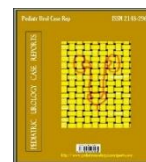


**PEDIATRIC UROLOGY CASE REPORTS**

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<http://www.pediatricurologycasereports.com>**A rare cause of chronic urinary retention in children, Fowler's syndrome: A case report****Agnese Roberti<sup>1</sup>, Francesco Turrà<sup>1</sup>, Maria Esposito<sup>1</sup>, Anna Maria Cotrufo<sup>1</sup>, Federica Marinoni<sup>2</sup>, Giovanni Di Iorio<sup>1</sup>**<sup>1</sup>Department of Surgical Sciences, Pediatric Urology Unit, Santobono- Pausilipon Children's Hospital, Naples, Italy.<sup>2</sup>Department of Pediatric Surgery, "V. Buzzi" Children Hospital, Milano, Italy.**ABSTRACT**

In 1986, Fowler and colleagues described a condition currently known as Fowler's syndrome in young women with unexplained urinary retention associated with polycystic ovary syndrome. Herein, we present a case of a 13-year-old woman diagnosed with Fowler's Syndrome as a result of clinical, radiological and urodynamic electromyogram (EMG) findings. Fowler's syndrome should be considered in the differential diagnosis in children with unexplained chronic urinary retention.

**Key Words:** Chronic urinary retention, Fowler's syndrome, polycystic ovary, persistent suprapubic pain, child.

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**Introduction**

Fowler's Syndrome is a clinical condition described in 1988 that includes certain pathologies such as urinary retention, electromyographic abnormalities of the striated urethral sphincter, and polycystic ovaries [1,2]. This syndrome is usually diagnosed in young women with unexplained urinary retention, which occurs as an isolated phenomenon and is very rare in children [3]. Some etiologic factors, including a history of general anesthesia for varying reasons,

prolonged use of opiates, and delivery are accused in the development of this condition [4]. In addition, the management of urinary retention, of which the cause is not fully explained, is very challenging and our understanding of this problem has made little progress since the syndrome was first described [1,5]. So far, the main treatment for restoring micturation in this syndrome is sacral neuromodulation [6].

We report a case of Fowler's Syndrome in a 13-year-old girl who should be considered in cases of chronic pelvic pain and unexplained chronic urinary retention without spinal cord injury.

**Case report**

A 13-year-old girl was presented to our unit with persistent suprapubic pain and urinary retention. Four months ago, abdominal

computerized tomography (CT) scan and exploratory laparoscopy were performed to investigate pelvic pain and were negative for digestive pathologies.

At birth, she was diagnosed as CAKUT (Congenital Anomalies of the Kidneys and of the Urinary Tract) -like complex uropathy with duplicity of the right excretory system, left kidney dysplasia, left vesicoureteral reflux (VUR) and left ureterocele. In the first years of life, it was understood that she had received endoscopic treatment at other centers for mentioned urinary conditions.

Despite surgical endoscopic treatments, the residual function of the left kidney was 10% in renal scintigraphy with Tc-99m mercaptoacetyl triglycine (MAG3) with compensatory function of the contralateral kidney.

Her growth was steady and her menarche was 9 years old. Four months after the laparoscopic exploration, the patient returned to our attention for chronic abdominal pain associated with decreased urinary frequency, prolonged urination time and a gradual complete lost of her ability to void.

The physical examination showed a normal abdomen and external genital findings with a height of 1.59 m and a weight of 47 Kg. The blood tests were negative for inflammatory or renal pathologies.

A pelvic ultrasound scan (US) did not find any pathology that could explain urinary retention, but identified multiple follicular cysts in the bilateral ovaries [Fig. 1]; therefore a hormonal panel was ordered and showed a lower progesterone value (<0.05 ng/ml).

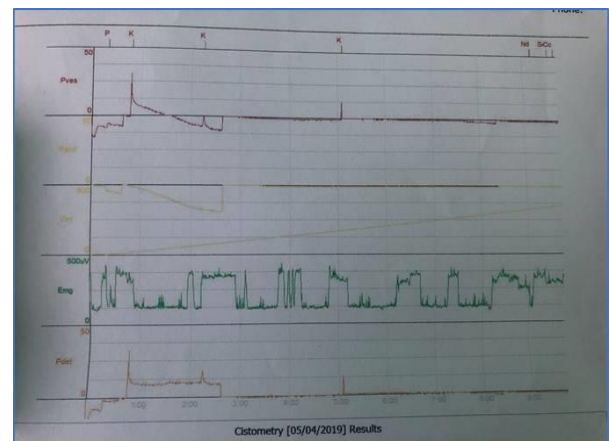
The voiding cystourethrography (VCUG) was performed and showed no vesicoureteral reflux and closed bladder neck.

In the urodynamic study, we found an insensitive, hypotonic bladder and a sphincter

EMG (Electromyogram) showing low detrusor activity and high pressures of the perineal plane associated with abnormalities of electromyography at level of striated urethral sphincter [Fig.2].

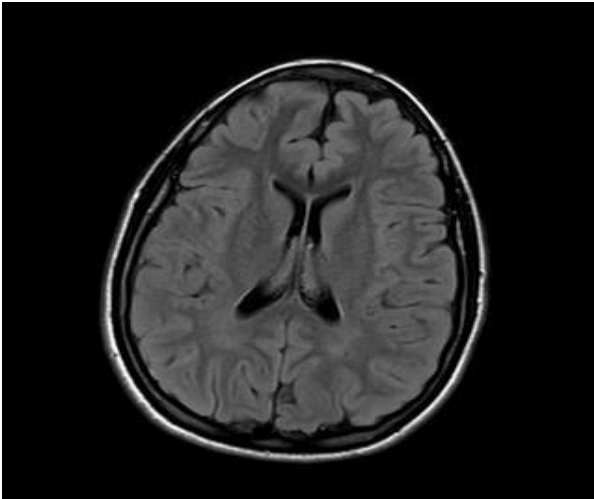


**Fig. 1.** The pelvic ultrasound showed multiple follicular cysts in ovaries.



**Fig. 2.** At urodynamic study: asensitive and hypotonic bladder; the sphincter electromyogram (EMG) showed a decreased detrusor activity and high pressures of the perineal plane with abnormalities of electromyography at level of striated urethral sphincter.

A pediatric neurologist reported no neurological conditions in physical examination and a magnetic resonance imaging (MRI) of the brain and spine and the somatosensory-evoked potentials (SEP) were normal [Fig. 3, Fig. 4].



**Fig. 3.** Magnetic resonance imaging (MRI) of the brain was normal.

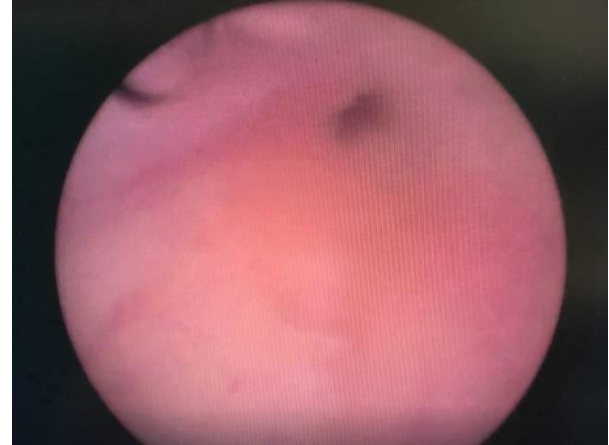


**Fig. 4.** Magnetic resonance imaging (MRI) of the spine was normal.

Cystoscopy and uteroscopy were performed in the evaluation of suspected operative urinary tract injury and there was an increase in urethral sphincter muscle with trabecular and hyperemic bladder, while vagina and uterus were normal [Fig. 5].

Psychological and psychiatric evaluations showed no significant problems, but she showed a stable mood associated with some depressive aspects due to her hospitalization. Prophylactic antibiotic therapy and clean intermittent catheterization (CIC) were recommended. Two months later, a good

tolerance of CIC was achieved with gradual reduction of pain symptoms in the suprapubic region. In addition, a neurostimulation of the tibial nerve was initiated.



**Fig. 5.** Uteroscopy showed a normal vagina and uterus.

After that, she suffered from suprapubic pain and recurrent symptomatic afebrile urinary infections caused by multi-resistant bacterial strains such as *Klebsiella*, *Pseudomonas* and *Escherichia* spp.

Finally, after 3 months of hospitalization, based on clinic history (chronic urinary retention, hypoprogesterinemia and bilateral polycystic ovary), and on urodynamic findings that showed an asensitive and hypotonic bladder, we have reached the diagnosis of Fowler's syndrome.

### Discussion

In 1988, Fowler et al. [1] reported that the urinary retention or voiding difficulty due to an uncertain causal disorder in young women is associated with a characteristic electromyographic (EMG) abnormality in the striated urethral sphincter muscle. In addition, the majority of young women had a history or clinical features of polycystic ovaries [1]. The incidence rate is 2 / 10,000,000 per year and affects only women aged 20-30 years [7].

However, as in the case presented here, FS is rare in childhood [4].

Even though its physiopathology is not completely understood, this disorder seems to be due to a primary failure of relaxation of external urethral sphincter, which results in inhibition of detrusor function [2, 8]. Several etiological factors have been proposed to explain the occurrence of urinary retention. Initially, the hormonal channelopathy theory was proposed by Fowler's group [9, 10]. This theory suggested that reduction in serum progesterone associated with polycystic ovary syndrome (PCOS) resulted in autonomic electrical activity which cause an instability of the sphincter muscle membranes. Other researchers have suggested that urine retention in these women is the result of maladaptive behavior and has no organic origin [8]. FS was reported in two sisters with PCOS and therefore genetic susceptibility to polycystic ovaries was suggested to be a risk factor for the development of the disease [7]. In addition, these patients usually come up with a history of surgical procedures [11]. The case we presented here had a bilateral polycystic ovary and low progesterone levels. In addition, it was found that she had benefited from endoscopic treatment of the left VUR and the ureterocele. The clinical manifestations of Fowler's syndrome are retention of sterile urine of more than 1 L, an asensitive bladder, and poor tolerance of clean intermittent self-catheterization that does not help straining [4]. The diagnosis of Fowler's syndrome remains challenging, even more complicated in children. Dysfunctional voiding, primary bladder neck obstruction, idiopathic detrusor underactivity, and chronic intestinal pseudo-obstruction should be considered in the differential diagnosis of chronic functional urinary retention [8]. A typical clinical history,

evaluation of the sphincter volume, sphincter pressure profilometry and electromyography are the determinants of Fowler's syndrome [8]. The fact that the only tool that can reliably diagnose Fowler's syndrome is a concentric EMG needle electrode makes it difficult to diagnose in everyday practice [8]. In patients with Fowler's syndrome, it was also found that abnormal EMG activity of the external anal sphincter commonly coexists along with abnormal external urethral sphincter EMG activity [8, 12]. In the present case, in addition to our clinical findings, a urodynamic study with EMG helped us to diagnose.

There are several methods for the treatment of Fowler's syndrome. One of these is the process of emptying the bladder by intermittent self-catheterization [11]. This is usually helpful in alleviation of urinary retention; however, it may increase the frequency of UTI, as in our case. Another treatment is botulinum toxin injections that temporarily restores voiding without serious side effects [13]. Finally, sacral neuromodulation has been proposed as a safe and effective management option [2, 11]. In conclusion, Fowler's syndrome is a very rarely diagnosed pathology in children. However, it should be considered in the differential diagnosis in children with unexplained chronic urinary retention.

### ***Compliance with ethical statements***

*Conflicts of Interest: None.*

*Financial disclosure: None.*

*Consent: All photos were taken with parental consent.*

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