Benign Focal Epilepsies in Children

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EPILEPSY BOARD REVIEW MANUAL

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Statement of Editorial Purpose

The Epilepsy Board Review Manual is a study guide for trainees and practicing physicians preparing for board examinations in epilepsy. Each manual reviews a topic essential to the current management of patients with epilepsy.

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Benign Focal Epilepsies in Children

Natanya M. Mishal, MD, and Sonya G. Wang, MD

INTRODUCTION

Epilepsy has classically been defined as 2 or more unprovoked seizures occurring more than 24 hours apart. In 2013, the International League Against Epilepsy (ILAE) expanded the definition to also include those with a diagnosis of an epilepsy syndrome and patients with 1 unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after 2 unprovoked seizures, occurring over the next 10 years.1

The ILAE 2010 revised classification system divides seizures into 3 groups based on clinical and electroencephalography (EEG) data: generalized, focal, and unknown (which includes epileptic spasms).2 Generalized seizures rapidly engage bilaterally distributed networks, whereas focal seizures originate in discretely localized or more widely distributed subcortical or neocortical structures limited to 1 hemisphere.

Previously defined as “simple” or “complex,” focal-onset seizures can either occur with preserved or impaired consciousness. Focal seizures are further categorized on the basis of clinical manifestations, including focal motor activity and motor automatisms, sensory symptoms, autonomic changes, and higher cortical or psychic symptoms such as “deja-vu” or affective changes. Generalized seizures involve impaired consciousness and may be convulsive, with bilateral motor manifestations, or nonconvulsive, as in the case of absence seizures (Table 1).2

An epilepsy syndrome is characterized by a complex of clinical features, signs, and symptoms including, for example, age of onset, seizure semiology, EEG findings, and outcome.3 Symptomatic-focal or localization-related epilepsy, now replaced with the terms structural/metabolic in the revised classification system, refers to epilepsy resulting from brain injury (eg, due to trauma, stroke, or infection), immune-mediated causes, or structural disease (such as cortical dysplasias, neoplasms, or vascular malformations).2 In contrast, benign or idiopathic focal epilepsies are a group of electroclinical syndromes that occur in otherwise healthy and developmentally normal children and are characterized by focal onset seizures in the absence of underlying structural brain abnormalities. The term benign implies that the seizures are easily treated (or require no treatment at all), that there is spontaneous remission prior to adulthood, and that there is a lack of neurologic sequelae in the majority of patients.4 This last point is not always entirely true, as some children have mild neuropsychological impairment even with classic syndrome findings, as will be discussed below.

This group of benign focal epilepsies includes benign childhood epilepsy with centrotemporal spikes, Panayiotopoulos syndrome, Gastaut-type idiopathic childhood occipital epilepsy, and photosensitive occipital lobe epilepsy. There are several less well-defined and rarer syndromes, including childhood epilepsy with affective symptoms, benign childhood epilepsy with parietal spikes and frequent giant somatosensory-evoked poten-
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Table 1. Classification of Epileptic Seizures

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<tr>
<th>Focal</th>
<th>Generalized</th>
<th>Unclassified</th>
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<tbody>
<tr>
<td>Without impaired consciousness (motor, sensory, autonomic, psychic)</td>
<td>Absence</td>
<td>Epileptic spasms</td>
</tr>
<tr>
<td>With impaired consciousness</td>
<td>Tonic-Clonic</td>
<td></td>
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<tr>
<td></td>
<td>Myoclonic</td>
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Evolution to generalization

Childhood epilepsy with centrotemporal spikes, also known as benign epilepsy with centrotemporal spikes (BECTS) or benign rolandic epilepsy, is the most common focal epilepsy syndrome in childhood and accounts for 15% of all childhood epilepsies. Incidence ranges from 10 to 20 cases per 100,000 children younger than 15 years of age. There is a high genetic predisposition, and overall the syndrome has a slight male predominance. The onset of seizures occurs between ages 3 and 13 years, with a peak incidence between 7 and 9 years.

Clinical Manifestations
The characteristic ictal symptoms of BECTS correspond anatomically to the inferior pre-central motor gyrus and post-central sensory gyrus surrounding the central (or Rolandic) fissure. This area corresponds with the face and oropharynx, but as the focus moves more superiorly along the homunculus, seizures can also involve the upper and, less frequently, the lower extremity.

Sensory auras may precede the seizures. Auras are described as unilateral numbness or paresthesias of the tongue, lips, gums and/or inner cheek, stiffness of the jaw or tongue, or a choking sensation. These initial symptoms may not be reported by younger patients or when the seizures arise exclusively out of sleep.

Most focal seizures are characterized by unilateral orofacial tonic or clonic contractions, most often affecting the labial commissure. Fifty-three percent are associated with oropharyngeal/laryngeal signs of speech arrest or grunting, gargling or guttural sounds, and drooling. It is unclear whether the sialorrhea is due to increased salivation, swallowing disturbance, or both. Less frequently, the seizure can involve clonic movements or sensory abnormalities of the ipsilateral extremity, with the arm affected more commonly than the leg.

Approximately 80% of the seizures in BECTS are reported to be focal. The remaining 20% reported by parents to be generalized are likely a result of focal symptoms going unnoticed during sleep and the seizure subsequently evolving to secondary generalization.
The majority of seizures are nocturnal, occurring shortly after sleep-onset or just before awakening in the periods of non-rapid eye movement (non-REM) sleep in 55% to 65% of patients. A smaller percentage of patients solely have seizures during wakefulness, and even less have both nocturnal and diurnal seizures. Seizures in BECTS typically last seconds to minutes, with a maximum duration of around 3 minutes. That said, focal motor or hemiconvulsive status epilepticus can occur, especially in younger children. Generalized convulsive status epilepticus is rare.

**ETIOLOGY**

The etiology of BECTS is presumed to be genetically determined, with predisposition linked to chromosome 15q14. However, the details and exact mode of inheritance of the syndrome have yet to be characterized. Interestingly, the characteristic centrotemporal spikes on EEG have demonstrated an autosomal dominant pattern without association to other aspects of the clinical syndrome. Furthermore, 2% to 3% of school-aged children have these epileptiform discharges, and most never develop epilepsy. This suggests that the pathogenesis is multifactorial, likely comprised of genetic and environmental influences that act synergistically to generate the clinical phenotype we identify as BECTS.

**DIAGNOSTIC EVALUATION**

When clinical features appear to be consistent with BECTS, characteristic EEG findings will help to confirm the diagnosis. It has been thought that neuroimaging is not necessary when clinical and EEG features are typical and a diagnosis is certain. There is, however, a higher percentage of abnormal magnetic resonance imaging (MRI) findings in patients with BECTS, including hippocampal asymmetries and white matter abnormalities. That said, the relationship is unclear and the abnormal findings seem to be unrelated to the pathophysiology of BECTS.

**EEG FINDINGS**

The syndrome of BECTS is named for its hallmark EEG pattern. The background EEG is usually normal with evidence of classic interictal centrotemporal spikes (CTS) that have distinctive characteristic features. The main spike is a high-voltage, diphasic or negative component followed by a prominent slow wave of lower amplitude. Spikes are typically located in both central and temporal regions, but may shift or occur exclusively in 1 location. Furthermore, CTS may be found in alternative locations in approximately 20% of patients, with occipital spikes being most common initially. Horizontal dipoles are common, with maximal negativity in the centrotemporal regions and frontal positivity. CTS may be seen unilaterally, bilaterally as independent right or left discharges, or, less commonly, as synchronous bilateral discharges. They may occur independently or, more commonly, in clusters. Spike frequency can also be variable, with potential for abundant occurrence of up to 4 to 20 times per minute. An example of a typical EEG found in a patient with BECTS is shown in Figure 1.

Ictal patterns are rarely documented on EEG, but those that have captured events have demonstrated that seizures mostly originate from the centrotemporal region with frequent spread over the entire ipsilateral hemisphere or even to the whole brain. Postictal slowing is usually not prolonged.

Discharges increase as a patient proceeds sequentially into drowsiness and through the sleep stages. The main source of spike activation in
the early sleep stages is thought to be physiological enhancement of the sleep-promoting systems, whereas the increase just prior to waking may be due to increased microarousals. Despite the increased spike frequency in sleep, EEG sleep architecture is preserved.

After sleep, the next most common form of activation of CTS is contralateral somatosensory...
stimulation of the hands or feet. This is called somatosensory evoked potentials or extreme somatosensory evoked spikes and occurs in 10% to 20% of patients. Any type of mechanical or electrical stimulation will elicit the response. Percussion of the distal arms or legs with the fingertips or a reflex hammer is usually sufficient to elicit the spikes and is especially practical during routine EEGs. Overall there is poor correlation between the frequency and location of interictal spikes and the clinical picture in regards to seizure severity, duration, frequency, and prognosis. However, several studies suggest correlations between spike frequency and neuropsychological deficits in the realms of development, academic performance, memory, and attention, for example. Others have investigated the relationship between interictal discharges and psychological functioning. As an example, one study examined 21 children with BECTS and found a strong association between spike frequency and mood and behavior problems, including depression, aggression, conduct, executive functioning, and anxiety. Although there appears to be ample evidence, the findings of these studies should be interpreted with caution, as there are biases and confounding factors (eg, antiepileptic treatment or seizure frequency) that may affect neuropsychological testing results.

MANAGEMENT

Antiepileptic drug (AED) treatment is often not needed for BECTS, particularly if the seizures are infrequent, are focal without impaired consciousness, or are only nocturnal. Consideration of AEDs should occur for patients with frequent seizures, regular secondary generalization, early onset, or comorbid conditions. In one study, AED therapy was associated with a reduction in generalized but not focal seizures. According to the literature, carbamazepine is the most common initial AED used, but anecdotally many physicians choose to start medications such as oxcarbazepine or levetiracetam because they have fewer side effects. Rarely, certain AEDs such as lamotrigine, phenobarbital, and even carbamazepine can exacerbate BECTS, causing aggravation of clinical seizures, appearance of atypical absence seizures with myoclonic and atonic components, worsening of the EEG appearance, and neuropsychological deterioration. It has been suggested that children with diffuse interictal sharp and slow-wave discharges, rather than just sharp waves, are more likely to be at risk. All reported cases of exacerbation resolved with discontinuation of the inciting drug.

PROGNOSIS

Even without treatment, BECTS has an excellent prognosis. The cumulative number of lifetime seizures is generally low, with 10% to 20% of patients experiencing only one seizure. Approximately 60% of patients experience between 2 and 5 seizures total, with onset prior to age 3 years having a positive predictive value for recurrence. When seizures occur more frequently, they often present in clusters separated by intervals of up to a few months. However, there is an additional small subset of up to 20% of patients who report daily seizures. The natural course of the syndrome shows remission by mid-adolescence, typically within 2 to 4 years of seizure onset. As noted above, later age of onset (after age 4 years) is associated with achieving remission at an earlier age. Overall there is a less than 2% risk of going on to have seizures during adulthood. That said, EEG abnormalities may persist long after seizures cease and thus reliance on EEG findings is not
recommended for consideration of discontinuing medications.21

Neuropsychological deficits, particularly linguistic abnormalities and behavior problems, may present during the “active phase” of the disease, but most improve following seizure remission and EEG normalization. Rarely, on the order of less than 1%, patients may evolve to atypical and more severe forms of the syndrome (see following section).24,40

ATYPICAL EVOLUTION

It has been proposed that BECTS falls on one end of a spectrum of disorders comprised of epilepsy, abnormal EEG patterns, and language, cognition, and behavior impairments. Other disorders in this spectrum include atypical childhood epilepsy with centrotemporal spikes, epileptic encephalopathy with continuous spike-and-wave during sleep (CSWS), and Landau-Kleffner syndrome (LKS).

Atypical childhood epilepsy with centrotemporal spikes, also referred to as atypical benign focal childhood epilepsy (ABFCE) or “pseudo-Lennox syndrome,” is characterized initially by infrequent, focal and mostly nocturnal seizures similar to BECTS. However, this progresses to periods of multiple daily atonic seizures that can last a few weeks at a time. During these clinically severe weeks, the EEG shows a pattern of almost continuous slow spike-and-wave discharges during sleep, similar to what is seen in patients with epilepsy with CSWS. Prognosis can still be favorable, provided that there is no significant deterioration during frequent or prolonged seizures. CSWS syndrome, on the other hand, is marked by an increase in seizure frequency, global cognitive decline, motor impairment, and behavioral problems.40,43

LKS, or acquired epileptiform aphasia, is a syndrome that causes epilepsy and progressive psychomotor and behavioral disturbances beginning between 3 and 9 years of age.44 The syndrome presents as a regression of developmental milestones, beginning with receptive and then expressive language skills. Approximately 70% of patients have seizures, though they are infrequent. Most seizures in LKS are described as nocturnal simple focal motor seizures similar to those seen in BECTS. Initially there may be variability among EEGs, with findings of multifocal, unilateral, or bilateral independent temporal or temporo-parietal spikes, slow waves, or generalized discharges.46 Later, essentially all patients develop bilateral (or focal temporo-parietal) spike-and-wave discharges during much of non-REM sleep, defined as electrographic status epilepticus of slow-wave sleep. The outcome is unpredictable, with varying degrees of residual deficits.47

PANAYIOTOPoulos SYNDROME

Panayiotopoulos syndrome (PS) is a benign idiopathic focal childhood epilepsy syndrome characterized by mainly autonomic seizures.24 The age of onset is between 1 and 14 years, but approximately 75% of patients have their first seizure between ages 3 and 6 years, with an overall peak mean age of 5 years. PS is also referred to as early-onset benign childhood occipital epilepsy, in contrast to its “late-onset” counterpart known as Gastaut-type idiopathic childhood occipital epilepsy (described below).48 Boys and girls are equally affected.49 Prevalence is reported to be around 13% in 3 to 6 year olds with afebrile seizures, and 6% in the 1- to 15-year-old age-group in the original cohort of Panayiotopoulos in 1988. The numbers may be even higher if children with atypical presentations are included.49

CLINICAL MANIFESTATIONS

Autonomic seizures and autonomic status epilepticus are the hallmark clinical features of PS.
The most frequent ictal manifestation is emesis, occurring in 70% to 80% of seizures. However, aberration of any part of the autonomic nervous system may be present. Nausea and/or retching may precede the vomiting in 15% to 20% of patients, and may occur without subsequent vomiting in 5%. Usually a patient will vomit 3 to 5 times, but some do so repeatedly for hours. Brief irregularities in breathing or cardiac rhythm (such as tachycardia) are common. Though cardiorespiratory arrest is rare, it has been reported and thus PS should be considered a potential cause of sudden unexpected death in epilepsy. Common autonomic manifestations other than emesis include pallor (28%–94%), urinary incontinence (19%), cyanosis (12%), hypersalivation (10%), and mydriasis (7%). Less commonly reported symptoms include fecal incontinence (3%), intestinal motility abnormalities (3%), miosis (2%), and elevated temperature (2%). Syncope-like manifestations, described as sudden loss of muscle tone and unresponsiveness, are also commonly reported. Finally, behavioral changes such as restlessness, agitation, terror, or quietness can occur as auras at the seizure onset.

Two-thirds of the seizures occur during sleep, either nocturnal sleep or during daytime naps, particularly soon after sleep onset. Seizures in PS are typically long, with almost half lasting longer than 30 minutes and even up to many hours, therefore constituting autonomic status epilepticus. Of the seizures that last less than 30 minutes, the mean duration is still prolonged at 9 minutes. Convulsive status epilepticus is rare.

ETIOLOGY
As in BECTS, there is likely an underlying genetic predisposition to PS. A number of patients will report unprovoked seizures in family members, but usually there is no family history of similar seizures. There is also a high prevalence of febrile seizures (~17%).

DIAGNOSTIC EVALUATION
The most useful diagnostic test is EEG, and the classic findings for PS are described below. Although brain imaging is usually normal, MRI is often warranted given the unusual autonomic ictal features and high frequency of status epilepticus.

It can often be difficult to recognize autonomic manifestations as epileptic events. Consequently, PS may be undiagnosed or incorrectly diagnosed, for example, as gastroenteritis, atypical migraine, motion sickness, or even encephalitis when emesis is associated with a deteriorating mental status or generalized convulsions. When ictal symptoms include syncopal-like episodes, changes in cardiac rhythm, or pallor, for example, patients may first be referred for a cardiology consultation and/or work-
up. Having a sound understanding of the clinical manifestations of PS is important in order to avoid unnecessary testing and a delay in diagnosis.

**EEG FINDINGS**

The interictal EEG in PS is characterized by a (usually) normal background with focal and, more commonly, multifocal high-amplitude sharp and slow-wave complexes. Occipital spikes are most common, but there is high variability and shifting in terms of location. Two-thirds of patients have at least one EEG with occipital spikes; the remainder have extra-occipital spikes (most commonly in frontal and centrotemporal regions), rare brief generalized discharges only, or even normal EEGs. A sample EEG from a child with PS is shown in Figure 2.

Spikes are accentuated by sleep; thus, if an awake-only EEG is normal (10% of patients), a repeat EEG should be performed to capture sleep. Patients will rarely have classic occipital paroxysms of high-amplitude sharp and slow wave complexes with eye closure. This is known as “fixation off sensitivity” and occurs when there is elimination of central vision and fixation. The paroxysms are eliminated or markedly attenuated when the eyes are opened. Occipital photosensitivity is minimal.

Ictal EEG recordings are rare. When seizures are recorded, the ictal findings include rhythmic theta or delta activity with intermixed small spikes that usually have focal onset at a posterior region, though this is not universal. Clinical manifestations may start minutes after the electrographic seizure onset.

There is no correlation between frequency, location, and persistence of interictal spikes and seizure severity, duration, frequency, or prognosis. As in BECTS, EEG abnormalities can persist long after clinical remission.

**MANAGEMENT**

AED therapy is not recommended if seizures are brief or infrequent. According to the literature, recur-
rent seizures are often treated with carbamazepine or valproic acid (particularly if the EEG shows evidence of spike-and-wave discharges rather than just spikes), whereas patients with prolonged seizures are prescribed rectal diazepam as abortive therapy. However, as with BECTS, many physicians use oxcarbazepine as first-line therapy because of its reportedly more favorable side-effect profile. Many patients or families often request treatment because of the dramatic impact and length of the seizures.

PROGNOSIS

Seizures in PS are infrequent. Most patients will have 2 to 5 seizures total, and one-third have only a single seizure. Five percent have more than 10 seizures, but even those patients have favorable outcomes. The vast majority of patients achieve remission within 1 to 2 years of seizure onset. Of those that continue to have seizures, approximately 20% develop another type of infrequent seizure pattern, such as BECTS, and will remit by the mid-teenage years. The risk for developing epilepsy as an adult is probably similar to that of the general population.

Some patients may have subtle neuropsychological deficits during the active phase of their syndrome, most often involving minor visual, visuo-perceptual, attention, and memory functions. It is unclear if these deficits are syndrome-related or adverse effects of AEDs. Regardless, these deficits appear to be transient, and long-term cognitive outcome is favorable.

IDIOPATHIC CHILDHOOD OCCIPITAL EPILEPSY (GASTAUT-TYPE)

Gastaut-type, or late-onset idiopathic childhood occipital epilepsy (ICOE-G), is a rare pure occipital epilepsy syndrome that accounts for 2% to 7% of benign focal seizures in childhood. It is considered to occur 5 times less frequently than PS. Age of onset is between 3 and 15 years, but most cases begin at age 8 or 9. Males and females are equally affected.

CLINICAL MANIFESTATIONS

Seizures are purely occipital and manifest as both visual and nonvisual clinical symptoms. Visual seizures generally occur during times of wakefulness, though longer ones can be reported during sleep or after awakening as well. Hallucinations are the most common ictal manifestation and are often elementary in nature. Thirty percent of patients experience elementary hallucinations as their only seizure type. Positive elementary visual hallucinations are often described as small multicolored circular patterns that begin in one peripheral field, enlarge, multiply, and then frequently move horizontally toward the other visual field. These hallucinations usually last 5 to 20 seconds and very rarely up to 3 minutes. Negative elementary visual seizures can also occur, where there is sudden onset of ictal blurred vision or blindness. These can last somewhat longer, more on the order of 3 to 5 minutes.

Complex visual hallucinations (such as faces and figures) and visual illusions (such as micropsia, palinopsia, and metamorphopsia) may follow elementary hallucinations in less than 10% of patients as a result of anterior ictal spreading. The hallucinations do not have emotional features as temporal lobe seizures often do.

Interestingly, the main visual features for each patient recur stereotypically from episode to episode. In some instances, a change in lighting (from light to darkness or vice versa) has been reported as a stimulus that may trigger a seizure. Importantly, consciousness is intact during visual symptoms, but may become impaired later in the seizure.
Nonvisual occipital symptoms may occur at seizure onset or after an elementary visual hallucination. Eye deviation occurs in 70%. Eye deviation is often associated with ipsilateral version of the head and, if severe enough, may progress to a hemi- or generalized convulsion. Forced eyelid closure and blinking can occur when consciousness is impaired and usually signals impending secondary generalization. Other nonvisual symptoms include pain and sensory hallucinations of ocular movements. Additionally, ictal headaches can occur in association with, or less frequently, just prior to, other symptoms. Postictal headache is common, occurring in approximately half of patients, and can appear similar to a migraine headache lasting 30 minutes to several hours.

ETIOLOGY

Between 20% and 40% of patients report a family history of unprovoked seizures. Additionally, family members with migraine headaches are reported in 12% to 16% of patients. This suggests that there likely is an underlying genetic predisposition, but to date no studies have found a link. It is very rare for more than one member of a family to have similar Gastaut-type seizures.

DIAGNOSTIC EVALUATION

As in all the benign focal epilepsy syndromes of childhood, imaging is normal by definition and a diagnosis can be achieved based on clinical presentation and EEG results alone. However, the electroclinical findings seen with ICOE-G do not exclude a symptomatic epilepsy syndrome, such as an occipital epilepsy secondary to a cortical malformation. Therefore, neuroimaging with high-resolution MRI is indicated to evaluate for subtle lesions. EEG may prove useful in differentiating ICOE-G from symptomatic occipital epilepsies as well. Often the background is abnormal in the symptomatic group, with possible occipital polymorphic delta activity, bursts of sharp waves or fast activity, multifocal spikes, or polyspike-and-wave discharges.

EEG FINDINGS

The EEG in ICOE-G is characterized by a normal background with interictal high-voltage occipital spike-and-wave complexes or sharp waves, though some may have spikes only in sleep and a few may have normal EEGs. Extraoccipital spikes may also occur (most commonly in the posterior temporal areas), but less frequently than in PS. Interictal occipital paroxysms that may show “fixation-off sensitivity” are common, occurring in more than 80% of patients. As described above, these occur when a patient closes his or her eyes and the EEG displays long runs of high-amplitude sharp-and-slow-wave complexes in the occipital head regions. Discharges often increase during non-REM sleep. Photic stimulation usually does not affect the occipital paroxysms and the effect of hyperventilation is variable.

Just prior to seizure onset there is a decrease in the interictal occipital discharges. This is followed by an ictal period of sudden-onset occipital fast spikes and/or rhythms of lower amplitude than the interictal spikes. Generally the fast spike activity correlates with elementary visual hallucinations, whereas the slower semi-rhythmic activity correlates with complex visual hallucinations. There are usually no postictal abnormalities. Please see Figure 3 for a sample EEG from a patient with ICOE-G.

MANAGEMENT

Even though seizures are brief, it is generally recommended that patients with Gastaut-type childhood occipital epilepsy be treated due to frequent seizures (see below). Carbamazepine
has been used most commonly, with dramatic response in 90% of patients.\textsuperscript{24,63}

PROGNOSIS

If untreated, most patients experience frequent brief seizures. They can occur weekly, daily, or even multiple times per day in severe cases. That said, propagation to hemi- or generalized convulsions is rare and occurs once per month or year, or even less frequently.\textsuperscript{24} Remission seems to occur within 2 to 4 years of seizure onset in 50% to 60% of patients.\textsuperscript{13,64}

\textbf{Figure 3.} Interictal electroencephalograms (longitudinal bipolar montage) of an 11-year-old child with ictal visualizations of objects or letters, left gaze deviation, and postictal headache. (A) Frequent right occipital sharp waves are seen, maximal at the O2 electrode. They are present during wake and sleep, though the discharges become more irregular and higher voltage during sleep. (B) Eye closure causes an increase in discharge frequency, consistent with “fixation-off sensitivity.”
Neuropsychological testing found no statistically significant differences in function between 21 children with Gastaut-type childhood occipital epilepsy and age-matched controls, but there was a trend toward just slightly lower performance scores for attention, memory, and intellectual function in the affected children.70

**IDIOPATHIC PHOTOSENSITIVE OCCIPITAL LOBE EPILEPSY**

Idiopathic photosensitive occipital lobe epilepsy (IPOLE) belongs to a group of syndromes known as reflex epilepsies. Seizures are visually induced and start around puberty.71

**CLINICAL MANIFESTATIONS**

Seizures are evoked by visual stimuli such as watching television or playing video games.72 The seizures begin with initial visual symptoms, often brightly colored rings or spots in the periphery that are fixed or flashing and may move across and into the opposite visual field.72 Alternatively, patients may report negative visual symptoms of ictal blindness or blurring. The initial visual phase may be followed by head or eye deviation, usually toward the side of the visual manifestations.73 Half of patients report epigastric discomfort, nausea, or vomiting. Both ictal and postictal headaches are common. Seizures may last for several minutes.71

**ETIOLOGY**

The etiology of IPOLE is not known. As in many of the other benign focal epilepsy syndromes of childhood, one-third of patients report a family history of epilepsy or a personal history of febrile seizures.73 A few patients have a history of BECTS or CTS on EEG, but the association is unclear.74

**EEG FINDINGS**

The background EEG in IPOLE is characterized by the presence of interictal spikes or spike-and-wave discharges in the occipital regions, with predominance at the mid-occipital (Oz) electrode. Spikes may be unilateral or bilateral and synchronous or asynchronous. That said, patients may demonstrate these spontaneous occipital spikes, only have spikes during sleep, or have entirely normal EEGs.75 Photic stimulation often provokes an occipital photoparoxysmal response, but the paroxysm may also be generalized. Even without a true photoparoxysmal response, photic stimulation can produce abnormally high amplitudes.72 A seizure typically follows a photoparoxysmal response, whereby there is a buildup of spikes or spike-and-wave discharges that can shift between occipital regions.71 An EEG from a child with IPOLE is shown in Figure 4.

**MANAGEMENT**

Treatment should first and foremost involve the avoidance of known triggers. This may not always be practical, but patients can try to at least somewhat modify their triggers by increasing the distance from the television, using a screen filter, or using eye glasses, for example.

Patients with infrequent and mild seizures may not need drug treatment. However, treatment should be offered to those with marked photosensitivity and disabling seizures. Sodium valproate has been used most commonly.72,73 One study reported complete resolution of photosensitivity in 54% of patients and marked reduction in another 24% with the use of valproate.76

**PROGNOSIS**

IPOLE generally has a favorable prognosis. Most patients have only a few infrequent seizures
Photic Stimulation

Figure 4. Electroencephalogram (longitudinal bipolar montages) from a 14-year-old girl with photosensitive occipital lobe epilepsy. (A) Photic stimulation at an 8-Hz flash frequency provokes a broadly distributed photoparoxysmal response, with maximal activity in the biposterior regions. (B) Further photic stimulation provokes biposterior rhythmic sharp waves, spikes, and spike-and-wave complexes. As the potentials evolve into a 3- to 4-Hz spike-and-wave rhythm and the amplitudes increase (arrow), the child states that she sees flashing lights, and then later says she feels dizzy and feels the urge to vomit. The seizure migrates between the left and right occipital and posterior temporal regions, and resolves with focal slowing in the right temporal chain (especially posteriorly) (not shown).
OTHER BENIGN FOCAL EPILEPSIES OF CHILDHOOD

There are patients with benign focal epilepsy syndromes with clinical and electrographic findings similar to the syndromes described above, but atypical features make it difficult to diagnose them as such and instead have led to the description of alternate syndromes. In this section, we discuss childhood epilepsy with affective symptoms, benign childhood epilepsy with parietal spikes and frequent giant somatosensory-evoked potentials, and focal epilepsy in infancy with midline spikes and waves during sleep.

CHILDHOOD EPILEPSY WITH AFFECTIVE SYMPTOMS

This benign focal epilepsy syndrome has features in common with PS and BECTS. It is very rare and has been reported in fewer than 40 patients in the literature. Onset is between age 2 and 9 years, with males and females equally affected.77,78

Seizures consist of behavioral manifestations, such as screaming or terror, autonomic disturbances including pallor, sweating, hypersalivation, abdominal pain, chewing or other automatisms, and speech arrest. There may or may not be mild impairment of consciousness. Seizures are brief, on the order of 1 to 2 minutes, and they can occur multiple times per day. There is no temporal association, as seizures can occur any time during sleep or wakefulness.77,78

The interictal EEG is characterized by high-amplitude frontotemporal and parietotemporal spikes that are potentiated by sleep. Ictal EEGs tend to be stereotypical for each patient and consist of frontotemporal, centrotemporal, or parietal discharges.12

A history of febrile seizures is reported in one-fifth of patients. Patients respond very well to AEDs, and remission occurs within 1 to 2 years from onset. As in some of the other syndromes above, behavioral problems may be prominent during the active stage, but improve as the patient approaches remission.13

BENIGN CHILDHOOD EPILEPSY WITH PARietAL SPIKES AND FREQUENT GIANT SOMATOSENSORY-EvOKED POTENTIALS

The seizure semiology in this condition involves deviation of the head or body to one side, often without impairment of consciousness. Seizures are diurnal and occur infrequently.13

The EEG is characterized by parietal spikes and frequent giant somatosensory-evoked spikes (GSES), where discharges are elicited by tactile stimulation. These discharges are also potentiated by sleep.29,79 However, GSES are not specific to this syndrome, as they occur in 10% to 20% of patients with BECTS and a few patients with PS.49,79 Furthermore, they have also been documented in patients with no seizures.80

Remission occurs within 1 year of seizure onset, though EEG abnormalities may persist for a longer period.13,79 The development of patients is thought to be normal.

FOCAL EPILEPSY IN INFANCY WITH MIDLINE SPIKES AND WAVES DURING SLEEP

Seizure onset in this syndrome begins before age 3 and affects males and females equally. Clinical seizure semiology consists of staring, behavioral arrest, cyanosis, upper extremity stiffening, and loss of consciousness. Rarely, there may be unilateral motor features, clonic convulsions, or automatisms. Seizures are mainly diurnal and average duration is 1 to 5 minutes.12,13 Seizures are infrequent, occurring approximately 1 to 3 times per year.

The interictal EEG is characterized by midline small singular spikes followed by bell-
shaped slow waves only seen during non-REM sleep.\textsuperscript{12}

There is often a strong family history of different types of epilepsies, but with benign syndromes reported most commonly.\textsuperscript{13}

Generally, the prognosis is excellent. In most patients, the seizures remit and the EEG normalizes before the age of 4 years.\textsuperscript{13} Development is reported to be normal.

**SUMMARY AND OVERLAP OF THE CLINICAL SYNDROMES**

The syndromes described above have distinctive clinical and electroencephalographic characteristics (Table 2) that make specific diagnoses easy to achieve. Despite the distinctness, many patients have overlapping features, which raises questions as to whether these are truly distinct syndromes or perhaps part of an electroclinical spectrum. As an example, one study identified 24 children with ictal emesis characteristic of PS but with CTS on EEG characteristic of BECTS. According to this study, 20% actually had both seizure types concurrently, 17% had Rolandic seizures with ictal emesis, and 17% initially with PS seizures later progressed to typical BECTS seizures.\textsuperscript{81} Multiple studies have also found that 19% to 50% of patients with idiopathic childhood occipital epilepsy had a mix of presentations, both clinically and...
electrographically, between PS and ICOE-G.\textsuperscript{82–84} As discussed in the section on atypical evolution of BECTS, rarely patients with these benign focal epilepsy syndromes progress to more severe epileptic encephalopathies associated with CSWS such as LKS. The reason for this is not known, but this derailment from the typical course of the benign syndrome further suggests both overlap between and evolution from one syndrome to another.\textsuperscript{85}

The general prognosis and time to remission is also comparable among the syndromes. Though it is likely multifactorial, the etiology of all of these syndromes seems to be at least partly genetically determined given the association with different forms of epilepsies in family members.

Despite the wealth of information as displayed above, there are still many clinical, epidemiological, etiological, pathophysiological, diagnostic, electrophysiological, management, and outcome details of these benign focal epilepsy syndromes that have yet to be determined and warrant further investigation. From a clinical point of view, it is important that we as providers are able to identify the phenotypic presentations and differentiate them from other syndromes, both epileptic and nonepileptic. In doing so, we will be able to provide more precise diagnoses that have management, prognostic, and potentially significant psychobehavioral implications for our patients.

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