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Letter to editor

Not only enuresis: do not disregard organic disorders

Pietro Ferrara[1,2], *Giovanna Magli*[3], *Elena Malavolta*[3], *Emidio Procaccini*[4], *Luca Massimi*[5], *Antonio Gatto*[6]

[1]Department of Medicine and Surgery, Università Campus Bio-Medico, Roma, Italy; [2]Operative Research Unit of Pediatrics, Fondazione Policlinico Universitario Campus Bio-Medico, Roma, Italy; [3]Institute of Pediatrics, Catholic University, Rome, Italy; [4]Department of Neuroscience, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy; [5]Pediatric Neurosurgery, Fondazione Policlinico Universitario A. Gemelli IRCCS, Rome, Italy; [6]Department of Pediatrics, Fondazione Policlinico Universitario A. Gemelli IRCCS, Roma, Italy

To the Editor,

We recently faced an unusual case of Nocturnal enuresis (NE). NE is a common condition in children that can cause low quality of life for the child and his/her family. NE is defined as intermittent bedwetting with any frequency while sleeping in children, according to the International Children's Continence Society (ICCS). NE in children without any other lower urinary tract symptoms (LUTS) and without a history of bladder dysfunction is defined as monosymptomatic NE (MNE). Children with concomitant daytime incontinence are defined nonmonosymptomatic NE (NMNE) (1). Three major pathogenetic mechanisms have been established as essential and they are nocturnal polyuria, detrusor overactivity, and associated with failure to awaken in response to bladder sensations. Each theory can be supported by various studies, but no one is reported to explain bedwetting in all children (2). Psychopathology is important too: 20-30% of the children with NE have at least one psychological/psychiatric disorder at rates two times higher than non-wetting children.

A 7-year-old female was referred to the Operative Research Unit of Pediatrics, Fondazione Policlinico Universitario Campus Bio-Medico for the recent onset of sphincter troubles and recurrent low urinary tract infections. The patient presented urinary urgency associated with daytime incontinence, bedwetting almost every night in the previous 3 months, and sometimes encopresis. Moreover, the patient had sleep disorders such as snoring and sleep talk. The patient had achieved bladder continence at the age of 3 and was diagnosed secondary NMNE. We investigated any possible lumbar trauma or any psychological or social problem, but nothing was found. The physical examination turned out to be normal. The first diagnostic evaluation included a bladder ultrasound that showed detrusor wall thickness slightly increased compared to normal values. First of all, we prescribed deamino-delta-D-arginine vasopressin (dDAVP) 120 mcg associated with oxybutynin 5 mg and educational therapy. After 3 months of treatment, the patient had no improvement. Although the neurological examination was negative, as a consequence of the onset of sphincter troubles without a history of traumatic lesions, we decided to perform a magnetic resonance imaging (MRI) of the spine. Unexpectedly, MRI highlighted the presence of hydrosyringomyelia, extended from D6 to D10, with a maximum diameter of 4 mm. Moreover, MRI revealed lipoma of the terminal filum and the presence of a synovial cyst in the right interapophyseal join space between L5-S1. These findings could explain sphincter disorders and urological manifestations of the patient. The patient underwent a neurosurgical evaluation and currently, she is waiting for a possible surgical treatment.

NE in children without any other LUTS and without a history of bladder dysfunction is defined as MNE. A child with symptoms such as urgency, hesitancy, frequency, day-time incontinence, or fecal

incontinence is said to have NMNE, which is more frequently associated with abnormalities of the urinary tract. This distinction is important since it affects the treatment strategy.

The asset to NE should be systematic and clinical-oriented in order to identify the NE subtype. A detailed medical history should be obtained, focusing on current symptoms, presence of intercurrent day-time urinary incontinence, previous trauma, and comorbid disorders, especially encopresis, constipation, and psychological and neurological disorders.

Encopresis is defined as repeated involuntary fecal soiling in the underpants in children older than 4 years of age. In the 90% of cases, encopresis is a functional disorder secondary to retentive constipation. In the 5-10% of cases, encopresis is the underlying symptom of an organic cause that can be anatomic, neurologic, metabolic, or iatrogenic. The association between NE and encopresis should pay attention to organic conditions.

Family history should also be investigated and supplemental questionnaires should be used to support the diagnosis. Furthermore, a complete physical examination is required, including palpation of the abdomen, anal and genital region examination. The child with NE should be examined carefully for neurological and spinal abnormalities in order not to miss an underlying neurological cause.

Further investigations, such as bladder sonography and urine flow measurement (uroflowmetry), can be required as first diagnostic approach in order to identify neuropathic bladder or sphincter dysfunctions.

In our case, the patient presented a secondary NMNE and comorbid disorders such as encopresis. The physical and neurological examination was silent, and no psychological or social problems intercurred. Bladder ultrasound showed detrusor wall thickness of 0.7 cm, slightly increased compared to normal values (which is 0.3 cm when the bladder is full and 0,5 cm when the bladder is empty), thus initial therapy with dDAVP associated with oxybutynin was started (3,4). However, after 3 months of treatment, the patient had no improvement. At this point, the suspicion of an underlying neurologic cause was high.

In the pediatric age, a wide spectrum of spinal cord lesions could be associated with abnormal urological manifestation, such as trauma, spina bifida occulta, tethered cord, intramedullary epidermoid cyst, synovial cysts, intramedullary tumors, hydrosyringomyelia and Chiari-I malformation (5,6).

Anytime a neurologic cause is suspected, MRI of the spine is fundamental for the diagnosis.

In our case the MRI highlighted the presence of hydrosyringomyelia from D6 to D10, lipoma of the terminal filum, and the presence of synovial cyst between L5-S1.

Syringomyelia is a disorder in which a fluid-filled cyst forms within the spinal cord and, in particular, hydromyelia is a focal dilatation of the central canal. In children, syringomyelia usually develops in the setting of congenital abnormalities, such as Chiari-I malformation or tethered cord, but it can also be secondary to meningitis, spinal trauma, and tumors. Each of these conditions leads to a disturbance of normal CSF flow dynamics. Syringomyelia is frequently discovered on incidental spinal cord imaging, but it can also be symptomatic causing different motor and sensory symptoms and signs depending on the syrinx's location. Symptoms of autonomic bladder and bowel dysfunction are very uncommon until end-stage spinal cord dysfunction. Lumbar spine synovial cysts are benign growths adjoining the facet joints that may induce low back pain, lumbar radiculopathy, and neurological deficit. Filum terminale lipoma (FTL), or tight filum terminale, is known to cause spinal cord tethering, which results in various spinal symptoms collectively called tethered cord syndrome (TCS).

In summary, the significance of our case is to remark the difference between primary and secondary NE. In particular, in secondary NE, although the neurological examination is normal, if the initial therapy is not effective, any possible organic disorder must be investigated.

Ethic Committee: This study was conducted in accordance with the Ethics Committee of the Pediatrics unit of Fondazione Policlinico Universitario Campus Bio-Medico, with no protocol number.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity

interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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Correspondence:
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Elena Malavolta, MD
Institute of Pediatrics, Catholic University, Rome, Italy
Largo Francesco Vito 1, Rome, 00168 Italy
E-mail: elena.malavolta96@gmail.com