CLINICAL CASE

Nonneurogenic neurogenic bladder (Hinman syndrome): two different treatments for the same problem

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Abstract Nonneurogenic neurogenic bladder, also known as Hinman syndrome, is a rare entity characterized by nonneurogenic urinary bladder dysfunction. It is a diagnosis of exclusion and is assigned to those children that present with clinical, radiologic, and urodynamic characteristics of a neurogenic bladder with external sphincter dyssynergia, but with no evidence of any neurologic alteration. In 1973, Hinman and Baumann were the first to report a case series of 14 children with nonneurogenic neurogenic bladder and treated these patients with bladder re-education and hypnosis. Bladder re-training and medical treatment have been recognized as effective management. However, when there is established damage to the upper urinary system, or risk thereof, surgery is preferred over conservative treatment. Due to the low incidence of the disease, there is still no consensus as to the most adequate treatment or management, making it essential to describe every case possible in order to gain the necessary knowledge for providing the best treatment for patients with this syndrome.

We present the cases of two boys with Hinman syndrome, 10 and 12 years old, for whom conservative treatment was not sufficient given that they already presented with upper urinary system damage. They underwent successful surgical treatment that was individualized for each case, resulting in 2 different solutions for the same problem.

Vejiga neurogénica no neurogénica (síndrome de Hinman). Dos tratamientos diferentes a un mismo problema

Resumen El síndrome de vejiga neurogénica no neurogénica, también conocido como síndrome de Hinman, es una entidad rara caracterizada por disfunción vesical miccional no neurogénica. Su diagnóstico es hecho por exclusión y se asigna a aquellos niños que tienen las características clínicas, radiológicas y urodinámicas de una vejiga neurogénica con disinyergería del
Nonneurogenic neurogenic bladder syndrome, also known as Hinman syndrome, is a rare entity characterized by nonneurogenic urinary bladder dysfunction. It generally presents in children in early infancy or in preschool age. Its diagnosis is made by exclusion and is assigned to those children that have clinical, radiologic, and urodynamic characteristics of a neurogenic bladder with external sphincter dyssynergia, but with no evidence of a neurologic alteration. Hinman syndrome generally causes recurrent urinary infections, urinary incontinence, vesicoureteral reflux, hydronephrosis, and acute and chronic renal failure. In 1973, Hinman and Baumann were the first to report a case series of 14 patients with nonneurogenic neurogenic bladder syndrome that they treated with bladder reeducation and hypnosis. Some authors have established the effectiveness of bladder retraining and medical treatment for its management, while others have employed different treatments such as Botox application in the external sphincter with good results. However, when there is already damage to the upper urinary tract, or risk thereof, more aggressive surgical treatment is preferred over conservative treatment in order to prevent chronic renal damage. Due to the low incidence of the disease, there is still no consensus as to the most adequate treatment or management. Surgical treatment is rare given that it is a last option.

Case presentations

Case 1
A 12-year-old boy had a past history of recurrent urinary infections from 7 months of age. He presented with continuous urinary incontinence and was previously treated with clean intermittent catheterization that apparently was not adhered to correctly, with no improvement. Laboratory test results showed serum creatinine of 0.7 mg/dl with urea of 49 mg/dl, and urinalysis reported leukocyturia, abundant bacteria, positive nitrites, and proteinuria of 300 mg/dl. Urine culture was positive for *Escherichia coli* (E. Coli) > 100,000 CFU, and antibiotic therapy was indicated. Cystography revealed a thickened bladder, with grade V right vesicoureteral reflux, and abundant residual urine (fig. 1). An ultrasound study identified severe right renal ectasia and moderate left ectasia (fig. 2). Magnetic resonance imaging of the spine was normal and evaluation by the neurosurgery department corroborated the lack of any neurologic alteration and cystoscopy established the absence of urethral valves. Bladder enlargement with the sigmoid colon and the creation of a Mitrofanoff stoma were performed, with clean intermittent catheter management through the stoma. The patient showed noticeable clinical...
improvement at the follow-up, with no more infections, reduced renal ectasia (fig. 3), disappearance of the vesicoureteral reflux (fig. 4), and complete continence between catheterizations.

Case 2
A 10-year-old boy began having urinary tract infections from the age of 3 years. He had been given numerous antibiotic treatments, and had a previous vesicostomy managed by a third party. Creatinine and urea were reported at 1.4 mg/dL and 83 mg/dl and the patient presented with bilateral renal ectasia (fig. 5) and recurrent urinary infections for which he was referred to our center. We began management with antibiotic therapy and then carried out a cystography study that showed grade V left vesicoureteral reflux (fig. 6). A kidney scintigram revealed reduced total renal function with greater involvement of the left kidney (fig. 7). A magnetic resonance imaging study of the spine was normal and no neurologic problem was found in the neurosurgical evaluation. A urodynamic study reported low intravesical pressure with no uninhibited contractions and cystoscopy revealed a permeable urethra. The decision was made to close the vesicostomy and to perform a left ureteral reimplantation and ureter remodeling. A Mitrofanoff stoma was created for intermittent catheterization. At follow-up, there was a significant reduction of dilation in the upper urinary tract (fig. 8) and improvement in the creatinine levels and bilateral renal function. The patient carried out intermittent catheterization with no problem.
Hinman syndrome is a pathology that can present in early infancy or in older children around 12 years of age. In some patients it is associated with psychologic factors, acquired personality disorders, or both. This syndrome is produced by an involuntary obstruction of the bladder exit at the external sphincter level with no evidence of a neurologic alteration. The detrusor muscle maintains a long period of hyperactivity in relation to the infravesical obstruction, which ends up producing detrusor decompensation. This, in turn, results in upper urinary tract damage that can produce chronic renal failure at an early age.1-3

The majority of patients initially present with urinary incontinence, recurrent urinary infections, micturition difficulty, and the presence of residual urine as the most frequent symptoms. If early diagnosis is not established the detrusor muscle and upper urinary system can become damaged, showing radiographic data of a thickened bladder, vesicoureteral reflux, hydrenephrosis, and loss of the renal parenchyma.4,5

Conservative treatment is recommended in the first stages of the disease that includes bladder retraining, a strict bladder voiding schedule, psychologic support, anticholinergic medication, and clean intermittent catheterization, all of which are effective if strictly adhered to by both the patient and family members,4 which is not always the case with all patients.

Given the failure of conservative management, a series of new slightly more invasive treatments has been described, such as Botox application to the external urethral sphincter.5 Mokhless reported on the application of Botox in the external urethral sphincter of 10 boys with nonneurogenic neurogenic bladder with good results in the intermediate period with no complications and a mean follow-up period of 10 months.5

Some authors have recently described the use of transcutaneous electrical bladder stimulation for treating Hinman syndrome. Rashid Sajid et al. reported that Hinman syndrome is a condition in which the bladder empties infrequently due to a lack of coordination between the sympathetic and parasympathetic activity causing a contraction in the external urethral sphincter muscle that produces infravesical obstruction. They evaluated 24 patients with Hinman syndrome, all above the age of 12 years, and divided them into 2 groups. The first group was managed only with traditional treatment and the second with transcutaneous neurostimulation plus traditional treatment. Urinary symptoms were evaluated after 12 weeks of treatment and there was noticeable improvement in the patients that received neurostimulation. However, the upper urinary tract and renal function were not evaluated, the follow-up period was very short, and the urinary symptoms did not completely disappear.8

Even with promising results, neuromodulation and Botox application must have long-term evaluations with controlled randomized studies in order to demonstrate their efficacy in children presenting with nonneurogenic neurogenic bladder syndrome.

Upper urinary tract protection and the prevention of progression into renal disease are the principal aims of nonneurogenic neurogenic bladder syndrome treatment. When initial treatment with bladder reeducation, psychologic support, clean intermittent catheterization, or some of the new techniques such as Botox application or transcutaneous neurostimulation have failed and the upper urinary tract is at risk, more aggressive treatment needs to be undertaken to protect the upper urinary tract and renal function, so that progression into renal disease is prevented. It has also been suggested that when there is established renal damage, conservative treatment should be abandoned and the patient should be managed with surgical treatment that protects renal function and prevents later deterioration.1-3

At present there are no case series or studies analyzing which is the best surgical technique for these types of patients. In the cases reviewed, bladder enlargement with sigmoid colon plus a Mitrofanoff stoma with intermittent catheterization has been reported.9
We described 2 different techniques that were individualized according to the clinical characteristics of our 2 patients. In the first case, we performed bladder enlargement with the sigmoid colon plus a Mitrofanoff stoma. In the second case, given that the patient presented with a low-pressure bladder and vesicoureteral reflux, we only performed a Mitrofanoff stoma and ureteral reimplantation. Both procedures had good results at the one-year follow-up, achieving the goal of protecting the upper urinary tract in the 2 patients.

Conclusions

Hinman syndrome, or nonneurogenic neurogenic syndrome, is a urinary dysfunction with a lack of bladder-sphincter coordination. This incoordination produces damage to the bladder and upper urinary tract if it is not adequately diagnosed and treated. Conservative management is accepted as initial treatment, but when it does not function, surgical treatment is necessary in order to prevent chronic renal failure.

Conflict of interest

The authors declare that there is no conflict of interest.

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References