

Transcript Details

This is a transcript of an educational program. Details about the program and additional media formats for the program are accessible by visiting: <https://reachmd.com/programs/frontlines-cidp/understanding-cidp-a-roadmap-to-diagnosis/33066/>

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Understanding CIDP: A Roadmap to Diagnosis

Announcer:

You're listening to *On the Frontlines of CIDP* on ReachMD. On this episode, we'll hear from Dr. Sami Khella, who's the Director of the Clinical Electrophysiology and a Professor of Clinical Neurology at the University of Pennsylvania. He'll be discussing challenges in diagnosing chronic inflammatory demyelinating polyradiculoneuropathy, or CIDP. Let's hear from Dr. Khella now.

Dr. Khella:

CIDP can present early on in two ways. The first, more typical way is that it's a slowly progressive neuropathy that goes on for more than two months—which is the minimum criteria needed to make the diagnosis, at least two months—and typically, it's more than that, where a patient may develop numbness and tingling proximal and distal weakness. So that's one of the key features of this illness.

The other way that it can present is like Guillain-Barre syndrome—an acute and fulminant presentation where the patient has the same sensory and motor complaints that develop very rapidly, leading to hospitalization and confusion with Guillain-Barre syndrome. And then, of course, the disease will continue to progress, unlike Guillain-Barre, where the progression should stop by definition by four weeks, and in the beginning, you really can't make the distinction between the two.

It can also be mistaken, when it's progressing chronically and slowly, with something like motor neuron disease or ALS because it may have minimal or no sensory symptoms. And the other thing in the differential diagnosis that you always have to keep in mind is a myelopathy—so that cord compression, especially a high cervical cord compression—can look exactly like CIDP, at least in the symptomatic phase and the symptomatic presentation, and not so much in the electrophysiologic evaluation. There are no specific biomarkers, and that's what makes this disease so hard to nail down. You need the clinical manifestations, you need the electrophysiologic manifestations, and you need to exclude other things—like I said, cervical myelopathy, multiple sclerosis, for example, or motor neuron disease, which you can then do by EMG.

But one of the interesting features of this illness is the subset of patients who don't respond typically to first-line therapies. And one of the first-line therapies, of course, is steroids or IVIG, and typically, CIDP is highly responsive to these drugs. And one of the diagnostic criteria, it's not a biomarker, but the rapid response. So if you don't have a rapid response to CIDP, either it's not CIDP or it's one of the subset illnesses that I mentioned, which are the nodopathies—so that you can have neurofascin, for example, or contactin—and you should be able to test for those. And so in some way, there is a subset of patients where the illness can be diagnosed not so much with a biomarker but with these proteins that you can measure.

Announcer:

That was Dr. Sami Khella talking about challenges in the diagnosis of CIDP. 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!