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ReachMD
www.reachmd.com
info@reachmd.com
(866) 423-7849

Identifying CIDP: A Guide to Risk Profiles and Clinical Presentation

Mr. Quigley:
Welcome to *On the Frontlines of CIDP* on ReachMD. I'm Ryan Quigley, and today I'm sitting down with Dr. Nicolas Dubuisson to discuss risk factors for chronic inflammatory demyelinating polyradiculoneuropathy, or CIDP. Dr. Dubuisson is a Postdoctoral Researcher at the Nuffield Department of Clinical Neurosciences at Oxford University in England as well as an MD of Neurology at Cliniques Universitaires Saint-Luc in Belgium.

Dr. Dubuisson, it's a pleasure having you on the program today.

Dr. Dubuisson:
Thank you very much. it's a pleasure to be here.

Mr. Quigley:
So to start us off, can you walk us through how CIDP typically presents and what sets it apart from other neuropathies?

Dr. Dubuisson:
What is important to know is that CIDP is divided in two main groups. So you have the typical CIDP, and then you have CIDP variants. For the typical CIDP, basically, the patient is usually coming with progressive or relapsing symptoms, such as weakness or sensory involvement. It's usually symmetric, and it's also involving the proximal and the distal part of the limbs. So that's also a really important point because it can differentiate them from the axonal polyneuropathy, for example, which are mainly distal, while CIDP is both proximal and distal. There is usually a sensory involvement, and there has to be at least two limbs involved, so that's also important for the diagnostic criteria. Finally, there is usually absent or reduced tendon reflexes in all limbs. And the timeframe is also important for typical CIDP because normally it has to be ongoing for at least eight weeks. And that's really important to differentiate them from Guillain-Barre syndrome, for example, where the onset is more acute and all of the symptoms are peaking within four weeks and after that it's gradually improving. In CIDP, it's continually progressing over eight weeks and even beyond that. That's for typical CIDP.

Then you have all the variants. So basically, there are five different ones. You have the distal CIDP where the problems, either sensory or motor, are predominantly involving the distal part of the limbs and usually the lower limbs. You have the multifocal CIDP, which can be both sensory and motor. It's usually asymmetric, so not like the typical CIDP I was referring to earlier. It's upper limb predominant, and it's usually involving more than one limb. You have the focal CIDP, which is only involving one limb—so you can imagine that the diagnosis here is a bit more complicated to do—the motor CIDP, where there is only motor features, and then the sensory CIDP, which is encompassing people with only sensory symptoms.

So basically, with that, you have the broad picture of the symptoms a CIDP patient can have. And to differentiate them from other neuropathies, if it's symmetric, that's probably CIDP, while asymmetrical features of progressive neuropathy could be vasculitis, for example. The fact that there is motor-predominant symptoms and sensory symptoms are a bit behind or not as pronounced is also an argument for CIDP. On the contrary, yes, you have neuropathies, such as diabetic neuropathy, where the sensory features are predominant. If you have also proximal and distal, that's also something that can differentiate CIDP from other neuropathies, such as hereditary or axonal neuropathies, where they are mostly distal.

The response to immunotherapy, of course, is also an argument to think about CIDP. On the contrary, hereditary, toxic, or metabolic neuropathies won't respond at all to this type of treatment. And finally, autonomic symptoms are usually present for amyloidosis, diabetic autonomic neuropathy, or even Guillain-Barre syndrome, while they are usually mild or absent for CIDP. So that's the most important thing to discuss here.

Mr. Quigley:
Now, as you just broke down for us, this is a very complex disease. If we zero in on demographics, what do we know about the role of age and sex in CIDP prevalence?

Dr. Dubuisson:
The first thing to know is that CIDP can occur at any age. It is more common, I would say, in adults over 50. Why? Probably because, as other autoimmune disease, it's occurring in the second half of the adulthood because then the immune system is getting less and less competent with age increasing, and that might explain why some autoreactive T cells or B cells are mostly generated in older age and can create autoimmune disease.

In term of sex, there is clearly a male predominance. It's approximately 2:1. And actually, this is unexplained. We don't know why. It's a bit contraindicative because you know that in autoimmune diseases usually there is a female predominance, but here it's the other way around, and we don't really know why.

Mr. Quigley:
And are there any particular comorbidities or clinical flags that should heighten our suspicion for CIDP?

Dr. Dubuisson:
In terms of comorbidity, I can think about three different ones. So, basically, there has been some reports saying that there is an association between diabetes mellitus and CIDP, but on the contrary, there are also some reports saying that there is no association between both, so it's a bit controversial at this moment. Then monoclonal gammopathies are also something I'm looking for when I'm testing patients. So basically, what I do when a patient is coming is a blood test—that's the first part—I check for the presence of diabetes. I check for the presence of monoclonal gammopathies with serum protein electrophoresis. Why? Because in case of MGUS or other hematological neoplasia, you can have CIDP-like presentations, and so the presence of a paraprotein or monoclonal gammopathies at the broader term is actually something you need to think about when there is a patient in front of you having symptoms that can fit with a CIDP diagnosis because the treatment might be different. You have to send them to the hematologist, and then they will decide whether there is another treatment to have. The third comorbidity is a concomitant autoimmune disease eventually because you know that when you have one autoimmune disease, you have more risk to have a second one. So if the patient already has a few autoimmune diseases—that can involve the skin, the lung—then that might be a complementary argument to say my patient maybe has CIDP.

Mr. Quigley:
For those just tuning in, you're listening to *On The Frontlines of CIDP* on ReachMD. I'm Ryan Quigley, and I'm speaking with Dr. Nicolas Dubuisson about the complex risk profile behind CIDP.

So, Dr. Dubuisson, let's continue to explore potential risk factors. As I understand it, there's growing interest about whether infections might act as triggers or risk modifiers. What does the evidence suggest?

Dr. Dubuisson:
That's a good question because I think there is no doubt that Guillain-Barre syndrome can be triggered by infectious factor, so there has been a lot of publication about that, so no doubt there. For CIDP, on the contrary, it's a bit less definitive I would say. There have been some reports saying that there was some CIDP-onset following a viral infection or even influenza vaccination, but it's complicated with CIDP because, first, it's a chronic disease, so of course you might have the patient in front of you a few weeks or maybe a few months after disease onset, so it's complicated to assess the causality between both. But at this moment I think that's a possibility, and it definitely cannot be ruled out. But I would say the evidence for now are a bit scarce.

Mr. Quigley:
And now we know that CIDP isn't considered a hereditary condition, but are there any genetic predispositions we should be aware of?

Dr. Dubuisson:
Well, as you said, CIDP is an acquired disease, so there is no single genetic variant responsible for CIDP. That's for sure. But there might be some genetic factors influencing immune regulation, for example. So a patient could have some variants in their genes that, combined with a specific triggering environmental factor, could lead to the disease. But for the moment, there has not been a lot about that. I would say that there is no reason now to achieve a genetic test for a patient with CIDP. The only reason where I would say you need to do it is when the clinical presentation is not typical and you might miss some hereditary neuropathies that sometimes are mimicking CIDP—for example, Charcot-Marie Tooth disease. Then, of course, you can do the genetic testing.

And what is interesting also about genetics is now a team in the Netherlands have shown that some variants are actually important in

term of response to treatment. So it's not in the routine clinical workup right now, but in the future, you might imagine to check for the presence of several variants and say, "Okay, this patient can respond better with IVIG, whereas this patient can respond better to another treatment," etc. So that's where we are with genetics for the moment.

Mr. Quigley:
And as we approach the end of our program, Dr. Dubuisson, do you have any final thoughts you'd like to share about recognizing and managing risk factors for CIDP for other clinicians who may be listening to this program?

Dr. Dubuisson:
Well, I think the main message of this talk is quite clear. First, the clinicians need to be aware of the presence of this disease and need to know that CIDP exists, and they might be able to spot the patient with this disease. So as I said, progressive, symmetrical, and motor-predominant neuropathy might make you think about CIDP because early recognition is really crucial in this disease, and delayed treatment can of course lead to irreversible damages, so that's really important.

Then, as soon as you have a clinical phenotype fitting with CIDP, program a blood test, check for comorbidities, and at this point, if you don't feel comfortable to go forward, then refer the patient to a neuromuscular referring center. That's really important because the earlier we have the patient in our center, the earlier we can do the treatment. And there are new treatments coming, so patients might benefit from clinical trials. There have been treatments on complement inhibitors and FcRn blockers for the moment, and so that's really interesting. And this type of treatment won't be available in all the centers—only in neuromuscular reference center. At least in Europe it's how it works. So if you don't feel comfortable with this disease, please refer it to a neuromuscular center.

Mr. Quigley:
Yes. And to your point, there is a lot of movement on treatment options for this disease, which is, of course, very encouraging. So I think that's a great way to round out our discussion, and I want to thank my guest, Dr. Nicolas Dubuisson, for walking us through the key risk factors for CIDP. Dr. Dubuisson, it was excellent having you on the program today.

Dr. Dubuisson:
Thank you. It was a pleasure. And thank you for the invitation.

Mr. Quigley:
For ReachMD. I'm Ryan Quigley. 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.