Carpe Diem – Seize the Day Blog

Editor's Note: Content presented in the Carpe Diem – Seize the Day Blog is for awareness and ***informational purposes only, and it is not meant to be a diagnostic tool.***
Epilepsy describes a spectrum of neurological disorders. These disorders involve recurring seizures caused by abnormal electrical activity in the brain. There are many different types of epilepsy and diagnosing the specific type of epilepsy you may have can help you and your doctor better understand what to expect in terms of seizure progression, challenges in learning and physical development, and treatment options.

There are three major categories of seizures: focal, generalized, or unknown onset. Other types of epilepsy are known as syndromes, or collections of specific signs and symptoms that point to a certain medical condition.

Each type of epilepsy has unique characteristics. Some syndromes are considered benign, meaning children will eventually become seizure-free with age, while other types of epilepsy are lifelong conditions.

Focal Epilepsies and Syndromes
Some types of epilepsy are referred to as localized, which means they are known by the location in the brain where seizures originate. A related term is focal epilepsy, which indicates seizures that have abnormal electrical activity focused in one area of the brain.

Temporal Lobe Epilepsy
Approximately 60 percent of people with focal epilepsy have temporal lobe epilepsy (TLE), or seizures that originate in the temporal lobe of the brain. The temporal lobe is located on the sides of the brain, near the ears, and is responsible for processing sound and language as well as memories relating to sound and vision. About one-third of TLE or other cases of focal epilepsy are resistant to anti-epileptic drugs (AEDs). Resective surgery may be recommended, and other types of palliative neuromodulatory devices, such as vagus nerve stimulation may also be therapeutic.

TLE usually develops between late adolescence and early adulthood, often after a head injury or febrile (fever-induced) seizure. For women with TLE, hormonal changes during the menstrual cycle can sometimes increase seizure activity. Focal onset seizures are most common for people with TLE, though some people may experience prolonged seizures or, in rare circumstances, status epilepticus (multiple, rapid seizures).

Frontal Lobe Epilepsy
Frontal lobe epilepsy is the second most common form of focal epilepsy after TLE. FLE may be inherited, or it may be caused by a structural problem such as a birth defect, an abnormal blood vessel, trauma, or scarring caused by infection. In about 50 percent of FLE cases, no cause is ever determined.

The frontal lobes of the brain are large, and they help you with reasoning, paying attention, regulating emotion, organizing, and solving problems. FLE seizure symptoms can vary widely, depending on the function of the affected part of the lobe. It is also possible for a seizure to begin in the frontal lobe and proceed without symptoms until the seizure spreads to other parts of the brain, leading to a tonic-clonic seizure. ("Tonic" refers to muscle stiffness and "clonic" refers to muscle jerking.) FLE is usually responsive to medications, but in cases where AEDs are ineffective, surgery or vagus nerve stimulation may help.

Occipital Lobe Epilepsy and Parietal Lobe Epilepsy

The occipital lobes are in the back of the brain and are primarily responsible for vision. The parietal lobes are located on the top and upper sides of the head. They are known as the "association cortex" because it is where perception becomes reality — sounds are recognized as words, visual images are created, and touch becomes associated with an object. Epilepsy that originates in the occipital and parietal lobes is much less common than TLE and FLE. Seizures that begin in these lobes are usually idiopathic (of unknown cause). In both types of epilepsy, ASMs are the first treatment option. If medication fails, surgery may be recommended.

Panayiotopoulos Syndrome

Also known as early onset occipital epilepsy, panayiotopoulos syndrome (PS) commonly begins in early childhood, typically between the ages of 3 and 10 years. PS affects children of all genders equally, and its cause is unknown. As many as 6 percent of children who have nonfebrile seizures (those not caused by fevers) have this type of epilepsy. PS frequently stops two to three years after the first seizure, though PS may turn into juvenile myoclonic epilepsy (JME) in some cases.

Children with PS will typically have focal seizures that can spread to a generalized seizure. Seizures in PS often last between one and 30 minutes, and more than half of the seizures occur during sleep. Pale skin, a sick feeling, and vomiting are typical symptoms during a PS seizure. Some children may also have tonic-clonic movements (convulsions). If seizures are infrequent, medication may not be needed. However, if they are needed, ASMs are usually effective at controlling PS seizures. Neurologists often teach parents how to initiate rescue therapy and create an emergency plan for children with PS.

Benign Rolandic Epilepsy

Benign rolandic epilepsy (BRE) — also known as benign epilepsy with centrotemporal spikes (BECTS) — usually begins around ages 6 to 8 years. Boys are slightly more likely to have BRE than girls are. BRE accounts for approximately 15 percent of all epilepsies in children.
Benign rolandic epilepsy is characterized by numbness, twitching, or tingling of the face or tongue. Seizures may inhibit speech and cause drooling. The child remains conscious during the seizure, which are infrequent and occur mostly at night. AEDs may be prescribed if the seizures happen during the day or disrupt sleep, but many children do not need medication. Seizures stop by early adolescence in almost all children with BRE.

Generalized Epilepsies and Syndromes
Generalized types of epilepsy feature seizures that do not originate in or remain confined to one lobe or area of the brain. Generalized epileptic syndromes tend to be idiopathic. Idiopathic generalized epilepsies account for one-third of epilepsy cases.

Childhood Absence Epilepsy
Childhood absence epilepsy (CAE) accounts for 2 percent to 8 percent of childhood epilepsy cases. Childhood absence epilepsy typically begins between the ages of 3 and 11 years, most frequently between ages 5 and 8. One-third of children with CAE have a family history of seizures, suggesting that the cause may be genetic. Siblings of children with CAE have a 10 percent chance of developing epilepsy.

Children with CAE experience absence seizures (formerly known as petit mal seizures). Often, the seizures take place while the child is exercising. The child is not aware or responsive during seizures, and may stare, blink, or roll their eyes up. You may notice a chewing motion or other repetitive movements. Seizures usually last less than 15 seconds, after which the child immediately returns to normal. The child is usually not aware they have had a seizure. Seizures may be infrequent or happen as often as one hundred times a day.

Some children with CAE have concentration and memory problems before seizures start. Rarely, children who have very frequent seizures may develop learning difficulties.
At least two-thirds of children with CAE respond to treatment, and their seizures will cease by mid-adolescence. If ASMs are not effective, the ketogenic diet may help children with CAE. However, 10 percent to 15 percent of children with CAE will develop other seizure types during adolescence — typically myoclonic seizures (myoclonic meaning short periods of jerking movements), generalized tonic-clonic seizures (formerly known as grand mal seizures), or both.

Juvenile Myoclonic Epilepsy
JME, also known as Janz syndrome, is the most common generalized epilepsy syndrome. This syndrome usually begins between the ages of 6 and 26 years, commonly between the ages of 12 and 16. Mild myoclonic seizures, generalized tonic-clonic (GTC), or clonic-tonic-clonic seizures (GTC seizures that begin with a clonic phase) are the most common types of JME. Myoclonic seizures tend to occur immediately upon waking in the morning. Absence seizures may be the first type of seizure most people with JME experience, although this type happens less often. Photosensitivity (seizures triggered by flashing or flickering light) affects approximately 45 percent of people with JME. Photosensitive seizures usually show on an electroencephalography (EEG) test. Most cases of juvenile myoclonic epilepsy are treatable with ASMs. Most people with JME need to remain on medication for life.

Juvenile Absence Epilepsy
Juvenile absence epilepsy (JAE) is similar to childhood absence epilepsy; however, it starts later in childhood (generally between ages 10 and 16) and is usually a lifelong condition. Between 1 percent and 2 percent of people with epilepsy have juvenile absence epilepsy. Although it is rare to have a family history of seizures, the cause of JAE is thought to be genetic. People with JAE experience absence seizures lasting from 10 to 45 seconds. They generally experience less than one absence seizure per day. Seizures often happen during exercise. Around 80 percent of those

with JAE will also have tonic-clonic seizures. The risk of absence status epilepticus (also known as nonconvulsive status epilepticus), in which seizures can last minutes or even hours, is higher in people with JAE.

Children with JAE generally develop normally, though they may experience learning difficulties if they have frequent seizures. Some children with JAE also have concentration and memory problems before seizures start; these issues often improve after AEDs are started. AEDs work well to treat JAE and generally must be taken for life.

Lennox-Gastaut Syndrome

Lennox-Gastaut syndrome (LGS) is an uncommon epilepsy syndrome. Between 1 percent and 4 percent of children with epilepsy have LGS. LGS occurs somewhat more often in males compared to females. Lennox-Gastaut syndrome often starts between the ages of 3 and 5. Atonic seizures, also known as drop attacks, are most common in children with LGS. Seizures may happen multiple times a day and cause the child to suddenly drop to the ground. Drop attacks may be perceived as a trip or the result of poor balance. Injuries are common, making the seizures upsetting for the child. Atypical absence seizures and tonic seizures are also common, especially at night, but children with LGS may experience other types of seizures as well.
LGS is very difficult to treat and is often referred to as intractable or refractory. Some AEDs can be effective, and the ketogenic diet may help. Surgery may be recommended if diet and medication do not work, though surgical treatment will not stop the seizures altogether.
Children with LGS tend to have moderate to severe learning difficulties, with some children exhibiting developmental delays before their first seizure. Some children who have West syndrome (infantile spasms) will develop LGS.

Progressive Myoclonic Epilepsies

The progressive myoclonic epilepsies are a group of rare syndromes characterized by a combination of tonic-clonic and myoclonic seizures. Disorders that fall under this category include:

- Lafora disease
- Mitochondrial encephalopathies
- Severe myoclonic epilepsy of infancy (also referred to as Dravet syndrome)
- Unverricht-Lundborg disease (also known as Baltic myoclonus)

The cause is often hereditary but may be unknown. Progressive myoclonic epilepsies affect males and females equally and start at different ages, depending on the specific condition.
In progressive myoclonic epilepsies, seizures are difficult to control. As the condition progresses, people with PME accumulate cognitive (thinking and memory) and motor (movement) disabilities. Medications may be successful at first, but effectiveness declines over months or years as the disease progresses.

Other Epilepsy Syndromes

While the syndromes described above are the most commonly diagnosed, there are many others that cause epilepsy. Examples include Angelman syndrome, Doose syndrome (myoclonic astatic epilepsy), neurocutaneous syndromes (such as Sturge-Weber syndrome), Rasmussen's syndrome, and Rett syndrome

Information for this blog was provided by Brooke Dulka, Ph.D., and Kelly Crumrin

Editor's Note: The Carpe Diem – Seize the Day Blog will be distributed and posted weekly.
Always remember – **CARPE DIEM – SEIZE THE DAY!**

Steve.Hutton@epilepsy-ohio.org