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Treatment of Micturitional Disorders in the Shy-Drager Syndrome With Oxybutynin Chloride and Self Intermittent Catheterization

P. Politi, S.D. Sandri, V. Marino, F. Fanciullacci, F. Catanzaro, and A. Zanollo

Department of Urology, Magenta Hospital, Milan, Italy

Patients with Shy-Drager syndrome complain of frequency, urgency, and urge incontinence associated with urinary retention. We report our experience with three cases. Urodynamic evaluation showed detrusor hyperreflexia with low amplitude detrusor contractions. Symptoms have been well controlled by means of oxybutynin chloride and intermittent self catheterization.

Key words: urge incontinence, urinary retention, detrusor hyperreflexia, anticholinergic treatment, intermittent self catheterization

INTRODUCTION

The association of orthostatic hypotension with other signs of neuronal degeneration of central nervous system is defined as Shy-Drager syndrome [Shy and Drager, 1960; Bannister and Oppenheimer, 1972; Rubenstein and Yahr, 1977].

This syndrome is classified as a neuronal degenerative disorder because at autopsy there is a loss of autonomic neurons in the brainstem with or without degeneration of the motor and extrapyramidal systems; clinically, there is a derangement of autonomic functions, manifested predominantly by orthostatic hypotension.

In some cases autonomic dysfunction is associated with Parkinsonian syndrome owing to striatonigral degeneration; in others it is asssociated with olivopontocerebellar degeneration. The disease is progressive and has a poor prognosis. Death is mainly caused by intercurrent infections of the respiratory tract at about 10 years from the onset. Few cases have a better prognosis [Schwarz, 1967; Kluysken et al, 1977; Allain et al, 1979; Singh and Fahn, 1980].

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Address reprint requests to P. Politi, Divisione di Urologia, Ospedale di Magenta, Via Donatore di Sangue, 50, 20013 Magenta (Milano), Italy.

MATERIALS AND METHODS

During the last 2 years we have seen three patients (1 male, 2 females) with Shy-Drager syndrome complaining of micturitional disturbances. These included frequency, nocturia, urgency, urge incontinence, difficulty in voiding, and urinary retention. All had recurrent urinary tract infections, with episodes of hyperpyrexia in two cases. The male patient suffered loss of libido and impotence, while the females complained of decreased libido and lack of sexual pleasure. All the patients have been evaluated with history, physical examination, laboratory examinations, excretory urogram, cystourethrogram, cystoscopy, and urodynamic evaluation including uroflowmetry, cystometry, electromyography, and urethral pressure profile.

Water cystometry was performed at a filling rate of 50 ml/min employing two 10-F urethral catheters: one for filling and one for pressure recording. Abdominal pressure was simultaneously recorded through a 12-F rectal catheter with the tip covered by a fingerstall.

Electromyography from the external urethral sphincter was recorded with a coaxial needle electrode introduced, in the male, through the perineum and directed toward the tip of the forefinger, positioned at the prostatic apex; in the female it was placed parallel to the urethra starting 1/2 cm lateral to the external meatus. Correct position was revealed by recording of motor unit potentials either audible at the loudspeaker or visible on the oscilloscope. Urethral pressure profile was measured with constant inflow catheter withdrawal. A 10-F catheter with side holes at 5 cm from the tip was used. A water inflow of 2 ml/min was maintained with a peristaltic pump during constant mechanical withdrawal at a rate of 2 mm/sec. The same rate was used for paper speed of the recorder in order to measure the functional urethral length.

RESULTS

Laboratory and radiographic examinations did not reveal any pathological changes of the upper urinary tract.

Cystourethrography showed no reflux and no urethral obstruction in the female patients. In the male patient, incomplete opening of the bladder neck was observed. Furthermore, endoscopic examination showed slight mucosal trabeculation in all the cases and ruled out prostatic enlargement in the male patient.

Cystometry showed in every case a low compliance hyperreflexic bladder with frequent low amplitude detrusor contractions and reduced bladder capacity with early appearance of voiding sensation (Fig. 1A). No patient had detrusor-sphincter dyssynergia. During micturition maximum detrusor pressure ranged from 10 to 14 cm. H_2O and abdominal efforts to void were seen in all the cases. Post micturitional residue was about 50% of bladder capacity (Table I). Cystometry, performed 20 minutes after the injection of emepronium bromide 50 mg intramuscularly (IM) showed increased bladder capacity with improvement of the compliance and disappearence of the bladder contractions (Table II, Fig. 1B).

Postmicturitional residue increased as well. Urethral pressure profile after the pharmacological test revealed no morphological test revealed no morphological and pressure differences from the preliminary evaluations, as shown in Tables I and II.

All patients were advised to perform intermittent self catheterization and were put under oxybutynin treatment (5 mg TID). Follow-up studies (from 6 to 12 months) showed increased bladder and urge incontinence (slight stress incontinence was present in female patients). All patients were able to perform intermittent self catheterization (4–6 times/day) and were free from U.T.I.

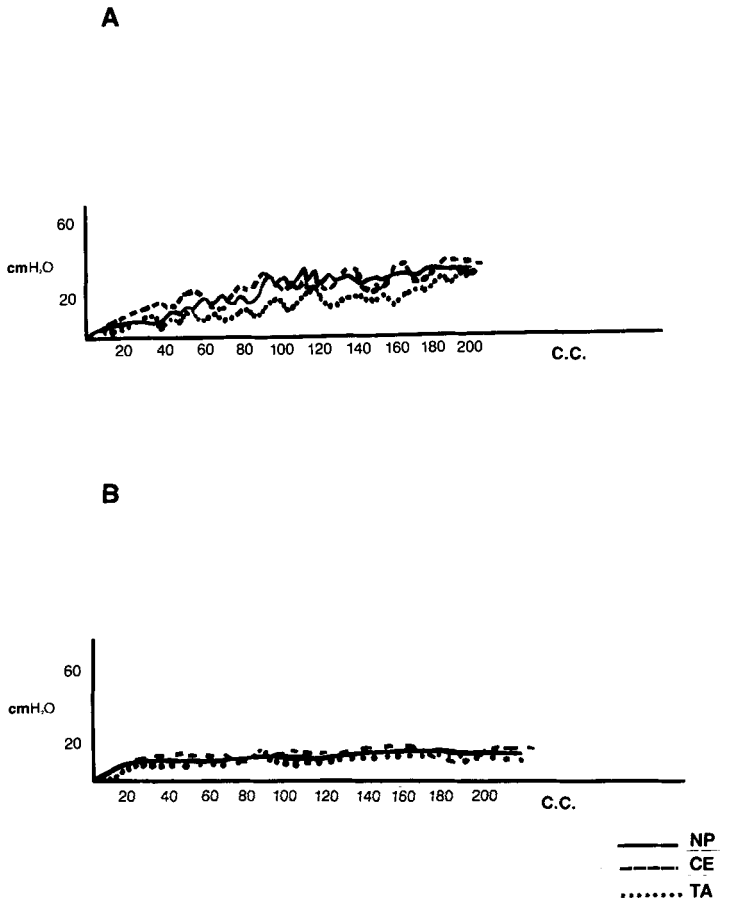

Fig. 1. A. subtracted detrusor pressure; B. after emepronium bromide.

TABLE I. Urodynamic Data

Patients	Age	Sex	Volume of involuntary detrusor contraction onset	Cystometric capacity	Bladder pressure at maximum cystometric capacity
N.P.	57 yr	M	110 ml	230 ml	40 cm H_2O
C.E.	52 yr	F	130 ml	180 ml	45 cm H_2O
T.A.	61 yr	F	110 ml	200 ml	38 cm H_2O

Volume voided	Postvoid residual	Maximum urethral closure pressure	Functional urethral length
100 ml	120 ml	55 cm H_2O	48 cm
100 ml	80 ml	30 cm H_2O	23 cm
80 ml	120 ml	35 cm H_2O	23 cm

TABLE II. Urodynamic Data After Emepronium Bromide (50 mg IM)

Patients	Age	Sex	Volume of involuntary detrusor contraction onset	Cystometric capacity	Bladder pressure at maximum cystometric capacity
N.P.	57 yr	M	190 ml	300 ml	14 cm H_2O
C.E.	52 yr	F	200 ml	280 ml	22 cm H_2O
T.A.	61 yr	F	220 ml	350 ml	18 cm H_2O

Volume voided	Postvoid residual	Maximum urethral closure pressure	Functional urethral length
50 ml	250 ml	50 cm H_2O	46 cm
20 ml	260 ml	30 cm H_2O	23 cm
20 ml	330 ml	34 cm H_2O	23 cm

Because of worsening of the neurological symptoms, the two females cases, after same time, needed help from family to perform intermittent catheterization.

DISCUSSION

Micturitional disorders (urge incontinence with urinary retention) in our patients were similar to those described by other authors [Lockart et al., 1981; Wulfsohn and Rubenstein, 1981; Vereecken et al, 1985]. We never found alterations of the perineal muscles reflexes as observed by Sakuta [1978], except for mild reduction of anal sphincter tone in only one case. Nevertheless, EMG findings were normal. Some polyphasic potentials were found, but they were of normal amplitude and duration.

No sensory changes were noted. Our urodynamic data agree with those of Lockart [1981] and Vereecken [1985], showing a pattern of detrusor hyperreflexia without sphincter dyssynergia but with poor detrusor contractility, explaining the urinary retention. Nevertheless, as with Wulfsohn [1981], we found good results with anticholinergic treatment and intermittent self catheterization. We preferred to increase pharmacologically the capacity of the hyperreflexic bladder because results from surgery have been disappointing [Wulfsohn, 1981].

Sexual and voiding disorders in these patients are most likely secondary to degeneration of the autonomic system typical of the disease.

The reduced detrusor contractility and the consequent lack of bladder neck opening during micturition might be explained by the recent finding of reduction of acetylcholinesterase-positive nerves with age in general and in particular with Shy-Drager syndrome [Gilpin et al, 1984]. This autonomic derangement may explain also the reduced urethral closure pressure in all the patients, with consequent slight stress incontinence in the female patients.

This problem should be investigated further in order to obtain a better understanding of the disease and to improve the clinical results.

REFERENCES

Allain H, Bentue Ferrer D, Brissel R, Mortsot D, Madigand M, Menaut P (1979): Syndrome de Shy-Drager: Approchè physiopathologique et phamacologique à propos d'un cas. Nouvelle Presse Med 8:2745.

Bannister, R, Oppenheimer DR (1972): Degenerative disease of the nervous system associated with autonomic failure. Brain 95:457.

Gilping SA, Kirby RA, Gosling J, Milroy EJG, Turner-Warwick KT (1984): Quantitative analysis of acetylcholinesterase plus nerves in the diagnosis of neurogenic bladder dysfunction. Proceedings 14th International Continence Society Meeting, Innsbruck, September 13–15, 1984, p 265.

Kluysken Y, Bossaert L, Sneock J, Martin JJ (1977): Idiopathica orthostatic hypotension and the Shy and Drager syndrome: Physiological studies in four cases; pathological report of one case. Acta Cardiol 32:317, 1977.

Lockhart JL. Webster GD, Sheremata W, Camuzzi FA, and Carrion HC (1981): Neurogenic bladder dysfunction in the Shy-Drager syndrome. J Urol 126:119.

Low PA, Thomas JE, Dyck PJ (1978): The splanchnic autonomic outflow in Shy-Drager syndrome and idiopathic orthostatic hypotension. Ann Neurol 4:511.

Rubenstein AE, Yahr MD (1977): Adult onset anatomic dysfunction coexistant with familial dysautonomia in a consanguineous family. Neurology 27:168.

Sakuta M, Nakanishi T, Tokokura Y (1978): Anal muscle electromyograms differ in amyotrophic lateral sclerosis and Shy-Drager syndrome. Neurology 28:1289.

Schwarz GA (1967): The orthostatic hypotension syndrome of Shy-Drager. A clinicopathologic report. Arch Neurol 16:123.

Shy GM, and Drager GA (1960): A neurological syndrome associated with orthostatic hypotension; a clinical-pathologic study. Arch Neurol 2:511.

Singh N, Fahn S (1980): Electrophysiologic studies in the Shy-Drager syndrome. Neurology 30:394.

Vereecken RL, Goovaerts J, Knockaert D, Joossens J, Sansen W (1985): Urological aspects of the Shy-Drager syndrome: A urodynamic and telemetric investigation. Eur Urol 11:65–68.

Wulfsohn MA, Rubenstein A (1981): The management of Shy-Drager syndrome with propantheline and intermittent self-catheterization: A case report. J Urol 126:122.

EDITORIAL COMMENT

In our experience with nine patients, Shy-Drager syndrome was most often associated with an open vesical neck, suggestive of a sympathetic lesion. In addition, most patients also had detrusor hypertonia with autonomous waves suggestive of parasympathetic decentralization. Only one-third had demonstrable detrusor hyperreflexia. Moreover, neurologically, our cases presented us with an extrapyramidal clinical picture.

In the present series, the authors noted that their female patients had stress urinary incontinence and decreased urethral closure pressure despite an effective oxybutynin chloride treatment. It would be interesting to know if a cystogram was performed to verify whether the bladder neck was not indeed open, verifying a type III incontinence.

The cystometry (Fig. 1A) was interpreted by the authors as detrusor hyperreflexia with frequent and low-magnitude contractions. They might, however, represent autonmous waves suggestive of a lower, rather than upper, motor neuron lesion.

Y. Berger, MD

REPLY BY AUTHORS

When low-amplitude detrusor contractions are present in a low-compliance bladder, it is always difficult to distinguish between autonomous waves and hyperreflexic contractions. A parasympathetic drug is effective in reducing both as recently shown [Sandri, 1985]. We described our cystometric curve as hyperreflexic because the detrusor contractions were of amplitude greater than 15 cm H_2O, following the Standardization Committee of the I.C.S.

Nevertheless, as written in this paper, the anatomic lesion of the disease might explain the appearance of both the urodynamic picture: hyperreflexia because of damage and areflexia with autonomous waves because of reduction of acetylcholinesterase positive nerves in the bladder. Obviously a mixed picture (detrusor hyperreflexia with low-amplitude contractions) may also occur.

The review of the cystograms confirmed that the bladder neck was closed in resting position in all our patients.

The urinary stress incontinence might be explained by reduced urethral closure pressure and increased bladder pressure during filling. The improvement after oxybutynin treatment might be attributed to the increase of bladder compliance.

REFERENCES

Sandri SD (1985): Pharmacologic tests in low compliance areflexic bladder. London: I.C.S. Annual Meeting, September 1985.