# Chronic Inflammatory Demyelinating Polyneuropathy

## Updates in diagnosis and management.

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Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an immune-mediated neuropathy syndrome comprising gradually progressive proximal and distal weakness, large-fiber sensory disturbances, areflexia, and features of acquired demyelination (eg, slowed conduction velocities, conduction block, temporal dispersion, and onion-bulb formation).[1,2] CIDP likely represents a heterogeneous group with similar clinical features; however, the term remains useful because of similarities in confirmatory diagnostic features, shared inflammatory demyelination pathophysiology, and responsiveness to immune therapy. The lack of a single specific biomarker, presence of several mimickers and chameleons, and pressure on healthcare providers to find treatable causes of disability increase the difficulty of diagnosing and managing CIDP and its variants. These factors, among others, have led to the paradox of simultaneous 'overdiagnosis'[3] and 'underdiagnosis'[4] of CIDP. Success in managing CIDP may depend on individual patient factors and type of CIDP and often requires a trial of immunotherapy using objective treatment endpoints. The purpose of this review is to enhance and update the reader's ability to diagnose and manage CIDP and its variants with recent and practical evidence-based principles.

## Diagnosis

### Typical Presentation, History, and Neurologic Examination

CIDP should be suspected when a person has features of disrupted myelinated nerve fiber function. In typical CIDP, fibers with the most myelin are preferentially affected earliest and most severely. Dysfunction of these fibers evolves over at least 8 weeks, by definition, either progressively or in a relapsing-remitting fashion. Because motor (type 1a/Aδ), Golgi tendon organ (Ib/Aδ), and large, myelinated sensory fibers (type II/ Aδ) have the most myelin, people with classic CIDP present with symmetric weakness, areflexia, and loss of vibration, proprioception, touch, and muscle spindle sensations. Fiber caliber, and therefore, amount of myelin, is greater in proximal segments, resulting in both proximal and distal weakness (polyradiculoneuropathy), although distal symptoms may be more severe because all distal nerve signals must also pass through the proximal segments. Sparing of pain and temperature sensation (types III/Aδ and IV/C fibers) and autonomic fibers is classic, because these are thinly myelinated or unmyelinated.[5]

Chronic, progressive or relapsing-remitting, proximal and distal large-fiber symptoms with small-fiber sparing should raise CIDP as a diagnostic consideration after an initial clinical encounter. These features can be gleaned mostly from a clinical history that may include difficulty walking or climbing stairs, imbalance (particularly with eyes closed in the shower or in dark environments), and "dead" or prickling numbness of the limbs. Pain, especially when described as "burning," is considered atypical and raises caution against a diagnosis of typical CIDP. In practice, neuropathic pain is present in a substantial minority of persons with typical CIDP (20%-41% in a single-center study[6]) but occurs later in the disease course, usually long after large-fiber impairment. CIDP should not be the first diagnosis to consider if there are only distal, painful, sensory-predominant symptoms without weakness, areflexia, or large-fiber dysfunction. Generally, systemic symptoms (eg, rash, weight loss, and cardiorespiratory or gastrointestinal symptoms) should also dissuade a clinician from CIDP diagnosis.

There are no uniquely specific neurologic physical examination signs or laboratory findings in CIDP, although hyporeflexia or areflexia should be confirmed. Palpable, hypertrophic nerves rarely occur, but may also be present in hereditary demyelinating neuropathies. The examination and laboratory studies should be used to investigate for systemic signs and symptoms, mostly to exclude mimics. Assessment of monoclonal proteins, especially lambda, may be useful to identify polyneuropathy, organomegaly, endocrinopathy, M-protein, and skin changes (POEMS) syndrome or IgM- monoclonal gammopathy of undetermined significance (MGUS)-associated distal acquired demyelinating symmetric and sensory (DADS) neuropathy. Serum testing for antibodies against nodal or paranodal epitopes should be considered in some cases.

### CIDP Variants

CIDP variants share a common pathophysiology of inflam-

matory demyelination with repeated demyelination and remyelination resulting in stacks of Schwann cell processes (ie, onion-bulb formation) and a responsiveness to immune therapy with typical CIDP. Otherwise, variants defy the typical clinical syndrome of symmetrical proximal and distal weakness (polyradiculoneuropathy), areflexia, and sensory loss in some particular respect. Although variants seem to be more rare than typical CIDP, 84 of 460 individuals with confirmed CIDP in a recent series initially had atypical presentations, with substantial proportions later developing typical CIDP.[7]

The number of CIDP variants is daunting and continues expanding (Table e1). CIDP variants should enter the neurologist's mind when 1) signs and symptoms are specifically localized to a select population of large, myelinated nerve fibers or roots (usually motor or sensory proprioceptive fibers); 2) there is relative sparing of thinly myelinated or unmyelinated nerves; 3) upper motor neuron features are lacking; and 4) other common causes of the clinical phenotype have been excluded. Although clinical features vary considerably, the unifying feature of these variants is inflammatory demyelination. Acute CIDP has ongoing active disease that requires ongoing immunotherapy.[8] Multifocal CIDP has typical demyelinating features confined to localized body segments, and is often upper limb predominant.[9] Sensory CIDP has demyelinating features on both motor and sensory electrodiagnostic (EDX) testing,[10] whereas motor CIDP has no measurable electroclinical sensory involvement.[11] Chronic immune sensory polyradiculopathy (CISP) has normal nerve conduction, and CISP-plus has neurophysiologic findings discordant with observed clinical features; in both, sensory nerve roots are predominantly affected.[12,13] Chronic immune sensory and motor polyradiculopathy (CISMP) predominantly involves nerve roots and spares distal nerves.[14]

### Practical Review of EDX Criteria

The European Academy of Neurology (EAN)/Peripheral Nerve Society (PNS) 2010 clinical diagnostic and EDX guidelines[2] are the most widely applied for CIDP and CIDP variant diagnosis (Table e1). These criteria are sensitive (73%-91%) and specific (66%-88%) in comparison to other available criteria,[15] and were most recently revised in June 2021.[16] This second revision emphasizes diagnosis of variants (no longer calling them atypical) and narrows diagnostic categories to CIDP or possible CIDP (probable CIDP was removed). As with all diagnostic criteria, there are limitations that can lead to underdiagnosis (false negatives) or overdiagnosis (false positives). To avoid either, the first and most important priority is to apply these criteria only in the appropriate clinical scenario. Secondly, application of these EDX criteria in people with mixed processes in which the severity of axonal features exceeds demyelinating features (ie, low compound motor action potential [CMAP] and sensory nerve action potential [SNAP] amplitudes) should be approached with extreme caution. Slowed conduction velocity and prolonged distal latency occur with axonal destruction in any neuropathy because of loss of the fastest-firing large nerve fibers and secondary segmental demyelination.[17] Lastly, the CIDP variants may be more challenging to correctly diagnose because these do not conform to the clinical or EDX patterns of typical CIDP, relying on smaller number or limited populations of involved nerves. Other supportive tests (eg, cerebrospinal fluid [CSF] analysis, nerve imaging, somatosensory evoked potentials [SEPs], response to treatment, or nerve biopsy) can increase confidence in diagnosis of CIDP variants.

## Misdiagnosis of CIDP

### Avoiding Overdiagnosis

The most likely cause of CIDP overdiagnosis is overinterpretation of EDX findings. A recent study at an academic referral center estimated that only 0.34% of patients who met EDX criteria for CIDP presented with a distal symmetric neuropathy phenotype.[18] Despite this, a substantial proportion of those referred to an academic center for refractory CIDP had a distal symmetric polyneuropathy phenotype and EDX findings attributable to other common etiologies (eg, nerve compression) rather than demyelination.[3] For others, features of axon loss (eg, conduction slowing) had been overemphasized.[3] The practical point is to ensure methods of nerve conduction studies (NCS) are performed properly at the required limb temperature, and rely only on unequivocal evidence of demyelination to confirm CIDP. When demyelinating features are called partial, borderline, possible, or otherwise do not completely fulfill criteria, other diagnoses should be considered.

False-positive EDX criteria for CIDP may truly occur when unequivocal EDX demyelination is present but caused by another demyelinating disorder. This may occur in conditions in which axon degeneration and demyelination coexist. Electroclinical mimics of CIDP that may fulfill EDX criteria for CIDP include POEMS syndrome, IgM-monoclonal gammopathy-associated neuropathy with or without myelin-associated glycoprotein (MAG) antibodies, hereditary or acquired amyloidosis (see *Neuromuscular Amyloidosis* in this issue), multifocal motor neuropathy with conduction block (MMN-CB), neurolymphomatosis, severe diabetic polyneuropathies, and hereditary demyelinating neuropathies (eg, Charcot-Marie-Tooth type 1). In our clinics, POEMS syndrome is often the most likely mimic in incorrectly diagnosed CIDP that has not responded to immune therapies, because both CIDP and POEMS present with progressive demyelinating neuropathies.[19] The presence of pain, thrombocytosis, or a monoclonal protein should increase suspicion of and initiate a search for an osteosclerotic myeloma.

### Avoiding Underdiagnosis

Overdiagnosis of CIDP has been emphasized recently because it leads to misuse of immune therapies with high costs

and risks. Underdiagnosis also occurs, and more than two-thirds of consecutive patients referred to a tertiary specialist clinic for a nonCIDP diagnosis met clinical and EDX criteria for CIDP.[4] The most common diagnostic chameleon in this group was Guillain-Barré syndrome (GBS), an understandable finding considering the similarities between GBS and CIDP, and potential need for follow-up with a neuromuscular disease specialist after hospital discharge. Underdiagnoses have also been attributed to 1) atypical CIDP variants (eg, multifocal CIDP, diabetic polyneuropathy or radiculoplexus neuropathy) in which demyelinating features were missed or called axonal; 2) satisfaction-of-search errors in which hereditary neuropathy and CIDP co-occurred; and 3) other circumstances in which proximal weakness was overlooked and laboratory findings (eg, Lyme antibodies) were overemphasized.[4] These findings highlight the need to closely link the clinical scenario with the EDX findings, which is helped by obtaining the history and examination at the same time as EMG and NCS. Reliable communication of neurophysiologic test results is also essential. Other atypical CIDP variants such as CISP, CISP-plus, and CISMP do not have EDX findings of demyelination on routine testing and may be missed. Clinical suspicion for these diagnoses and special evaluations of SEPs, CSF analysis, MRI of proximal nerves, or nerve pathology are needed to identify these variants.[12-14] If uncertainty exists about CIDP variants or nonreponse to treatment, neuromuscular specialist referral may be required.

### Future Directions for Diagnosis

As previously mentioned, the coexistence of severe axon loss and demyelinating findings may cause a floor effect in which most motor and sensory responses are absent. When this floor effect is reached, a demyelinating neuropathy cannot be identified and diagnosis is challenging. An R1 latency of the blink response longer than 13 ms has been shown to be a reliable sign of demyelination even in the setting of low or unobtainable ulnar CMAP amplitudes due to secondary axon loss. R1 latency improvement after treatment correlated to clinical improvement measured by neuropathy impairment score (NIS).[20] This finding is not specific to CIDP but may assist with evaluation for the presence and response to treatment of a demyelinating process with severe secondary axonal loss.

The role of CSF protein levels in CIDP diagnosis has come into question since the 2010 criteria were published, after overreliance on elevated CSF protein was identified as a major contributor to CIDP overdiagnosis.[3] A systematic review of over 20 high-quality studies published between 1960 and 2017 in which CSF protein was evaluated in people with and without neurologic illness showed an inelastic upper limit of 45 mg/dL should be replaced with an age-adjusted upper limit of 60 mg/dL at age 50 or more. Physicians also need to understand that elevated CSF protein levels indicate involvement of proximal nerve segments or roots that are not specific to CIDP but also occur in inherited neuropathy, radiculoplexus neuropathies, POEMS syndrome, sarcoidosis, and other conditions.

A new wave of discovery has begun after the initial recognition and continued investigation of IgG4 antibodies directed toward epitopes in neurofascin 155 (NF-155), contactin-1, and contactin-associated protein-1 (CASPR1), which all localize adjacent to or at the nodes of Ranvier. Testing for these antibodies in the appropriate clinical setting (Table e1) has become increasingly useful to inform prognosis and guide therapeutic choices. Involvement of these antibodies may be recognized by the presence of prominent sensory features with ataxia, upper limb onset or predominance, very high CSF protein levels, and the presence of tremor in the case of anti-NF-155. Early evidence suggests conventional therapies may be less effective in these conditions, whereas rituximab or other immunoglobulin-depleting therapy may be more effective.[21,22]

## Management

Proven treatments and standards of care for typical CIDP and most variants are intravenous immune globulin (IVIG), corticosteroids, and plasmapheresis. These have remained the best available options for over 20 years. Novel immune therapies and modifications to existing first-line agents are being examined, however. There is also an established role for physiotherapy and occupational therapy in treatment of CIDP.

### Therapy Targets

Goals of immune therapy should be established before starting medication. We counsel patients that stabilization is the first goal, and that improvement may follow stabilization. Setting realistic expectations is especially important when longstanding, untreated disease results in axon loss that may take months to years to recover or may never recover at all. We also inform patients that although several treatment regimens have been shown effective for people with CIDP as a group, each individual will require an individualized dose and duration of treatment to induce remission. An individualized approach may initially seem taxing on finances and time, but likely saves funds and improves outcomes compared to uniform application of standard dosing protocols.[23] Several reliable outcome measures in CIDP have been demonstrated and used in research studies.[24,25] In routine clinical practice, treating providers should find methods that can be easily and systematically used in their office during a standard follow-up visit, such as a combination of patient-questionnaire inventories (eg, Dyck score, inflammatory Rasch-built overall disability Scale [I-RODS], overall neuropathy limitations scale [ONLS]), manual motor testing or dynamometry, combined measures (neuropathy impairment score), or periodic repeat EDX testing. The minimum clinically important difference (MCID) for each measure may be set as a target for therapy, and if improvement is not achieved or measures show worsening, this can serve as a directive for changing therapy.

**TABLE. EAN/PNS ELECTRODIAGNOSTIC GUIDELINE ON CIDP–2ND REVISION 2021 CHECKLIST[a]**

| Criterion | | Definition | | Nerves | | | | Total |
|---|---|---|---|---|---|---|---|---|
| | | | | Median | Ulnar | Fibular | Tibial | |
| A | Distal latency | Prolonged ≥50% (1.5x) of UL | | Exclude CTS | | | | |
| B | Conduction velocity | Reduced ≥30% (0.7x) of lower limit | | | Exclude UNE andMGA | | | |
| C | F-wave latency | Normal CMAP amplitude | Prolonged ≥30% (1.3x) of UL | | | | | |
| | | Reduced (≥20%) CMAP amplitude | Prolonged ≥50% (1.5x) of UL | | | | | |
| D | F-Wave absence | Absence with CMAP amplitude ≥20% of lower limit | | | | | | |
| E | Partial motor conduction block | Proximal CMAP amplitude ≥50% Reduced compared to distal CMAP amplitude | | | | | b | |
| F | Abnormal temporal dispersion | CMAP duration increases >30% between proximal and distal sites | | | | | | |
| G | Distal CMAP duration | Prolonged interval between initial onset and baseline return of last negative peak | | >6.5 ms | >6.6 ms | >7.5 ms | >8.7 ms | |

Abbreviations: CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; CMAP, compound muscle action potential; CTS, carpal tunnel syndrome; EAN, European Academy of Neurology; PNS, Peripheral Nerve Society; UNE, ulnar neuropathy at elbow; MGA, Martin-Gruber (median-ulnar) anastomosis; UL, upper limit.

[a]Second revision provided via personal communication from Dr. Peter Van Den Bergh. These criteria must only be applied for patients who meet clinical criteria for typical CIDP: symmetric proximal and distal weakness in 4 limbs and sensory disturbance in 2 or more limbs. Temperature must be maintained at 33° C at the palm and 30° C at the lateral malleolus. Tally the box for each confirmed criterion in the corresponding nerve, then sum them in the total column. Typical CIDP is diagnosed if there are ≥2 points in rows A, B, C, E, or F or ≥1 point in row D or G plus 1 point in any other row. Possible CIDP is diagnosed if clinical criteria for CIDP are met, there are 2 supportive criteria from cerebrospinal fluid analysis, nerve imaging with ultrasound or MRI, response to treatment, or a positive nerve biopsy, and ≥1 point in row E and ≥2 points in rows A and B or reduced sensory nerve action potential (SNAP). [b] Tibial conduction block should not routinely be relied upon for diagnosis because of the presence of 50% depression in normal individuals.

We recommend seeing patients after a prespecified treatment period—often 3 months. During this follow-up visit, we evaluate with the prespecified measure first and then take the clinical history to remain as objective as possible.

### Standard Immune Therapies

*IVIG.* The use of IVIG as a first-line therapy for CIDP is supported by high-quality data from multiple randomized controlled trials. It is likely as effective as plasmapheresis, but better tolerated by many.[26,27] A clinical trial established that IVIG dosing of 1 g/kg over 1 to 2 days every 3 weeks for up to 24 weeks is more effective than placebo for improving disability and grip strength and increasing time between relapses.[28] This trial provides a reasonable regimen to use when starting immune therapy.

Some persons with CIDP may not respond to regimens of 3- to 4-week dosing intervals or may have improvement followed by wearing off of benefits during these intervals. Often these individuals improve with lower, more frequent doses. This more frequent dosing approach was used in a controlled trial of IVIG vs. plasmapheresis.[27] Consequently, when beginning IVIG treatment of newly diagnosed persons with CIDP or when treating refractory CIDP, we may prescribe doses of 0.4 g/kg once or, in severe cases, twice weekly, for short durations (eg, 4-8 weeks), and then weekly thereafter. This is done alone or sometimes in combination with intravenous methylprednisolone (IVMP) 1,000 mg. Reassessment for improvement should still be done at approximately 3-month intervals because earlier reassessment may miss subtle improvements. If there is improvement with these intensive dosing regimens, we maintain or scale down the IVIG dose slowly. If no treatment response is noted, alternative diagnoses are considered.[24] Use of combined IVIG and IVMP for treatment induction is currently being investigated in a large randomized controlled trial,[29] and was well-tolerated in a recent pilot study.[30]

Adjustment of IVIG can be difficult and sometimes patients cannot be easily weaned from a weekly to biweekly schedule because of worsening symptoms and neurologic deficits. In such cases, a 1.5-week protocol can be achieved with an alternating dosing interval to maintain infusions consistently on the

same day of the week (eg, scheduling infusions every 10th day then every 11th day, on an alternating basis of Monday the first week and Thursdays the following week or something similar).

Subcutaneous immune globulin (SCIG) is a relatively new means of administering immunoglobulin therapy that was effective and tolerable in a large randomized controlled trial with a long-term extension beyond 24 weeks.[31] There are 2 published doses (0.4 g/kg and 0.2 g/kg), which were apparently equally effective in the initial trial. In the extension study, however, approximately half of those who had dose reduction from 0.4 g/kg to 0.2 g/kg experienced relapse.

We determine immunoglobulin therapy duration by clinical response, attempting to slowly reduce it as individuals reach a plateau or have sequential improvements over a 6- to 8-month period. Tapering is done by decreasing frequency of dosing by 1 week at a time (eg, every 1 week to every 2 weeks) over 3-month periods to allow for a new steady state to be reached. If objective worsening occurs as therapy is tapered, we return to the most recent previously successful dosing.

Adverse events with IVIG are uncommon, but include thrombotic events, renal injury, headache/chemical meningitis, and allergic reactions. Dosing should be adjusted in persons with pre-existing renal disease, and premedication may be considered to avoid some unwanted effects.

*Corticosteroids.* Corticosteroids remain useful immunotherapy in CIDP because of their low cost, widespread availability, and convenience of use and dosing. Corticosteroids are effective, although the data supporting efficacy is not as strong as for immunoglobulin therapy,[26] and long-term adverse events occur. High-dose monthly oral dexamethasone produced fewer adverse events than daily use in a comparison to daily prednisolone and was no less effective.[26] Periodic intravenous methylprednisolone with dosing adjusted to clinical response is equally effective and produced fewer adverse effects than daily oral prednisone in a small, single-center retrospective study.[32]

*Plasmapheresis.* Plasmapheresis is well-established and effective immunotherapy in CIDP with the relative advantage of working quickly. Plasmapheresis may be more useful in those with very severe disability.[26,27,33] We have had personal experience of plasmapheresis being very effective in treatment of ataxia and tremor in some cases of recently described variants caused by antiNF-155, antiCASPR-1, and anticontactin-1—the nodopathies (NF-155/CASPR-1/contactin-1) that may be otherwise refractory to immunoglobulins and corticosteroids.

### Future Directions in Immune Therapy

In a 3-center retrospective study of 11 people with probable or definite CIDP who did not respond to conventional immune therapies, 9 of 11 had improved Inflammatory Neuropathy Cause and Treatment (INCAT) disability scores and gait function after treatment with rituximab. In these individuals, first signs of improvement occurred within 3 months of rituximab initiation, and maximum improvement occurred at 2 to 18 months.[22] Only 3 of the 11 individuals were tested for NF-155 antibodies, which were found in 1 of those 3 people. In a retrospective study of 200 people who had chronic immune neuropathy, 48 had typical or atypical CIDP, or paraproteinemic demyelinating neuropathy with or without nodal/paranodal or MAG antibodies. These individual's conditions had been refractory to conventional immune therapies, but 85.4% of them who received rituximab, cyclophosphamide, or bortezomib as monotherapy or in combination responded to treatment. The INCAT-overall disability sum scores (ODSS) after these therapies were higher than in people with similar conditions who received different agents used as second-, third-, or fourth-line therapies.[34] These studies, and others, suggest the need for new clinical trials to evaluate these immune therapies, and in routine clinical practice, provide some basis for trial in those with confirmed CIDP refractory to treatment.

## Key Points

- CIDP is an inflammatory demelinating neuropathy; as such, damage is predominantly to large myelinated fibers. As a result, CIDP usually presents with muscle weakness and sensory ataxia, and rarely with pain or autonomic findings.
- CIDP overdiagnosis is common and often related to over-interpretating EDX findings as demyelination or overrelying on mildly elevated CSF protein and other non-specific findings seen in mimicking disorders.
- Even when demyelination is confirmed with EDX, non-response to treatment warrants a search for alternative diagnoses with POEMS syndrome at the top of the list.
- Co-existing axon loss and demyelination may occur in long-standing CIDP and makes EDX testing more challenging, but the R1 latency may be useful to establish the presence of demyelinating disease in this scenario.
- An objective target for immune therapy should be established before initiating treatment for CIDP to guide therapy. Dosing should be individualized based on such targets.
- The mainstays of therapy are immunoglobulins, corticosteroids, or plasmapheresis, but novel emerging immune therapies may be used in refractory cases as further high-quality evidence is obtained. ■

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**Disclosures**
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**TABLE e1. CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY VARIANTS**

| Variant | Clinical presentation | Location of pathology | Differential diagnosis |
|---|---|---|---|
| Acute (A-)[8] or subacute (S-)[35] onset chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) | Rapid <8 weeks (A-CIDP) or 4-8 weeks (S-CIDP); proprioceptive loss and ataxia more than in Guillain-Barré syndrome (GBS)[8] | Proximal and distal nerves and nerve roots as in typical CIDP | GBS, acute myelopathy |
| Multifocal CIDP (Lewis-Sumner syndrome, MADSAM)[7,9] | Asymmetric, relapsing, or progressive, more often sensorimotor than sensory-only, and multiple mononeuropathy | Sensory and motor nerves >nerve root involvement, with patchy presentation, upper/lower limbs equally affected | Systemic small vessel vasculitides, hereditary liability to pressure nerve palsy (HNPP), cervical or lumbosacral radiculoplexus neuropathy, perineurioma |
| Sensory CIDP[10,36] | Progressive sensory loss with preserved strength, electrophysiology shows subclinical motor involvement that is nonlength dependent, and two-thirds progress to typical CIDP within a mean 5 years[7] | Postganglionic sensory nerves and motor nerves with more upper than lower limb involvement | Myeloneuropathy (dorsal columns), sensory ganglionopathy |
| Motor CIDP[2,7,11] | Progressive proximal and distal weakness with preserved sensation and sensory sparing on electrophysiology | Proximal and distal motor nerve fibers lower>upper limb involvement | Multifocal motor neuropathy with conduction blocks (MMN-CB), motor neuron disease (MND) |
| Chronic immune sensory polyradiculopathy (CISP)[13] | Sensory ataxia with normal strength, EMG, and nerve conduction studies (NCS) but abnormal sensory evoked potentials (SEPs) | Preganglionic sensory roots | Myeloneuropathy, sensory ganglionopathy |
| Chronic immune sensory polyradiculopathy plus (CISP-plus)[12] | Sensory ataxia with minimal distal "numbness" or weakness that may appear to have mild, distal axonal EMG/NCS findings and abnormal SEPs | Predominantly preganglionic sensory roots with some involvement of postganglionic segments | May have electrodiagnostic appearance of axonal distal symmetric polyneuropathy, paraneoplastic or metabolic myeloneuropathies |
| Chronic immune sensory and motor polyradiculopathy (CISMP)[14] | Proximal weakness and sensory loss with hyporeflexia or areflexia and lower>upper limb involvement | Root level inflammatory demyelination that spares distal segments | Infiltrative, neoplastic, or structural polyradiculopathy |

36. Rajabally YA, Wong SL. Chronic inflammatory pure sensory polyradiculoneuropathy: a rare CIDP variant with unusual electrophysiology. *J Clin Neuromusc Dis*. 2012;13(3):149-152.