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Looking into Lennox-Gastaut Syndrome

Announcer:
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Your host is Dr. Charles Turck and will be joined by Dr. Keith Starnes. Here's Dr. Turck now.

Dr. Turck:
Learning difficulties, development delays, and behavioral problems are just a few of the considerable effects children with Lennox-Gastaut Syndrome may experience. And with no cure available, practical and emotional support, along with a deep understanding of this disease are all essential in helping our youngest patients and their caretakers. That's why those topics and more will be the focus of today's discussion.

Welcome to *NeuroFrontiers* on ReachMD, I'm Dr. Charles Turck and joining me is Dr. Keith Starnes, a Pediatric Neurologist at The Mayo Clinic in Rochester, Minnesota. Dr. Starnes, thanks for being here today.

Dr. Starnes:
Thank you so much for having me.

Dr. Turck:
So, how about we start at the beginning, Dr. Starnes: what are the signs and symptoms of a child with Lennox-Gastaut Syndrome, or LGS, for short?

Dr. Starnes:
Yeah, so LGS is a severe form of childhood onset epilepsy and it is associated with a variety of seizure types. Most commonly, these include tonic and atypical absence seizures, but also we commonly see patients that atonic seizures, sometimes called "drop attacks", myoclonic seizures, focal seizures and generalized convulsions. The seizures can be quite frequent and they're very often difficult to control with medications. LGS is usually, although not always, associated with cognitive impairment and particular patterns on the EEG. It normally begins before the age of 8, but it's probably most common between ages 3 and 5. It comprises about 4% of all cases of childhood epilepsy, but because it can be difficult to treat, we do commonly encounter it in our epilepsy practice.

Dr. Turck:
Now, do we know what exactly causes LGS, and if not, what are some of the proposed causes?

Dr. Starnes:
There's not one universal cause; there are several potential causes, which include genetic conditions, structural brain injuries, malformations of cortical brain development and infections, like meningitis. However, no cause is found in up to 40% of cases. Up to a quarter of patients with LGS have a history of infantile spasms, however, that's probably due to common underpinnings that cause both conditions, rather than one condition leading to the other.

Dr. Turck:
Now, Dr. Starnes, how do you go about diagnosing LGS? Are there any particular signs or questions that aid in the diagnosis?

Dr. Starnes:
Yeah, it's usually based upon historical factors, clinical and EEG criteria. In general, it's important to think about LGS in any child that has multiple seizure types and intellectual disability, however, there are no universally agreed upon criteria for the diagnosis. Some have

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been proposed however, and one potentially useful metric is the refractory epilepsy screening tool for LGS or "REST-LGS", for short. This combines some of the criteria that I mentioned before in a more standardized way and it uses a scoring system to identify children who may have LGS. Again, this has not been agreed upon as a universal diagnostic tool, but it can be useful to identify patients. And identifying LGS early is important because there is some evidence that early treatment can alter the course of the disease and improve long-term outcomes.

Dr. Turck:
Now, LGS lies firmly within the province of pediatric neurology, but are there any members of other health disciplines or groups that play a role in the treatment of LGS?

Dr. Starnes:
Yeah, every case of LGS is different, but a team-based approach is very important. Depending upon the underlying causes of the condition and the needs of the individual patient, members of allied health can be very important and play a crucial role. Nutrition is often important for patients when we're considering diet therapy. Other disciplines including child life, nursing, and case workers can be very important for patient and their families as well. And, depending upon the comorbidities and treatment options that we're considering, other medical disciplines including neurosurgery and genetics can be quite important, as well.

Dr. Turck:
For those just tuning in, you're listening to *NeuroFrontiers* on ReachMD. I'm Dr. Charles Turck and I'm speaking with Dr. Keith Starnes about Lennox-Gastaut Syndrome, or LGS for short. So, Dr. Starnes, we were just speaking about how we may establish an LGS diagnosis, and now let's shift our focus and talk about treatment options. What does the treatment landscape look like?

Dr. Starnes:
Y'know, the mainstay of treatment for most epileptic conditions, including LGS, is medications. And with LGS, seizures can be quite difficult to control with medications, and the optimal therapy is uncertain, so medications are often tried, which include Valproic Acid, Lamotrigine, Topiramate, Rufinamide, Clobazam, and Felbamate and many others, but those are the ones that are most often used to treat LGS. None of those have shown any clear superiority over the others. When medications are tried and a patient continues to have seizures, we start to think about other things, and diet, in particular, is one thing that can be considered. Ketogenic diet can be quite effective for reducing seizure frequency in LGS. There's several variations of the diet, but they're all based upon the principal of high fat, moderate protein, and low carbohydrate intake. For the actual ketogenic diet, it usually requires inpatient admission for three to four days to get it started. And it is important to realize that diet therapy is meant to be used as an adjunctive therapy with medications and usually doesn't replace them. Something else a lot of patients are interested in is medical marijuana. Cannabidiol or "CBD", for short, is often used to treat LGS; it has an FDA improved indication for this, and it's been tested in children with LGS and it's shown to be effective for drop attacks. CBD, of course, is derived from the marijuana plant and the pharmaceutical formulation is a purified CBD extract with minimal amounts of THC, which is the psychoactive component. We do often use it with LGS, as I mentioned, but it's important to remember that it is similar to other seizure medications in that it's helpful for some patients and less helpful for others. It also has important interactions with other anti-seizure medications, and it also has the potential to cause side effects. And another treatment option that we sometimes think about in children with LGS is surgery, in particular, corpus callosotomy, which is a surgical separation of the connection between the two hemispheres of the brain, can be very effective in reducing the frequency of drop seizures. Additionally, vagus nerve stimulation, which is a surgically implanted device that delivers electricity to the brain through the vagus nerve as it passes through the neck can sometimes be helpful.

Dr. Turck:
And since there's no cure for LGS, what are some promising research areas; are there any novel treatments on the horizon that you're excited about?

Dr. Starnes:
Yeah, there's several medications that are currently being trialed which are potentially effective. Some of which, might actually be available in the next year or so. And there are some other non-pharmaceutical options under investigation. One that I'm particular excited about is non-invasive brain stimulation, which is emerging as an effective treatment for several neurological conditions and could be effective in a variety of epilepsy syndromes, hopefully, to include LGS.

Dr. Turck:
And finally, Dr. Starnes, as we know, most children with LGS will grow up to have seizures, which can take a considerable toll on a patient's overall quality of life, so what are some resources available for family members and caretakers as they navigate supporting their loved ones with LGS?

Dr. Starnes:

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So, many of my patients find benefit from joining online support groups on various social media. However, two groups that are really helpful are the LGS Foundation and the Epilepsy Foundation. They're both wonderful resources for family members and patients with epilepsy, not only with LGS, but a variety of conditions. And, in particular, it's helpful to use those, I think, for families to connect with other family members who've been affected by LGS, and that can be a really useful resource.

Dr. Turck:
Well, I'm sure that all of this information is going to serve our audience tremendously, and it's a wonderful way to round out our discussion on Lennox-Gastaut Syndrome. Dr. Starnes, it was great having you on the program.

Dr. Starnes:
Thank you so much for having me.

Announcer:
This episode of *Neurofrontiers* was sponsored by Greenwich Biosciences, the US leader in the field of cannabinoid prescription medicines. To access other episodes in this series, visit reachmd.com/neurofrontiers, where you can Be Part of the Knowledge. Thanks for listening!

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