BMJ Best Practice

Chronic inflammatory demyelinating polyradiculoneuropathy

Straight to the point of care

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Table of Contents

Summary

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an acquired demyelinating peripheral neuropathy of presumed autoimmune aetiology.

Proximal and distal symmetrical weakness (often without pain) is typical of CIDP. Symptoms of weakness and sensory loss progress beyond 8 weeks. CIDP variants ('atypical' CIDP) can result in proximal or distal asymmetrical weakness with sensory loss, predominantly distal weakness with sensory loss, pure motor symptoms, or pure sensory symptoms, sometimes associated with ataxia.

Diagnosis is based on a combination of clinical history, symptoms, examination findings, and electrodiagnostic testing. However, a treatment trial should not be withheld if electrodiagnostic criteria are not met, if history, symptoms, and clinical examination findings are consistent with the diagnosis.

Initial treatment is with intravenous immunoglobulin, corticosteroids, or plasma exchange. If there is insufficient response to first-line treatment, an alternative agent should be tried, followed by combination therapy. Lack of at least a partial response to one or two first-line agents should lead to re-evaluation of the diagnosis; an alternative immunosuppressant may be added to treat confirmed refractory CIDP. Long-term immunosuppressive therapy may be needed to prevent relapse.

Differentiation from Guillain-Barre syndrome (GBS) is important, because treatment and management are different for the two conditions, and corticosteroids may worsen GBS.

Definition

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an acquired demyelinating peripheral neuropathy of presumed immune-related aetiology. The course is usually either chronic progressive (>8 weeks), relapsing and remitting, or stepwise. The clinical phenotype is typically characterised by symmetrical proximal and distal weakness, distal large-fibre sensory loss, and absent or reduced reflexes.

THEORY

Epidemiology

One systematic review provided an estimated pooled incidence rate for CIDP of 0.33 per 100,000 person-years, and an estimated pooled prevalence rate of 2.81 per 100,000 people. There is substantial heterogeneity in incidence and prevalence across individual studies, due in part to use of different diagnostic criteria and under-recognition of CIDP variants.[2] [15] [16]

CIDP can occur at any age, with most common age of onset being between 40 and 60 years.[1] [5] CIDP is more common in men, with reported male-to-female ratios of between 1.4 and 4.4.[1] [2] [15] There are no known differences between ethnic groups or geographical regions.

Aetiology

There are no clearly defined genetic or environmental risk factors.[2] Infection, immunisation, or surgical procedures precede CIDP in approximately 10% to 30% of patients.[4] [6][17] There is an increased incidence of CIDP in patients with diabetes mellitus, and severity of CIDP may be worse in these patients.[18] [19] [20] CIDP can also occur in association with infection, including HIV.[19] [20] About 10% to 15% of patients have a benign monoclonal gammopathy of undetermined significance (MGUS), most often with an IgA or IgG paraprotein; MGUS may affect disease course and response to treatment.[2] [19] [20] CIDP may also co-exist with other autoimmune diseases, such as systemic lupus erythematosus, connective tissue disease, inflammatory bowel disease, thyroid disease, sarcoidosis, glomerulonephritis, chronic active hepatitis, or graft-versus-host disease.[19] [20]

Pathophysiology

Evidence suggests that CIDP is an autoimmune disease directed against peripheral nerve myelin. Nerve biopsies from patients with CIDP suggest underlying cell-mediated immunity as a mechanism of disease. Endoneurial infiltrates consist mainly of CD4+ and CD8+ T cells and macrophages.[21] [22] Immunoglobulin and complement have been immunolocalised to myelin sheaths of affected nerves, and a variety of cytokines, chemokines, co-stimulatory molecules, adhesion molecules, growth factors, and transcription factors are up-regulated in biopsies from patients with CIDP.[2] Humoral immunity also probably plays a role (especially given the discovery of disease mediated by antiparanodal and antinodal antibodies), but auto-antibodies directed against myelin proteins and lipids, such as gangliosides, sulfatides, galactocerebroside, and myelin protein zero, have been found in only a minority of patients.[2]

Classification

European Academy of Neurology/Peripheral Nerve Society (EAN/PNS)[1]

The 2021 EAN/PNS guideline defines two levels of diagnostic certainty for CIDP:[1]

- CIDP
- Possible CIDP.

The most common presentation is typical CIDP.[1] [2]

CIDP variants are:[1]

- Distal CIDP (also known as distal acquired demyelinating symmetrical neuropathy)
- Multifocal CIDP (also known as multifocal demyelinating neuropathy with persistent conduction block, Lewis-Sumner syndrome; multifocal acquired demyelinating sensory and motor neuropathy [MADSAM]; multifocal inflammatory demyelinating neuropathy)
- Focal CIDP
- Motor CIDP
- Motor-predominant CIDP
- Sensory CIDP
- Sensory-predominant CIDP.

See Diagnostic criteria .

Case history

Case history #1

A 55-year-old man presents with a 4-month history of numbness in both feet and hands. Soon after this numbness, he developed symmetrical leg and arm weakness. The weakness progressed such that on the day of admission he could not lift himself up from the commode or raise his arms over his head. He notes mild shortness of breath, but no bowel or bladder incontinence, dysarthria, dysphagia, or diplopia. General examination is normal. Neurological examination shows normal mental status and cranial nerves. He has symmetrical 2-3/5 (Medical Research Council [MRC] scale) strength proximally and 3-4/5 strength distally in his arms. His legs have 3-4/5 strength proximally and 4-5/5 strength distally. Sensation is moderately decreased in a stocking-glove distribution to pin, touch, and vibration, with mild proprioceptive loss in his toes. Deep tendon reflexes are absent and Babinski's reflex is negative. Co-ordination is intact except as related to weakness. His gait is hesitant with mild lordosis but otherwise normal. He sways mildly in the Romberg position.

Other presentations

While the most common presentation of typical CIDP is of chronic progressive symmetrical weakness, patients may present with a relapsing and remitting or stepwise course, which are more common with earlier age of onset.[3][4][5] [6] Subacute presentation over 4-8 weeks may make distinction between CIDP and Guillain-Barre syndrome difficult.[3] [5] [7] A subacute monophasic presentation is more common in children, adolescents, and young adults, who often respond well to immunomodulating therapy.[8] [9]

There are several CIDP variants. Pure sensory presentation may occur, with distal sensory loss and pain or with a sensory ataxia, but no muscle weakness. In sensory-predominant CIDP, motor demyelination is found on electrodiagnostic studies and weakness may develop as the course becomes more chronic.[1]

Patients with pure motor CIDP have relatively symmetrical proximal and distal weakness, but normal sensation clinically and electrodiagnostically. Care must be taken to differentiate from multifocal motor neuropathy, in which weakness is asymmetrical and mainly affects the upper limbs. Corticosteroids may cause deterioration in patients with motor CIDP.[1]

Distal CIDP (also called distal acquired demyelinating symmetrical neuropathy) must be distinguished from the neuropathy associated with myelin-associated glycoprotein.[1] [10] [11] Besides the lack of proximal weakness, this presentation differs from classic CIDP in that there is often a monoclonal protein, cerebrospinal fluid protein is usually not elevated, the course is rarely relapsing/remitting, and there is often a poor response to immunosuppressive medication (when a monoclonal protein is present).[10]

CIDP that presents with multifocal asymmetrical weakness and numbness that begins distally, often in one limb, and spreads proximally to involve several limbs is termed multifocal CIDP (also known as multifocal demyelinating neuropathy with persistent conduction block, Lewis-Sumner syndrome; multifocal acquired demyelinating sensory and motor neuropathy [MADSAM]; multifocal inflammatory demyelinating neuropathy).[1] [12] [13]

Rare cases may present with asymmetrical weakness and sensory loss only of the arms, known as focal CIDP or focal upper limb demyelinating neuropathy.[1] [14]

Approach

Diagnosis is based on patients meeting defined clinical criteria. Multiple clinical, electrophysiological, and laboratory criteria have been suggested with varying degrees of sensitivity and specificity.[1] [23][24] [25] While most sets of criteria have specificities approaching 100%, sensitivities are around 60% to 70%.[23] [24] [26] More recent criteria are less restrictive than earlier sets, resulting in higher sensitivities (80% to 85%) while retaining high specificities. Sensitivity may be higher if two sets of criteria with different parameters are combined.[27] Motor conduction block in a non-compressible site is highly sensitive.[26] Testing more nerves and proximal sites may improve sensitivity.[28] [29] Fulfilling the criteria for established diagnostic guidelines may predict a higher treatment response rate than that for patients who do not fulfil these criteria.[30]

Diagnostic criteria were updated in the second revision of the European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) guideline on CIDP (published in 2021) in order to increase specificity, and these criteria are considered the most useful for clinical practice and patient care. Diagnostic categories (based on level of diagnostic certainty) are CIDP and possible CIDP.[1] These criteria update and replace criteria from the 2010 first revision of the guideline.[31]

Despite good sensitivity and specificity of diagnostic criteria for CIDP, misdiagnosis is common, particularly for patients with CIDP variants. This can be due to poorly performed or misinterpreted nerve conduction studies, or non-adherence to electrodiagnostic criteria, and can lead to delay in effective treatment.[1] [2] [16]

Typical features of CIDP include a chronic progressive or relapsing/remitting or stepwise course over at least 8 weeks, resulting in symmetrical proximal and distal weakness, sensory loss in all four limbs, and hyporeflexia or areflexia in all four limbs. Predominantly distal weakness (known as distal acquired demyelinating symmetrical neuropathy), asymmetrical weakness, focal weakness, and subacute presentation over 4-8 weeks are alternative presentations indicating CIDP variants.[1] Electrodiagnostic evidence of demyelination is mandatory to make the diagnosis of CIDP.

Clinical evaluation

CIDP can occur at any age, with most common age of onset being between 40 and 60 years, and it is more common in men.[1] [2] [5] [15]

Drug or toxin exposure that is likely to have caused the neuropathy (e.g., diphtheria, buckthorn, amiodarone, tacrolimus) may rule out a diagnosis of CIDP.[32] [33] Hereditary demyelinating neuropathy based on family history, foot deformity, retinitis pigmentosa, optic atrophy, hearing loss, liability to pressure palsy, self-mutilation, significant central nervous system (CNS) manifestations, or presence of significant bowel or bladder sphincteric complaint may also point to an alternative diagnosis.[34] [35] [36]

Routine neurological examination is required. Weakness is seen in nearly all patients, and gait difficulty is common. Hyporeflexia or areflexia and sensory dysfunction (numbness, paraesthesias) are hallmarks of CIDP.[1][3] [16] Pain may be a feature; the prevalence of pain due to CIDP at any time during the disease course was estimated as 46% in one systematic review.[37]

Rarely, patients present with only sensory symptoms, typically distal paraesthesias and dysaesthesias, but may have sensory ataxia, which can also lead to gait difficulty.[1] [38] [39] Other uncommon manifestations include dyspnoea, vision loss, cranial nerve complaints (e.g., facial weakness, dysarthria, or dysphagia), erectile dysfunction, orthostatic hypotension, and urinary incontinence, urgency, or hesitancy; however, these are also symptoms and signs of alternative diagnoses.[40][41] Nerve

hypertrophy may be detected in a few patients. CNS findings such as papilloedema or spasticity have been reported, but are quite rare. Pure motor presentation may also occur.[1]

Investigations

Nerve conduction studies should be performed for all patients with suspected CIDP. Supportive investigations for diagnosis include cerebrospinal fluid (CSF) analysis, imaging studies with ultrasound or magnetic resonance imaging (MRI), response to treatment, and nerve biopsy; these may be helpful when electrodiagnostic studies are inconclusive or result in a diagnosis of 'possible CIDP' in patients who meet the clinical criteria for CIDP.[1]

Electrodiagnostic evaluation

- Electrodiagnostic evidence of demyelination is mandatory to make the diagnosis and should be ordered for all patients with a clinical suspicion of CIDP. The most important test is nerve conduction studies.
- Diagnostic criteria are mainly based on finding a combination of slowed conduction velocities, prolonged distal latencies, prolonged F-wave latencies, and conduction block in one or more motor nerves.[1]
- Sensory criteria are now also included in electrodiagnostic criteria.[1]
- Patients with a suggestive history and neurological examination, and supportive criteria consistent with a diagnosis of CIDP, should not be excluded from treatment if they do not fulfil electrophysiological criteria, as long as nerve conduction studies show some evidence of demyelination. Furthermore, especially for patients with CIDP variants, sensitivity may be improved by studying more than four motor nerves, utilising proximal stimulation in the upper limbs, and including sensory nerves and somatosensory evoked potentials.[1] Nerve conduction studies should be repeated if doubt still exists about the diagnosis.
- For patients with pure sensory CIDP, nerve conduction studies may show only mild abnormalities not meeting typical electrodiagnostic criteria; elevated CSF protein, abnormal somatosensory evoked potentials, and demyelination on nerve biopsy may be present.[38] [39]
- In one study of patients referred to a tertiary care centre for refractory CIDP, one of the main reasons for therapeutic failure was an incorrect alternative diagnosis. The importance of nerve conduction studies was emphasised, as many patients with alternative diagnoses (e.g., amyotrophic lateral sclerosis, idiopathic neuropathy, small-fibre neuropathy) had no demyelinating features and clinically lacked distal leg weakness, vibratory sensory loss, and widespread areflexia. CIDP mimics were also often misdiagnosed.[42]

CSF evaluation

- CSF analysis should be considered for patients with suspected CIDP when clinical and electrophysiological testing is not definitively diagnostic. It is also advised if infection or lymphoproliferative disease is suspected or possible.[1]
- In >90% of patients with CIDP, CSF analysis typically shows albuminocytological dissociation: elevated protein levels with a normal leukocyte count (<10 leukocytes/mm^3). CSF protein levels of >45 mg/dL are abnormal; levels of >100 mg/dL are not unusual, and much higher levels can be found. However, cautious interpretation is needed, especially for patients with diabetes mellitus or spinal stenosis, and in young or older patients.[1] [43] [44] [45]
- Cell counts of >10 leukocytes/mm^3 should raise suspicion of infection or malignancy. Cell counts may be up to 50 leukocytes/mm^3 in HIV infection.[46] [47]

- The sensitivity of CSF analysis for detecting CIDP variants is unknown.[1] [48]

Imaging

For patients who fulfil clinical and electrodiagnostic criteria for a diagnosis of CIDP, imaging is not needed for diagnosis and therefore is not recommended.[1]

Before concluding that abnormalities on imaging are supportive of a diagnosis of CIDP, other diseases should be considered and excluded. These include multifocal motor neuropathy, demyelinating Charcot-Marie-Tooth disease, IgM paraproteinaemic neuropathy (especially with anti-myelin-associated glycoprotein [MAG] antibodies), polyneuropathy-organomegaly-endocrinopathy-M-protein-skin changes (POEMS) syndrome, diabetic radiculoplexus neuropathy, amyloid neuropathy, neuralgic amyotrophy, leprosy, neurofibromatosis, and neurolymphomatosis.[1]

Nerve ultrasound

- Nerve ultrasound is recommended as a diagnostic tool for adult patients who fulfil the diagnostic criteria for possible CIDP.[1]
- The most common finding on ultrasound is multifocal nerve enlargement.[1] [49] [50] This can help to distinguish CIDP from Charcot-Marie-Tooth disease type 1, in which nerves are usually diffusely enlarged.[51] [52]
- The presence of multifocal nerve enlargement correlates with electrodiagnostic findings.[49] [50] [53] One study showed motor nerve conduction velocity to be inversely correlated with cross-sectional area in nerve segments with electrodiagnostic evidence of demyelination.[53] Nerves with increased cross-sectional area were seen in patients with a more severe course of CIDP as manifest by longer disease duration, lower Medical Research Council (MRC) sum score, higher Inflammatory Neuropathy Cause and Treatment (INCAT) score, and progressive disease.[53] [54]
- Diagnosis may be more likely if there is nerve enlargement of at least two sites in proximal median nerve segments/and or the brachial plexus (mimics must be excluded).[1]
- Ultrasound may be used to assess treatment response.[55] Enlarged nerves may become smaller or normalise with remission of disease.[56] Patients with active CIDP that is refractory to treatment will generally have enlarged nerves without significant change over time.[56]
- There is a lack of evidence on the use of ultrasound in paediatric patients.[1]

MRI

- MRI spine and plexus with and without contrast may be considered for adult patients when electrodiagnostic studies are inconclusive or for those who fulfil diagnostic criteria for possible CIDP, and when ultrasound is unavailable or unclear.[1]
- Without gadolinium enhancement, hypertrophy of lumbosacral or cervical nerve roots or of the lumbosacral or brachial plexus is the most common abnormality. Rarely, hypertrophy involves cranial nerves; it is more common in long-standing disease with a relapsing-remitting course. Enhancement may be present and suggests inflammation.[1] [57]
- There is a lack of evidence on the use of MRI in paediatric patients.[1]

Clinical trial of therapy

- Most patients with CIDP will have a response to treatment with a first-line monotherapy (intravenous immunoglobulin, corticosteroids, or plasma exchange).[1] [58] [59] [60] If the first monotherapy is ineffective, an alternative first-line agent should be tried.
- Lack of at least a partial response to one or two first-line immunosuppressive agents should lead to consideration of an alternative diagnosis: for example, autoimmune nodopathies.[1]

Nerve biopsy

- Nerve biopsy is of limited diagnostic value, and is unnecessary for most patients.[3] It should only be performed in certain circumstances, mainly if the diagnosis is unclear and alternative diagnoses have not been fully excluded after nerve conduction studies, lumbar puncture, laboratory investigations, and imaging (if indicated). It may be helpful in cases where electrodiagnostic studies are inconclusive.[1] [61] Biopsy should also be considered If there is a high suspicion of an infiltrative process such as seen in lymphoma, malignancy, amyloidosis, or sarcoidosis.[1]
- Although the sural or superficial peroneal nerve is often biopsied, a biopsy from a clinically affected nerve will be more informative.[4] [21]
- Classic findings include unequivocal or predominant evidence of segmental demyelination, remyelination, or onion bulb formation by electron microscopy or teased fibre analysis. Demyelination often occurs paranodally or near nodes of Ranvier. Inflammatory cells (may or may not be present) are typically both epineurial and endoneurial.[1] [3] [6] [41]
- Evidence of axonal degeneration is common, and includes axonal loss and myelin ovoid formation. [6] [21] [22] [40]

Other laboratory tests

- Enzyme-linked immunosorbent assay (ELISA) or Western blot can be used to detect auto-antibodies. A small subset of patients have antibodies against nodal or paranodal proteins, which are usually of the IgG4 subclass; these include contactin-1 (CNTN1), neurofascin-155 (NF155), contactin-associated protein 1 (Caspr1), and neurofascin isoforms NF140/186.[62] [63] Such patients are classified as having an autoimmune nodopathy, which presents with a distinct phenotype including rapid onset, ataxia, tremor, and poor response to intravenous immunoglobulin and corticosteroids.[1] Response to cyclophosphamide or rituximab has been reported.[64] [65] [66]
- A range of tests is recommended to identify other potential conditions (e.g., HIV, diabetes mellitus, monoclonal gammopathy, malignancy, hepatitis, sarcoidosis, systemic lupus erythematosus, mixed connective tissue disease) or to establish an alternate diagnosis if the criteria for CIDP are not met.[1]

History and exam

Key diagnostic factors

disease progression (common)

- Most patients have either a chronic progressive course or a relapsing-remitting course over >8 weeks.[3][5] [6]
- Rarely, patients present with subacute onset over 4-8 weeks with a monophasic course.[5] [7] [8] This presentation is more common in children.[5] [9]

weakness (common)

- Over 90% of patients report weakness that is typically symmetrical, involves all four limbs, and affects proximal and distal muscles.[3] [4][5] [6]
- Rarely, weakness affects arms more than legs or is asymmetrical.[12] [14] [67]
- Pure motor syndromes occur rarely.[1] [68][69]
- Distal CIDP (also known as distal acquired demyelinating symmetrical neuropathy) presents with predominant distal weakness.[1] [10]

altered sensation (common)

- Numbness or paraesthesia is noted in about 75% of patients.[3] [4][5] [6] The prevalence of pain due to CIDP at any time during the disease course was estimated as 46% in one systematic review.[37]
- Nearly all patients have symmetrical distal sensory loss to large- and small-fibre modalities.[3] [4][5] [6]
- Rarely, patients present with only sensory symptoms, typically distal paraesthesias and dysaesthesias.[1] [38] [39]
- Pure sensory syndromes occur in <10% of patients and may be either mixed large- and small-fibre or predominantly large-fibre, in which sensory ataxia is the major manifestation.[38] [39] [70] [71] In pure sensory syndromes, nerve conduction studies may show only mild abnormalities not meeting typical electrodiagnostic criteria. Elevated cerebrospinal fluid protein, abnormal somatosensory evoked potentials, and demyelination on nerve biopsy may be present.[38] [39]

decreased deep tendon reflexes (common)

- Hyporeflexia or areflexia is a required component in most sets of diagnostic criteria and occurs in >90% of patients.[3] [4][5] [6]

Other diagnostic factors

incoordination (common)

- Gait difficulty is usually due to weakness. However, presentation with a sensory ataxia is also described rarely in patients.[70] [71]

age 40 to 60 years (common)

- Can occur at any age (although rare in childhood), but the most common age of onset is between 40 and 60 years.[1] [5]

preceding infection (uncommon)

- Because of the chronic nature of the disease, it can be difficult to document a temporal relationship, but most studies suggest that 10% to 30% of patients have a preceding infection.[4] [6] [17]

absence of exposure to neuropathy-causing drugs (uncommon)

- Drug or toxin exposure that is likely to have caused the neuropathy (e.g., diphtheria, buckthorn, amiodarone, tacrolimus) may rule out a diagnosis of CIDP.[32] [33]

dyspnoea (uncommon)

- Shortness of breath requiring intubation occurs in <5% of patients.[4][70] [72]

facial weakness (uncommon)

- Occurs in about 15% of patients.[4][5] [6]

dysarthria (uncommon)

- Occurs in about 15% of patients.[4][5] [6]

dysphagia (uncommon)

- Occurs in about 15% of patients.[4][5] [6]

urinary incontinence (uncommon)

- Occurs in about 25% of patients and is often mild.[73] [74]

urinary urgency or hesitancy (uncommon)

- Occurs in about 25% of patients and is often mild.[73] [74]

impotence (uncommon)

- Occurs in about 25% of patients.[73] [74]

orthostatic hypotension (uncommon)

- Occurs in about 25% of patients.[73] [74]

papilloedema (uncommon)

- May be present but is usually mild.[75] [76] [77]

vision loss (uncommon)

- May be present but is usually mild.[75] [76] [77]

spasticity (uncommon)

- May be present but is usually mild.[75] [76] [77]

Risk factors

Weak

male sex

- CIDP is more common in men than in women, with reported male-to-female ratios of between 1.4 and 4.4.[15]

autoimmune diseases

- May co-exist with other autoimmune diseases, such as systemic lupus erythematosus, connective tissue disease, inflammatory bowel disease, thyroid disease, sarcoidosis, glomerulonephritis, chronic active hepatitis, or graft-versus-host disease.[19] [20]

diabetes mellitus

- There is an increased incidence of CIDP in patients with diabetes mellitus, and severity of CIDP may be worse in these patients.[18] [19] [20]

infection

- Can occur in association with infection, including HIV.[19] [20]

monoclonal gammopathy of undetermined significance (MGUS)

- About 10% to 15% of patients have a benign MGUS, most often with an IgA or IgG paraprotein; MGUS may affect disease course and response to treatment.[2] [19] [20]

Investigations

1st test to order

Test	Result
nerve conduction studies • All patients with a clinical suspicion of CIDP should have electrodiagnostic studies. • Diagnostic criteria are mainly based on finding a combination of slowed conduction velocities, prolonged distal latencies, prolonged F-wave latencies, and conduction block in one or more motor nerves. Sensory nerve conduction studies help support the diagnosis.[1] • A diagnosis of CIDP should not be completely excluded if a patient does not fulfil electrophysiological criteria, as long as nerve conduction studies show some evidence of demyelination. Furthermore, especially for patients with CIDP variants, studying more than four motor nerves, utilising proximal stimulation in the upper limbs, and including sensory nerves and somatosensory evoked potentials may improve sensitivity.[1] Nerve conduction studies should be repeated if doubt still exists about the diagnosis. • In purely sensory CIDP, nerve conduction studies may show only mild abnormalities not meeting typical electrodiagnostic criteria.[38] [39] • In one study of patients referred to a tertiary care centre for refractory CIDP, one of the main reasons for therapeutic failure was an incorrect alternative diagnosis. The importance of nerve conduction studies was emphasised, as many patients with alternate diagnoses (i.e., amyotrophic lateral sclerosis, idiopathic neuropathy, small-fibre neuropathy) had no demyelinating features and clinically lacked distal leg weakness, vibratory sensory loss, and widespread areflexia. CIDP mimics were also often misdiagnosed.[42]	**slow conduction velocities; prolonged distal latencies; prolonged F-wave latencies; conduction block in non-compressible sites**

Other tests to consider

Test	Result
cerebrospinal fluid (CSF) evaluation • CSF analysis should be considered for patients with suspected CIDP when clinical and electrophysiological testing is not definitively diagnostic. It is also advised if infection or lymphoproliferative disease is suspected or possible.[1] • In >90% of CIDP patients, CSF analysis typically shows albuminocytological dissociation, consisting of elevated protein levels with a normal leukocyte count (<10 leukocytes/mm³). CSF protein levels of >45 mg/dL are abnormal; levels of >100 mg/dL are not unusual, and much higher levels can be found. However, cautious interpretation is needed, especially for patients with diabetes mellitus or spinal stenosis, and in young or older patients.[1] [43] [44] [45] • Cell counts >10 leukocytes/mm³ should raise suspicion of infection or malignancy. Cell counts may be up to 50 leukocytes/mm³ in HIV infection.[46] [47] • The sensitivity of CSF analysis for detecting CIDP variants is unknown.[1] [48]	**albuminocytological dissociation**
nerve ultrasound • For patients who fulfil clinical and electrodiagnostic criteria for a diagnosis of CIDP, imaging is not needed for diagnosis and therefore not recommended.[1] Nerve ultrasound is recommended as a diagnostic tool for adult patients who fulfil the diagnostic criteria for possible CIDP.[1] • The most common finding on ultrasound is multifocal nerve enlargement.[1] [49] [50] This can help to distinguish patients with CIDP from those with Charcot-Marie-Tooth disease type 1, in which nerves are usually diffusely enlarged.[51] [52] • The presence of multifocal nerve enlargement correlates with electrodiagnostic findings.[50] [53][78] One study showed motor nerve conduction velocity to be inversely correlated with cross-sectional area in nerve segments with electrodiagnostic evidence of demyelination.[53] Nerves with increased cross-sectional area were seen in patients with a more severe course of CIDP, as manifested by longer disease duration, lower Medical Research Council (MRC) sum score, higher Inflammatory Neuropathy Cause and Treatment (INCAT) score, and progressive disease.[53] [54] • Diagnosis may be more likely if there is nerve enlargement of at least two sites in proximal median nerve segments/and or the brachial plexus (mimics must be excluded).[1] • Ultrasound may be used to assess treatment response.[55] Enlarged nerves may become smaller or normalise with remission of disease. Patients with active CIDP that is refractory to treatment will generally have enlarged nerves without significant change over time.[56] • There is a lack of evidence on the use of ultrasound in paediatric patients.[1]	**multifocal nerve enlargement of at least two sites in proximal median nerve segments and/or the brachial plexus; nerve enlargement is defined by cross-sectional area of median nerve >10 mm² at forearm, >13 mm² upper arm, >9 mm² interscalene (trunks), or >12 mm² for nerve roots**

Test	Result
MRI spine and plexus with and without contrast • For patients who fulfil clinical and electrodiagnostic criteria for a diagnosis of CIDP, imaging is not needed for diagnosis and therefore not recommended.[1] • MRI may be considered for adult patients when electrodiagnostic studies are inconclusive or who fulfil diagnostic criteria for possible CIDP, and when ultrasound is unavailable or unclear.[1] • Without gadolinium enhancement, hypertrophy of lumbosacral or cervical nerve roots or of the lumbosacral or brachial plexus is the most common abnormality. Rarely, hypertrophy involves cranial nerves; it is more common in long-standing disease with a relapsing-remitting course. Enhancement may be present and suggests inflammation.[1] [57] [79] [80] • There is a lack of evidence on the use of MRI in paediatric patients.[1]	**enlargement and/or increased signal intensity of nerve root(s) on T2-weighted MRI sequences**
clinical trial of therapy • Most patients with CIDP will have a response to treatment with a first-line monotherapy (intravenous immunoglobulin, corticosteroids, or plasma exchange).[1] [58] [59] [60] If the first monotherapy agent is ineffective, an alternative first-line agent should be tried. • Lack of at least a partial response to one or two first-line immunosuppressive agents should lead to consideration of an alternative diagnosis: for example, autoimmune nodopathies.	**clinical response**
nerve biopsy • Nerve biopsy is of limited diagnostic value, and is unnecessary for most patients.[3] It should only be performed in certain circumstances, mainly if the diagnosis is unclear and alternative diagnoses have not been fully excluded after nerve conduction studies, lumbar puncture, laboratory investigations, and imaging (if indicated). It may be helpful in cases where electrodiagnostic studies are inconclusive.[1] [61] Biopsy should also be considered If there is a high suspicion of an infiltrative process such as seen in lymphoma, malignancy, amyloidosis, or sarcoidosis.[1] • Although the sural or superficial peroneal nerve is often biopsied, a biopsy from a clinically affected nerve will be more informative.[4] [21] • Classic findings include unequivocal or predominant evidence of segmental demyelination, remyelination, or onion bulb formation by electron microscopy or teased fibre analysis. Demyelination often occurs paranodally or near nodes of Ranvier. Inflammatory cells (may or may not be present) are typically both epineurial and endoneurial.[1] [3] [6] [41] • Evidence of axonal degeneration is common and includes axon loss and myelin ovoid formation.[6] [21] [22] [40]	**demyelination and/or remyelination; axonal degeneration**
enzyme-linked immunosorbent assay (ELISA) or Western blot to detect auto-antibodies • A small subset of patients have antibodies against nodal or paranodal proteins, which are usually of the IgG4 subclass; these include contactin-1 (CNTN1), neurofascin-155 (NF155), contactin-associated protein 1 (Caspr1), and neurofascin isoforms NF140/186.[62] [63] Such patients are classified as having an autoimmune nodopathy, which presents with a distinct phenotype including rapid onset, ataxia, tremor, and poor response to intravenous immunoglobulin and	**may show anti-CNTN1, anti-NF155, anti-Caspr1, or anti-NF140/186 antibodies**

Test	Result
corticosteroids.[1] Response to cyclophosphamide or rituximab has been reported.[64] [65] [66]	
other tests • A range of tests is recommended to identify other potential conditions (e.g., HIV, diabetes mellitus, monoclonal gammopathy, malignancy, hepatitis, sarcoidosis, systemic lupus erythematosus, mixed connective tissue disease) or to establish an alternate diagnosis if the criteria for CIDP are not met.[1]	**positive result guides alternate diagnosis**

Differentials

Condition	Differentiating signs / symptoms	Differentiating tests
Guillain-Barre syndrome (GBS)	• GBS typically progresses for <4 weeks. In cases that progress for 4-8 weeks, distinction can be difficult, because these cases mimic acute- or subacute-onset CIDP.[7][81] [82] • Some subacute presentations of CIDP can be monophasic, as in GBS.[4][8] [40] • GBS is associated with more frequent antecedent infection, respiratory failure, cranial nerve involvement, motor greater than sensory involvement, and autonomic dysfunction.[81] [82] [83][84]	• Demyelinating features on nerve conduction studies tend to be less severe.[83] [85] • Nerve ultrasound may show less nerve enlargement with GBS compared with subacute CIDP.[51] [84] [86]
Charcot-Marie-Tooth disease (CMT)	• Also known as hereditary motor and sensory neuropathy. • Early age of onset, slow progression of weakness, skeletal deformities (e.g., pes cavus, hammer toes), and extremely distal weakness.[34]	• Electrodiagnostic features include uniform slowing of conduction velocities and usually absence of conduction block.[87] • Terminal latency index, the modified F ratio, and dispersion of the compound muscle action potential amplitude may also differentiate.[88] [89] • Nerve ultrasound shows more diffuse nerve enlargement, especially in CMT1A, compared with multifocal cross-sectional area enlargement in CIDP.[51] [52] [86] • Nerve biopsy more often shows areas of abnormally thickened or folded (tamaculous) myelin or areas of hypomyelination in CMT.[90]
Anti-myelin-associated glycoprotein (anti-MAG) neuropathy	• Patients tend to be older, more often male, have a more indolent course, and have only distal weakness or sensory ataxia with gait imbalance or action tremors.[11] [91]	• Conduction block and abnormal temporal dispersion rarely occur.[92] • Electrodiagnostic finding of prolonged distal latencies out of proportion to slowed conduction velocities is a hallmark.[93]

Condition	Differentiating signs / symptoms	Differentiating tests
	• Most common presentation is a distal acquired symmetrical demyelinating (DADS) phenotype.[11] [91]	• The presence of anti-MAG antibodies by enzyme-linked immunosorbent assay (ELISA) and Western blot is seen in essentially all patients.[11] • Widely spaced myelin is the classic pathological finding.[11] • Patients do not respond as well to corticosteroids, intravenous immunoglobulin, or plasma exchange.[94] • Rituximab has been widely used and is the treatment of choice despite limited evidence of efficacy.[11] [94] [95]
Monoclonal gammopathy of unknown significance (MGUS)-associated neuropathy	• Patients with CIDP and MGUS (usually IgA, IgG, or IgM) tend to be older and have more distal weakness, a more indolent course, and more sensory involvement.[11] [62] • IgA/IgG MGUS without CIDP has a better response to plasma exchange compared with IgM MGUS in the context of the distal variant of CIDP.[96]	• Testing should include serum protein electrophoresis and immunofixation, spot urine immunofixation for light chains, and serum free light chains.[1] [70] • For IgA or IgG lambda paraproteinaemia, testing of vascular endothelial growth factor (VEGF) serum levels is indicated, especially in patients with clinical suspicion of polyneuropathy-organomegaly-endocrinopathy-M-protein-skin changes (POEMS) syndrome.[1] • For IgM paraproteinaemia, testing for anti-MAG antibody is warranted.[1] • If immunoglobulin level is >15 g/L (>1.5 g/dL), check for a haematological malignancy.[97]
Multifocal motor neuropathy	• Distal asymmetrical weakness and the lack of sensory signs or symptoms are seen.[98] • European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) guidelines consider this a distinct entity from CIDP.[1]	• Electrodiagnostic studies show multifocal conduction block as the major demyelinating feature. Sensory nerve conduction studies are normal.[98] • Anti-GM1 antibodies are found in 30% to 80% of patients.[99] [100] • Nerve ultrasound often shows milder, more asymmetrical nerve

Condition	Differentiating signs / symptoms	Differentiating tests
		• enlargement with greater side-to-side intra-nerve variability.[86] [101] • Responds to treatment with intravenous immunoglobulin, but no response to corticosteroids or plasma exchange.[62] [98]
Autoimmune nodopathy	• Demyelinating polyneuropathy with acute or subacute aggressive onset, resembling Guillain-Barre syndrome or acute-onset CIDP.[63] • Low-frequency tremor, ataxia disproportionate to the sensory involvement or other cerebellar features, or predominantly distal weakness.[63] • May be associated with nephrotic syndrome.[63] • Central nervous system demyelination has been reported.[102] [103]	• Antibodies (mostly IgG4 subtypes) present against nodal and paranodal antigens, such as neurofascin-155 (NF155), contactin-1 (CNTN1), contactin-associated protein 1 (Caspr1), and neurofascin isoforms NF140/186.[62] [63] • Very high CSF protein levels may be present.[63] • Poor response to intravenous immunoglobulin and corticosteroids.[1] • Responds to cyclophosphamide or rituximab.[64] [104]
Chronic immune sensory polyradiculoneuropathy (CISP) and CISP-plus	• Chronic and progressive sensory ataxia. Preferentially affects large myelinated fibres of the sensory nerve roots proximal to dorsal root ganglions.[105] • CISP-plus was described in patients with CISP and mild distal weakness due to the extension of involvement distal to the dorsal root ganglions.[106] • There is not enough evidence to determine whether CISP is demyelinating or related to sensory CIDP.[1]	• Electrodiagnostic studies typically show normal nerve conduction. In CISP-plus, nerve conduction studies may show mild abnormality due to axonopathy.[105] [106] • Abnormal somatosensory evoked potential.[105] • Nerve root enlargement on MRI.[105] • Elevated CSF protein.[105] • Lumbar sensory rootlet biopsy can show inflammatory hypertrophic changes.[105] • Response to intravenous immunoglobulin or corticosteroids.[62] [105]
Neuropathy associated with lymphoproliferative disorders	• Neuropathy associated with multiple myeloma, polyneuropathy-organomegaly-endocrinopathy-M-protein-skin changes (POEMS) syndrome, osteosclerotic myeloma, Waldenstrom's macroglobulinaemia,	• Serum and urine immunofixation, quantitative immunoglobulin levels, skeletal survey, and bone marrow biopsy will usually find the haematological abnormality.[97] • Testing of vascular endothelial growth factor

Condition	Differentiating signs / symptoms	Differentiating tests
	and Castleman's disease tends to be predominantly distal.[107] [108] Neuropathy associated with cryoglobulinaemia and lymphoma can be multifocal.[109]	(VEGF) serum levels, especially in patients with IgA or IgG lambda paraproteinaemia, is indicated.[1] • Nerve biopsy may be needed in lymphoma.[109] • Electrodiagnostic studies may have features of axonal degeneration and demyelination, although axonal degeneration may predominate.[107] [108] [109] • Nerve ultrasound may show nerve enlargement only at the entrapment sites with distinct echogenicity pattern (hypoechoic fascicles with hyperechoic interfascicular tissue).[86] [110] • POEMS syndrome is associated with more severe reduction in lower extremity compound muscle action potential and sensory nerve action potential amplitudes, less temporal dispersion and conduction block, fewer prolonged distal motor latencies, higher terminal latency index in median nerve, and more evidence of active denervation in a length-dependent manner.[111] [112] [113]
Diabetic neuropathies	• Typical neuropathy is more axonal than demyelinating, and is distal, symmetrical, and sensory more than motor.[3] • Proximal diabetic neuropathies are often asymmetrical and can be painful.[114] • Acute diabetic neuropathy is often related to rapid glycaemic reduction.[115] [116] • Diabetic neuropathy and CIDP may co-exist.[117]	• Electrodiagnostic studies show primarily axon loss in diabetes mellitus, but secondary demyelinating features can be present. [117] [118]
Vasculitic neuropathy	• Manifests as subacute, progressive, asymmetrical sensorimotor	• Laboratory tests for systemic vasculitis may include antinuclear or antineutrophil

Condition	Differentiating signs / symptoms	Differentiating tests
	polyneuropathy, or mononeuropathy multiplex. • May be associated with systemic lupus erythematosus.[119] • May mimic multifocal CIDP (Lewis-Sumner syndrome).[12] • May present as part of systemic vasculitis or remain clinically restricted to the peripheral nerve.	cytoplasmic antibodies, erythrocyte sedimentation rate, CRP, anti-double-stranded DNA, cryoglobulin, etc. • Electrodiagnostic studies show axon loss, although pseudoconduction block can be seen with early lesions.[120] • Repeat nerve conduction studies often show later features typical of axon loss. • Nerve and/or muscle biopsy may be needed to document evidence of nerve inflammation.[121]
Drug-induced neuropathy	• Look for onset of disease in relation to onset of drug use. • Weakness tends to be predominantly distal.[32] [33] • Amiodarone, perhexiline, arsenic, glue sniffing, and buckthorn shrub poisoning are among toxins known to cause demyelinating neuropathy.[32]	• Nerve biopsy may show perineural or Schwann cell lysosomal inclusions with amiodarone and perhexilene.[32] [33]
Mitochondrial disorders (MNGIE syndrome)	• Neuropathy is predominantly distal. • Look for associated features: gastrointestinal neuropathy, ophthalmoplegia, cachexia, hearing loss, or leukoencephalopathy.[122]	• Muscle biopsy shows mitochondrial pathology due to multiple mitochondrial deletions (cytochrome oxidase-negative fibres and ragged red fibres).[122] • Laboratory tests show serum lactic acidosis and markedly elevated serum thymidine levels.[122]
Leukodystrophies	• In metachromatic leukodystrophy and Krabbe's leukodystrophy, the presenting features may be of a polyneuropathy indistinguishable from CIDP.[35] • In adrenoleukodystrophy, central nervous system signs and symptoms predominate.[35]	• Electrodiagnostic findings are more uniform in the leukodystrophies.[35] • Brain MRI findings are prominent and nerve biopsy usually shows Schwann cell inclusions in leukodystrophies.[35]
Infection-associated neuropathy	• Look for signs and symptoms of infection	• HIV test, hepatitis serology, or Lyme serology may be positive.

Condition	Differentiating signs / symptoms	Differentiating tests
	(e.g., HIV, hepatitis, Lyme disease).	• Cerebrospinal fluid pleocytosis with >10 cells/mm³.[46] [47]
Sarcoidosis	• Cranial neuropathy is most common.[123] • Peripheral nerve involvement consists of granulomatous neuropathy (rare) and non-granulomatous small-fibre neuropathy (more common).[124] • Granulomatous neuropathy may present as distal symmetrical polyneuropathy and asymmetrical polyradiculoneuropathy.[124] • Small-fibre neuropathy can manifest as non-length-dependent paraesthesias and pain.[124]	• Electrodiagnostic studies show primarily axonal loss of large myelinated fibres, but secondary demyelinating features can be present.[124] • Non-caseating granulomas are rarely found in nerve biopsies. • Endoneurial or epineurial inflammatory infiltrates may be present.[124]
Thyroid disease	• Neuropathy with hypothyroidism or hyperthyroidism is predominantly distal and sensory, and usually improves with correction of thyroid status.[40]	• May consider thyroid function studies and/or thyroid auto-antibody testing.[40][125]
Amyloidosis	• Typically, presentations include distal axonal sensory neuropathy, small-fibre neuropathy, carpal tunnel syndrome, or autonomic neuropathy. Rare presentations of multifocal neuropathy with demyelinating features have been reported.[126] [127]	• Amyloid present on nerve biopsy.[128] • Electrodiagnostic studies may have features of axonal degeneration and demyelination, although axonal degeneration may predominate.[126] [127] • Transthyretin mutation analysis for familial amyloidosis.[127] [128]

DIAGNOSIS

Criteria

European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) criteria[1]

Multiple clinical, electrophysiological, and laboratory criteria have been suggested, with varying degrees of sensitivity and specificity.[1] [23] [24][25] [41] [83][129] While most sets of criteria have specificities approaching 100%, sensitivities are around 60% to 70%.[24] [26] [23] Sensitivity may be higher if two sets of criteria with different parameters are combined.[27] Motor conduction block in a non-compressible site is highly sensitive.[26] Testing more nerves and proximal sites may improve sensitivity.[28] [29] Patients who fulfil diagnostic criteria, especially those with a higher degree of demyelination, have a predicted greater treatment response rate than patients who do not fulfil these criteria.

Revised diagnostic criteria were published by the EAN/PNS in 2021, replacing those published in 2010.[1] [31]

Clinical diagnostic criteria

a. Typical CIDP

All of the following criteria are met:

- Progressive or relapsing, symmetrical, proximal and distal muscle weakness of upper and lower limbs, and sensory involvement of at least two limbs
- Developing over at least 8 weeks
- Absent or reduced tendon reflexes in all limbs.

The term 'acute-onset CIDP' (A-CIDP) represents typical CIDP that rapidly progresses within 4 weeks and mimics GBS.

b. CIDP variants

One of the following, but otherwise as for typical CIDP (tendon reflexes may be normal in unaffected limbs):

- Distal CIDP: distal sensory loss and muscle weakness predominantly in lower limbs. (Distal neuropathy with IgM paraprotein and anti-MAG antibodies [anti-MAG neuropathy] is considered outside the scope of CIDP as most patients have specific electrodiagnostic and pathological findings, and intravenous immunoglobulin and corticosteroids are not effective.)
- Multifocal CIDP: sensory loss and muscle weakness in a multifocal pattern, usually asymmetrical, upper limb predominant, in more than one limb.
- Focal CIDP: sensory loss and muscle weakness in only one limb.
- Motor CIDP: motor symptoms and signs without sensory involvement.
- Sensory CIDP: sensory symptoms and signs without motor involvement.

c. Disorders not classified as CIDP

- Chronic immune sensory polyradiculopathy (CISP): not included in the CIDP variant classification because there is not enough evidence to determine if CISP is demyelinating or related to sensory CIDP.
- Autoimmune nodopathies: not regarded as CIDP variants because they have distinct clinical features, no overt inflammation or macrophage-mediated demyelination, and respond poorly to treatments for CIDP (especially intravenous immunoglobulin).

Motor nerve conduction criteria

(1) Strongly supportive of demyelination:

At least one of the following:

- Motor distal latency prolongation ≥50% above upper limit of normal values (ULN) in two nerves (excluding median neuropathy at the wrist from carpal tunnel syndrome), or
- Reduction of motor conduction velocity ≥30% below lower limit of normal values (LLN) in two nerves, or
- Prolongation of F-wave latency ≥20% above ULN in two nerves (≥50% if amplitude of distal negative peak compound muscle action potential [CMAP] <80% of LLN), or

- Absence of F-waves in two nerves (if these nerves have distal negative peak CMAP amplitudes ≥20% of LLN) + ≥1 other demyelinating parameter in ≥1 other nerve, or
- Motor conduction block: ≥30% reduction of the proximal relative to distal negative peak CMAP amplitude, excluding the tibial nerve, and distal negative peak CMAP amplitude ≥20% of LLN in two nerves; or in one nerve + ≥ 1 other demyelinating parameter except absence of F-waves in ≥1 other nerve, or
- Abnormal temporal dispersion: >30% duration increase between the proximal and distal negative peak CMAP (at least 100% in the tibial nerve) in ≥2 nerves, or
- Distal CMAP duration (interval between onset of the first negative peak and return to baseline of the last negative peak) prolongation in ≥1 nerve + ≥1 other demyelinating parameter in ≥1 other nerve

    - (Low frequency filter [LFF] 2 Hz) median >8.4 ms, ulnar >9.6 ms, peroneal >8.8 ms, tibial >9.2 ms
    - (LFF 5 Hz) median >8.0 ms, ulnar >8.6 ms, peroneal >8.5 ms, tibial >8.3 ms
    - (LFF 10 Hz) median >7.8 ms, ulnar >8.5 ms, peroneal >8.3 ms, tibial >8.2 ms
    - (LFF 20 Hz) median >7.4 ms, ulnar >7.8 ms, peroneal >8.1 ms, tibial >8.0 ms.

(2) Weakly supportive of demyelination

As in (1) but in only one nerve.

Sensory nerve conduction criteria

(1) CIDP

- Sensory conduction abnormalities (prolonged distal latency, or reduced sensory nerve action potential [SNAP] amplitude, or slowed conduction velocity outside of normal limits) in two nerves.

(2) Possible CIDP

- As in (1).
- Sensory CIDP with normal motor nerve conduction studies needs to fulfil (a) or (b):

    - sensory nerve conduction velocity <80% of LLN (for SNAP amplitude >80% of LLN) or <70% of LLN (for SNAP amplitude <80% of LLN) in at least two nerves (median, ulnar, radial, sural nerve), or
    - sural sparing pattern (abnormal median or radial SNAP amplitude with normal sural nerve SNAP amplitude) (excluding carpal tunnel syndrome).

Supportive criteria

Response to treatment, imaging, cerebrospinal fluid (CSF) analysis, or nerve biopsy may be supportive in patients who fulfil clinical criteria for CIDP, but whose electrodiagnostic criteria only allow for possible CIDP.[1]

Diagnostic categories

The 2021 guideline classifies CIDP into two categories (CIDP and possible CIDP).[1] The category of probable CIDP (as in the 2010 guideline) was removed because the diagnostic accuracy of criteria for probable and definite CIDP did not significantly differ.[1]

Typical CIDP

- Typical CIDP
    - Clinical criteria + motor conduction criteria in two nerves + sensory conduction abnormalities in two nerves; or
    - Possible typical CIDP + at least two supportive criteria
- Possible typical CIDP
    - Clinical criteria + motor conduction criteria in one nerve + sensory conduction abnormalities in two nerves; or
    - Clinical criteria + motor conduction abnormalities not fulfilling CIDP motor conduction criteria in one nerve + sensory conduction abnormalities in two nerves + objective response to treatment + one other supportive criterion

Distal CIDP

- Distal CIDP
    - Clinical criteria + motor conduction criteria in two upper limb nerves + sensory conduction abnormalities in two nerves; or
    - Possible distal CIDP + at least two supportive criteria
- Possible distal CIDP
    - Clinical criteria + motor conduction criteria in one upper limb nerve + sensory conduction abnormalities in one nerve; or
    - Clinical criteria + motor conduction criteria in two lower limb nerves only + sensory conduction abnormalities in two nerves (possible distal CIDP only, cannot be upgraded by supportive criteria)

Multifocal or focal CIDP

- Multifocal or focal CIDP
    - Clinical criteria + motor conduction criteria in two nerves + sensory conduction abnormalities in two nerves; or
    - Possible multifocal or focal CIDP + at least two supportive criteria
- Possible multifocal or focal CIDP
    - Clinical criteria + motor conduction criteria in one nerve + sensory conduction abnormalities in two nerves
    - Focal CIDP fulfilling clinical criteria + motor conduction criteria in one nerve + sensory conduction abnormalities in one nerve (possible focal CIDP only, cannot be upgraded by supportive criteria)

Motor CIDP

- Motor CIDP

- Clinical criteria + motor conduction criteria in two nerves + normal sensory conduction in four nerves; or
- Possible motor CIDP + at least two supportive criteria

- Possible motor CIDP

  - Clinical criteria + motor conduction criteria in one nerve + normal sensory conduction in four nerves

Motor-predominant CIDP

- As in motor CIDP but with sensory conduction abnormalities in two nerves

Sensory CIDP

- Possible sensory CIDP

  - Clinical criteria + sensory conduction criteria (possible sensory CIDP only, cannot be upgraded by supportive criteria). Motor conduction must be normal in at least four nerves.

Sensory-predominant CIDP

- Possible sensory-predominant CIDP

  - Clinical criteria + sensory conduction abnormalities in two nerves + motor conduction abnormalities in two nerves or motor conduction criteria fulfilment in one nerve

- Sensory-predominant CIDP

  - Clinical criteria + sensory conduction abnormalities in two nerves + motor conduction criteria fulfilment in two nerves.

Approach

Management of CIDP should encompass a multi-modality approach, focusing on treating the underlying inflammatory disease process and maximising quality of life. Early detection and treatment minimises secondary axonal loss and prevents further impairment.[130] Multi-disciplinary care should include access to a neurologist, pain specialist, physiotherapist, occupational therapist, orthotist, prosthetist, and psychologist, as required.[1] As CIDP may co-exist with other conditions (e.g., diabetes mellitus, monoclonal gammopathy of undetermined significance [MGUS], HIV, or connective tissue diseases), checking for and treating such conditions is essential and may improve outcomes for CIDP.[19]

The treatment strategy for CIDP is dependent on disease course and severity. Patients with mild disease (weakness that does not interfere with daily activities) may be monitored for deterioration without treatment.[131] Most other patients will respond to some form of immunosuppressive therapy. For patients with possible CIDP that does not fulfil clinical or electrodiagnostic criteria, a treatment trial with one of the first-line therapies, with objective measurement of response, may be used to establish a diagnosis.[1]

Initial therapy

Patients in whom the disease has a significant impact on function and quality of life should be treated with immunosuppressant or immunomodulatory therapy. Approximately 75% to 85% of patients will respond to monotherapy with intravenous immunoglobulin (IVIG), corticosteroids, or plasma exchange.[1] [58] [60] [132] [133] [134][135]

Corticosteroids

Corticosteroids are effective for treating CIDP.[1] [60] [134] High-dose pulsed methylprednisolone or dexamethasone are often tried first as they are easy to administer, have few adverse effects, have rapid onset of benefit, and are inexpensive.[1][134] [136] Efficacy and improvement in disability after treatment with pulsed high-dose dexamethasone or typical daily prednisolone regimens are equivalent; however, daily oral prednisolone may cause many more adverse effects, and may take longer to show benefit.[1] [134] [135][136]

Intravenous immunoglobulin (IVIG)

IVIG is another effective option that is often used as first-line treatment because of its ease of use, rapid onset of benefit, and low incidence of adverse effects.[1] [59] [131] [132] [135][137] There is no evidence for a difference in treatment efficacy between IVIG preparations.[1] [138] If efficacious, IVIG can be repeated from every 2 to 6 weeks.[1] To optimise therapy, the dose should be reduced before the frequency of administration is lowered, to achieve maximal benefits with minimal amounts of IVIG. However, care must be taken to avoid deterioration before the next dose.[1][131] [132]

Plasma exchange

Plasma exchange is effective and relatively safe, resulting in significant short-term improvement in disability, clinical impairment, and motor nerve conduction velocity in patients with CIDP.[1] [133] However, some patients who show improvement subsequently deteriorate.[133] Plasma exchange requires good vascular access and specialised equipment, which can make it less convenient than other treatments, especially in paediatric patients.[1] [133] [139]

Choice of initial therapy

Randomised controlled trials comparing efficacy between corticosteroids, IVIG, and plasma exchange do not show a significant benefit of any one over the others.[1] [2] [135] The final decision on which treatment to use is determined by availability, risks, contraindications, and disease severity; however, because corticosteroids and IVIG are easier to administer and are generally better tolerated, they are usually recommended ahead of plasma exchange.[1] If significant adverse effects occur with an initial therapy, an alternative should be substituted.

Corticosteroid therapy is not recommended as first-line treatment for patients with motor CIDP, due to evidence of deterioration after such therapy.[1][68][69] [135]

Partial or no response to initial therapy

It may take up to 3 months to determine treatment effectiveness. Lack of at least a partial response to one or two initial agents should lead to reconsideration of the diagnosis.[2]

If there is a partial but insufficient objective response to the initial agent (i.e., daily activities are still affected by distal weakness and paraesthesias), this agent should be continued and a second or third initial agent added to the regimen.[1]

If there is no objective response to the chosen initial agent within a few weeks to months (and the diagnosis of CIDP is confirmed), an alternative initial agent (corticosteroids, IVIG, or plasma exchange) should be tried before considering combination therapy.[1] [2] [140]

Refractory to combination therapy with initial agents

For patients whose condition does not improve sufficiently in response to a combination of two initial agents (i.e. two of corticosteroids, IVIG, and plasma exchange), re-evaluation of the diagnosis and referral to a specialist centre is appropriate. In one study, 32% of patients referred for CIDP had been incorrectly diagnosed.[16] In a study of patients referred to a tertiary care centre for refractory CIDP, reasons for therapeutic failure included an incorrect alternative diagnosis and inadequate immunotherapy. In patients who had a poor response to treatment, inadequate first-line therapy or failure to add a second or third agent were frequently observed. The authors also stressed the importance of electrodiagnostic studies to distinguish true CIDP from mimics.[42]

Alternative immunosuppressants

Once the diagnosis of CIDP is confirmed, an alternative immunosuppressant should be added. Although there is limited evidence, rituximab, cyclophosphamide, ciclosporin, azathioprine, or mycophenolate may be considered as an alternative agent after failure of initial treatments, or as add-on medication.[1] [141] [142] Any combination of initial and alternative agents can be used, and decisions should be based on severity of disease and adverse-effect profile.[1]

Guidelines recommend against using interferon beta-1a, methotrexate, fingolimod, alemtuzumab, bortezomib, etanercept, fampridine, fludarabine, immunoadsorption, interferon alfa, abatacept, natalizumab, and tacrolimus.[1]

In one retrospective analysis, approximately 25% of patients refractory to standard therapy showed a response to an alternative immunosuppressant; the authors noted that adverse effects should be monitored closely.[142]

Ciclosporin is well tolerated, drug levels can be monitored to guide treatment, and it usually produces benefit within 3-6 months.[1] [141] Combining ciclosporin with plasma exchange can make it difficult to achieve therapeutic levels of ciclosporin.

Cyclophosphamide (often in combination with a corticosteroid) may be used to treat refractory disease.[1] In one meta-analysis, cyclophosphamide yielded a response rate of 68% in patients with refractory CIDP; however, in practice, cyclophosphamide should be used with caution due to its toxicity, and patients must be monitored for adverse effects.[143]

Rituximab may be considered in refractory CIDP or CIDP associated with other autoimmune diseases or haematological diseases (e.g. monoclonal gammopathy).[62] [141][144] [145] Rituximab may also be considered with nodal/paranodal antibodies (e.g. IgG4 anti-contactin-1 or anti-neurofascin-155 antibodies) after failure of corticosteroids or IVIG.[64] [104] [146] [147] It is recommended for children instead of cyclophosphamide because of a better adverse-effect profile.[1]

Azathioprine is another good additional agent that is usually well tolerated.[1] [141] [148] However, onset of action may take 6-18 months, so it is not recommended for initial treatment of relapse or active refractory cases unless combined with other agents.

Mycophenolate is used similarly to azathioprine as an additional agent when rapid improvement is not needed.[1] Some, but not all, studies have shown benefit.[149] [150]

Maintenance therapy

Maintenance therapy may require long-term use of an initial agent, an alternative agent, or a combination of two or more initial or alternative agents, depending on which treatments have produced a response in the acute phase of treatment. IVIG and corticosteroids are the most common agents used. Plasma exchange is generally not recommended long term for patients in whom IVIG and corticosteroids are ineffective, because of the need for vascular access.[1]

Long-term therapeutic options must be chosen on an individual basis, taking into account adverse effects and response to treatment, because there are few clinical trial data to help guide the practitioner. With a good response to an initial agent, the patient usually has up to 1-2 years of therapy (depending on agent used) with a slow taper. If there is an exacerbation of the disease during taper, the dose should be increased to the initial dose that resulted in a good response.[1] Some patients may require 5-15 years of treatment, but eventual withdrawal should be considered, as approximately one-third of patients will go into remission and not require subsequent therapy. Thus, the need for continued maintenance therapy should be re-assessed every 6-12 months.[1]

IVIG

IVIG as maintenance treatment is associated with few adverse effects. The optimal dose and interval for IVIG maintenance treatment are not known. Once the patient is stable, treatment can be tapered based on clinical experience, by either reducing the dose or increasing treatment interval. The adjustment should be done every 6-12 months in the first 2-3 years of treatment.[1]

Subcutaneous immunoglobulin (SCIG) is approved for maintenance therapy in patients with CIDP who are receiving a stable dose of IVIG. There is insufficient evidence to recommend SCIG for initial therapy.[1] SCIG is typically given once weekly, but can be given over 2-3 days for larger doses. There is insufficient evidence that a higher dose of SCIG is superior to a lower dose, but the relapse rate was lower in the higher-dose group.[1] [151] [152] When switching from IVIG to SCIG, it is advised to use the

same mean dose per week.[1] SCIG may be beneficial for patients with adverse effects or treatment-related fluctuations from IVIG that are not resolved by premedication or dose adjustment.[153] SCIG can be administered at home by the patient, and eliminates problems associated with venous access. Some patients may, however, find it difficult to self-administer SCIG due to weakness; others may experience local infusion site reactions.[1]

Corticosteroids

Long-term corticosteroid treatment may induce significant adverse effects (e.g., osteoporosis, gastric ulceration, diabetes mellitus, cataracts, avascular necrosis of long bones, arterial hypertension). However, high-dose pulsed corticosteroids are associated with fewer adverse effects than daily oral dosing.[1]

Alternative agents

The most commonly used alternative medications during maintenance therapy are azathioprine, mycophenolate, and ciclosporin, although evidence of effectiveness is of low certainty. These alternative agents are used to decrease the dose or frequency of immunoglobulin or corticosteroids, or when patients experience adverse effects from those therapies.[1]

Ciclosporin usually shows benefit within 3-6 months, while azathioprine and mycophenolate may take up to 6-18 months. They may all increase the risk of malignancy when used for >10 years. Other alternatives should only be considered after these drugs have been shown to be ineffective.

Due to lack of evidence of efficacy and/or adverse safety profiles, guidelines recommend against using the following agents: methotrexate, tacrolimus, interferon beta-1a, interferon alfa, fingolimod, alemtuzumab, bortezomib, natalizumab, etanercept, abatacept, fampridine, fludarabine, and immunoadsorption.[1]

Stem cell transplantation

Several case reports and series have described the use of stem cell transplantation for treatment-resistant chronic CIDP, but evidence for efficacy is limited.[1] [154] [155] [156] This treatment may have life-threatening adverse effects, with significant morbidity and potential mortality. Therefore it should only be considered in specialised CIDP centres as a last resort for patients with severe chronic CIDP that is unresponsive to all other therapies or who have intolerable adverse effects with more conventional treatments.[1]

Symptomatic therapies

Pain associated with CIDP should be assessed and treated. It may be neuropathic or nociceptive, and a consequence of CIDP, or unrelated to it. Neuropathic pain may be more common in conditions that mimic CIDP (e.g., polyneuropathy-organomegaly-endocrinopathy-M-protein-skin changes [POEMS] syndrome, vasculitis, diabetes, amyloidosis, Charcot-Marie-Tooth disease 1B), so alternative diagnoses should be considered.[1]

All patients with neuropathic pain should be offered symptomatic therapy. Based on current guidelines, pregabalin, gabapentin, tricyclic antidepressants (e.g., amitriptyline), and serotonin-noradrenaline reuptake inhibitors (venlafaxine or duloxetine) are commonly used as first-line agents.[1] [157] [158] Tramadol and other opioid analgesics are recommended as second- and third-line agents, respectively.[157] Guidance from the US Centers for Disease Control and Prevention notes that evidence for efficacy of opioid therapy for neuropathic pain is limited, and that opioids should only be considered

once other options have been tried, and if the expected benefits are anticipated to outweigh risks to the patient.[159] Slow upward dose titration may minimise adverse effects of pain medications, particularly those associated with sedation.

Fatigue is common in CIDP regardless of the disease activity state, and worsening fatigue over time is associated with poor quality of life and increased disability. However, fatigue should not be used as a diagnostic sign for CIDP. Reducing sedative use, better sleep hygiene, and treating depression may help with CIDP-related fatigue.[160] [161]

Depending on the severity of the weakness associated with CIDP, physiotherapy, occupational therapy, and orthotic evaluation may be needed. Referral to one or more of a physical medicine and rehabilitation physician, a psychiatrist or psychologist, a pain management consultant, and a podiatrist should be considered on an individual basis.[1]

A home exercise programme under the supervision of a physiotherapist may be of benefit, but this has not been adequately studied. Only one randomised controlled trial with a measure of functional ability as a primary outcome measure includes patients with CIDP.[162] Patients in the exercise group (home exercise programme consisting of strengthening, stretching, and aerobic conditioning exercises) had improved average muscle scores after 6 weeks, as well as improved scores on the role limitation (physical) scales of the SF-36, a measure of ability to do work or daily activities.

Treatment algorithm overview

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Acute (summary)		
no significant impact on function and quality of life		
	1st	observation
significant impact on function and quality of life		
	1st	initial monotherapy: IVIG, corticosteroid, or plasma exchange
	adjunct	pharmacotherapy for neuropathic pain
	adjunct	allied health referral
partial or no response to initial monotherapy		
partial response	1st	combination therapy with 2 initial agents
	adjunct	pharmacotherapy for neuropathic pain
	adjunct	allied health referral
no response	1st	alternative initial agent
	adjunct	pharmacotherapy for neuropathic pain
	adjunct	allied health referral
refractory to combination therapy with 2 initial agents		
	1st	continue initial agent that produced partial response
	plus	addition of alternative immunosuppressant
	adjunct	pharmacotherapy for neuropathic pain
	adjunct	allied health referral

Ongoing		(summary)
response to treatment		
	1st	maintenance therapy
	adjunct	pharmacotherapy for neuropathic pain
	adjunct	allied health referral
no response to treatment		
	1st	supportive care

Treatment algorithm

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Acute

no significant impact on function and quality of life

1st **observation**

» Patients with mild disease (weakness that does not interfere with daily activities) may be monitored for deterioration without treatment.[131]

significant impact on function and quality of life

1st **initial monotherapy: IVIG, corticosteroid, or plasma exchange**

Primary options

» methylprednisolone: 500 mg intravenously once daily for 4 days every month for 6 months
Some centres may use alternative dose regimens; consult your local protocols.

OR

» dexamethasone: 40 mg orally once daily for 4 days every month for 6 months

OR

» prednisolone: 1-2 mg/kg/day orally until clinical response (usually 4-12 weeks), followed by slow taper over months, maximum 60 mg/day

OR

» normal immunoglobulin human: 2 g/kg intravenously initially given in divided doses over 2-5 days
Repeated courses may be required. Some centres may use alternative dose regimens; consult your local protocols.

» Approximately 75% to 85% of patients will respond to monotherapy with intravenous immunoglobulin (IVIG), corticosteroids, or plasma exchange.[1][58] [60] [132] [133] [134] [135]

Acute

» Corticosteroids are effective for treating CIDP.[1] [60] [134] High-dose pulsed methylprednisolone or dexamethasone are often tried first as they are easy to administer, have few adverse effects, have rapid onset of benefit, and are inexpensive.[1] [134] [136] Efficacy and improvement in disability after treatment with pulsed high-dose dexamethasone or typical daily prednisolone regimens are equivalent; however, daily oral prednisolone may cause many more adverse effects, and may take longer to show benefit.[1] [134][135] [136]

» IVIG is an effective option, with ease of use, rapid onset of benefit, and low incidence of adverse effects.[1] [59] [131] [132] [135] [137] There is no evidence for a difference in treatment efficacy between IVIG preparations.[1] [138] If efficacious, IVIG can be repeated from every 2 to 6 weeks.[1] To optimise therapy, the dose should be reduced before the frequency of administration is lowered, to achieve maximal benefits with minimal amounts of IVIG. However, care must be taken to avoid deterioration before the next dose.[1][131] [132]

» Plasma exchange is effective and relatively safe, resulting in significant short-term improvement in disability, clinical impairment, and motor nerve conduction velocity in patients with CIDP.[1] [133] However, some patients who show improvement subsequently deteriorate.[133] Plasma exchange requires good vascular access and specialised equipment, which can make it less convenient than other treatments, especially in paediatric patients.[1] [133] [139] Initial regimen is generally 5 exchanges over 2 weeks, with further dosing based on response.

» Randomised controlled trials comparing efficacy between corticosteroids, IVIG, and plasma exchange do not show a significant benefit of any one over the others.[1] [2] [135] The final decision on which treatment to use is determined by availability, risks, contraindications, and disease severity; however, because corticosteroids and IVIG are easier to administer and are generally better tolerated, they are usually recommended ahead of plasma exchange.[1] If significant adverse effects occur with an initial therapy, an alternative should be substituted.

» Corticosteroid therapy is not recommended as first-line treatment for patients with motor CIDP, due to evidence of deterioration after such therapy.[1][68] [69] [135]

Acute

adjunct **pharmacotherapy for neuropathic pain**

Treatment recommended for SOME patients in selected patient group

Primary options

» gabapentin: 300-1200 mg orally three times daily

OR

» pregabalin: 50-100 mg orally three times daily

OR

» amitriptyline: 10-150 mg orally once daily at bedtime

OR

» venlafaxine: 75-225 mg orally (extended-release) once daily

OR

» duloxetine: 30-60 mg orally once daily

Secondary options

» tramadol: 50-100 mg orally (immediate-release) every 4-6 hours when required

Tertiary options

» oxycodone: 5-15 mg orally (immediate-release) every 4-6 hours when required

» All patients with neuropathic pain should be offered symptomatic therapy.

» Based on current guidelines, pregabalin, gabapentin, tricyclic antidepressants (e.g., amitriptyline), and serotonin-noradrenaline reuptake inhibitors (venlafaxine or duloxetine) are commonly used as first-line agents.[1][157][158]

» Tramadol and other opioid analgesics (e.g., oxycodone) are recommended as second- and third-line agents, respectively, for neuropathic pain.[157] Guidance from the US Centers for Disease Control and Prevention notes that evidence for efficacy of opioid therapy for neuropathic pain is limited, and that opioids should only be considered once other options

Acute

have been tried, and if the expected benefits are anticipated to outweigh risks to the patient.[159]

» With all of these medicines, a slow upward titration of dose may help to avoid adverse effects, especially those related to sedation.

adjunct **allied health referral**

Treatment recommended for SOME patients in selected patient group

» Depending on the severity of the weakness, physiotherapy, occupational therapy, and orthotic evaluation may be needed.

» Referral to one of more of a physical medicine and rehabilitation physician, a psychiatrist or psychologist, a pain management consultant, and a podiatrist should be considered on an individual basis.[1]

» A home exercise programme under the supervision of a physiotherapist may be of benefit.[162]

» Reducing sedative use, better sleep hygiene, and treating depression may help with CIDP-related fatigue.[160] [161]

partial or no response to initial monotherapy

partial response **1st** **combination therapy with 2 initial agents**

» If there is a partial but insufficient objective response to the initial agent (i.e., daily activities are still affected by distal weakness and paraesthesias), this agent should be continued with a second initial agent added to the regimen.[1]

» Choice of combination therapy is based on availability, risks, and the severity of disease. See above for more information about treatment options (i.e., initial monotherapy: IVIG, corticosteroids, or plasma exchange).

adjunct **pharmacotherapy for neuropathic pain**

Treatment recommended for SOME patients in selected patient group

Primary options

» gabapentin: 300-1200 mg orally three times daily

OR

Acute

» pregabalin: 50-100 mg orally three times daily

OR

» amitriptyline: 10-150 mg orally once daily at bedtime

OR

» venlafaxine: 75-225 mg orally (extended-release) once daily

OR

» duloxetine: 30-60 mg orally once daily

Secondary options

» tramadol: 50-100 mg orally (immediate-release) every 4-6 hours when required

Tertiary options

» oxycodone: 5-15 mg orally (immediate-release) every 4-6 hours when required

» All patients with neuropathic pain should be offered symptomatic therapy.

» Based on current guidelines, pregabalin, gabapentin, tricyclic antidepressants (e.g., amitriptyline), and serotonin-noradrenaline reuptake inhibitors (venlafaxine or duloxetine) are commonly used as first-line agents.[1][157][158]

» Tramadol and other opioid analgesics (e.g., oxycodone) are recommended as second- and third-line agents, respectively, for neuropathic pain.[157] Guidance from the US Centers for Disease Control and Prevention notes that evidence for efficacy of opioid therapy for neuropathic pain is limited, and that opioids should only be considered once other options have been tried, and if the expected benefits are anticipated to outweigh risks to the patient.[159]

» With all of these medicines, a slow upward titration of dose may help to avoid adverse effects, especially those related to sedation.

adjunct **allied health referral**

Treatment recommended for SOME patients in selected patient group

MANAGEMENT

Acute

» Depending on the severity of the weakness, physiotherapy, occupational therapy, and orthotic evaluation may be needed.

» Referral to one or more of a physical medicine and rehabilitation physician, a psychiatrist or psychologist, a pain management consultant, and a podiatrist should be considered on an individual basis.[1]

» A home exercise programme under the supervision of a physiotherapist may be of benefit.[162]

» Reducing sedative use, better sleep hygiene, and treating depression may help with CIDP-related fatigue.[160] [161]

no response — **1st** — **alternative initial agent**

» It may take up to 3 months to determine treatment effectiveness. Lack of at least a partial response to one or two initial agents should lead to reconsideration of the diagnosis.[2]

» If there is no objective response to monotherapy with an initial agent within a few weeks to months, an alternative initial agent (i.e., corticosteroids, IVIG, or plasma exchange) should be tried. See above for more information about treatment options (i.e., initial monotherapy: IVIG, corticosteroids, or plasma exchange).

adjunct — **pharmacotherapy for neuropathic pain**

Treatment recommended for SOME patients in selected patient group

Primary options

» gabapentin: 300-1200 mg orally three times daily

OR

» pregabalin: 50-100 mg orally three times daily

OR

» amitriptyline: 10-150 mg orally once daily at bedtime

OR

» venlafaxine: 75-225 mg orally (extended-release) once daily

OR

Acute

» duloxetine: 30-60 mg orally once daily

Secondary options

» tramadol: 50-100 mg orally (immediate-release) every 4-6 hours when required

Tertiary options

» oxycodone: 5-15 mg orally (immediate-release) every 4-6 hours when required

» All patients with neuropathic pain should be offered symptomatic therapy.

» Based on current guidelines, pregabalin, gabapentin, tricyclic antidepressants (e.g., amitriptyline), and serotonin-noradrenaline reuptake inhibitors (venlafaxine or duloxetine) are commonly used as first-line agents.[1][157][158]

» Tramadol and other opioid analgesics (e.g., oxycodone) are recommended as second- and third-line agents, respectively, for neuropathic pain.[157] Guidance from the US Centers for Disease Control and Prevention notes that evidence for efficacy of opioid therapy for neuropathic pain is limited, and that opioids should only be considered once other options have been tried, and if the expected benefits are anticipated to outweigh risks to the patient.[159]

» With all of these medicines a slow upward titration of dose may help to avoid adverse effects, especially those related to sedation.

adjunct **allied health referral**

Treatment recommended for SOME patients in selected patient group

» Depending on the severity of the weakness, physiotherapy, occupational therapy, and orthotic evaluation may be needed.

» Referral to one of more of a physical medicine and rehabilitation physician, a psychiatrist or psychologist, a pain management consultant, and a podiatrist should be considered on an individual basis.[1]

» A home exercise programme under the supervision of a physiotherapist may be of benefit.[162]

Acute

» Reducing sedative use, better sleep hygiene, and treating depression may help with CIDP-related fatigue.[160] [161]

refractory to combination therapy with 2 initial agents

1st **continue initial agent that produced partial response**

» For patients whose condition does not improve sufficiently in response to a combination of two initial agents (i.e. two of corticosteroids, IVIG, and plasma exchange), re-evaluation of the diagnosis and referral to a specialist centre is appropriate.[16] [42]

» The initial agent to which a patient has shown a partial response should be continued. See above for more information about treatment options (i.e., initial monotherapy: IVIG, corticosteroids, or plasma exchange).

plus **addition of alternative immunosuppressant**

Treatment recommended for ALL patients in selected patient group

Primary options

» ciclosporin: consult specialist for guidance on dose

OR

» rituximab: consult specialist for guidance on dose

OR

» cyclophosphamide: consult specialist for guidance on dose

Secondary options

» azathioprine: consult specialist for guidance on dose

OR

» mycophenolate mofetil: consult specialist for guidance on dose

» Once the diagnosis of CIDP is confirmed, an alternative immunosuppressant should be added. Although there is limited evidence, ciclosporin, rituximab, cyclophosphamide, azathioprine, or mycophenolate may be

Acute

considered as an alternative agent after failure of initial treatments, or as add-on medication.[1] [141] [142] Any combination of initial and alternative agents can be used, and decisions should be based on severity of disease and adverse-effect profile.[1]

» Guidelines recommend against using interferon beta-1a, methotrexate, fingolimod, alemtuzumab, bortezomib, etanercept, fampridine, fludarabine, immunoadsorption, interferon alfa, abatacept, natalizumab, and tacrolimus.[1]

» In one retrospective analysis, approximately 25% of patients refractory to standard therapy showed a response to an alternative immunosuppressant; the authors noted that adverse effects should be monitored closely.[142]

» Ciclosporin is well tolerated, drug levels can be monitored to guide treatment, and it usually produces benefit within 3-6 months.[1] [141] Combining ciclosporin with plasma exchange can make it difficult to achieve therapeutic levels of ciclosporin.

» Cyclophosphamide (often in combination with a corticosteroid) may be used to treat refractory disease.[1] In one meta-analysis, cyclophosphamide yielded a response rate of 68% in patients with refractory CIDP; however, in practice, cyclophosphamide should be used with caution due to its toxicity, and patients must be monitored for adverse effects.[143]

» Rituximab may be considered in refractory CIDP or CIDP associated with other autoimmune diseases or haematological diseases (e.g. monoclonal gammopathy).[62] [141][144] [145] Rituximab may also be considered with nodal/paranodal antibodies (e.g. IgG4 anti-contactin-1 or anti-neurofascin-155 antibodies) after failure of corticosteroids or IVIG.[64] [104] [146] [147] It is recommended for children instead of cyclophosphamide because of a better adverse-effect profile.[1]

» Azathioprine is usually well tolerated.[1] [141] [148] However, onset of action may take 6-18 months, so it is not recommended for initial treatment of relapse or active refractory cases unless combined with other agents.

» Mycophenolate is used similarly to azathioprine as an additional agent when rapid

Acute

improvement is not needed.[1] Some, but not all, studies have shown benefit.[149] [150]

adjunct **pharmacotherapy for neuropathic pain**

Treatment recommended for SOME patients in selected patient group

Primary options

» gabapentin: 300-1200 mg orally three times daily

OR

» pregabalin: 50-100 mg orally three times daily

OR

» amitriptyline: 10-150 mg orally once daily at bedtime

OR

» venlafaxine: 75-225 mg orally (extended-release) once daily

OR

» duloxetine: 30-60 mg orally once daily

Secondary options

» tramadol: 50-100 mg orally (immediate-release) every 4-6 hours when required

Tertiary options

» oxycodone: 5-15 mg orally (immediate-release) every 4-6 hours when required

» All patients with neuropathic pain should be offered symptomatic therapy.

» Based on current guidelines, pregabalin, gabapentin, tricyclic antidepressants (e.g., amitriptyline), and serotonin-noradrenaline reuptake inhibitors (venlafaxine or duloxetine) are commonly used as first-line agents.[1] [157] [158]

» Tramadol and other opioid analgesics (e.g., oxycodone) are recommended as second- and third-line agents, respectively, for neuropathic pain.[157] Guidance from the US Centers for Disease Control and Prevention notes that evidence for efficacy of opioid therapy for

Acute

neuropathic pain is limited, and that opioids should only be considered once other options have been tried, and if the expected benefits are anticipated to outweigh risks to the patient.[159]

» With all of these medicines, a slow upward titration of dose may help to avoid adverse effects, especially those related to sedation.

adjunct **allied health referral**

Treatment recommended for SOME patients in selected patient group

» Depending on the severity of the weakness, physiotherapy, occupational therapy, and orthotic evaluation may be needed.

» Referral to one or more of a physical medicine and rehabilitation physician, a psychiatrist or psychologist, a pain management consultant, and a podiatrist should be considered on an individual basis.[1]

» A home exercise programme under the supervision of a physiotherapist may be of benefit.[162]

» Reducing sedative use, better sleep hygiene, and treating depression may help with CIDP-related fatigue.[160] [161]

Ongoing

response to treatment

1st **maintenance therapy**

» Maintenance therapy may require long-term use of an initial agent, an alternative agent, or a combination of two or more initial or alternative agents, depending on which treatments have produced a response in the acute phase of treatment. IVIG and corticosteroids are the most common agents used. Plasma exchange is generally not recommended long term for patients in whom immunoglobulin and corticosteroids are ineffective, because of the need for vascular access.[1]

» With a good response to an initial agent, the patient usually has up to 1-2 years of therapy (depending on agent used) with a slow taper. If there is a disease exacerbation during taper, the dose should be increased to the initial dose that resulted in a good response.[1] Some patients may require 5-15 years of treatment, but eventual withdrawal should be considered as approximately one-third of patients will go into remission and not require subsequent therapy. Thus, the need for continued maintenance therapy should be re-assessed every 6-12 months.[1]

» IVIG as maintenance treatment is associated with few adverse effects. The optimal dose and interval for IVIG maintenance treatment are not known. Once the patient is stable, treatment can be tapered based on clinical experience, by either reducing the dose or increasing treatment interval. The adjustment should be done every 6-12 months in the first 2-3 years of treatment.[1] In patients with a complete or near-complete response (distal weakness may not fully improve), IVIG should be tapered off and a trial on no immunosuppressants should be instituted.

» Subcutaneous immunoglobulin (SCIG) is approved as CIDP maintenance therapy in patients on a stable dose of IVIG. SCIG is typically given once weekly, but can be given over 2-3 days for larger doses. There is insufficient evidence that a higher dose of SCIG is superior to a lower dose, but the relapse rate was lower in the higher-dose group.[1] [151] [152] When switching from IVIG to SCIG, it is advised to use the same mean dose per week.[1] SCIG may be beneficial for patients with adverse effects or treatment-related fluctuations from IVIG that are not resolved

Ongoing

by premedication or dose adjustment.[153] SCIG can be administered at home by the patient, and eliminates problems associated with venous access. Some patients may, however, find it difficult to self-administer SCIG due to weakness; others may experience local infusion site reactions.

» Long-term corticosteroid treatment may induce significant adverse effects (e.g., osteoporosis, gastric ulceration, diabetes mellitus, cataracts, avascular necrosis of long bones, arterial hypertension). However, high-dose pulsed corticosteroids are associated with fewer adverse effects than daily oral dosing.[1] A corticosteroid taper may involve lowering the dose, reducing the frequency, or both; refer to local guidance on tapers.

» Ciclosporin, azathioprine, and mycophenolate can all be used as alternative agents to decrease the dose or frequency of corticosteroids or immunoglobulin, or when patients experience adverse effects from those therapies, although evidence of effectiveness is of low certainty.[1]

» Ciclosporin usually shows benefit within 3-6 months, while azathioprine and mycophenolate may take up to 6-18 months. They may all increase the risk of malignancy when used for >10 years. Other alternative agents should only be considered after these drugs have been shown to be ineffective.

» Due to lack of evidence of efficacy and/or adverse safety profiles, guidelines recommend against using the following agents: methotrexate, tacrolimus, interferon beta-1a, interferon alfa, fingolimod, alemtuzumab, bortezomib, natalizumab, etanercept, abatacept, fampridine, fludarabine, and immunoadsorption.[1]

adjunct **pharmacotherapy for neuropathic pain**

Treatment recommended for SOME patients in selected patient group

Primary options

» gabapentin: 300-1200 mg orally three times daily

OR

» pregabalin: 50-100 mg orally three times daily

OR

MANAGEMENT

Ongoing

» amitriptyline: 10-150 mg orally once daily at bedtime

OR

» venlafaxine: 75-225 mg orally (extended-release) once daily

OR

» duloxetine: 30-60 mg orally once daily

Secondary options

» tramadol: 50-100 mg orally (immediate-release) every 4-6 hours when required

Tertiary options

» oxycodone: 5-15 mg orally (immediate-release) every 4-6 hours when required

» All patients with neuropathic pain should be offered symptomatic therapy.

» Based on current guidelines, pregabalin, gabapentin, tricyclic antidepressants (e.g., amitriptyline), and serotonin-noradrenaline reuptake inhibitors (venlafaxine or duloxetine) are commonly used as first-line agents.[1][157][158]

» Tramadol and other opioid analgesics (e.g., oxycodone) are recommended as second- and third-line agents, respectively, for neuropathic pain.[157] Guidance from the US Centers for Disease Control and Prevention notes that evidence for efficacy of opioid therapy for neuropathic pain is limited, and that opioids should only be considered once other options have been tried, and if the expected benefits are anticipated to outweigh risks to the patient.[159]

» With all of these medicines, a slow upward titration of dose may help to avoid adverse effects, especially those related to sedation.

adjunct **allied health referral**

Treatment recommended for SOME patients in selected patient group

» Depending on the severity of the weakness, physiotherapy, occupational therapy, and orthotic evaluation may be needed.

» Referral to one of more of a physical medicine and rehabilitation physician, a psychiatrist or psychologist, a pain management consultant,

Ongoing

and a podiatrist should be considered on an individual basis.[1]

» A home exercise programme under the supervision of a physiotherapist may be of benefit.[162]

» Reducing sedative use, better sleep hygiene, and treating depression may help with CIDP-related fatigue.[160] [161]

no response to treatment

1st **supportive care**

» Absence of response to any treatment is highly unlikely. If this happens, the diagnosis should be reviewed. Nearly all patients respond, at least partially, to some kind of pharmacological therapy.

» Supportive care is the standard treatment for patients with an incomplete response.

Emerging

Antibodies against the neonatal Fc receptor

CIDP is associated with pathogenic IgG auto-antibodies. The neonatal Fc receptor (FcRn) extends the half-life of endogenous IgG by binding and transporting it back to the cell surface to prevent it from being degraded by lysosomes. Efgartigimod, a humanised IgG1-derived Fc fragment that competitively inhibits FcRn, is approved for the treatment of myasthenia gravis and is being investigated in CIDP.[163] The anti-human FcRn monoclonal antibody rozanolixizumab has been assessed in a phase 2 trial for CIDP.[164] No antibodies have yet been approved to treat CIDP.[165]

Serum neurofilament light chain

Serum neurofilament light chain (sNfL) levels are correlated with axonal damage, and so have the potential to be used as a serum biomarker of disease activity and treatment response in patients with CIDP. Studies have demonstrated that higher sNfL levels are associated with an active disease state (non-responders and patients who relapse after intravenous immunoglobulin withdrawal), and treatment-naive patients beginning induction therapy had higher levels of sNfL than those on maintenance therapy or in long-term remission without treatment. Larger studies will be needed to determine how to potentially utilise this to guide management of CIDP.[153] [166] [167] [168]

Patient discussions

Advise patients about exercise, diet, foot care, driving, and lifestyle management as required, and provide information about support groups.[1]

Physical limitations are related only to degree of disability from weakness or gait difficulty. Care must be taken to avoid falls and other injuries. Physiotherapy and occupational therapy, and orthotist and wheelchair specialist evaluations, may be appropriate.

Patients on long-term corticosteroid therapy should be advised to follow a low-fat, low-sodium diet and limit their intake of simple sugars. Patients on immunosuppressants should be made aware of their lowered immunity, the need for frequent laboratory monitoring, and that they should avoid receiving live vaccines.

FOLLOW UP

Monitoring

Monitoring

Depending on the severity of the disease, follow-up may need to be from once monthly to every 3-6 months to measure improvement in strength. Follow-up visits should address functional difficulties due to weakness or gait difficulty, as well as complications of treatment.

Repeat nerve conduction studies are usually not helpful because abnormalities may persist even after clinical improvement. However, if new signs or symptoms develop, repeat nerve conduction studies may show deterioration if there is a relapse. In addition, an increase in the total number of demyelinating features or the development of new demyelinating features following repeat studies may signal an increased risk of relapse after discontinuation of intravenous immunoglobulin.[174]

Comparisons of serial nerve ultrasound results can be useful to assess response to treatment in patients with CIDP, as a reduction in nerve size can suggest effective therapy. Patients with active CIDP that is refractory to treatment will generally have nerves that are enlarged, or nerves that have remained enlarged without significant change over time.[56] However, larger studies are needed before this approach is used routinely in management.

Patients on immunosuppressants should have routine full blood count and metabolic panels to look for leukopenia, thrombocytopenia, anaemia, electrolyte abnormalities, hepatic failure, or renal insufficiency.

Complications

Complications	Timeframe	Likelihood
respiratory failure	**short term**	**low**
Respiratory failure requiring intubation is rare and occurs in <10% of patients with CIDP. It is likely to be due to a combination of oropharyngeal and diaphragmatic weakness.[4][70] [72] Intensive care unit monitoring is warranted in cases with impending respiratory failure.		
aspiration pneumonia	**short term**	**low**
Rare, and likely to occur in people with oropharyngeal and diaphragmatic weakness or respiratory failure. Prophylactic intubation to protect the airway in patients with severe swallowing difficulties may be necessary.		
autonomic dysfunction	**variable**	**high**
Reports have highlighted mild autonomic dysfunction in up to 75% of patients with CIDP.[73] [74] However, severe dysfunction is rare and occurs in <10% of patients. Signs and symptoms can include palpitations, flushing, sinus tachycardia, urinary incontinence and urgency, diarrhoea, impotence, orthostatic hypotension, and altered sweating. Detailed autonomic testing such as R-R interval testing, Valsalva manoeuvre, tilt testing, and sympathetic skin responses may uncover mild abnormalities in patients suspected of having autonomic involvement.		
quadriplegia	**variable**	**low**
Severe weakness resulting in quadriplegia is only described rarely.[173] Long-term complications of quadriplegia include risk of deep vein thrombosis, decubitus ulcers, pain, and contractures.		

Prognosis

The majority of patients respond to therapy with improvement in strength, sensation, and gait. About 75% to 80% of patients will initially respond to intravenous immunoglobulin (IVIG), corticosteroids, or plasma exchange within the first few months of therapy.[58] [59] [60] [64] [133] However, response may be incomplete and patients may be left with residual deficit. Distal weakness and paraesthesias commonly do not resolve completely.

Several studies have looked at long-term prognosis and found that most patients continue to do well over 5-10 years, with about 75% of patients having no or only minor symptoms.[4][5] [40] [70] [71] [169]

Approximately one-third of patients will have medication-free remission. About 10% of patients have a poor outcome with either severe disability or death.

FOLLOW UP

Prognostic factors

Several studies have looked at prognostic factors.[4] [5] [71] [169] [170] Factors associated with a negative prognosis include older age, progressive course, central nervous system involvement, a high number of fibres showing active demyelination, and axonal loss on nerve biopsy. Factors associated with a positive prognosis include younger age, relapsing or subacute presentation, and proximal weakness.

Response to IVIG

A monophasic or relapsing-remitting (not chronic progressive) course and a twofold increase in cerebrospinal fluid protein typically predicts a good response to IVIG.[171] Another study showed that CIDP in patients with diabetes mellitus tends to be more severe, but has a good response to IVIG and fewer relapses than in patients without diabetes mellitus.[172]

Diagnostic guidelines

Europe

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 (https://www.ean.org/research/ean-guidelines/guideline-reference-center)

Published by: European Academy of Neurology/Peripheral Nerve Society **Last published:** 2021

Treatment guidelines

Europe

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 (https://www.ean.org/research/ean-guidelines/guideline-reference-center)

Published by: European Academy of Neurology/Peripheral Nerve Society **Last published:** 2021

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Regardless of the language in which the content is displayed, numerals are displayed according to the original English-language numerical separator standard. For example 4 digit numbers shall not include a comma nor a decimal point; numbers of 5 or more digits shall include commas; and numbers stated to be less than 1 shall be depicted using decimal points. See Figure 1 below for an explanatory table.

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This approach is in line with the guidance of the International Bureau of Weights and Measures Service.

Figure 1 – BMJ Best Practice Numeral Style

5-digit numerals: 10,000

4-digit numerals: 1000

numerals < 1: 0.25

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