A challenging review of childhood incontinence: rare complications of dysfunctional elimination syndrome in an epileptic boy

Özgü Aydoğdu[1], Berk Burgu[1], Serap Teber[2], Şemsa Altugan[3], İlker Gökçe[1], Gülhiz Deda[2], Tarkan Soygür[1]

Departments of [1]Pediatric Urology, [2]Pediatric Neurology, and [3]Pediatric Nephrology, Ankara University Faculty of Medicine, Ankara, Turkey

SUMMARY: Aydoğdu Ö, Burgu B, Teber S, Altugan Ş, Gökçe İ, Deda G, Soygür T. A challenging review of childhood incontinence: rare complications of dysfunctional elimination syndrome in an epileptic boy. Turk J Pediatr 2011; 53: 100-103.

A multidisciplinary approach is mandatory in defining the cause and directing the treatment of childhood urinary incontinence. Both pediatricians and pediatric urologists should carefully evaluate a child with incontinence for possible overlapping etiologies, before labeling him or her as a refractory case.

We report an epileptic boy with dysfunctional elimination syndrome (DES) with associated rare complications of giggle incontinence and idiopathic urethritis, proving the need for a good voiding diary, patient history and structured symptom scores.

Key words: giggle incontinence, dysfunctional elimination syndrome, urinary incontinence, epilepsy.

Incontinence in childhood is a common symptom and requires a structured evaluation. Several overlapping causes can make the diagnosis challenging, and physicians can only cope with this problem with a detailed history and appropriate diagnostic tools. We report an epileptic boy with dysfunctional elimination syndrome (DES) with associated rare complications of giggle incontinence and idiopathic urethritis. The challenging diagnostic approach to illuminate the overlapping causes of incontinence is discussed herein. The possible interaction of laughing, incontinence, epileptic seizures, and urethritis in childhood is discussed in terms of cause, clinical outcome and therapeutic approach.

Case Report

A nine-year-old boy was first evaluated by the Department of Pediatric Nephrology and consulted to Pediatric Urology for urinary incontinence and refractory DES, which had been present for six years. According to the detailed medical history, blood spotting in the underwear and urinary incontinence during laughter were also present. All his symptoms were refractory to urotherapy that had been started six months ago. The uroflowmetric evaluation revealed a staccato pattern and significant residual volume. The total symptom score of voiding problems[1] was high and showed severe daytime incontinence (wetting pants), squatting and fecal soiling.

Besides anticholinergic (tolterodine, 2 mg/day) therapy, alpha-blocker (doxazosin, 0.03 mg/kg/day) was initiated since he had significant post-voiding residual volume. After one month, daytime incontinence, fecal soiling and blood spotting had diminished. However, the main complaint of the family that occurred on a daily basis -- the loss of large volumes of urininduced by laughter (giggle incontinence) had persisted despite the successful treatment of DES.

The patient was also diagnosed as epilepsy associated with nocturnal focal motor seizures. He had sudden, bilateral and asymmetrical tonic posturing of the limbs lasting 20-30 seconds, 1-2 times a week. The classification of the seizure

was thought to be supplementary motor seizure of the frontal lobe. The electroencephalogram (EEG) showed bilateral frontal sharp waves. Valproic acid therapy was initiated and he has not experienced any seizure for two years. The presence of epilepsy and the association of his seizures with urinary incontinence made the diagnosis more challenging in this patient. Micturition can be an autonomic sign in epileptic seizures. However, in another entity, termed 'reflex epilepsy', seizures can be triggered by micturition. To fully trace and clarify the causative factor, a video monitoring EEG was performed while the child was forced to laugh. Clinically, after laughing, he voided involuntarily, compatible with giggle incontinence. He was conscious, and no other motor, sensory or autonomic symptoms were demonstrated. The EEG did not show any ictal activity like fast activity, rhythmic spike waves or rhythmic slow activity, and was consistent with the interictal EEG consisting of bilateral frontal sharp waves (Fig. 1). The persistent incontinence associated with laughter was interpreted as giggle incontinence and not associated with the epilepsy. Methylphenidate (1.2 mg/kg/day) was initiated, and the giggle incontinence episodes totally diminished. After six months of tolterodine and doxazosin, all symptoms of DES disappeared without any relapse; he has remained under methylphenidate treatment for approximately nine months and is totally dry.

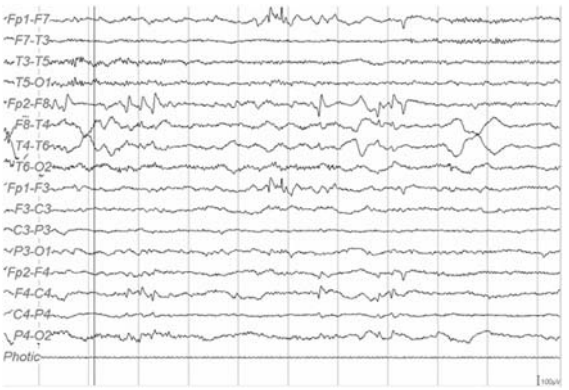

Fig. 1. The EEG shows sharp waves on bilateral frontal regions, which appear asynchronous. Electromyographic artifact is noted in temporal regions because of laughing *(low filter: 1 Hz, high filter: 35 Hz)*.

Discussion

The history is the most important component in the evaluation of a patient with incontinence. A thorough systemic approach is critical in defining the cause and directing the treatment. Most of the anatomic and neurogenic causes of incontinence can be eliminated by a careful history[2]. The wet child we present here is a very good example proving the need for a good voiding diary, patient history and structured symptom scores. A multidisciplinary approach is mandatory, and both pediatricians and pediatric urologists should not label a DES child as a refractory case before fully evaluating for overlapping etiologies of childhood incontinence.

The epileptic boy had not only severe DES causing daytime incontinence and fecal soiling, but also urinary incontinence during seizures and giggle incontinence.

Patients with DES often begin having symptoms 3-6 months after establishing urinary control. Children, who are not yet able to inhibit the voiding reflex, stay dry by voluntarily contracting their external sphincter during bladder contractions. Usually, persistence of this learned behavior leads to voiding problems. These patients often void staccato or fractionated patterns. Increased or decreased urinary frequency, urgency, urge-associated incontinence, enuresis, and fecal soiling are some of the common symptoms seen in the complex of DES. Instability of the bladder, increased bladder wall thickness, high post-void residual volumes, and recurrent urinary tract infections are often sequelae of this condition[3,4].

Idiopathic urethritis and giggle incontinence have previously been reported in association with DES[2,5]. Blood spotting in the underwear presented an additional challenge in diagnosing this patient. Idiopathic urethritis of childhood is a common pathology characterized by terminal hematuria, and urethrorrhagia is a common symptom characterized by red stains in the underwear[5,6]. A clear etiology has not been established and treatment alternatives vary. However, it has been previously speculated that DES is highly suspected to be the cause, and higher cure rates had been reported when children with urethritis were treated according to dysfunctional voiding guidelines, as in our

patient[5]. The main rationale for the use of alpha- blockers in children with DES is to minimize residual urine volume. However, use of alpha- blockers to overcome the symptoms of idiopathic urethritis has been reported. Herz et al.[5] compared the use of alpha-blockers and urotherapy to antibiotics and analgesics in children with idiopathic urethritis, and reported a higher success rate for the first group.

Giggle incontinence or enuresis risoria can be defined as complete bladder emptying occurring only with giggling or laughing while awake[2,7]. This was first described in 1959 by MacKeith[8] with further elaboration in 1984 by Williams[9]. This rare condition primarily affects girls. The urine is generally normal and the upper tracts are not affected. Concurrent dysfunctional voiding symptoms can be present in up to 95% of the patients[2,7]. Some authors conclude that detrusor instability in this group of patients contributes to wetting[2,10]. Although rare in boys, the typical history taken from the mother, i.e. 'He laughs so hard that he wets his pants' was enough to investigate this child for giggle incontinence. Giggle incontinence in this boy persisted despite the successful treatment of other DES symptoms. Methylphenidate, which is a neuro-stimulant drug that should be cautiously prescribed in an epileptic boy, was effectively used for the treatment of giggle incontinence[10,11]. Even at minimum dose, total dryness was achieved.

A detailed history revealed that this nine-year-old epileptic boy had urinary incontinence during his seizures. Incontinence is a common symptom in transient loss of consciousness. Micturition can be an autonomic sign in epileptic seizures. The supracallosal part of the medial frontal gyrus has been reported to include one of the suprapontine micturition centers; therefore, incontinence can potentially be a sign of frontal lobe seizure[12]. As our patient did not have nocturnal seizures for two years and was wet only during the day, we did not consider frontal lobe epilepsy as the cause of the giggle incontinence. However, seizures can also be triggered by micturition in another entity, termed 'reflex epilepsy'. While seizures observed in reflex epilepsy are no different from those in non-reflex epilepsy, they need specific stimuli, such as prolonged standing, emotion/pain and increased thoracic pressure caused by coughing, for micturition and defecation to occur[2,7]. A history of presyncope with diaphoresis, nausea and pallor as prodromal signs are strongly suggestive of reflex epilepsy, while tongue biting, muscle aches and cyanosis are more often associated with a true epileptic seizure[2,10]. It is difficult to ascertain whether this child was wet after an epileptic seizure or whether the micturition itself caused by laughter triggered the reflex epilepsy in this male patient with giggle incontinence. Similar to the chicken and egg story, urinary incontinence provides no additional evidence for or against the diagnosis of a true epileptic seizure. Romme et al.[13] showed that urinary incontinence can be present in 19% of the patients with reflex epilepsy. We thought that performing a video monitoring EEG while forcing the child to laugh with jokes, with the aim to record the simultaneous EEG, might help to distinguish between the possible diagnoses. The persistent incontinence associated with laughter was interpreted as giggle incontinence and not associated with the epilepsy. We then concluded that the giggle incontinence and epilepsy were present together coincidentally in this patient. However, it must be kept in mind that secondary enuresis nocturna may be a sign of a frontal lobe epilepsy.

The majority of the physicians dealing with pediatric incontinence refer to urodynamics or other imaging studies such as magnetic resonance imaging when there is a refractory case. These generally require anesthesia or sedation in the pediatric age group. Even in such a complex incontinence case with concomitant neurological problems, complete dryness was achieved with noninvasive diagnostic tools, underlining the fact that accurate history and bladder diaries generally provide sufficient information.

In conclusion, for the management of DES in children, clinicians should be aware of rare concomitant disorders, including giggle incontinence and idiopathic urethritis. Incontinence can be challenging, especially when it is associated with epilepsy. Since giggle incontinence is very rare in childhood, the reflex epilepsy should be discussed in the differential diagnosis. The majority of childhood incontinence cases, even with concomitant neurological problems, do not require invasive diagnostic tools.

REFERENCES

1. Akbal C, Genc Y, Burgu B, et al. Dysfunctional voiding and incontinence scoring system: quantitative evaluation of incontinence symptoms in pediatric population. J Urol 2005; 173: 969-973.
2. Chandra M, Saharia R, Shi Q, Hill V. Giggle incontinence in children: a manifestation of detrusor instability. J Urol 2002; 168: 2184-2187.
3. Chandra M. Common voiding disorders. In: Burg FO, Wald ER, Ingelfinger JR (eds). Current Pediatric Therapy. Vol. 16. Philadelphia: W. B. Saunders Co; 1999: 837–840.
4. Chandra M. Voiding and its disorders in children. In: Trachtman H, Gauthier B (eds). Pediatric Nephrology. Australia: Harwood Academic Publishers; 1998: 217–229.
5. Herz D, Weiser A, Collette T, et al. Dysfunctional elimination syndrome as an etiology of idiopathic urethritis in childhood. J Urol 2005; 173: 2132-2137.
6. Docimo SG, Silver RI, Gonzalez R, et al. Idiopathic anterior urethritis in prepubertal and pubertal boys: pathology and clues to etiology. Urology 1998; 51: 99-102.
7. Glahn BE. Giggle incontinence (enuresis risoria). A study and an aetiological hypothesis. Br J Urol 1979; 51: 363-366.
8. MacKeith RC. Micturition induced by giggling? Cataplexy. Arch Dis Child 1959; 34: 358.
9. Williams, D. I. Giggle incontinence. Acta Urol Belg 1984; 52: 151-153.
10. Sher PK, Reinberg, Y. Successful treatment of giggle incontinence with methylphenidate. J Urol 1996; 156: 656-658.
11. Sher PK. Successful treatment of giggle incontinence with methylphenidate. Pediatr Neurol 1994; 10: 81.
12. Sekido N, Akaza H. Neurogenic bladder may be a side effect of focus resection in an epileptic patient. Int J Urol 1997; 4: 101-103.
13. Romme JJ, van Dijk N, Boer KR, et al. Influence of age and gender on the occurrence and presentation of reflex syncope. Clin Auton Res 2008; 18: 127-133.