

-From the Board-

IT'S BACK TO SCHOOL TIME AGAIN, a time that is never easy for any parent. By the time you hunt down the requested school supplies, spend hours picking out new school clothes and finding the perfect backpack, you're exhausted. But for those parents who have a child with LGS, they are often faced with extreme anxiety and questions; will the teacher understand my child's needs? Will they want to learn about LGS? Will I have enough time to explain everything about the disorder and the special needs that go with it?

The key to this change is communication. It is imperative that communication starts prior to the school year and continues throughout. It is not uncommon for teachers and school administrators to be unfamiliar with Lennox-Gastaut syndrome, including the diverse characteristics of the disorder. Therefore, you may be obligated to "educate the educators" on the disorder and what they can expect from your child in an array of areas. It is also crucial to review your child's IEP and highlight goals and issues that the teacher should be aware of immediately, as well as your child's health care plan.

As a parent, you may receive mixed reactions from teachers in regards to the LGS diagnosis. You will find that some teachers have been supportive and asked for more information to learn as much as possible while others you encounter seem scared by the prospect of numerous seizures and behavior issues. In any case, keeping those open lines of communication is crucial to understanding, acceptance and peace of mind for all involved.

To download the LGS Foundation's "Guide to LGS" to distribute to your child's teacher, please visit:

<http://www.lgsfoundation.org/education.html>



National Month-Long Campaign Set to Launch October 6th with a Grand Finale Celebration Set for November 5th in New York City

Atonic [drop-attack] seizures are often synonymous with Lennox-Gastaut syndrome. They are one of the most devastating types of seizures which are caused by a sudden burst of electricity to the brain and loss of muscle tone, often resulting in a dangerous fall. Children are more commonly affected by drop-attacks than adults, and are usually required to wear a helmet or protective headgear for their safety.

Helmets for Hope is a campaign developed by the LGS Foundation to raise awareness of severe epilepsy disorders, including those which are characterized by drop-attack seizures. One of the main objectives of the campaign includes gathering support from professional athletes who wear helmets in their careers, in an effort to help erase the stigma associated with children who are required to wear helmets in their daily lives. Other objectives of the campaign include raising funds for epilepsy research and programs to benefit families affected by catastrophic or severe forms of epilepsy.

The Helmets for Hope campaign is set to launch on October 6th, 2009 and will run for a one-month long period with a grand finale celebration and fundraiser set for November 5th. The finale will be held at *The Providence*, 311 West 57th St in New York City, and will be co-hosted by the American Epilepsy Outreach Foundation.

For more information on the campaign and the event fundraiser, including tickets and sponsorship opportunities, please go to www.lgsfoundation.org/helmetsforhope.html.

In the News

NON-SURGICAL NERVE-STIMULATION DEVICE PROVES EFFECTIVE AGAINST INTRACTABLE EPILEPSY

In a recently completed clinical trial, a unique nerve-stimulation treatment for intractable epilepsy reduced the number of seizures by more than 50 percent. In the March 2009 edition of the journal *Neurology*, UCLA neurology professor Christopher M. DeGiorgio and colleagues report the results of the long-term pilot trial, which demonstrated the effectiveness of the new treatment, called trigeminal nerve stimulation (TNS).

The results, though preliminary, are very encouraging, DeGiorgio said. Those participating in the trial for three months saw a 66 percent reduction in the number of seizures, those participating for six months saw a 56 percent reduction and those who completed one year saw a 59 percent reduction in seizures. One of the subjects who participated for a full year had a 90-percent reduction in seizures.

The trigeminal nerve extends into the brain from the face and forehead and is known to play a role in seizure inhibition. The stimulator, about the size of a large cell phone, attaches to a belt or can slip into a pocket. Two wires from the stimulator are passed under the clothing and connected to electrodes attached to the forehead by adhesive. The electrodes, which can be covered by a cap or scarf, transmit an electrical current to the nerve.

"People with intractable epilepsy who have continuing seizures are often drug-resistant," DeGiorgio said. "In addition, anti-seizure drugs can have significant side effects on thinking and alertness."

Epilepsy brain surgery can be very effective, he said, but some patients are not ideal candidates because there is no single focal point in the brain for their seizures.

A larger clinical trial to further test for safety and effectiveness is now underway. The investigators hope that eventually a device can be permanently implanted above the eyebrow that would stimulate the trigeminal nerve and replace the external device.

"TNS is a promising alternative mode of neurostimulation because the trigeminal nerve can be stimulated in minimally invasive fashion," DeGiorgio said. "The major branches of the trigeminal nerve in the face are located close to the surface of the skull; that allows physicians to assess response prior to permanent implantation of a future device."

"For all of these reasons, we need to find non-drug and non-surgical alternatives," he said. "The results of our study are very encouraging and support further investigation into the safety and efficacy of TNS."

SINGLE GENE MUTATION RESPONSIBLE FOR "CATASTROPHIC EPILEPSY"

Catastrophic epilepsy - characterized by severe muscle spasms, persistent seizures, mental retardation and sometimes autism - results from a mutation in a single gene, said Baylor College of Medicine researchers in a report that appears in the July 8th issue of the *Journal of Neuroscience*.

The BCM department of neurology team replicated the defect in mice, developing a mouse model of the disease that could help researchers figure out effective treatments for and new approaches to curing the disease, said Dr. Jeffrey Noebels, professor of neurology, neuroscience and molecular and human genetics at BCM and director of the Blue Bird Circle Developmental Neurogenetics Laboratory at BCM, where the research was performed.

"While many genes underlying various forms of childhood epilepsy have been identified in the past decade, most cause a disorder of 'pure' seizures," said Noebels. Why some children have a more complicated set of disorders beginning with major motor spasms in infancy followed by cognitive dysfunction and developmental disorders such as autism remained a mystery until the discovery by the BCM team that a mutation in only a single gene explains all four features of catastrophic epilepsy.

A gene known as Aristaless-related homeobox or ARX has a specific mutation called a triplet repeat, which means that a particular genetic (in this case, GCG) is repeated many times in the gene. When the researchers duplicated this particular mutation in specially bred mice, the animals had motor spasm similar to those seen in human infants. Recordings of their brain waves showed that they had several kinds of seizures, included absence epilepsy and general convulsion. They also had learning disabilities and were four times more likely to avoid contact with other mice than their normal counterparts. This behavior is similar to that seen in children with autism or similar disorders in the same spectrum.

"The new model is an essential tool to find a cure for the disorder," said Noebels.

"Mutation of the ARX gene was previously known to affect interneurons, a class of cells that inhibit electrical activity in the brain," said Dr. Maureen Price, the report's lead author and an instructor in neurology at BCM.

When researchers evaluated the brains of the adult mice with the mutated gene, they found that a special class of interneurons had never developed in specific brain regions.

"Further study will allow use to pinpoint which brain region is linked to the autistic-like behavior," said Price.

Two members of the research team - Dr. James Frost, professor of neurology at BCM, who developed the concept of the special mouse, and Dr. Richard Hrachovy, also a professor of neurology at BCM - are pioneers in the study of human infantile spasms.

"At present there is no proven cure to offer children with this specific epilepsy", said Noebels. "We now have new clues into the mechanism and have already initiated studies with a new class of drugs not previously explored for this disorder." The new drug testing is supported by the private foundation People Against Childhood Epilepsy.

LGS Patient Spotlight

Our daughter Angel Dominica was born in 1979. She was delivered by C-section after a 55 hour labor, which her neurologists have suggested caused the LGS. Her Apgar scores were fine, and she developed normally until March of 1982 when she had her first grand mal seizure. Except for chronic ear infections and the placement of "tubes" in her ears, she was, and still is, a very healthy child.

After that first seizure, she continued to have all the types of seizures that Lennox-Gastaut patients go through. Watching your child have hundreds of seizures a day is heart wrenching. I think only LGS parents truly understand the depth of emotional pain we go through.



ANGEL

Angel has had various injuries due to falls from the seizures, since we never know when a seizure will happen. The common thing about this syndrome is the unexpected. Angel has been on many types of seizure medications. As we all know with Lennox-Gastaut syndrome, the medications do not always control the seizures, and we have learned to accept a certain amount of seizure activity in exchange for Angel being able to function well. She has been on Felbatol and Depakote for many years now, and this combo has worked well.

Angel had the Vagus Nerve Stimulator placed in her upper chest when it was new on the market. Unfortunately, it never controlled the seizures, and due to her petite size, the device caused her constant pain. The VNS was removed about 6 months after insertion, and the copper wire remains in her neck. Around age 23, Angel began to display behavioral changes not consistent with her usual sunny personality. As I research more about LGS, I see that the behavior, not for the better, can be "normal" for LGS patients.

She is currently in a day program for developmentally disabled adults, which is scheduled much like a typical school day. She is able to choose classes ranging from computers to ceramics, wood shop, organic gardening, music appreciation and more. We feel fortunate to live in California, a state which has wonderful programs for our kids.

I am the main caregiver for Angel, and have not been able to work outside the home, as I am continually called by her school due to the seizures. All of our choices, such as where we live, where we work (including health insurance), what type of vehicle we drive, etc, are based on what is best for Angel first. Our focus is to protect Angel, and give her the best possible life.

So far, this 27 year journey of continual seizures has been something that we never could of done, or wanted to do, without our strong faith in a loving God. Every day is different, and I expect the unexpected dealing with this disorder. With the constant, abiding love of my husband, I am able to forge ahead every day.

-Written by Gail, Angel's mother.

First Annual Wine Reception Fundraiser A Great Success!

On May 29th 2009, the LGS Foundation held their first Annual "Wine Reception Fundraiser" at Martha Clara Vineyards, located on the North Fork of Long Island, NY.

The event would not have been a success without the support from all of the individuals who attended, as well as the numerous local businesses who donated products to help raise funds for the LGS Foundation in the silent auction.

A special thank you to Wendy and Anthony Flammia and Jennifer Ruta who spoke about their experiences for our newest video which was debuted at the fundraiser.

LGS and Art Therapy

Art therapy is a mental health profession that uses the creative process of art making to improve and enhance the physical, mental and emotional well-being of individuals of all ages. It is believed to help the artist resolve problems, increase self awareness, express feelings and manage behavior. Art therapy is also believed to reduce stress, which can consequently lower seizure frequency in children and adults with Lennox-Gastaut syndrome.

Vincent van Gogh, Lewis Carroll, and Michelangelo were all profound artists who lived with a form of epilepsy. It is believed that their disabilities may have played a role in their creative process.

The LGS Foundation is currently developing an art therapy program for LGS patients. To find out more information, please visit us at www.lgsfoundation.org/arttherapy.html.



Van Gogh, "Starry Night" (1889)

Donate

To make a donation to the LGS Foundation, please visit www.lgsfoundation.org/donate.html. Your generous support is greatly appreciated.



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LGS Foundation

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