

Case Presentation: Pediatric Neurology Clinic

A 13-year-old boy presents to the pediatric neurology clinic, accompanied by his mother, to discuss vagus nerve stimulation (VNS) as a potential treatment for epilepsy. The boy's mother provides the majority of his medical history. At 9 years of age, he developed bacterial meningitis. Approximately 1 year later, he was diagnosed with epilepsy after he presented with generalized tonic-clonic seizures. Since that time, the patient has been treated with multiple antiepileptic medications that have been ineffective or poorly tolerated. On his current seizure medication regime of sertraline, oxcarbazepine, levetiracetam, and lamotrigine, his seizure frequency has decreased from daily to once weekly. His mother would like to discuss VNS as a treatment option for him. She read about VNS on the internet as a potential treatment for epilepsy.

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, mother and son were discharged after a routine hospital stay. The boy was not hospitalized until his bacterial meningitis infection at age 9 years, after which time he has suffered chronically from refractory epilepsy and depression. His vaccinations are up-to-date.

Previous to his current medication regimen, the patient had tried phenytoin, topiramate, carbamazepine, valproic acid, and zonisamide, each of which was either ineffective or poorly tolerated.

There is no family history of neurologic illness, including seizures, tuberous sclerosis, 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 tobacco. A complete 14-topic review of systems was obtained and was positive only for what is described in the history of present illness.

On physical examination, he is a well-nourished boy in no acute distress. He is afebrile. His blood pressure is 105/70, pulse is 88, and respiratory rate is 14.

On neurologic examination, the boy makes poor eye contact. However, when prompted, he does follow commands and repeats and names appropriately.

Cranial nerve testing reveals PERRLA; optic discs are sharp. Extraocular muscles are intact. Facial strength and sensation are normal. His tongue is midline.

On motor examination, he has normal tone. He moves his arms and legs with normal power. Deep tendon reflexes are 2/4 throughout. Plantar responses are flexor bilaterally.

Coordination is normal on finger–nose–finger and heel–knee–shin testing bilaterally. His gait is narrow-based and steady.

The patient has had a brain MRI and long-term EEG monitoring which failed to identify a surgically amenable lesion. The long-term EEG monitoring did not reveal nonepileptic seizures.

You discuss with the boy and his mother that his epilepsy has been refractory to multiple seizure medications. Furthermore, extensive testing has not revealed that he is a candidate for epilepsy surgery. In this circumstance, he would be a candidate for VNS therapy. This would be an adjunctive treatment to his seizure medications that, according to an American Academy of Neurology guideline, might help reduce his seizure frequency.¹ VNS may have the additional effect of helping his depression, as this has been seen in adults.¹ You explain how VNS is thought to reduce seizure frequency. You discuss both short-term side effects, such as infection, and long-term side effects, including exercise intolerance, cough, and hoarseness.

After their questions are answered, the boy and his mother decide to pursue VNS therapy, and a referral to neurosurgery is made. You arrange for a neurologic follow-up after his procedure so you can monitor for side effects, adjust VNS settings, and teach mother and son how to use the magnet. You advise that he should continue his current seizure medication regimen.

Questions

1. This young patient is a candidate for VNS therapy because:
 - A. Multiple antiepileptic medications have failed to be of help
 - B. He has the appropriate seizure type
 - C. He is not a cortical surgery candidate
 - D. All of the above

The correct answer is D.

2. Children:
 - A. May have different adverse effect profiles with VNS therapy than adults
 - B. Respond better to VNS therapy than adults
 - C. Should receive VNS therapy even if there are sleep apnea problems
 - D. Should always receive VNS therapy before a surgery evaluation

The correct answer is A.

3. VNS therapy may treat seizures and syndromes such as:
 - A. Partial-onset seizures
 - B. Lennox-Gastaut syndrome
 - C. Dravet syndrome
 - D. All of the above

The correct answer is D.

Diagnosis Coding

In this case study, the only stated diagnosis is “epilepsy.” In the history it is mentioned that the patient presented with generalized tonic–clonic seizures, that the epilepsy is refractory, and that it occurred after bacterial meningitis. It is presumed that in an actual patient record, the type of epilepsy would be named as a diagnostic statement. For the purposes of this coding analysis the assumption is made that the diagnostic statement in the record is generalized epilepsy, that it is refractory, and that it is due to childhood bacterial meningitis.

In ICD-9-CM² and ICD-10-CM,³ diagnosis coding for epilepsy asks for the seizure localization or syndrome and whether the epilepsy is refractory (intractable). In ICD-9-CM,² the presence of status epilepticus is a separate code. In ICD-10-CM³ this information is incorporated into each epilepsy code. In this case, another code is added for the cause of this syndrome.

The ICD-9-CM² code for this patient visit is:

345.11 Generalized convulsive epilepsy, with intractable epilepsy
326 Late effects of intracranial abscess or pyogenic infection

The ICD-10-CM³ code for this patient visit is:

G40.319 Generalized idiopathic epilepsy and epileptic syndromes,
intractable, without status epilepticus
G09 Sequelae of inflammatory diseases of the central nervous system

In summary, when coding for epilepsy diagnoses, one should *document* and provide answers to these questions:

1. What is the type, syndrome, or localization?
2. Is the epilepsy refractory?⁴
3. Is there status epilepticus?
4. Is the epilepsy a sequela to a previous injury or illness?

These questions are all easily answered for a patient such as the one presented here and also easily placed in a diagnostic statement such as a problem list. It is especially important to document the intractable (refractory) aspect because the patient may not get approval for the VNS without that information.

In addition to the questions for diagnosis coding, take these additional criteria into account with regard to AAN quality measures,⁴ to improve the likelihood of approval for VNS:

1. Consideration for surgical therapies for intractable epilepsy
2. Documentation of EEG and MRI results if appropriate
3. Query and counseling about antiepileptic medication side effects and specific safety issues
4. Counseling for women of childbearing age

Procedural Coding

The initial clinic visit could be a Consultation code, CPT® 99243-99245, depending on the patient's insurance and whether the child was referred for the visit. The notes above lack the exam details to qualify for a moderate-level 99244 or high-level 99245 visit. The moderate- or high-level service could be met best if signs from eight organ systems were documented in the note, or if a 23-element neurologic single-system exam were documented.

The VNS programming can be coded in one of three ways depending on how much programming was performed and how long the programming took:

95971, programming fewer than four parameters

95974 with modifier 52, programming four or more parameters, 30 minutes or less

95974, programming four or more parameters, more than 30 minutes

CPT © 2010 American Medical Association. All rights reserved. CPT is a registered trademark of the American Medical Association.

1. Morris GL III, Gloss D, Buchhalter J, Mack KJ, Nickels K, Harden C. Evidence-based guideline update: Vagus nerve stimulation for the treatment of epilepsy. Report of the Guideline Development Subcommittee of the American Academy of Neurology. *Neurology*® [Publ online ahead of print August 28, 2013].
2. Centers for Disease Control and Prevention. International classification of diseases, ninth revision, clinical modification (ICD-9-CM). www.cdc.gov/nchs/icd/icd9cm.htm.
3. Centers for Disease Control and Prevention. International classification of diseases, tenth revision, clinical modification (ICD-10-CM). www.cdc.gov/nchs/icd/icd10cm.htm.
4. Fountain NB, Van Ness PC, Swain-Eng R, Tonn S, Bever CT Jr; for the American Academy of Neurology Epilepsy Measure Development Panel and the American Medical Association–Convened Physician Consortium for Performance Improvement Independent Measure Development Process. Quality improvement in neurology: AAN epilepsy quality measures. Report of the Quality Measurement and Reporting Subcommittee of the American Academy of Neurology. *Neurology*® 2011;76:94–99.

The AAN develops these clinical case examples as educational tools for neurologists and other health care practitioners. You may download and retain a single copy for your personal use. Please contact guidelines@aan.com to learn about options for sharing this content beyond your personal use.

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.

© 2013 American Academy of Neurology