CHAPTER 18

STARING SPELLS

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Staring spells are a frequent reason for referral to pediatricians and pediatric neurologists. Teachers often fear the presence of seizures and encourage parents to seek specialist advice for their child’s episodes of inattention. As discussed in this chapter, these children rarely have epilepsy.

Differential Diagnosis
The differential diagnosis of staring spells is usually limited to the following conditions:

• Daydreaming (inattention)
• Absence seizures
• Complex partial seizures

For children with staring spells, the physician’s first responsibility is to differentiate daydreaming or inattentive staring spells from seizures. This distinction can usually be easily established by determining the characteristics of a child’s spells.

Unresponsiveness owing to inattention (daydreaming) usually occurs during periods of inactivity or when a child finds activities less interesting. Daydreaming almost never occurs when a child is physically active. In contrast, seizures occur at any time and may interrupt play. Children who are daydreaming may “ignore” verbal stimulation, such as calling their names, or arm waving, but usually quickly respond to tactile stimulation, such as touching or tickling (ie, “the touch test”). Children with absence seizures or complex partial seizures will not respond to the touch test. As discussed in this chapter, other clinical features and investigations are specific to each of these conditions. Clinically, however, establishing whether staring spells interrupt play activities in combination with the “touch test” are excellent methods of determining which children require further evaluation.

Inattentive Staring Spells
(Daydreaming)
All children daydream. Those whose daydreaming leads to concern among their parents or educators are more likely to have comorbid difficulties. Children with attention-deficit hyperactivity disorder (ADHD) spend more time off task and are therefore more likely to be referred. Similarly, children with learning difficulties (LD) may have difficulty maintaining attention during tasks they find particularly challenging. Both ADHD and LD are more common in children with Tourette’s syndrome (TS), as are obsessive–compulsive behaviors. Children with TS, therefore, often have periods of daydreaming in the classroom, as ADHD and LD may make schoolwork less compelling than their obsessive thoughts. Staring spells in the context of a child with TS are more likely to concern a teacher, as the associated tics may be misinterpreted as automatisms. These spells should not prove diagnostically difficult as they are easily terminated with the “touch test,” and the tics can be briefly interrupted on request.

Children with developmental delay or other neurodevelopmental difficulties may have staring spells with hand-flapping or other self-stimulatory behavior. It may prove more difficult to interrupt these children, but with effort their attention can usually be regained.

A particularly challenging group of patients are young female toddlers whose parents report staring spells associated with rhythmic pelvic and trunk movements. These spells invariably occur while the child is sitting or lying and represent self-stimulation (masturbation). The diagnosis is easier to establish if the spells are videotaped and if the parents demonstrate that the spells can be interrupted. Surprisingly, many parents are distressed by this diagnosis.

Another group of children who are occasionally referred because of unusual staring spells are children with the “Alice in Wonderland” phenomenon. During these migraine-related events, children experience distortions of time or space. They may perceive that some items in
their surroundings are either increased or decreased in size or distorted in shape. The children are aware that the event is a misperception but may appear to stare, as they are either scared or perplexed by the experience. The events occur in children who have a family history of migraine or who will subsequently develop more typical migraine headaches. The “Alice in Wonderland” events do not coincide with headache. It is important to make a correct diagnosis, as these children require reassurance and do not need further evaluation. The hallmark of all these spells is that they can be interrupted by touch or tickle and that they do not interrupt otherwise interesting activities.

Seizures

Seizures are discussed in Chapters 15–30. For children referred for staring spells, only complex partial and absence seizures need to be considered. Helpful clinical features that distinguish between complex partial and absence seizures are presented in Table 18-1.

Partial seizures are divided into simple partial seizures, where there is no alteration of alertness, and complex partial seizures, where consciousness is altered but not necessarily lost. Among the generalized seizures, only absence seizures present as staring spells. Absence is further subdivided into typical and atypical absence seizures. For those with typical absence, a further subdivision into a number of epilepsy syndromes, based on seizure and electroencephalogram (EEG) characteristics, allows more accurate prediction of the response to therapy and prognosis.

Typical Absence Seizures

Childhood Absence Epilepsy

Childhood absence epilepsy (CAE) seizure (or petit mal seizures) must be differentiated from other absence syndromes to develop appropriate management strategies (Table 18-2). The clinical features of childhood absence seizures are:

- Onset between ages 4 and 10 years
- Normal neurologic and developmental status
- Frequent brief absence seizures with abrupt and significant impairment of consciousness

- EEG discharges of generalized spike-and-wave complexes at 3 Hz lasting 4 to 20 seconds.

Question the diagnosis of CAE if there are any of the following factors:

- Other seizures, such as generalized tonic-clonic seizures (GTCS) or myoclonic seizures before or during the active stage of absences
- Eyelid or perioral myoclonia
- Mild or no impairment of consciousness during the 3 Hz discharges
- Brief EEG 3 Hz to 4 Hz spike-wave discharges of less than 4 seconds or multiple spikes (> 3)
- Visual (photic) or other sensory precipitation of clinical seizures

Seizures in CAE are characterized by brief episodes of altered consciousness, with an abrupt onset and termination. Each attack usually lasts approximately 10 seconds, and patients may have up to 100 seizures per day. During the seizure, the child suddenly stops activity and stares ahead. When the seizure ends, previous activities are resumed as if nothing occurred and the child may even complete an interrupted word. Patients are usually unaware of the seizure.

CAE occurs more frequently in girls (60 to 70%) and a positive family history exists in 15 to 44%. In pediatric studies of twins with CAE, 84% of their monzygotic twins had 3 Hz spike-wave discharges on EEG, and typical absence seizures were found in 75%.

The incidence rate for CAE in children younger than 15 years of age is 6/100,000 to 8/100,000. Community-based studies have determined the prevalence of CAE in children with epilepsy who were younger than 16 years of age to be 10 to 12%.

CAE is primary or idiopathic and therefore children do not require neuroimaging studies. The most prominent precipitant is hyperventilation, and the diagnosis should be questioned if a seizure cannot be provoked by 3 minutes of hyperventilation. Therefore, when absence seizures are suspected, the child should undertake 3 minutes of hyperventilation. This is most easily accomplished by asking the child to blow a tissue or blow and count each breath. The alteration in consciousness may be subtle and the procedure should be videotaped for documentation of clinical features. If a seizure is suspected during this procedure, the physician should say a word, such as a color, which the child is then asked to repeat upon termination of the spell. Those who have had a seizure will not recount the word. The most common error made in reaching a diagnosis is failing to conduct a sufficiently long trial of hyperventilation. Children (and physicians to some degree) require constant encouragement to complete the full 3 minutes, which should be timed.

<table>
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<tr>
<th>TABLE 18-1. Differentiation between Absence and Complex Partial Seizures</th>
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<td>Duration</td>
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<td>Aura</td>
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Juvenile Myoclonic Epilepsy of Janz

Juvenile myoclonic epilepsy of Janz occurs in neurologically normal adolescents who present with early morning GTCS and myoclonic jerks, usually involving the shoulders and upper arms. About one-third of patients with juvenile myoclonic epilepsy have absence seizures, which may be associated with milder impairment of consciousness. The EEG usually shows frequent polyspike discharges.

Atypical Absence Seizures

Atypical absences have a less abrupt onset and ending and are often associated with changes in tone. The seizures usually occur in children with neurologic and developmental deficits and are frequently seen as part of the spectrum of the Lennox-Gastaut syndrome. Seizure frequency often increases with drowsiness and varies from a few a day to almost continuous. The EEG shows generalized spike-and-wave discharge at less than 3 Hz.

Atonic atypical absences may also be seen with GTCS and myoclonic seizures in myoclonic astatic epilepsy, and atypical absences may occur as the only type of seizure in patients with continuous spike-waves in slow sleep. For these patients, valproate and lamotrigine are usually the antiepileptic drugs (AEDs) of choice.

Complex Partial Seizures

Complex partial seizures (CPS) may be preceded by an aura (ie, a simple partial seizure) or may manifest as an alteration of consciousness from the onset. Staring spells associated with CPS usually have a less abrupt onset than those associated with absence. CPS almost always last longer than absence seizures, typically 1 to 2 minutes, although longer seizures can occur. The automatisms that occur in CPS are often more complex than the eyelid fluttering or lip smacking seen in absences. Typical automatisms in CPS include fumbling with clothes, repetitive swallowing or chewing, and purposeless hand or body movements, although lip smacking can occur. As the seizure ends, the child is confused or irritated and may need to sleep. When violence occurs in the context of CPS, it is random and is not meaningfully directed at individuals or objects. CPS may begin at any age and are more likely to reflect underlying structural brain pathology. The distinctions between absence seizures and CPS are outlined in Table 18-1.
Frontal Lobe Absence Seizures
Partial seizures originating in the frontopolar or medial surface of the frontal lobe may produce frontal lobe absence-type seizures, which appear similar to absence seizures with altered consciousness but involve minimal motor activity. Patients may have a complete loss of consciousness or their responses may be slowed and inappropriate. These seizures can be brief or can last hours to days. Postictally, patients may not remember the event, although they may have appeared alert during the seizure. During a seizure, the EEG shows disorganized generalized spike-wave activity.

Evaluation
For children whose spells can be reliably interrupted, attention should be paid to possible comorbid conditions, such as ADHD or LD, with appropriate investigations such as psychological and educational evaluations (Figure 18-1). For almost all these children, the diagnosis is clinical and no further evaluation is required.

All children with suspected seizures should have an EEG. For those with suspected absence seizures, hyperventilation should be part of the protocol. When CPS are suspected, the child should be sleep-deprived on the evening before the EEG. Sleep deprivation may help provoke a seizure, and obtaining an EEG in awake, drowsy, and stage II sleep states increases the chance of recording a focal epileptic discharge.

Routine EEG may fail to record an epileptic discharge, although this is less likely with CAE because of the high frequency of the seizures. When routine EEG is uninformative, prolonged video EEG monitoring may be helpful.

Because CAE is idiopathic, children do not require neuroimaging studies. Atypical absences are secondary or symptomatic and require further investigation. For those with CPS, magnetic resonance imaging should be performed, as underlying pathology may be detected, especially if the seizures all originate from the same focus.

Management
Children with daydreaming spells almost never require intervention. Those with ADHD or LD may require treatment with medication or classroom modifications. For those who have comorbid disorders, such as Tourette’s syndrome, additional interventions may be required. For example, if obsessive thoughts are problematic, therapy with a selective serotonin reuptake inhibitor may be beneficial.

When epilepsy is confirmed, the child should be treated with an antiepileptic drug (AED). Figure 18-2 outlines the spectrum of activity of most of the commonly used AEDs. The most effective medications for absence seizures are ethosuximide, sodium valproate, and lamotrigine. Benzodiazepines, such as clobazam, may also be helpful. Differentiating absence seizures from CPS is essential, as some AEDs, such as carbamazepine, vigabatrin, and tiagabine, may be used for treating CPS but may aggravate absence seizures.

Absence Seizures
In our population of children with absence seizures, initial drug treatment was successful in 52 (60%) of 86 patients, with control less likely if a patient also had GTCS or myoclonic seizures. Terminal remission was more likely if the initial drug was successful than if it had failed (69% vs 41%; p < .02), and children whose initial drug treatment had failed were more likely to have JME and to develop intractable epilepsy. Although sodium valproate was more likely to be effective than other AEDs, many physicians prefer to start with ethosuximide when treating CAE, as it
is often better tolerated. Valproate is, however, a better choice for treating other absence syndromes and atypical absence seizures. When ethosuximide is prescribed, the usual starting dose is 10 to 15 mg/kg/d. The dose can be given once daily but is usually better tolerated when given twice daily. The dose is increased up to about 40 mg/kg/d depending on seizure control and side effects. Ethosuximide is available as large 250 mg capsules or as a suspension of 250 mg/5 mL. Commonly encountered adverse effects include gastrointestinal upset, drowsiness, hiccups, nightmares, and behavior changes. Side effects are seldom severe. Ethosuximide is not effective against GTCS and, therefore, should not be prescribed for children who have additional seizure types.

Valproate is an excellent AED, with effectiveness against a broad spectrum of seizures (see Figure 18-2). An effective starting dose is 20 mg/kg/d divided twice daily. In general, compliance is better with twice than three times daily dosing. The dose can be increased according to seizure response but can often be maintained at this starting dose. As the dose of valproate is increased, the incidence of tremor increases. Other side effects that are less related to dose (or blood levels) are nausea, weight gain, alopecia, thrombocytopenia, hepatotoxicity, pancreatitis, and ovarian cysts. The concern over fatal hepatotoxicity is most significant for those under 2 years of age and with metabolic disorders. Valproate is available as 250 mg tablets and as enteric-coated 125, 250, and 500 mg tablets. It is also available as 125 mg sprinkle tablets and as a suspension of 250 mg/5 mL.

Lamotrigine is also effective for the treatment of absence seizures. The major adverse effect associated with lamotrigine is skin hypersensitivity, which can occur in up to 4% of children. This risk is significantly lowered if the initial dose is low and the rate of titration over the first 8 weeks is slow. This results in a rather complicated titration schedule. For children with absence seizures, delaying the achievement of an effective dose for up to 4 to 8 weeks is often unacceptable. For patients on no other AED, the starting dose is 0.2 mg/kg/d administered once or twice daily. If patients are also taking an enzyme-inducing AED (eg, carbamazepine), start with 0.3 mg/kg/d for the first 2 weeks. The dose can then be doubled for weeks 3 and 4. Thereafter, the dose can be increased at weekly intervals by 1 mg/kg/d up to as much as 15 mg/kg/d. For patients on valproate, the dose should be approximately halved. Lamotrigine is available as chewable or dispersible 2 and 5 mg tablets and as 25, 100, and 150 mg regular tablets. Other formulations are available in some countries.

Patients who do not respond to their first AED may do well on a combination of valproate and either ethosuximide or lamotrigine. The role of newer AEDs, such as levetiracetam, in treating absence seizures has not yet been established.

Children who remain seizure-free for 1 year and who do not have a seizure induced by 3 minutes of hyperventilation might be weaned off their AED over approximately a 1-month period.

Complex Partial Seizures

There is a wider choice of AEDs available for children with CPS (see Figure 18-2). Most pediatric neurologists choose carbamazepine as their drug of first choice, although oxcarbazepine may have similar effectiveness without the potential for hepatotoxicity and hematoxidity. In addition, the risk of hypersensitivity skin reaction is lower with oxcarbazepine. Carbamazepine induces the hepatic enzymes responsible for its metabolism. The starting dose is usually 5 mg/kg/d divided twice daily. The dose should be increased at 5-day intervals to achieve a dose of around 15 to 20 mg/kg/d over 10 to 14 days. Further increases should be clinically determined by seizure control and
adverse effects. Common side effects include drowsiness, dizziness, diplopia, and lethargy. It is available as 100 and 200 mg “chewtabs,” a suspension of 100 mg/5 mL, 200 mg regular tablets, and 200 and 400 mg controlled-release tablets. Carbamazepine does not appear to be more effective than other AEDs in controlling CPS. The choice of medication is, therefore, primarily based on the potential for adverse effects.

**Summary**

The management of children with staring spells seldom proves difficult when approached in an organized fashion. Spells that can be interrupted by touch or tickling (“touch test”) or that only occur in relatively “boring,” less-stimulating circumstances are almost always due to daydreaming or inattention.

Children whose staring spells interrupt activities and are not easily terminated are more likely to have seizures. Childhood absence seizures can be precipitated by 3 minutes of hyperventilation, last less than 20 seconds, and are abrupt in onset and termination. Complex partial seizures may be preceded by an aura, last at least 1 minute, and are usually followed by a postictal phase of drowsiness or irritability. The appropriate treatment of seizures requires this differentiation to allow a rational choice of medications. Children whose seizures fail to respond to the first AED probably require referral to a pediatric epileptologist.

**Suggested Readings**
