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Practical Perspectives in Myasthenia Gravis #3: Therapeutic Selection and Monitoring Efficacy—Matching Mechanisms to Patients

Dr. Castro:
Hello. I'm Dr. Diana Castro, and this is CE on ReachMD. My colleague, Dr. Jonathan Strober and I are taking our previous conversation in this series one step further, and now we're breaking down therapy selection and monitoring our patients with generalized myasthenia gravis.

Dr. Strober:
I am so glad we're building on this discussion, Diana. Let's start our talk today with what treatment looks like for our patients. So can you tell me what standard of care is for adults with myasthenia gravis?

Dr. Castro:
Sure. So the standard of care obviously very different sometimes with our practice, because we have to do what we have to do, but the standard of care is corticosteroids, IVIg, plasmapheresis when needed—and usually that will come in acute situation in myasthenia crisis—immunosuppressors, biologics, complement inhibitors, FcRn receptor inhibitors. So we now, like you were saying before, we have way more options. Our options before were corticosteroids, IVIg, and plasmapheresis, and that was it, pretty much, because the oral medications, they were never my favorite medications.

We also use FcRn antagonists. I use it mainly in patients where—most of the time I don't do steroids, I'm going to be honest, because they can cause so many issues. And for example, patient has diabetes, like you said before, I'm not going to be able to put them, so I put them on IVIg. The problem with IVIg is that they may respond, but once you start trying to get them off, or try to space them up a little bit, they will start relapsing. So you're going to need something else.

So what else can you use? FcRn receptor antagonists not only are for steroid sparing, but also for the IVIg because it becomes—it's a whole thing having to inject yourself once a week, or having somebody coming to your house do it once a month, right? So I think the FcRn antagonists are a really nice way to move away from that.

And then in terms of monitoring, we were talking before about the scales, and I think the 10 and above children and adults and teenagers, we can use those scales to guide us in terms of the prognosis. Is this patient responding? Is this patient going to respond over time, really? Or is this patient the one that is super hard and we're going need to use everything?

So Jonathan, when you think about treatment selection in myasthenia gravis, especially in pediatrics, how do you frame that first conversation about management with the families? I mean, you're great with the families, but there are a lot of challenges that we face treating these patients.

Dr. Strober:
I mean, I agree. Like, I think it's not just as simple as saying, here is the treatment. We have so much that we can offer nowadays, and there are different ways of providing that for our patients. So it really has blossomed into this conversation of like, okay, so we have this IVIg, it's a once a month IV. We have rituximab, which I think we use a little bit more in pediatrics and maybe than in adults. And however you decide to give that, because there's no great standard of care for any of this. And then we have these newer IV medications that are every other week or once a month, if we can with some of these. Or now they're moving to subcutaneous which are approved for adults but not yet approved in pediatrics. So unfortunately, that kind of gets taken away, but there's still needles. So there still is that conversation with these families of, well, if you don't want an IV or sub-Q injectable, there are these other options and things that we've used in the past. Steroids being the big one, but it's not something that we're going to want to use long term. Maybe if they're super responsive to steroids, and you can get them down to the teeniest of doses, maybe, maybe, maybe you feel comfortable doing that in a pediatric population, maybe some of the adolescents. But it's still, in adolescents, especially, acne is a big issue, right? Things that maybe in adults, we don't worry about as much, because adults can deal with some of those things a little bit more than these adolescents who are already going through a lot of these changes. We think about long-term steroids and bone health and other issues, just infection risk, that sort of thing. So we talk about steroids.

I talk about the Imuran — mycophenolate, which I've used because it's what we had in their orals. And in those patients, where they're just like, ‘I do not want any needles. I hate needles. And I’m needle-phobic, I just can't do that,’ I mean, sometimes you're just stuck with it. And I think that's a little different. In most adults, I think you can kind of talk with them and kind of work it through. And you can kind of get away with that, oh, we have medication that we can put on to make you numb and prevent you won't feel it. And maybe in an adolescent and an adult can get that kind of concept, whereas, a younger kid just needle is—just forget it. And it becomes a really hard thing.

I think we try to think about thymectomy. We know that the earlier you do thymectomy, the higher chance that you're going to put a patient into remission in the adolescent population, especially. So that's kind of a nice thing. And then steroids kind of screw that up a little bit, right? Because poor wound healing if they're on steroids. So it's another factor that we have to put into play if we're trying to get them into the thymectomy situation.

And so really have to think about—we talked about PLEX, but that it requires a lot more. And so we tend to use it more for patients who are in crisis or really not doing well and not responding well to other things.

Dr. Castro:
I wanted to make a comment about thymectomy. I think it's very important to know now most of the time they are done with a robotic approach, so they don't have to do the full sternotomy we used to have to do for ___ back then. So it's important to remember that.

I do use thymectomy in definitely if we get MRI of the chest and the neck — I'm getting also neck included, because sometimes there are some areas that you miss in that area and then—or CT. Some people do CT scan. But I want to try to avoid the radiation for that kid. So I think it's important — thymectomy is one of those that is definitely an important piece for us to use as treatment.

Dr. Strober:
I think people do get scared of thymectomy, especially in pediatric patients. But I agree—we've been doing thoracoscopic thymectomies for a long time now, and it's a much better recovery and a much lower risk of complications, for sure. So it's definitely something that I think we need to put there. Because if I think when I talk to other people, or see the data, like thymectomy is often put off for a long time.

Dr. Castro:

Yeah. Yeah.

Dr. Strober:

But it may be something that should be thought about a little earlier. And I know in patients who are ocular only, we don't even really talk about that much, but there are some kids have really bad ocular disease that maybe would benefit from something like that. We don't know. There's still so much we have to learn, and so that’s what we have to find out.

But I think the nice thing is that we talked about is that there are a lot of new treatments out there. So we think about maybe we can put things off. So we have two approved drugs in the pediatric population. I mentioned there's the eculizumab, or the complement inhibitor that's given IV every other week, with newer ones being developed and approved for adults, but not yet in kids. And then there's the FcRn inhibitors, where there is one approved for kids 12 and over, which is wonderful that we have that.

I think one of the big hurdles that we have when we treat our patients is insurance, unfortunately, and so if you have something that's actually approved for a specific disease and this specific population, it's much easier to get insurance coverage for those patients.

So now we have nipocalimab, which has just been approved for 12 and over — the FcRn inhibitors, and actually the recent meetings they’ve been presenting the data and it really found to be well tolerated and really work the way they work in adults. So the antibody levels are dropping in these patients, as they are in adults, so showing in terms of what they need to do, they're doing. And then those patients are actually doing better in their quality measures, which is great. And it's sustained. It's a sustained treatment. And they actually were able to go from every other week, which is the standard that they use in adult population. Some patients were actually responding to once-a-month injections.

So I think that we're able to kind of use these new tools because they're being tested in kids, which I think is also — I have to give a shout-out to all the companies who are doing all these studies now and in the pediatric age groups, which we didn't have when we were young, budding child neurologists, and learning the ways. So the companies are really caring about making sure these drugs are there for the kids, and then we can use them and not have to fight for these patients to get treatments that would be really beneficial for them.

Dr. Castro:

Yeah, I'm going to make a comment about that, because I suffered before with— me, I have an experience with girls—with Hispanic girls, African American girls that will not respond to anything. I mean, I had them since they were 8 years, 10 years of age. And I keep treating, treating. Even thymectomy. And I was always waiting and waiting to get to that age that I could use something else, right? And this, when nipocalimab came and approved age 12 and above, I was like, Oh, God, I actually put a patient really quick on it, because I don't have to wait until they're 18.

So when you say about the companies, I think it's important the work they have done, because these studies are not easy to enroll. These patients are not everywhere, right? It's a rare disease, still a rare disease. So I think it's important the work they have done to be able to provide us and the patients with more options.

Dr. Strober:

And I think just to make it harder to enroll is that they need to be antibody positive, so— or to treat with them, we kind of need to be treating our antibody-positive patients until we can get more data in seronegative patients. We've been talking a lot about antibodies today, um so I think going back to that, being able to use these medications on our patients, like now, they need to be antibody positive.

And, again, we talked about the FcRn inhibitors, as well as the eculizumab, the complement inhibitors. The one thing about the complement inhibitors that we need to remind people, if they don't already know, is that the patients need to be immunized against meningococcemia, because it can occur in patients who are getting the complement inhibitors if they're not immunized. And if they're not immunized, you can put them on antibiotics while you give them the vaccine and let the vaccine kick in, so they have to be on antibiotics for a while. Which, again, people need to remember there are certain antibiotics that are contraindicated in patients with myasthenia gravis because they affect the neuromuscular junction.

So there's just a lot to think about, and a lot to remember when using these new therapies.

Dr. Strober:
So when it comes to monitoring response in children, we all use clinical scales, but what else should we be paying attention to that those tools might miss?

Dr. Castro:
Yeah, I think you mentioned something before, but I think it's so, so important, the

history, right? What are you getting from the parents? What are you getting from the kid? You mentioned something because when you ask, let's say, I'm going to give you a quick example about constipation. Do you have constipation? ‘No.’ How often do you have a development? ‘Every week.’ So that is more than constipation. So when you ask these questions, you have to ask where are you having those symptoms, right? Is that at home, or is happening more in the school? Whenever you're doing more activity? Many kids are active — soccer, tennis, like a thousand things. So you can ask for specific areas of the day or activities that they have to try to get a really good feedback. It's important to spend the time with the family getting that right.

It's important also to differentiate other things that kids may have, because kids may have ADHD, kids may have other kind of behavioral issues. So sometimes parents may try to minimize these symptoms, so we have to kind of sit with them and try to make them understand, differentiate. This could be related to ADHD, but these fatigue that comes and goes, or the fact that he wakes up in the morning fine, and then at night time is completely destroyed, that is not normal for anybody, right?

I remember a case of a girl who actually the first — well, one of the first things that mom saw was that she started school and she would come home and have the most horrible temper tantrums. But they wouldn't happen on the weekend. But it was like a daily temper tantrum, and we start talking and talking. Well, she ended up having myasthenia gravis. She was holding throughout the day, but when she came home, she felt more comfortable, and she was expressing everything she had to express, right?

So I think these kinds of questions are the ones that are going to get you to the answers you want. I don't think the tools help us more to move forward.

How is the patient responding? But we have to spend time with the families. I think that's my opinion. What do you think about that?

Dr. Strober:
Yeah, I agree. And I think it's not just about how they're functioning, but how are they feeling. I think we all get a little shy when we want to ask our patient — our adolescent, right, how are they doing with friends? Do they have a boyfriend or a girlfriend? Right? I mean, in adults, we talk about sexual life, which adolescents have a sex life, and something that we kind of, in pediatrics, we have a hard time

talking about. But it's something that we need to have that conversation with our older kids, just to make sure we're not missing something. Or do they have friends? Do they have a friend group? Do they have support at school? How are they doing in a social situation? So I think that we have to think about that as well.

Because having myasthenia sets them apart, right, just like any other chronic disease, they're dealing with treatments. They're getting infusions or injections or medication. They're not feeling well. Are they missing school? How is that affecting their overall mental health? And I think we need to make sure to ask about that too.

Dr. Castro:
Yeah, that's such an important point. I didn't mention that, but it's, to me, one of the most important points. But also to that, that's why I don't do steroids most of the time, because there is a lot going on for these poor kids, and you don't want them gaining weight. You don't want them having acne or hair. The poor girls start growing hair everywhere. So these kinds of things are important to think about. I think medicine is not just prescribing a medication; medicine is a holistic approach of the patient and the family, and that's something that many people are forgetting.

Dr. Strober:
Yeah. And on that note, it's unfortunately all the time we have for today. Thank you, Diana, for joining me, and thanks to our audience for listening. We hope you picked up some new ideas and practical tips to support your care of young patients living with myasthenia gravis.

So it's important to check with the patient and the family throughout the day. Are there difference that the patients are reporting throughout the day? But also, are they behaving differently in different settings? Right? For example, with ADHD, you may behave one way in school compared to another at home or another in clinic. So all of those are key areas to explore.