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Original research

Comparison of the diagnostic accuracy of the 2021 EAN/PNS and 2010 EFNS/PNS diagnostic criteria for chronic inflammatory demyelinating polyradiculoneuropathy

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ABSTRACT

Objectives To compare the sensitivity and specificity of the 2021 European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) diagnostic criteria for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) with those of the 2010 European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS).

Methods Sensitivity and specificity of the two sets of criteria were evaluated in 330 patients with CIDP and 166 axonal peripheral neuropathy controls. Comparison of the utility of nerve conduction studies with different number of nerves examined and of the sensitivity and specificity of the two criteria in typical CIDP and its variants were assessed.

Results EFNS/PNS criteria had a sensitivity of 92% for possible CIDP and 85% for probable/definite CIDP, while the EAN/PNS criteria had a sensitivity of 83% for possible CIDP and 74% for CIDP. Using supportive criteria, the sensitivity of the EAN/PNS criteria for possible CIDP increased to 85% and that of CIDP to 77%, remaining lower than that of the EFNS/PNS criteria. Specificity of the EFNS/PNS criteria was 68% for possible CIDP and 84% for probable/definite CIDP, while the EAN/PNS criteria had a specificity of 88% for possible CIDP and 98% for CIDP. More extended studies increased the sensitivity of both sets of criteria by 4%–7% but reduced their specificity by 2%–3%. The EFNS/PNS criteria were more sensitive for the diagnosis of typical CIDP while the EAN/PNS criteria were more specific for the diagnosis of distal and sensory CIDP.

Conclusions In our population, the EAN/PNS criteria were more specific but less sensitive than the EFNS/PNS criteria. With the EAN/PNS criteria, more extended nerve conduction studies are recommended to obtain an acceptable sensitivity while maintaining a high specificity.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ The sensitivity and specificity of the newly published 2021 EAN/PNS diagnostic criteria for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) are unknown and their diagnostic accuracy has never been compared with that of the 2010 EFNS/PNS criteria.

WHAT IS THIS STUDY ADDS

⇒ The EAN/PNS criteria were more specific but less sensitive than the EFNS/PNS criteria. With the EAN/PNS criteria, more extended nerve conduction studies are recommended to obtain an acceptable sensitivity while maintaining a high specificity.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ These results demonstrate the possibility of usefully and reliably utilizing the EAN/PNS diagnostic criteria for CIDP. Further studies are needed to replicate our findings.

INTRODUCTION

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a heterogeneous neuropathy characterised by a chronic relapsing or progressive course, an elusive pathogenesis and response to immune therapies.¹ After the first formal definition of CIDP in 1975,² numerous sets of diagnostic criteria have been proposed, but no single diagnostic biomarker has been found to date, and diagnosis still relies on clinical manifestations and nerve conduction studies, possibly supported by some additional diagnostic examinations.

Since their first formulation in 2006, the diagnostic criteria for CIDP proposed by the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS)³ have proven to provide

an optimal balance between sensitivity (78.5–96.7%) and specificity (79.3%–96.1%),^{4–6} thus emerging as the best criteria compared with the numerous others proposed criteria.^{7–14} The EFNS/PNS criteria in the 2010 first revision¹⁵ were also confirmed to have high accuracy values (sensitivity of 73.2–76.8% and specificity of 84.2–88.2% for definite/probable CIDP).⁵ Despite their worldwide acceptance and use in research,¹⁶ several studies showed that misdiagnosis of CIDP commonly occurs, particularly in CIDP variants,^{17–19} even in patients fulfilling diagnostic criteria based on correctly interpreted test.^{17–20} A second revision of the EFNS/PNS criteria has been published (now named the European Academy of Neurology/Peripheral Nerve Society [EAN/PNS] criteria) aiming at improving the specificity of the criteria.²¹ The most relevant changes include the reduction of the levels of diagnostic certainty to two categories (CIDP and possible CIDP), the definition of specific electrodiagnostic criteria for each CIDP variant, the inclusion of sensory nerve conduction studies among the mandatory electrodiagnostic criteria, and the exclusion of chronic immune sensory polyradiculopathy (CISP) and autoimmune nodopathies from the diagnosis of CIDP.²¹

The objective of this study was to evaluate the sensitivity and specificity of the 2021 EAN/PNS criteria in comparison with the 2010 EFNS/PNS criteria in a large population of patients with typical CIDP and its variants and controls. Comparison of nerve conduction studies with different number of nerves examined was also made as was comparison of the sensitivity and specificity of the two sets of criteria in typical CIDP and variants.

MATERIALS AND METHODS

Study population

Patients with CIDP

Patients with CIDP were included from the Italian CIDP database, a web-based registry where, so far, data from 524 patients fulfilling the 2010 EFNS/PNS criteria for possible, probable or definite CIDP, including 414 with typical CIDP, 37 with distal acquired demyelinating symmetric polyneuropathy (DADS), 25 with Lewis-Sumner syndrome (LSS), 29 with motor CIDP and 19 with sensory CIDP are included. The database also includes data of 70 patients with a clinical diagnosis of CIDP (51 with typical CIDP, 10 with DADS, 2 with LSS, 4 with motor CIDP and 3 with sensory CIDP) made by neurologists expert in peripheral neuropathies, but not fulfilling the same criteria. Diagnosis of CIDP was made by the treating neurologist and reviewed by the coordinating centre (PED and EN-O). Data monitoring included diagnosis revision, suspect double entries, missing data and plausibility checks. The reasons for suspecting CIDP when nerve conduction studies were not diagnostic were also reported by the treating neurologist and included, beside a clinical history and presentation consistent with CIDP, abnormality of supportive tests (cerebrospinal fluid (CSF) analysis, ultrasound or magnetic resonance (MR) imaging of the nerves and plexus, sensory conduction studies or somatosensory-evoked potentials (SSEP), nerve biopsy and response to previous therapy) and a relapsing course of the disease.²²

At enrollment, all patients underwent a detailed clinical history.²² This information was integrated with the data reported in the medical records. Results of previously performed examinations, including CSF analysis, nerve ultrasound or brachial/lumbosacral plexus MR examination, SSEP and sural nerve biopsy, were reported when available. The results of nerve conduction studies performed during the course of the disease were included. Response to treatment was defined as a subjective improvement that was objectively confirmed by an increase in at

least 2 points in the Medical Research Council sum score (range 0–60) or at least 1 point in the Inflammatory Neuropathy Cause and Treatment disability score (range 0–10).^{21 23}

At the time of inclusion, patients with an alternative diagnosis for the neuropathy or high titres of anti-MAG (myelin-associated glycoprotein) antibodies (over 7000 Bühlmann Titer Units by Bühlmann method)²⁴ or without available nerve conduction studies were excluded.

Controls

To determine the specificity of the two sets of criteria, we used the clinical and electrophysiological data from 166 control patients suffering from sensory or sensorimotor or motor axonal peripheral neuropathy regularly followed at our outpatient peripheral neuropathy clinic (Humanitas Research Institute). The control population included patients with diabetic peripheral neuropathy (n=72), chemotherapy-induced neuropathy (n=41), idiopathic neuropathy (n=30), vasculitic neuropathy (n=13), IgG paraproteinemic neuropathy (n=5), vitamin B12 deficiency neuropathy (n=5). Similar control populations have been used in previous CIDP studies.^{4–6} Differently from the EFNS/PNS criteria, the EAN/PNS criteria recommend the use of separate sets of electrophysiological criteria for the diagnosis of typical CIDP and its variants.²¹ To evaluate specificity of the EAN/PNS criteria, the clinical neuropathy presentation of each control patient was classified according to the EAN/PNS clinical criteria and the most appropriate set of EAN/PNS electrodiagnostic criteria was applied based on the control patient clinical presentation (eg, the EAN/PNS electrodiagnostic criteria for sensory CIDP were applied in control patients with a clinical presentation of pure sensory polyneuropathy).

Study design

In view of the retrospective design, electrophysiological studies had been performed in a non-standardised manner, the number of motor nerves studied having varied from two to eight. We preferred to analyse pretreatment electrophysiological studies, if these were not available, then a later or post-treatment study was selected. The extensiveness of the study of arm nerves varied from the distal forearm segment only, to a full-length study up to Erb's point. Patients were also managed in a non-standardised fashion, as ancillary examinations and treatments were selected according to the clinical judgement of the treating physician. Although this variability may be suboptimal for research purposes, it likely reflects real-life clinical practice. The compound muscle action potentials (CMAPs) were evoked from the median nerve (stimulating at wrist, elbow and, in some cases, axilla, Erb's point and recording at the abductor pollicis brevis muscle), ulnar nerve (stimulating at wrist, below elbow and, in some patients, above elbow, axilla, Erb's point and recording at the abductor digiti minimi muscle), common peroneal nerve (stimulating at ankle and fibular neck and recording at the extensor digitorum brevis muscle) and tibial nerve (stimulating at ankle and popliteal fossa and recording at the abductor hallucis muscle). Sensory nerve conduction studies were performed along the median, ulnar and sural nerves, and prolonged distal latency, or reduced sensory nerve action potential (SNAP) amplitude, or slowed conduction velocity outside of normal limits were evaluated. All nerve conduction studies were performed at a temperature of at least 33°C at the palm and 30°C at the external malleolus. Age-dependent reference values for sural SNAP amplitude were considered. Results were analysed according to each laboratory's range of normal values, and presence of demyelinating range values determined for each relevant parameter. Prolongation of F-wave

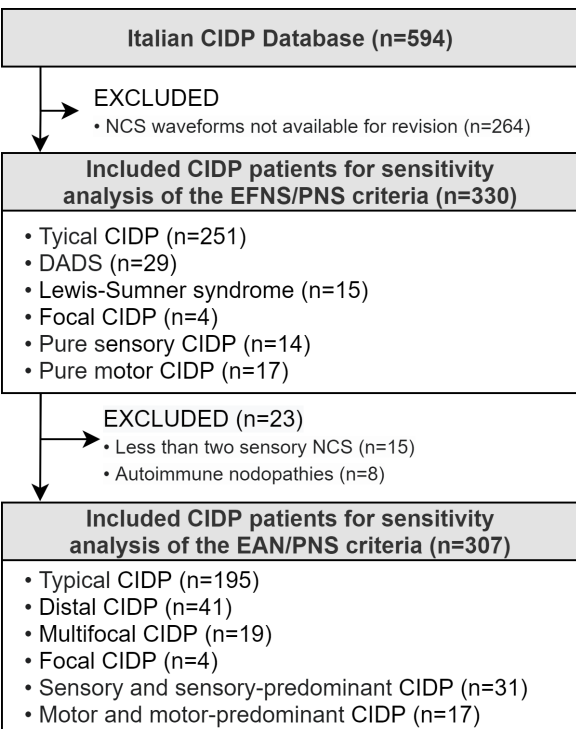


Figure 1 Flowchart of patients' selection for sensitivity analyses of the EFNS/PNS and EAN/PNS criteria for CIDP. CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; DADS, distal acquired demyelinating symmetric neuropathy; EAN/PNS, European Academy of Neurology/Peripheral Nerve Society; EFNS/PNS, European Federation of Neurological Societies/Peripheral Nerve Society; n, number; NCS, nerve conduction studies.

latency was calculated using the corrected cut-off of $\geq 20\%$.²⁵ To evaluate distal CMAP duration prolongation, nerve conduction study waveforms of the CIDP and control patients were reviewed and measurements were redone following the indications of the two sets of criteria.^{15 21} Patients for whom nerve conduction study waveforms were not available for revision were excluded from the study analyses. To evaluate sensitivity and specificity of the EAN/PNS criteria, the analysis of the distal CMAP duration prolongation was performed taking into account the low frequency filter used by the individual centres.²¹ For each patient, the clinical and electrophysiological data were reviewed to determine fulfilment of the two sets of criteria. Sensitivity and specificity analyses were first ascertained in all included patients regardless of the number of nerves examined, in order to test the two sets of criteria using real-life data, and then repeated in the patients and controls with at least four motor and four sensory nerves examined ('extended nerve conduction study protocol'). Since the EAN/PNS criteria mandatorily require sensory nerve conduction studies for the definition of typical CIDP and variants,²¹ patients with less than two tested sensory nerves were excluded from this analysis. We also excluded, from the analysis of the sensitivity of the EAN/PNS criteria, patients with anti-paranode antibodies or with a diagnosis of CISP.²¹

Sensitivity and specificity of the two sets of criteria were also specifically assessed in each of the individual CIDP clinical forms, and comparison between the two sets of criteria was made. Given the absence of precise clinical diagnostic criteria for the CIDP variants in the 2010 EFNS/PNS guidelines, for this analysis, we compared our previously published criteria for atypical CIDP²⁶ and those of the EAN/PNS.²¹

Statistical analysis

The comparative diagnostic gain in sensitivity and specificity, achieved with the use of the more sensitive or specific set of criteria, was calculated using a McNemar test. Sensitivity and specificity were compared between patients with typical CIDP and patients with CIDP variants with the Fisher's exact test for both sets of criteria separately. Diagnostic accuracy was calculated as the proportion of correctly classified subjects (TP+TN) among all subjects (TP+TN+FP+FN). All tests were two-tailed and the significance level was set to 0.05. Analyses were performed with SAS V.9.4 (SAS Institute, Cary, North Carolina). Sensitivity and specificity were assessed taking into account the number of nerves examined, in line with previous studies.

RESULTS

Among the 594 patients with CIDP included in the database, 264 patients for whom nerve conduction study waveforms were not available for revision were excluded, leading to a final study population of 330 patients. Of those, 202 had undergone an extended nerve conduction study. All these patients were used to determine the sensitivity of the EFNS/PNS criteria (figure 1). Twenty-three patients were not included in the calculation of sensitivity of the EAN/PNS criteria (figure 1) as studied with fewer than two sensory nerves (n=15) or because had positivity for neurofascin-155 (Nfasc155) or neurofascin-140/186 (Nfasc140/186), or contactin-1 (CNTN1), or contactin-associated protein 1 (Caspr1) (n=8) antibodies that were tested in 267 patients.^{27 28} Of the 166 control patients, 69 had undergone an extended nerve conduction study. Demographic and clinical characteristics and number of nerves examined with nerve conduction studies in patients with CIDP and controls are summarised in table 1. All control patients had clinical features that were consistent with the EAN/PNS clinical criteria for the CIDP variants. Since none of the control patients had a clinical presentation consistent with the EAN/PNS clinical criteria for typical CIDP, the specific set of criteria for typical CIDP could not be included in the analysis of specificity of the EAN/PNS criteria. Table 2 shows how the EAN/PNS electrodiagnostic criteria were applied to control patients based on their clinical neuropathy presentation.

Table 3 shows sensitivity of the two sets of criteria. Sensitivity of the EFNS/PNS criteria was 85% for probable/definite CIDP and 92% for possible CIDP, while sensitivity of the EAN/PNS was 74% for CIDP and 83% for possible CIDP. Using extended nerve conduction study protocol, sensitivity of the EFNS/PNS criteria rose to 90% for probable/definite CIDP and 96% for possible CIDP, while sensitivity of the EAN/PNS rose to 81% for CIDP and 88% for possible CIDP. When we also considered supportive criteria, sensitivity of the EAN/PNS criteria for possible CIDP increased to 85%, resulting in six additional patients fulfilling the criteria, but still the EFNS/PNS criteria were more sensitive (92% vs 85%) (table 3). In patients who have had diagnostic upgrading with the supportive criteria, considering the two groups together (EFNS/PNS and EAN/PNS), the supportive criteria that most frequently allowed a diagnostic improvement were response to treatment in 23 (88%) of 26 treated patients, CSF analysis in 21 (78%) of 27 tested patients and nerve imaging in 3/5 (60%). Nerve biopsy was performed in only one patient and was not diagnostic. Sensory electrophysiology improved the diagnosis with the EFNS/PNS criteria in 6 (21%) of 29 tested patients.

When we restricted the analysis to patients who could access the diagnosis with both sets of criteria, thus excluding those with anti-paranode antibody positivity or with less than two sensory

Table 1 Demographic, clinical characteristics and number of nerves examined with nerve conduction study in CIDP patients and controls

| | CIDP patients (n=330) | Control patients (n=166) |
|---|-------------------------|--------------------------|
| Gender, male, n (%) | 220/330 (67%) | 114/166 (69%) |
| Age at onset, mean (SD) | 47.9 (17) | 61.9 (13) |
| N of motor nerves examined at NCS, mean (range) | 5.6 (2–8)*; 5.7 (2–8) † | 4.5 (2–8) |
| N of sensory nerves examined at NCS, mean (range) | 4.1 (0–6)*; 4.3 (2–6)† | 3.4 (2–6) |
| Response to treatment, n (%) | 250/295 (85%) | |
| Increased CSF proteins/tested, n (%) | 197/259 (76%) | |
| Nerve biopsy; positive/tested, n (%) | 11/29 (38%) | |
| Nerve imaging; positive/tested, n (%) | 36/46 (78%) | |
| INCAT at enrolment, mean (range) | 2.6 (0–10) | |

* Patients included in the analyses of the EFNS/PNS criteria.
 † Patients included in the analyses of the EAN/PNS criteria.
 CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; CSF, cerebrospinal fluid; EAN/PNS, european academy of neurology/peripheral nerve society; EFNS/PNS, european federation of neurological societies/peripheral nerve society; INCAT, inflammatory neuropathy cause and treatment disability scale; NCS, nerve conduction studies; N, number.

nerve conduction studies available, who cannot fulfil the EAN/PNS criteria for CIDP, we found that 282/307 (92%) patients examined fulfilled the EFNS/PNS criteria (possible CIDP), in comparison with 254/307 (83%) patients who met the EAN/PNS criteria (possible CIDP) (McNemar test; $p < 0.001$). There were 29 missed diagnoses (possible CIDP) with the EAN/PNS electrodiagnostic criteria, consisting in 25 patients fulfilling the motor electrodiagnostic criteria but not the sensory ones,

Table 2 EAN/PNS electrodiagnostic criteria applied to control patients based on their clinical neuropathy presentation and consistent with the EAN/PNS clinical criteria for typical CIDP or its variants

| Clinical diagnosis of control patients; (n of control patients) | Set of EAN/PNS electrodiagnostic criteria applied in each control patient; (n of control patients) |
|---|--|
| Diabetic sensorimotor polyneuropathy (17) | EDX criteria for distal CIDP (34) |
| Chemotherapy-induced sensorimotor polyneuropathy (8) | |
| Idiopathic sensorimotor polyneuropathy (4) | |
| Vitamin B12 deficiency sensorimotor polyneuropathy (3) | |
| Paraproteinemic sensorimotor polyneuropathy (2) | |
| Vasculitic sensorimotor multineuropathy (5) | EDX criteria for multifocal CIDP (5) |
| Diabetic sensory polyneuropathy (55) | EDX criteria for sensory CIDP (123) |
| Chemotherapy-induced sensory polyneuropathy (33) | |
| Idiopathic sensory polyneuropathy (22) | |
| Vasculitic sensory polyneuropathy (8) | |
| Paraproteinemic sensory polyneuropathy (3) | |
| Vitamin B12 deficiency polyneuropathy (2) | |
| Idiopathic motor polyneuropathy (4) | EDX criteria for motor CIDP (4) |

CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; EAN/PNS, european academy of neurology/peripheral nerve society; EDX, electrodiagnostic; EFNS/PNS, european federation of neurological societies/peripheral nerve society; N, number.

two patients with a 30–100% proximal-to-distal CMAP duration increase in one tibial nerve, one patient with a >50% proximal-to-distal CMAP amplitude reduction in one tibial nerve and one patient with distal CMAP duration prolongation above the EFNS/PNS but below the EAN/PNS cut-off for demyelination. One patient with a 40% proximal-to-distal CMAP amplitude reduction in one motor nerve (ulnar nerve), without other demyelinating parameters, met the EAN/PNS criteria but not the EFNS/PNS criteria for possible CIDP. The mean number of sensory nerves examined in the 25 patients who missed the diagnosis using the EAN/PNS criteria because of the lack of sensory nerve abnormalities was 3.7 (range 2–6), with 13 patients with at least four nerves examined. All the 25 patients had a clinical phenotype of typical CIDP with persistent sensory symptoms and signs. The difference between the EFNS/PNS and the EAN/PNS criteria remained statistically significant when comparing the sensitivity of the two sets of criteria for the diagnosis of probable/definite CIDP (or CIDP using EAN/PNS criteria): 263/307 (86%) fulfilled the EFNS/PNS criteria in comparison with 227/307 (74%) (McNemar test; $p < 0.001$).

Table 4 shows the specificity of the two sets of electrodiagnostic criteria. Specificity of the EFNS/PNS criteria was 68% for possible CIDP and 84% for probable/definite CIDP, while specificity of the EAN/PNS criteria was 88% for possible CIDP and 98% for CIDP. Using extended nerve conduction study protocol, specificity of the two sets of criteria decreased only slightly, possibly as a result of the high number of tested nerves in our control patients group (table 1). Compared with the EFNS/PNS criteria, the EAN/PNS criteria were more specific for the diagnosis of possible CIDP (McNemar test; $p < 0.001$) and probable/definite CIDP (McNemar test; $p < 0.001$). The higher specificity of the EAN/PNS criteria (possible CIDP) resulted in 34 additional control patients correctly identified as not having CIDP, including 13 with a >50% proximal-to-distal CMAP amplitude reduction in one tibial nerve, 9 with a 30–100% proximal-to-distal CMAP duration increase in one tibial nerve, seven control patients with distal CMAP duration prolongation above the EFNS/PNS but below the EAN/PNS cut-off for demyelination, and five fulfilling the motor electrodiagnostic criteria but with normal sensory conduction studies. One patient with a 40% proximal-to-distal CMAP amplitude reduction in one motor nerve (ulnar nerve), without other demyelinating parameters, was correctly identified as true negative by the EFNS/PNS criteria but not by the EAN/PNS criteria. Using extended nerve conduction study protocol, difference between the two sets of criteria remained statistically significant (possible CIDP, McNemar test, $p = 0.001$; probable/definite CIDP, McNemar test, $p = 0.004$).

The diagnostic accuracy of the EFNS/PNS criteria (84% possible CIDP; 85% probable/definite CIDP) was similar to that of the EAN/PNS (85% possible CIDP; 82% CIDP).

Table 5 shows sensitivity of the EFNS/PNS and EAN/PNS electrodiagnostic criteria in patients with typical CIDP and its variants. Compared with the EAN/PNS criteria, the EFNS/PNS criteria were more sensitive for the diagnosis of typical CIDP both in the category of possible (92% vs 81%, $p < 0.001$), and probable/definite diagnosis (86% vs 76%, $p < 0.001$), and when using supportive criteria (91% vs 78%, $p < 0.001$). There was no difference in the sensitivity of the EFNS/PNS and EAN/PNS criteria for the diagnosis of the CIDP variants.

Table 6 shows specificity of the EFNS/PNS and EAN/PNS electrodiagnostic criteria in patients with typical CIDP and its variants. Compared with the EFNS/PNS criteria, the EAN/PNS criteria were more specific for the diagnosis of possible distal CIDP (88% vs 70%,

Table 3 Sensitivity of the EFNS/PNS and EAN/PNS criteria

| | EFNS/PNS electrodiagnostic criteria(Possible CIDP) | EFNS/PNS electrodiagnostic criteria(Probable/DefiniteCIDP) | EAN/PNS electrodiagnostic criteria(Possible CIDP) | EAN/PNS electrodiagnostic criteria(CIDP) |
|------------------------------|---|--|--|---|
| Total, n (%) | 303/330 (92%) | 282/330 (85%) | 254/307 (83%) | 227/307 (74%) |
| Extended NCS protocol, n (%) | 194/202 (96%) | 182/202 (90%) | 175/198 (88%) | 161/198 (81%) |
| | EFNS/PNS criteria for Possible CIDP using supportive criteria | EFNS/PNS criteria for Probable/Definite CIDP using supportive criteria | EAN/PNS criteria for Possible CIDP using supportive criteria | EAN/PNS criteria for CIDP using supportive criteria |
| Total, n (%) | 303/330 (92%) | 299/330 (91%) | 260/307 (85%) | 237/307 (77%) |
| Extended NCS protocol, n (%) | 194/202 (96%) | 192/202 (95%) | 177/198 (89%) | 167/198 (84%) |

$p=0.046$), and for the diagnosis of sensory CIDP both for the category of possible sensory or possible sensory-predominant (88% vs 63%, $p=0.004$) and sensory predominant CIDP diagnosis (98% vs 81%, $p=0.005$).

DISCUSSION

Diagnosis of CIDP is made primarily on the basis of clinical features combined with electrodiagnostic testing, and supported by ancillary exams, and the exclusion of other diseases that may mimic CIDP. Due to the lack of a pathognomonic diagnostic biomarker of the disease, diagnostic criteria for CIDP have been based mainly on the electrodiagnostic demonstration of peripheral nerve demyelination. In the last years, the EFNS/PNS criteria have largely replaced the other criteria for CIDP due to their increased sensitivity and still high specificity and have been broadly adopted both for research and clinical purposes.¹⁶ Still, misdiagnosis of CIDP has been reported to occur in almost half of the cases.¹⁸ Several pitfalls related to overdiagnosed and underdiagnosed have been identified^{18–20 29 30} and the detrimental consequences of CIDP misdiagnosis have been reported by various studies and include undertreatment and overtreatment, increased disease burden and costs.^{18 31} The EAN/PNS criteria were made with the aim of improving specificity of the EFNS/PNS criteria.²¹ They also attempted to improve sensitivity by allowing access to diagnosis to patients with typical CIDP who do not fulfil minimal electrodiagnostic criteria but show an objective response to treatment with at least one other positive supportive criterion for diagnosis.²¹

In this study, we applied both sets of criteria to a large cohort of patients with CIDP and controls. Our control group consisted of patients with axonal peripheral neuropathy, most with diabetic polyneuropathy that, as reported in the literature, may have similar electrophysiological abnormalities compared with CIDP.^{32 33} All our control patients had a clinical presentation that was consistent with the EAN/PNS clinical diagnostic criteria for the CIDP variants. This is not surprising since patients with axonal peripheral neuropathy generally do not have proximal weakness while they frequently present a length-dependent sensory or sensorimotor peripheral neuropathy consistent with

the clinical criteria of sensory CIDP or distal CIDP. Five of our control patients had a sensorimotor multineuropathy that was consistent with the clinical criteria of multifocal CIDP, while four patients had a pure motor polyneuropathy that was consistent with the clinical criteria of motor CIDP. As shown by other studies, misdiagnosis of CIDP occurs more commonly in patients classified as CIDP variants.^{18 34} This underlies the importance of the use of the instrumental and laboratory tests in the differential diagnosis of CIDP, as it has been further stressed in the EAN/PNS criteria, which recommend a specific list of investigations to be performed in each CIDP clinical form.²¹

The present study demonstrates the higher specificity of the EAN/PNS criteria in comparison with those of the EFNS/PNS (98% vs 84% for probable/definite CIDP (or ‘CIDP’ with EAN/PNS) and 88% vs 68% for possible CIDP; $p<0.001$), with specificity values for the EFNS/PNS criteria similar to what previously observed.^{4–6} On the other hand, sensitivity of the EAN/PNS criteria was lower than that of the EFNS/PNS criteria (74% vs 85% for probable/definite CIDP (or ‘CIDP’ with EAN/PNS) and 83% vs 92% for possible CIDP), with the EFNS/PNS criteria allowing a significant additional number of diagnosed patients ($p<0.001$). The inclusion by the EAN/PNS criteria of patients fulfilling clinical criteria for typical CIDP but not minimal electrophysiological criteria, in presence of an objective response to therapy plus another supportive criterion,²¹ only provided a small diagnostic gain. The additional gain in sensitivity of the EAN/PNS criteria using supportive criteria was 2% (from 83% to 85%) for possible CIDP and 3% (from 74% to 77%) for CIDP, with the EAN/PNS criteria still showing a reduced sensitivity compared with the EFNS/PNS (77% vs 91% for probable/definite CIDP (or ‘CIDP’ with EAN/PNS) and 85% vs 92% for possible CIDP). This may reflect the fact that the EAN/PNS criteria limited the recommendation only to patients with typical CIDP,²¹ with only six additional diagnoses (11%) from a total of 53 patients fulfilling clinical criteria for typical CIDP or its variants but not minimal motor electrodiagnostic criteria. The main reason for the infrequent diagnostic improvement in typical CIDP patients in our population was the mandatory presence of sensory nerve conduction abnormalities required by the EAN/

Table 4 Specificity of the EFNS/PNS and EAN/PNS electrodiagnostic criteria

| | EFNS/PNS electrodiagnostic criteria (possible CIDP) | EFNS/PNS electrodiagnostic criteria (definite or probable CIDP) | EAN/PNS electrodiagnostic criteria (possible CIDP) | EAN/PNS electrodiagnostic criteria (CIDP) |
|------------------------------|---|---|--|---|
| Total, n (%) | 113/166 (68%) | 140/166 (84%) | 146/166 (88%) | 162/166 (98%) |
| Extended NCS protocol, n (%) | 45/69 (65%) | 58/69 (84%) | 59/69 (86%) | 67/69 (97%) |

CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; EAN/PNS, european academy of neurology/peripheral nerve society; EFNS/PNS, european federation of neurological societies/peripheral nerve society; N, number; NCS, nerve conduction study.

Table 5 Sensitivity of the EFNS/PNS and EAN/PNS criteria in typical CIDP and its variants

| | EFNS/PNS electrodiagnostic criteria for possible CIDP | EFNS/PNS electrodiagnostic criteria for probable/definite CIDP | EFNS/PNS criteria for probable/definite CIDP using supportive criteria | EAN/PNS electrodiagnostic criteria for possible CIDP | EAN/PNS electrodiagnostic criteria for CIDP | EAN/PNS criteria for CIDP using supportive criteria | P value |
|---|---|--|--|--|---|---|---------------|
| Typical CIDP, n (%) | 232/251 (92%) | 217/251 (86%) | 229/251 (91%) | 158/195 (81%) | 149/195 (76%) | 153/195 (78%) | <0.001*, †, ‡ |
| DADS/distal CIDP, n (%) | 24/29 (83%) | 22/29 (76%) | 24/29 (83%) | 35/41 (85%) | 29/41 (71%) | 31/41 (76%) | |
| LSS/multifocal CIDP and focal CIDP, n (%) | 17/19 (89%) | 17/19 (89%) | 17/19 (89%) | 17/23 (74%) | 14/23 (61%) | 16/23 (70%) | |
| Sensory CIDP, n (%) | 13/14 (93%) | 10/14 (71%) | 12/14 (86%) | 27/31 (87%)§ | 21/31 (68%)¶ | 22/31 (71%) | |
| Motor CIDP, n (%) | 17/17 (100%) | 16/17 (94%) | 17/17 (100%) | 17/17 (100%) | 14/17 (83%) | 15/17 (88%) | |

*EFNS/PNS electrodiagnostic criteria for possible CIDP versus EAN/PNS electrodiagnostic criteria for possible CIDP.
 †EFNS/PNS electrodiagnostic criteria for probable/definite CIDP versus EAN/PNS electrodiagnostic criteria for CIDP.
 ‡EFNS/PNS criteria for probable/definite CIDP using supportive criteria versus EAN/PNS criteria for CIDP using supportive criteria.
 §Patients with electrodiagnostic testing of sensory CIDP and sensory predominant CIDP/clinical diagnosis of sensory CIDP.
 ¶Patients with electrodiagnostic testing of sensory predominant CIDP/clinical diagnosis of sensory CIDP.
 CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; DADS, distal acquired demyelinating symmetric neuropathy; EAN/PNS, european academy of neurology/peripheral nerve society; EFNS/PNS, european federation of neurological societies/peripheral nerve society; LSS, lewis-sumner syndrome; N, number.

PNS criteria, with only 9 (24%) of the 37 patients with typical CIDP having this criterion. Another reason, limited to our study population, was the small number of patients who underwent nerve biopsy or nerve imaging.

Our study shows that among the supportive criteria for the diagnosis of CIDP, response to treatment and CSF analysis were particularly helpful in improving the diagnosis with both sets of criteria, in line with what we previously reported.³⁵ Only a few of our patients, however, had undergone nerve imaging or nerve biopsy.

The comparative analysis of the sensitivity and specificity of the two sets of criteria for the diagnosis of typical CIDP and its variants showed that the EFNS/PNS criteria are more sensitive for the diagnosis of typical CIDP, while the EAN/PNS criteria are more specific for the diagnosis of possible distal CIDP and sensory CIDP, both for the category of possible sensory or sensory-predominant and sensory-predominant CIDP diagnosis. The increased specificity of the EAN/PNS criteria for the diagnosis of distal CIDP and sensory CIDP represents an important step forward given that most of the overdiagnosis of CIDP occurs

in patients with an ‘atypical’ clinical presentation.¹⁸ There was no difference between the two sets of criteria in the diagnosis of the other variants, although this may be explained considering the small number of patients in these diagnostic groups.

Overall, our study shows that the diagnostic accuracy of the EFNS/PNS criteria (84% possible CIDP; 85% probable/definite CIDP) is similar to that of the EAN/PNS (85% possible CIDP; 82% CIDP), with the EAN/PNS criteria showing an improvement in specificity (20% for possible CIDP and 14% for probable/definite CIDP (or ‘CIDP’ with the EAN/PNS criteria)) with a reduction in sensitivity (about 11% for possible CIDP and 9% for probable/definite CIDP (or ‘CIDP’ with the EAN/PNS criteria)) compared with the EFNS/PNS criteria. We found that the extended nerve conduction study protocol improved the diagnostic sensitivity of both sets of criteria, with the EAN/PNS criteria being about 10% less sensitive than the EFNS/PNS criteria. Similarly to previous studies,⁴⁻⁶ this was achieved with a decline in specificity that in our study was only minimal, likely due to the high number of motor and sensory nerves examined in our controls group (table 1). Using the EAN/PNS criteria,

Table 6 Specificity of the EFNS/PNS and EAN/PNS criteria in typical CIDP and its variants

| | EFNS/PNS electrodiagnostic criteria for possible CIDP | EFNS/PNS electrodiagnostic criteria for probable/definite CIDP | EAN/PNS electrodiagnostic criteria for possible CIDP | EAN/PNS electrodiagnostic criteria for CIDP | P value |
|---|---|--|--|---|------------------|
| DADS/distal CIDP, n (%) | 85/122 (70%) | 103/122 (84%) | 30/34 (88%) | 33/34 (97%) | 0.046* |
| LSS/multifocal CIDP and focal CIDP, n (%) | 8/13 (62%) | 11/13 (89%) | 4/5 (75%) | 5/5 (100%) | |
| Sensory CIDP, n (%) | 17/27 (63%) | 22/27 (81%) | 108/123 (88%)† | 120/123 (98%)‡ | 0.004* 0.005§ |
| Motor CIDP, n (%) | 3/4 (75%) | 4/4 (100%) | 4/4 (100%) | 4/4 (100%) | |

*EFNS/PNS electrodiagnostic criteria for possible CIDP versus EAN/PNS electrodiagnostic criteria for possible CIDP.
 †Patients with electrodiagnostic testing of sensory CIDP and sensory predominant CIDP/clinical diagnosis of sensory CIDP.
 ‡Patients with electrodiagnostic testing of sensory predominant CIDP/clinical diagnosis of sensory CIDP.
 §EFNS/PNS electrodiagnostic criteria for probable/definite CIDP versus EAN/PNS electrodiagnostic criteria for CIDP.
 CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; EAN/PNS, european academy of neurology/peripheral nerve society; EFNS/PNS, european federation of neurological societies/peripheral nerve society.

the extended nerve conduction study protocol offered the best balance between sensitivity (88% for possible CIDP and 81% for CIDP) and specificity (86% for possible CIDP and 97% for CIDP); therefore, with these criteria, it is recommendable to perform more extended studies to obtain an acceptable sensitivity while maintaining a very high specificity.

In a disease causing severe disability and for which therapies may be expensive, as CIDP, both underdiagnosis and overdiagnosis are inconvenient. The former cause undertreatment and increased disease burden, the latter increased costs and unnecessary exposure to treatment side effects. When we looked at the effect, in terms of diagnostic gain, of the individual changes that have been made in the EAN/PNS criteria from the EFNS/PNS criteria, some have proved to be disadvantageous while others to be effective. The inclusion of sensory nerve conduction studies among the mandatory criteria for diagnosis proved to be disadvantageous in our study population, having led to 25 missed diagnoses with only five additional control patients identified despite the fact that the number of sensory nerves examined in our patients was relatively high (mean 3.7; range 2–6). The exclusion of tibial nerve from the conduction block criterion and the increase to at least 100% of the cut-off for abnormal temporal dispersion in the tibial nerve were instead effective, resulting the former in 13 additional control patients identified with only one missed diagnosis and the latter in nine additional control patients identified with only two missed diagnoses. The same applies to the introduction of separate criteria for four different low frequency filters to measure distal CMAP duration prolongation, which led to seven additional control patients identified with only one missed diagnosis.

Limitations of our study include its retrospective nature and the inclusion of CIDP patients recruited from tertiary referral centres with the risk of selection bias of more serious cases. In the Italian CIDP database patients were selectively enrolled, thus the diagnostic accuracy figures found in our study might be overestimated. No diagnostic biomarker for CIDP exists at present; therefore, similarly to other studies,^{4,6} for the diagnosis of CIDP, we relied on the expert opinion of the treating physician. We attempted to minimise diagnostic errors by reviewing the diagnosis in each patient but the possibility that some of the included patients were misdiagnosed cannot be excluded. Most of the patients included in the database were enrolled before the publication of the EAN/PNS criteria; therefore, response to treatment was measured using only an impairment or a disability measure. Our study did not include the electrodiagnostic criteria for typical CIDP in the analysis of the specificity of the EAN/PNS criteria. This was due to the type of control patients included in our study. Only a few peripheral neuropathies have a clinical presentation similar to typical CIDP and all are demyelinating, so that their inclusion in the study would have significantly reduced the observed diagnostic accuracy figures.

In summary, our study showed that the EAN/PNS criteria are more specific but less sensitive than the EFNS/PNS criteria. More extended nerve-conduction studies improved the diagnostic sensitivity of the EAN/PNS criteria maintaining a very high specificity.

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Correction notice This article has been corrected since it was first published. It has been made open access under a CC BY licence. 19th May 2023.

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Contributors PED and ADL contributed to the conception of the research project, performed the statistical analysis, wrote the first draft of the report and reviewed the report. FM, DC, RF, CB, AM, MF, GC, LB, AS, GAM, GA, SM, ML, GL, ES, EP, CS, MC, LG, SCP, EB contributed to the conception, organisation and execution of the research project, reviewed and commented on the statistical analysis and the report. ENO conceived, organised and designed the study, reviewed and commented on the statistical analysis, reviewed the report. PED and ENO accept full responsibility for the work and the conduct of the study, had access to the data, and controlled the decision to publish.

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Competing interests Pietro Emiliano Doneddu has received travel grants to attend scientific meetings from CSL Behring and Kedrion. Fiore Manganelli reports personal fees for scientific events from CSL Behring and has received travel grants to attend scientific meetings from CSL Behring and Kedrion. Dario Cocito has received honoraria for lecturing from Shire, CSL Behring, and Kedrion and travel grants to attend scientific meeting from Shire, Kedrion, and CSL Behring. Raffaella Fazio has served on scientific advisory boards for CSL Behring and has received travel grants from Kedrion and CSL Behring to attend scientific meeting. Chiara Briani has served on scientific advisory boards for Pfizer, Alnylam, and Akcea, and has received travel grants from Kedrion and CSL Behring to attend scientific meeting. Anna Mazzeo has received travel grants from Kedrion and CSL Behring to attend scientific meeting. Massimiliano Filosto has served on scientific advisory boards for CSL Behring, Sanofi and Amicus and has received travel grants from Sanofi, Biogen, Kedrion and CSL Behring to attend scientific meeting. Giuseppe Cosentino has received travel grants to attend scientific meetings from CSL Behring and Kedrion. Marco Luigetti has received travel grants to attend scientific meetings from Kedrion. Girolama Alessandra Marfia has received travel grants to attend scientific meetings from CSL Behring and Kedrion. Giuseppe Liberatore has received travel grants to attend scientific meetings from CSL Behring and Kedrion. Erdita Peci has received travel grants to attend scientific meetings from CSL Behring. Eduardo Nobile-Orazio reports personal fees for Advisory or Scientific Board from ArgenX—Belgium, Takeda—Italy and USA, CSL-Behring - Italy and USA, Janssen—USA, Kedrion—Italy, LFB—France, Roche—Switzerland, Sanofi - USA. The other authors declare no conflict of interest.

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Ethics approval The study was approved by the Ethical Committee of each participating Center. Participants gave informed consent to participate in the study before taking part.

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Data availability statement Data are available upon reasonable request. Anonymised data used for this study are available upon reasonable request from the corresponding author.

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