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ReachMD

www.reachmd.com

info@reachmd.com

(866) 423-7849

Turning Flares Into Function: Flag Uncontrolled Disease

Dr. Wolfe:

Thanks for joining us again on CE on ReachMD, and I'm Dr. Gil Wolfe. Here with me today is Dr. Christyn Edmundson.

So far, we've talked about setting and defining targets in gMG, but what happens when we're not getting there? In this episode, we'll talk about how to recognize when disease control is slipping and how to respond before crisis hits. Let's start with what uncontrolled really means in this context.

In practical terms, Dr. Edmundson, how do you define uncontrolled generalized MG at the bedside? What are the key patterns or signals that tell you a patient's current regimen just isn't working well enough

Dr. Edmundson:

That's a great and very important question. There is no single thing that tells you this patient has uncontrolled disease, but some things that I look for include the following: having persistent MG-ADL score greater than 4 or 6 or a lot of ongoing bulbar or limb weakness. Also, if a patient is having frequent flares, sort of frequent worsening of their MG-ADL score over time, where they're like limping along, but every couple of months, their disease really worsens in a way that's impacting their life. Another is to look at patients who are having high treatment burdens. So, patients who are on long-term, particularly moderate- or high-dose steroids, or if they're requiring frequent IVIG or plasmapheresis, which can be very time consuming for patients.

It's also important to note that any episode of myasthenic crisis makes a patient uncontrolled by definition. If they're having crisis where they're needing respiratory support, that is a patient that needs their treatment escalated.

Some bedside checkpoints that you can use are things like asking a patient if myasthenia gravis still materially limits their daily life, asking them if it feels like their treatment regimen is unstable, or asking them if they have any red flags for crisis, like are they having trouble chewing, swallowing, breathing in a way that's escalating and really suggests that they're deteriorating in a way that needs more active management.

So, let me ask you this, Dr. Wolfe, once you have identified uncontrolled disease, what does escalation look like in an outpatient practice, and are there steps that clinicians can sort of assert to help control myasthenic disease before crisis occurs?

Dr. Wolfe:

Those are great points you made at the bedside, in the clinic, or in the hospital to assess patients where disease control isn't quite good enough. So, what do you do? I mean, we know there are pretty common triggers for myasthenia gravis, the most common of which are infections or lack of adherence with medication, so you really want to confirm that they're taking medications the way they're supposed to. Rule out triggers such as infections and so forth, optimize pyridostigmine as much as possible. That drug really should not be given at a dose more than 480 mg total a day, however. And you may have to escalate immunosuppression if the patient really is not well

controlled. You may have to switch between different agents if one agent is not that well tolerated, if they haven't responded that well to an initial agent and so forth. And you really have to keep in the back of your mind steroid-sparing strategies throughout because a lot of these patients are going to be on prednisone or other corticosteroids. And the side effect burden from those is so great.

You may have to initiate or switch between targeted therapies. Like switching, say, between a complement inhibitor and a neonatal FC receptor antagonist. And then, judiciously, you should use plasma exchange or even go back to intravenous gamma globulin for stabilizing disease or providing rescue therapy if the patient is really in dire straits.

Well, I think we nailed it. Thanks so much, and we'll see you next time.