

Seizures

FOR MOST PEOPLE, seizures are a frightening thought. They can be simply described as a kind of sudden static, or bolt of electricity in the brain. Watching a seizure for the first time can be alarming, but learning what happens during a seizure reduces our anxiety and helps us learn how to react with calm and understanding. Your daughter with RS may never have a seizure in her life, or she may have seizures that are well controlled with medication. Knowledge is your best ally.

Epilepsy is a broad term that is used to describe recurring seizures or the potential for recurrence of seizures. Seizures are called by various names including fits, spells, convulsions, and attacks. A seizure is a sign of a disorder and in and of itself is not an illness or a disorder. You can not “catch” epilepsy. It does not cause mental retardation, is not a mental illness, and is not anyone’s fault.

THE CAUSE

WHILE MUTATIONS IN THE *MECP2* gene are found in ninety-five percent of girls with classic RS, how these mutations cause RS is not known. We do not understand why some children with RS have seizures while others do not. However, since RS involves the nervous system, it is not surprising that seizures occur. If seizures do occur, the onset tends to be between two and ten years of age. However, bear in mind that seizures are often over-reported in RS. Only fifty-percent of those with RS truly have epileptic seizures. Other “seizures” are a consequence of motor dysfunction (stiffening, rigidity, tremors), autonomic dysfunction (cold, blue extremities, facial flushing, pupil dilatation or enlargement), awake breathing abnormalities (breath-holding, hyperventilation, blank staring), gastrointestinal disturbances (acid reflux), or other behaviors seen in girls with RS. These non-epileptic episodes would not be expected to be responsive to antiepileptic drugs (AEDs).

A VISIT TO THE NEUROLOGIST

IF YOU SUSPECT THAT YOUR DAUGHTER is having seizure activity, it is wise to consult with a doctor who specializes in the treatment of seizures. It will put your mind at ease and it will assure that she gets a thorough evaluation of her seizure status. Check with other families in your area for the name of a qualified child neurologist. If your daughter does have seizures, you will be in frequent contact with him, so it is important



*“In our lives
there is
bound to be
some pain,
surely as
there are
storms and
falling rain;
just believe
that the one
who holds
the storms
will bring
the sun.”*

— ANONYMOUS

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that you are satisfied with his knowledge and experience, approachability, and willingness to consider you as partners in your daughter's care. Seizure control is an evolutionary process and it is peculiar to each different individual. What works for another child may not work for yours, and what once worked for your child may not always work.

SEIZURES HAPPEN

EVERY BRAIN CELL IS AN ELECTROCHEMICAL UNIT that generates a very small electrical current. The brain sends out electrical signals through our nerves. The brain receives signals along the nerves from all parts of the body. Normally, this electrical activity is well regulated and organized. Sometimes there is sudden excessive or erratic electrical activity. When this happens, it may result in a seizure. The electrical disturbance that accompanies this seizure may arise from any part of the brain. The kind or type of seizure may indicate the area in the brain where the electrical disturbance arises.

IS IT A SEIZURE?

A CLINICAL SEIZURE RESULTS from the excessive, synchronous discharge of brain cells (neurons), which brings about a change in movement or behavior. If no change is seen in behavior or movement, it is not considered a clinical seizure. Rarely, a seizure discharge may be recorded during an EEG (electroencephalogram, the recording of ongoing brain activity) without any obvious clinical change in a person. More frequently, random EEG abnormal discharges are recorded in many girls. These are not seizures and in themselves do not require antiseizure medication.

In RS, it can be difficult to determine whether she is having a clinical seizure or autonomic responses that look like a seizure. Often, seizurelike behaviors are seen, but seizure activity is not seen on the EEG during these events. Girls with RS have vacant spells that resemble absence seizures, but are not. Often there are jerky movements and eye rolling that are associated with seizures, but in RS they are involuntary movements that are not seizures.

NON-EPILEPTIC EVENTS IN RS

THE FOLLOWING EVENTS MAY LOOK LIKE TYPICAL seizure behavior, but in RS, they are often *not* seizure related:

Motor Events

- Twitching
- Jerking
- Head turning
- Falling
- Trembling
- Staring/vacant spells

Other Events

- Dilated pupils
- Laughing/screaming episodes
- Breathing: apnea and hyperventilation
- Nighttime awakenings

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Jessica gets the shakes when she first wakes up in the mornings or after a daytime nap. It is like she is really scared and surprised by any movements. She does not walk and so when we pick her up we give her big cuddles (she hangs onto us so tightly) and reassure her until she collects herself. If we sit her on the floor, she grabs hold of whatever she can to stabilize herself and has a look of fear on her face. We have put it down to her poor balance and think that while lying down she feels secure and comfortable but when brought upright into sitting her balance reactions are "confused." It takes her anywhere between five and fifteen minutes to stop shaking. We have also noticed that if she is afraid by a gross motor activity that may push her a bit hard she will get the shakes; when this happens, we just back off and let her recover. We have discussed it with doctors, who don't believe it is seizure-related.

Management of Non-Epileptic Events

- Recognize any potentially treatable medical or surgical disorders
- Consider the use of atypical neuroleptic drugs such as Risperdal or Seroquel
- Consider the use of anti-anxiety medications such as Buspar or Diazepam
- Consider the use of antidepressants (SSRIs, tricyclics)

DIAGNOSING A SEIZURE

THE DIAGNOSIS OF EPILEPSY (SEIZURES) IS MADE on the basis of the parent's description of seizures, physical and neurological examinations, and the electroencephalogram (EEG) findings. An EEG is a test to measure and record the electrical activity generated by the brain. It does not measure intelligence. Small electrodes are placed over the scalp and held in place with tape or a special paste. Your daughter will experience no pain or discomfort during an EEG recording, but she must remain still during placement of the electrodes. Therefore, she may become upset and cry. In some situations, she may need medication to make her drowsy in order to record her brain activity during sleep.

The EEG will show changes that may indicate an abnormality in one or many areas of the brain, localize the specific area of the brain that is involved, help determine the type of seizure, and reveal the kind of medication that can best control the type of seizure.

In individuals who do not have epilepsy, the EEG recordings resemble squiggly lines with waves that are similar in height. In most people with seizures, abnormalities are seen as little bursts of electrical activity, called "sharp waves" or "spikes," that interrupt normal rhythm.

The EEG records only the electrical activity present at the time the EEG is being recorded. Therefore, a seizure will not be recorded unless she has a seizure during the recording. However, if she has had a recent seizure, the EEG tracing may show changes that are helpful to the physician in determining appropriate treatment. An EEG measures waves on the surface and outer layers of the brain only. If the EEG does not show seizure activity during what you think may be a seizure, it doesn't mean that seizures are not occurring. It could be that they are taking place deep within the brain and cannot be measured by conventional means.

The neurologist will determine if an EEG is necessary. The EEG should be recorded in both the awake and sleep states since the abnormal activity may be seen in either or both. With medication, the seizures will probably decrease in frequency or be completely controlled. This does not mean the EEG will necessarily be normal.

The EEG looks different when she is awake and asleep, and some abnormalities may only be seen when she is drowsy or sleeping. Some children have a normal waking EEG and a very abnormal sleep EEG.

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Routine repetition at periodic intervals is not necessary, but repeating the EEG may be necessary if seizures change in character, severity or frequency. Children with RS frequently have abnormal EEG patterns. This abnormal pattern may include activity called epileptiform abnormalities that may be recorded in persons who have seizures. The information supplied by the EEG may help define the specific seizure type. The physician can then choose the most appropriate anticonvulsant drugs. However, a normal EEG does not rule out the diagnosis of epilepsy or mean that seizures have not occurred. On the other hand, if there is no history of a seizure, an abnormal EEG does not make the diagnosis of epilepsy.

THE EEG IN RETT SYNDROME

WHILE THE EEG IS USUALLY ABNORMAL IN RS, there is no diagnostic pattern. EEG patterns frequently seen in RS include generalized slowing, rhythmic slow activity (reported as “theta” activity), and focal and generalized spikes and sharp waves.

READING THE EEG

THE EEG CAN PICK UP “ARTIFACTS,” such as muscle twitches and eye blinks that are not coming from the brain. It is not abnormal to see these on an EEG. The abnormalities that are important include spikes, slowing, and evidence of seizures. They may be either focal (localized to a specific area of the brain) or generalized (seen all over the brain).

Spikes are abnormal discharges from brain cells. The abnormal discharges may involve many brain cells and may result in an EEG seizure discharge and seizure. When spikes are seen in a specific area of the brain, it may indicate where the seizure began. Multifocal spikes give an indication that there are many abnormal areas of the brain.

Slowing of the EEG is determined when it is compared to the normal rhythm of the EEG, which varies with the age of the child and whether she is awake, drowsy, or asleep. Generalized slowing is often seen in children with chronic brain dysfunction.

Evidence of seizures refers to the association of specific abnormalities on the EEG with specific seizure types.

AMBULATORY EEG

IN SOME CASES, IT IS NECESSARY TO OBTAIN a twenty-four hour EEG to see if the behavior correlates with EEG seizure discharge. This is usually performed with a small tape cassette worn with a belt around the waist that is attached to small electrodes placed on the scalp. The cassette can record an EEG for twenty-four hours without changing the tape. When the person who is observing the child feels that she is having a seizure, the adult pushes a button, which records a mark on the tape. The physician then compares the EEG tracings with what was observed to see if the reported events are actual seizures.

VIDEO EEG

THE BEST WAY TO ANALYZE POSSIBLE SEIZURE ACTIVITY is to both see and record the events. Video EEG monitoring uses a camera to record movements and behavior while the EEG is also recording. The physician is then able to compare all of the child’s behaviors to see which, if any, are epileptic seizures. These may require treatment to prevent recurrent seizures.

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GENERAL EEG FINDINGS BY STAGE

Stage I

Normal or minimal background slowing

Stage II

Background slowing

Rhythmic theta

Rare epileptiform discharges

Epileptiform discharges may occur before the onset of clinical seizures

Stage III

More background slowing

Abundant epileptiform discharges, mostly during sleep

Loss of developmental features

Stage IV

Continuous epileptiform activity OR

Lack of epileptiform discharges and faster background activity

TYPES OF SEIZURES IN RETT SYNDROME

EPISODIC ELECTRICAL EVENTS CAN OCCUR in different parts of the brain. The type of seizures they produce will differ depending on what area of the brain is affected and the direction and speed as the event spreads. Each type of seizure may require a different medication. Girls and women with RS may experience generalized and partial seizures, but there is not one specific type of seizure seen in RS. Your child may have only one type of seizure or she may have more than one type. It is important to remember that some children with RS never have seizures. Most seizure disorders respond well to medication.

GENERALIZED SEIZURES

TONIC/CLONIC SEIZURES: These seizures have been called “grand mal” and are now referred to as generalized tonic/clonic seizures. When the EEG is recorded during a generalized seizure, the seizure activity seems to start all over the brain all at once. Since the whole brain is involved, the seizure may involve all muscles and motor functions with loss of consciousness. Generalized seizures can be “large” and convulsive, with muscle movements such as jerking or stiffening, or “small” and nonconvulsive with alteration of consciousness but no jerking movements. If she loses consciousness, she may fall and may then have rhythmic jerking of all extremities or stiffening followed by rhythmic jerking. The tonic phase is when stiffening occurs. Since all the muscles are contracted, the chest muscles also contract, and breathing may become difficult. She may cry out because of air rushing out of her lungs, but she is not in pain. Lack of oxygen causes a bluish tinge around the lips and face. Saliva may cause a gurgling sound in the throat. The jaw becomes tightly clenched. The clonic phase then begins with rhythmic jerking and tightly clenched fists. The arms, legs and head may flex and then relax. This usually lasts no more than a few minutes. When she regains consciousness, she may be sleepy for one or two hours afterward. After rest, she should be able to go back to her usual activities.

Absence Seizures: This type of seizure, which has been called “petit mal,” starts suddenly. It is characterized by brief staring spells without a preceding warning. There may be

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head-bobbing or eye blinking. Absence seizures generally last just a few seconds and end as abruptly as they began. Afterward, she returns to her usual activity without a period of confusion or sleepiness. Absence seizures can be confused with complex partial seizures because they both involve staring. It is important to differentiate the type of seizure to determine the type of medication that will work best.

Myoclonic Seizures: Formerly called minor motor seizures, these events consist of abrupt jerks of muscle groups and involve brief, sudden, twitchlike movements of one or more extremities. They may take many forms. A foot may kick out, a hand may fly forward. Myoclonic seizures may arise from deep structures in the brain stem that control tone and posture, causing an abrupt increase in a muscle group that brings about a sudden movement of that part of the body. However, myoclonic jerks are not always seizures. When falling asleep, most healthy individuals experience a sudden jerk and awaken with a startle. This is a normal sleep phenomenon.

Akinetic/Atonic Seizures: This type of seizure is as sudden as a myoclonic seizure. However, it is characterized by a sudden loss of tone or posture. If standing, the child may suddenly become limp and drop to the floor. If sitting, one may simply see loss of tone with the head falling forward or backward. These seizures are brief. The child returns to her usual activity immediately afterward. These seizures may also be referred to as “drop attacks.”

PARTIAL SEIZURES

THE SEIZURE IS CALLED PARTIAL when at the beginning of the seizure the electrical disturbance is limited to one part of the brain. The electrical disturbance may spread to involve the whole brain. If this occurs, the seizure has become secondarily generalized.

Simple Partial Seizure: These seizures may involve movement with rhythmic jerking of one extremity of one side of the face or body, or they may involve the senses, with a particular tingling, burning or abnormal sensation in any part of the body. There is no alteration of consciousness.

Complex Partial Seizure: During this type of seizure there is some alteration of consciousness. The child is unable to make meaningful responses or her usual responses. The seizures may be characterized by confusion, loss of alertness, and staring episodes, either alone or combined with automatic behavior such as picking at the clothes, smacking the lips, or random nonpurposeful movements of the arms or legs.

RECOGNIZING A SEIZURE

IT IS SOMETIMES DIFFICULT TO TELL THE DIFFERENCE between autonomic responses associated with RS and seizure activity. Some breath-holding episodes, cyanotic spells (turning blue), jerky tremors, inattention, and eye-rolling movements ordinarily associated with seizures are not seizures in RS. You should observe the sequence of events that occur. For example:

- Is breath-holding followed by jerky movements or vice versa?
- Observe the movements. Are they rhythmic or random?
- How long does the episode last?
- Does she sleep afterward?
- Observe her eyes. Does she stare vacantly or have eye rolling? Eye deviation?
- Does her head drop?

Report these observations to your child's physician.

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All of these events have been observed during breath-holding episodes as well as during seizures. During a breath-holding spell, the child does not breathe, turns blue (cyanotic) or white (pallid) and may lose consciousness. Sometimes, breath-holding spells result in a seizure. Breath-holding spells are not seizures by themselves and are not serious.

It may be necessary to obtain an EEG with video monitoring of the breathing or movement pattern to establish whether or not the events are seizures and need to be treated. This is very important to establish, because in addition to their beneficial effects, all AEDs have potential side effects.

In Ashley's case, severe breathing episodes and seizures do have some similar characteristics, but generally are quite different. Her eyes are a big clue to seizures. Her eyes will freeze, and have been known to roll upward, or in one direction only. In breathing episodes, her eyes are free to move around. Ashley's face becomes quite flushed during seizures. There is more movement in her arms during seizures. There is a tightening during breathing episodes, but not as much movement as there is in seizures. Ashley appears frightened or stressed during seizures. I do not notice this during severe breathing episodes. Also, her lips will often turn blue in a seizure but not in a breathing episode. She has had severe apnea where her lips will turn blue, but these episodes do not resemble a seizure.

Karina has this sort of shaking her body, grimacing her face and stiffening and jerking her arms and legs. It lasts one or two minutes, and then she relaxes again. She tends to bend over forward when she is walking, and sideways when she is sitting. It happens frequently, many times a day, but some days more than others. She started this when she was about eight years old. It is not epilepsy. She has that too, and it definitely is different.

We took video of a student during her "seizures" and the neurologist said most of what we were seeing was "involuntary motions" dealing with a breathing problem. This girl looks like a falling tree when she has these involuntary movements. Her seizures are a little different.

If too much comes at Dani too fast or if we leave the TV on with no lights and the light from the TV blinks, she will have a seizure. She is sensitive to loud noises too.

Brit has had seizures since four months, but they have never been evoked by sound, visual or tactile stimuli. I remember this because we could take her anywhere and we would play games and scare her like "boo" and hide around a corner and she would giggle. Now, however, ripping a paper towel off in front of her has given a seizure, along with a glimmer of sunlight on a therapist's watchband, even a cough or sneeze. Closing the car door is hard to do quietly. Going over the bumps in the sidewalk triggers a seizure as well. She began this hypersensitivity at age five and a half.

WHAT TO DO WHEN A SEIZURE OCCURS

IT MAY BE HARD TO STAY CALM during your child's seizure, particularly the first time. Most parents get to be pros at it over time. Once a seizure has begun, there is actually little you can do to stop it. You do not need to do CPR. There is nothing you can do to stop the stiffness or start the breathing. Mouth-to-mouth resuscitation will not work because her chest will not expand. You can gently support her.

During the seizure

- Do not put anything in her mouth
- Do not restrain her
- Do not call an ambulance unless the seizure continues for more than five to ten minutes

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- Do try to put her on her side
- Do put something soft under her head
- Do loosen tight clothing around her neck
- Do remove sharp objects from the immediate area

After the seizure

As the seizure stops, she will usually let out a sigh and go into a deep sleep, known as the “post-ictal” period. This period may vary depending on the length of the seizure, but it usually lasts from ten minutes to a couple of hours. If she wants to sleep long, it is OK. Sleep is a healthy way for her brain to recover from the overexertion.

- Do stay with her until she is awake and alert
- Do be comforting and reassuring
- Do allow her to return to activities if she is all right

WHEN TO CALL FOR HELP

IF THE CLONIC (JERKING) PHASE OF THE SEIZURE lasts more than five to ten minutes, it is advisable to call an ambulance. Check with your child’s own neurologist to determine how long you should wait.

MEDICATIONS

EEG ABNORMALITIES ARE COMMON IN RS. The goal is to treat the seizure disorder, not the EEG. If your child has an abnormal EEG but does not have a history of seizure like activity, she does not necessarily need to take anticonvulsant medication. Even some individuals who have no neurological problems and never have seizures may show abnormalities on the EEG at some time. In research studies of RS, nearly all girls with RS showed an abnormal EEG, while only from one-third to one-half of them had epileptic seizures.

If she does have seizures, there are many different AEDs are used for specific seizure types. There is not a specific medication for the treatment of seizures in RS. A pediatric neurologist needs to know what type(s) of seizure a child is having before prescribing medication. The primary objective of drug therapy is to control the seizures with the least possible drug side effects. Fortunately, AEDs are generally safe and severe side effects are rare or very infrequent. All side effects should be reported to your physician. Any change in behavior, including slowing (oversedation), hyperactivity, lack of coordination, or other behaviors of concern to you are appropriate to bring to the physician’s attention.

Either we don’t medicate and Jenn has lots of seizures and is “out of it” most of the time, or we medicate and her seizures are minimized but the trade-off is the side effects and her not being “with it” some of the time. Nothing’s perfect and I can’t expect the impossible. I know there’s an adjustment phase with new medications as the body needs time to learn how to work with them.

There are many drugs to prevent and treat seizures, but it is not entirely clear how or why they work. We do know how these drugs are absorbed and metabolized, and we know about their side effects. Some drugs are more effective than others for different seizure types.

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Once the seizure type has been identified, the choice of drugs is made on the basis of the drug's effectiveness, cost, the child's age, and other drugs she may be already taking and any drug allergies or sensitivities.

The objective of AED therapy is to use the lowest dose of drug with the least number of side effects to achieve the best seizure control. Sometimes higher doses are necessary, and more than one drug may have to be used to achieve seizure control. It is important to remember that all drugs, including aspirin, have side effects along with their beneficial effects. Side effects may occur during the first few days on the drug until the body adjusts. Some drugs may require dose adjustment or discontinuation if the side effects are unpleasant.

1. The *Benzodiazepines* (Valium, Klonopin, Tranxene, Versed, and Ativan) are grouped together as a class of AEDs: The benzodiazepines potentiate the neuroinhibitory effect of the GABA neurotransmitter. Valium (diazepam), Ativan (lorazepam), and Versed (midazolam), are used to treat status epilepticus (discussed later). Klonopin, Tranxene, Valium and Ativan are also used in long-term treatment of seizures. The drugs in this class are useful for absence seizures, but are most effectively used to treat myoclonic and atonic seizures. Since each of these drugs can cause drowsiness, irritability and hyperactivity, they are usually chosen as "add-on" drugs when other drugs do not bring the seizures under control by themselves. The body can develop tolerance for drugs of this type, so the dose may need to be increased to maintain a therapeutic effect. Diastat or properly formulated generic Valium may be used per rectum by parents to interrupt prolonged seizures (ten minutes) or serial seizures. The use of rectal diazepam (Diastat) has proven to be safe when used properly and has helped reduce the number of ER visits. This drug can be given at home by the parents as directed by their child's physician.
2. *Carbamazepine* (Tegretol) is used for simple and complex partial seizures and generalized tonic/clonic seizures. It does not interfere negatively with behavior and learning and has no cosmetic side effects. Tegretol should be started at a low dose and increased each week for the first several weeks until the appropriate therapeutic blood level is reached. Side effects include drowsiness, dizziness, blurred vision, lethargy, nausea/vomiting, lack of coordination, decreased white blood count, and decreased platelets. One good side effect of Tegretol is elevation of mood. Tegretol may make atypical absence worse.
3. *Ethosuximide* (Zarontin) is used for akinetic/atonic seizures and is most valuable in treating generalized absence seizures. It has no effect on partial seizures. Side effects include drowsiness, dizziness, GI upset, headache, hiccough, hyperactivity, and nausea/vomiting. Zarontin can cause allergic reactions, but these complications are very rare. In most cases, it is safe, well tolerated, and effective.
4. *Gabapentin* (Neurontin) is a well-tolerated "add on" medication for partial complex and generalized tonic/clonic seizures. It does not interfere with other medications, so it is helpful for those girls taking multiple drugs. It has a short duration of action necessitating multiple daily doses. The most common side effects are sedation, fatigue, dizziness, ataxia, nystagmus, headache, nausea, and weight gain.
5. *Lamotrigine* (Lamictal) is used for the treatment of partial and generalized seizures as a primary drug or as an "add on" drug, used in combination with other drugs. It should be used cautiously with Depakote/Depakene® because of a possible likelihood of hypersensitivity reaction (skin rash). It is useful for patients who cannot control their seizures adequately with current medication or who experience unacceptable side effects. Side effects include skin rash, dizziness, headache, double vision and unsteadiness.

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Interactions with other antiepileptic drugs may alter its duration of action. From available studies it has been concluded that oxcarbazepine and lamotrigine do not affect cognitive function in healthy volunteers or in adults with newly diagnosed epilepsy.

6. *Levetiracetam* (Keppra) is approved by the Food and Drug Administration as adjunctive treatment of partial seizures in adults. Effectiveness in generalized seizures has been reported. It has been successfully used in children. Side effects include somnolence and dizziness. Behavior problems are reported and may necessitate discontinuation.
7. *Oxcarbazepine* (Trileptal) is chemically related to carbamazepine. Oxcarbazepine is approved for the treatment of partial seizures in adults and children aged 4 years and older. Side effects associated with this antiepileptic drug include somnolence and ataxia. More severe side effects such as liver dysfunctions seen with carbamazepine are not reported for oxcarbazepine.
8. *Phenytoin* (Dilantin) is used for tonic/clonic and simple and complex partial seizures. This drug can cause allergic reactions, so if a skin rash develops after the first two to three weeks, the child should be seen by the physician immediately. Dilantin may cause mood changes and lethargy. Overgrowth of the gums reportedly occurs in about half of the children who have therapeutic blood levels. When taken over a long period of time, Dilantin can cause the development of coarse facial features and more extensive growth of body hair. Side effects include tremor, anemia, loss of coordination, double vision, nausea/vomiting, confusion, and slurred speech.
9. *Phenobarbital* (Luminal) is used for tonic/clonic and simple and complex partial seizures. Phenobarbital is ineffective for absence seizures. Since it is metabolized slowly, it is usually given only once per day. Allergic reactions are possible, so the child should be observed carefully for skin rashes. The most important adverse side effects are in behavior and learning. Other side effects include drowsiness, lethargy, and hyperactivity, which can result in changes in behavior and learning.
10. *Primidone* (Mysoline) is used for tonic/clonic and simple and partial complex seizures. It is metabolized by the body into phenobarbital. The child should be carefully observed as the drug can cause hyperactivity and behavior problems. To avoid sedative effects and personality changes, Mysoline must be started at a low dose and increased very slowly over several weeks. Side effects include drowsiness, appetite loss, irritability, nausea/vomiting, dizziness, and loss of coordination.
11. *Tiagabine* (Gabatril) appears to be an effective antiepileptic drug for partial seizures but may exacerbate certain types of generalized seizures. Its use is limited by short duration of action, decreased rate of absorption when given with food, and lower levels in the evening than in the morning. Side effects include generalized weakness, cognitive disturbances, and depression.
12. *Topiramate* (Topamax) is used for partial or partial complex seizures and generalized seizures. It may also be useful for difficult-to-control generalized seizures (atonic) and infantile spasms. When Topamax is combined with Dilantin®, the dose of Dilantin may need to be increased. If Dilantin or Tegretol is added or withdrawn, the dose of Topamax may need adjustment. Side effects include lethargy, agitation, headaches, drowsiness, lack of coordination, nervousness, dizziness, and arm/leg tingling. At higher doses, loss of appetite and weight loss can be seen. Side effects include confusion, psychomotor slowing, and difficulty with concentration. It is important to observe closely for the risk of kidney stones and maintain adequate hydration. Oligohidrosis (decreased sweating),

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infrequently resulting in hospitalization, has been reported in association with Topamax use. Decreased sweating and an elevation in body temperature above normal characterized these cases. Some of the cases were reported after exposure to elevated environmental temperatures. The majority of the reports have been in children. Patients, especially pediatric patients, treated with Topamax should be monitored closely for evidence of decreased sweating and increased body temperature, especially in hot weather. Caution should be used when Topamax is prescribed with other drugs that predispose patients to heat-related disorders; these drugs include, but are not limited to, other carbonic anhydrase inhibitors and drugs with anticholinergic activity.

13. *Valproic acid* (Depakene) and *Divalproex Sodium* (Depakote) are used for tonic/clonic, absence and myoclonic, and simple and complex partial seizures. These two drugs seem to work better after the child has taken them for a couple of weeks. When either of these drugs is stopped, it continues to work for several weeks. Each of these are very safe drugs, but should be used with caution in children under the age of two, and preferably alone. Side effects include nausea/vomiting, indigestion, sedation, dizziness, hair loss, tremor, lack of coordination, weight loss and/or gain, and changes in liver function.
14. *Zonisamide* (Zonegran) has been used for the treatment of a broad spectrum of seizure types including generalized seizures (tonic/clonic, absence, infantile spasms and myoclonic seizures) and all partial seizures. Side effects include somnolence, cognitive problems, and anhidrosis (lack of sweating).

The medication should never be stopped abruptly. Your child should continue with her medication even if she has not had a seizure for quite a while. She may continue to be at risk for having seizures. She will not become dependent on the drug; anticonvulsant medications are not addicting. If she can keep control of the seizures for a number of years, she may be able to be taken off these medications without recurrent seizures. But stopping the drug must always be done slowly and with a doctor's careful supervision.

Over time, we built Jenn up to dosages of 800 mg Tegretol, 200 mg Phenobarbital, and 20cc Zarontin in one day just to control the seizure activity. Now, Jenn needs only 5cc of Tegretol per day.

GENERIC OR BRAND NAME

ABSORPTION RATES OF ANTICONVULSANT MEDICATIONS may vary with different manufacturers and there may also be some difference in their metabolism. This may cause varying blood levels, which may either allow seizures to occur or lead to toxicity. Using brand name drugs instead of the cheaper generic forms helps avoid these problems. And, always stick to the brand made by the same manufacturer.

VITAMINS AND CALCIUM

THE ADDITION OF VITAMINS TO A BALANCED DIET is of no value in the treatment of seizures. There is concern that AEDs may effect calcium metabolism and bone mineralization. This is not proven for RS. Supplementation with oral calcium should be done under the supervision of a physician.

MEASURING DRUG LEVELS

THE TERM "DRUG LEVEL" REFERS to the amount of medication in the blood. Drug levels are usually measured two to three weeks after a new drug is started or the dose changed. The amount of drug in the blood will indicate if she is in a therapeutic range and

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make sure her dose is not toxic. If there is toxicity, especially when she is on more than one drug, the blood test can help determine which drug should be decreased. In general, if the child seems to be doing well, blood levels are repeated once or twice a year.

OTHER TREATMENTS

KETOGENIC DIET

SPECIAL DIETS MAY BE RECOMMENDED by your daughter's physician as a last resort when anticonvulsants have proven ineffective. The ketogenic diet, one of the oldest treatments for epilepsy, is one which is very high in fat. The diet provides the minimal amount of protein necessary for growth and virtually no carbohydrates. Most of the calories consumed come from fat, using butter or cream. It is a very restrictive diet and can be difficult to begin and maintain. The diet simulates the effects of prolonged starvation by causing the body to burn fat rather than carbohydrate as its main energy source. The ketone bodies, which are the result of this fat metabolism, are utilized as an energy source by the brain. Why this results in improved seizure control is unknown.

The diet begins with several days of fasting. *The ketogenic diet must never be attempted except under strict medical supervision* and by a team which can provide support for the family during the difficult period of adjustment after the diet is initiated. The diet can be dangerous if not done properly. Food on the diet is strictly limited and not always appealing. A typical meal might consist of a small amount of meat, fish, poultry or cheese, a serving of fruit, an additional serving of fat such as butter or mayonnaise and a serving of heavy whipping cream. While some say the family ends up eating in the closet and the diet is too restrictive, others say the effort is worth it when seizures are under control for the first time. The diet should be given a four to six week trial with ketosis well maintained. If seizure frequency is significantly reduced, the diet can be maintained and an attempt try to reduce and stop AEDs made.

Katie has been on the ketogenic diet for almost two years now with wonderful results. However, when she gets sick, she can sometimes have breakthrough seizures.

Potential side effects include reduction in bone mass, kidney stones and thinning of the hair. Increases in plasma lipids have been reported, but their significance is not clear.

Sugar-free supplements of multivitamins and calcium need to be provided. Medicines such as antibiotics and daily used substances like toothpaste must be monitored for carbohydrate/sugar intake.

The majority of children with few seizures will achieve seizure control with one medication. When the first medication fails to control the seizures, a second medication is added. The ketogenic diet should be considered only for children who have more than two seizures per week despite treatment with at least two different anticonvulsant medications. The diet may also be used when the frequency of seizures, despite medications, interferes with the child's daily function, or when serious adverse side effects result from medications.

The decision about whether to use the ketogenic diet should be made by informed parents and their physician. The diet is not a cure-all, nor is it intended for everyone with epilepsy.

A form of diet less disruptive to family eating is the MCT diet, in which a special oil is added and she can eat most foods. Still, at least sixty percent of calories must come from fats and carbohydrates must be limited.

Seizures

Meghan had amazing success on the diet. Her seizures were cured from it—no meds, normal diet now. It wasn't easy but it was worth it. Meg didn't seem to mind it too much. She wasn't happy to give up McDonald's nuggets, but she did OK. It can be very tough for a child who is able to feed herself, but I think sometimes as a parent you have to look at the big picture and realize the two years of restrictions and unhappiness are worth it in the long run if you get a cure. You will know in a couple months if it will work.

We did the ketogenic diet for two and a half years. It gave Angela a ninety-five percent improvement in seizure control. Angela developed a rare side effect on the diet that forced her to discontinue. But were it not for that, I'd have her back on the diet in a heartbeat. It isn't easy, it is stressful, it's a huge commitment, but for all that, it is very worth the benefits we saw. We got to have our girl "back" for a whole year and it was probably the happiest time in our lives.

VAGUS NERVE STIMULATOR

THE VAGUS NERVE STIMULATOR (VNS) is a device that is implanted under the skin of the chest with wires that wrap around the left vagus nerve in the neck. It is used to control seizures that are unresponsive to traditional anticonvulsant medications. The VNS sends electrical signals that are applied to the vagus nerve in the neck for transmission to the brain. The vagus nerve is one of the primary communication lines from the major organs of the body to the brain. When parents, caregivers, or teachers sense that a seizure is imminent, they pass a magnet over the VNS generator in the chest, which activates a painless electrical stimulation, thus interrupting the seizure. The most common side effects include hoarseness, a prickling feeling on the skin, shortness of breath, and increased coughing. These side effects diminish over time. As with any surgery, there is a risk of infection. Benefits include reduction in seizures and reduction in AEDs.

Julianna's VNS was turned on and she didn't appear to feel a thing. When the neurologist stimulated her for the second time and tried the magnet on her, she didn't even flinch. I can't even tell when it is going off.

Desire had no trouble at all with the surgery and went home the same day. They waited two weeks after surgery to turn it on. The only negative side effect was mild coughing when we used the magnet, but that stopped after the first two weeks. The other side effects were increased energy, increased focus, and the return of her personality. It was pretty awesome. When we had the implant done, Desire was averaging between fifty to seventy small seizures a day and a seizure requiring Diastat every other day. After all of the adjustments, she has now been seizure-free for two weeks.

The VNS is helping Alexis. It is not a quick fix and it takes some time for it to work. I do see a difference in the amount of seizures a day; they are much less than before the implant. Alexis still has one or two through the night. I would tell anyone considering the VNS that it is a personal choice for family and your daughters. For us, it has been a roller coaster ride so far. It takes a great deal out of us caring for her, but it is more the seizures causing the care and the problems, not the VNS. The better side is less seizures and no drugs, so that is a good thing.

STATUS EPILEPTICUS

A SEIZURE THAT LASTS A VERY LONG TIME (thirty minutes or more) is referred to as status epilepticus (SE). "Convulsive status epilepticus" refers to tonic/clonic seizures, and "nonconvulsive status epilepticus" refers to an episode of absence spells, staring spells, or periods of confusion that last for more than a half hour. In most children who have SE, the cause is not known. The most common cause of convulsive status in a person who already has seizures is a blood drug level which is too low to control seizures.

Seizures

Status epilepticus may occur from missed doses, interaction with another drug that has interfered with the drug's effectiveness, or substitution of a generic drug that is not well absorbed in the bloodstream.

Very prolonged SE can cause brain damage. However, it may not be the seizures themselves, but the underlying cause of the seizures, such as infection, trauma, or tumors that may cause brain damage. Most children with SE recover without significant new deficits.

When convulsive SE occurs, it is important to bring the seizures to an end as quickly as possible. Most likely, this will take place in the emergency room of the hospital. Here, she will be observed, blood will be drawn, and oxygen will be given if needed. She will probably get an IV (intravenous line) to supply fluids and to give AEDs into the vein if it becomes necessary. This is the fastest way to get medicine to the brain where it is needed. A number of medications may be used, but Ativan and Valium, which are quick-acting AEDs, are usually given first. Their effectiveness may wear off quickly and another seizure may occur. An additional drug such as phosphenytoin (IV Dilantin) is then given, which is slower to start acting but lasts longer.

Most of the time, the SE can be controlled within a half hour to an hour of arrival at the hospital. In prolonged cases, it may be necessary to give large doses of medication or general anesthesia, which usually stops the seizures. While these episodes are frightening to watch, most children recover well and do not have lasting damage, even from prolonged seizures.

When nonconvulsive SE occurs, it is more difficult to detect because it does not involve body movements. The child may seem detached or just "not herself." The only way to know if nonconvulsive status is happening is to do an EEG, which will show constant spike-wave abnormalities. It is treated with AEDs given into the vein, and the child then returns to her normal state.

FINAL THOUGHTS

The goals of treating seizures include:

1. To identify epileptic seizures and to distinguish them from non-epileptic behaviors.
2. To prevent further seizures, especially prolonged or frequently recurring seizures, in order that the individual can enjoy the best quality of life, and do all that she is capable without untoward side effects from the treatment, particularly oversedation, adverse behavior, or cognitive dysfunction, and motor impairments.
3. Use the most effective treatment with the least side effects and expense.
4. Maintain her seizure free for a period of time sufficient (often two-three years) to be able to discontinue (slowly) the treatment and have her remain seizure free.
5. To improve not only the quality of life the girl with RS but also that of her family.

"I exist as I am, and that is enough."

— WALT WHITMAN