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To:

Compassionate Allowances Program Office
Social Security Administration

From:

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Proposed Condition Name

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

Alternate Names

- Chronic Relapsing Polyneuropathy
 - Chronic Polyradiculoneuritis
 - Immune-Mediated Demyelinating Polyneuropathy
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Summary

CIDP is an immune-mediated peripheral neuropathy characterized by progressive or relapsing weakness and sensory loss in both proximal and distal muscles, typically evolving over at least eight weeks [1,2]. The condition is caused by chronic inflammation and demyelination of peripheral nerves and nerve roots, leading to slowed conduction velocity and impaired nerve signal transmission.

CIDP is considered the chronic counterpart of Guillain-Barré Syndrome (GBS), but with a longer course, frequent relapses, and potential for severe long-term disability if untreated [2,3]. Early recognition and treatment are critical to prevent irreversible axonal loss.

Description of Condition

The pathophysiology of CIDP involves autoimmune-mediated damage to myelin sheaths and Schwann cells of peripheral nerves [2]. This demyelination causes weakness, sensory impairment, and reflex loss, often symmetrically and involving both upper and lower extremities.

Unlike GBS, CIDP symptoms evolve slowly or in a stepwise fashion over more than eight weeks and may persist or recur for years [1,3].

Diagnostic Testing

Clinical Diagnosis:

- Symmetric weakness in both proximal and distal muscles of upper and lower limbs
- Areflexia or hyporeflexia
- Symptom progression >8 weeks [1]

Confirmatory Testing:

- **Nerve Conduction Studies (NCS) / EMG:** Marked slowing of conduction velocities, prolonged distal latencies, conduction block, temporal dispersion [2,3]
- **Lumbar Puncture:** Elevated cerebrospinal fluid (CSF) protein with normal or slightly elevated white cell count [1]

Supportive Imaging/Testing:

- MRI of nerve roots or plexus (may show hypertrophy or gadolinium enhancement)
- Nerve biopsy in atypical cases (shows segmental demyelination/remyelination, onion bulb formations) [3]

Physical Findings

- Symmetrical proximal and distal weakness
- Loss or reduction of deep tendon reflexes
- Sensory deficits (vibration, proprioception, touch)
- Gait instability, positive Romberg sign
- In severe cases: muscle atrophy, foot drop, or claw hand deformity [2]

ICD-10 Code

- **G61.81** — Chronic inflammatory demyelinating polyneuritis

Onset

Typically, insidious onset over weeks to months; may follow infections, vaccinations, or autoimmune triggers. Some cases occur without a clear precipitant [1].

Course / Progression

- **Progressive Form:** Steadily worsening over months to years without remission.
- **Relapsing-Remitting Form:** Episodes of worsening followed by partial recovery.
- **Monophasic Form:** Single episode lasting >8 weeks followed by stabilization.

Without treatment, CIDP often leads to severe disability, including inability to walk, perform daily activities, or maintain employment [3,4].

Treatment

First-Line Therapies:

- Intravenous immunoglobulin (IVIG)
- Corticosteroids (oral or pulse therapy)
- Plasma exchange (plasmapheresis) [3]

Maintenance / Immunosuppressive Therapy:

- Azathioprine, mycophenolate mofetil, cyclosporine, rituximab in refractory cases [2,4]

Rehabilitation:

- Physical and occupational therapy to maintain mobility and function
- Orthotic support for foot drop or hand weakness

Rationale for Compassionate Allowance

- **Chronic, disabling condition** with significant impairment in ambulation and self-care
- **Objective diagnostic criteria** with confirmatory testing available [1,3]
- **Severe socioeconomic impact** due to prolonged disease course and frequent relapses

- **Treatment urgency**—delayed care leads to permanent nerve damage [2,4]

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