# A Retrospective Study of Ultrasound Accuracy for the Diagnosis of Chronic Inflammatory Demyelinating Polyneuropathy

Nicholas H. Crump* and Michael S. Cartwright†

*Departments of Neurology and Medicine, Austin Health and University of Melbourne, Victoria, Australia; and †Department of Neurology, Wake Forest School of Medicine, Winston-Salem, North Carolina, U.S.A.



**Introduction:** Ultrasound is emerging as a useful tool for the evaluation of immune-mediated neuropathies because it can provide high-resolution anatomic information to complement electrodiagnostic data. Nerve enlargements are commonly found in chronic inflammatory demyelinating polyneuropathy (CIDP), and their presence likely useful in diagnosis, particularly if multifocal.

**Methods:** In this study, the authors undertook a retrospective chart review to identify ultrasound findings in patients with CIDP previously studied in a single busy neurodiagnostic laboratory.

**Results:** Of the 50 cases identified from 2000 to 2017, individuals with a confirmed diagnosis of CIDP (21 cases) were more likely to have multiple sites of enlargement, as well as more pronounced nerve enlargement, than patients who were subsequently found to have an alternate cause of neuropathy (22 cases). The presence of any moderately enlarged nerve segment predicted definite CIDP with sensitivity of 81% and specificity 77%.

**Conclusion:** This study demonstrates that ultrasound can be of diagnostic utility in patients with suspected CIDP, even when conducted in a nonstandardized real-world setting.

**Key Words:** Chronic inflammatory demyelinating polyneuropathy, Ultrasound, Nerve ultrasonography, Diagnosis, Electrodiagnosis.

(J Clin Neurophysiol 2022;39: 312–316)

Recent studies confirm that neuromuscular ultrasound (NMUS) is beneficial in the diagnosis of immune-mediated neuropathies such as chronic inflammatory demyelinating polyneuropathy (CIDP), adding information regarding nerve structure and possibly the pathophysiological processes at play to the functional information obtained from nerve conduction studies and EMG.[1,2] Focal nerve enlargements and variable nerve size are very suggestive, if not characteristic findings in CIDP, and form the basis of proposed diagnostic scores and testing protocols.[3–17] Despite the lack of consensus regarding the optimal diagnostic approach to CIDP using ultrasound, all studies have shown nerve enlargement as measured by increased nerve cross-sectional area (CSA)—in particular, if multifocal—can distinguish CIDP from healthy controls and other differential diagnoses.

This retrospective study explored the accuracy of ultrasound for the diagnosis of CIDP when performed as a component of routine neurodiagnostic testing. This group of patients was not enrolled in a systematic study, with no fixed clinical or ultrasound protocol applied. Instead, they had variable amounts of ultrasound performed during routine clinical care to help establish their diagnoses. Therefore, this retrospective chart review assesses the diagnostic accuracy of ultrasound in a real-world setting.

The authors have no funding or conflicts of interest to disclose.
Some of these data were presented as a poster at the American Academy of Neurology Annual Meeting, Los Angeles CA, April 23, 2018.
Supplemental digital content is available for this article. Direct URL citations appear in the printed text and are provided in the HTML and PDF versions of this article on the journal's Web site (www.clinicalneurophys.com).
Address correspondence and reprint requests to Nicholas H. Crump, MBBS, FRACP, Austin Health and University of Melbourne, Studley Road, Heidelberg, Victoria 3084, Australia, Australia; e-mail: ncrump@student.unimelb.edu.au.

ISSN: 0736-0258/20/3904-0312
DOI 10.1097/WNP.0000000000000782

## METHODS

The ethics committee of Wake Forest Baptist Medical Center (WFBMC) approved our study protocol, and we proceeded to conduct a retrospective chart review to identify all patients with CIDP studied with ultrasound at WFBMC from January 2000 to August 2017. Patients were identified as potential subjects if they had a coded diagnosis of CIDP in the electronic medical record (ICD-10 G61.8 or ICD-9 357.81) and any recorded attendance at the WFBMC diagnostic neurology laboratory. A brief chart review was then undertaken to confirm the performance of NMUS; if so, we proceeded to a more detailed chart review to extract a standardized dataset of clinical, electrodiagnostic, and ultrasonographic findings. Clinical data included duration of disease, history of immunomodulatory or immunosuppressant treatment, and the current clinical state as determined by the Medical Research Council (MRC) sum score as described by Kleyweg et al.[18] from closest recorded examination findings and estimated Inflammatory Neuropathy Cause and Treatment Overall Disability Sum Score (ODSS)[19] from documented history. Ultrasound data (CSA, echogenicity, abnormal vascularity) was recorded and compared with laboratory-derived normal values to determine whether segments were enlarged.[20] We defined any enlargement if the CSA was higher than the upper limit of normal, and moderate enlargement if $>1.5$ times the upper limit of normal.

## RESULTS

One hundred forty-eight patients were initially identified as having a diagnostic code of CIDP and previously attending the WFBMC diagnostic neurology laboratory. Fifty of these 148

patients had ultrasound performed and formed the subjects for this analysis. The indication as to why ultrasound was performed was unable to be ascertained from the records.

Despite being coded as CIDP, analyses of the clinical and electrophysiological data made it clear that not all subjects had a CIDP clinical phenotype (either typical or atypical) or met the definite or probable electrodiagnostic European Federation of Neurological Societies/Peripheral Nerve Society (PNS) criteria for CIDP.[21,22] We subsequently allocated the 50 cases into three groups:

- 21 with a confirmed diagnosis of CIDP, meeting definite or probable European Federation of Neurological Societies/PNS criteria
- 22 with a clear alternative diagnosis
  - Eight with Guillain–Barré syndrome/acute inflammatory demyelinating polyneuropathy with no relapse or progression
  - Five with paraprotein-related neuropathy without CIDP features on nerve conduction studies
  - The remaining nine with multifactorial axonal neuropathies, such as from diabetes or renal impairment
- Seven with an "uncertain" diagnosis of CIDP. All these cases had atypical phenotypes, along with either limited electrophysiological testing or insufficient evidence of chronicity (unclear documentation, lost to follow-up, or pending further review at the time of data extraction).

Subject demographics are outlined in Table 1. There was no statistically significant difference between the groups, except for comorbid diabetes (lower in the confirmed CIDP group, higher in the uncertain group). Interestingly, all groups had similar rates of treatment with IVIg or corticosteroids both at the time ultrasound was performed and at any stage. Severity was similar at the time of assessment across the three groups (as determined by MRC sum score or Inflammatory Neuropathy Cause and Treatment ODSS).

There was wide variability in the number of nerves (range, one to seven), and segments (range, 2–17) tested. Thirty-seven subjects (74%) had assessment of one or both median nerves, with at least two segments assessed in all but one case, and 34 subjects (68%) had evaluation of one or both ulnar nerves (again with at least two segments assessed in all but one case). Other nerves tested included radial (six subjects), fibular (10), tibial (nine), sural (two), and brachial plexus.[8]

Table 2 summarizes the key ultrasound findings from subjects with confirmed CIDP, uncertain CIDP, and those with a clear alternative diagnosis. Our analysis focused on documentation of nerve size and focal enlargements, as documentation of echogenicity and abnormal vascularity was insufficient for analysis. Abnormalities on ultrasound were common in our confirmed CIDP cohort, with 19 of 21 subjects having at least one abnormal finding. Most subjects had a mix of both normal and enlarged nerve CSAs, with only one having moderate (>150%) enlargement in all sites tested (16 of 16). Another two subjects had nerve enlargement at all sites tested (four and six, respectively), with both having 50% of sites moderately enlarged.

Analysis was planned along the lines of previously published diagnostic scores and protocols,[5,7,8,13,14,16] though the nonstandardized testing and often limited sites assessed with ultrasound restricted use of these approaches. However, differences between patients with and without CIDP could be quantified on several metrics, despite the variable data acquisition.

Table 3 summarizes the diagnostic accuracy of parameters in our group that distinguished confirmed CIDP from patients with an alternate diagnosis. Ultrasound data obtained were mostly from the median and ulnar nerves, but many parameters are shown to have reasonable diagnostic accuracy. These included two or more enlarged segments, any moderately enlarged segment, and any enlarged median nerve segment (excluding the wrist).

Analysis of previously published "quick test" protocols using our data was undertaken. Applying the protocol of Goedee et al.[16] (enlargement of any from median nerve in mid-forearm or upper arm; any trunk of brachial plexus) in our dataset yielded a sensitivity of 81% and specificity of 82%. This is despite only two subjects from the entire cohort having all three of the requisite segments tested. The protocol of Jang et al.[7] (enlargement of any from median nerve in mid-forearm or upper arm; ulnar nerve in upper arm; tibial nerve in calf; and vagus nerve in carotid sheath) had a sensitivity of 81% and specificity of 77%;

**TABLE 1.** Subject Demographics

| | Confirmed CIDP | Not CIDP | "Uncertain" CIDP | *P** |
|---|---|---|---|---|
| Number subjects (N) | 21 | 22 | 7 | |
| Age at test (mean, years) | 49.4 | 53.4 | 51.9 | 0.756 |
| Gender—male (N, %) | 14 (66.7%) | 12 (54.5%) | 5 (71.4%) | 0.614 |
| BMI (mean, kg/m$^2$) | 30.9 | 28.9 | 27.4 | 0.357 |
| Duration of symptoms (mean, months) | 45.8 | 37.8 | 35 | 0.887 |
| Comorbid type 2 diabetes (N, %) | 5 (23.8%) | 9 (40.9%) | 5 (71.4%) | 0.074 |
| Ever on disease-modifying treatment (N, %) | 9 (42.9%) | 11 (50.0%) | 3 (42.9%) | 0.881 |
| On treatment at the time of NMUS (N, %) | 5 (23.8%) | 6 (27.3%) | 2 (28.6%) | 0.954 |
| MRC sum score (mean) | 53.8 | 56.1 | 55.8 | 0.562 |
| INCAT ODSS (mean) | 3.0 | 3.1 | 3.7 | 0.780 |

*Continuous variables tested by one-way analysis of variance; discrete by Chi-square.

CIDP, chronic inflammatory demyelinating polyneuropathy; INCAT, Inflammatory Neuropathy Cause and Treatment; NMUS, neuromuscular ultrasound; ODSS, Overall Disability Sum Score.

**TABLE 2.** Ultrasound Findings in Each Diagnostic Group

| | Confirmed CIDP ($N = 21$) | Not CIDP ($N = 22$) | "Uncertain" CIDP ($N = 7$) |
|---|---|---|---|
| Mean number of nerves tested | 3 | 3 | 3.4 |
| Mean number of segments tested | 8.3 | 5.1 | 7.7 |
| Mean number of abnormal segments | 5.6 (66.9%) | 1.6 (31.0%) | 5.4 (70.4%) |
| Mean number of segments >1.5× ULN | 3.0 (36.0%) | 0.4 (7.1%) | 2.6 (33.3%) |
| Subjects with 2+ abnormal segments | 18 (85.7%) | 7 (31.8%) | 6 (85.7%) |
| Subjects with any segment >1.5× ULN | 17 (81.0%) | 5 (22.7%) | 4 (57.1%) |
| Any abnormal median nerve segment | 17 (81.0%) | 6 (27.3%) | 4 (57.1%) |
| % in subjects with median nerve tested | 89.5% | 50% | 66.7% |
| Any abnormal median nerve segment (except wrist) | 16 (76.2%) | 4 (18.2%) | 4 (57.1%) |
| % in subjects with median nerve tested | 84.2% | 33.3% | 80% |
| Any enlargement median or ulnar at upper arm | 14 (66.7%) | 3 (13.6%) | 5 (71.4%) |
| % in subjects with median or ulnar tested | 82.4% | 42.9% | 100% |
| Zaidman type 0 (no enlargement) | 1 (4.8%) | 10 (45.4%) | 0 (0%) |
| Zaidman type 1 (mild enlargement) | 9 (42.9%) | 4 (18.2%) | 2 (28.6%) |
| Zaidman type 2 (regional enlargement) | 2 (9.5%) | 0 (0%) | 1 (14.3%) |
| Zaidman type 3 (diffuse enlargement) | 6 (28.6%) | 0 (0%) | 3 (42.9%) |

CIDP, chronic inflammatory demyelinating polyneuropathy; ULN, upper limit of normal.



again despite no subject with vagus nerve assessed and only nine with tibial nerve parameters. There is obviously a potential for an increased false-negative rate for applying both these protocols (which are abnormal if one segment is enlarged, so the "test" can still be positive even if not all segments are scanned) in our dataset given the missing data points. However, this would result in more conservative observed sensitivities.

Our a priori plan to assess more involved protocols (Ultrasound Pattern Sum Score,[13] Bochum Ultrasound Score/Nerve Ultrasound Protocol,[14] variability scores[8]) was not possible because of insufficient data from this retrospective nonstandardized cohort. For example, only five subjects out of the entire cohort had more than two of the four components of the Bochum Ultrasound Score assessed.

Assessment for variability in nerve size (which alongside focal nerve enlargement is the most described abnormality in CIDP) was tricky in this cohort, with nonstandardized assessments making analyses by previously published methods (such as intranerve and internerve CSA variability[8] and heterogeneity score[13]) not possible. However, analyses as per the schema of Zaidman[5] (based on median and ulnar assessment only) was possible. This proposes four patterns based on NMUS findings: no enlargement (type 0); mild enlargement (no sites with moderate enlargement, type 1); regional enlargement (at least one site normal and one moderate, type 2); and diffuse enlargement (all sites enlarged, at least one moderate, type 3). In our cohort, confirmed CIDP patients are spread across the range of enlargement patterns (as has been shown in most other published cohorts),[6–12,15–17] whereas subjects without CIDP have either no enlargement (type 0) or only mild (type 1) changes.

There was no enlargement pattern or parameter clearly different between typical and atypical CIDP phenotypes, although the subject numbers are small. However, all NMUS parameters identified from the overall cohort were also able to distinguish between CIDP and those with an alternate diagnosis for both typical and atypical phenotype with reasonably similar

**TABLE 3.** Utility of Specific Ultrasonographic Parameters in Distinguishing Between Subjects With Confirmed CIDP ($N = 21$) and Those With an Alternate Diagnosis ("Not CIDP", $N = 22$)

| | Sensitivity (%) | Specificity (%) | Accuracy (%) | PPV (%) | NPV (%) | *P* (Fisher Exact Test) |
|---|---|---|---|---|---|---|
| Subjects with 2+ enlarged segments | 85.7 | 68.2 | 76.7 | 72.0 | 83.3 | 0.0005 |
| Subjects with any segment >1.5× ULN | 81.0 | 77.3 | 79.1 | 77.3 | 81.0 | 0.0001 |
| Any enlarged median nerve segment | 81.0 | 72.7 | 76.7 | 73.9 | 80.0 | 0.0005 |
| Any enlarged median nerve segment (excluding at the wrist) | 76.2 | 81.8 | 79.1 | 80.0 | 78.3 | 0.0002 |
| Enlargement of either median or ulnar nerve at upper arm site | 66.7 | 86.4 | 76.7 | 82.4 | 73.1 | 0.0005 |
| Enlargement pattern 2–3 (regional/diffuse) vs. 0–1 (none/mild) as per Zaidman 2013 | 44.4 | 100.0 | 68.8 | 100.0 | 58.3 | 0.0042 |
| Goedee diagnostic protocol | 81.0 | 81.8 | 81.4 | 81.0 | 81.8 | 0.0001 |
| Jang diagnostic protocol | 81.0 | 77.3 | 79.1 | 77.3 | 81.0 | 0.0002 |

CIDP, chronic inflammatory demyelinating polyneuropathy; NPV, negative predictive value; positive predictive value; ULN, upper limit of normal.

levels of accuracy (Table 4). The exception was that the Goedee protocol was less sensitive for atypical CIDP, possibly reflecting that it involves the lowest number and most specific set of nerve enlargement sites.

Given the high prevalence of diabetes in these subjects, we further analyzed the most diagnostic ultrasound findings according to diabetes status in each of the three groups (see **Table 1**, **Supplemental Digital Content 1**, http://links.lww.com/JCNP/A129). Interestingly, the rate of ultrasound abnormalities was higher with comorbid diabetes across all diagnostic groupings, but particularly for confirmed CIDP and the uncertain group. Furthermore, subjects with an alternate diagnosis and who were not diabetic had very low rates of ultrasound abnormalities. Our dataset did not include information on diabetic severity or treatment.

There is insufficient data to accurately assess correlations between ultrasound parameters and disease severity, duration, and outcomes, as well as with findings from nerve conduction studies. Of the two confirmed CIDP subjects with no ultrasound abnormalities, one was in remission off treatment (ODSS zero, MRC sum score 60, zero of three abnormal nerve segments with ultrasound) and the other had mild residual clinical changes on treatment (ODSS two, MRC sum score 54, zero of eight abnormal segments). However, other definite CIDP subjects with either ODSS of zero or MRC sum score of 60 had a variable number and extent of NMUS enlargements detected.

## DISCUSSION

The limitations of this retrospective study are evident: heterogeneity in ultrasound data obtained, lack of analysis of ultrasound parameters aside from nerve enlargements, absence of a fixed protocol, variable completeness of clinical documentation from which severity scores could be extrapolated, wide ranges in treatments and durations, and nonstandardized patient groups (both with and without CIDP) without clear indication for the addition of NMUS to routine electrophysiological testing. Furthermore, the structure of the retrospective case review meant no cases with an inherited neuropathy were included for analysis or comparison. All these are potential sources of bias and contribute to a restricted dataset. However, despite these limitations, we could still demonstrate that ultrasound changes are common in patients who have a diagnosis of CIDP, with apparent differences in the degree and extent of nerve enlargements between subjects with CIDP and those with an alternate diagnosis. In particular, this is despite the majority of the ultrasound data coming from the more easily assessed median and ulnar nerves, without many of the less routinely assessed nerves and segments that make up other published diagnostic protocols. Furthermore, despite the data being obtained over an extensive period, from different sonographers with variable levels of experience and utilizing different devices, the diagnostic benefit could still be demonstrated.

The "uncertain" CIDP group is interesting from a real-life management and diagnostic perspective. However, conclusions from this data need to be drawn with caution given the outcome of these patients was generally unknown, or diagnostic workup was incomplete, and hence their exclusion from analysis of diagnostic accuracy. The small size of this group (seven patients) and nonstandardized data are clearly limitations. The high rate of comorbid diabetes is of potential significance in this group given ultrasound abnormalities are more prevalent in these subjects. Still, the patterns of ultrasound findings in the uncertain CIDP group are very similar to the confirmed CIDP group (Table 2), although this group has a lower rate of moderately enlarged (>1.5× upper limit of normal) nerves and median nerve changes. Although it would be interesting to ascertain the response to treatment in this "uncertain" group, the limitations of the

**TABLE 4.** Utility of Specific Ultrasonographic Parameters in Distinguishing Between Subjects With Typical CIDP Phenotypes ($N = 9$), Atypical CIDP ($N = 19$), and Those With an Alternate Diagnosis ("Not CIDP", $N = 22$)

| | Phenotype | Sensitivity (%) | Specificity (%) | Accuracy (%) | PPV (%) | NPV (%) | *P* (Fisher) |
|---|---|---|---|---|---|---|---|
| Subjects with 2+ enlarged segments | Typical | 88.9 | 68.2 | 74.2 | 53.3 | 93.8 | 0.0059 |
| | Atypical | 84.2 | 68.2 | 75.6 | 69.6 | 83.3 | 0.0013 |
| Subjects with any segment >1.5× ULN | Typical | 77.8 | 77.3 | 77.4 | 58.3 | 89.5 | 0.0001 |
| | Atypical | 73.7 | 77.3 | 75.6 | 73.7 | 77.3 | 0.0001 |
| Any enlarged median nerve segment | Typical | 77.8 | 72.7 | 74.2 | 53.9 | 88.9 | 0.0166 |
| | Atypical | 73.7 | 72.7 | 73.2 | 70.0 | 76.2 | 0.0048 |
| Any enlarged median nerve segment (excluding at the wrist) | Typical | 77.8 | 81.8 | 80.7 | 63.6 | 90.0 | 0.0033 |
| | Atypical | 68.4 | 81.8 | 75.6 | 76.5 | 75.0 | 0.0016 |
| Enlargement of either median or ulnar nerve at upper arm site | Typical | 66.7 | 86.4 | 80.7 | 66.7 | 86.4 | 0.0068 |
| | Atypical | 68.4 | 86.4 | 78.1 | 81.3 | 76.0 | 0.0005 |
| Enlargement pattern 2–3 (regional/diffuse) vs. 0–1 (none/mild) as per Zaidman 2013 | Typical | 42.9 | 100.0 | 81.0 | 100.0 | 77.8 | 0.0263 |
| | Atypical | 52.9 | 100.0 | 74.2 | 100.0 | 63.6 | 0.0013 |
| Goedee diagnostic protocol | Typical | 88.9 | 81.8 | 83.9 | 66.7 | 94.7 | 0.0005 |
| | Atypical | 68.4 | 81.8 | 75.6 | 76.5 | 75.0 | 0.0016 |
| Jang diagnostic protocol | Typical | 77.8 | 77.3 | 77.4 | 58.3 | 89.5 | 0.0118 |
| | Atypical | 88.9 | 77.3 | 82.5 | 76.1 | 89.5 | 0.0001 |

CIDP, chronic inflammatory demyelinating polyneuropathy; NPV, negative predictive value; positive predictive value; ULN, upper limit of normal.

retrospective chart review did not allow this to occur (and in fact, was part of the definition of the grouping). A key question is whether ultrasound abnormalities in patients with an unclear diagnosis on clinical or electrophysiological grounds is predictive of treatment response, as found by other groups interested in this area.[23,24] Disappointingly, three of the four subjects with more ultrasound enlargements (all with $\geq 3$ enlargements, and at least one moderate) had a trial of treatment before being lost to follow-up.

Attempts to correlate ultrasound findings with specific disease parameters were limited in this dataset, aside from a higher rate of abnormalities in diabetic patients. However, even with limited subjects, we were able to demonstrate diagnostic utility for both atypical phenotypes as well as subjects with typical CIDP.



## CONCLUSIONS

In this real-world, nonstandardized study of ultrasound in patients, who at some stage were considered to possibly have CIDP, we found clear differences in ultrasound parameters between those who had a subsequently confirmed diagnosis CIDP compared with those with an alternate cause of neuropathy. These differences are similar for both typical and atypical CIDP phenotypes and are still found in the presence of comorbid diabetes. This study further adds to the increasing body of literature that finding nerve enlargements with ultrasound is characteristic of CIDP, and thus of likely diagnostic utility.

Protocols based on ultrasound assessment of easily accessible upper limb nerves (which can be scanned and measured along their entire length) should be further pursued in studies of patients with CIDP, particularly in the diagnostic setting.

## ACKNOWLEDGMENTS

Data extraction, collation, and statistical analysis were undertaken by N.H.C. All authors otherwise contributed equally to the study design, data interpretation, and manuscript preparation.

## REFERENCES

1. Telleman JA, Grimm A, Goedee S, Visser LH, Zaidman CM. Nerve ultrasound in polyneuropathies. Muscle Nerve 2018;57:716–728.
2. Gallardo E, Noto Y, Simon NG. Ultrasound in the diagnosis of peripheral neuropathy: structure meets function in the neuromuscular clinic. J Neurol Neurosurg Psychiatry 2015;86:1066–1074.
3. Kerasnoudis A, Tsivgoulis G. Nerve ultrasound in peripheral neuropathies: a review. J Neuroimaging 2015;25:528–538.
4. Padua L, Paolasso I, Pazzaglia C, et al. High ultrasound variability in chronic immune-mediated neuropathies. Review of the literature and personal observations. Rev Neurol (Paris) 2013;169:984–990.
5. Zaidman CM, Harms MB, Pestronk A. Ultrasound of inherited vs. acquired demyelinating polyneuropathies. J Neurol 2013;260:3115–3121.
6. Grimm A, Heiling B, Schumacher U, Witte OW, Axer H. Ultrasound differentiation of axonal and demyelinating neuropathies. Muscle Nerve 2014;50:976–983.
7. Jang JH, Cho CS, Yang KS, Seok HY, Kim BJ. Pattern analysis of nerve enlargement using ultrasonography in chronic inflammatory demyelinating polyneuropathy. Clin Neurophysiol 2014;125:1893–1899.
8. Padua L, Granata G, Sabatelli M, et al. Heterogeneity of root and nerve ultrasound pattern in CIDP patients. Clin Neurophysiol 2014;125:160–165.
9. Scheidl E, Bohm J, Simo M, Bereznai B, Bereczki D, Aranyi Z. Different patterns of nerve enlargement in polyneuropathy subtypes as detected by ultrasonography. Ultrasound Med Biol 2014;40:1138–1145.
10. Di Pasquale A, Morino S, Loreti S, Bucci E, Vanacore N, Antonini G. Peripheral nerve ultrasound changes in CIDP and correlations with nerve conduction velocity. Neurology 2015;84:803–809.
11. Grimm A, Decard BF, Axer H, Fuhr P. The ultrasound pattern sum score—UPSS. A new method to differentiate acute and subacute neuropathies using ultrasound of the peripheral nerves. Clin Neurophysiol 2015;126:2216–2225.
12. Athanasopoulou IM, Rasenack M, Grimm C, et al. Ultrasound of the nerves—an appropriate addition to nerve conduction studies to differentiate paraproteinemic neuropathies. J Neurol Sci 2016;362:188–195.
13. Grimm A, Vittore D, Schubert V, et al. Ultrasound pattern sum score, homogeneity score and regional nerve enlargement index for differentiation of demyelinating inflammatory and hereditary neuropathies. Clin Neurophysiol 2016;127:2618–2624.
14. Kerasnoudis A, Pitarokoili K, Haghikia A, Gold R, Yoon MS. Nerve ultrasound protocol in differentiating chronic immune-mediated neuropathies. Muscle Nerve 2016;54:864–871.
15. Merola A, Rosso M, Romagnolo A, Peci E, Cocito D. Peripheral nerve ultrasonography in chronic inflammatory demyelinating polyradiculoneuropathy and multifocal motor neuropathy: correlations with clinical and neurophysiological data. Neurol Res Int 2016;2016:9478593.
16. Goedee HS, van der Pol WL, van Asseldonk JH, et al. Diagnostic value of sonography in treatment-naive chronic inflammatory neuropathies. Neurology 2017;88:143–151.
17. Niu J, Cui L, Liu M. Multiple sites ultrasonography of peripheral nerves in differentiating Charcot-Marie-Tooth type 1A from chronic inflammatory demyelinating polyradiculoneuropathy. Front Neurol 2017;8:181.
18. Kleyweg RP, van der Meche FG, Schmitz PI. Interobserver agreement in the assessment of muscle strength and functional abilities in Guillain-Barre syndrome. Muscle Nerve 1991;14:1103–1109.
19. Merkies IS, Schmitz PI, van der Meche FG, Samijn JP, van Doorn PA. Clinimetric evaluation of a new overall disability scale in immune mediated polyneuropathies. J Neurol Neurosurg Psychiatry 2002;72:596–601.
20. Cartwright MS, Passmore LV, Yoon JS, Brown ME, Caress JB, Walker FO. Cross-sectional area reference values for nerve ultrasonography. Muscle Nerve 2008;37:566–571.
21. 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;17:356–363.
22. Hughes RA, Bouche P, Cornblath DR, 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. Eur J Neurol 2006;13:326–332.
23. Hartig F, Ross M, Dammeier NM, et al. Nerve ultrasound predicts treatment response in chronic inflammatory demyelinating polyradiculoneuropathy-a prospective follow-up. Neurotherapeutics 2018;15:439–451.
24. Herraets IJT, Goedee HS, Telleman JA, et al. Nerve ultrasound improves detection of treatment-responsive chronic inflammatory neuropathies. Neurology 2020;94:e1470–e1479.