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Prioritizing Patient-Centered, Multidisciplinary Care in CIDP

Ashley:
You're listening to *On the Frontlines of CIDP* on ReachMD. I'm Ashley Baker, and today I'm joined by Dr. Andrew Lee to discuss the importance of patient-centered care in chronic inflammatory demyelinating polyneuropathy, or CIDP. Dr. Lee is a Professor of Ophthalmology, Neurology, and Neurosurgery at Weill Cornell Medical College as well as the Chairman of the Department of the Blanton Eye Institute at Houston Methodist Hospital.

Dr. Lee, it's a pleasure to have you on the program.

Dr. Lee:
Thanks for having me.

Ashley:
To start us off, Dr. Lee, can you explain what CIDP is and how it typically presents?

Dr. Lee:
CIDP is a chronic disease. That's the C, which means it's months to years in duration. It's an inflammatory disorder, which means its hallmark is inflammation, which is the type of body response to our immune system reacting to this disease. And it's a demyelinating disorder, which means it removes the covering of the nerve. So the covering is the myelin—that's like insulation on wires. And it's a polyneuropathy because it involves both sensory and motor nerves. So it's a chronic, inflammatory, demyelinating polyneuropathy—CIDP.

It normally presents with weakness and numbness, and for a neuro-ophthalmologist, the way it comes to us as it can affect the sensory nerves for the face and the vision, the optic nerve—although that's rare. And the motor nerve that we are involved in is the eye movement nerves, and so it can cause ophthalmoplegia, which means your eyes don't move properly, and it can cause the symptom of diplopia. So it's an ascending polyneuropathy. It affects both motor and sensory nerves, and that's normally how it presents.

Ashley:
And how does variability and disease progression create the need for a patient-centered approach?

Dr. Lee:
So, as with a lot of inflammatory disorders, there's variability both in the presentation and the severity of illness. So it can affect the central nervous system predominantly or the peripheral nervous system. And neuro-ophthalmology is a bridge between those two areas of the nervous system, and it can be different in its severity, which means some people can have life-threatening illness and they can't breathe or walk—that kind of severity—to milder forms of disease, especially the sensory forms where they don't have any paralysis. So that variability is what's key to patient-centered approaches for not just this disorder, but all disorders, but especially for CIDP because every case must be individualized, and that's because there's no real cure. So the treatment paradigms have to match both the severity and the variability in the disorder.

Ashley:
Now, why is multidisciplinary collaboration so important when diagnosing and treating these patients?

Dr. Lee:
So it's really important that we have multidisciplinary care for CIDP because it may affect both the motor nerves and the sensory nerves, and that means patients might need to have intensive care unit type of complexity level of services if they're having difficulty breathing,

and you need neuro-ophthalmologists like myself if they have the eye component of it. So in the acute setting, the acute inflammatory demyelinating polyneuropathy, that has an eponym called Guillain-Barré syndrome. And if it causes the double vision and the ophthalmoplegia, we call that the Miller Fisher variant. And so both the acute form, AIDP, the Guillain-Barré syndrome, and the chronic form, CIDP, can present with eye findings, and so you might need an eye doctor, a neurologist and a critical care specialist all involved in the case at the same time. We're going to have to have imaging studies to make sure it's not something else. You might need to have electrophysiologic studies, EMG and nerve conduction studies, to confirm the diagnosis, so we need those types of neurophysiologists. And then we might need a spinal tap to look for inflammation in the spinal fluid that will help confirm the diagnosis. So it really is a multidisciplinary care team. And, of course, we need the hospitalists if the person has to be hospitalized, and that sometimes occurs in patients who have severe disease.

Ashley:
For those just tuning in, you're listening to *On the Frontlines of CIDP* on ReachMD. I'm Ashley Baker, and I'm speaking with Dr. Andrew Lee about personalizing CIDP management.

So, Dr. Lee, when it comes to treating patients with CIDP, what role does shared decision-making play?

Dr. Lee:
As with other disorders that are inflammatory and autoimmune, you have a balance between the treatment and the side effects of those treatments. So our traditional treatments of immunosuppression are steroids and other immunosuppressive-type agents. In the acute setting, we might use IVIg or plasma exchange, but in the maintenance phase of the disorder, because it's chronic and there's no cure, patients have to be on agents that suppress the immune system. And so it's really critically important that the patients also be involved in the care team, but we have to involve the primary care physician, the neurologist, the neuroimmunologist, and the critical care team because not only is the disease a problem, but also the treatment and its side effects.

So there are a whole myriad of complications that can occur from corticosteroids, for example. They might have glaucoma or cataract from the treatment. So even though the disorder doesn't cause cataract and glaucoma, the steroids can cause both those disorders, so we need the eye doctor to be involved even after the acute phase to monitor for the side effects. Same with the primary care physician. Steroids can cause high blood pressure, high glucose, and osteoporosis. And, of course, the stronger immunosuppressive agents can lead to secondary infections, so it's critically important that we have the patient participate to make an informed decision about the risk and benefit of treatment versus the side effect profile and what they're willing to tolerate to suppress their disorder.

Ashley:
And for patients who are in periods of remission or functional stability, how can we individualize long-term care?

Dr. Lee:
So the same issue is, if you take someone off immunosuppression and don't use any steroids, they might get a relapse. However, you're weighing that against the benefits of avoiding the side effects. So it's really critically important that we, one, make sure that any relapse is adjudicated as a real relapse and not some side effect of the drug. And the second is, if they really are in a remission, following that person to ensure that they do not get a relapse. And so that decision-making about functional status and whether someone is truly in a remission requires communication amongst the team but also involvement of the patient. And it's really much better to be off treatment if you're in remission, and so that is the goal.

Ashley:
As we approach the end of our program, Dr. Lee, do you have any final thoughts you'd like to share with clinicians who manage CIDP?

Dr. Lee:
Yeah. So there's really two forms of inflammatory demyelinating polyneuropathy. They present with ascending polyneuropathies, weakness, and sensory loss. It can have other features. You need to rule out those other conditions. There are both electrophysiologic criteria and clinical criteria for making the diagnosis of IDP. In the acute setting, that is AIDP. And you should be cognizant then it can affect the central nervous system as well as the peripheral nervous system. If it's affecting the eyes and causing double vision, that has an eponym Miller Fisher variant, but CIDP is the chronic form, and they're both inflammatory demyelinating polyneuropathies. But CIDP doesn't have an eponym like Guillain-Barré. It's just called CIDP. And the ophthalmoplegia of CIDP has no eponym. In the acute form, that's called Miller Fisher, but it's just called ophthalmoplegia in CIDP. And it's really critical that we have a team approach to these patients because it can affect any nerve, and that is the dangerous part about the polyneuropathy. It can be life-threatening, and so we need to be vigilant about relapses and ensure remission with chronic immunosuppression and other treatments to give the patient the best quality of life we can.

Ashley:

With those key takeaways in mind, I want to thank my guest, Dr. Andrew Lee, for joining me to discuss individualized care in patients with CIDP. Dr. Lee, it was great having you on the program.

Dr. Lee:
Thanks for having me.

Ashley:
For ReachMD, I'm Ashley Baker. To access this and other episodes in our series, visit *On the Frontlines of CIDP* on ReachMD.com, where you can Be Part of the Knowledge. Thanks for listening.