



Current practice patterns in CIDP: A cross-sectional survey of neurologists in the United States

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ABSTRACT

To evaluate how neurologists make decisions regarding chronic inflammatory demyelinating polyneuropathy (CIDP), we conducted a cross-sectional quantitative survey of 100 community neurologists in the United States. Only 13% cited using the European Federation of Neurological Societies/Peripheral Nerve Society guideline. In addition, variability in treatment approaches existed regarding the dose of IVIg used, the length of IVIg therapy before determining response, the outcome measures used to determine IVIg response, and the protocol for weaning off therapy. Forty-three percent reported giving doses that were lower than the recommended IVIg loading dose for CIDP. Many reported giving nonspecific patient education about the rationale of IVIg use and treatment duration. The finding that approximately half of community neurologists endorsed electrodiagnostic criteria that do not support CIDP diagnosis indicated difficulties relying heavily upon neurophysiologic studies in diagnostic guidelines. More education on CIDP diagnosis and treatment and a clear, actionable, clinically focused guideline would enhance best practices, particularly in the midst of high information flow and multiple guidelines.

1. Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a treatable disease caused by autoimmune inflammation of the peripheral nerves, yet accurate diagnosis and successful treatment of CIDP can be challenging [1,2]. The diagnosis is determined by multiple factors (ie, clinical history, electrodiagnostic studies, neurological examination, pathological examination, cerebrospinal fluid studies, and the absence of other causes of neuropathy) that may be confusing to treating physicians. The primary goal of treatment is to improve function (eg, reverse weakness and sensory loss) and maintain long-term remission without overtreating the patient. Balancing these goals to achieve optimal disease control may be difficult, even for seasoned neurologists. It has long been suspected that there may be overuse and/or misuse of long-term therapy for CIDP [3]. As IVIg is an effective yet expensive treatment, it is imperative that patients are neither denied treatment nor maintained on treatment unnecessarily. To assess this issue, we set out to learn how neurologists are making clinical decisions regarding CIDP in the midst of high information flow and multiple guidelines.

Thus, we performed an online survey with 100 board-certified community neurologists to better understand the practice patterns of community neurologists from university- and nonuniversity-affiliated practices in the CIDP field.

2. Materials and methods

2.1. Study design and methods

This study was designed to collect quantitative cross-sectional survey data from community neurologists. A 10-min online survey was administered to investigate the patterns of treatment among 100 neurologists who manage patients diagnosed with CIDP. This group included treaters from university- and nonuniversity-affiliated practices. Survey questions were developed by all 4 authors (DG, PN, JDE and JK) and finalized through discussion and input from all. The online survey underwent a pilot-test with 2 community neurologists to ensure questions were worded appropriately, response sets were comprehensive, no salient questions had been omitted, etc. Each of the 2 test physicians

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progressed through the online survey while simultaneously participating in a telephone interview with PN. Information from these 2 pilot tests was not included in the data analysis for the 100 community neurologists.

The 42-item self-administered questionnaire (Appendix A) was designed to obtain information about the guidelines, clinical features, and electrophysiologic and laboratory evaluations that the respondents typically use for diagnosing CIDP and deciding on initial therapy. If the chosen therapy was IVIg, we explored their choices for typical induction and maintenance IVIg dosages, interval of administration, length of therapy, weaning protocols, and how they identify and treat relapses. In addition, the respondents were asked about the outcome measures they typically use for determining response to therapy and about their approach to patient education, especially regarding disease and expectations for treatment. For brevity, we refer to corticosteroids as “steroids” throughout this manuscript.

All quantitative survey data were collected in December 2016. The 100 community neurologists were recruited from the e-Rewards Medical panel, which consisted of physicians who opted to be panel members and who were paid for their time and opinions. E-Rewards Medical is a leading provider of an online survey platform to the professional healthcare community. Email invitations for participation in this study were sent from e-Rewards to a sample of 1200 of its panelists, who remained anonymous to the investigators in this study. To qualify to participate, a respondent had to specialize in neurology; practice in the United States; have been in practice for at least 2 years since residency; and have treated or have consulted on at least 2 CIDP patients per year. Physicians practicing in the state of Vermont were excluded due to legal restrictions against online survey participation. All potential respondents were recruited by email invitation, which provided a general description of the survey topic (i.e., “CIDP Patient

Management”) and a link by which to access the online survey. Each invitation contained a unique identification that prevented any one respondent from taking the survey more than once. No patient data were obtained, and no questions were asked of the participants that would help in identifying them. All participant data were de-identified. Hence, this study was exempt from requiring institutional review board approval under United States Code of Federal Regulations Title 45 Part 46.101(b)(2). The study did receive a formal Letter of Exemption from the Chesapeake IRB.

2.2. Statistical analysis

Descriptive statistics were performed using *t*-tests to evaluate differences across ratio variables, including those between board certification subgroups, practice settings of the community neurologists, and years in practice. Statistical significance was assessed at the alpha level of < 0.05. Descriptive analyses were performed using SPSS (Version 23.0). Data analysis was done by PN. Grifols and the authors of this manuscript did not have knowledge of physician identity.

3. Results

3.1. Demographics of community neurologists

The online survey of the 100 community neurologists took place December 7 to 14, 2016. The respondents represented 31 of the 50 states in the United States (Fig. 1). Table 1 describes the demographics data of the respondents. The average survey-completion time was 12 min. About half (49%) of the respondents were in a university-affiliated practice; the other 51% were not affiliated with a university. Ninety-six percent of the neurologists were board-certified with some declaring



WV, West Virginia.

This map of the United States of America highlights the states from which the study participants came. N values represent the number of community neurologists who participated in the quantitative online survey phase.

Fig. 1. Geographic distribution of 100 community neurologists in this study.

Table 1
Demographic information of online survey respondents (100 community neurologists).

Number of CIDP patients per year	
Mean (SD)	31 (42.8)
Median	20
Range	2–321 ^a
Primary practice setting	
Not university-affiliated	51%
University-affiliated	49%
Board certifications (may be ≥ 1 per respondent)	
Neurology	96%
Electrodiagnostic medicine	26%
Neuromuscular medicine	22%
Primary practice type	
Solo	17%
Single-specialty partnership or group (≥ 2 physicians)	38%
Multi-specialty partnership or group (≥ 2 physicians)	45%
Number of years since residency/training	
Mean (SD)	17 (9.4)
Median	14
Range	3–38

CIDP, chronic inflammatory demyelinating polyneuropathy; SD, standard deviation.

^a Two respondents reported seeing 150 patients, 1 respondent reported seeing 175 patients, and 1 respondent reported seeing 321 patients.

additional board certifications in electrodiagnostic medicine (26%) and neuromuscular medicine (22%). Collectively, this was an experienced group of neurologists, having practiced an average of 17 years (SD, 9 years) since residency and/or training.

3.2. Diagnosing CIDP

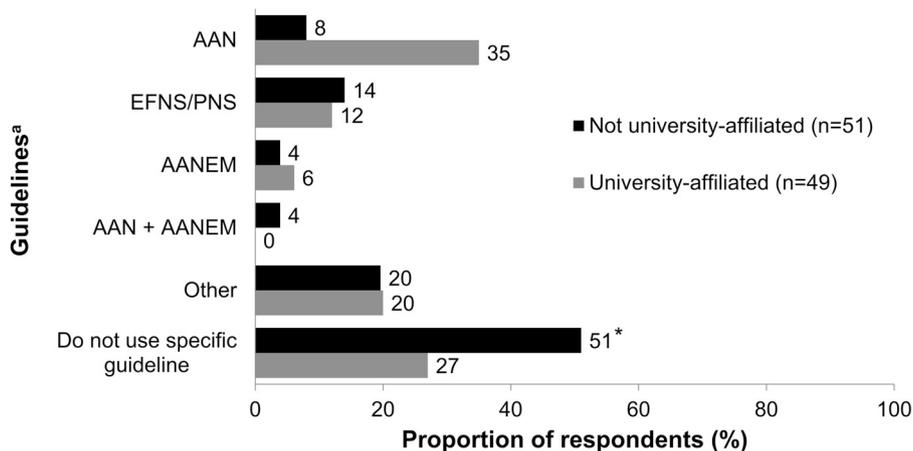
Fig. 2 shows the proportions of community neurologists who indicated using a particular guideline or no specific guideline for diagnosing CIDP based on their practice setting. Twenty-seven percent (13 of 49) of university-affiliated and 51% (26 of 51) of nonuniversity-affiliated neurologists reported that they do not routinely use a specific guideline when diagnosing CIDP. Even though the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS)

guideline [4,5] is accepted globally for both clinical and research purposes due to its balance between high sensitivity (81%–99%) and specificity (61%–97%) for CIDP, [6–9] only 13% (6 university- and 7 nonuniversity-affiliated) of the 100 community neurologists reported using the EFNS/PNS guideline. In contrast, 21% (17 university- and 4 nonuniversity-affiliated) reported using the older 1991 American Academy of Neurology (AAN) criteria [10], which was created for research purposes and can have 100% specificity for CIDP but a sensitivity as low as 3.6% [9].

When the data were analyzed based on the number of board certifications of each respondent, a higher proportion of community neurologists with multiple board certifications (74% of 34) stated they use a specific guideline when diagnosing CIDP than those with a single neurology board certification (55% of 66). Still, only 12% of the 34 neurologists with multiple board certifications reported using the EFNS/PNS guideline.

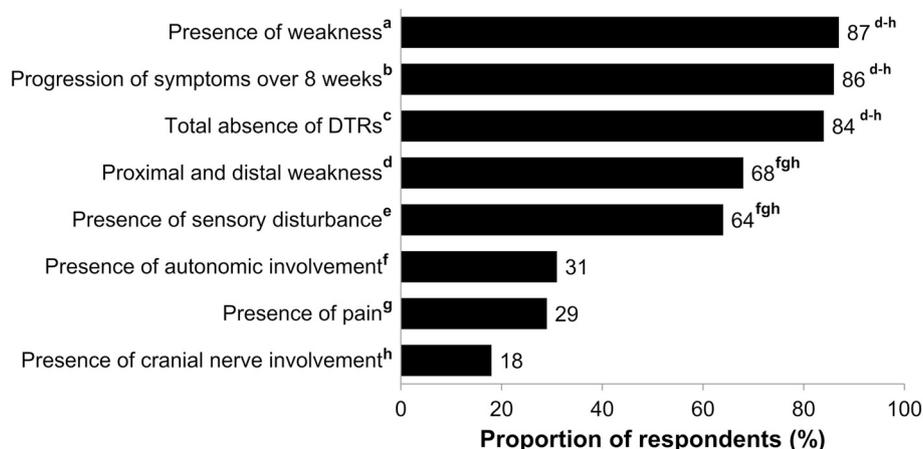
When asked about the clinical features used to diagnose CIDP (Fig. 3), community neurologists tended to rely on 4–5 clinical features. The 3 most frequently chosen features were: weakness (87%), progression of symptoms over 8 weeks (86%), and total absence of deep tendon reflexes (84%). Unexpectedly, many of the respondents cited features that are not typically associated with CIDP, such as autonomic involvement (31%), pain (29%), and cranial nerve involvement (18%). Although the majority of physicians (56%–87%) recognized the 4 most useful electrophysiological features as slow nerve conduction velocity, prolonged distal latency, temporal dispersion of CMAP, and prolonged F waves (Fig. 4), approximately half endorsed extraneous features that do not help in making a CIDP diagnosis (e.g., absent distal latency motor unit number estimate, and jitter on single fiber EMG). These preferences did not differ significantly based on the respondents' practice setting or board certification (data not shown).

As for the ability to recognize various presentations of CIDP, community neurologists identified the following as well-recognized presentations of CIDP (Fig. 5): “pure motor CIDP” (75%), “typical CIDP” (69%), “pure sensory CIDP” (65%), and “asymmetric CIDP” (60%). As anticipated, 98% of neurologists were aware that CIDP can present in atypical forms, but many may not appreciate the clear phenotypes that have been described. For example, 47% selected “Miller-Fisher CIDP.” Miller-Fisher is a variant of Guillain-Barré Syndrome, not CIDP. In addition, more than one third of the respondents considered “pain” (37%) and “fatigue” (35%) to be indicative of medically significant inflammatory neuropathies requiring treatment (data not shown), yet



Survey question: What is the specific guideline you use when diagnosing CIDP?
^a The guidelines listed are grouped unaided responses.
 Asterisk indicates statistically significant difference of $P < .05$.
 AAN, American Academy of Neurology; AANEM, American Association of Neuromuscular & Electrodiagnostic Medicine; CIDP, chronic inflammatory demyelinating polyneuropathy; EFNS, European Federation of the Neurological Societies.

Fig. 2. Proportion of neurologists indicating they use particular guideline(s) when diagnosing CIDP (N = 100).



Survey question: Which of the following clinical features do you rely on when determining diagnosis of CIDP? (Select all that apply). Responses did not differ significantly by board certifications held or whether university-affiliated. Percentage values sum to more than 100%, because some respondents selected multiple answers. Superscript letters to the right of data values indicate statistically significant difference of $P < .01$ from the corresponding clinical features to the left of the vertical axis. CIDP, chronic inflammatory demyelinating polyneuropathy; DTR, deep tendon reflex.

Fig. 3. Clinical features that neurologists rely on when diagnosing CIDP (N = 100).

pain and fatigue are less reliable prognostic indicators and are generally not used to guide dosing decisions [11,12].

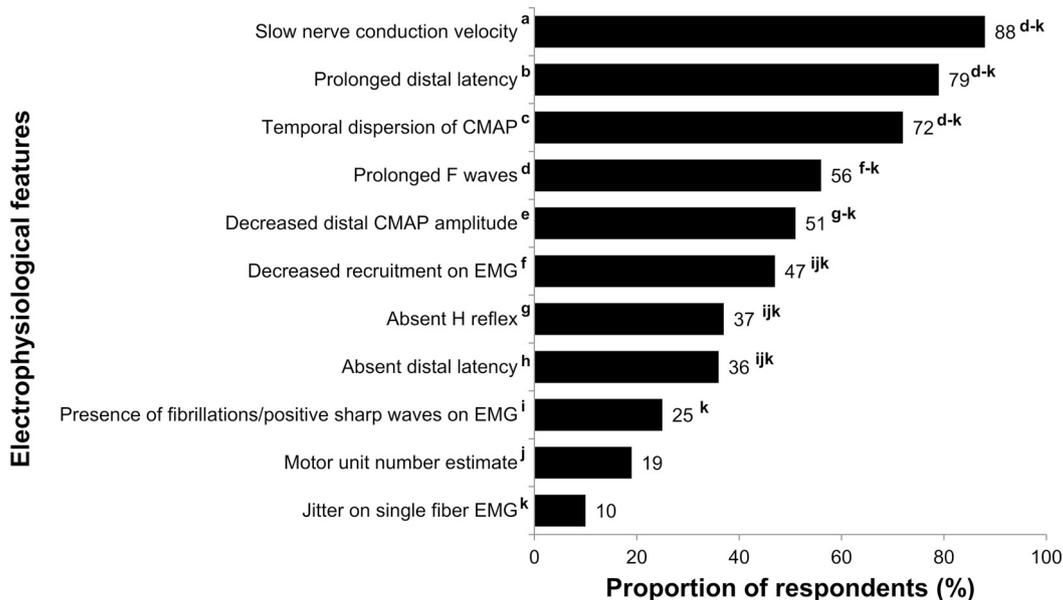
3.3. Initial therapy of choice

As for therapy preferences, 44% of the community neurologists reported using IVIg alone as their first choice of therapy (Fig. 6). Of note, 24% reported using steroids and IVIg together in first-line, even though the EFNS/PNS guideline does not support combination therapy for initial treatment [5]. Steroid alone was the first choice of therapy for

20%, but only a quarter of those respondents reported their preference for steroids was related to lower cost.

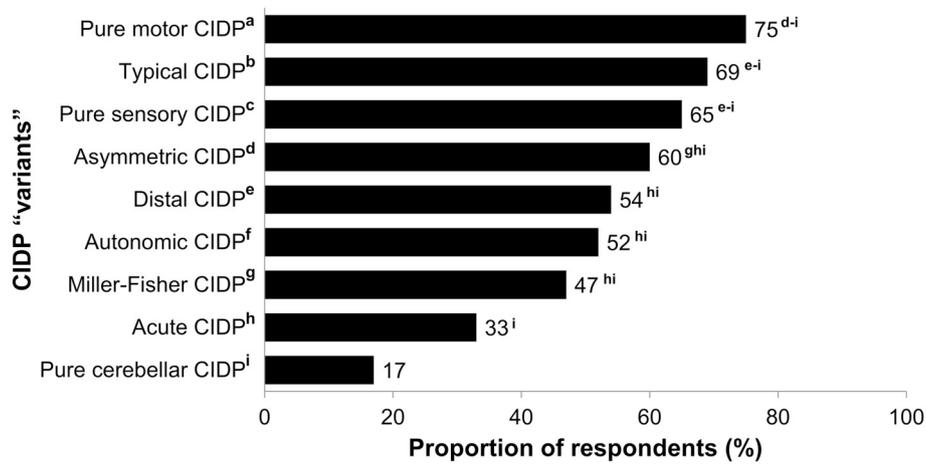
3.4. IVIg dosing for induction and maintenance

For calculating IVIg dosages, 67% of all community neurologists reported using the patient's actual weight, 25% preferred adjusted or ideal weight, and 8% chose a set dosing regimen in adult patients regardless of weight. When the data were analyzed based on practice setting, a statistically significant higher proportion of nonuniversity-



Survey question: Which of the following electrophysiologic features do you rely on when making a diagnosis of CIDP? (Select all that apply). Percentage values sum to more than 100%, because some respondents selected multiple answers. Superscript letters to the right of data values indicate statistically significant difference of $P < .01$ from the corresponding electrophysiological features to the left of the vertical axis. CIDP, chronic inflammatory demyelinating polyneuropathy; CMAP, compound muscle action potential; EMG, electronmyography.

Fig. 4. Electrophysiological features that neurologists rely on when diagnosing CIDP (N = 100).

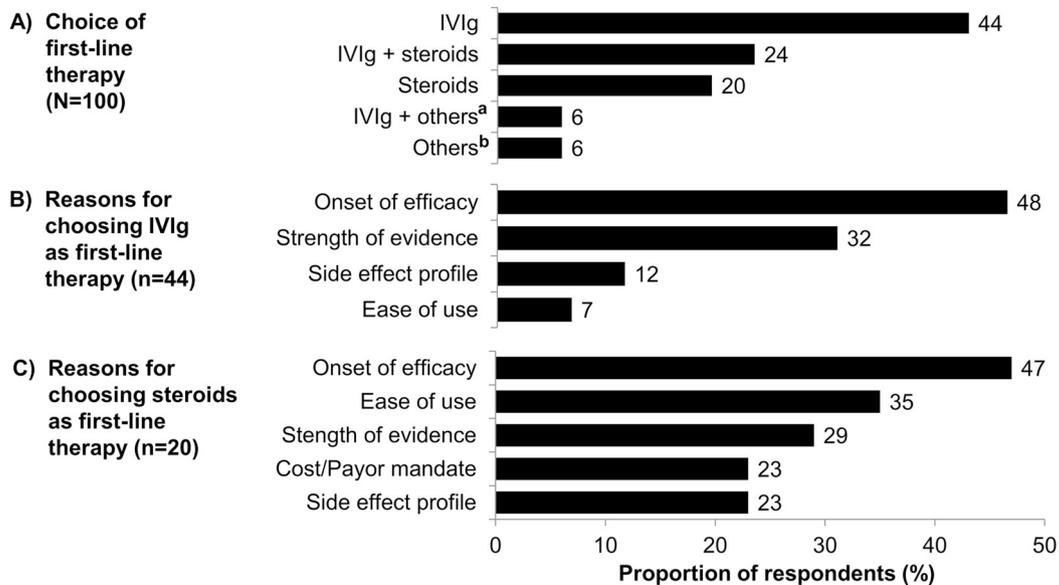


Survey question: Which of the following are well-recognized presentations of CIDP? (Select all that apply). Percentage values sum to more than 100%, because some respondents selected multiple answers. Superscript letters to the right of data values indicate statistically significant difference of $P < .01$ from the corresponding presentations to the left of the vertical axis. CIDP, chronic inflammatory demyelinating polyneuropathy.

Fig. 5. Percent of neurologists who considered each of the following as a well-recognized presentation of CIDP.

affiliated neurologists (78% of 51) reported using actual weight than those in university-affiliated practices (55% of 49) ($P < .05$). For the initial IVIg treatment, only 55% of the 100 respondents declared using the recommended loading dose of 2.0 g/kg; whereas, 39% reported using a loading dose of 0.3 kg to 0.6 kg per month, every 2–4 weeks, which is a dosing regimen more commonly used in immune globulin replacement. For the subsequent maintenance period, 44% reported ordering an average dose of 1.0 g/kg every 3 to 5 weeks for an average of 4.2 months (SD, 2.7; median, 3.0; range, 1–18 months). About 40%

of respondents reported treating their patients for at least 3 months before determining treatment response, and 25% reportedly give IVIg for 6 months. A small minority (5%) said that they give IVIg for ≥ 12 months before assessing initial response. Of note, 43% of the respondents stated that they had used a maximum IVIg induction dose of < 2.0 g/kg in patients who failed to respond. Forty-two percent reported ordering a maximum loading dose of 2.0–2.9 g/kg, but some preferred higher doses of 3.0–3.9 g/kg (4%), 4.0–4.9 g/kg (6%), and ≥ 5.0 g/kg (4%). The average reported maximum dose for IVIg



^a“IVIg + others” included IVIg + plasmapheresis (3%), IVIg + mycophenolate mofetil (2%), IVIg + rituximab (1%).
^b“Others” included plasmapheresis (3%), azathioprine (1%), cyclosporine (1%), plasmapheresis + methotrexate + cyclosporine (1%).
A) Choices of first-line therapy among all survey respondents. Survey question: Which one of the following do you typically use as a first-line therapy to treat CIDP? If you use a combination therapy, please select from the list the therapies you use in combination as a first-line therapy. Answer choices: a. steroids, b. plasmapheresis, c. IVIg, d. methotrexate, e. azathioprine, f. mycophenolate mofetil, g. cyclosporine, h. rituximab, i. bone marrow transplant. Responses did not differ significantly by board certifications held or by university affiliation. **B)** Grouped reasons for choosing IVIg among neurologists who chose IVIg as first-line therapy. Survey question: Please briefly tell us why [IVIg] is typically your choice of first-line therapy to treat CIDP.
C) Grouped reasons for choosing steroids among neurologists who chose steroids as first-line therapy. Survey question: Please briefly tell us why [steroids] is typically your choice of first line therapy to treat CIDP. Percentage values sum to more than 100%, because some respondents cited multiple reasons.
 CIDP, chronic inflammatory demyelinating polyneuropathy; IVIg, intravenous immunoglobulin.

Fig. 6. Preferences for first-line therapy of CIDP.

induction was 2.9 g/kg (SD, 8.0; median, 2.0; range, 0.1–70 g/kg) given at an average frequency of 4.1 weeks (SD, 3.3; median, 4.0; range 1–24 weeks).

As for how to determine treatment response, the most frequently selected outcome measures were Medical Research Council manual muscle strength testing (74%), gait assessment (68%), patient self-reported improvement (64%), and grip strength (61%). The Inflammatory Neuropathy Cause and Treatment scale for disability was chosen by about 30% of the 100 community neurologists, and the patient-reported inflammatory Rasch-built overall disability scale was chosen by only 17%. Six percent of the respondents chose “none of these.” Even more unanticipated was that 21% had made decisions to increase IVIg dosing via phone conversation with their patients in the absence of any objective data.

3.5. Weaning off IVIg

When asked to select the best description(s) of how the respondents counsel patients during initiation of IVIg treatment for CIDP, about 1 out of 3 community neurologists reported not discussing weaning off of IVIg therapy, and approximately 1 out of 10 tell patients that they will be on IVIg for life (data not shown). Twenty-two percent tell their patients “that we will give one dose to see if it works.” Although 70% reported telling their patients “that we will treat for X time to see if IVIg works,” only 39% reported telling their patients “that we will attempt to discontinue IVIg at some point in the future.”

When asked about the percentage of CIDP patients the respondents are able to wean off IVIg, the neurologists reported that approximately 38% of their patients (SD, 22%; median, 30%; range 5%–100%) can be weaned from IVIg therapy. Once weaned, about two thirds of patients return to the office for follow-ups within 3 months; the remaining one third have no definite follow-up plan and are seen only “as needed,” which may contribute to the reluctance of patients to wean off IVIg.

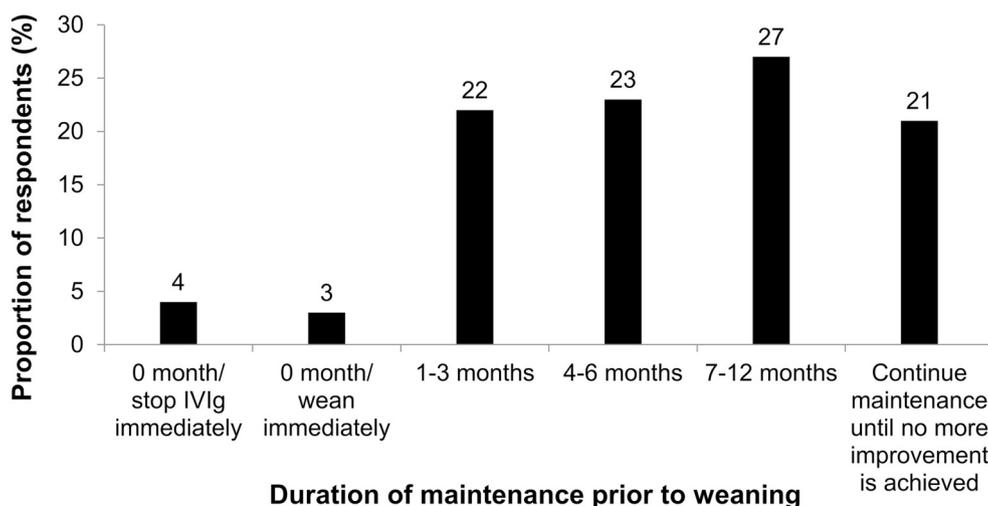
There was no clear consensus among community neurologists on the duration of maintenance prior to weaning (Fig. 7). When we performed a subgroup analysis on the data based on practice setting, university-affiliated neurologists were less likely to commit patients to IVIg

for > 3 months if the patient did not exhibit any sign of improvement on IVIg (data not shown).

4. Discussion

This was a cross-sectional quantitative survey study. We sought to understand the treatment patterns of 100 community neurologists with regard to CIDP diagnosis and treatment. There was variability among the respondents on the approach to CIDP diagnosis and treatment, as well as how to determine treatment response, length of IVIg therapy before determining response, when to wean from IVIg, and how to wean. The majority did agree on IVIg as their first choice of therapy for CIDP and steroids as their second choice. They also relied heavily on subjective outcomes (e.g., how patients feel) to determine treatment response. Many respondents were not familiar with the EFNS/PNS guideline; the majority of them reported not using any particular guideline. Many respondents also seemed to have a difficult time recognizing the electrodiagnostic criteria that support a CIDP diagnosis. In addition, they reported frequently giving IVIg doses that are lower than those recommended for CIDP, and sometimes they leave patients on treatment for long periods without attempting to wean. Some reported providing nonspecific patient education regarding the rationale for IVIg use and the appropriate duration of treatment. Others tended to not schedule follow-up visits after weaning, which may contribute to problems with weaning. The majority of community neurologists had familiarity with the clinical presentations of typical CIDP, but many thought that atypical phenotypes were more various than what have been described in guidelines. The observations that community neurologists cited electrodiagnostic criteria not supportive of a CIDP diagnosis and endorsed phenotypes not related to CIDP suggest that CIDP guidelines relying too heavily on electrophysiologic measures may not be helpful in community practice. There is also a need for more education about CIDP diagnosis and treatment, especially on atypical CIDP phenotypes.

We will not discuss details about evidence-based dosing regimens for treatments of CIDP here, as this topic has been thoroughly discussed elsewhere [14]. However, it is worth pointing out that regardless of the



Survey question: Once a good and maximal response to IVIg is reached, how long do you typically continue the maintenance dose for the patient before you start to wean the patient from IVIg, if at all? Answer choices: a. I do not continue maintenance but stop IVIg immediately; b. I do not continue maintenance but begin weaning immediately; c. Continue maintenance at least one but no more than three months and then wean; d. Continue maintenance at least three but no more than six months and then wean; e. Continue at least 6 but not more than 12 months and then wean; f. Continue until no more improvement is achieved.
IVIg, intravenous immunoglobulin.

Fig. 7. IVIg maintenance and weaning practices among neurologists (N = 100).

choice of CIDP therapy, treating CIDP requires a short-term induction period with the main objective being maximal recovery of functional disabilities and a long-term maintenance period in which the goal is to prevent deterioration, or ideally achieve remission [5,15]. The 2010 EFNS/PNS guideline [5] recommends IVIg, steroids, or plasma exchange as first-line therapies to be offered to patients with CIDP and states that IVIg has the strongest evidence supporting its use. While the majority of published evidence [16–20], support using a total loading dose of 2.0 g/kg IVIg over 2–5 consecutive days, almost half of the respondents reported using a maximum IVIg induction dose of < 2.0 g/kg in patients who failed to respond. This lower dosing regimen leads us to wonder whether those patients did not respond because they were truly nonresponsive to IVIg or because they had received suboptimal loading doses of 0.3 kg to 0.6 kg per month, every 2 to 4 weeks, which is a replacement regimen for immunodeficiency [13], not an autoimmune dosing regimen needed for patients with CIDP.

As for steroid use, the recently published *Cochrane Database of Systematic Reviews* [2] stated that the evidence supporting steroid use is low yet noted that steroids are commonly used due to their broad availability, low cost, and ease of use. One would assume that the choice to use steroids is cost-related, but less than a quarter of the community neurologists who opt to use steroids in first-line said that they would do it for cost/payer mandate.

The art of weaning off therapy is another important topic for education. There is no guideline-recommended weaning protocol. Some publications have suggested it may be better to decrease the dosages and maintain constant intervals to avoid large peak/trough IVIg concentrations, while others choose to increase the interval between treatments [15,21]. It is reasonable to consider setting clear expectations with patients an essential part of practicing medicine. Patients should be aware of how the treatment regimen would be administered, including specific details regarding how attempts will be made to wean therapy and under what conditions weaning is appropriate. We agree with other authors [12] that it is important to explain the need to reduce and withdraw from treatment before treatment is initiated. Sometimes, patients are hesitant to wean IVIg, but they are more likely to comply with regular trials of weaning if the conversation takes place early and they are informed of warning signs that might indicate they are not tolerating weaning. Unfortunately, expectations of treatment are not always discussed clearly in real-world practice. One third of the community neurologists in this survey reported not discussing weaning with their patients, and about one tenth tell patients that they will be on IVIg for life.

Findings from neurological exams, increased loss of function, and changes in patient status based on patient self-report were considered the 3 most important items community neurologists use when determining the need to re-treat a CIDP patient. Once a therapeutic regimen is established, it is important that neurologists also use objective outcomes measures, such as grip strength, walking speed and manual muscle testing, to determine treatment response. Many community neurologists reported relying on subjective measures, such as pain and fatigue, to determine treatment response. One out of 5 community neurologists in our quantitative survey reported having made decisions to increase IVIg dosing based on patient reports via phone without an in-person confirmation of the subjective report.

There are numerous reasons why physicians may not apply all of the criteria in guidelines in community practices. Difficulties in translating guidelines into clinical practice can result from the complexity of the guidelines themselves, the necessity to incorporate other potentially less familiar test results (nerve conduction studies, MRIs, etc.), insufficient physician education regarding the existence of guidelines, the time demands of busy clinical practices, the lack of attention to comorbidities, or the misalignment between the recommendations by the treating physicians and the preferences of the patients. Therefore, physician education alone may not increase utilization of guidelines. Proactive strategies to improve CIDP guideline adherence might entail

simplifying the diagnostic criteria for CIDP and addressing knowledge gaps, such as providing suggestions for maintenance dosing, duration and methods of weaning. It may also be helpful to organize formal, regularly held discussions between community neurologists and CIDP disease experts to facilitate information exchange regarding diagnostic and therapeutic strategies.

4.1. Limitations

This survey was focused predominantly on CIDP diagnosis and use of IVIg in treatment. It did not assess all of the barriers for why physicians do not follow clinical practice guidelines, such as time constraints, the relative rareness of a CIDP diagnosis, the multiplicity of neurologic diseases with which a community neurologist must be familiar, patient demands for treatment among many others. Because guidelines do not recommend combination therapy as the initial treatment for CIDP, we did not collect detailed information on the use of steroid and IVIg. We also did not collect information regarding weaning off steroids. It would have been interesting to explore these aspects of CIDP management.

A small number of physicians reported caring for an extremely large number of CIDP patients, which was unexpected but analyses without the responses from these outliers did not substantially change the results. This study reports the opinions of physicians in the United States, and thus the results may not be extrapolated to the behaviors of physicians from other countries. The nature of the e-Rewards panel may have affected how the panel filled out the form, although it is difficult to know if it would create any specific biases. That said, having representation from the majority of 50 states gives a general flavor of how CIDP is being treated throughout the country.

The results reflect the perceptions of the survey respondents. Barriers to adherence in different situations may reflect the variety of experiences of the respondents.

5. Conclusions

CIDP treatment varied among the community neurologists in this study. The majority of neurologists in our quantitative survey were not familiar with the EFNS/PNS guidelines and did not routinely use any particular guideline to diagnose or treat CIDP. The majority of community neurologists were familiar with the clinical presentation of typical CIDP but were not familiar with atypical phenotypes. They often administered IVIg doses that are lower than the recommended doses for both induction and maintenance; they also dosed less frequently. The community neurologists relied heavily, and sometimes exclusively, on subjective patient reports. In addition, they reported using combination therapy (steroids and IVIg) and often provided nonspecific information on the duration and expectations of IVIg treatment. These findings led us to believe there remains a need for a guideline on CIDP diagnosis and treatment that is brief, clear, and actionable, in particular stressing the need for objective outcomes when determining response to therapy. Furthermore, it may be impactful to pursue proactive changes (eg, more IT innovations that support clinical decisions, enhanced training for medical students, residents, fellows, and practicing physicians on guideline usage, patient education through CIDP Foundation, and greater access to recognized experts) to support guideline adoption and adherence.

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Declaration of interest

DG is an employee of Grifols.

JK reported personal fees for his scientific consultancy on various projects with Grifols.

JDE reported personal fees for his advisory board participation with Baxalta and speakers' bureau engagement with Grifols, other from GBS/CIDP Foundation International, and grants from Elsevier, outside of the submitted work.

PN reported personal fees from One Research, LLC during the conduct of this study.

Author's contributions

DG, JK, and JDE designed the study and generated the questionnaires.

PN consulted on study design and services for data collection and data analyses.

DG and JDE provided critical review of the manuscript. All authors participated in the interpretation of the data and approved the final draft.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.jns.2018.11.031>.

References

- [1] J.M. Vallat, C. Sommer, L. Magy, Chronic inflammatory demyelinating polyradiculoneuropathy: diagnostic and therapeutic challenges for a treatable condition, *Lancet Neurol.* 9 (4) (2010) 402–412.
- [2] A.L. Oaklander, M.P. Lunn, R.A. Hughes, I.N. van Schaik, C. Frost, C.H. Chalk, Treatments for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP): an overview of systematic reviews, *Cochrane Database Syst. Rev.* 1 (2017) CD010369.
- [3] H.S. Patwa, V. Chaudhry, H. Katzberg, A.D. Rae-Grant, Y.T. So, Evidence-based guideline: intravenous immunoglobulin in the treatment of neuromuscular disorders: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology, *Neurology* 78 (13) (2012) 1009–1015.
- [4] Force. E-PJT, 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, *J. Peripher. Nerv. Syst.* 10 (3) (2005) 220–228.
- [5] P.Y. Van den Bergh, R.D. Hadden, P. Bouche, D.R. Cornblath, A. Hahn, I. Illa, C.L. Koski, J.M. Leger, E. Nobile-Orazio, J. Pollard, 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.* 17 (3) (2010) 356–363.
- [6] Y.A. Rajabally, A.J. Fowle, P.Y. Van den Bergh, Which criteria for research in chronic inflammatory demyelinating polyradiculoneuropathy? An analysis of current practice, *Muscle Nerve* 51 (6) (2015) 932–933.
- [7] Y.A. Rajabally, G. Nicolas, F. Pieret, P. Bouche, P.Y. Van den Bergh, Validity of diagnostic criteria for chronic inflammatory demyelinating polyneuropathy: a multicentre European study, *J. Neurol. Neurosurg. Psychiatry* 80 (12) (2009) 1364–1368.
- [8] C.L. Koski, M. Baumgarten, L.S. Magder, R.J. Barohn, J. Goldstein, M. Graves, K. Gorson, A.F. Hahn, R.A. Hughes, J. Katz, et al., Derivation and validation of diagnostic criteria for chronic inflammatory demyelinating polyneuropathy, *J. Neurol. Sci.* 277 (1–2) (2009) 1–8.
- [9] A. Breiner, T.H. Brannagan 3rd, Comparison of sensitivity and specificity among 15 criteria for chronic inflammatory demyelinating polyneuropathy, *Muscle Nerve* 50 (1) (2014) 40–46.
- [10] D.R. Cornblath, A.K. Asbury, J.W. Albers, T.E. Feasby, et al., Research criteria for diagnosis of chronic inflammatory demyelinating polyneuropathy (CIDP). Report from an Ad Hoc Subcommittee of the American Academy of Neurology AIDS Task Force, *Neurology* 41 (5) (1991) 617–618.
- [11] J.A. Allen, R.A. Lewis, CIDP diagnostic pitfalls and perception of treatment benefit, *Neurology* 85 (6) (2015) 498–504.
- [12] Y.A. Rajabally, Long-term immunoglobulin therapy for chronic inflammatory demyelinating polyradiculoneuropathy, *Muscle Nerve* 51 (5) (2015) 657–661.
- [13] J.S. Orange, E.M. Hossny, C.R. Weiler, M. Ballow, M. Berger, F.A. Bonilla, R. Buckley, J. Chinen, Y. El-Gamal, B.D. Mazer, et al., Use of intravenous immunoglobulin in human disease: a review of evidence by members of the Primary Immunodeficiency Committee of the American Academy of Allergy, Asthma and Immunology, *J. Allergy Clin. Immunol.* 117 (4 Suppl) (2006) S525–S553.
- [14] I. Kleyman, T.H. Brannagan 3rd, Treatment of chronic inflammatory demyelinating polyneuropathy, *Curr. Neurol. Neurosci. Rep.* 15 (7) (2015) 47.
- [15] M.E. Adrichem, F. Eftimov, I.N. van Schaik, Intravenous immunoglobulin treatment in chronic inflammatory demyelinating polyradiculoneuropathy, a time to start and a time to stop, *J. Peripher. Nerv. Syst.* 21 (3) (2016) 121–127.
- [16] J.R. Mendell, R.J. Barohn, M.L. Freimer, J.T. Kissel, W. King, H.N. Nagaraja, R. Rice, W.W. Campbell, P.D. Donofrio, C.E. Jackson, et al., Randomized controlled trial of IVIg in untreated chronic inflammatory demyelinating polyradiculoneuropathy, *Neurology* 56 (4) (2001) 445–449.
- [17] R.A. Hughes, P. Donofrio, V. Bril, M.C. Dalakas, C. Deng, K. Hanna, H.P. Hartung, N. Latov, I.S. Merkies, P.A. van Doorn, et al., Intravenous immune globulin (10% caprylate-chromatography purified) for the treatment of chronic inflammatory demyelinating polyradiculoneuropathy (ICE study): a randomised placebo-controlled trial, *Lancet Neurol.* 7 (2) (2008) 136–144.
- [18] A.F. Hahn, C.F. Bolton, D. Zochodne, T.E. Feasby, Intravenous immunoglobulin treatment in chronic inflammatory demyelinating polyneuropathy. A double-blind, placebo-controlled, cross-over study, *Brain* 119 (1996) 1067–1077 Pt 4.
- [19] M. Vermeulen, P.A. van Doorn, A. Brand, P.F. Strengers, F.G. Jennekens, H.F. Busch, Intravenous immunoglobulin treatment in patients with chronic inflammatory demyelinating polyneuropathy: a double blind, placebo controlled study, *J. Neurol. Neurosurg. Psychiatry* 56 (1) (1993) 36–39.
- [20] N. Thompson, P. Choudhary, R.A. Hughes, R.M. Quinlivan, A novel trial design to study the effect of intravenous immunoglobulin in chronic inflammatory demyelinating polyradiculoneuropathy, *J. Neurol.* 243 (3) (1996) 280–285.
- [21] Y.A. Rajabally, H. Seow, P. Wilson, Dose of intravenous immunoglobulins in chronic inflammatory demyelinating polyneuropathy, *J. Peripher. Nerv. Syst.* 11 (4) (2006) 325–329.