

# Live Q&A Session

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[00:00:04] **Krissy Dilger:** Hello, everyone, and thank you for joining our ADEM Together event. This is the Q&A portion with Dr. Linda Nguyen from UT Southwestern. So, we can start off with some questions that we've already gotten. But also feel free to share questions in the chat or in the Q&A section in Hopin. So first off, what is ADEM and how is it connected to other rare neuroimmune disorders?

[00:00:39] **Dr. Linda Nguyen:** That's a great question. So, to understand ADEM, it's important to know what it stands for. So ADEM stands for acute disseminated encephalomyelitis. So, what does that mean? If you break it down, acute means rapid onset of a few days to a few weeks and then disseminated meaning it affects multiple parts of the central nervous system. Encephalomyelitis breaks down into the brain and then the spinal cord. So, it's a disease that affects the - can affect the brain and spinal cord and then it's actually very common presentation in children and can be the first event for the onset of multiple sclerosis.

[00:01:28] **Krissy Dilger:** Thank you. And is it possible to have ADEM and another diagnosis such as MOGAD?

[00:01:38] **Dr. Linda Nguyen:** That's great. So, yeah, I was mentioning multiple sclerosis as one of the possible evolving diagnoses. But actually, in children, in kids that present with ADEM a lot of them can have what we call MOGAD. So, if you have the positive antibody for MOG, and you present with ADEM, you are classified as MOGAD with just an ADEM presentation and so yes, they can have it concurrently.

[00:02:12] **Krissy Dilger:** Thank you. Our next question, how does ADEM present in children versus adults? Are there different treatment approaches with children compared to adults?

[00:02:25] **Dr. Linda Nguyen:** Yeah. So, I was alluding to ADEM being very common in children in terms of the age profile, it's very, very uncommon for adults to present with something we call ADEM. If they were present with ADEM in adults, we would try to figure out some underlying cause for it. And in terms of how they present, there's specific criteria to be diagnosed with ADEM, you have to have different lesions in the brain and/or spinal cord and you have to have altered mental status. So, if adult or pediatric patient fits with that definition, then they can be diagnosed with ADEM and so a similar presentation really. And then in terms of treatments, because it's thought to be a similar disease process in adults or kids, we treated them acutely the same way with acute immunotherapy like steroids, IVIG and if they're pretty severe plasma exchange.

[00:03:30] **Krissy Dilger:** Thank you. So, this next question is from a community member. I was diagnosed 14 years ago with ADEM, what advancements have been made in diagnosing and treating ADEM and any advancements on finding a cause?

[00:03:49] **Dr. Linda Nguyen:** That's a great question. We have learned a lot over the past decade or two. Unfortunately, there's no biomarker that can say you have ADEM or not or some other diagnosis based on a

clinical diagnosis with MRI and clinical symptoms. However, in the past decade, there has been the recent knowledge about MOG antibody-mediated disease. So, in that way, we do know one etiology for ADEM since 14 years ago. In terms of treatment, there hasn't been significant advancements there. Most of it is similar in terms of the acute regimen we typically use, which is as I mentioned, steroids, IVIG, and plasma exchange.

[00:04:40] **Krissy Dilger:** Thank you. This next question - what causes symptoms to return? How can I as a parent understand if symptoms are returning, an attack is happening, or what is typical child actions?

[00:04:57] **Dr. Linda Nguyen:** That's a great question. This is something we always get asked. How can we know what will happen in the future after my kid presents with this one-time episode? So just in general ADEM is usually a monophasic presentation. So, in the majority of patients probably 75% they don't have another attack or a relapse. So that's a good thing. And so that's why we don't start them on long-term immunosuppression as opposed to if they were to be diagnosed with multiple sclerosis. And then, it's thought that ADEM is triggered by a viral infection. So, in a lot of patients, 75% of them, they have a preceding viral infection that triggers and confuses the immune system to cause the ADEM to present.

[00:05:45] What causes symptoms to return? We don't actually know, we can't really predict, unfortunately, at onset, what separates between the relapsing individuals and the monophasic individuals. Certainly, if you have the MOG antibody and persistent MOG antibody in your blood, you can have a higher risk for developing relapse in the future. Now, how can you know if symptoms are returning? So usually in patients, 70-90% they return completely to normal, they're able to carry out the expected activities for a kid. So, if you're noticing symptoms coming back, such as they're acting differently or they're more agitated or they have a focal neurological deficit, like they are having trouble walking, definitely let your neurologist know because that would be very unusual.

[00:06:53] **Krissy Dilger:** Thank you. So, if someone had an ADEM attack, should they be tested for MOGAD?

[00:07:03] **Dr. Linda Nguyen:** That's a great question. So actually, in those present with ADEM about a 40-50% of them can be MOG positive. So, we highly recommend all patients be tested for the MOG antibody when they present with ADEM, or even if they're following up and it hasn't been tested in the initial presentation, it'll be a good idea to get that tested.

[00:09:29] **Krissy Dilger:** Thank you. And this next question is about ADHD. So how is ADEM-induced ADHD treated?

[00:07:42] **Dr. Linda Nguyen:** That's a good question because we expect a lot of our patients to not have any neurological disability once they've had their initial attack but we're finding out that in the long-term because they had a hit on the brain, some of them can have over time learning disabilities, including cognitive problems that they grow into ADHD. In terms of the treatment, there isn't necessarily any difference in the management. If you were diagnosed with ADHD, you would treat it symptomatically with ADHD medications, if needed.

[00:08:29] **Krissy Dilger:** Thanks. So, should precaution be taken for dental care with ADEM? This person's family member is six years post-ADEM with residual TM bladder, pain, and temperature issues, also lost some eyesight. ADEM was full spine into brain.

[00:08:55] **Dr. Linda Nguyen:** So, I'm not exactly sure what this question was trying to ask if it's like anesthesia for dental care or whatnot. If it's referring to that, it shouldn't make a difference in terms of special precautions.

[00:09:18] **Krissy Dilger:** Thank you. Are ADEM relapses in MOG milder than the onset attack or how do they compare?

[00:09:28] **Dr. Linda Nguyen:** That's a good question. So, it's hard to say, because usually actually when people present with ADEM initially and they have MOG, they usually present with a different phenotype than ADEM. They present with optic neuritis most commonly, or they can present with any other focal neurological symptom. And sometimes it can be milder and then sometimes it can be more severe. So, we really don't know in terms of the severity of the next attack. But in general, I would have to say if you have MOG, it's thought to be much milder than if you had something called aquaporin-4 positive NMO or even multiple sclerosis.

[00:10:25] **Krissy Dilger:** I'm just trying to see what questions we have coming in. So, this person just is describing their symptoms, but I am assuming they want to know how this can be helped or treated. So, they have bladder issues and weakness of legs four years after their ADEM attack. Is there anything that they can try to resolve these issues or help and is this typical of ADEM?

[00:10:59] **Dr. Linda Nguyen:** So, I was mentioning most patients aren't left with any neurological deficit, but certainly, there are a small subset that have a very severe presentation and can be left with long-lasting neurological deficits such as bladder issues and weakness. And so, in terms of the management since four years of disease, right now, usually the persistent symptoms is the sequelae of the damage that has already been done. And there isn't any certain therapy right now to regenerate the damaged spinal cord and brain tissue, but certainly supportive treatment with continued care with urology, neurology, physical therapy, and rehab medicine is very important to maintain strength and ability to function and live a good life.

[00:12:04] **Krissy Dilger:** Thank you. This person wants to know. Are headaches typical post-ADEM? When should I be concerned if I'm having a headache and how can these be treated?

[00:12:24] **Dr. Linda Nguyen:** Sorry, there was a different connectivity issue there. But I am able to see the question. And so, in terms of the question about headaches post-ADEM, so ADEM can present with headaches very commonly in our subset of patients. About 30% of them can have headaches, but usually, the headaches go away. Now, if there is a persistent headache afterwards, I think it may become more of a medic -- like we have to think about a preventative therapy to try to help with the headaches in terms of trying headache medications if headache are persisting. It's not usually thought to be a sign of persistent inflammation, meaning we have to control it with immunotherapies. But oftentimes if they have headaches afterwards and they continue to have headaches, we try to do supportive treatment with headache medications.

[00:13:22] **Krissy Dilger:** Thank you. We just got another question in, so I got it here. This person wants to know what can cause a flare up in nerve pain.

[00:13:37] **Dr. Linda Nguyen:** So, in terms of if you had nerve pain as a result of the ADEM or any other acquired demyelinating disease, certainly flare ups can be caused by if you're overheating, overexerting yourself, having any stressors in your body, or being ill with viral infection or any sort of infection. Anything that stresses your body can flare up your symptoms, whether it be nerve pain, or difficulty walking, or vision. And so, what's thought to be underlying that is because when you have ADEM, it's thought to be an attack on your myelin and myelin is the outer covering of your nerve endings. And so, when that myelin gets damaged, the nerve signal, the conduction of the electron signal from your brain to any part of your body gets disrupted and that transmission can be slowed down. And when there's any stress that communication gets further delayed and then you can have a flare up of your symptoms.

[00:14:52] **Krissy Dilger:** Quite related to the last question. This person wants to know if viruses irritate nerves?

[00:15:00] **Dr. Linda Nguyen:** So certainly, if we're talking about viruses directly invading into our nerves and attacking it and causing pain, yes, that can happen. But if we're talking about viral infection worsening the nerve pain, it can just be because of stress to the body rather than a direct invasion of the virus on the nerves.

[00:15:33] **Krissy Dilger:** I think we're still seeing if any more questions come in because we've actually reached the end of what we've got so far, we really flew through these ones. But I think maybe we can give it a second just to see if anyone listening in wants to throw out you a question. Now is the time we've got an expert here. So, we'll just give it a few minutes. I guess in the meantime if there's anything we haven't asked about yet that you think is important for someone to know with ADEM that you would like to talk about, we'd be happy to hear what your perspective is.

[00:16:17] **Dr. Linda Nguyen:** I think I blew way too fast through the questions. I'll take my time more if there's any new questions coming on. So, it's sometimes hard to get at because children when they're sick can be very irritable. They can have behavioral change and wondering what is part of the illness of the virus infection or some other infection versus the ADEM response with an attack on the brain and spinal cord. So, a lot of the times it would be an exaggeration of what we would think in terms of how the kids are acting. So, for instance, if a kid presented with a cold, they had fever, cough, congestion, typical 3, 4 days but they're not getting better and they're starting to act a little strange towards the end of it, whether it be a few days after the onset of the fever or a week after the resolution of the febrile illness. At that point, they should get checked out because a lot of the times ADEM can present that way. I was alluding to around 70% of patients have a preceding viral infection. So just keep that in mind, certainly kids can be very irritable when they're sick. But I think all parents would know when it's a little beyond just an illness.

[00:17:57] **Krissy Dilger:** Those are great points. I think it's important to be aware. We did receive one question. What types of health care professionals should I continue seeing post-ADEM? Should I still have regular appointments with my neurologist, psychologist, etc.?

[00:18:16] **Dr. Linda Nguyen:** So, this is a great question because I told you that a lot of patients, the majority of patients, it's a one-time event. It's monophasic and the majority of patients recover fully. So, do we even need to continue to see our experts or neurologists? I think it's important actually to do so. I mean, at our center - I'm sure different centers are different but at our center, we typically see kids post-ADEM through their elementary school years and into their high school years. Because what we're finding is as they grow and get more academic challenges as they increase their education years, they can start to develop cognitive problems and we want to be able to address and support you guys through that. And so, we do have our patients follow up for at least five years, post the initial event. And then in terms of regular appointments, usually it's a one-year check-in when you're post-ADEM, just to see how you're doing, make sure we're not seeing any unusual new symptoms. And then really just to tell you if we have any new research or new data points that help us identify whether your risk of relapse, what would be in the future, and what else to expect. So, it's always good and at least in our center to continue to check in.

[00:19:54] **Krissy Dilger:** Thank you. This is related to what you've recently been just talking about. But someone did write in, my child has been irritable, and their personality changed since their diagnosis a couple of months ago. Will these traits continue on? How can a parent deal with it? And I guess, also just to expand a little bit, like what kinds of plans can be put in place in schools, for instance, for children experiencing these issues?

[00:20:29] **Dr. Linda Nguyen:** So, in terms of the persistent symptoms, that can happen. But over time, usually, it continues to get better, and they're expected to get back to their regular self. So, if your child's still been

irritable and personality is different in the past couple months, I think to give it time would be important. But once you're passing the 3-6 months period and you're still noticing your child having difficulty, irritability that can be affecting his/her ability to perform in school, to interact socially, I think at that time point, it's important to think about if there's something else that can be done. Whether it'd be to see a psychologist, a behavioral specialist, or even to try medications to help with that irritability so that they can function as best as they can.

[00:21:27] So that's commenting on how to deal with it and it's very important to work with the school so they would know what to expect. Certainly, some kids may need good accommodations, or extra help, or an extra understanding from the teachers. So, it's important to try to let the school know what your child has undergone, what their diagnosis is and then having them get evaluated by the school to see what accommodations can best fit them so that they can excel.

[00:22:06] **Krissy Dilger:** I think these are all really important points and it is definitely a learning curve. I think it's really important also just to have put together that team of professionals because one person won't be able to just deal with this all on their own and having that care team can really make a difference in how the outcomes are for these children and occasionally adults with ADEM.

[00:22:37] **Dr. Linda Nguyen:** Absolutely. Yeah, we're privileged here at UT Southwestern to have a multidisciplinary clinic. But our motto is not a typical motto for other regions of Texas or even in the United States. So certainly, if there's still ongoing problems, it's important to continue to work with your neurologist to try to make sure that your child is getting all the support he or she needs.

[00:23:11] **Krissy Dilger:** That's great. So, this is the question, we have a five-year-old child recovering from ADEM and are thinking about getting him vaccinated for TBE since we live in a "tick-risk-area". Are there any risks with taking those types of vaccines post-ADEM? And I guess just to expand in general on vaccines and their safety with people with ADEM.

[00:23:39] **Dr. Linda Nguyen:** So, to answer this question, I think early on in the early studies, they thought there was an association between getting the vaccine and then having a higher incidence of ADEM presenting. But I think as they follow on the studies in larger studies, that earlier study couldn't be confirmed. They couldn't find a consistent increased risk of having ADEM if you were vaccinated or not vaccinated for wide variety of the vaccines, including all of the recommended pediatric vaccines.

[00:24:17] And really to understand what the vaccine related ADEM, what does that mean? So, to better understand that and to get to the answer, I think it's important to know that ADEM is thought to be a post-infectious or if it's vaccine, post-vaccine related phenomenon where your immune system gets confused. So, it's fighting off the infection, it's mounting a response to the vaccine and then it unfortunately gets confused and it's trying to do what it's best at doing, protect you, but then it gets confused and attacks parts of your brain and spinal cord. So, it's really just your immune system acting appropriately. But unfortunately, it gets a little confused and attacks your brain and spinal cord.

[00:25:12] So it's not necessarily any type of vaccine that can do it. It's a rather random act. So, whether it be the TBE tick vaccines or any other vaccines or any infection that you catch in the environment, it's really random. And so, I wouldn't say it's any vaccine response, it's just your immune system fighting off and mounting on response to vaccines and randomly by chance, unfortunately also attacks your brain.

[00:25:48] **Krissy Dilger:** Thank you. That's I think a hot topic for sure. And the more information that we can get out there around this issue, the more informed people can be when they're making choices.

[00:26:01] **Dr. Linda Nguyen:** I mean, in general, for our kids, we know -- we follow them, they go through the years, they require different vaccines as they age, and we have no contraindication or precaution for vaccines. Now, if they're continuing on immunosuppressive therapies for other reasons, if they have another attack or they develop something else, then certainly, that's a different discussion. But no, we don't have any caution about getting vaccines.

[00:26:35] **Krissy Dilger:** Thank you. So, this person is a parent, and they just would like to know how they can explain what is going on with ADEM in their child, so that the child siblings can understand? And this particular person I believe was having some trouble with the family dynamics post-infection or post-ADEM I should say, and how the irritability with their child who experienced ADEM has when their siblings are not understanding of their condition.

[00:27:16] **Dr. Linda Nguyen:** Definitely it's hard for parents to understand even if they're siblings to understand what's going on with their own child. So, to have the siblings also understand can be a lot tough on the family. I think in terms of explanation, it's important to know that this was a direct hit on the brain which controls everything, controls your emotions, controls your actions, your ability to see, to write, to walk. So, this is a real phenomenon that has happened to their sibling. So, to understand that I think is an important part of understanding the sequelae of what could happen because their brain has been damaged and they're recovering. In order for them to recover, they can have manifestations including irritability that may be more than they had before. So, it's important to take that into account the important organ in their body, the brain was hit and sometimes they can't control how they're behaving afterwards.

[00:28:43] **Krissy Dilger:** That's a great point. And I think that's a tricky area for sure. And something that it must be so hard for families experiencing this because ADEM, all of these disorders, they don't just affect the person they can change, how everyone in their life, at least in their immediate family has to live in the future.

[00:29:14] **Dr. Linda Nguyen:** I think it's important to understand that that can happen, the changes in irritability or mood or personality, that can certainly happen. But having additional stressors certainly doesn't help in terms of the environment, but it's important to continue to get that support for the child that's experiencing those symptoms to make sure we can help them navigate through it and deal with it and such that they're able to interact with their siblings, play with their siblings, interact with other peers.

[00:29:54] **Krissy Dilger:** Yeah, I agree. So, I think we actually have run out of questions for today but thank you so much for joining us. We really appreciate it. I think there was some really great information provided in this little chat and we hope to continue the conversation in the future.

[00:30:21] **Dr. Linda Nguyen:** It was a pleasure. Thank you.