Journal of the Neurological Sciences 443 (2022) 120478

Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Review Article

Tips in navigating the diagnostic complexities of chronic inflammatory demyelinating polyradiculoneuropathy

Richard A. Lewis [a,*], Pieter A. van Doorn [b], Claudia Sommer [c]

[a] Cedars-Sinai Medical Center, Los Angeles, CA, USA
[b] Department of Neurology, Erasmus MC, University Medical Center Rotterdam, Rotterdam, Netherlands
[c] University of Würzburg, Würzburg, Germany

ARTICLE INFO

Keywords:
Chronic inflammatory demyelinating polyradiculoneuropathy
Clinical outcome measures
Biomarkers
Diagnosis
Disease activity
Impairment

ABSTRACT

The 2021 guideline of the European Academy of Neurology/Peripheral Nerve Society on chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) includes important revisions to the previous 2010 guideline. This article highlights the new criteria and recommendations for the differential diagnosis of CIDP. In the revised guideline, the CIDP spectrum has been modified to include typical CIDP and four well-characterized CIDP variants, namely distal, multifocal/focal, motor and sensory CIDP, replacing the term 'atypical' CIDP. To improve the diagnosis of CIDP, the revised guideline attempts to improve the specificity of the diagnostic criteria for typical CIDP and the four CIDP variants. Specific clinical and electrodiagnostic (including both motor and sensory conduction) criteria are provided for typical CIDP and each of the CIDP variants. The levels of diagnostic certainty have been changed to CIDP and possible CIDP, with the removal of probable CIDP (due to the lack of difference in the accuracy of the electrodiagnostic criteria for probable CIDP) and definite CIDP (due to the lack of a gold standard for diagnosis). If the clinical and electrodiagnostic criteria allow only for a diagnosis of possible CIDP, cerebrospinal fluid analysis, nerve ultrasound, nerve magnetic resonance imaging, objective treatment response, and nerve biopsy can be used as supportive criteria to upgrade the diagnosis to CIDP. Although the revised guideline needs to be validated and its strengths and weaknesses assessed, using the guideline will likely improve the accuracy of diagnosis of CIDP and variants of CIDP, and aid in distinguishing CIDP from conditions with similar features.

1. Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a term for a syndrome or group of disorders although it does not include all immune demyelinating neuropathies. In the late 1970s, the term CIDP was coined to distinguish it from, but also relate it to, acute inflammatory demyelinating polyneuropathy, the most common form of Guillain-Barré syndrome (GBS).

A joint taskforce of the European Federation of Neurological Societies (EFNS) and the Peripheral Nerve Society (PNS) published their initial guideline on the management of CIDP in 2005 [1,2], followed by the first revision in 2010 [3,4]. The 2010 EFNS/PNS guideline [3,4] has been the gold standard for diagnosis and treatment of CIDP. Criteria published in the 2010 guideline have been used for patient enrolment in almost all clinical trials of CIDP [5], and have a relatively high diagnostic accuracy (sensitivity 73–95%, specificity 91–96% [6–8]), although the range of specificity is broader when a diagnosis of possible CIDP is included.

As the understanding of CIDP has evolved since publication of the 2010 EFNS/PNS guidelines, a second revision was developed and published in 2021 [9,10]. Using available clinical evidence, the guideline taskforce attempted to make practical sense of the diagnosis and management of typical CIDP and its variants. Major changes to the diagnostic terminology include replacing the term 'atypical' CIDP with four well-characterized CIDP variants, and specifying clinical, electrodiagnostic and supportive diagnostic criteria for typical CIDP and each CIDP variant [9,10]. This article focuses on the diagnosis of CIDP according to these revised criteria [9,10].

2. Differentiation of CIDP from other conditions

The diagnosis of CIDP in patients who present with weakness and

* Corresponding author at: Cedars-Sinai Medical Center, 127 S. San Vicente Blvd - #A6600, Los Angeles, CA 90048, USA.
E-mail address: Richard.Lewis@cshs.org (R.A. Lewis).

https://doi.org/10.1016/j.jns.2022.120478
Received 18 May 2022; Received in revised form 29 September 2022; Accepted 23 October 2022
Available online 27 October 2022

sensory disturbances is often complex, and involves distinguishing it from other conditions that mimic CIDP [9,10]. Traditionally, CIDP was viewed as a single neuropathy that was distinct from other neuropathies such as GBS, monoclonal gammopathy of undetermined significance (MGUS), POEMS (polyneuropathy- organomegaly-endocrinopathy-M-protein-skin changes) syndrome, multifocal motor neuropathy (MMN) and vasculitis. A more modern view recognizes several subgroups within the CIDP spectrum. In 2019, Bunschoten et al. proposed a spectrum that included typical CIDP and distal, multifocal/focal, motor and sensory CIDP variants, with some overlap with immunoglobulin IgG4 nodal-paranodal antibody neuropathies and monoclonal gammopathies [11]. This spectrum has been further updated (Fig. 1) following publication of the 2021 European Academy of Neurology (EAN)/PNS CIDP guideline [9,10].

Acute-onset CIDP continues to be included as a subtype of typical CIDP. Conversely, autoimmune nodopathies [i.e., antibodies against contactin associated protein 1 (Caspr1), contactin 1 (CNTN1), neurofascin (NF)-186 and NF155], anti-myelin associated glycoprotein (MAG) neuropathy, POEMS syndrome and MMN are not included as part of the CIDP syndrome (Fig. 1), as they have distinct clinical, laboratory, electrodiagnostic, pathological and treatment features [9,10]. Chronic immune sensory polyradiculopathy [12–14] is also currently considered to be separate from CIDP (Fig. 1), as it is not clearly a demyelinating CIDP variant, and requires further investigation [9,10].

3. Diagnosis of typical CIDP and its variants

In the revised 2021 EAN/PNS guidelines [9,10], the term 'typical CIDP' is retained but the term 'CIDP variants' replaces 'atypical CIDP', with well-characterized CIDP variants —distal, multifocal/focal, pure motor and pure sensory CIDP (Fig. 1) — being added. All forms of CIDP have some common features, including progression or relapse over a period of >8 weeks, areflexia, nerve changes indicative of segmental demyelination and, generally, a response to immunomodulation with intravenous immunoglobulin (IVIg), corticosteroids or plasma exchange. However, the pathogenic mechanisms and clinical presentation may differ between variants, with each form having some unique characteristics that modify their differential diagnosis. To that end, the 2021 guidelines include tables of differential diagnostic considerations for each variant as well as tables that advise on diagnostic investigations useful in evaluating each potential variant (Tables 1 and 2 and Fig. 2).

When diagnosing CIDP and its variants, specific clinical and electrodiagnostic criteria must be met. Among the changes in the 2021 guideline [9,10], the levels of diagnostic certainty have been reduced to two (CIDP and possible CIDP) from three (definite, probable, possible CIDP) in the 2010 guideline [9,10]. These changes were made as there were no significant differences in the accuracy of the electrodiagnostic criteria for probable CIDP [6,15], and to avoid using the term 'definite CIDP' due to the lack of a gold standard for diagnosis.

3.1. Clinical criteria

When diagnosing CIDP, the first question to ask is 'Is the clinical history and examination consistent with CIDP?' [9,10]. This is evaluated by assessing whether the patient exhibits weakness and sensory disturbance in keeping with typical or variant CIDP. It is necessary to identify the clinical phenotype of CIDP based on its motor and sensory characteristics due to differences in their 'red flags' and differential diagnoses (Table 1) and diagnostic workflow (Fig. 2 and Table 2).

3.1.1. Typical CIDP

Typical CIDP is the most common form of CIDP. It presents with symmetrical motor and sensory signs and symptoms affecting at least two limbs that follow a progressive or relapsing-remitting pattern for ≥8 weeks [9,10]. On examination, there is proximal and distal muscle weakness of the upper and lower limbs, sensory involvement and absence or reduced tendon reflexes in all limbs. The 2009 Koski criteria for CIDP noted that the typical CIDP phenotype (symmetrical proximal and distal weakness with areflexia) was virtually pathognomonic for CIDP, with nerve conduction studies confirming, but not adding to, diagnostic accuracy [16].

Acute CIDP develops more rapidly, with considerable weakness within 4 weeks, and initially may be diagnosed as GBS. However, unlike patients with GBS, patients will continue to deteriorate for ≥8 weeks or

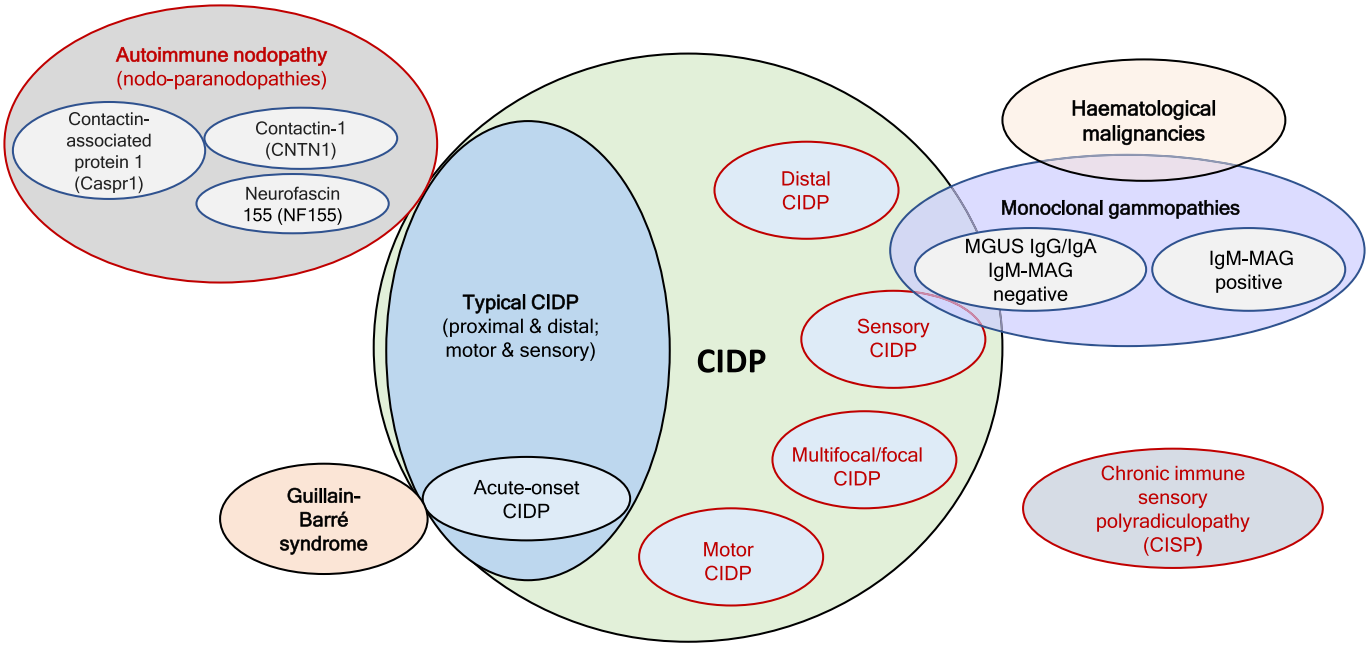

Fig. 1. Changes in the spectrum of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) based on the 2021 second revision of the European Academy of Neurology/Peripheral Nerve Society guideline [9,10]. Red text/ovals indicate changes made in the 2021 guideline. *Ig* immunoglobulin, *MAG* myelin-associated glycoprotein, *MGUS* monoclonal gammopathy of undetermined significance. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Table 1
Red flags and examples of potential alternative diagnoses for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) based on the second revision of the European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) guideline [9,10].

Red flags for considering an alternative diagnosis	Examples of potential alternative diagnoses
Typical CIDP Onset <4 weeks (possible GBS); subacute, low-frequency tremor, marked ataxia, distal predominance (possible autoimmune nodopathy[a]); diabetes (possible diabetic NP); IgA or IgG monoclonal gammopathy (possible multiple myeloma, AL amyloidosis, POEMS syndrome)	GBS; multiple myeloma; osteosclerotic myeloma; POEMS syndrome; AL amyloidosis, ATTRv-PN; HIV-related NP; hepatic NP; uraemic NP; actual or functional vitamin B_{12} deficiency, autoimmune nodopathy[a]; CANOMAD
Distal CIDP[b] Pain and autonomic features predominant (possible AATRv NP); subacute, low-frequency tremor, marked ataxia, distal predominance (possible autoimmune nodopathy phenotype); family history of CMT or AATR); diabetes (possible diabetic NP); IgA or IgG monoclonal gammopathy (possible multiple myeloma, AL amyloidosis, POEMS syndrome); IgM monoclonal gammopathy (possible anti-MAG NP)	Anti-MAG IgM NP; IgG paraproteinemia (multiple myeloma); diabetic NP; hereditary NP (e.g., CMT type 1, X-linked or type 4, metachromic leukodystrophy, Refsum disease, adrenomyeloneneuropathy, ATTRv-PN); POEMS syndrome; vasculitic NP; autoimmune nodopathy[a]; CIAP
Multifocal /focal CIDP[c] Pain (possible diabetic radiculopathy/plexopathy, neuralgic amyotrophy); normal sensation (possible MMN); only 1 nerve in 1 limb affected (possible nerve entrapment/tumour); family history of HNPP; signs of vasculitis (possible vasculitis NP)	Diabetic radiculopathy/plexopathy; entrapment NPs; HNPP; MMN; neurological amyotrophy; peripheral nerve tumours (e.g., lymphoma, perineurioma, schwannoma, neurofibroma); vasculitic NP (mononeuritis multiplex)
Motor CIDP Dyspnoea, dysarthria and dysphagia (possible MND, myasthenia gravis); family history of hereditary motor NPs; asymmetrical weakness at onset (possible MMN); elevated serum creatine kinase levels (possible myositis)	Hereditary motor NPs (e.g., distal hereditary motor NPs, spinal muscular atrophy, porphyria); MND; neuromuscular junction disorders (e.g., myasthenia gravis, Lambert-Eaton syndrome); advanced MMN; inflammatory myopathies
Sensory CIDP Family history of hereditary motor NP; diabetes (possible diabetic NP); vitamin B_{12} deficiency, chemotherapy (possible sensory neuronopathy); IgM monoclonal gammopathy (possible anti-MAG NP); normal motor and sensory conduction (possible CISP)	CANVAS; CISP; dorsal column lesions (due to paraneoplastic syndromes, syphilis, or copper or vitamin B_{12} deficiency); hereditary sensory NPs; idiopathic sensory NP; sensory NP; toxic NPs (e.g., due to chemotherapy or vitamin B_6 toxicity)

AATR transthyretin amyloidosis, *AATRv-PN* variant AATR polyneuropathy, *AL* amyloid light-chain, *CANOMAD*, chronic neuropathy ophthalmoplegia M-protein agglutination disialosyl antibodies, *CANVAS* cerebellar ataxia, neuropathy and vestibular areflexia syndrome; *CIAP* chronic idiopathic axonal polyneuropathy, *CISP* chronic immune sensory polyradiculopathy, *CMT* Charcot-Marie-Tooth disease, *GBS* Guillain-Barré syndrome, *HNPP* hereditary NP with liability to pressure palsies, *Ig* immunoglobulin, *MAG* myelin-associated glycoprotein, *MMN* multifocal motor NP, *MND* motor neuron disease, *NP* neuropathy, *POEMS* polyneuropathy-organomegaly-endocrinopathy-M-protein-skin changes.

[a] Especially the presence of contactin-associated protein 1 (Caspr1), neurofascin (NF) 155 or contactin (CNTN) 1 IgG4 antibodies. Autoimmune nodopathies is the term proposed by the EAN/PNS taskforce for conditions associated with antibodies against these nodal-paranodal cell-adhesion molecules. These conditions are also referred to as nodo-paranodopathies.

[b] Also known as distal acquired demyelinating symmetric NP (DADS).

[c] Also known as Lewis-Sumner syndrome, multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), multifocal acquired demyelinating NP with persistent conduction block, multifocal inflammatory demyelinating NP.

relapse three or more times after initial improvement [17–19].

3.1.2. Distal CIDP

Distal CIDP presents as distal sensory loss and weakness predominantly in the lower limbs, with gait instability [9,10]. The upper limbs may also be affected by distal sensory loss and weakness.

Importantly, patients with distal neuropathy who have IgM paraprotein and anti-MAG antibodies (i.e., anti-MAG neuropathy) are not classified as distal CIDP [20–22]. Anti-MAG neuropathy is not considered to be a type of CIDP, as electrodiagnostic and pathological findings are specific in most patients, and it does not usually respond to IVIg or corticosteroids [9,10]. Testing for anti-MAG may, therefore, be a useful additional investigation in differentiating distal CIDP from anti-MAG neuropathies. Furthermore, a recently developed diagnostic score may prove useful in discriminating between patients with anti-Mag neuropathy and those with anti-MAG antibodies with a CIDP-like presentation, thus aiding in the treatment of these conditions [23].

In patients with distal CIDP with lambda light chain associated IgA or IgG paraproteins, testing of vascular endothelial growth factor (VEGF) serum levels can help differentiate distal CIDP from suspected POEMS syndrome.

3.1.3. Multifocal CIDP

Multifocal CIDP presents with motor and sensory symptoms that are usually asymmetrical and predominant in the upper limbs [9,10]. The upper limbs are usually affected first. The lower limbs may become involved later or are sometimes affected from the onset [24,25]. Relative to other types of CIDP, the cranial nerves, including oculomotor, trigeminal, facial, vagal and hypoglossal nerves, are probably more frequently involved [22,26–31].

Focal CIDP, which is rare, generally affects the brachial or lumbosacral plexus. But may also affect individual peripheral nerves [32,33]. Antinuclear antibody (ANA) and antineutrophil cytoplasmic antibody (ANCA) tests may help differentiate multifocal/focal CIDP from other diseases; for example, the presence of positive ANA indicates mononeuropathy multiplex rather than CIDP.

3.1.4. Motor CIDP

Motor CIDP presents with relatively symmetric proximal and distal weakness, and normal sensation (both clinically and electrodiagnostically [34,35]) [9,10]. In contrast, sensation is abnormal in typical CIDP and other CIDP variants, and weakness is asymmetrical and mainly affects the upper limbs in MMN.

In patients with clinical signs of motor CIDP who have some

Table 2
Summary of clinical and electrodiagnostic criteria for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) based on the second revision of the European Academy of Neurology/Peripheral Nerve Society guideline [9,10].

Clinical criteria		Electrodiagnostic criteria (no. of nerves)		Diagnosis and level of certainty
Weakness	Sensory disturbance	Motor conduction criteria[a]	Sensory conduction criteria[b]	
Typical CIDP				
Symmetrical in 4 limbs, proximal and distal	In ≥2 limbs	Met in ≥2	Abnormal in ≥2	Typical CIDP
		Met in 1	Abnormal in ≥2	Possible typical CIDP (upgrade to typical CIDP if 2 SC[c])
		Non-diagnostic abnormalities		Possible typical CIDP if objective treatment response +1 other SC
Distal CIDP				
Predominantly in lower limbs, distal	In ≥2 limbs	Met in ≥2 (in upper limbs)	Abnormal in ≥2	Distal CIDP
		Met in 1 upper limb	Abnormal in ≥2	Possible distal CIDP (upgrade to distal CIDP if 2 SC[c])
		Met in lower limb only	Abnormal in ≥2	Possible distal CIDP
Multifocal/focal CIDP				
In ≥2 limbs (multifocal); in 1 limb (focal)	In distribution affected nerves	Met in ≥2	Abnormal in ≥2	Multifocal CIDP
		Met in 1	Abnormal in ≥2	Possible multifocal CIDP (upgrade to multifocal CIDP if 2 SC[c])
		Met in 1	Abnormal in 1	Possible focal CIDP
Motor or motor-predominant CIDP				
Symmetrical in 4 limbs, proximal and distal	Normal sensory conduction	Met in ≥2	Normal in 4	Motor CIDP
		Met in 1	Normal in 4	Possible motor CIDP (upgrade to motor CIDP if 2 SC[c])
		Met in ≥2	Abnormal in ≥2	Motor-predominant CIDP
		Met in 1	Abnormal in ≥2	Possible motor-predominant CIDP (upgrade to motor-predominant CIDP if 2 SC[c])
Sensory or sensory-predominant CIDP				
None	Symmetrical in 4 limbs	Met in ≥2	Abnormal in ≥2	Sensory-predominant CIDP
		Met in 1	Abnormal in ≥2	Possible sensory-predominant CIDP (upgrade to sensory-predominant CIDP if 2 SC[c])
		Normal in 4	Abnormal in ≥2	Possible sensory CIDP[d]

CMAP compound muscle action potential, *LLN* lower limit of normal, *SC* supportive criteria, *SNAP* sensory nerve action potential, *ULN* upper limit of normal, ↑ increase/prolongation, ↓ decrease.

[a] Features strongly supportive of demyelination include ≥1 of the following: motor distal latency ↑ to ≥50% above ULN in 2 nerves (except that due to carpal tunnel syndrome); ↓ in motor conduction velocity to ≥30% below the LLN in 2 nerves; ↑ in F-wave latency to ≥20% of ULN (≥50% if the amplitude of distal negative peak CMAP <80% of LLN); absence of F-waves in 2 nerves (in nerves with a distal negative peak CMAP amplitude ≥20% of LLN) + ≥1 other demyelinating parameter in ≥1 other nerve; motor conduction block (≥30% ↓ of the proximal relative to distal negative peak CMAP amplitude, excluding the tibial nerves, and distal negative peak CMAP amplitude ≥20% of LLN in 2 nerves; or in 1 nerve + ≥1 demyelinating parameter in ≥1 other nerve except absence of F-waves in ≥1 other nerve); abnormal temporal dispersion (>30% ↑ in duration between the proximal and distal negative peak CMAP in ≥2 nerves [≥100% ↑ in the tibial nerve]); or distal CMAP duration ↑ in ≥1 nerve (median, ulnar, peroneal and tibial values at low frequency filter bandpass of 2, 5, 10 or 20 Hz) + ≥1 other demyelinating parameter in ≥1 other nerve.

[b] Sensory conduction abnormalities include ≥1 of the following in 2 nerves: ↑ in distal latency; ↓ SNAP amplitude, or slowed conduction velocity outside of normal limits.

[c] Includes objective treatment response and findings from nerve ultrasound, nerve MRI, nerve biopsy and cerebrospinal fluid.

[d] Normal motor nerve conduction studies plus either: sensory nerve conduction velocity < 80% of LLN (for SNAP amplitude >80% of LLN) or < 70% of LLN (for SNAP amplitude <80% of LLN) in ≥2 nerves (median, ulnar, radial, sural nerve); or sural sparing pattern (abnormal median or radial SNAP with normal sural nerve SNAP) [excluding carpal tunnel syndrome].

abnormalities in sensory conduction studies [36], the variant is known as motor-predominant CIDP. Of note, patients with motor CIDP may deteriorate after treatment with corticosteroids [34,36–38], similar to patients with MMN. Although the reason for this deterioration is currently unknown, the use of corticosteroids should be avoided in motor CIDP and IVIg is considered first-line treatment [9,10].

Creatine kinase levels, muscle biopsy and anti-acetylcholine receptor antibody testing may be useful in differentiating this variant from other possible conditions (Table 1). Other disorders that must be considered in the differential diagnoses are various genetic and non-genetic forms of motor neuron disease (MND). To diagnose motor CIDP, it is necessary to identify conduction abnormalities that meet EAN/PNS CIDP criteria.

3.1.5. Sensory CIDP

Sensory CIDP presents with sensory signs and symptoms that are symmetrical in four limbs, without the presence of motor weakness [9,10]. It is usually characterized by gait ataxia, impairments in vibration and position sensation, and changes in cutaneous sensation [39–41]. In patients with clinical signs of sensory CIDP who show motor nerve conduction slowing or motor conduction block, the variant is known as sensory-predominant CIDP [39,42,43].

Importantly, sensory CIDP is often a transient clinical stage that precedes the appearance of motor weakness. In long-term follow-up studies, approximately 70% of patients with sensory CIDP eventually developed motor weakness [37,44]. Paraneoplastic antibody screening may help differentiate sensory CIDP from paraneoplastic dorsal column lesions (Table 1).

Of note, although induction treatment with IVIg and maintenance treatment with subcutaneous Ig have been effective in treating other forms of CIDP [9,10], they have not yet been evaluated in randomized clinical trials in patients with sensory CIDP.

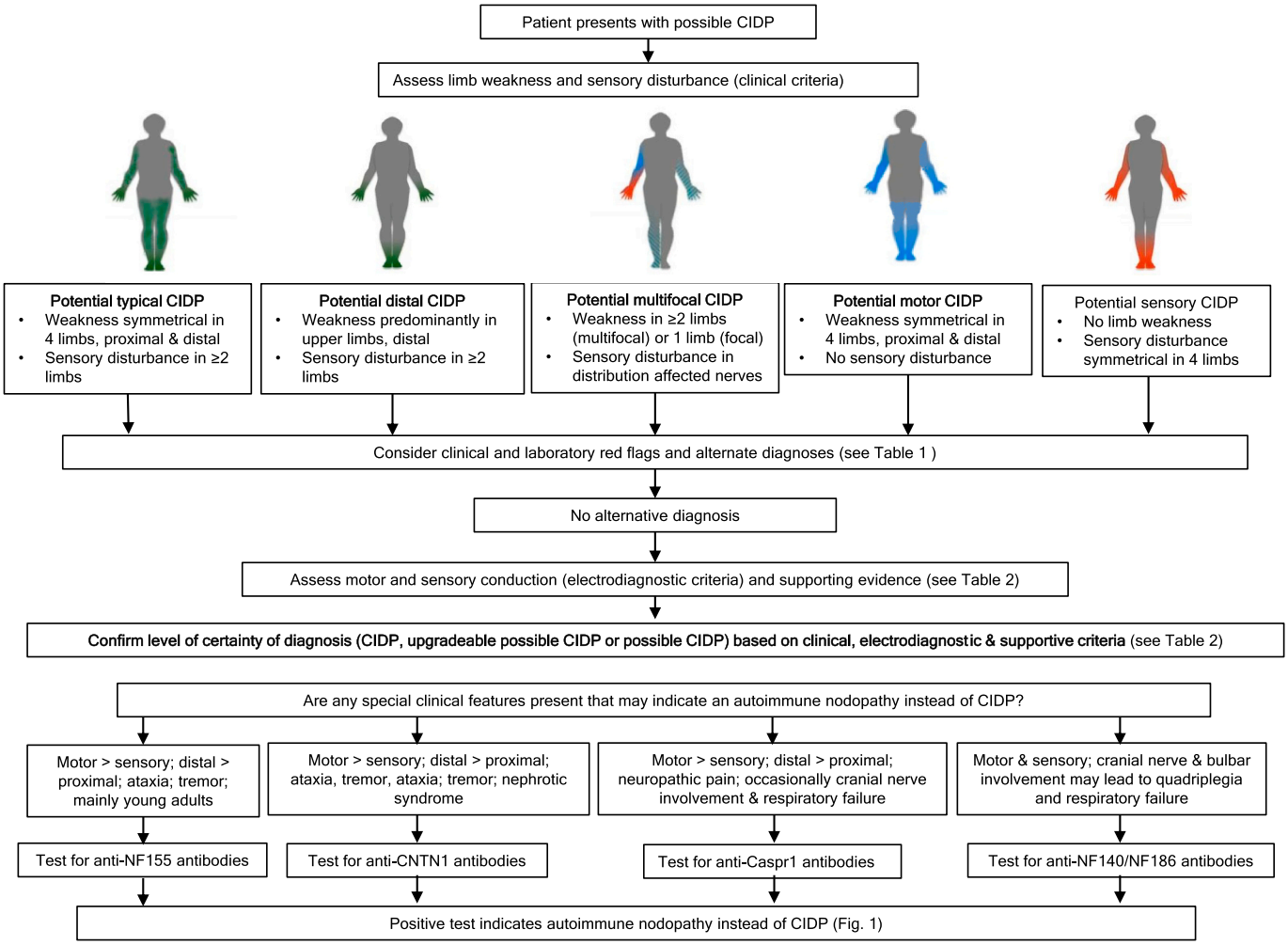

Fig. 2. Summary of key steps in the differential diagnosis of typical and variant chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) based on the 2021 second revision of the European Academy of Neurology/Peripheral Nerve Society guideline [9,10]. In the figures, green indicates motor-sensory, blue indicates motor and red indicates sensory involvement. *Caspr* contactin-associated protein, *CNTN* contactin, *NF* neurofascin. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

3.2. Electrodiagnostic criteria

If the clinical criteria for CIDP are met, the second question is 'Do nerve conduction studies indicate demyelination?' In the 2021 EAN/PNS guideline [9,10], the taskforce chose to include both motor and sensory conduction criteria to support the clinical diagnosis of typical and variant CIDP (Table 2), due to the long-standing clinical experience, widespread availability and relative inexpensiveness of electrodiagnostic testing.

The motor conduction criteria in the 2021 guidelines [9,10] (Table 2) are similar to those in the 2010 guidelines [3], with a few exceptions: 1) prolongation of F-wave latency is now defined as ≥20% [9,10] vs ≥30% [3] above the upper limit of normal; 2) motor conduction block is defined as a ≥ 30% [9,10] vs ≥50% [3] reduction of the proximal relative to distal negative peak amplitude; 3) distal compound muscle action potential duration prolongation is defined as median, ulnar, peroneal and tibial values at low frequency filter bandpass of 2, 5, 10 or 20 Hz [9,10] vs one set of values [3]. Importantly, the 2021 guidelines expanded the 2010 electrodiagnostic criteria by including sensory nerve conduction criteria specific for CIDP and defining criteria specific for its variants (Table 2).

3.2.1. Typical CIDP

The diagnosis of typical CIDP is made when the patient meets the clinical criteria and the motor conduction criteria are met in at least two nerves and sensory abnormalities are present in at least two nerves [9,10]. If motor conduction criteria are met in only one nerve and sensory conduction abnormalities are present in at least two nerves, a diagnosis of possible CIDP can be made (upgradeable to CIDP if at least two supportive criteria are met) (Table 2).

As up to one-fifth of patients with clinically typical CIDP do not fulfil minimal electrodiagnostic criteria for possible CIDP (one nerve abnormality), such patients may be diagnosed as possible typical CIDP if they show an objective response to any of the three proven CIDP treatments (IVIg, corticosteroids and plasma exchange) along with at least one other supportive criterion (Table 2) [9,10].

3.2.2. Distal CIDP

A clinical diagnosis of distal CIDP is confirmed if motor conduction criteria are fulfilled in at least two upper limb nerves and sensory conduction abnormalities are present in at least two nerves [9,10]. The distal negative peak compound muscle action potential amplitude should be ≥1 mV. If other criteria are met, but motor criteria are met in only one upper limb nerve or two lower limb nerves, the diagnosis is possible distal CIDP.

3.2.3. Multifocal/focal CIDP

A clinical diagnosis of multifocal CIDP is confirmed if motor

conduction criteria are met in at least two nerves in at least two limbs with sensory conduction abnormalities present in at least two nerves of the affected limbs [9,10]. If other criteria are met but motor criteria are fulfilled in only one nerve, the maximum certainty of the diagnosis is possible multifocal CIDP.

A clinical diagnosis of focal CIDP is confirmed if motor conduction criteria are met in at least two nerves in one limb with sensory conduction abnormalities present in at least two nerves of the affected limb [9,10]. If motor and sensory conduction criteria are fulfilled in only one nerve, the maximum certainty of the diagnosis is possible focal CIDP.

3.2.4. Motor and motor-predominant CIDP

A clinical diagnosis of motor CIDP is confirmed if motor conduction criteria are fulfilled in at least two nerves and sensory conduction is normal in all of at least four nerves (median, ulnar, radial, and sural) [9,10]. If other criteria are met but motor criteria are fulfilled in only one nerve, the diagnosis is possible motor CIDP.

If sensory conduction abnormalities are present in two nerves, the diagnosis is motor-predominant CIDP if motor conduction criteria are fulfilled in least two nerves, and possible motor-predominant CIDP if motor conduction criteria are fulfilled in only one nerve [9,10].

3.2.5. Sensory and sensory-predominant CIDP

A diagnosis of possible sensory CIDP can be made if sensory conduction abnormalities are found in two nerves and motor conduction is normal in all of at least four nerves (median, ulnar, peroneal, and tibial) [9,10].

In patients with motor conduction abnormalities in addition to sensory conduction abnormalities in two nerves, the diagnosis is sensory-predominant CIDP if motor conduction criteria are fulfilled in two nerves, and possible sensory-predominant CIDP if motor conduction criteria are fulfilled in one nerve [9,10].

3.3. Supportive criteria

If clinical and electrodiagnostic criteria allow only for a diagnosis of possible CIDP, several additional tests – cerebrospinal fluid (CSF) analysis, nerve ultrasound, nerve magnetic resonance imaging (MRI), nerve biopsy and objective treatment response – can be performed to support the diagnosis (Table 2) [9,10]. However, based on assessments of their diagnostic accuracy, these tests cannot be used as primary diagnostic criteria for CIDP.

3.3.1. Cerebrospinal fluid analysis

CSF analysis may not be as useful as previously thought in the diagnosis of CIDP. Due to its unproven independent diagnostic value in typical CIDP and its uncertain sensitivity in CIDP variants [8,29], the taskforce was unable to make a formal recommendation to use CSF analysis in diagnosing CIDP [9,10]. If clinical and electrodiagnostic criteria for CIDP have already been met, CSF analysis may not be required [9,10]. However, CSF analysis should be considered to exclude other diagnoses or to support a diagnosis of CIDP when 1) the diagnostic criteria for possible CIDP but not CIDP are met; 2) the patient has weakness and sensory disturbances with an acute or subacute onset; 3) there is a suspected or possible infectious or malignant cause.

High CSF protein levels are generally used as a positive indicator of CIDP. However, elevated levels of protein in the CSF should be interpreted cautiously in patients with diabetes or those aged >50 years (due to their higher normal CSF protein levels [45,46]). A diagnosis of CIDP or possible CIDP should not be made based on an elevated CSF protein in the absence of clinical and/or electrodiagnosis criteria (due to the risk of misdiagnosis [47]) [9,10]. On the other hand, normal CSF protein levels do not exclude a diagnosis of CIDP or a response to immune treatment.

Unfortunately, rigorous cut-off values for CSF protein levels in CIDP cannot be established due to the lack of sufficient data. The clinical experience of the physician and the laboratory's normative CSF protein values should be relied upon to guide the diagnosis. Nonetheless, very high CSF protein levels (i.e., ≥1 g/L) strongly suggest the presence of CIDP [48], whereas levels below 1 g/L are more frequently associated with misdiagnosis [47]. Of note, a recent investigation of oligoclonal IgG bands in CSF found no evidence of a CSF-restricted humoral response in patients with CIDP [49].

3.3.2. Nerve ultrasound

As nerve enlargement is not specific to CIDP, nerve ultrasound has only moderate diagnostic accuracy in CIDP and is considered a supportive criterion [9,10]. Nerve ultrasound has the advantages of being a non-invasive, easily-repeated, relatively low-cost procedure that is widely available. However, interpreting the results requires expertise, practice and the establishment of intra-laboratory normal values.

Parameters evaluated during nerve ultrasound include the cross-sectional nerve size, fascicle size, nerve vascularity, echogenicity and epineurium thickness [47]. A diagnosis of CIDP may be more likely if there is nerve enlargement in at least two sites in proximal median nerve segments and/or the brachial plexus [50–53]. Normative values for the cross-sectional area of the median nerve are available, with nerve enlargement being shown by values of >10 mm^2 at the forearm, >13 mm^2 at the upper arm, >9 mm^2 for the interscalene (trunks) and > 12 mm^2 for nerve roots [54,55].

Although not featured in the 2021 CIDP guideline, muscle ultrasound may provide additional information. In a recent study in 80 patients with typical or variant CIDP [56], distal muscles showed increased echointensity, indicating fibrosis and fatty infiltration due to secondary axonal damage, which correlated with disease severity. Fasciculations, which are a potential marker of active axonal damage, were frequently observed in distal muscles. Although further studies on specificity and sensitivity are required, echointensity information from muscle ultrasound may prove to be a prognostic, and potentially even diagnostic, marker for CIDP.

3.3.3. Nerve MRI

Nerve MRI is a supportive criterion in patients fulfilling the diagnostic criteria for possible CIDP when nerve ultrasound is not available or when nerve ultrasound results are non-contributory [9,10].

A diagnosis of CIDP may be more likely if there is nerve enlargement and/or increased signal intensity of nerve roots on T_2-weighted MRI sequences (DIXON/STIR, coronal and sagittal planes) [50,57–60]. Preferably, evaluation includes a quantitative assessment of spinal nerve root sizes (nerve root diameter immediately next to the ganglion, measured as coronal plane height with a cut-off value of >5 mm) or semi-quantitative scoring of abnormalities of the spinal nerve roots and trunks as normal, possibly abnormal or clearly abnormal [9,10]. However, as nerve enlargement and increased signal intensity are associated with conditions that mimic CIDP, it is important to consider other potential diagnoses.

Nerve ultrasound is currently preferred over nerve MRI due to the low inter-rater reliability, lack of objective cut-off values, relatively high cost and selection bias of nerve MRI in current studies [9,10]. However, the value of nerve MRI in the differential diagnosis of CIDP may improve as its specificity is enhanced by more precise measurement criteria. In a systematic comparison of the nerve architecture of the brachial plexus in a recent study [61], significant differences in quantitative MRI parameters were shown between patients with CIDP, MMN and MND, which may reflect differences in underlying pathophysiological mechanisms. The investigators took great care in precisely measuring the nerve root and calculating the values, which may be necessary when determining which measurements to take with nerve MRI.

3.3.4. Immunological testing

Testing for serum monoclonal proteins is strongly advised in patients with clinical suspicion of CIDP, with further evaluation recommended if a gammopathy is found [9,10]. As monoclonal gammopathies can be

associated with neuropathies presenting a clinical picture resembling CIDP (e.g., anti-MAG IgM paraproteinemic neuropathy, POEMS syndrome, multiple myeloma), it is vital that the neurological and oncological conditions are correctly identified to enable appropriate treatment [9,10].

Nodal/paranodal or MAG antibody testing should be considered in patients who fulfil criteria for CIDP and present with particular characteristics or do not respond well to proven effective treatments [9,10]. Antibody testing is associated with relatively low costs, and positive results have significant implications for diagnosis and treatment; however, nodal/paranodal antibody testing is not widely available.

Testing for nodal and paranodal autoantibodies in patients with clinical suspicion of CIDP should be considered when nodal and paranodal (anti-NF155, anti-CNTN1, anti-Caspr1) and possibly anti-NF140/186) antibody testing is available and meets quality standards [9,10]. The proviso for 'availability' and 'quality standards' is included in recognition that not all laboratories can provide such testing. Clinical hints of paranodopathies include an onset similar to that of GBS, tremor and poor response to IVIg and corticosteroids. Antibodies to the proteins of the paranodal complex, including anti-NF155/−86, anti-CNTN1 and anti-Caspr1, may be present [62]. The predominant antibody subtype may be IgG4 (not complement dependent) or, in more acute cases, IgG3 (complement dependent). The screening test is serum binding on teased nerve fibres, with results confirmed by a specific enzyme-linked immunosorbent assay (ELISA). The importance of correctly diagnosing paranodopathies is highlighted by the case of a 30-year-old male who developed severe progressive distal neuropathic pain and sensory deficits, was diagnosed with CIDP and, despite IVIg treatment, was unable to walk within a few months [63]. Binding assays showed autoantibodies against Caspr1 with IgG4 as the predominant autoantibody subclass. Following four cycles of rituximab, which has proven effectiveness in treating paranodopathies [64,65], the patient was pain free and could walk without aid [63].

All patients fulfilling CIDP diagnostic criteria should be tested for a paraprotein. Patients who are IgM-positive should undergo anti-MAG antibody testing. Anti-MAG antibody testing is widely available, with Bühlmann test ELISA and locally validated ELISA, Western blot or immunohistochemistry assays being recommended. A high anti-MAG antibody titre (>7000 Bühlmann units) [66] strongly suggests a diagnosis other than CIDP with a potentially different response to treatment.

3.3.5. Nerve biopsy

Nerve biopsies should not routinely be performed to diagnose CIDP [9,10]. Due to their low diagnostic accuracy, invasive nature and the availability of other diagnostic means, their use should be limited to unusual cases when all other investigations are non-diagnostic [9,10].

In the past, nerve biopsies were often conducted and have provided important information about CIDP [67–70]. For example, macrophage clusters around vessels are markedly increased in patients with CIDP relative to patients with hereditary neuropathies and healthy controls, suggesting a possible diagnostic marker for CIDP [67]. Currently, however, nerve biopsies should be used only in carefully selected patients when CIDP is suspected but cannot be confirmed with clinical, laboratory, imaging and electrodiagnostic studies, there is little or no response to treatment, skilled and specialized physicians and laboratories are available, and symptoms are severe enough to justify the potential complications.

3.3.6. Treatment response

A response to treatment with a standard immunomodulatory agent may assist in confirming a diagnosis of CIDP, as IVIg (high-certainty evidence), and corticosteroids and plasma exchange (both moderate-certainty evidence) have been shown to improve impairment [71]. Objective treatment response is considered to be a supportive diagnostic criterion for CIDP in patients in whom clinical, electrodiagnostic and other supportive criteria allow only for a diagnosis of possible CIDP (Table 2) [9,10].

As current immunomodulatory treatments are not specific for CIDP, treatment response must be considered carefully in tandem with clinical and electrophysiological evidence to avoid over-diagnosing CIDP [9,10]. In patients with possible CIDP, an objective response to treatment increases the probability of a CIDP diagnosis, but a response could also occur if another autoimmune condition was present. Importantly, treatment failure may or may not indicate a misdiagnosis of CIDP, as failure may be due to other causes such as inadequate treatment dosage or duration [72]. In patients who do not respond to at least one of the three proven effective CIDP treatments, it is appropriate to review the CIDP diagnosis and consider other conditions before considering other immunosuppressive therapies.

The guideline defines an objective treatment response as an improvement in at least one measure of disability and at least one measure of impairment. Although the changes required to define improvement have not been adequately validated, those which have been used in clinical trials can serve as a guide (Table 3) [9,10].

4. Discussion

The revised 2021 EAN/PNS guideline aimed to improve the specificity of the diagnostic criteria for typical CIDP and each of the CIDP variants [9,10]. In this article, we reviewed key diagnostic-related revisions in the guideline including the addition of four well-characterized CIDP variants, changes in specific clinical, electrodiagnostic (including the addition of sensory conduction criteria) and supportive criteria for typical CIDP and each of the CIDP variants, and revisions in the levels of diagnostic certainty for typical and variant CIDP.

Improvements in the guidelines for the diagnosis of CIDP were necessary, as CIDP, especially the variant forms, continued to be commonly over- or under-diagnosed [47,72–74]. Problems associated with misdiagnosis include not recognizing the key clinical signs, especially of CIDP variants, misinterpretation of nerve conduction studies, lack of adherence to electrodiagnostic criteria, over-reliance on supportive findings such as CSF protein levels, and failure to exclude other causes of polyneuropathy.

The addition to the guidelines of specific criteria for the four variant forms of CIDP should aid in the diagnosis of these conditions. Although typical CIDP with its proximal and distal weakness may be relatively easy to diagnose, it is more difficult to diagnose CIDP variants. For example, in a review of the accuracy of CIDP diagnosis in 59 patients

Table 3
Examples of disability and impairment scales that may be used to assess an objective response to treatment for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) [9,10].

Measure	Change in measure possibly indicating improvement[a]
Disability measure	
I-RODS [76–78]	↑ of ≥4 centile points
INCAT disability scale [79,80]	↓ of ≥1 point
Impairment measure	
MRC sum score [78,80,81]	↑ of ≥2–4 points[b]
mISS score [80,82]	↓ of ≥2 points
Neuropathic Impairment score [83]	
Grip strength (handheld dynamometry) [80,84–86]	Martin Vigoritmeter: ↑ of ≥8–14 kPa[b] Jamar dynamometer: ↑ of ≥10%[c]

I-RODS Inflammatory Rasch-built Overall Disability Scale, *INCAT* Inflammatory Neuropathy Cause and Treatment, *mISS* modified INCAT Sensory Sum, ↑ increase, ↓ decrease.

[a] These changes have been used in clinical trials and can serve as a guide, but have not been adequately validated.

[b] Higher values may improve diagnostic specificity.

[c] Diagnostic specificity improved if values averaged over 3 consecutive days [84].

[47], 28 patients (47%) were misdiagnosed. All patients with typical CIDP were accurately diagnosed, meeting the 2010 EFNS/PNS diagnostic requirements for CIDP [3,4], and all misdiagnosed patients had a variant form of CIDP [47].

Despite recent advances in the understanding of CIDP, many questions remain about its diagnosis and treatment, as well as other related aspects (e.g., epidemiology and healthcare burden). To reduce the high rates of over- and under-diagnosis of CIDP, it is vital that neuromuscular experts carefully review an individual's clinical findings and electrophysiological test results, and factor in supportive diagnostic criteria when considering a diagnosis of CIDP.

Supportive criteria include an objective response to treatment with one of the three immunomodulatory agents (i.e., IVIg, corticosteroids and plasma exchange) that have proven effective in the treatment of CIDP. However, as other auto-immune conditions may also respond to these treatments, treatment response as a supportive diagnostic criteria needs to be considered carefully along with the clinical and electrophysiological findings. Furthermore, as a fraction of non-responders to at least one of these treatments may still have CIDP, additional testing is needed to rule out disorders that mimic CIDP before considering other immunosuppressive treatment strategies. Despite extensive efforts to objectify treatment response, there are clinical scenarios that cannot be addressed by clinical measures alone [75]. A treatment response test can be diagnostically convenient when the differential diagnosis is narrow and there is a high probability of improvement as assessed by a measurable objective outcome; however, problems occur when 'benefit' is loosely defined [75]. Questions still remain regarding how treatment response should be defined, how long each treatment should be continued and how many of the treatment options should be used before deciding that a lack of treatment response has occurred.

It is hoped that future studies will establish diagnostic biomarkers that can aid in the differential diagnosis of CIDP and other neuropathic disorders, thereby further improving the diagnostic criteria. The establishment of biomarker criteria could also serve as a surrogate marker for treatment response and disease progression in clinical trials, as well as in clinical practice.

5. Conclusion

In conclusion, use of the diagnostic criteria in the revised 2021 EAN/PNS guideline should enable more accurate diagnosis and treatment of CIDP and its variants in clinical practice. In addition, it will allow patient populations to be appropriately defined in clinical trials, thereby improving our understanding of CIDP. The revised guideline still needs to be validated and its strengths and weaknesses assessed. Until then, we need to utilize the revised guideline, recognize pitfalls that can lead to a delayed or incorrect diagnosis of CIDP, and be particularly vigilant before diagnosing a CIDP variant.

Contributions

All authors contributed equally to this publication in concept, writing and editing.

Funding

The manuscript was developed from the proceedings of a Grifols-sponsored virtual symposium, held on 2 November 2021 as part of the 2021 Peripheral Nerve Society Virtual Event.

Disclosures

R. Lewis is a consultant for Annexon, Argenx, CSL Behring, Grifols, J & J, Roche, Pfizer, Sanofi and Takeda, and has received honoraria from Akcea, Alnylam and Medscape.

P. A. van Doorn is a consultant for Annexon, Argenx, Hansa, Immunic, Roche, and Sanofi; has received honoraria from Grifols (all honoraria paid to the Erasmus MC research fund); and has received unrestricted research grants from the Netherlands Organisation for Health Research and Development (ZonMW), Baxter/Shire, Grifols, Prinses Beatrix Spierfonds, and Sanquin.

C. Sommer is a consultant for Alnylam, Air Liquide, Bayer, Immunic, Ipsen, LFB, Merz, Pfizer, Roche and Takeda, and has received honoraria from Alnylam, CSL Behring, Grifols, Lilly, Merck, Novartis, Pfizer and TEVA.

Acknowledgements

Editorial assistance was provided by Katherine Lyseng-Williamson on behalf of Content Ed Net (Madrid, Spain) with funding from Grifols SA (Barcelona, Spain).

References

[1] 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, https://doi.org/10.1111/j.1085-9489.2005.10302.x.

[2] R.A. Hughes, P. Bouche, D.R. Cornblath, E. Evers, R.D. Hadden, A. Hahn, I. Illa, C. L. Koski, J.M. Léger, E. Nobile-Orazio, J. Pollard, C. Sommer, P. Van den Bergh, P. A. van Doorn, I.N. van Schaik, 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. 13 (4) (2006) 326–332, https://doi.org/10.1111/j.1468-1331.2006.01278.x.

[3] European Federation of Neurological Societies/Peripheral Nerve Society, Guideline on management of paraproteinemic demyelinating neuropathies. Report of a Joint Task Force of the European Federation of Neurological Societies and the Peripheral Nerve Society–first revision, J. Peripher. Nerv. Syst. 15 (3) (2010) 185–195, https://doi.org/10.1111/j.1529-8027.2010.00278.x.

[4] P.Y. Van den Bergh, R.D. Hadden, P. Bouche, D.R. Cornblath, A. Hahn, I. Illa, C. L. Koski, J.M. Léger, E. Nobile-Orazio, J. Pollard, C. Sommer, P.A. van Doorn, I. N. van Schaik, 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, https://doi.org/10.1111/j.1468-1331.2009.02930.x.

[5] Y. Rajabally, A. Fowle, P. Van den Bergh, Which criteria for research in CIDP? An analysis of current practice, Muscle Nerve 51 (2014), https://doi.org/10.1002/mus.24496.

[6] 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, https://doi.org/10.1002/mus.24088.

[7] B. Tackenberg, J.D. Lünemann, A. Steinbrecher, E. Rothenfusser-Korber, M. Sailer, W. Brück, S. Schock, R. Zschenderlein, F. Zipp, N. Sommer, Classifications and treatment responses in chronic immune-mediated demyelinating polyneuropathy, Neurology. 68 (19) (2007) 1622–1629, https://doi.org/10.1212/01.wnl.0000260972.07422.ea.

[8] Y.A. Rajabally, G. Nicolas, F. Piéret, 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, https://doi.org/10.1136/jnnp.2009.179358.

[9] P.Y.K. Van den Bergh, P.A. Doorn, R.D.M. Hadden, B. Avau, P. Vankrunkelsven, J. A. Allen, S. Attarian, P.H. Blomkwist-Markens, D.R. Cornblath, F. Eftimov, H. S. Goedee, T. Harbo, S. Kuwabara, R.A. Lewis, M.P. Lunn, E. Nobile-Orazio, L. Querol, Y.A. Rajabally, C. Sommer, H.A. Topaloglu, European Academy of Neurology/Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint Task Force—Second revision, J. Peripher. Nerv. Syst. 26 (3) (2021) 242–268, https://doi.org/10.1111/jns.12455.

[10] P.Y.K. Van den Bergh, P.A. van Doorn, R.D.M. Hadden, B. Avau, P. Vankrunkelsven, J.A. Allen, S. Attarian, P.H. Blomkwist-Markens, D. R. Cornblath, F. Eftimov, H.S. Goedee, T. Harbo, S. Kuwabara, R.A. Lewis, M. P. Lunn, E. Nobile-Orazio, L. Querol, Y.A. Rajabally, C. Sommer, H.A. Topaloglu, European Academy of Neurology/Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint Task Force-Second revision, Eur. J. Neurol. 28 (11) (2021) 3556–3583, https://doi.org/10.1111/ene.14959.

[11] C. Bunschoten, B.C. Jacobs, P.Y.K. Van den Bergh, D.R. Cornblath, P.A. van Doorn, Progress in diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy, Lancet Neurol. 18 (8) (2019) 784–794, https://doi.org/10.1016/s1474-4422(19)30144-9.

[12] M. Sinnreich, C.J. Klein, J.R. Daube, J. Engelstad, R.J. Spinner, P.J. Dyck, Chronic immune sensory polyradiculopathy: a possibly treatable sensory ataxia, Neurology.

63 (9) (2004) 1662–1669, https://doi.org/10.1212/01.wnl.0000142507.12763.58.
[13] Y.A. Rajabally, S.L. Wong, Chronic inflammatory pure sensory polyradiculoneuropathy: a rare CIDP variant with unusual electrophysiology, J. Clin. Neuromuscul. Dis. 13 (3) (2012) 149–152, https://doi.org/10.1097/CND.0b013e31822484fb.
[14] A.M. Clerici, E. Nobile-Orazio, M. Mauri, F.S. Squellati, G.G. Bono, Utility of somatosensory evoked potentials in the assessment of response to IVIG in a long-lasting case of chronic immune sensory polyradiculopathy, BMC Neurol. 17 (1) (2017) 127, https://doi.org/10.1186/s12883-017-0906-2.
[15] P.Y. Van den Bergh, F. Piéret, Electrodiagnostic criteria for acute and chronic inflammatory demyelinating polyradiculoneuropathy, Muscle Nerve 29 (4) (2004) 565–574, https://doi.org/10.1002/mus.20022.
[16] 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, R.A. Lewis, G.J. Parry, P. van Doorn, D. R. Cornblath, Derivation and validation of diagnostic criteria for chronic inflammatory demyelinating polyneuropathy, J. Neurol. Sci. 277 (1–2) (2009) 1–8, https://doi.org/10.1016/j.jns.2008.11.015.
[17] L. Ruts, J. Drenthen, B.C. Jacobs, P.A. van Doorn, Distinguishing acute-onset CIDP from fluctuating Guillain-Barre syndrome: a prospective study, Neurology. 74 (21) (2010) 1680–1686, https://doi.org/10.1212/WNL.0b013e3181e07d14.
[18] M. Odaka, N. Yuki, K. Hirata, Patients with chronic inflammatory demyelinating polyneuropathy initially diagnosed as Guillain-Barré syndrome, J. Neurol. 250 (8) (2003) 913–916, https://doi.org/10.1007/s00415-003-1096-y.
[19] L. Ruts, R. van Koningsveld, P.A. van Doorn, Distinguishing acute-onset CIDP from Guillain-Barré syndrome with treatment related fluctuations, Neurology. 65 (1) (2005) 138–140, https://doi.org/10.1212/01.wnl.0000167549.09664.b8.
[20] S. Larue, F. Bombelli, K. Viala, J. Neil, T. Maisonobe, P. Bouche, L. Musset, E. Fournier, J.M. Léger, Non-anti-MAG DADS neuropathy as a variant of CIDP: clinical, electrophysiological, laboratory features and response to treatment in 10 cases, Eur. J. Neurol. 18 (6) (2011) 899–905, https://doi.org/10.1111/j.1468-1331.2010.03312.x.
[21] M.P. Lunn, E. Nobile-Orazio, Immunotherapy for IgM anti-myelin-associated glycoprotein paraprotein-associated peripheral neuropathies, Cochrane Database Syst. Rev. 10 (10) (2016) Cd002827, https://doi.org/10.1002/14651858.CD002827.pub4.
[22] T. Maisonobe, B. Chassande, M. Vérin, M. Jouni, J.M. Léger, P. Bouche, Chronic dysimmune demyelinating polyneuropathy: a clinical and electrophysiological study of 93 patients, J. Neurol. Neurosurg. Psychiatry 61 (1) (1996) 36–42, https://doi.org/10.1136/jnnp.61.1.36.
[23] P.E. Doneddu, M. Ruiz, E. Bianchi, G. Liberatore, F. Manganelli, D. Cocito, G. Cosentino, L. Benedetti, G.A. Marfia, M. Filosto, C. Briani, C. Giannotta, E. Nobile-Orazio, A diagnostic score for anti-myelin-associated-glycoprotein neuropathy or chronic inflammatory demyelinating polyradiculoneuropathy in patients with anti-myelin-associated-glycoprotein antibody, Eur. J. Neurol. n/a(n/a). doi:https://doi.org/10.1111/ene.15296.
[24] D.S. Saperstein, A.A. Amato, G.I. Wolfe, J.S. Katz, S.P. Nations, C.E. Jackson, W. W. Bryan, D.K. Burns, R.J. Barohn, Multifocal acquired demyelinating sensory and motor neuropathy: the Lewis-Sumner syndrome, Muscle Nerve 22 (5) (1999) 560–566, https://doi.org/10.1002/(sici)1097-4598(199905)22:5<560::aid-mus2>3.0.co;2-q.
[25] R.M. Van den Berg-Vos, L.H. Van den Berg, H. Franssen, M. Vermeulen, T. D. Witkamp, G.H. Jansen, H.W. van Es, H. Kerkhoff, J.H. Wokke, Multifocal inflammatory demyelinating neuropathy: a distinct clinical entity? Neurology. 54 (1) (2000) 26–32, https://doi.org/10.1212/wnl.54.1.26.
[26] S.J. Oh, G.C. Claussen, D.S. Kim, Motor and sensory demyelinating mononeuropathy multiplex (multifocal motor and sensory demyelinating neuropathy): a separate entity or a variant of chronic inflammatory demyelinating polyneuropathy? J. Peripher. Nerv. Syst. 2 (4) (1997) 362–369.
[27] A.P. Jeanjean, T. Duprez, P.Y. Van den Bergh, Massive peripheral nerve hypertrophy in a patient with multifocal upper limb demyelinating neuropathy (Lewis-Sumner syndrome), Acta Neurol. Belg. 101 (4) (2001) 234–238.
[28] M.D. Weiss, J.C. Oakley, G.D. Meekins, Hypoglossal neuropathy in Lewis-Sumner syndrome masquerading as motor neuron disease, Neurology. 67 (1) (2006) 175–176, https://doi.org/10.1212/01.wnl.0000223577.69111.2c.
[29] Y.A. Rajabally, G. Chavada, Lewis-sumner syndrome of pure upper-limb onset: diagnostic, prognostic, and therapeutic features, Muscle Nerve 39 (2) (2009) 206–220, https://doi.org/10.1002/mus.21199.
[30] M. Rabin, G. Mutlu, T. Stojkovic, T. Maisonobe, T. Lenglet, E. Fournier, P. Bouche, J.M. Léger, K. Viala, Chronic inflammatory demyelinating polyradiculoneuropathy: search for factors associated with treatment dependence or successful withdrawal, J. Neurol. Neurosurg. Psychiatry 85 (8) (2014) 901–906, https://doi.org/10.1136/jnnp-2013-306105.
[31] I.M. Lucke, L. Wieske, A.J. van der Kooi, I.N. van Schaik, F. Eftimov, C. Verhamme, Diagnosis and treatment response in the asymmetric variant of chronic inflammatory demyelinating polyneuropathy, J. Peripher. Nerv. Syst. 24 (2) (2019) 174–179, https://doi.org/10.1111/jns.12325.
[32] P.K. Thomas, D. Claus, A. Jaspert, J.M. Workman, R.H. King, A.J. Larner, M. Anderson, J.A. Emerson, I.T. Ferguson, Focal upper limb demyelinating neuropathy, Brain. 119 (Pt 3) (1996) 765–774, https://doi.org/10.1093/brain/119.3.765.
[33] P.Y. Van den Bergh, J.L. Thonnard, T. Duprez, E.C. Laterre, Chronic demyelinating hypertrophic brachial plexus neuropathy, Muscle Nerve 23 (2) (2000) 283–288, https://doi.org/10.1002/(SICI)1097-4598(200002)23:2<283::AID-MUS23>3.0.CO;2-Q.
[34] M. Donaghy, K.R. Mills, S.J. Boniface, J. Simmons, I. Wright, N. Gregson, J. Jacobs, Pure motor demyelinating neuropathy: deterioration after steroid treatment and improvement with intravenous immunoglobulin, J. Neurol. Neurosurg. Psychiatry 57 (7) (1994) 778–783, https://doi.org/10.1136/jnnp.57.7.778.
[35] M. Sabatelli, F. Madia, T. Mignogna, G. Lippi, L. Quaranta, P. Tonali, Pure motor chronic inflammatory demyelinating polyneuropathy, J. Neurol. 248 (9) (2001) 772–777, https://doi.org/10.1007/s004150170093.
[36] A. Pegat, W. Boisseau, T. Maisonobe, R. Debs, T. Lenglet, D. Psimaras, A. Azoulay-Cayla, E. Fournier, K. Viala, Motor chronic inflammatory demyelinating polyneuropathy (CIDP) in 17 patients: clinical characteristics, electrophysiological study, and response to treatment, J. Peripher. Nerv. Syst. 25 (2) (2020) 162–170, https://doi.org/10.1111/jns.12380.
[37] P.E. Doneddu, D. Cocito, F. Manganelli, R. Fazio, C. Briani, M. Filosto, L. Benedetti, A. Mazzeo, G.A. Marfia, A. Cortese, B. Fierro, S. Jann, E. Beghi, A.M. Clerici, M. Carpo, A. Schenone, M. Luigetti, G. Lauria, G. Antonini, T. Rosso, G. Siciliano, G. Cavaletti, G. Liberatore, L. Santoro, E. Peci, S. Tronci, M. Ruiz, S. Cotti Piccinelli, A. Toscano, G. Mataluni, L. Piccolo, G. Cosentino, M. Sabatelli, E. Nobile-Orazio, Atypical CIDP: diagnostic criteria, progression and treatment response. Data from the Italian CIDP Database, J. Neurol. Neurosurg. Psychiatry 90 (2) (2019) 125–132, https://doi.org/10.1136/jnnp-2018-318714.
[38] F. Eftimov, M.H. Liesdek, C. Verhamme, I.N. van Schaik, Deterioration after corticosteroids in CIDP may be associated with pure focal demyelination pattern, BMC Neurol. 14 (2014) 72, https://doi.org/10.1186/1471-2377-14-72.
[39] S.J. Oh, J.L. Joy, R. Kuruoglu, "chronic sensory demyelinating neuropathy": chronic inflammatory demyelinating polyneuropathy presenting as a pure sensory neuropathy, J. Neurol. Neurosurg. Psychiatry 55 (8) (1992) 677–680, https://doi.org/10.1136/jnnp.55.8.677.
[40] X. Ayrignac, K. Viala, R.M. Koutlidis, G. Taïeb, T. Stojkovic, L. Musset, J.M. Léger, E. Fournier, T. Maisonobe, P. Bouche, Sensory chronic inflammatory demyelinating polyneuropathy: an under-recognized entity? Muscle Nerve 48 (5) (2013) 727–732, https://doi.org/10.1002/mus.23821.
[41] K. Viala, T. Maisonobe, T. Stojkovic, R. Koutlidis, X. Ayrignac, L. Musset, E. Fournier, J.M. Léger, P. Bouche, A current view of the diagnosis, clinical variants, response to treatment and prognosis of chronic inflammatory demyelinating polyradiculoneuropathy, J. Peripher. Nerv. Syst. 15 (1) (2010) 50–56, https://doi.org/10.1111/j.1529-8027.2010.00251.x.
[42] A.R. Berger, S. Herskovitz, J. Kaplan, Late motor involvement in cases presenting as "chronic sensory demyelinating polyneuropathy", Muscle Nerve 18 (4) (1995) 440–444, https://doi.org/10.1002/mus.880180411.
[43] J.L. Thonnard, C. Detrembleur, P.Y. Van den Bergh, Assessment of hand function in a patient with chronic sensory demyelinating neuropathy, Neurology. 49 (1) (1997) 253–257, https://doi.org/10.1212/wnl.49.1.253.
[44] G.W. van Dijk, N.C. Notermans, H. Franssen, J.H. Wokke, Development of weakness in patients with chronic inflammatory demyelinating polyneuropathy and only sensory symptoms at presentation: a long-term follow-up study, J. Neurol. 246 (12) (1999) 1134–1139, https://doi.org/10.1007/s004150050531.
[45] H. Kobessho, K. Oishi, H. Hamaguchi, F. Kanda, Elevation of cerebrospinal fluid protein in patients with diabetes mellitus is associated with duration of diabetes, Eur. Neurol. 60 (3) (2008) 132–136, https://doi.org/10.1159/000144083.
[46] A. Breiner, P.R. Bourque, J.A. Allen, Updated cerebrospinal fluid total protein reference values improve chronic inflammatory demyelinating polyneuropathy diagnosis, Muscle Nerve 60 (2) (2019) 180–183, https://doi.org/10.1002/mus.26488.
[47] J.A. Allen, R.A. Lewis, CIDP diagnostic pitfalls and perception of treatment benefit, Neurology. 85 (6) (2015) 498–504, https://doi.org/10.1212/wnl.0000000000001833.
[48] H.C. Lehmann, D. Burke, S. Kuwabara, Chronic inflammatory demyelinating polyneuropathy: update on diagnosis, immunopathogenesis and treatment, J. Neurol. Neurosurg. Psychiatry 90 (9) (2019) 981–987, https://doi.org/10.1136/jnnp-2019-320314.
[49] M. Ruiz, M. Puthenparampil, M. Campagnolo, F. Castellani, A. Salvalaggio, S. Ruggero, E. Toffanin, M. Cacciavillani, P. Gallo, D. Franciotta, C. Briani, Oligoclonal IgG bands in chronic inflammatory polyradiculoneuropathies, J. Neurol. Neurosurg. Psychiatry 92 (9) (2021) 969–974, https://doi.org/10.1136/jnnp-2020-325868.
[50] H.S. Goedee, B.A. Jongbloed, J.H. van Asseldonk, J. Hendrikse, A. Vrancken, H. Franssen, S. Nikolakopoulos, L.H. Visser, W.L. van der Pol, L.H. van den Berg, A comparative study of brachial plexus sonography and magnetic resonance imaging in chronic inflammatory demyelinating neuropathy and multifocal motor neuropathy, Eur. J. Neurol. 24 (10) (2017) 1307–1313, https://doi.org/10.1111/ene.13380.
[51] H.S. Goedee, W.L. van der Pol, J.H. van Asseldonk, H. Franssen, N.C. Notermans, A. J. Vrancken, M.A. van Es, S. Nikolakopoulos, L.H. Visser, L.H. van den Berg, Diagnostic value of sonography in treatment-naive chronic inflammatory neuropathies, Neurology. 88 (2) (2017) 143–151, https://doi.org/10.1212/wnl.0000000000003483.
[52] A. Grimm, T.W. Rattay, N. Winter, H. Axer, Peripheral nerve ultrasound scoring systems: benchmarking and comparative analysis, J. Neurol. 264 (2) (2017) 243–253, https://doi.org/10.1007/s00415-016-8305-y.
[53] C.M. Zaidman, M.B. Harms, A. Pestronk, Ultrasound of inherited vs. acquired demyelinating polyneuropathies, J. Neurol. 260 (12) (2013) 3115–3121, https://doi.org/10.1007/s00415-013-7123-8.
[54] I.J.T. Herraets, H.S. Goedee, J.A. Telleman, R.P.A. van Eijk, J.T. van Asseldonk, L. H. Visser, L.H. van den Berg, W.L. van der Pol, Nerve ultrasound improves detection of treatment-responsive chronic inflammatory neuropathies, Neurology. 94 (14) (2020) e1470–e1479, https://doi.org/10.1212/wnl.0000000000008978.

[55] I.J.T. Herraets, H.S. Goedee, J.A. Telleman, R.P.A. van Eijk, C. Verhamme, C.G. J. Saris, F. Eftimov, N. van Alfen, J.T. van Asseldonk, L.H. Visser, L.H. van den Berg, L.W. van der Pol, Nerve ultrasound for diagnosing chronic inflammatory neuropathy: a multicenter validation study, Neurology. 95 (12) (2020) e1745–e1753, https://doi.org/10.1212/wnl.0000000000010369.
[56] A.L. Fisse, S. Fiegert, Z. Stoykova, J. Brünger, D. Athanasopoulos, T. Grüter, J. Motte, R. Gold, K. Pitarokoili, Increased muscle echointensity correlates with clinical disability and muscle strength in chronic inflammatory demyelinating polyneuropathy, Eur. J. Neurol. 28 (5) (2021) 1698–1705, https://doi.org/10.1111/ene.14716.
[57] S. Kuwabara, M. Nakajima, S. Matsuda, T. Hattori, Magnetic resonance imaging at the demyelinative foci in chronic inflammatory demyelinating polyneuropathy, Neurology. 48 (4) (1997) 874–877, https://doi.org/10.1212/wnl.48.4.874.
[58] A.J. Duggins, J.G. McLeod, J.D. Pollard, L. Davies, F. Yang, E.O. Thompson, J. R. Soper, Spinal root and plexus hypertrophy in chronic inflammatory demyelinating polyneuropathy, Brain. 122 (Pt 7) (1999) 1383–1390, https://doi.org/10.1093/brain/122.7.1383.
[59] Y.A. Rajabally, M.J. Knopp, D. Martin-Lamb, J. Morlese, Diagnostic value of MR imaging in the Lewis-Sumner syndrome: a case series, J. Neurol. Sci. 342 (1–2) (2014) 182–185, https://doi.org/10.1016/j.jns.2014.04.033.
[60] H.S. Goedee, W.L. van der Pol, J. Hendrikse, L.H. van den Berg, Nerve ultrasound and magnetic resonance imaging in the diagnosis of neuropathy, Curr. Opin. Neurol. 31 (5) (2018) 526–533, https://doi.org/10.1097/wco.0000000000000607.
[61] M.H.J. van Rosmalen, H.S. Goedee, R. Derks, F.L. Asselman, C. Verhamme, A. de Luca, J. Hendrikse, W.L. van der Pol, M. Froeling, Quantitative magnetic resonance imaging of the brachial plexus shows specific changes in nerve architecture in chronic inflammatory demyelinating polyneuropathy, multifocal motor neuropathy and motor neuron disease, Eur. J. Neurol. 28 (8) (2021) 2716–2726, https://doi.org/10.1111/ene.14896.
[62] A. Uncini, S. Kuwabara, Nodopathies of the peripheral nerve: an emerging concept, J. Neurol. Neurosurg. Psychiatry 86 (11) (2015) 1186–1195, https://doi.org/10.1136/jnnp-2014-310097.
[63] K. Doppler, L. Appeltshauser, C. Villmann, C. Martin, E. Peles, H.H. Krämer, A. Haarmann, M. Buttmann, C. Sommer, Auto-antibodies to contactin-associated protein 1 (Caspr) in two patients with painful inflammatory neuropathy, Brain. 139 (Pt 10) (2016) 2617–2630, https://doi.org/10.1093/brain/aww189.
[64] L. Querol, R. Rojas-García, J. Diaz-Manera, J. Barcena, J. Pardo, A. Ortega-Moreno, M.J. Sedano, L. Seró-Ballesteros, A. Carvajal, N. Ortiz, E. Gallardo, I. Illa, Rituximab in treatment-resistant CIDP with antibodies against paranodal proteins, Neurol Neuroimmunol Neuroinflamm. 2 (5) (2015), e149, https://doi.org/10.1212/nxi.0000000000000149.
[65] C. Painous, M. López-Pérez, I. Illa, L. Querol, Head and voice tremor improving with immunotherapy in an anti-NF155 positive CIDP patient, Ann Clin Transl Neurol. 5 (4) (2018) 499–501, https://doi.org/10.1002/acn3.539.
[66] G. Liberatore, C. Giannotta, B.P. Sajeev, E. Morenghi, F. Terenghi, F. Gallia, P. E. Doneddu, F. Manganelli, D. Cocito, M. Filosto, G. Antonini, G. Cosentino, G. A. Marfia, A.M. Clerici, G. Lauria, T. Rosso, G. Cavaletti, E. Nobile-Orazio, Sensitivity and specificity of a commercial ELISA test for anti-MAG antibodies in patients with neuropathy, J. Neuroimmunol. 345 (2020), 577288, https://doi.org/10.1016/j.jneuroim.2020.577288.
[67] C. Sommer, S. Koch, M. Lammens, A. Gabreels-Festen, G. Stoll, K.V. Toyka, Macrophage clustering as a diagnostic marker in sural nerve biopsies of patients with CIDP, Neurology. 65 (12) (2005) 1924–1929, https://doi.org/10.1212/01.wnl.0000188879.19900.b7.
[68] G.B. Kulkarni, A. Mahadevan, A.B. Taly, A. Nalini, S.K. Shankar, Sural nerve biopsy in chronic inflammatory demyelinating polyneuropathy: are supportive pathologic criteria useful in diagnosis? Neurol. India 58 (4) (2010) 542–548, https://doi.org/10.4103/0028-3886.68673.
[69] R.U. Haq, T.J. Fries, W.W. Pendlebury, M.J. Kenny, G.J. Badger, R. Tandan, Chronic inflammatory demyelinating polyradiculoneuropathy: a study of proposed electrodiagnostic and histologic criteria, Arch. Neurol. 57 (12) (2000) 1745–1750, https://doi.org/10.1001/archneur.57.12.1745.
[70] D.S. Molenaar, M. Vermeulen, R. de Haan, Diagnostic value of sural nerve biopsy in chronic inflammatory demyelinating polyneuropathy, J. Neurol. Neurosurg. Psychiatry 64 (1) (1998) 84–89, https://doi.org/10.1136/jnnp.64.1.84.
[71] 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 (1) (2017) Cd010369, https://doi.org/10.1002/14651858.CD010369.pub2.
[72] A. Kaplan, T.H. Brannagan 3rd, Evaluation of patients with refractory chronic inflammatory demyelinating polyneuropathy, Muscle Nerve 55 (4) (2017) 476–482, https://doi.org/10.1002/mus.25271.
[73] J.A. Allen, J. Ney, R.A. Lewis, Electrodiagnostic errors contribute to chronic inflammatory demyelinating polyneuropathy misdiagnosis, Muscle Nerve 57 (4) (2018) 542–549, https://doi.org/10.1002/mus.25997.
[74] M.C. Broers, C. Bunschoten, J. Drenthen, T.A.O. Beck, E. Brusse, H.F. Lingsma, J. A. Allen, R.A. Lewis, P.A. van Doorn, B.C. Jacobs, Misdiagnosis and diagnostic pitfalls of chronic inflammatory demyelinating polyradiculoneuropathy, Eur. J. Neurol. 28 (6) (2021) 2065–2073, https://doi.org/10.1111/ene.14796.
[75] J.A. Allen, The misdiagnosis of CIDP: a review, Neurol Ther. 9 (1) (2020) 43–54, https://doi.org/10.1007/s40120-020-00184-6.
[76] S.I. van Nes, E.K. Vanhoutte, P.A. van Doorn, M. Hermans, M. Bakkers, K. Kuitwaard, C.G. Faber, I.S. Merkies, Rasch-built Overall Disability Scale (R-ODS) for immune-mediated peripheral neuropathies, Neurology. 76 (4) (2011) 337–345, https://doi.org/10.1212/WNL.0b013e318208824b.
[77] I.N. van Schaik, V. Bril, N. van Geloven, H.P. Hartung, R.A. Lewis, G. Sobue, J. P. Lawo, M. Praus, O. Mielke, B.L. Durn, D.R. Cornblath, I.S.J. Merkies, Subcutaneous immunoglobulin for maintenance treatment in chronic inflammatory demyelinating polyneuropathy (PATH): a randomised, double-blind, placebo-controlled, phase 3 trial, Lancet Neurol. 17 (1) (2018) 35–46, https://doi.org/10.1016/s1474-4422(17)30378-2.
[78] Y.A. Rajabally, M. Ghasemi, Comparative value and determinants of suitability of outcome measures in treated chronic inflammatory demyelinating polyneuropathy, Muscle Nerve 61 (2) (2020) 182–186, https://doi.org/10.1002/mus.26747.
[79] I.S. Merkies, P.I. Schmitz, F.G. van der Meché, J.P. Samijn, P.A. van Doorn, Clinimetric evaluation of a new overall disability scale in immune mediated polyneuropathies, J. Neurol. Neurosurg. Psychiatry 72 (5) (2002) 596–601, https://doi.org/10.1136/jnnp.72.5.596.
[80] I.S. Merkies, S.I. van Nes, K. Hanna, R.A. Hughes, C. Deng, Confirming the efficacy of intravenous immunoglobulin in CIDP through minimum clinically important differences: shifting from statistical significance to clinical relevance, J. Neurol. Neurosurg. Psychiatry 81 (11) (2010) 1194–1199, https://doi.org/10.1136/jnnp.2009.194324.
[81] Medical Research Council, Aids to the Investigation of the Peripheral Nervous System, Her Majesty's Stationary Office, London, 1943.
[82] I.S. Merkies, P.I. Schmitz, F.G. van der Meché, P.A. van Doorn, Psychometric evaluation of a new sensory scale in immune-mediated polyneuropathies. Inflammatory neuropathy cause and treatment (INCAT) group, Neurology. 54 (4) (2000) 943–949, https://doi.org/10.1212/wnl.54.4.943.
[83] P.J. Dyck, C.J. Boes, D. Mulder, C. Millikan, A.J. Windebank, P.J. Dyck, R. Espinosa, History of standard scoring, notation, and summation of neuromuscular signs. A current survey and recommendation, J Peripher Nerv Syst. 10 (2) (2005) 158–173, https://doi.org/10.1111/j.1085-9489.2005.0010206.x.
[84] J.A. Allen, M. Pasnoor, M.M. Dimachkie, S. Ajroud-Driss, T.H. Brannagan, A. A. Cook, T. Walton, M.B. Fiecas, J.T. Kissel, I. Merkies, K.C. Gorson, R.A. Lewis, Quantifying treatment-related fluctuations in CIDP: results of the GRIPPER study, Neurology. 96 (14) (2021) e1876–e1886, https://doi.org/10.1212/wnl.0000000000011703.
[85] P.E. Doneddu, D. Mandia, F. Gentile, F. Gallia, G. Liberatore, F. Terenghi, M. Ruiz, E. Nobile-Orazio, Home monitoring of maintenance intravenous immunoglobulin therapy in patients with chronic inflammatory neuropathy, J. Peripher. Nerv. Syst. 25 (3) (2020) 238–246, https://doi.org/10.1111/jns.12396.
[86] E.K. Vanhoutte, N. Latov, C. Deng, K. Hanna, R.A. Hughes, V. Bril, M.C. Dalakas, P. Donofrio, P.A. van Doorn, H.P. Hartung, I.S. Merkies, Vigorimeter grip strength in CIDP: a responsive tool that rapidly measures the effect of IVIG–the ICE study, Eur. J. Neurol. 20 (5) (2013) 748–755, https://doi.org/10.1111/j.1468-1331.2012.03851.x.