JCSM
Journal of Clinical Sleep Medicine
http://dx.doi.org/10.5664/jcsm.2424

SLEEP MEDICINE PEARLS

Nocturnal Paroxysmal Events in an 8-Year-Old Girl

Lourdes M. DelRosso, M.D.; Lei Wang, M.D.; Romy Hoque, M.D.
Division of Sleep Medicine, Department of Neurology, Louisiana State University School of Medicine, Shreveport, LA

An 8-year-old girl with normal neurological development presented with nocturnal arm and leg jerking with occasional hyperventilation starting approximately 10 minutes after going to bed. The events started 6 months prior to presentation, occurred 2-3 times per week, and lasted a few minutes. Immediately after each event the patient was able to communicate normally with her mother. She denied recollection of the event. Since the start of the episodes the patient's academic performance has declined.

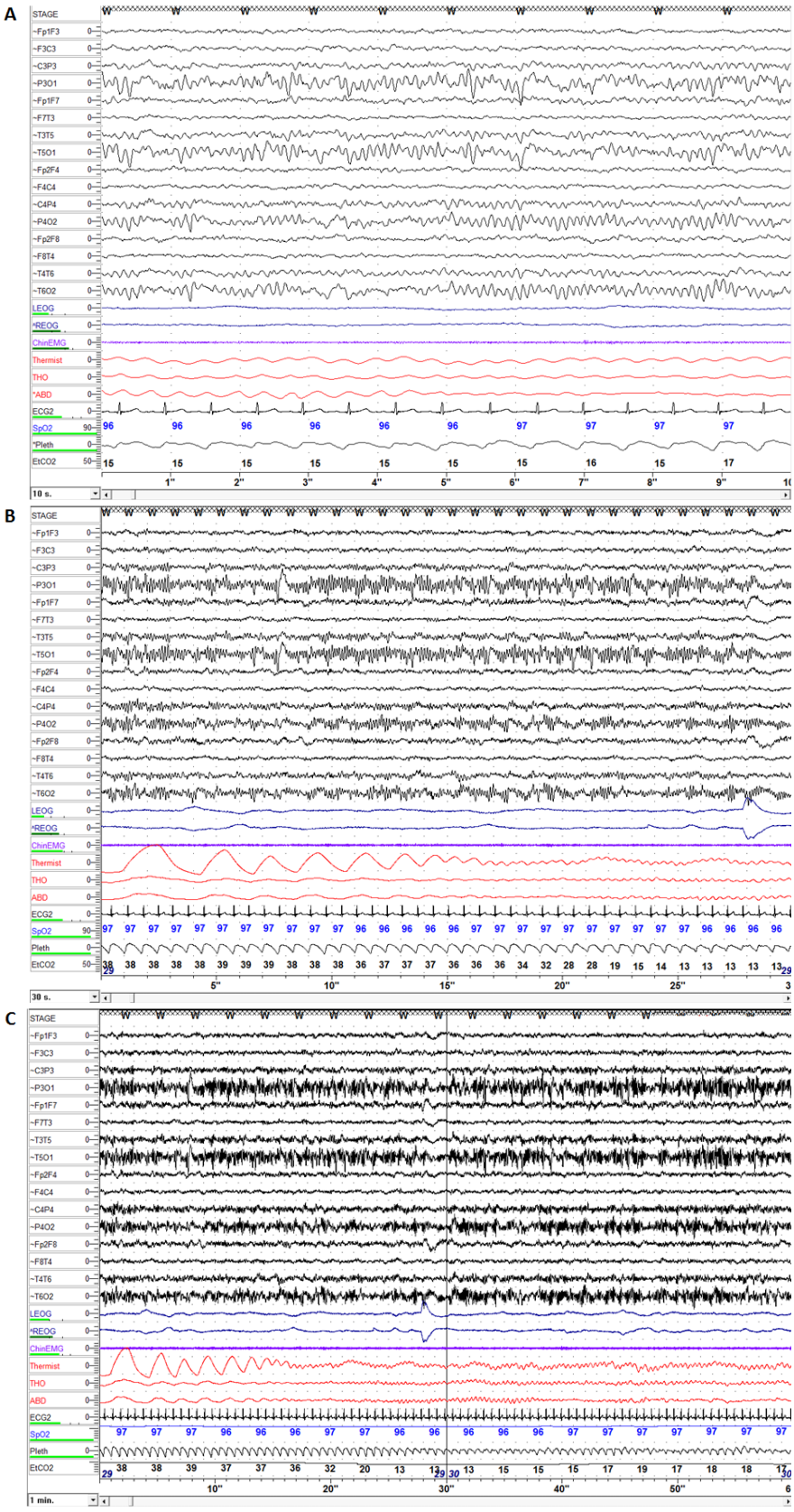
The patient has no family history of neurological disease. She was born via cesarean section at 36 weeks gestational age and had a prolonged stay in the neonatal intensive care unit due to respiratory distress. She lives with both parents and two siblings and has never slept in her own bed. She co-sleeps with both parents.

Prior to presentation the patient underwent the following tests, which were all normal: chest x-ray, pulmonary function tests, bronchoscopy, 24-h Holter monitor, magnetic resonance imaging of the head, and overnight polysomnogram (PSG). The following serum laboratory analysis were normal: basic metabolic panel, liver function tests, complete blood count, vitamin B12, thyroid stimulating hormone, folate, vitamin D, and lead.

Examination showed an active, cooperative girl with normal affect whose growth was appropriate for age. Physical and neurologic examinations were unremarkable without signs of physical abuse or sexual abuse. The mother recorded a home video of a typical nocturnal event (**Video 1**). The patient was admitted and evaluated by a multidisciplinary team (psychiatry, pediatrics, sleep medicine, and neurology). An extensive psychiatric evaluation including multiple interviews with the patient and her immediate family did not reveal depression, anxiety, psychosis, or personality disorders. The only identified stressor was school-related difficulties. We repeated a full diagnostic PSG with respiratory channels and full head electroencephalogram (EEG) montage with a single typical event captured during wakefulness (**Figure 1**): 24-h continuous video-EEG monitoring captured 3 more typical nocturnal events during wakefulness without change in the waking EEG background. Neither interictal epileptiform discharges nor background asymmetries were noted.

QUESTION
Given the history and accompanying video what is the most likely diagnosis?

Figure 1—Diagnostic polysomnogram with full head electroencephalogram (EEG) montage during a typical nocturnal event



(A) 10-sec epoch showing EEG wakefulness during the event. Note the persistent 9-10 Hz posterior predominant rhythm in the occipital derivations bilaterally. Also note the rapid shallow breathing associated with event shown in the thermistor, thoracic and abdominal channels. Hyperventilation episodes were corroborated with review of the accompanying video. **(B)** 30-sec epoch showing typical event arising from EEG wakefulness. **(C)** 1-min epoch showing the prolonged event with persistent EEG wakefulness throughout.

ANSWER: Non-epileptic seizures.

DISCUSSION

Non-epileptic seizures (NES) are seizure-like episodes without evidence of electrographic seizures. They are not uncommon in the pediatric population, are found in up to 38% of pediatric patients admitted for continuous video EEG, monitoring and are more common in girls.[1]

The most common presentation in children younger than 13 years of age is staring episodes. In children older than 13 years of age, the most common feature is jerking of the extremities. A history of stressors can be identified in the majority of patients, with the most common stressors being school difficulties, family problems, and/or interpersonal conflicts with teachers or friends.[2]

The differential diagnosis for this patient includes epileptic paroxysmal events that occur out of sleep in children, such as frontal lobe epilepsy (FLE), juvenile myoclonic epilepsy (JME), benign epilepsy with centrotemporal spikes (BECTS), and infantile spasms. FLE is more likely to occur during sleep than other seizure types. Only 30% of children with FLE have events during the day. FLE has a wide spectrum of manifestations ranging from brief nocturnal arousals to complex motor behaviors (e.g., ambulation, violent outbursts, rhythmic twisting/turning of the trunk/pelvis, bipedal automatisms, dystonic posturing). Some clinical features that are suggestive of FLE include: stereotyped movements, multiple ictal events across the night, and absent or mild postictal confusion.[3] JME commonly presents during adolescence and is characterized by myoclonic jerks occurring shortly after awakening or in early evening. BECTS presents between 3-13 years of age with sleep-related seizures. The seizures are either focal motor (involving the face and arm), oropharyngeal sensorimotor phenomenon (hypersalivation, guttural sounds, dysarthria, abnormal mouth movements, or abnormal mouth/tongue sensations), or generalized tonic-clonic in nature. Infantile spasms have a usual age of onset between 3 months and 1 year.[4]

The diagnosis of non-epileptic seizures is challenging even though history and interview can correctly characterize up to 86% of patients, video EEG, (the gold standard)[5] shows events arising from wakefulness without change in the background EEG.

Patients with pseudoseizures benefit from prompt diagnosis followed by psychotherapy with a close interaction between patient, family members, and the patient's health care providers.[6]

In our case, the patient expressed a desire to sleep in her own room. After communicating the diagnosis to the parents and the child, we instituted the following 4-week extinction therapy protocol for the patient to sleep in her own room. During the first week the mother slept in a recliner in the patient's room. During the second week, the mother was allowed 2 scheduled checks on the patient across the night, one at 10:00 pm and one at 02:00. During the third week the mother was allowed only one scheduled check at 10:00. From the fourth week onward, the child was allowed to sleep in her own room alone with no more check ups. Following successful completion of the extinction protocol, the patient has not had recurrence of her events. The authors cannot definitively rule out placebo effect from our therapy on possible FLE. The patient will have close follow-up with psychology and pediatrics.

CLINICAL PEARLS

1. NES are not uncommon in the pediatric population, found in up to 38% of pediatric patients admitted for continuous video electroencephalography monitoring.
2. The differential diagnosis for NES in children includes FLE, JME, BECTS, and infantile spasms.
3. A history of stressors can be identified in the majority of patients, with the most common stressors being school difficulties, family problems and/or interpersonal conflicts with teachers or friends
4. Institution of an extinction protocol was helpful in this particular patient.

CITATION

DelRosso LM; Wang L; Hoque R. Nocturnal paroxysmal events in an 8-year-old girl. *J Clin Sleep Med* 2013;9(2):178-180.

REFERENCES

1. Kutluay E, Selwa L, Minecan D, Edwards J, Beydoun A. Nonepileptic paroxysmal events in a pediatric population. *Epilepsy Beh* 2010;17:272-5.
2. Patel H, Scott E, Dunn D, Garg B. Nonepileptic seizures in children. *Epilepsia* 2007;48:2086-92.
3. Tomonoh Y, Yasumoto S, Ihara Y, et al. Diagnosing nocturnal frontal lobe epilepsy: a case study of two children. *Seizure* 2011;20:583-5.
4. Vendrame M, Kothare SV. Epileptic and nonepileptic paroxysmal events out of sleep in children. *J Clin Neurophysiol* 2011;28:111-9.
5. Reuber M, Mayor R. Recent progress in the understanding and treatment of nonepileptic seizures. *Curr Opin Psychiatry* 2012;25:244-50.
6. LaFrance WC Jr., Devinsky O. The treatment of nonepileptic seizures: historical perspectives and future directions. *Epilepsia* 2004;45 Suppl 2:15-21.

SUBMISSION & CORRESPONDENCE INFORMATION

Submitted for publication June, 2012
Submitted in final revised form July, 2012
Accepted for publication July, 2012
Address correspondence to: Lourdes DelRosso, M.D., Division of Sleep Medicine, Department of Neurology, Louisiana State University School of Medicine, 1501 Kings Highway, Shreveport, Louisiana 71103; E-mail: lordydel@yahoo.com

DISCLOSURE STATEMENT

This case was presented in part by Dr. Romy Hoque at the SLEEP 2012 meeting in Boston, Massachusetts. The authors have no conflict of interest or financial support to declare.