



which do not have an EEG correlate, are not considered epileptic seizures. The jerks of myoclonic seizures range from small movements of the face or hands to massive bilateral spasms that simultaneously affect the head, limbs, and trunk. Repeated myoclonic seizures may crescendo and evolve into a generalized tonic-clonic seizure. Although myoclonic seizures can occur at any time, clusters shortly after awakening are typical.

*Primary generalized tonic-clonic seizures* may begin with a few myoclonic jerks or abruptly with a tonic phase lasting 20 to 60 seconds and then a clonic phase of similar duration followed by a postictal state. Although there are usually no focal features, sometimes head turning occurs; this movement does not suggest a specific localization. If the onset is missed, it is often not possible to distinguish a primary generalized convulsion from a secondarily generalized one due to focal epilepsy.

### Idiopathic Generalized Epilepsy (Epilepsy Syndrome)

The idiopathic (primary) generalized epilepsies (IGEs) are likely polygenic resulting from a combination of mutations and polymorphisms in genes involved in thalamocortical circuitry. Different members of the same family often have dissimilar phenotypes. However, only rare IGE genes have been identified. A person with IGE has a 10% chance of passing the condition to a child. Most people with IGE have normal intelligence.

*Childhood absence epilepsy* (CAE; *pyknolepsy*, *petit mal epilepsy*) is the most common type of childhood epilepsy. It begins between 3 to 12 years of age with a peak at 7 years. Children with CAE have frequent absences (often hundreds per day) and are sometimes initially thought to have attention problems or to be daydreamers. Parents tend to report more absences at meal times, but that is due to closer observation; the absences are present throughout the day. One half of children with CAE have occasional GTC seizures. CAE is self-limited and seizures and the EEG abnormalities resolve by young adulthood. The absence seizures of CAE are typically provoked by hyperventilation, a useful procedure in the office setting and during an EEG.

*Juvenile myoclonic epilepsy* (JME) begins between age 8 and 20 years old. It is characterized by clusters of myoclonic seizures in the morning, starting shortly after awakening. The clusters typically persist for several to 30 minutes. The jerks are predominantly in the arms and last less than one second each. Consciousness is preserved. An affected teenager often fails to mention the morning jerks unless specifically asked. Sometimes there is a history of throwing breakfast utensils or toothbrushes due to the jerks. JME is often diagnosed after a morning GTC seizure. In JME, GTC seizures are particularly common after sleep deprivation or alcohol consumption during the prior night. People with JME are usually photic sensitive. That is, the seizures and EEG discharges are activated by flickering lights between 5 to 20 Hz (*photic-paroxysmal* or *photic-convulsive* response). This is a type of reflex seizure. Some people with JME also have absence seizures. The EEG is similar to that seen in CAE, but the generalized spike and slow wave discharges are slightly faster (3 to 4 Hz) and often have polyspike components. In contrast to CAE, the seizures in JME persist into adulthood and can be lifelong, although they lessen with age.

Less common IGE phenotypes include *juvenile absence epilepsy* (JAE) and *generalized tonic-clonic seizures alone* (GTCA). In JAE the predominant seizure type is absence with onset in the teenage years and, like JME, persists into adulthood. In GTCA the predominant seizure type is a convulsion, sometimes with a predilection for the morning. Less frequent absences or myoclonic seizures can occur in GTCA. Although onset of IGE is typically during childhood or teenage years, rare cases occur with onset in all ages of adulthood and are called *adult absence epilepsy* (AAE).

### Symptomatic Generalized Seizures (Seizure Types)

Symptomatic generalized seizures involve rapidly synchronized abnormal brain activity across the corpus callosum or involving midbrain structures occurring in brains with diffuse or multifocal dysfunction usually from early life.

*Drop seizures* can be either or both tonic and atonic. “Drop” implies that if the patient is upright they fall with no protective reflexes. Patients with drop seizures often suffer head injuries and should wear a helmet except when they are directly attended by a care provider, in a secure chair, or lying down. During a *tonic seizure*, the arms abruptly thrust forward at a 90 degree angle to the body and the entire body stiffens. Classically the fall is backwards. During an *atonic seizure*, tone is abruptly lost in the postural muscles and the patient falls forward.

*Atypical absence seizures* manifest as staring or mental slowing associated with a slow generalized spike and slow wave discharge (2.5 Hz or less) on the EEG. They may last minutes or even hours. Fluctuating levels of awareness and gradual onset and offset is described with atypical absences.

### Symptomatic Generalized Epilepsies (Epilepsy Syndrome)

Symptomatic generalized epilepsies occur in people with multifocal or diffuse brain dysfunction from early in life. There is usually an associated encephalopathy with some degree of developmental delay. In addition to all of the seizure types that occur in focal and idiopathic generalized epilepsy, people with symptomatic generalized epilepsies have tonic and atonic drop seizures and atypical absence seizures.

There are several unrelated types of epilepsy under the Symptomatic Generalized Epilepsy syndrome classification, with the most common being the *Lennox-Gastaut syndrome* (LGS).

The LGS is a common form of symptomatic generalized epilepsy due to diffuse or multifocal brain dysfunction. LGS presents from 2 to 10 years of age. Sixty percent have preexisting encephalopathy and developmental delay and 20% had infantile spasms (see later). LGS is responsible for 5% to 10% of childhood epilepsy. It is characterized by the combination of tonic or atonic seizures, myoclonic seizures, and atypical absences with the characteristic EEG pattern of 2.5 Hz or slower generalized spike and slow waves. During sleep there are bursts of diffuse fast rhythms on the EEG consistent with tonic or atonic seizures often with minimal clinical expression. Tonic-clonic and partial seizures also occur. Almost all people with LGS have developmental delay with associated behavioral disorders. LGS is a chronic condition requiring supervision; many ultimately live in group homes. If drop seizures are present, and the patient is ambulatory, a helmet should be prescribed for protection.