FOWLER'S SYNDROME UK

A Guide to Fowler's syndrome

What is Fowler's syndrome?

Fowler's syndrome is a condition in which women have problems passing urine. This is due to a difficulty in relaxing the urinary sphincter, which is the muscle controlling the flow of urine through the water pipe. Fowler's syndrome is one of the causes of chronic idiopathic urinary retention (CIUR).

The condition causes full or partial urinary retention and solely affects women*. The peak age of onset is between the ages of 15 and 30. It was first described by a team led by Professor Clare Fowler, in 1985.

* Women and those appearing female at birth (AFAB)

Fowler's syndrome and CIUR

Some women with long-standing or recurrent urinary retention may not have had the opportunity to have the tests to confirm Fowler's syndrome.

In this case the term Chronic (more than three months) Idiopathic (cause unclear) Urinary Retention, or CIUR, is often used. For these women most of the information in this leaflet and resources from FSUK will still apply.

More information:
www.fowlerssyndrome.co.uk
@fowlerssyndrome on Instagram and Facebook

About FSUK

Fowler's Syndrome UK (FSUK) is a national charity led by patients and top clinicians determined to raise awareness and to promote education and research into the debilitating and often under-diagnosed condition, Fowler's syndrome.

This guide is for anyone at the beginning of their diagnosis journey, receiving or exploring treatment options, or concerned about what could be causing their urine retention.



The scientific definition

A condition of chronic urinary retention affecting women, in the absence of urological, gynaecological and structural neurological causes, and characterised by idiopathic dysfunction of the external urinary sphincter demonstrated by at least one of the following:

- Elevated urethral pressure profile
- Abnormal findings in urethral sphincter electromyography including decelerating bursts
- Evidence for bladder outflow obstruction on video urodynamics testing
- Increased sphincteric volume on imaging



Causes

Normally a ring of bladder muscle (the urethral sphincter) keeps the water pipe (urethra) closed. In Fowler's syndrome this muscle goes into spasm, preventing urine flow out of the bladder. This reduces bladder activity, which in turn leads to reduced bladder sensation and reduced bladder muscle squeezing. What causes this sphincter to spasm is unknown.

Urinary retention in Fowler's syndrome can sometimes be triggered by surgical procedure, childbirth, opiate exposure or a sudden medical condition, such as an infection or illness.

Co-existing conditions

A significant proportion of patients have polycystic ovarian syndrome, endometriosis, or other gynaecological conditions.

There are also associations with Bladder Pain syndrome, Irritable Bowel syndrome, Functional Neurological disorder, Fibromyalgia, Ehlers Danlos syndrome, and other chronic pain syndromes or mental health difficulties such as anxiety and depression.

Demographics

The typical woman presents in her 20s-30s and may not pass urine at all, or may infrequently pass urine with an intermittent stream.

Fowler's syndrome has an incidence rate of 0.2 cases per 100,000 per year (Szymanski, 2021), suggesting that over 3000 women in the UK alone are living with the condition.

Due to the rarity of the condition, very little research has been carried out on diagnosing and treating the condition.

Symptoms

Many of the symptoms of Fowler's syndrome are caused by an inability to empty the urine that is stored in the bladder.

In a 2022 survey conducted by Fowler's Syndrome UK of those reporting to have the condition, the most common signs and symptoms in people with Fowler's syndrome are:



Inability to urinate

Fowler's syndrome may present itself as either full or partial inability to urinate or as frequent urination with an inability to fully void.

This means some women may not be able to pass all or part of the urine contained in the bladder, while others may have the sensation of needing to urinate more frequently than normal.

Some people need to strain or push to urinate and it may take longer to pass urine. Often people with Fowler's will hold larger amounts of urine in their bladders

Infection

Frequent urinary infections may be a problem for women suffering from Fowler's syndrome due to the bladder not emptying properly.

Women with Fowler's syndrome are at an increased risk of infection and sepsis. They are also at risk of developing antibiotic resistance due to the frequency with which they need antibiotics.

Pain

While the condition itself is painless, it can result in urinary infections and/or a need for a catheter, both of which can cause pain. Stomach, bladder and pelvic pain are often reported in Fowler's syndrome.

Pain may increase with specific movements and activities. Sexual intercourse can be painful and problematic.

Frequent bladder and urinary infections exacerbate and contribute to pain levels. Over 75% of survey respondents reported pain in either their bladder, urethra or pelvis.

Other symptoms

Fowler's syndrome can cause a loss of sensation, leading to difficulty knowing when the bladder is full.

Some women may experience a loss of bladder control, and also symptoms of urinary urgency and frequency and increased visits to the toilet.

Diagnosis

Fowler's syndrome is just one of many causes of chronic urinary retention in women. It is not easy to diagnose, with over70% reporting consulting more than four medical professionals before a diagnosis was made.

Women with suspected Fowler's syndrome may undergo varying tests, including:

- Ultrasound
- Flow rate test (uroflowmetry)
- Cystoscopy
- Urodynamic studies
- Urethral pressure profile
- Urethral Sphincter electromyography (EMG)
- Urethral sphincter volume



Treatments

Treatments for Fowler's Syndrome are being researched and developed.

There are several different treatment options, though some have limited results and may not be suitable for all patients.

Often patients have a poor urine stream but can still void almost normally. In these patients, your urology department will monitor your residual volume. If these volumes are low, no intervention is necessary.

Some people may be candidates for sacral nerve stimulation, which is the only treatment for Fowler's syndrome shown to restore voiding.

The patients with the most severe symptoms sometimes benefit from either the removal of the bladder and creation of a stoma or the use of the appendix to create an alternative route for emptying the bladder (Mitrofanoff).

Botox is currently being investigated as a treatment option.

Resources

General Practitioner

Will be able to refer you to specialists.

Continence Adviser / Specialist Nurse

Teach clean self-intermittent catheterisation techniques (if possible), advice on catheters, moral support.

Urology, Urogynaecology or Uro-Neurology services

May have experience on Fowler's Syndrome or may refer on to a specialist unit

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