,550 g, length of 46.5 cm, and amniotic fluid with meconium (++/4). The patient had already been submitted to prenatal ultrasonographic examination in the 31st week of gestation, which clearly showed persistent bladder distension and bilateral hydronephrosis with clinical suspicion of PUV.

After birth, the child presented with dripping urination, with bulging of the penile urethra on urinary straining. The examinations made on the first day of life showed the following: urea = 20 mg/dL; creatinine = 0.7 mg/dL. The ultrasonography showed renal parenchyma with preserved thickness and echogenicity, with bilateral hydronephrosis. The bladder presented thick walls with dilation of the distal third of the urethra.

The cystourethrogram performed on the third day of life showed pronounced urethral dilation with diverticulum, suggesting an anterior urethral valve (Figure 3). On the fourth day of life, the child underwent cutaneous vesicostomy, with hospital discharge on the fourth postoperative day.

After 18 months, a transurethral fulguration of the AUV with simultaneous closure of the vesicostomy was performed. During the surgery, urethral stenosis distal to the valve was observed, thus requiring open urethrotomy. The patient presented good postoperative outcome, with normal urinary stream. Later on, reconstructive urethroplasty was carried out, without intercurrent diseases. The late postoperative evaluation with MCUG and US revealed normal radiological aspect of the bladder and urethra, and improvement of hydronephrosis, with normal renal function, according to the serum levels of urea and creatinine (urea = 20 mg/dL and creatinine = 0.6 mg/dL).

Discussion

The routine utilization of ultrasonography in prenatal evaluation has currently become the best-known method for intrauterine diagnosis of several fetal anomalies, especially those related to the urinary tract.7 On this examination, unilateral or bilateral fetal hydronephrosis, associated or not with oligohydramnios, but followed by persistent bladder distension, is highly suggestive of infravesical obstruction.
This hypothesis is reinforced when the simultaneous identification of urethral dilation occurs. In this situation, PUV is the most important anomaly to be ruled out in male patients, since its incidence is estimated at 1/30,000 to 1/5,000 boys. Anterior urethral valve should be considered for differential diagnosis. AUV and urethral diverticula are rare, with a frequency eight times lower than the frequency of posterior urethral valve.

Anterior urethral valve, whose etiology is not completely known, may represent an incomplete fusion of a segment of the urethral plate or, an incomplete focal development of the corpus spongiosum with bulging of the urethral mucosa due to ineffective support, which manifests itself as urethral diverticulum. Some authors also consider the possibility that it represents an attempt of urethral duplication in the first 12 to 14 weeks of intrauterine life.

In the postnatal period, cases of anterior urethral valve usually occur in the first week of life. The signs and symptoms of urinary obstruction are the most often reported ones; with decrease in the strength of the urinary stream or dripping urination in about 40% of the patients. Serious obstruction concomitantly with hydronephrosis and azotemia may occur in newborns and infants. Another common clinical observation in infravesical obstruction through anterior urethral valve is the bulging of the penile urethra caused by the filling and distension of the urethral diverticulum during patient’s urinary straining. In general, dripping occurs. With the obstruction, the concomitant occurrence of bladder distension is common, and the newborn’s bladder is easily palpable.

Parallel to the analysis of serum urea and creatinine levels, micturating cystourethrography (MCUG) is of paramount importance for the diagnostic confirmation of the location of infravesical obstruction. In this case, the whole urethra has to receive contrast medium for adequate visualization and location of urethral diverticulum associated with PUV. MUCG also allows the identification of vesicoureteral reflux, which may be present in 30% of these patients. Ultrasonography may suggest the diagnosis of AUV through the visualization of the urethral dilation and it is useful in the evaluation of the upper urinary tract, since it identifies hydronephrosis of the urethra and the characteristics of the renal parenchyma, especially the presence and intensity of renal dysplasia.

Due to the various forms of urinary tract involvement, Firlit et al. created a classification that contemplates the correlation of the radiologic findings of the anterior urethral valve with the clinical and laboratory data. This shows the evolutorial aspect of urinary tract involvement, from mild (pronounced hydronephrosis of the urethra) to severe (renal insufficiency) disorders. This influences the attention to be given to the patient, especially in a long-term follow-up.
Several methods for the treatment of AUV have been proposed, ranging from temporary diversion to primary resection of the urethral diverticulum and valve, in association with necessary procedures for the treatment of possible complications that secondarily affect the upper urinary tract. However, most authors agree that transurethral resection of the valve should be made when the urethra has a diameter that permits the passage of the resector without causing any trauma. Nowadays, the small diameter of pediatric cystoscopes allows valve resection in the neonatal period, but the procedure should be prescribed according to the general status presented by the patient.

In newborns with altered urinary tract drainage, especially in association with urinary infection, some form of temporary diversion must be considered. In our setting, we have preferably used Blockson cutaneous vesicostomy as temporary urinary diversion in children. This provides fast and effective decompression of the upper urinary tract and prevents the potential complications of resection in small-caliber urethras, such as stenosis or inadequate resection. In addition, it allows the potential reestablishment of renal function in severely affected children. It is interesting to notice the reduction of the urethral diverticulum, secondary to urinary diversion, without the need of open surgery for adjustment of the urethral lumen in the patients presented here. On the other hand, it is possible to use previous vesicostomy as an entry for the resector for the anterograde fulguration of the valve, avoiding the manipulation of the urethra.

In conclusion, we recall the importance of anterior urethral valve in children whose prenatal ultrasonography suggests infravesical obstruction, along with the hypothesis of posterior urethral valve. The suspicion of AUV is stronger when urethral dilation is observed during postnatal urinary straining. We also point out that the utilization of vesicostomy as a temporary urinary diversion is recommended in these cases.

References


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