

Understanding Seizures/Epilepsy

By Lynne M. Kerr, M.D., Pediatric Neurologist, Division of Pediatric Neurology, University of Utah Medical Center

A seizure is the change in attention or behavior that occurs due to an episode of abnormal electrical activity in the brain. Epilepsy is a brain disorder in which a person has repeated seizures over time. For unknown reasons, individuals with Cornelia de Lange Syndrome (CdLS) have an increased incidence of epilepsy compared to the general population; seizures are observed in approximately 15 percent of children with the syndrome.

The first question a medical professional considers is whether the episode was in fact a seizure or some other kind of spell. In children with CdLS, reflux episodes causing arching may look like seizures. Other spells that mimic seizures include breath-holding, daydreaming, tics, and stereotypies, which are repetitive, apparently non-functional, possibly self-stimulatory behaviors. It is important to definitively diagnosis the episode as being due to a seizure, since other types of behavioral events will not respond to anti-epileptic medication and these medications can have unwanted, sometimes dangerous, side effects.

Conversely, sometimes children with no behavioral or consciousness changes will have an electroencephalogram (EEG) that is abnormal. An EEG is a non-painful test involving the application of electrodes to the head to record brain waves. Although there are exceptions, in general we do not treat children for abnormal EEGs without behavioral events that may be seizures. For example, abnormal EEGs without behavioral correlates are fairly common in children with autism, and this may be true in children with CdLS as well.

If the physician feels that the episodes that the child is having are likely to be seizures, she or he may perform an EEG. EEGs are not performed unless there is a strong clinical suspicion of a seizure disorder.

Since it's often difficult to tell if a particular episode is a seizure, family members may be asked to keep a record of the events for a few weeks, and even videotape the activity they are concerned about. A follow-up is then scheduled with the neurologist. If events that are concerning seizures are occurring fairly often, and a standard EEG, which is only 20-30 minutes long, does not show epileptiform activity, the neurologist may perform a longer EEG.



These can be done in two ways. The first is an ambulatory EEG where the EEG wires are worn while the individual performs their normal activities at home for several days. While the ambulatory EEG is in place, the individual or family is able to push a button to demonstrate when the activity of concern occurs which then can be correlated with the electrical activity in the brain. The second is an in-hospital video EEG where the behavior/consciousness changes that may be seizures are monitored with video while the individual wears an EEG montage.

As a member of the Clinical Advisory Board for the CdLS Foundation, and as a practicing pediatric neurologist, I have been asked many times about behavioral episodes and whether or not these are seizures. One example is an individual with episodes of aggression. It is not likely that behavior changes such as rage or aggression are due to seizure activity. However, if the questions persist it is possible to determine if events of concern are seizures or not by recording the brain's electrical activity during the event by either ambulatory EEG recording or a prolonged in-hospital EEG. The second example is a child whose heart rate went up to a high level and stayed that way for a while after being given food by his gastrostomy tube. The certainty that this testing can offer can allow the event of concern to be addressed directly and lays the question of underlying seizure to rest.

If seizures are diagnosed, the physician often orders further testing, including a brain MRI to determine if there are any structural changes in the brain such as scarring that may be causing the seizures. MRI is preferred to a CT scan because although individuals may need to be sedated for MRIs, the images obtained of the brain are much clearer and there is no exposure to radiation.

While the type of seizures and age of the child guides the choice of medication, convenience and expense for the family should be considered. All seizure medications have potential side effects, including changes in mood, activity level and learning. Every family should ensure that they have been thoroughly informed regarding the benefits and risks of the medicine being prescribed for their child.

Families should also be aware that if the first medication chosen causes unwanted behavior changes such as difficulty sleeping or difficulty focusing, they should contact their neurologist regarding changing medications. There are alternate therapies as well that are sometimes used when children have difficulty controlling seizures. These include a special diet (the ketogenic diet), epilepsy surgery, and implantation of a vagus nerve stimulator. Your neurologist may refer you to a specialized epilepsy center if



seizures are difficult to control.

Questions regarding events that may be seizures, which medication is the right medication, and possible side effects should be discussed with the child's neurologist. It's helpful to find a neurologist familiar with children with CdLS when possible.

For more information on seizure disorders, visit
www.medhomeportal.org, www.epilepsyfoundation.org or

www.epilepsy.com. Published First Quarter 2014

