Fowler's Syndrome

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Fowler's Syndrome

- Rare condition
- First described by Professor Clare Fowler in 1985 (Professor of Uro-Neurology, Institute of Neurology, UCL)
- Described by Professor Fowler as "the commonest cause of urinary retention in young women"
- Caused by failure of urethral sphincter to relax
- Other causes of urinary retention must be excluded for diagnosis to be made but there are positive features in the history and investigations that are also needed

Key clinical features

- **Female** – the condition has never been found in young men with urinary retention
- **Age between menarche and menopause** – peak age of onset is 26 yrs
- **No evidence of urological, gynaecological or neurological disease** – other causes of urinary retention should be excluded ideally by cystoscopy +/- MRI brain and spinal cord

Key clinical features

- **Retention with volume of >1000ml** – lesser volumes are less likely to be due to Fowler's syndrome
- **No sense of urinary urgency despite high bladder volumes**
- **Straining does not help emptying**
- **Sense of "something gripping" or difficulty on removing catheter used for drainage**

Key clinical features

- **No history of urological abnormalities in childhood or associated abnormalities of the urinary tract**

- **Association with PCOS and endometriosis** – up to half of patients affected have PCOS

Clinical history

- Urinary retention may be spontaneous or preceded by an event such as an obstetric, urological, gynaecological or surgical procedure. Childbirth may be a trigger.

- General anaesthetic use may be implicated as some women have had onset of urinary retention after procedures such as wisdom teeth extraction.

- Women may report difficulty voiding with an interrupted flow prior to onset of retention. UTIs may be common due to the bladder not emptying properly. Some women may experience back and suprapubic pain.

Fowler's Syndrome and Opiates

- Use of opiates may worsen voiding symptoms in those who have Fowler's Syndrome and are able to pass urine or they may trigger urinary retention.
- The action of opiates on the urethral sphincter and in causing retention is similar to what happens in Fowler's Syndrome without opiate use.
- Professor Fowler's theory: Abnormal sphincter EMG activity probably triggers excessive release of endorphins or encephalins in the spinal cord. These have the effect of suppressing sensations from the bladder resulting in no feedback to the higher brain centres and so inhibition of bladder contraction.

Investigations

- **DIAGNOSTIC** investigation is **urethral sphincter EMG** using a concentric needle electrode, to detect a particular type of abnormal activity which sounds like “helicopters” or “whales”

- Urethral sphincter EMG is only available in a limited number of centres in the UK

Investigations

Other tests include:

- **Urodynamics** – which would usually demonstrate a large capacity bladder without the usual sensations during the filling phase and an inability to pass urine after filling has stopped
- **Flow rate**
- **Residual volume** on ultrasound of bladder
- **Urethral pressure profile**
- **Urethral sphincter volume** measured by ultrasound – an overactive sphincter may enlarge due to continuous "muscle activity"

Management

- Treatments are still being researched and developed
- Monitoring residual volumes for those with low residual volume and ability to void almost normally
- Intermittent self-catheterisation for those with large residual volumes or long term catheter for those unable to self-catheterise
- Sacral nerve stimulation for severe cases – the only treatment shown to restore normal voiding.

Sacral Nerve Stimulation

- Similar to pacemaker
- Usually 2 staged implant procedure
- Stimulator is usually placed in buttock or abdomen if initial tests show that it is successful
- Long-term follow up at specialist centre is required.

- Complications include – leg pain, battery pain, lead displacement, lead fracture, loss of efficacy or battery site infection

Prognosis

- The condition is slowly being understood
- No absolute cure yet
- Aim of treatment is to ensure bladder emptying
- Bladder function may recover spontaneously in some women, especially those in whom symptoms are triggered after childbirth
- Some patients can suffer lifelong with debilitating effects on quality of life

References/Sources

- Fowlers Syndrome website
- National Hospital for Neurology and Neurosurgery Information
- Urinary retention PDF Fowler's Syndrome Website