Dr. Joseph Sirven: We have had now the opportunity to hear a detailed presentation about various facets of Lennox-Gastaut from our three terrific speakers today. What I want to do is explore a little bit in a conversational format some of the issues that are often asked by our patients, by the physicians about the condition. I’m going to start with Dr. Sheth. One of things, since this is syndromic, one of the questions that always comes up is when do you put that label? When do you code someone as Lennox-Gastaut?

Dr. Raj Sheth: That's a great question. The way we presented the information is with a very clear-cut triad that occurs somewhere between the ages of three and five. But Joe, you’re bringing up a point that really highlights the issue of recognizing it when it first happens. Remember what we said. We talked of this as a transition often times from infantile spasms. In that transition, it's very difficult to recognize change that may occur.

The first issue I would recommend is actually to think about could this patient have Lennox-Gastaut Syndrome. I think the EEG, if you have the clear-cut EEG tipoff of slow spike in wave, that’s very helpful. But remember, the EEG pattern evolves over time, so you may not see it in the early stages in its full form. You may actually see it more in its evolving form, so be very aware of that.

The second thing is that if you hear that a patient is dropping, as was mentioned earlier, drops really can take place from the Doose Syndrome, and if it isn’t that and the patient is a little older, then it becomes very clear that you may be dealing with the first emergence of Lennox-Gastaut Syndrome.

Dr. Joseph Sirven: Dr. Jarrar, what do you think is the most difficult thing about managing a Lennox-Gastaut patient?

Dr. Randa Jarrar: I think the hardest part is trying to address the child as a whole along with the family, and addressing all the comorbidities that we talked about. Seizures, although they are hard to control, may be the easiest part to address in this syndrome. Whereas you have the whole
family that is severely impacted by the diagnosis, the frequent seizures, the behavioral problems, the prospect of cognitive deterioration down then line.

Then you have the seizures. Then you have the child themselves, their care, what are you going to do with them down the line. Addressing all these things becomes your goal and it is crucial to agree with the family that you’re going to address all these aspects and set the goals so that you don’t set yourself up for failure.

You have to tell them initially that although we are going to try our best to achieve good seizure control, we don’t want to overly sedate the child. We don’t want to overly expose them to adverse effects and at the same time, we want to address their education, their behavior. We want to address your family and how is this impacting your family. And then how do we take this child from these years into adulthood?

Dr. Joseph Sirven: Dr. Vazquez, you did a nice job in presenting the various options, but when it comes to medications, to our readers and viewers, what do you prefer to use in terms of agents for Lennox-Gastaut and perhaps in what order, at least for you personally?

Dr. Blanca Vazquez: So we really live in an exciting time in the treatment of Lennox-Gastaut Syndrome because we have major drugs that are now studied in clinical trials. So rufinamide, clobazam, those are the two new additions to the program. Topiramate, lamotrigine, those are drugs that are in the newer generation of drugs that may have less of a burden of side effects and perhaps a better way to use in polytherapy.

I often choose the first drug based on the seizure type. Do they start with the infantile spasms? Do they need more of a drop attack type of medicine? Is it something that we need benzos for because it’s more of a rescue type of a strategy? My favorites really start with the least likely to have toxicity drug and then combine them.

Very often, the patients have a background of the valproic acid and then we use different drugs added to it, but it doesn’t have to be like that. I think that my main concern when the patient is having very frequent seizures is not having to use a medicine that is going not take us a long time like lamotrigine, but yet that’s a great option. Set yourself at a pace where you can titrate slowly and then get to your goal.

Lennox-Gastaut Syndrome is going to be a very chronic seizure type, you need to make your goals obtainable, and it doesn’t have to happen immediately.

Dr. Joseph Sirven: That’s good advice. Let me bring up a topic that I hate to bring up, and we really didn’t cover it. I’m going to ask you, Dr. Sheth, to help us on what do you tell patients or their family or caregivers about death, about sudden and unexpected death in epilepsy, or for that matter, just mortality as it pertains to the Lennox-Gastaut patient? Could you maybe make some comments there?
Dr. Raj Sheth: Joe, that’s a very important point. In the scheme of things, we’re not supposed to die before our children. In epilepsy, that may not necessarily be true. It think it’s sort of is a very hard conversation to have, when you’re managing seizures that are very difficult to control, to be adding one more burden to this family of telling them about the possibility of death, sudden, unexplained death.

It is something that should be broached. There is always a hesitation when you’ve given the family a very devastating diagnosis, when you’re managing seizures that are very difficult to control, to be adding one more burden to this family of telling them about the possibility of death, sudden, unexplained death.

They’re already terrified of the fact that one day the child is going to have a seizure and is going to be in a position that’s much compromised and possibly pass away. We try to encourage families to be the air under their child’s wing, not to put them in a glass bubble. In that context, it’s often times difficult to really focus in on an issue of this sudden, unexplained death.

On the other hand, it is very important to point out, particularly when it comes to patient with compliance, with trying medication, to really hone in on this issue that seizures and epilepsy may be benign most of the time, but on rare occasion, it can be fatal. It think it helps bring the focus back onto the reason we try to look for seizure control, the reason to try other medications and strategies. It's a very difficult situation.

Dr. Joseph Sirven: Great. Let me bring another situation not to the level of depth, but one that really hits caregivers and families. I’m going to ask Dr. Jarrar, how do you counsel our families about the issue of education, what the expectation is for school, and for that matter, just the rank and file behavioral issues? What do you say about those points?

Dr. Randa Jarrar: Well, like we talked about, the quality of life is very important in these kids. It is very important to realize that they have the cognitive problems early on at the time of diagnosis. Frequently, some of them have it even before you diagnosis them with Lennox-Gastaut Syndrome.

You have to address it in multiple ways. One needs to enlist the help of the school. You need to educate the families that they can ask the school to test their child so that they’re appropriately placed. Also, I think each of these kids should have baseline neuropsychometric evaluation when they are diagnosed so that you can figure out what their capabilities are, what they are able to participate in in school, and to place them appropriately, and also to understand that this is a dynamic process. The cognitive decline can be accumulative over time, so they need to be reevaluated and reassessed at different points in time.

Don’t think that this is a static process. It changes and evolves with time. Also, it’s very important to educate the school about the seizures themselves because you want the school at your side reporting to your how many seizures the child is having, to be familiar with these little atypical absences, the drop attacks, to care because the child may not be completely with it at school at different
points during their treatment depending on how well they’re doing with regards to their seizure control.

Also, injury prevention in the school environment is very, very important. You need to educate the school with regards to the adverse affects of medication so that they can observe them and report them back to you. Then there is the issue of behavior, which can be quite dramatic and very problematic. You can have excellent seizure control, but the quality of life could be poor just because behavioral control is still an issue.

I think in these situations, it’s very important to make sure that you have your child seeing a psychiatrist and a psychologist so that the best person that’s familiar with managing behavioral issues is addressing them.

Dr. Joseph Sirven: That’s very, very helpful. Dr. Vazquez, let me ask you another one of these tough questions. You presented a lot of options for choices of therapy, which included things beyond medications. I guess my question is what order perhaps do you choose to go beyond medication to those options of surgery, stimulation, or diet?

Dr. Blanca Vazquez: Fantastic. The goal, as you can see, is that medication is not everything. This is a comprehensive approach and we’re very lucky to have other options that we can perhaps decrease the burden of medication side effects. I often enlist the family and say this is my option for diet. Some families will say oh, I would love to do that. We can enlist the teachers, the caregivers, everyone involved is going to get educated, and we’ll make the diet happen.

But other families will say there is no way. I have three other kids. Carbs are all over. It’s really not an option. That’s when other options become available. I always look at the surgical option as something that I really want to polish and manage before I present because it might be over promising and you have to be very conscious about not promising something that you can’t deliver. Lennox-Gastaut resective surgery, there is a very small number of patients that we can offer it to. It’s worth looking at and it is very important that you do a comprehensive evaluation so you can study that option.

Each one of these other ways to treat epilepsy in the Lennox-Gastaut Syndrome has a very important role. Always try to look at quality of life. The families really have to be a part of these decisions.

Dr. Joseph Sirven: That’s great advice. Let me get to my final question to the group. As you have a lot of interested individuals, physicians, healthcare professionals, patients, caregivers, we’ll be looking to see this particular site roundtable about information with regards to Lennox-Gastaut. Really, what I’m going to ask you is to give us your big overarching piece of advice or take-home message to those that have been listening through all of the presentations with regards to management and care of these individuals. Dr. Sheth, I’m going to start with you. What kind of pearl advice would you like to leave for the audience?
Dr. Raj Sheth: I think the issue is that sometimes, this comes up from below your radar screen. I think when it presents, it’s really flying low. The American Academy of Neurology has a very important piece in patients that come to seizure clinics for month-in, month-out, year-in, year-out where they actually ask the neurologist to ask the question, is this patient an epilepsy surgery candidate patient.

What I would recommend for your patients is that in patients that have a generalized EEG discharge, to ask the question, and it’s intractable, to ask the question could this possibly be Lennox-Gastaut Syndrome that I’m dealing with? It may not have all the features involved just yet, but that would be important.

Having a diagnosis is very important to the family. It’s very important for management. You may not be able to cure it, but at least there’s a syndrome that gives some perspective as to what to expect longer term.

Dr. Joseph Sirven: Thank you. Dr. Jarrar, your thoughts?

Dr. Randa Jarrar: Well, I think it’s very important not to over diagnose and just like Dr. Sheth said, do not under diagnose because it’s very important to be able to get the diagnosis as early as possible. Another important part of this is to come up with a treatment plan, so that it is clear to the family and to yourself as well what are my treatment goals, so that you can address them one by one, and be able to make sure that the family is part of your treatment plan and is on your side. Try to get the school involved. Try to get all your other healthcare providers involved early on, so that you can achieve the best outcome.

Dr. Joseph Sirven: That’s very helpful. Thank you. And Dr. Vazquez, your final thoughts on this?

Dr. Blanca Vazquez: For physicians trying to treat patients with Lennox-Gastaut Syndrome, you have to individualize based on needs, tolerability, and quality of life. Assemble a team of people that are going to help you, the therapists, the teacher, the family and they are all going to help you make your treatment plans and help you along with the same recommendations that you gave us.

Dr. Joseph Sirven: I want to take the opportunity to thank our terrific presenters today. I hope to everyone who is reading this, listening to this, viewing this, that you find this helpful. Let us know about it on epilepsy.com. Visit the Facebook page. Visit our community site, our blog. Let us know what you like, what you didn’t like or the questions that you may have. We’re ready to answer them. Again, thank you very much for your attention and thank you for our wonderful presenters for fantastic and informative presentations today.