SLEEP MEDICINE PEARLS

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A 14-Year-Old Girl with Excessive Daytime Sleepiness and Facial Twitching

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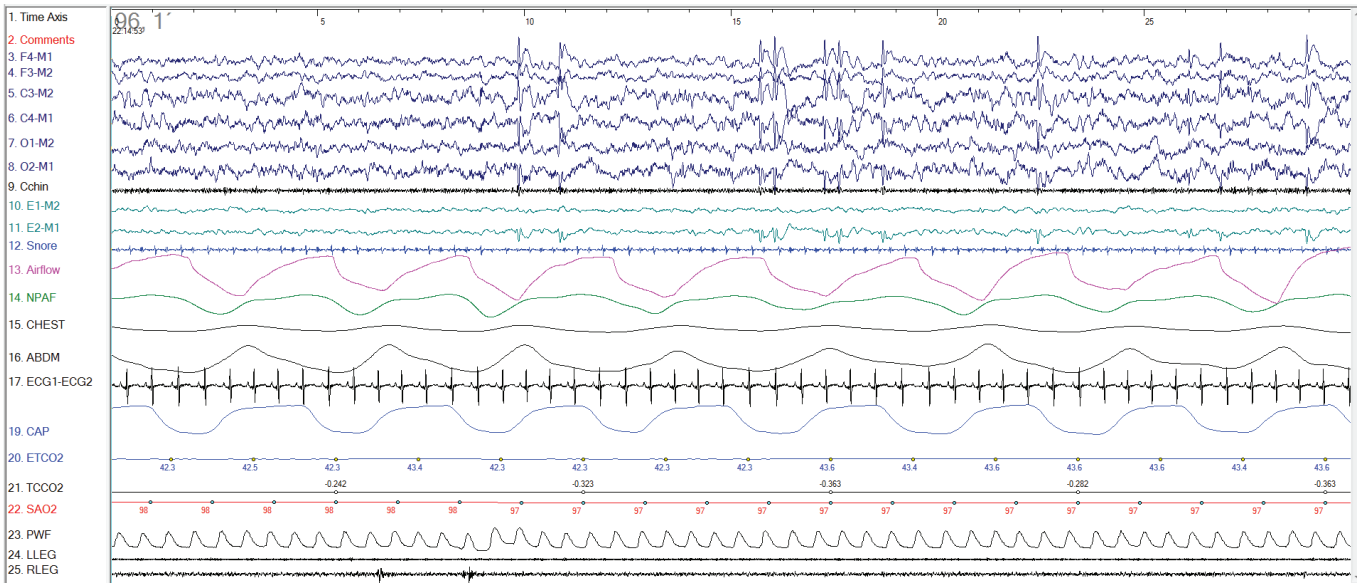
A 14 year-old girl, with history of asthma and tonsillectomy/adenoidectomy, presents for second-opinion evaluation of sleepiness starting at the age of 13 that was diagnosed as idiopathic hypersomnia by polysomnography (PSG) and multiple sleep latency test (MSLT) performed at another institution. The PSG revealed total sleep time (TST) 335 minutes, sleep efficiency 69%, apnea hypopnea index 0 events/hour of sleep, and REM sleep latency 4 min 43 sec. MSLT performed the next morning showed no sleep onset REM periods (SOREMPs); average sleep latency across 5 naps was 7 min 13 sec. She was diagnosed with idiopathic hypersomnia and prescribed methylphenidate. The parents did not want to start the medication until further evaluation.

The patient goes to bed around 21:00, awakens multiple times during the night, and arises in the morning at 05:00 on weekdays and 07:00 on weekends. During vacation periods from school, she typically sleeps from 22:00 until 09:00. She denies history of witnessed snoring/apnea, seizures, dream-enacting behavior, or cataplexy. During the day, she struggles to stay awake and tends to doze when sitting quietly. A modified Epworth Sleepiness Scale score without question 8 ("Stopped for a few minutes in traffic while driving") is 9/21. She does not take any medication. On physical examination, her oral airway revealed a Mallampati score of II with no tonsils appreciated. Otherwise, the neurological examination was unremarkable.

Repeat PSG showed sleep latency 50 min, sleep efficiency 84%, TST 483 min, REM sleep onset 90 min, and centrally located sharply contoured waves during wake and sleep. MSLT across 4 naps showed average sleep latency of 17 min 8 sec, with no SOREMPs noted. Upon further interview the mother revealed that the patient occasionally exhibits facial twitches during sleep. The patient was admitted to the epilepsy monitoring unit (EMU) for evaluation. **Figure 1** shows a discharge captured during PSG.

QUESTION: What is the child's diagnosis?

Figure 1—30-second epoch showing epileptic discharge.



ANSWER: Benign epilepsy of childhood with centro-temporal spikes (BECTS)

DISCUSSION

Benign epilepsy of childhood with centro-temporal spikes (BECTS) is a common pediatric epilepsy syndrome, with onset between 3 and 13 years, usually resolving by age 16. The classic seizures of BECTS occur shortly after falling asleep or upon awakening, are brief in duration lasting seconds to a few minutes, are rarely followed by postictal confusion or amnesia, and have semiology consisting of: unilateral numbness/paresthesias or clonic/tonic activity of face, lips or tongue; dysarthria; and drooling. Seizures in BECTS may be associated with excessive daytime sleepiness and subjectively poor sleep quality with longer latency to sleep.[1] Children with BECTs achieve remission regardless of anti-epileptic drug (AED) therapy, and prognosis is excellent even in those with frequent, troublesome seizures. If the events are sufficiently disturbing to either the parent or child, AEDs may be initiated.

The characteristic EEG manifestations are high amplitude spikes or sharps in an otherwise normal background with maximal negativity in the centro-temporal derivations, frequently cluster, and are markedly activated in sleep. Other features that may be present include prominent after-going slow waves or a horizontal dipole.[2]

In EEG recordings the vast majority of spikes are radial dipoles, manifesting as scalp negativity with no concurrent scalp positivity measured. In contrast, the classic horizontal dipole of BECTS consists of negativity in the centro-temporal areas, with concurrent positivity recorded from the frontal areas. The horizontal dipole is best seen on a referential montage with the centro-temporal component manifesting as an upgoing (negative) deflection, and the frontal component manifesting as a downgoing (positive) deflection. Our patient's EEG is a bit atypical, demonstrating an upgoing deflection in the left central derivation, along with upgoing deflections in the frontal derivations. Though apiculated, vertex waves of stage 1 sleep tend to be of longer duration than typical sharp waves of BECTS, but occasionally distinction may be difficult. On 24-h EEG of children with typical BECTS, 21% had a focus outside the centro-temporal area, and half lacked a horizontal dipole.[3]

BECTS is an electro-clinical syndrome in that EEG findings alone are insufficient to establish diagnosis. Typical centro-temporal discharges are seen in 0.7% of awake recordings in normal children without a history of seizures and are probably higher in sleep recordings.[4] The percentage of children with centro-temporal sharp waves who develop clinically apparent seizures is probably less than 10%, thereby making such discharges likely an incidental finding in those without spells or those whose spell semiology is not suggestive of BECTS.[5]

The diagnosis of idiopathic hypersomnia in this patient was given after an MSLT administered after 5.5 hours of sleep on prior PSG. Six hours is the minimum recommended night sleep duration for a valid MSLT. After sleep extension (with TST of 8 h on follow-up PSG), the objective sleepiness measured by the MSLT improved significantly to a mean latency of 17 minutes. However, subjective sleepiness remained.

The symptoms of facial twitching with electroencephalographic correlate were consistent with BECTS. It is the author's opinion that BECTS contributed to this patient's subjective daytime sleepiness. Treatment with diazepam 1 mg was initiated at bedtime with resolution of nocturnal awakening and daytime sleepiness.

SLEEP MEDICINE PEARLS

1. The classic seizures of BECTS occur shortly after falling asleep or upon awakening.
2. Seizures in BECTS may be associated with excessive daytime sleepiness and subjectively poor sleep quality with longer latency to sleep.
3. The characteristic EEG manifestations are high amplitude spikes or sharps in an otherwise normal background with maximal negativity in the centro-temporal derivations, frequently cluster, and are markedly activated in sleep.
4. 50% of those with typical BECTS may lack the classic horizontal dipole.

CITATION

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