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Gynaecological pathology in women with Fowler's syndrome

Roopali Karmarkar [a], Bahareh Abtahi [b], Mohammed Saber-Khalaf [b], Gwen Gonzales [b], Sohier Elneil [b,*]

[a] Department of Urogynaecology, Imperial College Healthcare NHS Trust, St Mary's Hospital, Praed Street, London W2 1NY, United Kingdom
[b] Department of Uro-Neurology, National Hospital of Neurology and Neurosurgery, London WC1B 3BG, United Kingdom

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ABSTRACT

Objectives: To assess the prevalence of gynaecological pathologies in women with Fowler's syndrome (FS) which is characterised by chronic urinary retention (CUR) secondary to failure of urethral sphincter to relax and allow normal voiding.

Study design: This was a case control study conducted at a tertiary referral centre specialised in managing women with FS. There were 41 patients with FS in the study group with CUR without mechanical obstruction of the urethra or neurological problem. All patients had raised maximum urethral closure pressure on urethral pressure profile, high urethral sphincter volume on ultrasound and complex repetitive discharges on eletromyography of the urethral sphincter. Normal voiding was established in these women after treatment with sacral neuromodulation. Fifty women without voiding dysfunction acted as control group. Data was obtained by using standard questionnaire for both the groups. Information was collected regarding gynaecological pathologies such as endometriosis, polycystic ovarian syndrome, menstrual abnormalities, ovarian cysts and subfertility and also regarding previous pregnancies. Analysis was performed using SPSS software from IBM Corporation.

Results: At least one gynaecological pathology was present in 33 (80%) patients with FS compared to 16 (32%) women from control group ($P < 0.001$). This included a higher incidence of endometriosis (29% versus 6%, $P = 0.003$), PCOS (24% versus 8%, $P = 0.041$) and subfertility (34% versus 8%, $P = 0.003$). The incidence of menstrual abnormalities and ovarian cysts was similar in both groups ($P > 0.05$).

Conclusion: Subfertility was more prevalent in women with FS in our study. Though the prevalence of different pathologies was higher in the FS group compared to the control group, it remained similar to that found in the normal female population in the published literature. Thus, it is not possible to state whether FS is caused by a hormonally based disorder. It took patients many years before they could get right diagnosis and treatment for FS. Early investigation of chronic urinary retention and referral to specialists for appropriate treatment in this small group of women can result in better health and improvement in their quality of life.

Introduction

Chronic urinary retention (CUR) is defined as a 'non-painful bladder, where there is a chronic high post-void residual' [1]. CUR in women may be due to one of two reasons: bladder outlet obstruction or detrusor under-activity. Mechanical obstruction of the urethra can be caused by leiomyomas, vaginal wall cysts, urethral diverticulae, urethral cysts or chronic constipation. Other known causes of CUR include neurological conditions such as multiple sclerosis, spinal injuries, spina bifida, cauda equina syndrome and tethered cord. Historically, in the absence of any demonstrable cause, women who were unable to void were thought to have a psychological disorder until Fowler's syndrome (FS) was described [2].

Fowler's syndrome (FS) is a condition seen in young women typified by painless chronic urinary retention (CUR) secondary to failure of urethral sphincter to relax and allow normal voiding. The diagnosis is made by electromyographic findings on the external urethral sphincter characterised by complex repetitive discharges (CRDS) and decelerating activity [3]. The primary urodynamic finding in patients with FS is reduced bladder sensation, large capacity bladder and detrusor underactivity. Fowler's syndrome

* Corresponding author at: Department of Uro-Neurology, National Hospital of Neurology and Neurosurgery, Queen Square, London WC1N 3BG, United Kingdom. Tel.: +44 02034472516.
E-mail address: sohier.elneil@uclh.nhs.uk (S. Elneil).

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typically occurs in post-menarche women in their second or third decade. Sacral neuromodulation is the only known treatment that can restore voiding in patients with Fowler's syndrome [4,5].

There are a number of associations with FS, published in the literature. Most notably, polycystic ovarian syndrome (PCOS) was first defined as possibly having an important role in women with FS. It was believed to occur in 64% of cases [6]. This relative association with PCOS suggested the possibility that a hormonally mediated response, like progesterone deficiency (a cell membrane stabiliser), may have been the trigger for poor external urethral sphincter (EUS) relaxation [6–8]. However, PCOS is a frequent ultrasonographic finding in healthy reproductive aged women which makes this association tenuous. The link with other gynaecological conditions with a hormonal basis, such as endometriosis, ovarian cysts and pregnancy, has not been completely studied. Thus the objective of this study was to look at whether there was an association between gynaecological pathologies and Fowler's syndrome which may form the pathophysiological basis for abnormal sphincter activity.

In our experience, most of these women suffer physically and psychologically for many years, and even decades, before they reach a diagnosis of Fowler's syndrome (FS) [6,9]. Thus, it is important to understand the relationship between this rare bladder condition and gynaecological conditions, as it may help physicians reach an earlier diagnosis and give patients quicker access to definitive care.

Materials and methods

This is a case control age-matched study to explore the different characteristics of female patients diagnosed with FS at a tertiary centre specialised in management of FS and compare them with normal women without urinary retention or voiding difficulty.

The departmental database of patients from 1995 till 2012 was studied and 66 patients were found to be suitable for the study. These patients were diagnosed with CUR. Urethral compression by a pelvic mass was ruled out by pelvic ultrasound examination. Complete neurological assessment was performed to exclude neurological causes of CUR. Investigations were then performed on these patients to diagnose Fowler's syndrome. All patients had raised maximum urethral closure pressure on urethral pressure profile, high urethral sphincter volume on ultrasound and CRDS on eletromyography of the urethral sphincter. All 62 patients underwent sacral neuromodulation and had successfully regained normal bladder function. For the purpose of this study, all patients in this group were given a detailed questionnaire and data was collected regarding presence of gynaecological pathologies such as endometriosis, PCOS, menorrhagia, ovarian cysts and subfertility and any previous pregnancies.

Controls for this study were provided by 50 women attending hospital for non-medical reasons such as healthcare professionals and patients' relatives and without any history of urinary retention or voiding difficulty, but with other comparable parameters. They filled the same questionnaire as the study patients with FS.

Ethical approval was not required as this was an analysis of ongoing clinical work for patients with FS and the control data was obtained by voluntary survey of women.

Statistical analysis was performed using SPSS software (IBM SPSS Statistics 22, IBM Corporation). For quantitative values, results have been expressed in the form of mean ± standard deviation and range. In qualitative data we used number and percentage. Fisher's exact test was performed to analyse the association of various gynaecological pathologies with the FS. P value <0.05 was considered statistically significant.

Results

Forty-one out of the 66 women with FS responded to our questionnaire. In the study group of 41 women, 40 were Caucasian and one was of African origin. The control group included 49 (98%) Caucasians and 1 (2%) of African origin. The demographics of the two groups are as shown in Table 1. For 17 patients with FS, it took more than 2 years to access definitive care, whereas in 8 patients it took almost 10 years before they were seen and treated. Two patients spent half their life without a diagnosis. The average interval between treatment of FS with SNM and data collection was 4.4 years for the study group.

At least one gynaecological disorder was pre-existent in 33 (80%) patients before the diagnosis of FS. The average time between the diagnosis of a gynaecological condition and subsequent onset of voiding dysfunction was 8.3 years. Eighteen (44%) patients felt that their voiding difficulty was influenced by the severity of their gynaecological condition(s). Three women had worsening of symptoms during their menses. In the control group, only 13 women (26%) had one or more gynaecological disorders.

The prevalence of various gynaecological pathologies observed in the study was as shown in Table 2. The findings showed a statistically significant relationship between the diagnosis of FS and endometriosis ($P = 0.003$), and FS and PCOS ($P = 0.003$). However, no relationship was found between FS and ovarian cyst ($P = 0.403$) and FS and menorrhagia ($P = 0.290$).

With regards to fertility and conception, 14 (34%) patients suffered from either primary or secondary subfertility from various causes and needed treatment to conceive. With regards to pregnancy, 18 (44%) patients did have one or more pregnancies before the diagnosis and treatment of FS in the study group; 9 (22%) of whom needed medical help to conceive. During pregnancy, 5 (12%) patients had persistent voiding difficulty. Voiding dysfunction was experienced by 10 (24%) patients after delivery. In the control group, 4 (8%) women suffered from subfertility and needed treatment to conceive. Subfertility was statistically higher in the FS compared to the control group ($P = 0.003$) (Table 2).

Comments

This study clearly showed that it took patients many years before they could get the right diagnosis and treatment for Fowler's syndrome. In our experience, these patients undergo various investigations during these years and undergo different surgical procedures such as urethral dilatation, urethrotomy, hysterectomy, myomectomy and treatment to endometriosis without relief from urinary retention. It may be possible that there is lack of awareness of FS amongst the medical community and this is clearly causing delay in referral to specialists in the field.

Over 80% of patients with FS had one or more gynaecological disorder. Statistical evaluation by Fisher's exact test showed that patients with FS had a comparatively higher prevalence of various gynaecological conditions than women in the control group ($P < 0.05$). It is difficult to say, whether or not, this was because

Table 1
Patient demographics.

Parameter	Patients with Fowlers syndrome	Control group
Age	32 ± 10 years	33 ± 10 years
Ethnicity		
Caucasian	40 (98%)	49 (98%)
African	1 (2%)	1 (2%)