Isolated urinary retention in young women, or Fowler's syndrome

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Urinary retention in women is much less common than in men, with a recorded incidence of 7 cases per 100,000, per year [1]. Structural causes, such as fecal impaction, uterine fibroids, and pregnancy can usually be excluded by a careful history and examination. Similarly, retention occurring secondary to the anticholinergic side effects of medication can readily be identified. When retention occurs as part of a neurological disorder, this is usually quite apparent from the history and examination, and with the current availability of magnetic resonance imaging scanning, structural neurological lesions can now be readily excluded.

Occasionally, clinicians are presented with a female with urinary retention in which no cause is apparent. Imaging studies of the cord and cauda equina show normal results and magnetic resonance imaging shows no abnormality to suggest a diagnosis of multiple sclerosis. Furthermore, the patient shows no evidence of other autonomic dysfunction. In the past, the physician faced with such a patient concluded that the patient's retention had a psychologic basis. Indeed, the majority of the urological literature on urinary retention in women refers to “psychogenic urinary retention” [2–9]. This view was challenged in the mid-1980s when a consistent abnormality in the urethral sphincter was found by electromyographic (EMG) testing in a group of women with otherwise unexplained retention [10]. There were similarities in the way many of these cases had presented: the women were young, the retention was painless, and apart from a seemingly high incidence of polycystic ovaries, the women were well. The association of painless urinary retention occurring as an isolated disorder in a young woman, polycystic ovaries, and abnormal urethral sphincter EMG was described in 1988 [11] and is now sometimes referred to as Fowler’s syndrome.

The abnormal activity found on EMG sampling from the striated urethral sphincter using a concentric needle electrode comprises two types: complex repetitive discharges and decelerating bursts. When heard over the audio output of the EMG machine, this latter activity has a distinctive quality that has been said to be reminiscent of whales’ song [12]. When complex repetitive discharge activity is analyzed by single-fiber EMG, the jitter between component potentials is so low that transmission of the nerve impulse can only be directly from muscle cell to muscle cell, so-called “ephaptic transmission” [13]. It is known that the striated urethral sphincter is hormonally sensitive as it undergoes changes perimenopausally, for example. The hypothesis is that the hormonal imbalance that many of these women have results in instability of the cell membranes of the muscle cells of the striated urethral sphincter, allowing the nerve impulses to follow a disordered, circuitous route. This prevents sphincter relaxation, which, under physiologic circumstances, is the first stage in the initiation of micturition.

In cats, increased afferent activity from the urethral sphincter causes inhibition of detrusor activity [14], which is thought to be the physiologic basis of the “guarding reflex,” whereby voluntary contraction of the sphincter results in a transient reduction of sensations of urgency and suppression of detrusor activity. It is hypothesized that it is upregulation of the guarding reflex as a result of persistent, involuntary contraction of the urethral sphincter, that accounts for retention in this group of women. Suppression of the sensation of the bladder filling fits well with the observation that the retention in these patients is, for the most part, painless.

There is other evidence that the urethral sphincter is abnormal in these patients. When EMGs of the urethral...
sphincter were performed, it was observed that, on palpation, the urethral sphincter on the anterior vaginal wall in many of these patients was enlarged. Estimating the volume of the sphincter in a group of these patients by transrectal ultrasound demonstrated sphincter enlargement [15], and recently, using transvaginal ultrasonography, this observation has been confirmed, together with a demonstrated increase in urethral pressure [16].

In order to define the natural history of this disorder, a detailed questionnaire was sent to the 216 women who were identified as having experienced urinary retention and who were found to have, by EMG testing, the abnormal urethral sphincters [17]. Replies were obtained from 112; 91 were suitable for analysis. The majority of the women had begun to experience urinary retention in their 20s and 30s (mean age 27 y). Most (82%) described abnormal voiding behavior before the onset of urinary retention, and usually this was in the form of experiencing prolonged periods between voids. It was not uncommon for the women to report passing urine only two or three times in a 24-hour period. In 65% there seemed to be a specific event that triggered the retention, whereas in the remainder the retention was spontaneous. The most common triggering event, occurring in 30% of all patients' retention, was a gynecological operation while under general anesthetic. However, 13% of all patients reported their retention to have been brought on by various other operations, including surgery on the knee, ear, or jaw. Childbirth was seen as the triggering event in 15% of all patients.

Fifty-eight percent had undergone a pelvic ultrasound test, and 58% of these women had been told that they had polycystic ovaries; an additional 24% had been found to have “cysts on the ovaries.” Most (58%) of the women had not experienced any recovery of bladder function and were managing their problem for the most part by intermittent self-catheterization. Interestingly, the majority of them (69%) reported difficulty in passing catheters. This reflected our observation that many of these women experience a painful “gripping” sensation when trying to remove the catheter after self-catheterization, presumably because of the high sphincter tone that women with abnormal EMG findings have. In some women this was so troublesome that they had resorted to a suprapubic catheterization.

An important aspect of the questionnaire dealt with the treatments that the patients had encountered. The patients had been referred from a wide variety of sources, and a correspondingly diverse range of treatments had been tried, including medications such as α-blockers and urethral dilatations using a general anesthetic. Local injections of botulinum toxin into the urethral sphincter had not been successful [18]. The only consistently successful treatment that the patients had experienced was the relatively recently introduced therapy of sacral neuromodulation. This can be used for a variety of voiding disorders [19–22] and involves the application of trains of electrical stimuli delivered either by a permanently implanted device or by a temporary wire passed percutaneously through the 3rd sacral foramen. This has been used effectively in our department in this group of women with a success rate approaching 70% [23]. Many of the patients experience dramatic results with this therapy; not infrequently a woman who had not passed urine urethraically for many years will find that approximately 12 hours after the onset of stimulation, the urge to void returns. Spontaneous voiding by detrusor contraction follows shortly after this.

The same EMG abnormality is sometimes found in women with obstructed voiding and it seems reasonable to propose that, although the underlying pathophysiology in the sphincter may be the same in women with retention and obstructed voiding both, it is the secondary effect the abnormal sphincter activity has on the detrusor that determines the clinical picture. The concept that the EMG abnormality is due to a relatively commonly occurring, hormonally determined channelopathy affecting the striated sphincter muscle is currently being researched. Studies of populations of women undergoing sphincter EMG show that this type of activity is not uncommon (8–10%) [24–25], but its expression as complete urinary retention is relatively rare. This highlights the need to examine the sphincter with other techniques that may establish the extent of its impaired relaxation.

References