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CASUISTRY

Late diagnosis of posterior urethral valves*



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KEYWORDS

Posterior urethral valves; Voiding dysfunction; Urethrocystoscopy; Electrofulguration

Abstract

Clinical problem: We diagnosed 8 patients with late-stage posterior urethral valves (PUV) between 1 and 14 years of age. Five patients complained of symptoms related to voiding dysfunction. The other 3 patients required urethrocystoscopy for other reasons (hypospadias fistulae, difficulty with catheterisation and high-grade vesicoureteral reflux [VUR]). A second review of the first 2 patients' medical history showed voiding dysfunction symptoms. All patients underwent preoperative ultrasonography: 3 patients had normal results and 5 had renal or vesical disorders. The diagnosis was reached through voiding cystourethrogram (VCUG), and 4 patients underwent urodynamic studies. The diagnosis was confirmed by urethrocystoscopy, performing valve electrofulguration. We performed urethrocystoscopy during the check-ups at 3–6 weeks and observed no stenosis. The symptoms disappeared for all patients after 20 months of follow-up. The patient with VUR was cured. The ultrasounds showed no progression of the renal involvement and showed improvement in the vesical involvement. The velocimetries during check-ups presented curves within normal ranges.

Discussion: Most children with PUV are diagnosed through ultrasound during the neonatal period. Some patients present PUV at later ages with diverse symptoms, which impedes its diagnosis. We should suspect PUV in male patients with symptoms of voiding dysfunction, either when they have normal or pathological results from ultrasounds or VCUG. We recommend performing urethrocystoscopy to rule out urethral obstruction.

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PALABRAS CLAVE

Válvulas de uretra posterior; Disfunción de vaciado; Uretrocistoscopia; Electrofulguración

Válvulas de uretra posterior de diagnóstico tardío

Resumen

Problema clínico: Diagnosticamos 8 pacientes de forma tardía de válvulas de uretra posterior (VUP) entre 1 y 14 años. Cinco pacientes consultaron por sintomatología relacionada con disfunción del vaciamiento vesical. Los otros 3 precisaron una uretrocistoscopia por otro motivo (fístula de hipospadias, dificultad de sondaje y RVU de alto grado), y al rehistoriar a los 2 primeros también presentaban sintomatología de disfunción de vaciado. Todos tenían ecografías preoperatorias: 3 fueron normales y 5 patológicas, con alteraciones a nivel renal o vesical.

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El diagnóstico se sospechó por cistouretrografía miccional seriada (CUMS) y 4 pacientes tenían estudios urodinámicos. El diagnóstico se confirmó por uretrocistoscopia, realizándose electrofulguración de las válvulas. Realizamos uretrocistoscopia de control a las 3-6 semanas sin observan ninguna estenosis. La sintomatología desapareció en el 100% de los pacientes tras 20 meses de seguimiento. El paciente con RVU se curó. Las ecografías no mostraron progresión de la afectación renal y presentaron mejoría de la afectación vesical. Las flujometrías de control mostraron curvas dentro de la normalidad.

Discusión: La mayoría de los niños con VUP se diagnostican ecográficamente en el periodo neonatal. Algunos pacientes manifiestan las VUP a edades más tardías con clínica diversa, lo que dificulta su diagnóstico. Debemos sospecharlas en pacientes varones con síntomas de disfunción de vaciado, tanto si tienen ecografías o cistouretrografía miccional seriada normales o patológicas y recomendamos realizar uretrocistoscopia para descartar obstrucción uretral. © 2015 AEU. Publicado por Elsevier España, S.L.U. Todos los derechos reservados.

Clinical problem

We present 8 patients diagnosed with late posterior urethral valves (PUV) at our center over the last 4 years (2009–2013). The age at which they were diagnosed with PUV was between 1 and 14 (median 6 years and a half). The reason why patients consulted was very diverse (Table 1), 5 of them presenting symptoms of voiding dysfunction. Patient 6 was controlled in outpatient clinics for high-grade vesicoureteral reflux, in which it was decided to perform endoscopic treatment of reflux. Patients 7 and 8 consulted for other reasons that required performing an exploratory urethrocystoscopy, and by rehistoring parents, children also had symptoms of dysfunctional emptying.

With regard to preoperative imaging tests (Table 1), all patients had ultrasound before surgery. In 3 patients they were normal and in the other 5 they were pathological, finding changes at the kidney and bladder level (Fig. 1). In patients with renal parenchymal involvement, renal scan was performed showing significant decrease in differential renal function. Serial voiding cystourethrography (SVCUG) was performed in 6 out of 8 patients. In 2 patients, a significant dilation of the posterior urethra was observed (Fig. 2), but in all studies we found pathological findings. In 4 patients with symptoms of voiding dysfunction, urodynamic studies were performed.

In all patients, the presence of PUV was confirmed by urethrocystoscopy, which were electrofulgurated (Fig. 3). To control the caliber of the urethra, a new urethrocystoscopy between 3 and 6 weeks (median 4 weeks) was performed after the first surgical procedure. In all patients, a urethra of good caliber was observed.

After more than a year of follow-up (mean 20 months), 100% of symptomatic patients have presented symptom disappearance. The child who had difficulty for probing initially presented persistent leaks, but to a lesser degree, and they have been controlled with a low dose of anticholinergics. The patient with high-grade VUR (patient 6) remained without infections and absence of reflux by SVCUG was found.

In all patients, ultrasound scans were performed, and in none of the children with renal involvement was there progression thereof. In patients with bladder wall thickening it decreased, although they do not yet present images compatible with normality. Flowmetries were performed and all presented curves within normality.

Discussion

PUVs are the most common cause of congenital obstruction of the urethra. 1,2 Currently, prenatal ultrasound diagnoses the vast majority of cases of PUV, 1-3 but sometimes, when the obstruction is mild, the ultrasound signs are more subtle and may go unnoticed. 1,2 This group of patients is usually diagnosed at later ages, because symptoms do not appear until the bladder dysfunction secondary to prolonged obstruction of the urethra is observed. 4,5

In our review, the median age at which the patients were diagnosed was 5, slightly lower than in other articles. 1,2,5 In symptomatic patients, the most frequent symptom was daytime incontinence, as in other series. 2,5

Ultrasound is basic in the initial study given the suspicion of lower urinary tract obstruction due to its high sensitivity and specificity. The alterations found were at 2 levels: alterations of the upper urinary tract (renal parenchymal involvement or dilation of the urinary tract) due to retrograde increase of pressure, and bladder disorders (increased thickness greater than 5 mm and of the trabeculation of the bladder wall), because of increasing collagen and detrusor muscle fibers, secondary to prolonged obstruction. We recommend applying for ultrasound before the beginning of the conservative treatment of voiding dysfunction, 1,5 and not only if we have not achieved an improvement in symptoms. In patients with ultrasound renal impairment, renal function should be documented by scintigraphy (DMSA) to know the degree of renal impairment from which we start and to be able to compare the evolution during the follow-up.

The next test to apply for is the SVCUG.^{1,5} The most typical finding is the dilation of the posterior urethra,⁷ which in our study was found only in 2 children. Other common findings that we found in our patients are, like those published in the literature,⁸ vesicoureteral reflux, trabeculated bladder, bladder diverticula, and urethral obstruction, or at the level of the sphincter. SVCUG remains the gold standard test for evaluating the urethra, it also provides data on the bladder and removes the operating factor dependent on ultrasound testing.⁹

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