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**To:**

Compassionate Allowances Program Office  
Social Security Administration

**From:**

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## Proposed Condition Name

**Autoimmune-Mediated Small Fiber Neuropathy**

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## Alternate Names

- Autoimmune Small Fiber Neuropathy (SFN)
  - Immune-Mediated Small Fiber Neuropathy
  - Immune-Related Painful Polyneuropathy
  - Idiopathic Small Fiber Neuropathy (when autoimmune cause suspected)
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## Summary

Autoimmune-mediated small fiber neuropathy (SFN) is a peripheral nerve disorder caused by immune-mediated injury to small myelinated (A $\delta$ ) and unmyelinated (C) fibers responsible for pain, temperature sensation, and autonomic functions [1,2]. This results in chronic neuropathic pain, sensory loss, and dysautonomia. SFN can be idiopathic or secondary to autoimmune diseases such as Sjögren's syndrome, sarcoidosis, systemic lupus erythematosus, or post-infectious immune activation [3,4].

In autoimmune forms, inflammatory and antibody-mediated mechanisms damage small fibers, sometimes in association with systemic autoantibodies [2,5]. This condition frequently leads to significant impairment of mobility, sleep, autonomic stability, and daily functioning.

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## Description of Condition

Autoimmune SFN is characterized by selective injury to small-caliber sensory and autonomic nerve fibers [1,2]. Damage disrupts pain and temperature perception, as well as autonomic control of heart rate, sweating, and gastrointestinal motility [3].

Symptoms often include burning pain, allodynia, hyperalgesia, distal sensory loss, and autonomic symptoms such as orthostatic intolerance, abnormal sweating, and gastrointestinal dysmotility [4,5].

Without effective treatment, symptoms can be persistent, progressive, and disabling. Even with treatment, many patients experience chronic pain and functional limitations [6].

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## Diagnostic Testing

### Neurological Examination:

- Preservation of reflexes and muscle strength with distal sensory abnormalities [1,3].

### Objective Confirmation:

- **Skin biopsy:** Intraepidermal nerve fiber density (IENFD) reduction on immunohistochemical staining (gold standard) [1,4].
  - **Quantitative sensory testing (QST):** Demonstrates impaired thermal and pain thresholds [2].
  - **Autonomic function testing:** Heart rate variability, sudomotor testing (QSART), tilt-table testing [3].
  - **Laboratory evaluation:** Screening for autoimmune markers (ANA, SSA/SSB, anti-ganglionic AChR antibodies), ESR/CRP [4,5].
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## Physical Findings

- Distal burning pain, tingling, or numbness (often stocking-glove distribution)
  - Allodynia (pain from normally non-painful stimuli)
  - Hyperalgesia (exaggerated pain response)
  - Loss of pinprick and temperature sensation
  - Autonomic instability: orthostatic hypotension, abnormal sweating patterns, GI dysmotility, urinary dysfunction [2,4]
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## ICD-10 Codes

- **G62.89** — Other specified polyneuropathies
  - **G62.9** — Polyneuropathy, unspecified
  - **G90.09** — Other idiopathic peripheral autonomic neuropathy
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## Onset

Autoimmune SFN may present **acutely, subacutely, or insidiously**, with symptom onset over days to months [3]. It can occur at any age but is most often diagnosed in adults between 30–60 years [1,4].

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## Course / Progression

- **Early:** Burning pain, paresthesia, intermittent autonomic symptoms [2].
  - **Intermediate:** Persistent pain, loss of temperature sensation, more pronounced autonomic involvement [4].
  - **Chronic:** Worsening sensory loss, severe dysautonomia, mobility limitations, and secondary depression/anxiety due to pain [5,6].
  - Many cases are progressive without immunotherapy, and even treated patients may have incomplete recovery [6].
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## Treatment

### Acute/Immunomodulatory:

- Corticosteroids (e.g., prednisone, methylprednisolone) [5]
- Intravenous immunoglobulin (IVIG) [2,5]
- Plasma exchange (for severe refractory cases)

### Chronic/Symptomatic:

- Neuropathic pain agents: gabapentin, pregabalin, duloxetine, tricyclic antidepressants [3]
  - Autonomic symptom management: midodrine, fludrocortisone, dietary modifications for orthostatic intolerance
  - Physical therapy for gait/balance support
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# Rationale for Compassionate Allowance

- **Severe functional impairment:** Neuropathic pain and autonomic instability frequently prevent gainful employment [2,4].
  - **Objective diagnosis available:** Confirmed by skin biopsy and autonomic testing [1,4].
  - **Chronic course with poor prognosis:** Many patients have lifelong disability despite treatment [5,6].
  - **Rapid recognition important:** Early access to benefits can support patients in obtaining needed medical care and adaptive equipment.
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