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Chronic Inflammatory Demyelinating Polyradiculoneuropathy

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Synonyms and related keywords: chronic inflammatory demyelinating polyneuropathy, CIDP, chronic acquired demyelinating polyneuropathies, CADP, acute inflammatory demyelinating polyradiculoneuropathy, AIDP, Guillain-Barré syndrome, GBS, chronic inflammatory demyelinating polyradiculoneuropathy, monoclonal gammopathies, multifocal motor neuropathy, MMN, acquired purely motor neuropathy, lower motor neuron-type deficits, multifocal sensorimotor neuropathy, sensorimotor mononeuropathy multiplex, Lewis-Sumner neuropathy, sensory predominant CADP, polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, POEMS, CADP associated with diabetes mellitus, sensorimotor disorder

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Background

The term chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) has been used to identify patients with a chronically progressive or relapsing symmetric sensorimotor disorder with cytoalbuminologic dissociation and interstitial and perivascular endoneurial infiltration by lymphocytes and macrophages. In many ways, CIDP can be considered the chronic equivalent of acute inflammatory demyelinating polyradiculoneuropathy (AIDP), the most common form of Guillain-Barré syndrome (GBS).

A number of variants of CIDP have been described that have immune or inflammatory aspects and electrophysiologic and/or pathologic evidence of demyelination in common. No consensus exists on the best approach to the nomenclature of these disorders. CIDP is a major subset of chronic acquired demyelinating polyneuropathies (CADP). In this context, CIDP is considered when patients have a symmetric proximal and distal motor predominant disorder.

CIDP variants include patients with predominantly sensory symptoms, those with a distal symmetric disorder (DADS), those with multifocal sensorimotor neuropathy or sensorimotor mononeuropathy multiplex with prominent conduction block (also known as Lewis-Sumner neuropathy), and those with CIDP with associated CNS demyelination or with other systemic disorders.

The following disorders are considered distinct from CIDP because they have specific pathophysiologic features and respond to treatments differently than do patients with CIDP: Demyelinating neuropathies associated with immunoglobulin M (IgM) paraproteins, including those with anti-myelin-associated glycoprotein (MAG) antibodies; polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome; and multifocal motor neuropathy.

Pathophysiology

CIDP is presumed to occur because of immunologic antibody-mediated reaction along with interstitial and perivascular infiltration of the endoneurium with inflammatory T cells and macrophages. The consequence is a segmental demyelination of peripheral nerves.

Human leukocyte antigens Dw3, DRw3, A1, and B8 occur more frequently in patients with CIDP than in the healthy population.

Cytoalbuminologic dissociation is a characteristic finding in cerebrospinal fluid (CSF) pointing to nerve root involvement. Occasionally, CSF studies reveal mild lymphocytic pleocytosis and elevation of gamma globulin level, but this is observed most frequently in HIV-positive patients.

Frequency

International

CIDP is uncommon. The prevalence of CIDP is difficult to ascertain but estimates have ranged from 0.8-1.9 cases per 100,000 population.

Mortality/Morbidity

CIDP most commonly has an insidious onset and either chronic progressive or relapsing course. Occasionally, complete

remissions occur. Quadriplegia, respiratory failure, and death have been described but are rare.

Race

No racial predilection has been identified.

Sex

Both sexes are affected. Of CADP variants, multifocal motor neuropathy has a male predominance of at least 2:1 based on a survey of the largest case series.

Age

CIDP may occur at any age, but it is more common in the fifth and sixth decades. Relapsing course is associated with younger age of patients (third and fourth decades). CIDP has been described in childhood.

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History

CIDP most frequently starts insidiously and evolves slowly, either in a slowly progressive (more than 60% of patients) or relapsing manner (approximately one third of patients), with partial or complete recovery between recurrences.

- Periods of worsening and improvement usually last weeks or months. Patients with a younger age of onset are said to have a higher frequency of relapsing course.
- Because of the insidious onset of CIDP, documenting precipitating illnesses or events is very difficult. However, preceding infection has been reported infrequently. Both respiratory and gastrointestinal infections have been cited, but no causative organism has been identified.
- Initial symptoms include weakness of the limbs, both proximal and distal, with proximal muscles affected at least as severely as distal.
- Sensory symptoms are common, such as tingling and numbness of hands and feet, but usually motor symptoms predominate.
- Only a small proportion of patients (approximately 16%) have a relatively acute or subacute onset of symptoms, with subsequent steadily progressive or fluctuating course.
- Children usually have a more precipitous onset of symptoms.
- Most experts consider the necessary duration of symptoms to be greater than 8 weeks for the diagnosis of CIDP to be made.
- Autonomic system dysfunction can occur; in such a case, the patient would complain of orthostatic dizziness, problems with bowel and bladder functions, and cardiac problems.

Physical

Pertinent findings are limited to the nervous system, except for cases of CADP associated with other diseases, as already mentioned. Depending on the associated systemic disorder, abnormalities on physical examination may be found in multiple organ systems. Patients should be examined in detail for signs of autoimmune, inflammatory, and neoplastic conditions.

- Cranial nerves
 - Cranial nerves may be involved, particularly CN VII, with paralysis of both upper and lower facial muscles. Diplopia can occur with the involvement of CN III, IV, or VI. Rarely, bulbar muscles (eg, palate, tongue) can be affected.
 - Papilledema with pseudotumor cerebri syndrome (eg, headaches, transient visual obscurations, pulsatile tinnitus, visual field defects) are observed rarely in patients with CIDP and are due to a very high CSF protein level (usually >1000 mg/mL).
- Gait frequently is abnormal.
- Type of gait depends on location of weakness and degree of proprioceptive loss. With significant weakness in the lower extremities, patients may walk with steppage (ie, high elevation of both feet to compensate for weakness of foot dorsiflexors) or a slapping gait (ie, due to deficit of proprioception in the feet).
- Children are said to have more profound gait abnormalities than adults.
- Motor system
 - Usually relatively symmetric weakness of both proximal and distal muscles is present in upper and lower extremities.
 - Muscle tone can be normal or decreased. Hypotonia, atrophy, and fasciculations may be present.
- Deep tendon reflexes: Reflexes characteristically are diminished or absent even in regions with only mild weakness.
- Sensory system
 - Large-diameter, heavily myelinated fibers are affected most severely, leading to proprioceptive and vibratory deficits.
 - Loss or decrease of pain (ie, pinprick) and temperature sensations is less common.
 - Stocking-glove distribution of sensory deficits is typical.
 - Neuropathic pain in affected extremities may occur.
- Coordination: Patients may have sensory ataxia with positive Romberg sign due to damage to the large nerve fibers that convey proprioception.
- Pathological reflexes: Pathological reflexes (eg, Babinski, Chaddock, Oppenheim) usually are not observed in patients with CIDP.

Causes

CIDP is most frequently an idiopathic illness, but it has been known to occur with several conditions. In those cases, the associated condition is included in the main diagnosis (eg, CIDP with systemic lupus erythematosus, CIDP with HIV infection) to separate those cases from the idiopathic variety. Most reported conditions associated with CIDP are listed below.

- CIDP associated with other disorders cannot be distinguished clinically from isolated CIDP except where mentioned in this section. The disease mechanisms for all these disorders are not known. They appear to be immunologic; in some instances, antibody-mediated mechanisms have been shown to play a role.
- Vasculitis does not seem to be involved; physiologic and pathologic differences can distinguish the multifocal variants of CADP from vasculitic mononeuritis multiplex. Vasculitic neuropathies cause wallerian degeneration with minimal signs, if any, of segmental demyelination on biopsy and electromyographic studies (EMG). The

multifocal variants of CADD have prominent conduction block and slowing—hallmarks of segmental demyelination.

- HIV infection: In these patients, mild lymphocytic pleocytosis and increased gamma globulin level in the CSF are seen frequently. CIDP has been observed with early disease and later on in the course of AIDS.
- Hodgkin lymphoma: Neuropathy associated with Hodgkin lymphoma is not caused by direct infiltration of the peripheral nerves but is a consequence of the autoimmune cascade that occurs with this disease; the mechanism is not completely clear.
- Paraproteinemias and/or plasma cell dyscrasias
 - CADD is seen with monoclonal gammopathies (eg, monoclonal gammopathy of unknown significance [MGUS]), most frequently gammopathy of IgM. Evidence exists to suggest that CADD with IgM MGUS has specific clinical and electrophysiologic characteristics. Patients usually have predominance of distal sensory symptoms that are greater than motor symptoms. Conduction slowing on nerve conduction testing is accentuated in distal nerve segments. Fifty percent of patients with IgM-associated neuropathies have antibodies directed against MAG, a protein found in noncompact myelin of peripheral nerves. Whether any clinical difference exists between patients with IgM gammopathy without anti-MAG antibodies and those with anti-MAG antibodies is not clear. In both cases, the response to immunosuppressive and/or immunomodulatory treatment is poor. However, encouraging reports describe a response to rituximab, a monoclonal antibody directed against B cells.
 - Some paraproteinemias occur as isolated phenomena and some are by-products of malignant cells, as in the cases of Waldenström macroglobulinemia and myeloma. In myeloma-associated neuropathy, the abnormal paraprotein usually consists mostly of lambda light chain component. Combination of osteosclerotic myeloma, organomegaly, endocrinopathy, M protein, sensorimotor neuropathy, and pigmentary skin changes is referred to as POEMS syndrome. In POEMS syndrome, the M protein is typically immunoglobulin G (IgG).
 - The association of CIDP with IgG or immunoglobulin A (IgA) gammopathy is less clear. IgG paraproteins can occur in 5% of the population and it is unclear that the incidence of IgG paraproteins is excessive in patients with demyelinating neuropathy. Patients with CIDP and IgG or IgA paraproteins have identical clinical and electrophysiologic features to patients with CIDP and no paraprotein. Response to treatment is also the same.
- Multiple sclerosis: Reports describe CNS white matter changes in patients with CIDP. Whether a true association exists between CIDP and multiple sclerosis remains unclear.
- Systemic lupus erythematosus
- Chronic active hepatitis (B or C): CIDP associated with hepatitis should be differentiated from cryoglobulinemic vasculitis. The latter causes either symmetric distal sensorimotor polyneuropathy or mononeuropathy multiplex but on pathologic examination shows wallerian degeneration and not the segmental demyelination seen in CIDP.
- Inflammatory bowel disease: CIDP has been described in association with Crohn disease and other inflammatory bowel conditions, although no direct correlation between the 2 afflictions is known. The mechanism of development of CIDP is presumed to be an autoimmune abnormality that is also causing the primary problem in inflammatory bowel disease, although the details are not known.
- Diabetes mellitus: Whether an increased incidence of CIDP occurs in patients with diabetes mellitus remains unclear. The most recent literature has not corroborated earlier reports of an association.
- Pregnancy: Pregnancy is known to exacerbate CIDP. Worsening usually occurs in the third trimester or in the postpartum period.

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Other Problems to be Considered

Variants of CADP

Multifocal motor neuropathy
Chronic ataxic polyneuropathy
Polyneuropathy associated with monoclonal gammopathies
Multifocal sensorimotor demyelinating neuropathy with persistent conduction block (ie, Lewis-Sumner syndrome)
CIDP associated with diabetes mellitus
Chronic inflammatory sensory polyneuropathy
Hereditary Motor and Sensory Neuropathies

WORKUP

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Lab Studies

- Test CSF on all patients in whom CIDP is suspected. Protein level is increased significantly in 80% of patients (usually between 50 and 200 mg/dL, but can be higher); 10% of patients also have mild lymphocytic pleocytosis (<50 cells) and increased gamma globulin (usually associated with HIV infection).
- CBC count, sedimentation rate, antinuclear antibody, biochemistry profile, and serum and urine immunoelectrophoresis are necessary to exclude important associated systemic disorders.
- In certain instances, genetic testing may be helpful. Examples include patients with positive family history, very insidious symmetric course of the disease, or some atypical features, including lack of treatment effect. In such cases, genetic testing for Charcot-Marie-Tooth disease might be indicated. Hereditary neuropathy with predisposition to pressure palsies can be suspected and tested for in some patients. Charcot-Marie-Tooth disease type 1X and adult-onset Charcot-Marie-Tooth disease type 1B have been confused with CIDP.

Imaging Studies

- Imaging studies can provide supportive evidence of CIDP.
- MRI of the spine with gadolinium enhancement may show enhancement of nerve roots.

Other Tests

- EMG is a critical test to determine whether the disorder is truly a peripheral neuropathy and whether the neuropathy is demyelinating. Findings of a demyelinating neuropathy are as follows:
 - Multifocal conduction block or temporal dispersion of compound muscle action potential (see [Image 1](#))
 - Prolonged distal latencies and dispersion of the distal compound motor action potential
 - Variable conduction slowing to less than 70% of normal
 - Absent or prolonged F wave latencies (see [Image 2](#))

- As the disease progresses, patients tend to develop secondary axonal degeneration.
- Reports exist of a predominantly axonal neuropathy with clinical course and response to treatment similar to those of CIDP. Most cases of axonal neuropathy are not immune or inflammatory. However, some patients with an aggressive axonal neuropathy have been treated effectively with immunosuppressive and/or immunomodulatory therapy, raising the question of whether a chronic axonal inflammatory neuropathy, akin to the acute axonal variants of GBS, may be present. The relationship of these chronic axonal variants to CIDP is unclear.

Procedures

- Peripheral (sural) nerve biopsy is considered as supportive evidence of CIDP.
 - Consider biopsy for those patients in whom the diagnosis is not completely clear, when other causes of neuropathy (eg, hereditary, vasculitic) cannot be excluded, or when profound axonal involvement is observed on EMG.
 - Some experts recommend biopsy for most patients prior to initiating immunosuppressive therapy, but more recent guidelines no longer recommend biopsy.

Histologic Findings

Tissue collected on biopsy of the sural nerve may demonstrate evidence of interstitial and perivascular infiltration of the endoneurium with inflammatory T cells and macrophages with local edema. Evidence exists of segmental demyelination and remyelination with occasional onion bulb formation, particularly in relapsing cases (see [Image 3](#)). Some evidence of axonal damage also is observed, with loss of myelinated nerve fibers. The inflammatory infiltrate with neutrophil infiltration is observed in only a minority of patients.

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Medical Care

Untreated, CIDP is characterized by accumulating disability that requires physical and occupational therapy, orthotic devices, and long-term treatment. Close follow-up care with a physician knowledgeable in the field is necessary to adjust treatment.

Surgical Care

Surgical and orthopedic consultation may be required for sural nerve biopsy or in severe disease with joint deformities that require corrective surgery.

Consultations

- Consult a neurologist for evaluation, diagnosis, and treatment of CIDP and possible associated diseases.
- Consult a physical medicine and rehabilitation specialist for physical and occupational therapy and evaluation for orthotic devices.
- Other consultations (eg, internist, hematologist, oncologist, rheumatologist) may be necessary if associated diseases are present. Some treatments may be best accomplished with the assistance of a hematologist or rheumatologist.

Diet

No specific diet has been proven beneficial in CIDP.

Activity

Activity generally depends on the level of disability. Encourage physical therapy and an active lifestyle.

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If associated conditions are identified (HIV infection, lupus, paraproteinemia, lymphoma), treat them accordingly. The mainstay of treatment is immunosuppressive or immunomodulatory intervention.

Drug Category: *Immunomodulatory/immunosuppressive agents*

These agents include intravenous immunoglobulin (IVIg), plasmapheresis, prednisone, azathioprine, methotrexate, mycophenolate, cyclosporine, and cyclophosphamide. Their use is based on the proposed pathogenesis of CIDP as an immune-mediated condition.

Drug Name	Plasmapheresis
Description	Two controlled and blinded studies have confirmed benefit of plasmapheresis. Proposed mechanism is removal of antibodies and complement components that are responsible for immune-mediated damage of peripheral nerves. Plasma removed from blood through method similar to dialysis. Requires 2 large-bore needles, one to remove whole blood and other to return blood cells with albumin and saline. Patients whose veins are not large enough for repeated needle insertions have double-lumen catheter placed, either Quinton catheter (can be kept in for few weeks) or Permacath (can remain inserted indefinitely). Has been shown to have similar efficacy as IVIg in treatment of CIDP.
	Based on body weight and size to determine plasma volume; commonly, patients undergo 3 plasma exchanges per wk for first 2 wk; after that, number and frequency of treatments

Adult Dose	determined by clinical response; response tends to last for 2-4 wk and must be repeated to sustain improvement; some patients require more frequent treatment; remission is uncommon if plasmapheresis is sole therapy
Pediatric Dose	Based on body weight and size
Contraindications	Documented hypersensitivity; contraindications to permanent line placement; heart disease; cardiac risk factors; coagulation problems; hypocalcemia; sensitivity to fluid imbalance
Interactions	Pheresis removes medications that are circulating in plasma; dosing adjustments may be necessary
Pregnancy	D - Unsafe in pregnancy
Precautions	Can be classified as device or procedure related Device-related risk factors include RBC hemolysis, overheating of blood, and inaccurate delivery of anticoagulant and/or replacement fluids Procedure-related risk factors include citrate-induced hypocalcemia; replacement with fluids depleted of coagulation factors, proteins, or electrolytes can lead to problems; replacement fluids containing plasma have capacity to transmit infection; allergic reactions can lead to anaphylaxis; hemorrhage and DIC can occur; activation of coagulation, complement, fibrinolytic cascades, and/or aggregation of platelets is potential risk Problems with vascular access can be significant, such as sepsis, penetration, and thrombosis with subsequent systemic embolism; use special caution in patients with heart disease or risk factors

Drug Name	Intravenous immunoglobulin (IVIg)
Description	Multiple clinical trials establish efficacy. Solution for IV infusion that is composed mostly of heterogenous human IgG but also small amounts of IgA and IgM. Its proposed mechanism of action based on thought that IVIg contains random set of antibodies that would neutralize immune factors, causing damage to peripheral nerve in CIDP. Used in infectious diseases to provide immediate passive immunity in situations in which time constraints do not allow development of active immunity via vaccination. Also used to treat multiple immune-mediated conditions, such as idiopathic thrombocytopenic purpura, GBS, and myasthenia gravis. Activates complement cascade and provides multitude of antibodies capable of neutralization of many microorganisms, toxins, viruses, and presumably autoantibodies. Latter mechanism possibly underlies effect in CIDP. Several studies showed significant benefit in CIDP; this makes it useful alternative to plasmapheresis. On average, improvement seen by day 10 and continues through day 42. Serum half-life approximately 21-29 d. Patients usually require repeated treatments every few weeks or months to maintain remission or treat recurrences.
Adult Dose	For treatment of CIDP, total dose is 2 g/kg, IV usually divided into 5 daily doses of 400 mg/kg; effect tends to be for 2-4 wk and must be repeated to sustain improvement; remission from IVIg as sole therapy is unusual (<10%)
Pediatric Dose	Not established; 2 g/kg (as in adults) recommended
Contraindications	Absolute: IgA deficiency Relative: Documented hypersensitivity; hypogammaglobulinemia; pregnancy; breastfeeding; diabetes; cardiac disease; elderly patients; hypovolemia; maltose, sucrose, or egg hypersensitivity; renal insufficiency; sepsis

Interactions	Patients receiving IVIg are not to receive live virus vaccines for 2 wk before infusion and for 3 mo after because antibodies in IVIg can prevent immunization effect
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Hypersensitivity (anaphylactic reactions have occurred); injection site reactions are possible as well as reaction to infusion, including chills, back pain, diaphoresis, fever, hypotension, myalgias, nausea, and vomiting; premedication with diphenhydramine (25-50 mg IV 30 min before infusion) recommended Renal impairment (increased risk of renal tubular necrosis, particularly in presence of hypovolemia) Crosses placenta (in increasing amounts after 30 wk gestation) and may be excreted into milk During few weeks after IVIg therapy, some blood tests, such as complement components levels, sedimentation rate, protein electrophoresis, and serologic tests for antibodies, are not reliable

Drug Name	Prednisone (Deltasone, Orasone, Meticorten)
Description	Oral corticosteroid that suppresses inflammation and immune responses by altering protein synthesis in cells. Naturally occurring hormone that crosses cell membranes to bind to cytoplasmic receptors. Some mechanisms of action in CIDP are altering mediator function at site of inflammation and suppressing immune response. Controlled trial demonstrated efficacy of oral prednisone.
Adult Dose	Doses vary; most patients are started on prednisone at 1 mg/kg/d PO initially (60-80 mg/d) Improvement can be anticipated within next 2 mo; further on, dosing converted to alternate-day treatment and then titrated to lowest effective dose that allows maintaining patient in remission; some clinicians use weekly high-dose PO or IV corticosteroids (500 mg Solu-Medrol) instead of daily or alternate-day prednisone and believe this is at least as effective and better tolerated
Pediatric Dose	Not established; because of severity of adverse effects in children, use lowest possible dose
Contraindications	Absolute: Cushing syndrome; fungal and some viral (measles, varicella) infections Relative: Documented hypersensitivity; breastfeeding; children; acute infection; cataracts; glaucoma; coagulopathy; diabetes; infections; severe hypertension; GI problems; recent surgery or vaccination; osteoporosis; psychosis
Interactions	Significant effects occur when taken together with a few medication groups, such as antithyroid and thyroid replacement medications (eg, hyperthyroidism increases and hypothyroidism decreases prednisone metabolism), anticoagulants (prednisone can increase coagulability and thus decrease effect), barbiturates (induce hepatic enzymes), cholinesterase inhibitors, and neuromuscular blockers (acutely prednisone can induce decreased neuromuscular junction conduction), diuretics, estrogens, NSAIDs, salicylates (GI adverse effects), vaccines, and others
Pregnancy	B - Usually safe but benefits must outweigh the risks.
	Pregnancy Category B, but hydrocortisone classified as category D, probably reflecting more experience with use in pregnancy; miscarriages, stillbirth, and malformations are known to occur with use of prednisone in pregnant women; monitor infants and treat for adrenal insufficiency; precautions

Precautions	<p>in children include growth retardation, altered bone development, and multiple other adverse effects, which should lead to use in children only when absolutely necessary and to precautions and careful monitoring</p> <p>Do not stop abruptly—taper off over period of time, length of which depends on duration and dose of treatment, because of potential for withdrawal effects</p> <p>Musculoskeletal system risks include osteoporosis, fractures, and avascular necrosis (institute prophylactic measures); myopathy can occur; cataracts are known adverse effect, as is exacerbation of glaucoma (follow-up care with ophthalmologist needed in long-term use); vigorous glucose control necessary in patients with diabetes mellitus because of tendency for increased blood glucose with prednisone use; ulcers are frequent complication (use prophylactic H2 blocker); increases risk for infections</p> <p>Weight changes, variety of psychiatric adverse effects, sleep disorders, amenorrhea, and other problems encountered; patients must be aware of potential problems</p>
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Drug Name	Azathioprine (Imuran)
Description	Purine analog that decreases metabolism of purines and also may inhibit DNA and RNA synthesis. Reduces disability and symptoms of CIDP by suppressing immune-mediated damage to nerves. A small trial did not show any beneficial effect but data are insufficient to draw conclusions.
Adult Dose	Initial dose: 50 mg PO qd, increased gradually to total daily dosage of 2-3 mg/kg/d PO Therapeutic dose of azathioprine difficult to determine for each patient; some evidence suggests that elevations of RBC volume (MCV) indicate therapeutic dosing Therapeutic response may take > 6 mo to become apparent
Pediatric Dose	Not established
Contraindications	Absolute: Pregnancy and breastfeeding Relative: Documented hypersensitivity; bone marrow suppression; liver or kidney problems; infection
Interactions	ACE inhibitors cause severe leukopenia and anemia; other immunosuppressants and vaccines also cause significant interactions; carbamazepine can worsen marrow suppression
Pregnancy	D - Unsafe in pregnancy
Precautions	<p>Can lead to various GI symptoms and ulcer formation with delayed healing (use with prophylactic agent to prevent ulcers); can lead to severe leukopenia, anemia, and thrombocytopenia (strictly monitor blood counts: obtain CBC count before treatment, every 1-2 wk for first few months, then monthly; in author's practice, WBC count of 3000/μL considered warning, requiring closer monitoring of WBC counts and infection precautions; 2000/μL considered sign to stop medication)</p> <p>As immune suppressant, places patients at risk for infections</p> <p>Monitor hepatic enzymes because of risk of liver failure (same frequency as monitoring of CBC count); caution should be used if new elevation of liver enzymes up to twice normal level noted; if stopping drug brings liver enzymes back to normal, drug can be tried again at later date, although with special caution; an idiosyncratic reaction can occur within days of initiation of treatment, including fever, jaundice, nausea and vomiting, and elevation of hepatic enzymes</p> <p>Discontinuation of drug usually results in complete resolution of symptoms; restarting drug does not always result in same reaction but should be considered carefully</p>

Studies showed significant risk to fetus (on occasion benefits for mother can outweigh risk to fetus)

Drug Name	Mycophenolate (CellCept)
Description	Prodrug for immunosuppressive agent mycophenolic acid. Inhibits lymphocyte purine synthesis by inhibiting enzyme inosine monophosphate dehydrogenase. Reports of efficacy but no large controlled trials.
Adult Dose	250 mg to 3 g/d PO; adjust dose depending on clinical effect
Pediatric Dose	Not established
Contraindications	Documented hypersensitivity; bone marrow suppression; infections; GI problems; phenylketonuria; pregnancy; breastfeeding; renal failure; vaccination
Interactions	Antacids decrease bioavailability; other immunosuppressive agents can worsen marrow suppression; thrombolytic agent or anticoagulant can increase risk of bleeding; has additive GI effect with salicylates and NSAIDs; can alter efficacy of oral contraceptives and vaccines
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Can lead to various GI symptoms and ulcer formation with delayed healing (use with prophylactic agent; however, antacids impair bioavailability); can lead to leukopenia, anemia, and thrombocytopenia (monitor blood counts); as immune suppressant, places patients at risk of infections; monitor creatinine and liver enzymes (liver toxicity is much less likely than with azathioprine); effect on human pregnancy has not been studied but had teratogenic effects in animal studies

Drug Name	Cyclosporine (Sandimmune, Neoral)
Description	Cyclic polypeptide consisting of 11 amino acids; effective in many autoimmune conditions. Inhibits first phase of T cell activation and does not affect humoral immunity. By suppressing T cells, may inhibit cell-mediated nerve damage at site of inflammatory/immune reaction. Small trial showed efficacy but data still insufficient to draw conclusions.
Adult Dose	5 mg/kg/d PO divided bid initially; increase dose according to response and monitoring of trough concentrations Trough and peak levels should be monitored to register efficacy and avoid toxicity; although no definitive desirable trough level has been identified specifically for CIDP, usual trough levels utilized for immunologic disorders are between 100 and 250
Pediatric Dose	Not established
Contraindications	Absolute: Documented hypersensitivity; breastfeeding Relative: Liver, biliary tract, or kidney disease; children (since no well-controlled studies have been performed); elderly patients; females of childbearing age; infections; hyperuricemia; pregnancy; radiation treatments; vaccinations; skin abnormalities
Interactions	Significant interactions exist with multiple medications, such as ACE inhibitors (causes severe leukopenia and anemia), other immunosuppressants, vaccines, antibiotics such as aminoglycosides and vancomycin (additive nephrotoxicity), androgens (decrease cyclosporine clearance), corticosteroids, estrogens, NSAIDs (additive GI adverse effects), neuromuscular blockers (prolonged effect), diuretics, and others

Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Monitor elderly patients particularly carefully because of risk of systolic hypertension and decreased CrCl; females of childbearing age should use contraception because of effects on fetus; known to be embryotoxic and fetotoxic, causing premature birth, low birth weight, and malformations; can be used if benefit for mother outweighs risks to fetus; excreted in breast milk, and breastfeeding is contraindicated; monitoring of creatinine and liver enzymes, bilirubin, and alkaline phosphatase important

Drug Name	Cyclophosphamide (Cytoxan)
Description	Cell-cycle phase-nonspecific antineoplastic agent and immunosuppressant that acts as alkylating agent.
Adult Dose	1-2 mg/kg/d PO or monthly pulse IV; dosing based on body size used; usually a 6-mo treatment period used; dose adjusted to reduce WBC count to 2000-3000/ μ L; reports describe high-dose ablative therapy (which reduces WBC counts to 0) having efficacy in patients with otherwise severe and refractory disease
Pediatric Dose	Not established
Contraindications	Absolute: Breastfeeding; dehydration; infection Relative: Documented hypersensitivity; bone marrow suppression; dental work; gout; hyperuricemia; hemorrhagic cystitis; pregnancy; radiation treatment; vaccinations
Interactions	Other immunosuppressive agents can worsen marrow suppression; thrombolytic agent or anticoagulant can increase risk of bleeding; has additive GI effect with salicylates and NSAIDs; enzyme inducers such as phenytoin and barbiturates can increase toxic effects; multiple medications, including prednisone, can alter efficacy; can alter efficacy of vaccines
Pregnancy	D - Unsafe in pregnancy
Precautions	Hemorrhagic cystitis is serious complication, and vigorous hydration has to be instituted in all patients; can lead to various GI symptoms and ulcer formation with delayed healing (use with prophylactic agent); can lead to leukopenia, anemia, and thrombocytopenia (monitor blood counts); as immune suppressant, places patients at risk for infections Teratogenic—use in pregnancy should be greatly discouraged, particularly in first trimester Variety of other adverse effects must be monitored, and patients must be aware of them, including alopecia, allergic reactions, infertility, myocarditis and pericarditis, nephrolithiasis, and secondary malignancy

Drug Category: *Antiepileptic medications*

In patients with CIDP, a variety of medications is used for treatment of neuropathic pain. Antiepileptic medications are quite effective. The 2 most frequently used medications, gabapentin and carbamazepine, are described.

Drug Name	Gabapentin (Neurontin)
Description	Known to effect to GABA, but exact binding site unknown. Also has effects on calcium channels. Mostly used for treatment of epilepsy and neuropathic pain; 100-, 300-, and 400-mg cap and 600- and 800-mg film-coated tab are available.
Adult Dose	Starting dose depends on patient age and renal function; initial maintenance dose usually 300 mg PO tid; slow increase in dose may minimize adverse effects

	Many patients require 1800-3600 mg/d to reach therapeutic effect; adjust dose for CrCl in patients with renal failure
Pediatric Dose	Not established; may use 30-60 mg/kg/d PO tid; as in adults, begin with lowest possible dose
Contraindications	Documented hypersensitivity; children; elderly patients; pregnancy; breastfeeding; renal impairment; driving or operating machinery
Interactions	Not appreciably metabolized; does not interfere with metabolism of most medications; cimetidine mildly decreases renal excretion of gabapentin; minimally decreases level of norethindrone (oral contraceptive pill component); give at least 2 h following Maalox administration (otherwise Maalox reduces bioavailability)
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Do not discontinue abruptly because of increased risk of seizure; dose adjustment required for patients with renal failure; caution in patients with jobs demanding high alertness, in elderly patients, and with driving because of potential to cause somnolence and fatigue; most common adverse reactions are somnolence, dizziness, ataxia, nausea, vomiting, and fatigue; has been shown to cause delayed ossification of several bones in skull, vertebrae, forelimbs, and hindlimbs in rodents; because animal reproduction studies are not always predictive of human response, use during pregnancy only if potential benefits justify potential risk to fetus

Drug Name	Carbamazepine (Tegretol)
Description	Blocks use-dependent sodium channels and inhibits sustained repetitive firing as well as reduces posttetanic potentiation of synaptic transmission in spinal cord. Potent enzyme inducer that can induce own metabolism. Used as anticonvulsant and for treatment of neuropathic pain. Available in chewable 100-mg tab, in tab of 200 mg, XR tab of 100, 200, and 400 mg, and as susp of 100 mg/5 mL.
Adult Dose	400 mg PO divided bid; slowly increase in divided doses (tid for regular, bid for XR)
Pediatric Dose	10-35 mg/kg/d PO bid/qid; as in adults, use/begin with lowest possible dose
Contraindications	Absolute: Documented hypersensitivity; agranulocytosis; bone marrow suppression; cardiac conduction block Relative: Renal impairment; atonic, myoclonic, or absence seizures; barbiturate or hydantoin hypersensitivity; cardiac disease including coronary artery disease; driving or operating machinery; alcoholism; breastfeeding; pregnancy; elderly patients; glaucoma; hematologic disease; hepatic disease; hyponatremia; jaundice; neonates; psychosis
Interactions	Induces hepatic microsomal enzymes, which, in turn, accelerate carbamazepine metabolism or metabolism of other drugs; interactions between carbamazepine and other anticonvulsants are variable and complicated; can decrease effectiveness of oral contraceptive pills, phenothiazines, antidepressants, barbiturates, corticosteroids, and antiretroviral protease inhibitors; in turn, enzyme-inducing medications can alter metabolism of carbamazepine; antineoplastic agents can exacerbate hematologic toxicity
Pregnancy	D - Unsafe in pregnancy
	Monitor WBC counts closely because of risk of agranulocytosis and aplastic anemia; monitor sodium levels because of

Precautions	<p>possibility of hyponatremia and SIADH; monitor liver enzymes because of possibility of elevation with risk of hepatic failure with or without hepatitis</p> <p>Patients should report any rash immediately—toxic epidermal necrolysis, Stevens-Johnson syndrome, and exfoliative dermatitis are observed with use</p> <p>Cardiac adverse effects include arrhythmia exacerbation, worsening of preexisting block, and new conduction block</p> <p>Neurological adverse effects include ataxia, diplopia, aseptic meningitis, confusion, drowsiness, and nystagmus</p> <p>Due to teratogenicity, should be used in pregnancy only if benefits outweigh risks; has been implicated in a number of fetal abnormalities, particularly neural tube defects</p>
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Drug Category: Tricyclic antidepressants

These medications are used frequently for the treatment of neuropathic pain. The most traditionally used medication, amitriptyline, is discussed.

Drug Name	Amitriptyline (Elavil)
Description	<p>Tertiary amine TCA known to decrease reuptake of serotonin and norepinephrine. Has been used more than other newer TCAs and has more proven benefits, although other TCAs, such as desipramine and nortriptyline, are also quite potent and have fewer adverse effects.</p> <p>Tab available in 10, 25, 50, 75, 100, and 150 mg.</p>
Adult Dose	10-150 mg/d PO can be used; start at low doses hs, then increase doses depending on clinical response
Pediatric Dose	Not established
Contraindications	<p>Absolute: Documented hypersensitivity; acute MI; heart block; prolonged QT; ileus; use of radiographic contrast metrizamide</p> <p>Relative: Alcohol or substance abuse; breastfeeding; pregnancy; agranulocytosis; use of anticholinergic medications; asthma; bipolar disorder; children; elderly patients; glaucoma; diabetes mellitus; driving or operating machinery; liver disease; GI disease; hematologic problems; thyroid problems; prostatic hypertrophy; urinary retention; Parkinson disease; seizures; thrombocytopenia</p>
Interactions	<p>Interacts with multiple medications; anticonvulsants may require increased doses for adequate seizure control; clonidine can lead to dangerous hypertension; antimuscarinics may cause additive anticholinergic effects; other psychiatric medications such as hypnotics and sedatives can worsen depressant effects and lead to hypotension, somnolence, and respiratory depression; MAOIs can lead to hypertension, hyperthermia, and seizures; increases pressor effects of sympathomimetics; thyroid hormones may increase receptor sensitivity to amitriptyline</p>
Pregnancy	D - Unsafe in pregnancy
Precautions	<p>Obtain ECG and detailed cardiac history prior to beginning treatment, since drug increases risk of conduction abnormalities; ECG also is suggested when dose raised to >75 mg/d; do not use in patients with risk factors or cardiac history</p> <p>Lowers seizure threshold and may induce seizures or compromise seizure control in patients with epilepsy; monitor level of anticonvulsants</p> <p>Decreased libido, impotence, ejaculatory dysfunction, galactorrhea, and gynecomastia may occur (make patients aware)</p> <p>Use in pregnancy only when benefits clearly outweigh risks;</p>

	<p>risk of malformation, developmental delay, and withdrawal at birth</p> <p>Variety of other adverse effects must be monitored, and patients must be aware of them, including anxiety, blurred vision, sedation, urinary retention, and withdrawal symptoms after rapid cessation</p>
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Drug Name	Pregabalin (Lyrica)
Description	Thought to have similar mode of action to gabapentin. Clinical trials have shown efficacy for diabetic neuropathy and shingles induced neuropathic pain.
Adult Dose	Starting dose: 150 mg/d PO divided bid/tid Maintenance dose: 300 mg/d PO divided bid/tid
Pediatric Dose	Not established
Contraindications	Documented hypersensitivity; children; elderly patients; pregnancy; breastfeeding; renal impairment; driving or operating machinery
Interactions	Not appreciably metabolized; does not interfere with metabolism of most medications; no effect on level of norethindrone (oral contraceptive pill component)
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Do not discontinue abruptly because of increased risk of seizure; dose adjustment required for patients with renal failure; caution in patients with jobs demanding high alertness, in elderly patients, and with driving because of potential to cause somnolence and fatigue; most common adverse reactions are somnolence, dizziness, ataxia, nausea, vomiting, and fatigue; edema and weight gain may occur

Drug Name	Duloxetine (Cymbalta)
Description	Duloxetine clinical trials have shown efficacy in diabetic neuropathy pain and for depression. It is a selective serotonin and norepinephrine reuptake inhibitor.
Adult Dose	Starting dose: 30 mg PO qd Maintenance dose: 60 mg PO qd in either one or two divided doses (30 mg bid)
Pediatric Dose	Not established
Contraindications	Documented hypersensitivity; uncontrolled narrow-angle glaucoma; within 14 d of stopping MAOI use (do not initiate MAOIs within 5 d of stopping duloxetine)
Interactions	CYP1A2 and CYP2D6 responsible for metabolism; interacts with paroxetine, fluoxetine, and quinidine (CYP2D6 inhibitors); TCAs, phenothiazines, antiarrhythmics (propafenone, flecainide) may interact (CYP2D6 metabolized drugs); alcohol-CNS acting drugs; MAOIs; gastric acidity drugs
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Observe closely for clinical worsening and suicidality when initiating treatment or following dosage change; gradually decrease dose when discontinuing, do not abruptly discontinue; caution with hepatic impairment or end-stage renal disease; recommended not to prescribe to patients with substantial alcohol use or evidence of chronic liver disease; may cause slight blood pressure increase; may activate mania or hypomania; common adverse effects include nausea, dry mouth, constipation, decreased appetite, fatigue, somnolence, and increased sweating

FOLLOW-UP

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Further Inpatient Care

- Most care is delivered on an outpatient basis, although patients may have to be admitted for a short stay for the initiation of plasmapheresis or IVIg treatment, because of complications of CIDP or treatment, or for inpatient physical therapy.

Further Outpatient Care

- Outpatient care consists of visits to specialists such as neurologists and physiatrists and of treatment visits for IVIg infusions or to the plasmapheresis unit.

In/Out Patient Meds

- Most medications are administered on an outpatient basis. IVIg can be administered as a home infusion or during an outpatient visit. An exception is plasmapheresis, which requires visits to a specialized pheresis center.

Complications

- If the disease becomes severe, swallowing and breathing functions can be affected. Aspiration pneumonia, atelectasis, and respiratory failure can occur.
- If autonomic function is involved, GI motility and bladder function can be abnormal. Orthostatic hypotension and cardiac conduction defects can occur.
- As already discussed, complications of treatment also must be considered.

Prognosis

- Some have suggested that patients with the relapsing disease have a better prognosis than patients with the chronic progressive course. Approximately 70% of patients are said to make relatively good recovery from their relapses, and close to 90% of patients respond to initial immunosuppressive therapy. Some patients do not respond to the usual treatments and accumulate significant disability. Some patients have only a short treatment effect and become treatment dependent.

Patient Education

- Refer to physical and occupational therapists and to a physiatrist for optimal outpatient therapy, orthotic devices, and adaptation at home.

MISCELLANEOUS

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Medical/Legal Pitfalls

- CIDP and its variants are uncommon disorders and can be difficult to diagnose and treat. Frequently, patients with this disorder require referral to a medical center specializing in neuropathic disorders.

Special Concerns

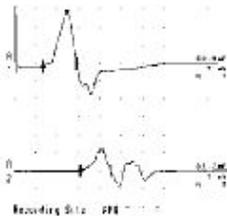
- Associated conditions need to be excluded and treated appropriately.

MULTIMEDIA

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Media file 1: [Electromyography of a patient with chronic inflammatory demyelinating polyradiculoneuropathy illustrating conduction block, temporal dispersion of compound muscle action potential, prolonged distal latencies, and slowed conduction.](#)



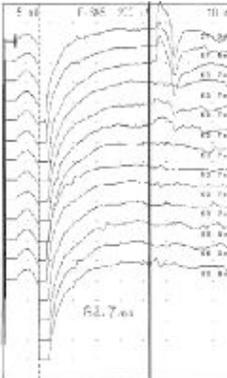
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STIMULUS SITE	STIM	CMAP	AMP	AREA
RFB wrist	8.4	2.40	10.11	1.1
RFB Elbow	15.4	7.04	25.04	1.1
RFB				

STIMULUS	CMAP	AMP	AREA
RFB wrist	20	6	0.0102
RFB wrist	200	6	0.0102
RFB Elbow			

Media type: Image

Media file 2: [Prolonged F wave latencies \(normal is <31\).](#)



[View Full Size Image](#)

Media type: Image

Media file 3: [Electron micrograph of the peripheral nerve of a patient with chronic inflammatory demyelinating polyradiculoneuropathy. Note "onion bulb" formation in the myelin sheath of the nerve fibers due to continuous demyelination and remyelination. Courtesy of A. Sima, MD, Department of Pathology, Wayne State University.](#)



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Media type: Image

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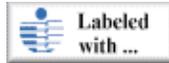
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