

Improving Quality of Life and Care in Lennox-Gastaut Syndrome: Best Practices for Collaborative, Family- and Patient-Centered Management



CMEO Podcast Transcript

Laurie M. Douglass, MD:

Hello. On behalf of CME Outfitters, I would like to welcome and thank you for joining us for today's educational activity entitled *Improving Quality of Life and Care in Lennox-Gastaut Syndrome: Best Practices for Collaborative, Family- and Patient-Centered Management*. Today's program is supported by an educational grant from Jazz Pharmaceuticals. This activity may include discussions of products or devices that are not currently labeled for use by the FDA [U.S. Food and Drug Administration]. All conflicts of interest have been mitigated.

I am Dr. Laurie Douglass. I'm Chief of Child Neurology at Boston Medical Center. I'm an associate professor of pediatrics and neurology at Boston Medical Center and Boston University Chobanian & Avedisian School of Medicine. And I'm joined by my colleague, Ika Noviawaty. Ika, could you introduce yourself to our audience?

Ika Noviawaty, MD:

Absolutely. My name is Doctor Ika Noviawaty. I'm a clinical associate professor at Boston University Medical School. I'm an Epilepsy Division Chief. Great to be here.

Laurie M. Douglass, MD:

Thank you, Ika. Also contributing powerful insights today to our discussion is Jennifer Griffin, who is the Director of Family Support of the Lennox-Gastaut Syndrome Foundation. Wonderful. Let's begin. Before we do so, however, I just want to inform our audience that we will address each other on a first name basis, as we are all colleagues. And now to frame the discussion today, let me start by reviewing our learning objectives for the program.

So, learning objective one: identify non-seizure features and comorbidities that may impact patients with LGS. And for the second: integrate strategies to address quality of life concerns of patients with Lennox-Gastaut syndrome and their caregivers. And lastly, for objective three: incorporate best practices for long-term planning as part of the transition from pediatric to adult care for patients with Lennox-Gastaut syndrome and their caregivers, including team-based, interprofessional care.

We're going to begin with highlighting the challenges in identifying the non-seizure features of Lennox-Gastaut syndrome. I'm going to begin with a question for the audience. So, the first question is: Which of the following non-seizure symptoms seen in Lennox-Gastaut syndrome do you consider to be the most burdensome? So A, cognitive impairment. B, behavioral issues. C, feeding difficulties. D, mobility issues.

And just thinking about this question, since all persons with Lennox-Gastaut syndrome are not the same, I think it really depends on your perspective and who you're thinking about when you're reading this question. As a clinician, most of the time I hear – from caregivers in particular – the most burdensome for them is the behavior. They sort of manage the other issues a little bit easier, 'cause it's the most disruptive to their home life.

Ika Noviawaty, MD:

I certainly agree with that, especially in adult setting. As the patient's parents started getting older, it becomes more and more of an issue.

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Laurie M. Douglass, MD:

Yeah, I agree. Ika, do you mind just sharing with our audience today what defines Lennox-Gastaut syndrome?

Ika Noviawaty, MD:

Lennox-Gastaut syndrome is a developmental and epileptic encephalopathy characterized by multiple seizure types, a characteristic EEG [electroencephalogram] pattern, and cognitive impairment. Importantly, it is a spectrum, and severity varies widely depending on etiology and also seizure burden. And this is why we certainly need to address Lennox-Gastaut syndrome patients as a holistic treatment and not just aiming to try to control their seizures.

Laurie M. Douglass, MD:

Would you like to comment on the different causes of Lennox-Gastaut syndrome?

Ika Noviawaty, MD:

Yes. Some of the different causes of Lennox-Gastaut syndrome could be genetic, brain injury, trauma, birth complications, infections, some sort of metabolic disorder, brain malformation, or neurocutaneous disorder. But for the most part, some of these patients could also have completely unknown cause and it could come as a surprise for parents.

Laurie M. Douglass, MD:

Thank you. As we dive a little deeper, let's discuss the diagnostic triad that is associated with Lennox-Gastaut syndrome. What do you think is most important for clinicians to remember here, Ika?

Ika Noviawaty, MD:

The diagnosis requires tonic seizures plus at least one additional seizure type and supportive EEG findings. But beyond diagnosis, the triad signals long-term complexity.

Laurie M. Douglass, MD:

Let's pause a little bit on the seizures themselves for a moment, because this slide really anchors the diagnosis. Can you walk us through what's essential for clinicians to recognize here?

Ika Noviawaty, MD:

So I think early recognition of the multiple seizure types and also the characteristic EEG pattern can help with diagnosing the patients early. And the earlier we can diagnose this Lennox-Gastaut syndrome, the sooner we can get them the appropriate treatment, and hopefully the outcome, especially the cognitive outcome and the seizure control outcome, can be better.

Laurie M. Douglass, MD:

Do you mind if I add in a little something here, Ika?

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Ika Noviawaty, MD:

Okay.

Laurie M. Douglass, MD:

From my pediatric experience, what's sometimes missed is if you do not do monitoring overnight, you will miss the tonic seizures. Many times they're subclinical or they're so subtle that unless you're doing that, you're going to miss them. Many times the tonic seizures are not happening during the day and parents don't witness them. So that's just something to keep in mind.

Ika Noviawaty, MD:

That's a great point. High-quality continuous video EEG monitoring certainly helps a great deal.

Laurie M. Douglass, MD:

Well, let's move into the non-seizure symptoms. Ika, what do you think clinicians should understand about cognitive impairment in particular about Lennox-Gastaut syndrome?

Ika Noviawaty, MD:

I think parents need to have a good baseline understanding that the degree of the cognitive challenge could be different in different patients. And I think it is our job as clinicians to be able to prepare them and also equip them with the necessary tools to manage the cognitive impairment side of the care.

Laurie M. Douglass, MD:

Yeah, I agree. And I think a lot of parents think that if seizures are controlled, then the cognitive impairment will improve. And I think over time though, they start to understand that the seizures and the cognitive impairment, though there's some connection, that there's not necessarily a significant improvement as the seizures change.

Ika Noviawaty, MD:

Exactly.

Laurie M. Douglass, MD:

As we talked about earlier, Ika, families often raise behavioral concerns with us. It obviously is a big part of their stress as a caregiver. Could you speak to this a little bit more?

Ika Noviawaty, MD:

So, one of the issues with behavioral challenges, it doesn't happen all the time, but certainly it can be unpredictable and not always directly correlating with the seizure frequency nor the seizure control. And another challenge that we face is it is difficult to refer Lennox-Gastaut patients with behavioral issues, especially the severe behavioral issues, to a psychiatrist. Not all psychiatrists are willing to see Lennox-Gastaut syndrome patients, especially in the adult world.

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Laurie M. Douglass, MD:

I think that's true. It's a little bit better on the pediatric side, but it's still challenging. I think it's one of the things that we're hoping is that we'll continue to educate our colleagues in the mental health field so they feel more comfortable and more equipped to manage these symptoms.

Ika Noviawaty, MD:

Absolutely. And I think having an interdisciplinary clinic in the same center can help too, because certainly the neurologist and the psychiatrist can directly communicate, and we can also coordinate other support staff such as social worker, community health workers to better understand how we can support the family.

Laurie M. Douglass, MD:

Agreed. Motor decline and loss of ambulation can also occur over time, which is increasing the dependence upon their caregiver and risk of injury. It really reinforces the need for proactive safety planning and education. And how do you think, Ika, these risks shape caregiver priorities?

Ika Noviawaty, MD:

So mobility issues, especially in adult clinic, certainly it's a priority for them to get referred to either a more assistive device or even a Botox clinic. Some parents noticed that the patient started to have more pain and contracture, which certainly also makes the day-to-day care even more difficult.

Laurie M. Douglass, MD:

I would say the other thing that I see, as you know, even though I'm pediatric, I do take care of a fair number of adults, is that as this person with Lennox-Gastaut is aging, despite being the child, they are getting bigger and their caregiver is often getting older.

Ika Noviawaty, MD:

Yes.

Laurie M. Douglass, MD:

And the burden on the family to come physically into the hospital or into the clinic for care really becomes greater and greater over time. So when they're small and parents are younger, it's less of a burden, but as they get older, it's much more difficult.

Onto another topic, which is always difficult, is thinking about the mortality risk and how it is elevated in this population. And the main one that we worry about is sudden unexpected death associated with epilepsy. And we know that parents live in fear. They're always being very vigilant. They really want to think about safety planning. It becomes very central in their lives. And thinking about this, this can be very burdensome. Sometimes there's a fear of really leaving the home. Maybe they'll not be in the right environment to watch for these signs, and it really is difficult. Unfortunately, it's a burden that they carry, really, lifelong. If you have any insight about this, I'd really welcome it.

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Ika Noviawaty, MD:

Absolutely. Some parents have thought about it and feel more psychologically prepared that they have a sick child whom probably will pass away prematurely. However, some parents prefer to be in denial state and just fail to do any safety planning or any long-term planning.

Laurie M. Douglass, MD:

Now we're going to move a little bit onto the next topic, which is quality of life encompasses safety, as we talked about. Comfort for the individual, communication, social inclusion. We talked a lot about isolation so far. Caregiver really supporting them so that they can continue in that role and they can be sustainable in that role. It's really not just about the seizure reduction.

On the next slide, this slide is a summary which was taken from the 2018 Lennox-Gastaut Syndrome Foundation Caregiver Survey, and it really helps to ground our discussion in who this population is. On the right of this particular slide, you'll notice that the darker blue, what we're seeing here is that 80% of individuals with LGS have severe delay. And the majority, 98%, have severe or moderate delay. There is a very small number who have a mild delay. The other thing you'll see is, although yes, there's a higher risk of mortality, when you look at what were the age of the persons living with Lennox-Gastaut, the caregivers responded of who they were caring for here. But although a fair number are young, you'll notice that 18% were 26 and older. I've been doing this a very long time, and I think that you're going to see that particular part of it, the 18+, the adult population with LGS growing.

So taken together, this really reminds us that LGS care must account for long-term care. We really have to think about this caregiver knowing that they're going to be caring for their child well into adulthood, and I have to really think about helping them to plan for well beyond the pediatric years. How does this age and developmental profile align with what caregivers tell you about their priorities and concerns over time?

Ika Noviawaty, MD:

This reflects exactly what families describe. Many caregivers are supporting adolescents and adults with severe developmental needs, often while aging themselves. So early on, family are focused on survival and how to manage the seizure, seizure control, but over time, the focus shifts to quality of life for the patient and themselves: safety, meaningful engagement, and long-term care planning. And I think I agree with you. If our pediatric neurologist or pediatric epileptologists can somewhat start opening up the conversation about all of these topics, it would be a much easier transition to adult clinic, because as we all know, a lot of adult neurologists are not as well-trained in addressing these issues.

Laurie M. Douglass, MD:

On the next slide, this kind of gives us a little deeper dive into our population of people living with Lennox-Gastaut syndrome to really get at the spread of their disability. So 70% are unable to read, and 65 to 70 are unable to write. I'm not surprised by the bar graph on the right. You'll see that the tallest bar here is the number of people experiencing a sleep disturbance. And then we've talked about the mobility, that's sort of the second-highest bar here with a large number of people, 70% or so, who are having difficulty with ambulation and requiring wheelchair or other adaptive equipment.

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So the key takeaways from this is that LGS care has to be multidisciplinary by default. It's more than just seizures, as we discussed. And we need to really think about screening for the non-seizure symptoms in a proactive way and helping families build management plans to think about their whole life so much more than just the epilepsy itself. Ika, how do caregivers talk about these challenges with you, thinking about communication, behavior, and sleep, and what do they most want from clinicians in terms of support?

Ika Noviawaty, MD:

Caregivers often tell us these are the hardest part of their daily life because they're constant. And not only that, different issues came at different time and it's not predictable for the most part. So they're not only exhausted, but they're also constantly worried. So seizures may come in clusters, but the communication struggles, behavioral challenges, and sleep disruptions are every day, and they affect the whole household. So family wants clinician to actually name this issue directly and treat them as medical and not just parenting or coping problems. Or even worse, some providers tend to think that the parents have been dealing with these issues long enough, and therefore they should be well-equipped or well-adjusted to it. Nobody gets adjusted to it. It's exhausting, and we just have to acknowledge that.

Laurie M. Douglass, MD:

Thank you so much. So next, I'd like to introduce a short clip from Jennifer Griffin on the medical complexity beyond seizures: what families are managing across these multiple comorbidities, the appointments, the therapies, the daily care needs, and the system-level barriers.

Jennifer Griffin:

We went from aspiration to urine retention to severe constipation and low blood pressure – I mean, all of these different issues were popping up. And even afterwards, there was an interesting thing that happened at CVS that I'll get into later, but I think I had two takeaways from this. And that's that when your child is taken to the hospital, step one is to get your child well enough so that they can leave the hospital. But after that, step two is a little bit more complex. I mean, sometimes there's something entirely new that has to occur to their treatment regimen.

Ika Noviawaty, MD:

That is such an important framing, because it captured what can get lost when we focus narrowly on seizure frequency. Families often say the biggest gap is coordination: no single place where the plan lives and no clear sense of who owns the follow-up.

Laurie M. Douglass, MD:

The next slide here, we really wanted to summarize for you the impact and quality of life for those living with Lennox-Gastaut syndrome. You can see how it encompasses multiple domains. When we think about cognitive and behavioral symptoms, many individuals have limited ability to communicate, which can drive frustration and aggression, and often it can be a form of unmet needs. So many times what we try to train families is the behavior isn't something that necessarily needs treatment with medicine. Many times it's figuring out what's the trigger. What's getting them frustrated? What are the settings where this is coming out?

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Sometimes they're expressing dissatisfaction. If they're nonverbal, the only way that they have to let people know that they don't like what's happening is to have an outburst. Certainly by using communication devices and other things we can alleviate some of that, but it requires a lot of detective work on the part of the caregiver, whatever community setting that that person is immersed in, and the health care team. But my biggest advice is don't just layer meds here. Really do a little bit of a detective dive to figure out the root cause. Ika, when families talk about quality of life, which of these domains tends to be most pressing for them, and what do you think they want us to do beyond seizure reduction?

Ika Noviawaty, MD:

Family often say communication, behavior, and sleep are the daily drivers, because they're just constant and it exhausts the whole family. Safety and mobility are huge, especially as kids grow and falls become more dangerous. And as you previously mentioned, parents are getting older and it's getting harder for them to take care of their kids with mobility issues. And many families want clinicians to ask directly and address these issues, and also acknowledge that these are indeed issues and it is difficult. I think by just hearing that it is a lot – it is exhausting to address all of these issues every day – sometimes can provide them some degree of relief.

Laurie M. Douglass, MD:

Thank you so much for that. Here is a very busy slide with lots of factors here, but it's really a lovely summary to think about what are the factors that contribute to the chronic traumatic stress for the caregiver. Obviously, there's a huge amount of sleep deprivation. You talked about the social isolation, and you can't have this discussion without thinking about the fear: the fear in the now and the fear about the future. So many times families say, "What is my child going to be like when they graduate high school? What are they going to be like as an adult?" And sometimes we can give a little bit of prognostic opinion there, but it isn't as if we have all the answers. Ika, what do you think caregivers want most from the clinicians? What are they hoping here, you think?

Ika Noviawaty, MD:

They definitely want acknowledgement, and they want to partner with us, and also be seen as people who also need support themselves. I think all of these measures can make a profound difference, and having more definitive plans in addressing issues that they have can also put them at ease in terms of moving forward with their life.

Laurie M. Douglass, MD:

Thank you. Next slide captures something we all see in practice every day, and I perhaps see it a little bit more than you do, Ika, just being in the pediatric realm. So, when I'm caring for children when they're very young, the focus tends to be for families on the seizures. They're very, very worried about seizure emergencies. They really operate in a fight or flight mode. They're very, very focused on the seizure action plan. As they get older, there's a shift. It's really about quality of life. Do you want to elaborate on that a little bit more since you're caring for the adult population?

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Ika Noviawaty, MD:

Absolutely. Over time, the center of gravity shifts, families begin to focus more on adult care setting: the inclusion, long-term quality of life, and what will happen as caregivers age themselves and their ability to provide care changes. So the key message here is that education and clinical messaging and long-term care planning can't be one size fits all. We certainly need to personalize that depending on the patient situation and the caregiver situation.

Laurie M. Douglass, MD:

I would say the shift also is different for different people. So going from highly, highly focused on the seizures to thinking about quality of life happens at different times for different families.

Ika Noviawaty, MD:

Absolutely. And I also see a lot of families who didn't used to have extra help from a PCA [personal care assistant], for instance. I think they tend to have a much harder time asking or verbalizing that need for help.

Laurie M. Douglass, MD:

Oh, I so appreciate that. One of my favorite things to tell families is that loving families make these decisions. Whether it be they care for their child fully and solely, which a lot of families choose to do, or whether they ask for help, those are still loving decisions to make.

Ika Noviawaty, MD:

Absolutely. And I also tell caregivers that loving yourself and caring for yourself is also a big part of caring for your child.

Laurie M. Douglass, MD:

Absolutely. Next, we're going to hear from Ms. Griffin. She's again going to speak to us this time on the ongoing learning curve caregivers face in Lennox-Gastaut syndrome and how the information needs don't end after diagnosis. This really evolves over time, and families are continually adapting as the seizures change, the comorbidities evolve, as their needs change around school and other life stages. So we'll pause here to view the clip.

Jennifer Griffin:

As caregivers of adults, we've been doing this for so many years that we get into these little routines, and sometimes we're grateful that a particular routine sometimes lasts a long time, and then chaos disrupts everything. And then you're not back at square one, but you're feeling like you've got a whole new learning curve.

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Laurie M. Douglass, MD:

That really reinforced that caregiver education isn't a single conversation, it is a longitudinal process. Ika, when families tell you they're on this constant learning curve, where do they most commonly get stuck, and what can clinicians do to make education more usable and less overwhelming for them?

Ika Noviawaty, MD:

Absolutely. Caregiver often says that the hardest part is that the information arrives during crisis when they're sleep-deprived and already scared, so they can't absorb it all at once. They also struggle when guidance is fragmented across specialties or when they're handed resources without help prioritizing what matters the most right now.

Laurie M. Douglass, MD:

Yeah. This slide is a reality check perhaps for me, for you, for other health care providers, and what families experience early after a Lennox-Gastaut syndrome diagnosis. Ika, when families are in that early phase, what do you think they most need to hear from clinicians to feel supported and able to engage in their child's health care?

Ika Noviawaty, MD:

They absolutely need clarity and validation. Someone naming what's happening without minimizing it. And then giving a short, prioritized plan: who to call, what to track, what to do if seizures change, and what supports to pursue first.

Laurie M. Douglass, MD:

Next, we have another short clip where Ms. Griffin gives us some insight as a caregiver on navigating emergency care.

Jennifer Griffin:

We had an emergency with my son a couple of weeks ago. He had cluster seizures, and I was giving him rescue meds and it didn't work, so I had to call 911. And he was taken by ambulance to the hospital, and multiple issues came up while we were in the hospital that reminded me that as prepared as I think I am, there was a lot that I had to learn.

Laurie M. Douglass, MD:

Ika, what are the most practical ways clinicians and care teams can make emergency care less isolating for families, both in the moment and after?

Ika Noviawaty, MD:

Families need a clear ED [emergency department]-facing seizure action plan and a sense that the neurology team will follow up and not just reacting to the crisis. Even a simple workflow – if you go to the ED, here's what to ask for, and here's how we'll reconnect – reduces fear.

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Laurie M. Douglass, MD:

On this slide here is some caregiver feedback: what health care providers should know, thinking about lifelong systems and support. Ika, how do caregivers describe these system barriers and what do they want clinicians to do differently as they plan for the future?

Ika Noviaty, MD:

I think having clinicians be their partners in terms of, say, if they need to transition to a group home or trying to tap into different resources for more support is key in this. Because let's just be honest: a lot of group homes and adult support, they may not be equipped to meet the complex need of individuals with Lennox-Gastaut syndrome. So it is very helpful for clinicians to discuss and name the realities and help parents to navigate them, and not just to tell them, "Okay, just go look into it yourself and see what you think," or, "You wouldn't know until you try it out." I think that would add another layer of the mental load.

Laurie M. Douglass, MD:

One of the things I hear from families that checklists aren't helpful – it's overwhelming. The other thing that they really want is they want a human. They want a human to help them navigate these systems. These systems are complicated. There are multiple magic doors. If you don't open the right magic door, it doesn't open the next magic door, and there's no roadmap for them to figure it out. Many times they turn to other parents, so I definitely refer them back to whatever support agencies are in the region. Certainly the Lennox-Gastaut Syndrome Foundation is helpful, but also local groups are super helpful here. And tapping into your Epilepsy Foundation and other support organizations is also really helpful to see if they have navigators for these families.

Ika Noviaty, MD:

I agree. And if they have bad experience in the day program or in a group home, sometimes by us connecting with those facilities and educating their staff, it can make a tremendous difference.

Laurie M. Douglass, MD:

On this slide, it's actually a beautiful representation from the caregiver's perspective of the many, many, many jobs that they have. They are responsible for pretty much every aspect of their child's life. And when you think about all the systems that they have to navigate, it can be overwhelming. They're the finance manager for their young person quite often. How do they handle that? They have to think about navigating insurance, and that gets really complicated. One of the things people aren't ready for, 'cause they usually are in pediatric care when this happens, is they change – even if they're on public health insurance, they move from a Medicaid to a Medicare program, and then all their seizure medications need prior authorizations all over again. Ika, when you look at this really complicated diagram, thinking about all the caregiver is managing, what do you think are the points that really make them hit overload, and what kinds of support change the day-to-day reality for them?

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Ika Noviawaty, MD:

Coordination. Coordination is definitely the key. So overload often happens when families are forced to be the care coordinator, the advocate, and also the nurse without any backup. So under the circumstance that they also have sleep disruption and constant vigilance, it just makes everything so much harder. So, with coordinated care and realistic care plans along with practical supports, that can make a huge difference.

Laurie M. Douglass, MD:

At this next juncture, I'd like you to watch this clip considering the caregiver's perspective around the emotional barriers: guilt, fear of judgment, the feeling that asking for help means failing. And we talked a little bit about this: that I think so many parents carry so much guilt, so much fear, like if they ask for help that they're giving up.

Jennifer Griffin:

Let people help you because they want to. And the reality of this experience reminded me that I, as the caregiver, absorb all of the responsibility for coordinating all of this. A lot of time, others can't help me. So if there's something that somebody is offering to you, be it a good friend offering to go pick up some groceries for you, or bring over dinner, or just take you out for a coffee, or something, rub your feet...oh my goodness, that's a dream. That doesn't happen in my life. But if something like that were to happen in my life, I would say, "Heck yes, help me out."

Laurie M. Douglass, MD:

Great. I think this clip is such an important reminder – reminds us that caregiver support is not optional. It really needs to be part of the clinical plan. Ika, what are practical ways clinicians can normalize support and make it easier for families to accept help?

Ika Noviawaty, MD:

One powerful step is to prescribe support the way we prescribe treatment. Name it directly, normalize it, and connect it to outcomes. For example, respite and peer support reduce burnout and help you sustain care long-term. And I think it is also good to always screen for the possibility of parents feeling the guilt. I've heard some parents stating that they feel like as if they did something wrong and therefore their child becomes sick. And others also feel that when they're asking for help, for more help, not only it feels like they're failing and unable to provide care for their child, but also feeling like as if the person who's going to come and help them is not going to be able to provide the best care for their child.

Laurie M. Douglass, MD:

So, we've talked about the clinical realities of Lennox-Gastaut syndrome, but none of it happens in a vacuum. Caregivers are really the constant in this system, and caregiver capacity often determines what's feasible in treatment. We're going to move into a little bit of a discussion, and I'm going to read a question for you. So what is your main goal when you treat individuals with Lennox-Gastaut syndrome? Is it achieving seizure freedom,

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improving patient motor skills, addressing behavior and sleep issues, reduce seizure frequency and severity, or is it minimizing the treatment side effects?

Ika, I'll start with you. Tell me how you usually prioritize these things.

Ika Noviawaty, MD:

I always prioritize the balance of the best possible seizure control and also addressing safety along with quality of life: not just for the patient, but also for the patient's caregiver. And also understanding that we need to be on the same page. Most of Lennox-Gastaut syndrome patients may never be able to be seizure-free, but I think the goal is to minimize the seizures so it's not a safety concern for the patients, and it doesn't look as threatening for the parents and other caregivers that they are so stressed all the time.

Laurie M. Douglass, MD:

Yeah, I think that's great. One of the ways that I approach it, and I think it's quite similar to what you said, is I sit down with the caregiver, and the young person if they're able to contribute to this, and we make goals. And we talk about what's getting in the way most of their quality of life. If they're telling me seizures are preventing them from doing the activities that they enjoy and being engaged or whatever it might be, we'll set some goals around seizures.

And like you said, it might be we're trying to reduce the amount of rescue medicine that the person is receiving, because if they're constantly in that emergent, urgent state, it's very hard to engage in life in a meaningful way. Or we're trying to reduce hospitalizations, ER [emergency room] visits. So that might be a simple way of looking at the seizures, like you said, decreasing the frequency and the severity, but it also has to be balanced with all that other stuff, what matters most. We don't want to give so much medicine that the person's sleeping all day and not engaging in life. And so it really is a matter of thinking about what matters for them as a unit.

Ika Noviawaty, MD:

Absolutely.

Laurie M. Douglass, MD:

So here we can see the gradual expansion of anti-seizure medication options, including the agents that carry an FDA-approved indication for Lennox-Gastaut, alongside many therapies that have been used in epilepsy care without a specific Lennox-Gastaut indication. Ika, what do caregivers typically experience as treatment evolves over the years?

Ika Noviawaty, MD:

Families often describe as an ongoing cycle of hope, adjustment, and trade-offs. They're tracking not only seizures, but alertness, mood, sleep, appetite, and behavioral, because a change that looks like progress on paper can be a setback at home if side effects worsen daily life.

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Laurie M. Douglass, MD:

Yeah, I agree. So here we can see some of the medications that are commonly used with Lennox-Gastaut syndrome and how they might impact quality of life. Seizures are certainly important to treat, but we need to think about how those treatments impact mood, cognition, sleep. We also need to think how the seizures are impacting those particular parts of the person's life. And because many patients with LGS are on polytherapy, obviously the results of this can be really compounded, and so polytherapy can be challenging. Ika, when caregivers talk about medication burden, what are the common quality of life trade-offs they describe?

Ika Noviawaty, MD:

Sedation and sleep disruption come up constantly, along with changes in behavioral, irritability, agitation, or emotional flatness. Caregivers often say, "We can handle some seizures if our child is awake and able to engage," but they really struggle when treatment makes the child feel disconnected or when the household becomes sleepless because of aggressive behavior or insomnia.

Laurie M. Douglass, MD:

That's exactly my experience as well. I have many parents, "Dr. Douglass, Dr. Douglass, when can we peel back some of this medicine?" Even though the child is having many, many seizures, they really want their child to be able to engage in life. And so, so important, what you said.

So here we're talking about the safety and tolerability of anti-seizure medicines for Lennox-Gastaut syndrome. And there's a lot here on this table, but the point here is to normalize proactive counseling and monitoring. Families do better when we set expectations upfront: what side effects to watch for, when to call, what labs or follow-up may be needed, and how quickly we expect to see benefit. Ika, what do caregivers most want clinicians to communicate when starting or changing an anti-seizure medicine?

Ika Noviawaty, MD:

They want clear expectations and a plan. What changes are expected versus concerning? And how long should we wait before deciding it's not working? And who do we contact after hours? And how do we balance side effects against seizure improvement without feeling like we're choosing between two bad options? And I also make the expectation very clear that every patient can respond differently to different medications, so certainly it's not one size fits all.

Laurie M. Douglass, MD:

So medication is one piece, but many patients also benefit from dietary therapy, device-based approaches, and when appropriate, even surgical evaluation. And alongside seizure-directed therapies, there are supportive services like physical therapy, occupational therapy, speech therapy, behavioral supports, and care coordination can materially improve function and family sustainability. Ika, what helps families most when the plan becomes complex and involves multiple disciplines?

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Ika Noviawaty, MD:

Families do best when someone helps them sequence and prioritize a clear roadmap: what we're focusing on now, what comes next, and who owns each part. All of these can reduce the overwhelming feeling. Families also benefit when the care team acknowledges bandwidth and design plans that are realistic for their home situation.

Laurie M. Douglass, MD:

Next, we're going to talk about shared decision-making. And having a framework for decision-making, oh gosh, I feel like it's never brought up enough and it's not brought up soon enough. So in practice, it's often done with the caregiver, but when possible, it really should be with the patient. And it can be started in little micro bits. Ika, what do you think makes your decision-making feel genuine to families rather than performative?

Ika Noviawaty, MD:

So I often tell patients and their families that a large portion of my recommendation is reliant on what they're telling me, their goals and how they feel with certain medications, certain treatment. And families feel that it's genuine when clinicians ask what a good week looks like, what they can realistically manage, and what trade-offs they're willing or not willing to accept. And when clinicians revisit decisions – is this still working with you? Families feel like partners instead of just a passenger. And I think that can improve compliance to medical recommendations greatly, and certainly can build rapport in the long run when we need to have an even more difficult conversation when the conditions deteriorate.

Laurie M. Douglass, MD:

Thank you for that. So supported decision-making often gets confused with shared decision-making. Supported decision-making is really a framework for where individuals' disabilities are supported in making their own choices to the greatest extent possible, rather than having decisions made entirely for them. So depending on the age and the level of intellectual impairment, supported decision-making may help patients with Lennox-Gastaut retain some agency in their treatment decisions, daily routines, and long-term planning. And I think each family has to decide what the decision-making framework is for them that makes the most sense, but whenever possible involving the youth is really, really helpful. Ika, how do caregivers respond to these concepts when they're planning for adolescence and adulthood?

Ika Noviawaty, MD:

Many caregivers want their loved one to have as much agency as possible, but they also worry about safety and legal responsibilities. So having this conversation sooner than later, even when it doesn't produce the final planning, it helps the patient and their caregiver to have more time to think about it.

Laurie M. Douglass, MD:

Thank you. So we're going to shift a little bit now into long-term planning and transition, because for many families, the question isn't only how do we manage seizures this month, but how do we build a sustainable life across decades? So this section will focus on holistic planning, medical development, educational, legal, and

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community supports so that transition is proactive rather than driven by crisis. So I'm going to start with a question. At what age do you generally begin transitioning patients with Lennox-Gastaut to adult care? I recognize this is going to be different for, I think, every provider, and sometimes it's individualized based on what's going on with that particular person living with Lennox-Gastaut. So is it 12 to 14 years, 15 to 17 years, over 18? Or some people might say, "I'm not involved in the transition process at all."

Ika, what do you think?

Ika Noviawaty, MD:

I think the actual transition should start at around 17 or 18 years old, but also unfortunately, it's highly driven by sometimes the family's reluctance to leave their pediatric neurologist whom they have known their whole life, and also driven by the availabilities of adult neurologists whom are willing to take these patients in their practice.

Laurie M. Douglass, MD:

Yeah, I think that's so true. Certainly planning early is very, very important. So it sort of depends on the family and where I think they are in being able to hear this information, but I'll start talking about, let's talk about planning. I let them know when I would definitely need to transfer the person, what kinds of things would be my pivot points. And we always talk about, let's do this as a plan rather than when we absolutely have to. So that's part one. And then we talk about it as a process – it's not just a sudden exiting of the pediatric clinic. I think that's important.

I usually talk about transitioning about primary care. When are they going to transfer their primary care? First, make sure that they have somebody, because what I find, the difference for pediatric versus adult is the primary care provider in the adult world absorbs a lot more than what they do in the pediatric world. In the pediatric world, I think the pediatric neurologist deals with the behavior, the sleep, the epilepsy, and so much of life planning. But in the adult world, I think a lot of that gets managed by other specialists and their primary care provider. So I always recommend transferring their neurological care after their primary care.

Which kind of brings us to the next slide, and I think these terms get really confusing. What is the difference between transfer and transition? So, transition is a developmental process, so helping the patient and the caregiver to build skills, confidence, and supports so that when they're going into adult-serving systems, they're ready and prepared. Whereas transfer is really the physical movement of going from pediatric-serving systems to adult-serving systems. Ika, what do you think a good transition looks like from the caregiver point of view?

Ika Noviawaty, MD:

A good transition feels planned and supported. Families know who the adult team is, what will happen with medications and emergency plans, how to access service, and who to contact when something changes, and they don't feel like they're starting over from zero. And I do agree transferring the primary care service first and then followed by transferring the neurology service is often the best measure to do this transition successfully.

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Laurie M. Douglass, MD:

Great. So without planning, families face delays in specialty care, poor management of comorbidities, and barriers to adult hospital access. Adulthood introduces new issues like guardianship, insurance changes, long-term care needs, and many caregivers worry about who will support their child when they no longer can. Ika, when families tell you, "We tried to transition and it didn't work," what are the most common breakdown points that you hear?

Ika Noviawaty, MD:

The biggest breakdown point is provider readiness and the system fragmentation. Families feel like adult system just simply don't understand the complexity, and caregivers are forced to educate new teams while managing crisis. And oftentimes, families are also not fully ready for the transition. So it is also important to acknowledge that we need to disclose to families that some adult providers may have limited experience with pediatric-onset epileptic encephalopathies, so therefore at least families will give the provider some time to build that rapport and trust in their care.

Laurie M. Douglass, MD:

One of my strategies – it doesn't always work – but I try to encourage my patients, just make an appointment, get it on the books, and you're still going to come back to see me because I'll work with your adult neurologist as a team. But as you know, wait times to get into adult neurology and certainly adult epilepsy are really long. And then we can come back and review, and I can give them some skills to move forward into the adult system, and it's a back and forth. It doesn't have to be an all at once. I know that doesn't work for everybody. Of course, when we're in a system that allows for both pediatric and adult care, we kind of have the best of both worlds.

Ika Noviawaty, MD:

Absolutely. And I feel that if just so happened a pediatric neurologist personally know the adult neurologist, sometimes by just doing a five-minute discussion about what are the family's expectations, what are their difficulties, what are their goals, it can help tremendously more so than just doing chart review.

Laurie M. Douglass, MD:

Thank you so much. So the next slide is a timeline of transition planning. So in the beginning, we start to have those discussions, then we begin to review testing and what things do we need to make sure are complete before there's an ultimate transfer so that when that person goes to the adult-serving system, really all of the pieces have been complete. For me, especially right now, genetics is a big thing and it's become more broadly available. It used to be for the youngest children, but that age range has expanded. And I know that access to genetic testing will go away for many when they turn 18, so I really, really work to get that testing. So number two here for me, as the pediatric person, is really important so that I know that they got all that we could during those early years.

Part three is to really think about decision-making frameworks, as we discussed. Is the caregiver going to be the sole decision-maker, something we refer to as guardianship? Or are they going to use a different framework of supported decision-making? And if they're going to do supported decision-making, I really have the youth

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practice it with me in the clinic as soon as they're able. "We're going to start a new medication for you today. Tell me what's important to you about your medicines." Well, they might say, "I really hate taking medicine three times a day." "Okay, great. Maybe I can give you something you only have to take once." So getting them involved in that discussion so that it's not just really prescriptive, that they are actually having some agency, as you said, Ika, before.

For four, refer support agencies and/or social worker for transition. That can't happen early enough. I am overwhelmed all the time by the number of 19-year-olds who have not been connected to the Department of Developmental Services, for example. And so finding those systems and who those people are can be really challenging. For step five, identification of adult providers – not just for the epilepsy, as we said. Many times in the adult-serving system, the epilepsy doctor will pretty much manage the epilepsy. The behavior? Who's going to do that? Is it going to be an adult psychiatrist? Is it simple enough that the family physician might feel comfortable taking over that care? So thinking about it. Sometimes these patients have trouble feeding, they have G-tubes, and who's going to manage that? It turns out, in the adult world in some hospitals, the gastroenterologist doesn't manage the G-tube. It might have to be the internist or the family physician. And thinking about functional evaluations is also really, really important. And then six, really putting together a plan with the family is so important. It doesn't have to be all on the health care provider. So pulling everyone you can from all your disciplines to really develop a joint plan, I think, is so important. So, Ika, when families are overwhelmed by this huge list of things they have to accomplish, what helps them to feel like it's doable?

Ika Noviawaty, MD:

Two things: sequencing and support. Families do better when the care team helps them prioritize what matters now, what can wait, and who can help with each task. And when clinicians acknowledge the emotional weight, some families aren't ready to talk about long-term planning, and that's a real issue.

Laurie M. Douglass, MD:

So as was said multiple times, a multidisciplinary team may include pediatric and adult neurology, nurses, advanced practice providers, social workers, community organizations like the Epilepsy Foundation, and of course, at the very center of this socio-ecological model or support network is really the youth who's living with the Lennox-Gastaut syndrome and their caregiver. Do you want to comment at all about this diagram?

Ika Noviawaty, MD:

Yeah, so support network is extremely important: having a village so that they don't feel isolated in their struggle. It's going to be very helpful in terms of long-term care.

Laurie M. Douglass, MD:

So we've talked about this diagram before. It's really so much information here. So this caregiver perspective slide makes the complexity visible. Transition isn't only medical, it's about the education, the vocational planning, the insurance, the eligibility requirements, legal decision-making. Where is the person going to live? And if they are living with their caregiver, how do they access respite? Families are mapping out an entire life system. Ika, when you speak with caregivers in this stage, what do they most want clinicians to do or stop doing so planning feels realistic and respectful?

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Ika Noviaty, MD:

I think the most important thing for them is for us clinicians to understand where they are in their stage of planning and goals and life in general. And then for them to be treated as a partner and not just clinicians dictating what should or should not be done, or what we think is best for them. I think that's the most important thing.

Laurie M. Douglass, MD:

Yeah, I agree. I think knowing what their goals are and what looks like a full life for their family, what I might want for my family is very different from theirs. So I think sitting down and figuring out what does a full life look like for the individual with the epilepsy and the whole family as a unit, what their priorities are, what their values are. And then thinking about who is in this support network. What's their family unit look like? Who else can come into this picture to support this person? And I find that also can be really helpful – is reminding them they don't have to be alone, and sometimes they forget who else might be there to lean on.

Ika Noviaty, MD:

That's a great point.

Laurie M. Douglass, MD:

So this slide highlights a practical resource from the Lennox-Gastaut Syndrome Foundation, which is called the C.A.R.E. Binder. It's designed to help families plan long-term adult care for Lennox-Gastaut syndrome. It includes key factors to consider during transition, how to prepare, and the available support services. Resources like this really matter because they reduce fragmentation. Instead of families reinventing the wheel, they have a structured guide to organize documents, decisions, and next-step plans. I have to say, I'm really excited to use this particular resource with my families.

Ika Noviaty, MD:

Absolutely. I think it helps for them to have such a comprehensive reference of resources.

Laurie M. Douglass, MD:

We're going to move into a little bit of a faculty discussion in the next minutes, and I thought I'd begin with when families hear transition, many feel overwhelmed. What are the first two concrete steps you want caregivers to take in the next three to six months to make transition feel doable? What would you say, Ika, about that?

Ika Noviaty, MD:

I think the first thing is it's good for caregivers to reflect what are the most troublesome barriers or challenges that they face with the care that they have right now, and what is their long-term goals, and how do they want to see that change or are they satisfied with where they are in their care? And maybe we just need to discuss about the long-term planning, about end-of-life planning.

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Laurie M. Douglass, MD:

Yeah, I think this is a loaded question, because transition isn't just about the health care transition, it's also about where are they going to live? Are they going to work? Are they going to a day program? Are they going to school? What are their social needs? And so it's a pretty loaded question, so I'm sorry I gave you a tough one. I think it really depends on the age and where they are along the transition process versus transfer, which we've talked about. Since we're health care providers, maybe it makes sense to think about health care transition. One of the things I begin with is where do you want your young adult to receive care? Do you want them to receive it in the community? Because then we need to think about finding a primary care provider who's close to where they're going to be living.

And do you want to have them have neurological care in the community as well? And we need to think about that. That takes a little bit of coordination. Versus, do you see yourself being in a large academic center? It might mean some travel for you. And then I will do that kind of plan. So just focusing on those basics, like what feels good to you? Where would you want your young person to be receiving care? So starting really at the basics and not dictating what they should do.

Ika Noviawaty, MD:

From the clinician side, what belongs in a one-page transition summary so adult providers can pick up care without families having to start from scratch? What do you think, Laurie?

Laurie M. Douglass, MD:

Well, it's funny, because I feel like this is best for an adult neurologist. I'll tell you what I like to put in my summary, and you can tell me if I'm right. So first I'd like to tell you, if I was sending my patient to you, is how old were they when seizures began? I'd like you to know the seizure types that are most problematic to the patient and to the family. I'd like you to know what they're currently taking for medications. I'd also like you to know what they failed and why they failed. Sometimes it's true: we can retry medicines, they work at an older age when they didn't when they were younger. But it's helpful, I think, to you to know that, because I think when you as an adult provider say, "Well, let's try this." And then the parent says, "Well, we already did that in the past." I think that can really hurt the trust. So I'd like you to know that.

I'd like you to know what testing has been done. I'd like you to know if I've done an EEG recently and what that showed. I usually will communicate pretty clearly, not just a rescue plan, but a sick day plan. I don't know that everyone thinks about this, but it's very common for people when they are ill to have a worsening of seizures. So if you don't have to do that from scratch and I've already experimented and had to have several iterations of those plans, I'd like to be able to give that to you.

And then because many of these people have developmental disabilities, I also want you to know them as a person. First of all, do they have any sensitivities? That might help you to understand what that visit needs to look like. Maybe they need to be scheduled as the first patient of the day where it's not so noisy, so busy, and so overwhelming, and that they don't have to wait. As we mentioned, behavior can be a big challenge for people with Lennox-Gastaut. And if you're running behind, it might actually become a really unsuccessful appointment.

And then I want you to know how do they communicate? How will you communicate with them? What things make them feel comfortable so that you start to be able to build a rapport with them? For example, maybe your

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patient really likes Marvel heroes. I'm going to let you know that, so that just mentioning a couple of key characters might make them brighten up, and then suddenly you're already off on your way for rapport building. I guess, I mean, there's so much more I could include, but I'll stop there to see what you have to say, Ika.

Ika Noviawaty, MD:

I would say that as clinicians, we are all trained to make very good medical-related summary, but I do agree with you. I think all of those nonmedical stuff is what matters the most in making the transition successful.

Laurie M. Douglass, MD:

So we've really discussed a lot today. And let's sum up with our SMART goals that we hope our audience will take away from this program. So first, I want you to put this information to action, consider the following goals, and set a timeframe that fits your work environment and a reasonable improvement target that aligns with your patient population. Really think about monitoring quality of life challenges for your patients and caregivers, and address these challenges with resources and supports when possible. I really encourage you to go online, search the supported agencies in your area, and start to build them into your practice. Incorporate seizures, non-seizure symptoms, and quality of life into treatment decisions. That's very, very important. And then lastly, prepare patients and caregivers for transition from pediatric to adult care by really having these discussions early and following key milestones for successful transfer of care.

So that's all we have time for today. You can find the slides from this activity and the course guide under the resources tab of this program as well as additional downloadable resources about Lennox-Gastaut syndrome. Don't forget to visit cmeoutfitters.com for more free resources and education for health care professionals and patients. To receive CME or CE credit for today's program, complete the post-test and evaluation. I'd really like to thank Dr. Noviawaty for joining me today, and Ms. Griffin for her fantastic insight. Thank you so much for this great program.

Ika Noviawaty, MD:

Thank you for having me, and I really enjoyed this great discussion.

Laurie M. Douglass, MD:

Thank you so much, Ika. I also want to thank CME Outfitters for their assistance in developing today's program. And have a great evening.