AMERICAN ACADEMY OF NEUROLOGY

Update: Medical Treatment of Infantile Spasms

Case Presentation Part I: Emergency Room Consultation

A 9-month-old boy presents to the emergency room with abnormal movements, and a neurology consult is requested. His mother had recently noticed abnormal movements such that he will have brief forward flexion of the head. This can be accompanied by either flexion or extension of both arms, brief eye-rolling, and a cry. She initially noticed this a few weeks back but did not think it was anything to worry about. However, in the last week the episodes have become more frequent and have even occurred multiple times in an hour. She called her pediatrician on the phone, and colic was suggested because of the cry. However, because of the increased frequency and severity of the events and decreased visual alertness she decided to bring the child to the emergency room. She has not noticed any prolonged tonic-clonic movements or skin color changes. She can not identify any clear triggers for these episodes. She notes that he has not met some of his developmental milestones.

His past medical history reveals that he was the product of a full-term pregnancy via vaginal delivery. His mother thinks his Apgar scores were normal. After the delivery, the mother and child were discharged after the routine hospital stay. The child has not been hospitalized since. He is up to date on his vaccinations.

The boy does not take any medications and does not have any known medication allergies.

There is no family history of neurologic illness, including seizures, tuberous sclerosis complex (TSC), or developmental problems.

The boy lives with his mother and father. There are no other children or pets in the home. Nobody in the house smokes.

A complete 14-topic review of systems was obtained and was positive only for these movements.

On physical examination, he is a well-nourished boy in no acute distress. He is afebrile. His blood pressure is 110/70, pulse is 88, and respiratory rate is 14.

His growth parameters, including length, weight, and head circumference, were at the 50th percentile for his age. There were no facial dysmorphisms.

Examination of his skin with a Wood’s lamp reveals no café au lait spots. He does not have a shagreen patch.

On neurologic examination, the boy has evidence of developmental delay. He babbles but does not say “mama” or “dada.” He cannot sit by himself. He does not pass objects from hand to hand. He did not appear as visually alert for age as he should have been.

Cranial nerve testing reveals PERRLA; optic discs are sharp. Extraocular muscles are intact as the boy tracks you in all directions. Facial strength is normal.

On motor examination, he has normal tone. He moves his arms and legs in a symmetric fashion.

Deep tendon reflexes are 2/4 throughout. Plantar responses are extensor bilaterally.

While examining the boy, you are able to witness one of his spells. His head flexes forward, his eyes roll up, and his arms extend. The spell starts as a fast phase that lasts 2 seconds and is followed by 4 to 5 seconds of his remaining in this position. He then returns to normal posture and starts to cry.

Basic labs from the emergency room reveal normal electrolytes.

You discuss with the boy's mother that he is having recurrent seizures that are concerning for infantile spasms. As earlier treatment has been associated with better outcomes, you recommend a hospital admission for an expedited workup, including EEG, brain MRI, and various serum and urine tests.

Case Presentation Part II: Hospital Follow-up

The boy is admitted and is overall the same when you return to visit with him. You discuss with the boy’s mother that his brain MRI was unremarkable. His urine organic acids, quantitative serum amino acids, TORCH infection titers, and lead level are normal. His EEG showed hypsarrhythmia.

You counsel his mother that this testing confirms your clinical suspicion that the boy is having infantile spasms. You discuss that there is evidence that early treatment is associated with better developmental outcomes. You counsel her that according to the AAN’s “Evidence-based Guideline Update: Medical Treatment of Infantile Spasms,”[1] treatment with the medication adrenocorticotropic hormone (ACTH) offers better short-term resolution of spasms and long-term developmental outcomes. You discuss possible side effects of this medication, including cerebral atrophy, electrolyte disturbances, and hypertension. After this discussion, she agrees to this treatment. He is started on ACTH 20 IU daily. She understands that studies have shown similar efficacy between low- and high-dose ACTH regimes. You advise that he should receive the first few doses while in the hospital so that he may be monitored for adverse effects and therapeutic response. A visiting nurse is arranged to give the intramuscular injections at home. The boy will also have an outpatient neurology clinic visit scheduled for 1 to 2 weeks after discharge, at which time he will have another EEG to look for improvement on medication.

Questions

1. For short-term treatment of infantile spasms, which of the following statements is correct?
A. Vigabatrin (VGB) is better than ACTH
B. Prednisolone is better than VGB
C. Either ACTH or VGB can be considered
D. Low-dose ACTH is better than high-dose ACTH
E. None of the above

The correct answer is C. VGB (Level C) or ACTH (Level B) can be considered although ACTH is more effective than VGB (Level C) in the short-term treatment of children with infantile spasms, excluding those with tuberous sclerosis complex). There is insufficient evidence to determine whether other forms of corticosteroids are as effective as ACTH (Level U). Low dose ACTH is probably as effective as high-dose ACTH (Level B).

2. Which statement is correct with regard to long-term outcomes?
A. Shorter lag time to treatment does not improve long-term developmental outcomes
B. Shorter lag time to treatment improves long-term seizure outcomes
C. Shorter lag time to treatment improves both long-term developmental and seizure outcomes
D. Successful short-term treatment of cryptogenic and symptomatic infantile spasms with ACTH or prednisolone leads to better long-term developmental outcomes than treatment with VGB
E. None of the above

The correct answer is E. Short lag time to treatment of infantile spasms with either hormonal therapy or VGB may be considered to improve long-term cognitive outcomes (Level C). Hormonal therapy with ACTH or prednisolone may be considered for use in preference to VGB in infants with cryptogenic infantile spasms to possibly improve developmental outcome (Level C).

3. Which of the following statements are incorrect regarding VGB?
A. It is a medication that is given intramuscularly
B. Retinal toxicity in infants cannot be tested
C. It was used in the United Kingdom Infantile Spasms Study (UKISS) study for patients with TSC
D. All of the above
E. Only A and C

The correct answer is D. VGB is an oral medication. Patients with TSC were excluded from the UKISS study. ERG 30-Hz flicker amplitude has proven to be a useful tool in predicting retinal toxicity in infants treated with VGB.[2,3]

Diagnosis Coding[4,5]

In the United States ICD-9-CM is the classification system used for diagnosis coding. ICD-10-CM is planned to replace ICD-9-CM, but the exact implementation date is uncertain at the time of this writing.

In Part I the stated known diagnosis is recurrent seizures and the stated suspected diagnosis is infantile spasms. The ICD-9-CM and ICD-10-CM code choices are for the current known diagnosis or symptoms. For recurrent seizures the codes for the billing claim for that day are:

ICD-9-CM

345.8 Other forms of epilepsy and recurrent seizures

Recurrent seizures, not otherwise specified

ICD-10-CM

G40.909 Epilepsy, unspecified, not intractable, without status epilepticus

Recurrent seizures, not otherwise specified

These are also the codes to be used to request the testing ordered. The suspected, but not confirmed, diagnosis is not used to request testing unless specifically required by the third-party payer (and never for Medicare).

There is one additional neurologic diagnosis possible, though it must also be included in the final diagnostic statement in the patient record. That diagnosis is developmental delay. The codes for this are:

ICD-9-CM

783.42 Delayed milestones

ICD-10-CM

R62.0 **Delayed milestone in childhood**

In Part II, the definite diagnosis is infantile spasms. The codes for the billing claim for this day are:

ICD-9-CM

345.60 Infantile spasms without mention of intractability

ICD-10-CM

G40.822 Epileptic spasms, not intractable without status epilepticus

Procedure Coding

Electroencephalography

Routine Electroencephalography (EEG)

EEG codes 95812-95822 include hyperventilation and/or photic stimulation when appropriate. Routine EEG codes 95816-95822 include 20 to 40 minutes of recording. Extended EEG codes 95812-95813 include reporting times longer than 40 minutes.

95812 Electroencephalogram (EEG) extended monitoring; 41-60 minutes

95813 greater than one hour

95816 Electroencephalogram (EEG); including recording awake and drowsy

95819 including recording awake and asleep

95822 recording in coma or sleep only

Electroencephalography: Coding Tips

Hyperventilation and photic stimulation procedures are not a mandatory part of EEG testing using codes 95812-95822. They are to be performed only when medically appropriate and not otherwise contraindicated. Hyperventilation and photic stimulation are bundled into the EEG service whenever they are performed – they cannot be coded separately.

95819 is not any routine EEG, it is for a planned awake/asleep study with or without sedation.

Code **95819** if an awake/asleep study was intended even if patient did not sleep.

Use **95816** if an awake only study is planned. However, one may upcode to **95819** if the patient falls asleep and the recording time is sufficient.

95822 ("coma and sleep" EEG) can be used for patients that are:

- Anesthetized
- Neonates

Evaluation and Management Coding

The patient is seen in consultation in the emergency room. The history is comprehensive, but the examination is only detailed. Thus the consultation would be coded as a level 3 consult or 99243. The follow-up evaluation includes a limited history and no examination. It would be best to code using time. If one assumes that 25 minutes was spent with the patient and about 20 minutes was spent in counseling and coordination of

care, the follow-up evaluations would be coded as a level 2 subsequent hospital day or 99232.

1. Go CY, Mackay MT, Weiss SK, et al. Evidence-based guideline update Medical treatment of infantile spasms Report of the Guideline Development Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. *Neurology* 2012;78:1974–1980.
2. Durbin S, Mirabella G, Buncic JR, Westall CA. Reduced grating acuity associated with retinal toxicity in children with infantile spasms on vigabatrin therapy. Investigative Ophthalmology and Visual Science 2009;50:4011–4016.
3. Westall CA, Nobile R, Morong S, Buncic JR, Logan WJ, Panton CM. Changes in the electroretinogram resulting from discontinuation of vigabatrin in children. Doc Ophthalmol 2003;107:299–309.
4. Centers for Disease Control and Prevention. International classification of diseases, ninth revision, clinical modification (ICD-9-CM). *www.cdc.gov/nchs/icd/icd9cm.htm.*
5. Centers for Disease Control and Prevention. International classification of diseases, tenth revision, clinical modification (ICD-10-CM). *www.cdc.gov/nchs/icd/icd10cm.htm.*

Disclaimer
This statement is provided as an educational service of the American Academy of Neurology. It is based on an assessment of current scientific and clinical information. It is not intended to include all possible proper methods of care for a particular neurologic problem or all legitimate criteria for choosing to use a specific procedure. Neither is it intended to exclude any reasonable alternative methodologies. The AAN recognizes that specific patient care decisions are the prerogative of the patient and the physician caring for the patient, based on all of the circumstances involved.