

Fowler's Syndrome Handbook



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Forward

Written by Dr Jalesh Panicker, Consultant Neurologist in Uro-Neurology, University College London Hospitals

Chronic urinary retention in women may not be very common, however it is a significant social problem that can have a tremendous impact on quality of life. An important cause of urinary retention in young women is Fowler's Syndrome, and the diagnosis can be established by undergoing specialist tests.

It is beneficial to understand the cause of urinary retention as treatments are now available for managing the condition, particularly sacral neuromodulation. The journey undertaken in finding a cause for retention and receiving treatment is often challenging however, and it is therefore our hope that this handbook will provide some information about Fowler's Syndrome and support your need to find the best way to manage your bladder symptoms.

Bladder Health UK supports patients with urinary retention and in addition to this handbook, their website offers further help and advice. (www.bladderhealthuk.org)

Bladder Health UK

Bladder Health UK gives support to all suffering from chronic bladder illness, together with their families and friends. We are the largest bladder patient support charity in the UK with the most widely used website for those suffering with bladder illness who are seeking information, help and support. We also have an active presence on social media.

Our Vision is:

To work in partnership with key stakeholders to aim for a future free from the pain, stigma and isolation of bladder illness.

We will do this by:

Ensuring people with bladder health issues can expect a consistent level of healthcare support and treatment.

Encouraging research into new treatments and personal management strategies.

Working pro-actively with other like-minded organisations and charities to raise the profile of bladder illness issues.

Our Mission Statement is:

To help people live positively with bladder illness through the provision of personalised support and care.

Commitment to our members:

We will....

Share your journey to better bladder health

Provide you with support and contact with others with similar conditions and concerns

Be a trustworthy source of information and advice on the most up-to-date and available treatment options.

Elevate patient concerns to ensure your voice is heard.

Give you the confidence to make the right choice for you and your bladder health.

Bladder Health UK is a registered charity and has been in existence since 1994. Our membership is increasing year-on-year and our telephone advice line receives a constant stream of enquiries from those in need of support and information. Our membership also includes health care professionals, who join us to learn more about our resource information and the current therapies and treatment options that are available to aid sufferers.

We work with many other like-minded charities, focus groups, health organisations and other medical bodies to improve patient care and share information.

Since the beginning the charity has produced a very informative magazine providing up-to-date information on the latest treatments, research, resources and story features. There is a facility to write and have letters, opinions and experiences published or pose questions to our medical advisers.

We have a network of support available throughout the UK for our members. The groups are run by volunteers who are fellow sufferers.



Contents

Acknowledgements and special thanks	2
Foreword written by Dr Jalesh Panicker	2
Bladder Health UK	3
Contents	4
Explanation of medical terms used	5
<i>Symptoms, The Urinary Tract</i>	
Introduction to Fowler's Syndrome	6
<i>What is Fowler's Syndrome? How common is Fowler's Syndrome? What causes Fowler's Syndrome? Symptoms of Fowler's, Syndrome, Associated Conditions</i>	
Investigations & Diagnosis	8
<i>Residual Volume, Flow Rate, Urodynamics, Urethral Pressure Profile</i>	
Treatment	10
<i>Clean Intermittent Self-Catheterisation, In-Dwelling Urethral Catheters, Sacral Nerve Stimulation, Supra-Pubic Catheters, Mitrofanoff Surgery, Urostomy or Urinary Diversion</i>	
Coping with Fowler's Syndrome	12
<i>Coping with Catheter-Related Infection, Coping with Changes to Body Image, Your Doctor, Your Relationships, Relaxation, Clothing, Showering & Bathing, Coping with Emergency Hospital Admission with Fowler's Syndrome, Travelling with Fowler's Syndrome, Coping Strategy, Support, Useful Resources</i>	
Frequently asked questions	15
Useful websites & resources	16
Appointment Notes	17
Contact Bladder Health UK	18

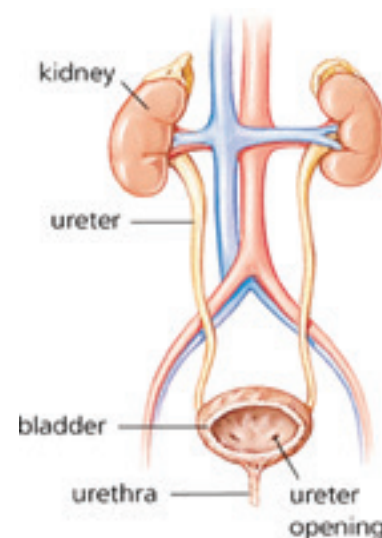
Explanation of medical terms used

Symptoms

- **Urinary retention (inability to pass urine normally)** – either being able to pass urine with difficulty leaving significant amounts still in the bladder or being unable to pass urine at all (complete retention)
- The absence of the normal sensation of urinary urgency expected with a full bladder.
- Pain and discomfort when the bladder reaches capacity and it's impossible to pass urine.

The Urinary Tract

- **Kidneys** - There are usually two kidneys in the body, situated just below the ribs, one on either side. They produce between 30-60ml of urine each hour which is carried from the kidneys via the ureters to the bladder.
- **Ureters** - These are two hollow, muscular tubes, about 24-30cm in length, through which urine travels from the kidneys to the bladder by muscular contractions. The tubes enter the bladder at an angle to prevent urine flowing back to the kidney.
- **Bladder** – This is a muscular bag which expands to store urine. When empty, the bladder is small, and the muscular walls expand as the bladder fills. Voiding occurs when the bladder muscle, the detrusor, contracts and the sphincters open.
- **Urethra** – This is the small outlet tube from the bladder through which urine is passed. In women, the opening of the urethra is at the top of the vaginal opening. Men have a longer urethra than women.
- **Sphincters** – These are two ring-like muscles surrounding the urethra. The external sphincter is under voluntary control and relaxes when you urinate.

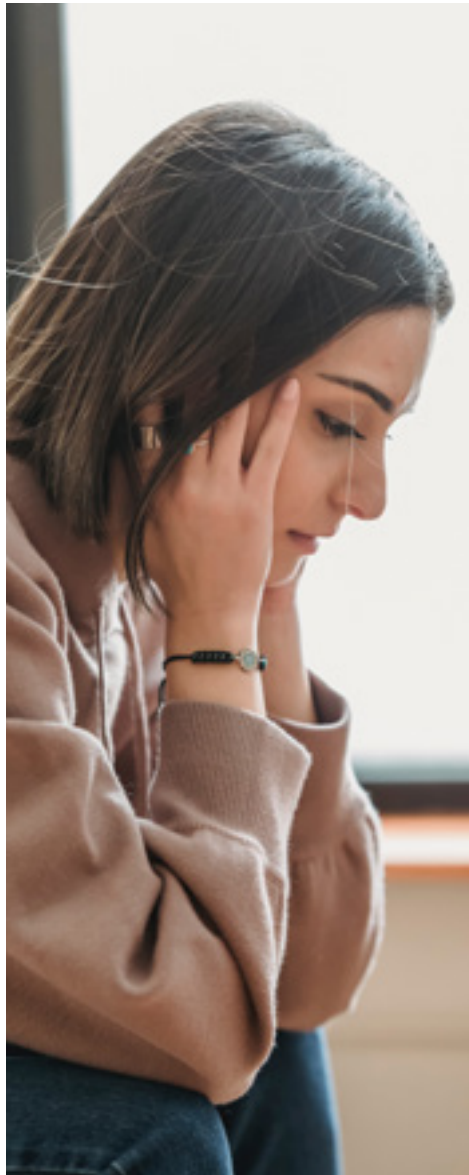


Introduction to Fowler's Syndrome

Fowler's Syndrome is a condition which was first described in 1985 by Professor Clare Fowler and colleagues. It is a cause of urinary retention (inability to pass water normally) in young women. While not common, it is a debilitating condition. The abnormality lies in the urethral sphincter and its failure to relax to allow urine to be passed normally. There is no neurological disorder associated with the condition. A significant proportion of those affected have associated polycystic ovaries and unexplained chronic pain.

The handbook has been written to help you:

- gain the knowledge required to understand your condition and symptoms.
- cope with your condition in a positive way.
- work with your doctor and other health professionals who will be treating you.
- enhance your quality of life.
- explain your condition to others who need to know and understand your symptoms.



What is Fowler's Syndrome?

Fowler's Syndrome is a cause of urinary retention in young women. Not all patients will be in complete retention. Some women can void, albeit with difficulty and report hesitancy and poor urinary stream. The symptoms caused by the inability to empty the bladder may include back pain, suprapubic pain (pain above the bladder) and dysuria (burning and discomfort whilst passing urine) as a result of urinary tract infection.

How common is Fowler's Syndrome?

Fowler's Syndrome is a rare disease despite being the most common cause of urinary retention in young females.

Fowler's Syndrome is listed on the Genetic and Rare Diseases Information Center at the following link <https://rarediseases.info.nih.gov/diseases/2365/fowlers-syndrome>

What causes Fowler's Syndrome?

The cause of Fowler's Syndrome remains unknown and is currently the subject of research.

Symptoms of Fowler's Syndrome

Fowler's Syndrome symptoms are caused largely by the inability to empty the bladder. Not all patients will be in complete urinary retention. Some women can void, albeit with difficulty and report hesitancy and poor urinary stream.

Women with Fowler's Syndrome are prone to developing urinary tract infections at which time they can experience suprapubic pain (pain above the bladder area), back pain and dysuria (discomfort or burning whilst passing urine).

Associated conditions

A significant proportion of women affected by Fowler's Syndrome have polycystic ovary syndrome.

For further information on Polycystic Ovary Syndrome visit the following link on the NHS website <https://www.nhs.uk/conditions/polycystic-ovary-syndrome-pcos/>

Investigations & Diagnosis

Residual Volume

An ultrasound scan of the bladder gives information on how much urine remains in your bladder after you have voided.

Flow Rate

If you are able to pass urine, a flow rate test is sometimes suggested. You will be asked to sit on a special toilet that measures the speed at which you pass urine and how long it takes you to void. You may subsequently be given a bladder scan to see how much urine remains after you have been to the toilet.

Urodynamics

Urodynamics is useful if you are in complete retention. You will have a small catheter placed into your bladder and one into your rectum. The bladder is slowly filled with saline (salt water) and is monitored for irregular spasms. When your bladder is full you are asked to try to pass urine. The catheters let the urology team see how much pressure your bladder muscle generates for a particular urine flow rate.

Urethral Pressure Profile

A catheter is inserted into the urethra and saline is infused slowly through the catheter. The catheter is then withdrawn and re-inserted (six times) into the bladder whilst the pressure of the urethral sphincter is measured. This test gives information on how much pressure is generated by the sphincter, and how overactive the muscle is.

Treatment

Once the diagnosis of Fowler's Syndrome has been established, you and your doctor should discuss treatment options, and agree the best approach to managing and treating your particular symptoms.

Before beginning any treatment, here are some questions you may find helpful to ask:

- How does the treatment work?
- How is it given?
- Are there any side effects or risks to this treatment, and what are they?
- Is the treatment known to have good results in other Fowler's Syndrome patients?
- What are the alternatives?

NB Urethral access treatment can prove painful and traumatic for sufferers and is therefore not recommended long-term



Clean Intermittent Self-Catheterisation

In order to fully empty the bladder, you will be asked to pass a catheter via the urethra to drain urine at regular intervals during the day to ensure that stagnant volumes of urine are not remaining in the bladder to cause infection. The catheters are single-use only and can be disposed of after use.

You will be taught the procedure by a nurse practitioner. She/he will also put you in touch with a dispensing appliance contractor who will be able to provide you with catheters and products going forward.

There are many different types of catheters available, and it might take a while to find the one that best suits you. You should be given the details of a dispensing appliance contractor by your clinician from whom you will be able to order further catheter supplies. Bladder Health UK can also provide information on dispensing appliance contractors if you are struggling.

In-Dwelling Urethral Catheter

A urethral catheter (also referred to as a 'Foley' catheter) is a closed sterile system with a catheter and a retention balloon that is inserted into the urethra to drain the bladder. The catheter will be connected to a drainage bag. You

will be able to order further drainage bags from a dispensing appliance contractor and also order securement devices. There are many different sized bags and securement devices to choose from and it can take a while to find the one that works well for you.

Surgical Interventions

Supra-pubic Catheter

If you are unable to empty your bladder, your consultant may suggest the insertion of a supra-pubic catheter.

A supra-pubic catheter is a catheter that is inserted into the bladder via an incision in the stomach just above the pubic bone. The procedure will usually be carried out under a local anaesthetic or sometimes under a general anaesthetic. The catheter is kept from falling out of position by a small balloon at its tip.

The urine can drain out from the catheter into a drainage bag (free drainage) or alternatively you can have a valve at the end of the catheter which can allow urine to be drained directly into the toilet or into a bag once your bladder feels full.

There are several types of drainage bags available. Leg bags are held in place with straps and a holster and can be worn under loose clothing during the day. During the

night, you will need a larger bag – a night drainage bag – which can be attached to a leg bag or alternatively, a nightstand overnight.

The catheter is usually changed at regular intervals depending on the type of catheter you have and your situation. Changes will usually be carried out by a nurse.

Sacral nerve stimulation (SNS)

Sacral nerve stimulation or sacral neuromodulation involves implanting a small medical device under the skin in the upper buttock. The device uses mild electrical stimulation of the sacral nerve that influences the behaviour of the bladder, sphincter and pelvic floor muscles.

If you are considered eligible for SNS, then the first stage would be the insertion of a temporary lead into a small naturally occurring hole in the lower spine using an external stimulator. Your response to the treatment will then be assessed over a seven-day period. For approximately two thirds of women voiding will be restored.

If the trial of the device is a success the stimulator will be placed internally in the buttock or abdomen.

You will generally be reviewed in clinic approximately four times a year once you have had the device implanted.

Mitrofanoff

Mitrofanoff surgery involves creating a channel using tissue from the appendix or bowel into the bladder. A valve is created where the channel meets the bladder to prevent the leakage of urine.

When you need to empty your bladder, you will be able to insert an intermittent catheter into the channel and then remove it when the bladder is empty.

Urostomy or Urinary Diversion

If other interventions do not prove successful, you may be asked to consider a Urostomy. A urostomy is an operation to create a channel through which you can pass urine following removal of the bladder.

Coping with Fowler's Syndrome

Coping with Catheter-Related Infection

Catheter associated urinary tract infection (CAUTI) is a common issue for those suffering from Fowler's Syndrome.

In-dwelling catheters can encourage bacteria to travel along catheter and cause infection in the bladder or the kidneys.

Some of the common symptoms of a CAUTI are:

- Burning or pain in the bladder
- Increased frequency of urination
- Feeling hot and running a temperature
- Blood in the urine
- Nausea

These symptoms are not always present if you have a catheter associated urinary tract infection.

Most CAUTI can be treated with antibiotics and by a change of catheter. Your doctor will be able to advise you on which antibiotic is best to use for the infection.

While you are symptomatic you can reduce your discomfort by

- Drinking plenty of water to dilute your urine.

- Alkalinising your urine by taking potassium citrate or bicarbonate of soda.
- Taking pain relief medication as prescribed by your doctor
- Using a heat pad or hot water bottle, if you are comforted by warmth; or a cold compress (such as a frozen pack of peas wrapped in a towel) on the site of your pain.
- Trying to distract your attention by watching television, playing a relaxation audiobook, or soothing, relaxing music.

How to Cope with the Emotional Impact and Changes to Your Body Image

Changes in your physical health will inevitably have some impact on your mental health. It can initially feel very overwhelming when you receive a diagnosis but please remember that you are not alone. Be kind to yourself and allow those up and down emotions.

Sharing your feelings with a counsellor is useful for some in helping them to come to terms with the changes in body image and lifestyle that a diagnosis of Fowler's Syndrome will inevitably bring with it.

Bladder Health UK provides an Advice Line for anyone with chronic bladder illness. We are always here to listen to anyone who is struggling emotionally.

Talk to your family and friends about Fowler's Syndrome. We provide useful leaflets which may help your family and friends to understand how best to support you and to give them a better understanding of the condition.

Joining a closed group on Facebook for sufferers of Fowler's Syndrome or sufferers of chronic bladder illness can help you to gain a better understanding of your condition and to get support on difficult days.

Your Doctor /GP

- It is important to build a good relationship with your doctor, in which respect BHUK can provide an information pack for your GP.
- If you feel that your doctor is unhelpful, or that your concerns are not being heard, you are free to look for another doctor, although this may mean moving to a different practice.
- Keep your doctor fully informed by keeping a diary and by being clear and concise about your symptoms and reactions to treatment.

- If you have questions for your doctor, write them down beforehand, which saves time and establishes priorities.
- If you have a lot of concerns, ask the receptionist for a longer appointment so that you need not feel rushed.
- If you feel that you need support, take a family member, or good friend with you to your appointment.

Your relationships

- Some of your symptoms may worry your partner, family and friends. Be open with them about your condition so that they can understand and help. It is important that those you love know that your occasional withdrawal from them does not mean a lack of interest in your relationship. Having Fowler's Syndrome can be hard – but being a partner, relative or friend can also be difficult, because they care and are concerned. Talking and explaining how you are feeling helps.
- If you find sexual intercourse painful, ask for your partner's help. Using lubricant can be helpful and, similarly, a change in position. If not, there are ways to satisfy each other without full penetration. With love and co-operation, a caring relationship can be rejuvenated.

- Be prepared to bury your pride and accept help from those who offer it.
- If your partner would like further information or support, to understand more about living with someone with Fowler's Syndrome, they can speak to us on the Bladder Health UK advice line. (0121 702 0820)

Relaxation

- Learning relaxation techniques can be very helpful. You can buy relaxation CDs or DVDs, or you may prefer to find a reputable therapist to help you learn self-hypnosis techniques. Your doctor or health visitor may be able to give you the necessary information.
- Aromatherapy can be a helpful way of relaxation.
- Listen to your body if it needs a rest!

Clothing

Choosing clothing that you feel comfortable in is the most important thing. Wide leg, loose trousers will accommodate a leg bag if you have an in-dwelling catheter or alternatively, a long skirt. If you like wearing a short dress or skirt, then you can order belly bags from your dispensing appliance contractor. Be patient - in time you will adapt to a new routine and find clothes that work well for you.

Showering & Bathing

In-Dwelling Catheters

If you have an in-dwelling catheter you should remember to keep your collection bag or flip flow attached to the catheter while in the bath or shower.

SNS Device

During the SNS trial, while the device externalised, you should not shower or bath. Once the permanent device is fitted and the site is well healed you can shower or bath (usually one week after the device is implanted).

Coping With Emergency Hospital Admissions with Fowler's Syndrome

If you are admitted to hospital as an emergency with retention it may be difficult to explain to staff what you suffer from. Our Fowler's Hospital Card which is designed to be small enough to be stored in a wallet or a purse will explain to staff exactly what you are suffering from and where they can quickly find information on Fowler's Syndrome.



Travelling with Fowler's Syndrome

When out and about with Fowler's Syndrome it is useful to take a bag with you of any supplies you would usually need when you are at home, together with a small towel or change of clothes in case you need them.

If you are on a long journey and have an in-dwelling catheter, it may be helpful to have a night bag attached to the catheter to avoid frequent changes.

If you have an SNS device fitted you will need to take the SNS pack and control with you and keep it with you at all times.

If you are traveling abroad, it may be helpful to get a letter from your GP or consultant explaining what Fowler's Syndrome is and why are you carrying medical supplies with you.

Coping Strategy

Fowler's Syndrome may put some limitations on life, which at times can be very frustrating. When you are in pain, uncomfortable or tired, - stop, rest and accept help.

Never be afraid to say 'Sorry, but I need a rest just now.' Once you find coping strategies to help you live as full a life as you can, the limits will seem to lessen. Being positive is a definite bonus.

Support

At times of difficulty, you may need questions answered and support from others who also have Fowler's Syndrome. Joining Bladder Health UK, where you can learn from others and share your experiences, can be very helpful. BHUK has a confidential advice line which is open daily. It also has other Fowler's Syndrome sufferers who are willing to speak to you if you are struggling.

Useful Resources

RADAR Keys

RADAR keys for disabled toilets are available from your local council, tourist information offices or contact:

Disability Rights UK - RADAR, 12 City Forum, 250 City Road, London EC1V 8AF

Tel: 020 7250 3222. Website: www.disabilityrightsuk.org

BHUK Invisible Disability Card

If you are in the queue for a toilet while out and about and would like to use a disabled toilet you can show your BHUK Invisible Disability Toilet Card which reminds anyone who challenges you that you are entitled to do so.

Fowler's Syndrome Hospital Card

A wallet-sized card you can carry in your purse explaining what Fowler's Syndrome is and where further information can be found if you need to be admitted as an emergency to hospital.

BHUK Can't Wait Card

Our Can't Wait Card gives priority access if you are waiting in a queue for a toilet when out and about.

Catheter Bag Covers

Catheter bag covers or tubie pads (for SPC users) can make things feel less medical. There are a number of on-line shops on Etsy that sell these such as Loulabellie, Sew & Zo and Sew Lovely Tubie.

Electric Heat Pads

Electric heat pads are available on Amazon and can help to relax the muscles and reduce pain during an infection.

Long hot water bottles are also useful, particularly if you are experiencing kidney pain as they tie around the waist. These are also available on Amazon.

Night Stands

If you have an in-dwelling catheter and use a night bag, you can obtain a night-stand to hook your bag onto over-night. Covered night-stands are also available so that the bag is hidden if you would prefer.



Frequently Asked Questions

As others try to understand the changes in you, you may be asked a number of questions such as:

What is Fowler's Syndrome?

Fowler's Syndrome is a cause of urinary retention in young women. The symptoms caused by the inability to empty the bladder may include back pain, suprapubic pain (pain above the bladder) and dysuria (burning and discomfort whilst passing urine) as a result of urinary tract infection.

What is it like to suffer from Fowler's Syndrome?

Living with Fowler's Syndrome is challenging, and the symptoms are unpredictable and change rapidly. There is a general lack of knowledge among the medical profession and the general public of the condition.

Useful websites

Bladder Health UK

Bladder Health UK provides a wide range of relevant information and resources. We have an Advice Line for anyone needing a little extra support and are active on social media.

University College London

Offers a website with information on Fowler's Syndrome at the following link

<https://www.ucl.ac.uk/ion/fowlers-syndrome>

Further Resources available from the Bladder Health UK include:

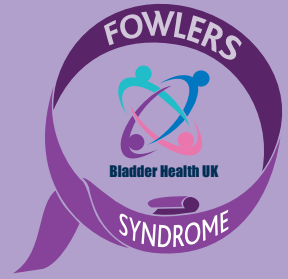
- **A members' magazine** – mailed to members three times per year, packed with features on chronic bladder illness from the latest worldwide research, members' letters and a section with questions answered by a member of BHUK's Medical Panel.
- **Medical Pack** – for GPs and health professionals containing information on Fowler's Syndrome.
- **Fact Sheets** – over 80 information sheets available on medication, treatments and complementary therapies.

- **The BHUK Library** – contains bladder information books for members to borrow.
- **Can't Wait Cards** – are provided on membership and available for purchase in several languages.
- **Fowler's Hospital Card** outlines the problems of bladder illness and provides a useful resource for members entering hospital.
- **Surgery Leaflet** – offering information about the different types of bladder surgery available.
- **BHUK Website** – information on Fowler's Syndrome, how to become a member and also offers a forum for generating discussion via the internet. www.bladderhealthuk.org
- **Social Media** – social media platforms on Facebook, Twitter and Instagram putting out information on chronic bladder illness on a daily basis.

Appointment Notes

Please photocopy this page for your own use.

Making notes during important appointments can help you to ensure that you speak about everything you want to but also that you remember all that was said. Use this page to write down anything prior to or during your next appointment that you feel is important to discuss.



This handbook has been produced and published by Bladder Health UK

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