Diagnosis of Epilepsy in Children

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Epilepsy is a major public health problem in all societies, with nearly 2 million Americans having some form of epilepsy. Approximately one-third of these individuals are children. Epilepsy is the most frequent reason for visits to a pediatric neurologist. This condition is a symptom of disordered brain function that results from a variety of different etiologies, some progressive, others static, and some of a genetic origin. In addition, children may have isolated seizures as a result of events such as infections, trauma, or exposure to neurotoxic agents. These symptomatic seizures are discussed elsewhere in this issue.

The diagnosis of epilepsy begins with identification of the seizure type. A seizure is defined as a clinical event caused by abnormal cerebral electrical activity that results in the disturbance of sensorium, motor activity, sensation, and/or autonomic function. Epilepsy is recurrent, unprovoked seizures. "Epileptic syndrome" is a term used to characterize certain seizure disorders, defined by the cluster of symptoms, signs, and findings on electroencephalograms (EEGs) and imaging studies. The epileptic syndrome suggests the etiology of the condition and the appropriate therapy, and predicts prognosis. As an analogy, the epileptic syndrome can be considered the "picture" and the clinical and laboratory findings can be thought of as its "colors."

The impact of epilepsy on the life of the individual and on society is substantial. Appropriate diagnosis and management can reduce the costs of diagnosis, and treatment lessens the effects of epilepsy on the child and the family.

**CLASSIFICATION OF EPILEPTIC SEIZURES**

The epilepsies are classified by two different methods, seizure type and etiology. The characterization of epilepsy based on the seizure type is easier and better reflects the possible underlying etiology (Table). Seizures are classified as either partial or generalized based on the site of onset of seizure activity in the brain. This is determined by clinical information, by findings on the EEG, or both.

Partial seizures have onset of the abnormal electrical activity in a specific locus of a cerebral hemisphere. This may remain restricted to the loci or may be followed by the spread of the activity to the rest of that hemisphere, or to the other hemisphere. Seizures that remain limited to one cerebral hemisphere and do not cause alteration of consciousness are referred to as simple partial seizures. The clinical presentation may range from convulsive motor activity, sensory alteration, disturbances of thought, to autonomic symptoms alone. Elaboration of the epileptiform
activity to the limbic region from either the temporal or the frontal lobes results in impairment of sensorium without complete loss of consciousness and may be associated with automatisms. These seizures are known as complex partial seizures. When the abnormal electrical activity involves both cerebral hemispheres, there is loss of consciousness with generalized convulsive movements. This is termed a secondarily generalized partial seizure.

Generalized seizures involve both cerebral hemispheres simultaneously from onset and manifest as an immediate loss of consciousness. They may present with impairment of sensorium alone, as in absence seizures, or with tonic and clonic, tonic, clonic, or myoclonic activity in a generalized fashion.

Based on the etiology, seizures are classified as secondary to an identifiable cause or idiopathic. Reactive or acute symptomatic seizures occur with (or secondary to) high fever, infections of the nervous system, metabolic disturbances, intoxication, head trauma, acute hypoxic and ischemic insults, and other acute encephalopathies. These seizures are usually generalized and present with tonic and clonic motor activity.

Epileptic syndromes are disorders that are primarily characterized by the type or types of seizures, clinical findings, and EEG. The epileptic syndromes are not specific diseases, and multiple etiologies can result in the same epileptic syndrome. Although findings on EEG are required for accurate diagnosis, a syndrome can be suspected based on a careful history and examination. The response to medication and the prognosis can be predicted for each epileptic syndrome. The identification of a specific epileptic syndrome also suggests underlying etiologies. This can increase cost-effectiveness for decisions about further evaluation, including whether neuroimaging is needed. For example, if the diagnosis of childhood absence epilepsy is made, there is no need for imaging studies because this epileptic syndrome is seldom associated with structural disease in the brain. In contrast, complex partial epilepsy can be associated with focal sclerosis, neoplasm, migration anomalies in the cortex, and other structural abnormalities, so imaging studies are generally indicated.

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<th>TABLE</th>
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<tr>
<td>Classification of Seizure Categories*</td>
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<tr>
<td>Partial (local, local) seizures</td>
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<tr>
<td>Simple partial seizures</td>
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<tr>
<td>With motor signs: focal motor, Jacksonian, versive, postural, phonatory</td>
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<tr>
<td>With somatosensory or special sensory symptoms</td>
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<tr>
<td>(simple hallucinations, eg, tingling, light flashes, buzzing): somatosensory, visual, auditory, olfactory, gustatory, vertiginous</td>
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<tr>
<td>With autonomic symptoms and signs</td>
</tr>
<tr>
<td>With psychic symptoms (disturbances of higher cerebral functions): dysphasic, dysnamic, cognitive, affective, illusions, structured hallucinations</td>
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<tr>
<td>Complex partial seizures (with impairment of consciousness)</td>
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<td>Simple partial onset followed by impairment of consciousness</td>
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<tr>
<td>With no other features</td>
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<tr>
<td>With simple partial features as in A1–A4</td>
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<tr>
<td>With automatisms</td>
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<tr>
<td>Partial seizures evolving to secondarily generalized tonic-clonic seizures</td>
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<tr>
<td>Generalized seizures</td>
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<tr>
<td>Absence seizures, with impairment of consciousness, with clonic, atonic, tonic, or autonomic components, or with automatisms occurring alone or in combination</td>
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<tr>
<td>Atypical absences, more pronounced changes of tone than in absence seizures; onset and/or cessation not abrupt</td>
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<tr>
<td>Myoclonic seizures (single or multiple)</td>
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<tr>
<td>Clonic seizures</td>
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<tr>
<td>Tonic seizures</td>
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<tr>
<td>Tonic–clonic seizures</td>
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<tr>
<td>Atonic seizures</td>
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<tr>
<td>Unclassified epileptic seizures</td>
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*Combinations of seizures listed here may occur.

COMMON EPILEPTIC SYNDROMES IN CHILDREN

Benign Partial Epilepsy

Benign childhood epilepsy with centerotemoral spikes (benign rolandic epilepsy of childhood) is a common epileptic syndrome seen in children between the ages of 3 and 15 years. It resolves by age 16 years. Approximately 15% of children with epilepsy who are younger than 15 years of age have this diagnosis. Generalized motor
seizures occur during sleep with occasional partial seizures when awake. The partial seizures often begin with sensory symptoms on one side of the face or tongue. This is followed by anarthria, drooling of saliva, and clonic motor activity of the facial and lingual muscles. The partial seizures may occasionally spread to become generalized tonic-clonic seizures. The seizures are generally brief and occur soon after falling asleep or early in the morning.

These children are generally healthy and the results of the neurologic examination are normal. However, the results of the EEG are abnormal and demonstrate characteristic frequent high amplitude spikes or sharp waves with following slow waves in the central and temporal leads from both hemispheres (Fig. 1). The spikes have a unique horizontal dipole that is negative in the temporal region and positive over the frontal region. The results of imaging studies in these patients are normal.

Benign rolandic epilepsy is inherited in an autosomal dominant fashion with variable penetrance. This epilepsy is diagnosed by the clinical description of seizures and the EEG. The positive family history is helpful when present.

Benign occipital epilepsy of childhood is a rare disorder seen in school-aged children. It is similar to benign rolandic epilepsy, but is characterized by visual experiences or blindness followed by clonic jerks on one side or automatisms with impairment of consciousness. Headaches frequently follow the seizures and have features similar to those of migraine headaches. The results of the EEG are often abnormal and show high amplitude spike and wave discharges from the occipital region. Most of these children have normal results on neuroimaging studies. However, because some of these children have structural lesions, including low-grade tumor in the occipital lobes, they need neuroimaging studies.

**Childhood Absence Epilepsy (Petit Mal Epilepsy)**

Absence epilepsy occurs in otherwise healthy school-aged children and is characterized by the occurrence of frequent “absences” of short duration. It begins between ages 4 and 8 years, and rarely before age 3. It constitutes approximately 8% of childhood epilepsies and is seen more frequently in girls than in boys.

The seizures occur without any warning and are periods of loss of consciousness (absences) that generally last seconds. When the spells are prolonged, automatism, such as eyelid blinking or clonic jerking of the limbs, occurs. The spells stop abruptly with no post-ictal lethargy or sleepiness. Children may have numerous spells in a day, but some can be brief and barely noticeable. Impairment of attention occurs during these spells, and children often present with a history of declining schoolwork. Some, especially those with a later age of onset, experience rare generalized motor seizures. The results of the neurologic examination are normal and the seizures can be activated by hyperventilation in the office.

The results of the EEG are normal, except during a seizure when the characteristic ictal pattern is seen. This consists of bursts of 3-Hz spike and wave discharges (Fig. 2). The seizures can be triggered by hyperventilation in the EEG laboratory. During sleep, fragments of generalized spike and wave discharges may be seen. The results of imaging studies are normal.

The diagnosis is suspected by the history and by demonstrating that seizures can be brought about by hyperventilation. The spells of absences are often confused with complex partial seizures. The latter can be differentiated from absence seizures by its clinical features: longer spells, automatisms, post-ictal lethargy, and sleepiness.
Definitive diagnosis is made on characteristic EEG findings during the seizure.

**Juvenile Myoclonic Epilepsy**

This syndrome is characterized by the occurrence of bilaterally synchronous single jerks or clusters of myoclonic jerks. The myoclonic jerks typically occur on falling asleep or on waking up. The jerks predominantly involve the arms, and, as a result, the patient often drops or throws objects. Rarely, the legs are involved, causing the child to fall. The seizures begin between ages 13 and 19 years. Most patients experience at least one generalized tonic-clonic convulsion. Fifteen percent of patients also have absence seizures, and 30% of these are photosensitive. Sleep deprivation is a potent provocative influence to precipitate the seizures.

The EEG abnormality in these patients is a frontally prominent, 4- to 6-Hz, bilaterally synchronous spike and wave plus generalized polyspike discharges. The spike and wave discharges often occur without associated myoclonic jerks. The symptom complex and EEG findings help identify this syndrome. The results of imaging studies are normal.

**Complex Partial Epilepsy**

Complex partial epilepsy affects all ages and is by far the most common type of intractable epilepsy. The epileptic seizures are characterized by impairment of sensorium, resulting from epileptiform activity beginning in the temporal, frontal, or occipital lobes. With onset in the temporal lobe, especially the mesial temporal area, the seizures often present with impairment of sensorium and psychic, autonomic, olfactory, and sometimes gustatory symptoms. An aura, usually a rising epigastric sensation, or an olfactory, gustatory, or another unusual feeling is reported more often by older patients. With impairment of sensorium, automatisms are seen. These occur in patterns of semipurposeful activity such as lip smacking, sucking movements, fumbling of hands, moaning, or verbal perseveration. Following the seizure, there is often a period of confusion, and sometimes nondirected aggressive behavior. Sleep for variable duration follows.

Seizures with frontal lobe onset are generally shorter and characterized by impairment of sensorium and postural or motor activity. The motor movements include head and eye deviation, posturing of the arms, falling, and incontinence. The post-ictal period is short. The partial complex seizures may spread and become a generalized motor seizure.

The inter-ictal EEG shows spike and wave discharges over the appropriate cortical region. In patients with seizures of temporal lobe onset, focal epileptiform discharges are seen over the anterior temporal regions. A sleep-deprived EEG and special EEG leads help improve the chances of finding the abnormal activity. Focal slowing of the background activity may be seen over the source of epileptic seizures. Focal abnormalities are seen less often in complex partial epilepsy of frontal lobe origin.

Structural abnormalities, such as cortical dysgenesis, cicatrices, cysts, vascular anomalies, and low-grade tumors, are seen more often with partial complex epilepsy. Hence, neuroimaging studies, preferably magnetic resonance imaging with contrast enhancement, should be performed in all patients with partial complex epilepsy.

**Infantile Spasms**

These are unique seizures seen only in infancy. They represent the response of a developing brain to a variety of insults. Infantile spasms can be cryptogenic (idiopathic), or, more often, they are symptomatic of a variety of underlying dis-
As with infantile spasms, the Lennox–Gastaut syndrome can be secondary to both cryptogenic and symptomatic diseases. Many patients have a history of infantile spasms. Neuroimaging scans, metabolic evaluations, and genetic studies are indicated, as determined by the clinical findings.

**DIAGNOSIS**

The first step in the diagnosis of epilepsy is determining that the paroxysmal spell in question is indeed a seizure. For the most part, the diagnosis of seizures is based on the descriptions offered by those who witness the spells. A detailed description of the spells, the circumstances surrounding them, and the symptoms prior to them helps distinguish seizures from other paroxysmal disorders. The description of the seizures also helps in determining type.

The next step is to determine whether the seizure is symptomatic of an acute disorder or whether it is unprovoked. Seizures that occur in children with acute illness require a detailed evaluation for infections of the central nervous system, metabolic disturbances, head injury, and intoxication. Most of these seizures are generalized motor convulsions and do not recur after the acute illness has resolved.

For the child with recurrent, unprovoked seizures, a complete history and physical examination will often uncover an epileptic syndrome. A detailed description of the seizure, as well as of precipitating events, is needed. One should note whether the child appears to have any warning or aura. The older child can often describe these symptoms, but the younger child cannot. Younger children can appear frightened or will seek out a parent as the seizure begins. The presence of an aura indicates a partial seizure. The description of the seizure, or ictus, is critical. Generally, parents report convulsive activity, brief alterations in muscle tone, or spells of altered sensorium. Convulsive activity should be noted as focal, generalized, or beginning focally and spreading (Jacksonian seizure). Any focal component suggests a partial seizure. Brief alterations in muscle tone, such as spasms, myoclonus, or drop attacks, often occur in clusters. The presence of automatisms occurs more often with partial complex seizures than with...
absence seizures. The frequency and duration of the seizures can also help with the diagnosis. One should always ask about other seizure types, in addition to that of the chief complaint. It is not unusual that when a child is seen for the first generalized motor seizure, a prior history of absence, myoclonic, or other seizures that the parents dismissed as being medically insignificant is discovered.

The post-ictal period is also important. Generalized motor seizures and partial seizures generally have some post-ictal symptoms, whereas absence seizures do not. Focal weakness (Todd’s paralysis) is typical of partial seizures.

The medical history should determine past injuries or illnesses that could be a cause for the seizures. Development and school performance should be reviewed. Delayed, arrested, or a declining course in these areas can be important clues to etiology. Similarly, a review of systems can occasionally discover related symptoms of an underlying cause. In many cases, the family history will determine a diagnosis of the familial epilepsies.

The physical examination should document growth parameters. A too large or too small head circumference needs further evaluation in the child with a seizure. The eye examination is also important in that it often reveals signs of metabolic, inherited, and acquired diseases. The funduscopic examination can show evidence of trauma, congenital infection, increased intracranial pressure, and abnormal pigmentation.

Neurocutaneous disorders such as tuberous sclerosis, neurofibromatosis, hypomelanosis of Ito, and Sturge–Weber syndrome have characteristic cutaneous and neurologic features. They often present as epilepsies with features of partial onset. Visceromegaly in patients with epilepsy should prompt evaluation for a storage disorder.

The pediatrician should feel confident in performing a screening neurologic examination. This would include evaluation of the cranial nerves, deep tendon reflexes, muscle strength and tone, coordination, mental status, and development. Focal findings are consistent with partial epilepsy and require neuroimaging studies.

Most children with recurrent, unprovoked seizures receive routine laboratory studies, but these are seldom useful. The most important studies are the EEG and neuroimaging scans. The findings on the inter-ictal EEG often diagnose the seizure type and help classify the epilepsy syndrome. The EEG should be obtained as soon as the diagnosis of a seizure is suspected, and before treatment if possible. However, anticonvulsant drugs usually do not eradicate seizure foci and one should not unnecessarily delay therapy for the purpose of obtaining an EEG, especially for patients who have generalized motor seizures. Post-ictal effects on the EEG may persist for up to a week following the seizure. During this time, spikes may be depressed and replaced with slowing. Focal slowing is valuable information, because it may indicate focal pathology.

The EEG may predict the recurrence risk after the first unprovoked seizure. However, an EEG with normal results does not exclude the diagnosis of epilepsy, and a small number of normal children with no history of seizures have epileptiform discharges on the EEG. The EEG laboratory and the quality of the interpretation are important concerns. Facilities and physicians experienced in studying children should be used when available. EEGs performed during spontaneous sleep, and with activation procedures such as hyperventilation or photic stimulation, often provoke abnormalities that may not be seen on a routine, awake EEG. Sleep deprivation is especially useful for patients with complex partial epilepsy. After the diagnosis of epilepsy is established, there is little need for “routine” EEG studies. The EEG should be repeated if the child has persistent seizures and an initial EEG that shows normal results, if there is a change in the seizure type, or when discontinuation of therapy is being considered. A persistently abnormal result on the EEG may suggest continued therapy.

Neuroimaging studies are indicated in all patients who have seizures with partial features, unexplained abnormal neurologic signs, or focal abnormalities on an EEG. Regardless of the seizure type, neonates and infants should also have an imaging study because of the frequent association with congenital and progressive diseases. Large studies have found that approximately 40% of children with epilepsy have abnormal findings on neuroimaging studies, whereas
52% of children with simple partial seizures and 15% with complex partial seizures have abnormal findings on computerized tomography (CT) scans. If the EEG shows focal slowing or the results of the neurologic examination are abnormal, almost two-thirds have an abnormal study. Conversely, if the examination and the EEG show normal results, the yield is only approximately 5% for having a scan with abnormal results. Although 40% of patients have abnormal findings, only 3% of these findings are of therapeutic significance. Clinical judgment should be used for the older child who has a generalized seizure disorder or rolandic epilepsy by EEG. Scans are rarely helpful in such cases and generally represent an unnecessary expense.

A magnetic resonance image of the brain with contrast enhancement is the preferred modality, especially for patients with complex partial epilepsies and simple partial seizures. The magnetic resonance imaging is more sensitive than CT and does not expose the child to ionizing radiation. In most studies, magnetic resonance imaging has approximately twice the sensitivity of CT for identifying mass lesions, and is even more sensitive for identifying focal gliosis and focal dysgenesis of the brain. Obtaining a magnetic resonance image can be problematic in small children because it often requires conscious sedation. Ultrasonography and CT are useful in certain situations, but do not generally provide as much information as magnetic resonance imaging. Even with an imaging study yielding normal results, one should consider repeating the study if the child shows a deteriorating course or worsening seizure control despite adequate therapy.

In some patients who have paroxysmal spells, the diagnosis of seizures can be difficult despite clinical evaluation and multiple EEGs. These patients should have EEG-video telemetry if the spells are frequent. EEG-video monitoring records both the clinical event and the EEG simultaneously. It is helpful in differentiating recurrent, mostly benign childhood paroxysmal disorders from epilepsy. For the test to be feasible from a practical standpoint, the spells need to be frequent. It should be performed at a center with qualified neurologists and technologists who have experience in prolonged EEG-video monitoring.

Neurologic consultation is indicated for neonates and infants with recurrent seizures, children and adolescents with recurrent seizures but normal results on EEG, and those who do not respond to therapy. Children with seizures and unexplained developmental or school problems should be referred, as should those with neurocutaneous disorders. Neurologic consultation should also be considered for children whose families need reassurance concerning the diagnosis or treatment plan.

In summary, the diagnosis of epilepsies is based on evaluation of clinical features. An EEG is indicated in all patients with epilepsy and helps in diagnosis and to classify the type of epilepsy. Normal results on EEG do not exclude the diagnosis of epilepsy if the clinical features are consistent with this diagnosis. Imaging studies are indicated for patients with partial complex or partial simple epilepsy.

REFERENCES

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