



How I Treat Chronic Inflammatory Demyelinating Polyneuropathy Podcast

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ABSTRACT

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired immune-mediated neuropathy that typically presents with progressive or relapsing, symmetric, proximal, and distal weakness of upper and lower limbs, sensory involvement of at least two limbs, and decreased or absent deep tendon reflexes. The symptoms of CIDP can be similar to those of other neuropathies, making diagnosis difficult, which can often lead to delays in correct diagnosis and treatment. The updated European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) 2021 guideline outlines a set of diagnostic criteria that help to identify CIDP with high accuracy and provides recommendations for the treatment of CIDP. The aim of this podcast, featuring Dr. Urvi Desai (Professor of Neurology, Wake Forest School of Medicine and Atrium Health Neurosciences Institute Wake Forest Baptist, Charlotte), is to discuss how the new guideline impacts diagnosis and treatment decisions in her

everyday clinical practice. Using a patient case study example, the updated guideline recommends assessing a patient for clinical, electrophysiological, and supportive criteria for CIDP, enabling a more straightforward diagnosis of either typical CIDP, a CIDP variant, or an autoimmune nodopathy. A second patient case study highlights how the new guideline no longer considers autoimmune nodopathies as CIDP, as patients with these disorders do not meet hallmark CIDP criteria. This leaves an unmet need in terms of guidance on how to treat this subset of patients. Although the new guideline has not necessarily changed treatment preference in clinical practice, the addition of subcutaneous immunoglobulin (SCIG) into the guideline now better reflects clinical practice. The guideline helps to define and categorize CIDP more simply and consistently, allowing quicker and more accurate diagnosis, leading to a positive impact on treatment response and prognosis. These real-world insights into the diagnosis and management of patients with CIDP could help guide best clinical practice and help facilitate optimization of patient outcomes.

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Key Summary Points

The newly updated European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) 2021 guideline now simplifies the clinical definition of CIDP diagnosis into “CIDP” and “possible CIDP” and also introduces the term “CIDP variants” to replace the previous term “atypical CIDP” when describing the various presentations of CIDP.

This podcast discusses how the new guideline impacts diagnosis and treatment decisions.

Using a case study example, the updated guideline recommends assessing a patient for clinical, electrophysiological, and supportive criteria for CIDP enabling a more straightforward diagnosis of either typical CIDP, a CIDP variant, or an autoimmune nodopathy, which may lead to a positive impact on treatment response and prognosis.

Another case study highlights that as autoimmune nodopathies do not meet hallmark CIDP criteria, they should no longer be considered CIDP. This leaves an unmet need in terms of guidance on how to treat this subset of patients.

Although the new guideline has not necessarily changed clinical treatment preference, the addition of SCIG into the guideline now better reflects clinical practice and the guideline helps to define and categorize CIDP more simply and consistently, allowing quicker and more accurate diagnosis, which may positively impact treatment response and prognosis.

DIGITAL FEATURES

This article is published with digital features, including the podcast audio, to facilitate under-

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TRANSCRIPT

Bill Maltas: I would like to begin with a warm welcome to our audience for today’s podcast. I am Bill Maltas, Senior Medical Science Liaison with CSL Behring Pharmaceuticals, and will serve as today’s moderator. I am honored to welcome Dr Urvi Desai, board certified physician in neurology, and currently professor of neurology at Atrium Health Wake Forest Baptist in North Carolina. I would also like to acknowledge that today’s podcast will be featured in the journal of *Neurology and Therapy*. The aim of today’s podcast is to highlight the 2021 European Academy of Neurology in association with the Peripheral Nerve Society (known as the EAN/PNS) 2021 guideline [1], and its impact to the approach of the diagnosis and treatment of chronic inflammatory demyelinating polyneuropathy (known as CIDP) in clinical practice, and to convey real-world perspective of the practical approach taken by physicians to diagnose and treat CIDP in everyday practice with the use of case studies.

Dr Desai: Thank you, Bill, for inviting me to talk about CIDP. I also want to thank the listeners of *Neurology and Therapy* for listening about this very important topic.

Bill Maltas: So, let us begin with the first question. What is chronic inflammatory demyelinating polyneuropathy (CIDP)?

Dr Desai: CIDP, or chronic inflammatory demyelinating polyneuropathy, is an immune-mediated neuropathy that typically presents with progressive or relapsing, symmetric, proximal, and distal weakness of upper and lower limbs, sensory involvement of at least two limbs, and decreased or absent deep tendon reflexes [1, 2]. This is how we can summarize CIDP in terms of clinical presentation.

CIDP is more common in males and can occur at any age, although commonly seen in patients aged 40–80, with a peak prevalence in patients aged 60–79 [3–5]. The incidence of CIDP varies between 0.15 and 0.70 cases per

100,000 person-years and prevalence of CIDP ranges from 0.7 to 7.7 cases per 100,000 people [3, 6–8]. This variation in range is partly due to different interpretations of diagnostic criteria, geographic variation, and the heterogeneous presentation of the disease itself [9]. Additionally, it is very important to note that CIDP misdiagnosis is common. Over-reliance on subjective patient-reported perception of treatment benefit, liberal electrophysiologic interpretation of demyelination, and placing an overstated importance on mild or moderate cytoalbuminologic dissociation are common diagnostic errors [10].

We should also note that, pathophysiologically, reversible conduction failure, demyelination, and secondary axonal loss can all contribute to CIDP, and may involve both humoral and cell-mediated mechanisms, with autoantibodies thought to be the primary effectors, although few target antigens have been identified [2, 11–13]. The pathophysiology of CIDP, however, is distinct from that of autoimmune nodopathies, which we will discuss a little bit later in the podcast, which is a separate category identified in the updated EAN/PNS 2021 guideline, which is mediated by immunoglobulin (Ig) G4 antibodies, and rarely IgG3 against nodal and paranodal antigens [14].

Bill Maltas: Thank you for that Dr Desai. How is CIDP diagnosed?

Dr Desai: CIDP diagnosis relies on clinical signs and electrodiagnostic criteria, and there have been many electrodiagnostic criteria published over the years, of which, the 2010 European Federation of Neurological Societies (EFNS) and Peripheral Nerve Society (PNS) deserves special mention. These 2010 EFNS/PNS criteria proposed diagnosis of CIDP into three categories; “definite”, “probable”, and “possible” CIDP based on how a patient’s disease course, electrodiagnosis, and clinical presentation fit certain criteria [9]. These clinical diagnostic criteria also stratified CIDP into “typical” and “atypical” CIDP [9]. However, the new updated 2021 EAN/PNS guideline that we just talked about now simplifies the clinical definition of CIDP diagnosis into “typical CIDP” and

“possible CIDP” [1]. This updated guideline also replaces the previous term “atypical CIDP”, which is very important to understand, with the new term “CIDP variant”, as the majority of CIDP previously categorized as “atypical” would fit into a known, well-recognized subtype of CIDP [1]. In addition to that, supportive criteria, including imaging (like ultrasound and magnetic resonance imaging), cerebrospinal fluid (CSF) analysis, nerve biopsy, and immunological testing and response to treatment, can also help to define the definitiveness of diagnosis and can be used to upgrade from the possible category to definitive category of CIDP [1]. I must say that applying this guideline in clinical practice thus allows the diagnosis of patients more simply, as either typical CIDP or a CIDP variant.

Bill Maltas: So how is “typical” CIDP defined by the guideline?

Dr Desai: So for the typical CIDP, first we need to fulfil the clinical criteria [1], that there is a progressive, symmetric, proximal and distal weakness, decreased or absent deep tendon reflexes, and sensory loss in at least two limbs, with a progressive clinical course over more than 8 weeks. Typical CIDP should fulfil this electrodiagnostic criteria as well, in at least two motor nerves and two sensory nerves. So how do we define this electrodiagnostic criteria? The motor nerve conduction criteria strongly supportive of demyelination include distal motor latency prolongation, slowing of conduction velocity, prolongation of F wave latency or absence of F waves, motor conduction block, abnormal temporal dispersion of the wave forms, or distal compound muscle action potential duration increase. However, each of these have specific parameters defined by electrodiagnostic criteria, and I would suggest the listener would refer to the literature for more details regarding each of these criteria (Supplementary Table 1). The sensory nerve conduction abnormalities would include prolongation of the sensory deep latency, reduced sensory nerve conduction potential amplitude, or slow conduction velocity.

I also want to make a point of differential diagnosis, especially in patients who have what appears to be an acute onset of a neuropathy. That we differentiate between Guillain-Barré syndrome vs acute onset CIDP, or acute CIDP which is defined as having an acute onset, but there is continued deterioration for more than 8 weeks, or relapse occurring at least three times following the initial improvement with the treatment, thus indicating a relapsing–remitting course.

Coming to possible CIDP diagnosis [1], this category fulfils, of course, the clinical criteria that we discussed in typical CIDP, which is progressive, symmetric, proximal and distal weakness, decreased or absent deep tendon reflexes, and sensory loss in at least two limbs, with a progressive clinical course over more than 8 weeks. However, it does not fulfil the electrodiagnostic criteria of a typical CIDP, meaning the motor nerve conduction criteria are seen in only one nerve and not two nerves (as required for typical CIDP), though sensory nerve conduction criteria are seen in two nerves. However, possible CIDP can be upgraded to typical CIDP if two supportive criteria are met. That is, objective improvement with intravenous immunoglobulin (IVIG), plasma exchange or steroids, imaging abnormalities, like ultrasound documenting nerve swelling at the site of conduction block, CSF analysis showing cytoalbumin dissociation, and pathological abnormalities as seen on nerve biopsy.

Bill Maltas: How are the CIDP variants, previously termed “atypical CIDP”, defined by the guideline?

Dr Desai: CIDP variants are very well-defined entities separate from typical CIDP, with specific clinical and electrodiagnostic phenotypes. To name these variants, one is a distal CIDP or distal acquired demyelinating symmetric neuropathy (DADS), which typically presents with weakness in the distal legs, and you can have sensory loss in distal arms and distal legs.

Another variant is multifocal CIDP or multifocal acquired demyelinating sensory and motor neuropathy (MADSAM)/Lewis-Sumner syndrome (LSS). The upper extremities are typically affected initially in this category, but

lower extremities would be affected later in the course of the disease.

Another variant is a focal CIDP, which occasionally would affect individual nerves, but more typically has brachial or lumbosacral plexus involvement.

Sensory CIDP; where you would have predominantly sensory symptoms and signs, including sensory ataxia, but no motor involvement, so no motor symptoms and signs, and it could be an early or transient stage before typical CIDP develops.

And last is motor CIDP, where there is a symmetric, progressive, proximal, and distal weakness, but there are normal sensations.

Bill Maltas: Are there any other updates to the guideline that affect the way you diagnose CIDP in your practice?

Dr Desai: The main change is that autoimmune nodopathies should no longer be considered CIDP as they do not meet the hallmark CIDP criteria. So, we know that the antibodies against the nodal and paranodal cell adhesion molecules like contactin-1, Caspr1, paranodal neurofascin-155, and a nodal neurofascin 140 or 186 have been discovered in a small subset of patients fulfilling the 2010 EFNS/PNS criteria for CIDP. However, this left an unmet need in terms of guidance on how to treat this subset of patients. The updated 2021 guideline, however, proposed to name these conditions as “autoimmune nodopathies” and not to regard them as CIDP variants because they have very distinct clinical features, there is no obvious inflammation on macrophage-mediated demyelination that is seen on the neuropathology in CIDP, and they respond poorly to the standard treatment, especially IVIG in particular [1].

Another point is that CIDP has also been associated with monoclonal gammopathies, for example, immunoglobulin G (IgG) or immunoglobulin A (IgA) monoclonal gammopathy of undetermined significance, as well as immunoglobulin (IgM) monoclonal gammopathy without antibodies to myelin associated glycoproteins (MAG). This new guideline states that there is insufficient evidence to consider CIDP associated with these diseases different from idiopathic CIDP [1].

Bill Maltas: Can you describe any case studies to illustrate how updates to the guideline affect diagnosis and treatment in real-life clinical practice?

Dr Desai: So, let us talk about a case which defines a typical CIDP. In September 2015, a 53-year-old female developed numbness and tingling in the fingers of both hands. She was diagnosed with carpal tunnel syndrome and underwent surgical procedures for bilateral carpal tunnel syndrome with no real improvement. After a further 10 months of continued symptoms, especially the involvement of the lower extremities, with progressive weakness in her bilateral, proximal, and distal lower extremities, she was finally referred to a neurologist.

Bill Maltas: What tests were performed and what were the outcomes of the tests?

Dr Desai: So, an electrodiagnostic study was performed, which met the electrodiagnostic criteria of CIDP. She also had a lumbar puncture for CSF protein analysis which showed that she had elevated CSF protein levels, which is a supportive criteria of CIDP diagnosis, as we just observed. The glucose and cell counts were normal, and the patient thus was diagnosed as having typical CIDP.

Bill Maltas: Were there any changes in the approach to CIDP diagnosis in clinical practice based on the updated EAN/PNS guideline, either in the context of this particular case, or just in your general experience?

Dr Desai: I should say that following the updated guidelines, assessing a patient for clinical, electrophysiological, and supportive criteria for CIDP enables a more straightforward diagnosis of either typical CIDP, or CIDP variant, or an autoimmune neuropathy, and that has been very helpful in the day-to-day clinical practice.

Bill Maltas: What therapies were used to treat this patient?

Dr Desai: So, for this patient, for induction, she was started on IVIG at 2 g/kg [body weight] and then she continued as a maintenance treatment on IVIG as 1 g/kg [body weight] every 3 weeks. She continued this therapy for around 3 years. She was then transitioned to subcutaneous immunoglobulin (SCIG). However, after

a year receiving weekly SCIG, the patient then switched back to IVIG as she noticed relapse of her symptoms.

Bill Maltas: How was the decision to switch to SCIG reached?

Dr Desai: We had discussions and they were held with the patient offering SCIG as maintenance therapy in line with the new recommendations and its associated benefit of flexibility in dose scheduling, convenience of administration, and thus greater independence [15–17]. The disadvantages of SCIG were also discussed, including the possibility of injection site reactions and some discomfort during the subcutaneous administration. I also educated the patient on the differences between the IVIG and SCIG pharmacokinetics, where SCIG helps maintain the better steady state IgG concentration. These discussions allowed the patient to make an informed decision about her choice of treatment. Once the patient had decided to switch to SCIG, I provided further guidance on the education and training on how to administer the medication independently.

Bill Maltas: And how did you determine efficacy of treatment?

Dr Desai: The patient continued follow-up visits in the clinic, with neurological examination to assess the clinical course. And we also monitor CIDP impairment and disability scales, including inflammatory neuropathy cause and treatment (INCAT), inflammatory Rasch-built overall disability scale (I-RODS), grip strength, and sometimes quantitative muscle strength to help sequentially monitor the patient's outcome in the clinic.

Bill Maltas: And how do you decide whether to revise a regimen or try a new treatment?

Dr Desai: It depends on how stable the patient is on their ongoing therapy, is there any concern for a relapse, as well as how the patient feels about the ease of administration, and how she or he is noticing benefit from the ongoing therapy.

Bill Maltas: What is your clinical experience with switching from intravenous IG to subcutaneous IG?

Dr Desai: So, this patient initially responded well to SCIG, but then felt a relapse of symptoms. The SCIG dose was increased from 0.2 to

0.4 g/kg [body weight], but the patient still had suboptimal response after 4 weeks (which was determined by electrodiagnostic assessment and symptom relapse), so we decided to switch back to IVIG. The patient was initially transitioned back to IVIG at 2 g/kg [body weight] over 4 days, which was then followed by 1 g/kg [body weight] every 3 weeks thereafter.

Bill Maltas: How would the presence of certain comorbidities change your approach of the current case study?

Dr Desai: Due to the possibility of severe allergic reactions, immunoglobulin (Ig) should not be given to patients who have had a previous allergic reaction to immunoglobulins, and due to the possibility of thrombotic events, I would also avoid the use of immunoglobulins if a patient has a history of cerebrovascular accident (CVA) or pulmonary embolism (PE).

Bill Maltas: Would the updated EAN/PNS guideline recommendations have changed your treatment approach?

Dr Desai: The updated guideline hasn't really altered my approach to treatment as such, because the updated guideline strongly recommends the use of corticosteroids, IVIG, or plasma exchange as the first-line of treatment. However, one addition was that the updated guideline added SCIG to the list of therapies for maintenance treatment of CIDP. It should also be noted that if objective response to a first-line therapy is inadequate or results in significant adverse events, the updated guideline recommends an alternative first-line treatment before considering combination of first-line or second-line therapies.

Bill Maltas: Could you describe a case where a patient's diagnosis was a little more complicated and where the updated guideline had an impact?

Dr Desai: Absolutely. So, this is a patient, who in January 2019, at age 68, she presented with a diagnosis of CIDP following 4–5 months of symptoms of distal numbness, pain, and proximal weakness. The patient initiated IVIG therapy at 2 g/kg [body weight], and then was maintained on IVIG 1 g/kg [body weight] every 3 weeks. However, she had adverse reactions to IVIG including headaches, and had a history of PE and CVA, so her IVIG treatments were

reduced to every 6 weeks by her referring previous neurologist. So, when she was referred to me, and I evaluated the patient, she had shown poor responsiveness to IVIG therapy, and also some concerns regarding the adverse effects (that we have discussed) that she already had developed, and she had ongoing symptoms of proximal weakness, neuropathic pain, ataxia, and occasional tremor. So, this constellation of the presentation raised the possibility of CIDP misdiagnosis and indicated the possibility of an autoimmune nodopathy instead, according to the clinical features described in the new guideline, and the poor responsiveness to her IVIG therapy [1].

Bill Maltas: What tests were performed, and what were the outcomes of those tests?

Dr Desai: Because of the concern for autoimmune nodopathy, we did the antibody profiling, and that revealed an abnormal antibody profile with neurofascin-155 IgM and contactin IgG antibodies. It should be noted that the contactin binding to IgG by western blot can be associated with a demyelinating neuropathy with distal or diffuse weakness (e.g., distal CIDP). Neurofascin-155 IgM antibody by western blot can be associated with chronic and acute neuropathies and with distal weakness, sensory loss and tremor, with nerve conduction showing prolongation of distal motor latencies and F wave latencies.

Bill Maltas: How does this diagnosis differ in light of the new guidelines?

Dr Desai: So as autoimmune nodopathies and monoclonal gammopathies do not meet the hallmark CIDP criteria, they should no longer be considered as CIDP. As we discussed, they have very different clinical presentation, the electrodiagnostic features could be different, but there is also different responsiveness to the standard therapies, and consideration should be given for alternative therapies, like rituximab, especially if the patient has not IgG1-, but predominantly IgG4-mediated antibody profile.

Bill Maltas: What else do you consider important practice in the treatment of patients with CIDP?

Dr Desai: Despite the availability of new guideline to help achieve optimal outcome for

the treatment of patients with CIDP, treatment and monitoring should be a multidisciplinary approach and it is important to involve the patient in the decision-making throughout the course of the disease, as we just saw with our patient who decided to switch from IVIG to SCIG, and then back to IVIG.

Bill Maltas: What are the signposts for patient dialogue?

Dr Desai: Explaining the disease heterogeneity, especially regarding the course of the disease and continued surveillance of disease progression to allow for adjustment to the frequency and the type of therapies I think are quite important.

Bill Maltas: How do you discuss selection of treatment?

Dr Desai: Discuss the pros and cons of each induction therapy with the patient, and based on the acuity of the patient's presentation, the degree of disability, as well as associated comorbidities.

Bill Maltas: What is considered best practice for managing expectations?

Dr Desai: It's very important to educate the patients about the disease heterogeneity and reassuring the patient that prognosis can be good if the disease is addressed and monitored closely and appropriately.

Bill Maltas: How do you discuss dose adjustments?

Dr Desai: Again, educating the patient about the possibility of disease relapse is important, which may require a dose adjustment, such as the frequency and dosing of IVIG or SCIG administration, or even a change in the therapy. In the case of suboptimal treatment response, as mentioned before, we need to discuss the consideration of other etiologies, such as autoimmune nodopathies. Once the patient has responded to treatment and appears to be in remission, we also should discuss the process of slowly withdrawing the treatment and continued monitoring of the patient with very watchful observation for any relapse.

Bill Maltas: What are the quality of life considerations?

Dr Desai: Quality of life issues are extremely important when considering therapies. We should aim that the patient should be living

independently, and their treatment regimens should have minimal impact on their social, economic, and quality of life.

Bill Maltas: What are your future perspectives in light of how the new guideline has affected clinical practice, and are there any unmet needs?

Dr Desai: Adherence to the precise diagnostic criteria set out in the new guideline, as well as continued education and increased awareness of CIDP by neurologists, will help to reduce the delay in CIDP diagnosis, and thus appropriate treatment. Appropriate choice of therapy in context with comorbidities is very important, as well as the tolerance of the therapies, as described in the new guideline will also improve patient outcomes. A greater understanding of the role of biomarkers and the underlying pathophysiological mechanisms of the disease in CIDP versus autoimmune nodopathies will help enable correct diagnosis. A greater availability of clinical trials in the CIDP space should also allow for newer therapies. And although the new guideline has not necessarily changed the treatment preference in clinical practice, they help to define and categorize CIDP more simply and consistently, allowing quicker and more accurate diagnosis. This will have a positive impact on treatment response and prognosis, with an eventual improvement in overall quality of life for our patients.

Bill Maltas: Thank you Dr Desai.

Dr Desai: Thank you very much.

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DATA AVAILABILITY

Not applicable.

REFERENCES

1. Van den Bergh PYK, van Doorn PA, Hadden RDM, et al. European Academy of Neurology/Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint Task Force-Second revision. *J Peripher Nerv Syst.* 2021;26(3):242–68.
2. Mathey EK, Park SB, Hughes RA, et al. Chronic inflammatory demyelinating polyradiculoneuropathy: from pathology to phenotype. *J Neurol Neurosurg Psychiatry.* 2015;86(9):973–85.
3. Broers MC, Bunschoten C, Nieboer D, Lingsma HF, Jacobs BC. Incidence and prevalence of chronic inflammatory demyelinating polyradiculoneuropathy: a systematic review and meta-analysis. *Neuroepidemiology.* 2019;52(3–4):161–72.
4. Oaklander AL, Lunn MP, Hughes RA, van Schaik IN, Frost C, Chalk CH. Treatments for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP): an overview of systematic reviews. *Cochrane Database Syst Rev.* 2017;1:CD010369.
5. Robertson EE, Donofrio PD. Treatment of chronic inflammatory demyelinating polyneuropathy. *Curr Treat Options Neurol.* 2010;12(2):84–94.
6. Dalakas MC. Advances in the diagnosis, pathogenesis and treatment of CIDP. *Nat Rev Neurol.* 2011;7(9):507–17.
7. Laughlin RS, Dyck PJ, Melton LJ 3rd, Leibson C, Ransom J, Dyck PJ. Incidence and prevalence of CIDP and the association of diabetes mellitus. *Neurology.* 2009;73(1):39–45.
8. Rajabally YA, Simpson BS, Beri S, Bankart J, Gosalakkal JA. Epidemiologic variability of chronic inflammatory demyelinating polyneuropathy with different diagnostic criteria: study of a UK population. *Muscle Nerve.* 2009;39(4):432–8.
9. Van den Bergh PY, Hadden RD, Bouche P, et al. European Federation of Neurological Societies/Peripheral Nerve Society guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society—first revision. *Eur J Neurol.* 2010;17(3):356–63.

10. Allen JA, Lewis RA. CIDP diagnostic pitfalls and perception of treatment benefit. *Neurology*. 2015;85(6):498–504.
11. Pollard JD, Armati PJ. CIDP—the relevance of recent advances in Schwann cell/axonal neurobiology. *J Peripher Nerv Syst*. 2011;16(1):15–23.
12. Berger M, McCallus DE, Lin CS. Rapid and reversible responses to IVIG in autoimmune neuromuscular diseases suggest mechanisms of action involving competition with functionally important autoantibodies. *J Peripher Nerv Syst*. 2013;18(4):275–96.
13. Koike H, Nishi R, Ikeda S, et al. Ultrastructural mechanisms of macrophage-induced demyelination in CIDP. *Neurology*. 2018;91(23):1051–60.
14. Koike H, Kadoya M, Kaida KI, et al. Paranodal dissection in chronic inflammatory demyelinating polyneuropathy with anti-neurofascin-155 and anti-contactin-1 antibodies. *J Neurol Neurosurg Psychiatry*. 2017;88(6):465–73.
15. van Schaik IN, Bril V, van Geloven N, et al. Subcutaneous immunoglobulin for maintenance treatment in chronic inflammatory demyelinating polyneuropathy (PATH): a randomised, double-blind, placebo-controlled, phase 3 trial. *Lancet Neurol*. 2018;17(1):35–46.
16. Allen JA, Gelin DF, Freimer M, Runken MC, Wolfe GI. Immunoglobulin administration for the treatment of CIDP: IVIG or SCIG? *J Neurol Sci*. 2020;408: 116497.
17. Berger M. Subcutaneous administration of IgG. *Immunol Allergy Clin North Am*. 2008;28(4): 779–802.