JAMA Diagnostic Test Interpretation

# Electrodiagnostic Testing for Diagnosing Polyneuropathy

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**A 50-year-old woman** with a history of seizures presented to the electromyography laboratory with progressive lower extremity pain and gait imbalance for approximately 5 years. A neurological examination demonstrated reduced pinprick sensation in both feet and absent vibratory sensation at the first toes. Mild weakness of ankle dorsiflexion and absent ankle jerk reflexes were noted bilaterally. She had a slightly wide-based gait, high-arched feet, and hammer toes. She had no known family history of polyneuropathy and no history of diabetes, alcohol misuse, known vitamin deficiency, hypothyroidism, or HIV. Nerve conduction studies (NCS) and needle electromyography (EMG) were performed. The results are shown in **Table 1** and **Table 2**.

**HOW DO YOU INTERPRET THESE TEST RESULTS?**

**A.** Results are consistent with axonal polyneuropathy

**B.** Results are consistent with demyelinating polyneuropathy

**C.** Results are consistent with polyradiculopathy

**D.** Results are normal

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**Table 1. Sensory and Motor Nerve Conduction Study Results**

| Nerve (recording site) | Peak latency, ms | Amplitude, μV | Segments | Distance, cm | Velocity, m/s |
|---|---|---|---|---|---|
| **Sensory nerve conduction studies** | | | | | |
| Left radial: anatomical snuff-box | | | | | |
| Forearm (wrist) | 2.60 | 8.6 | Forearm to wrist | 10 | 55 |
| Normal | ≤2.90 | ≥15.0 | | | ≥49 |
| Left sural: ankle | | | | | |
| Calf (ankle) | Absent | Absent | Calf to ankle | 14 | Not applicable |
| Normal | ≤4.20 | ≥5.0 | | | ≥41 |
| **Motor nerve conduction studies** | **Latency, ms** | **Amplitude, mV** | **Segments** | **Distance, cm** | **Velocity, m/s** |
| Left ulnar: abductor digiti minimi | | | | | |
| Wrist | 2.66 | 9.0 | Wrist to ADM | 7 | |
| Normal | ≤3.60 | ≥5.0 | | | |
| Below elbow | 6.35 | 8.1 | Below elbow to wrist | 19 | 51 |
| Normal | | ≥5.0 | | | ≥49 |
| Left peroneal: extensor digitorum brevis | | | | | |
| Ankle | 6.41 | 0.3 | Ankle to EDB | 8 | |
| Normal | ≤6.20 | ≥2.0 | | | ≥41 |

Abbreviations: ADM, abductor digiti minimi; EDB, extensor digitorum brevis.

**Table 2. Needle Electromyography Test Results**

| | | | Spontaneous activity | | | Voluntary motor units | | | |
|---|---|---|---|---|---|---|---|---|---|
| Muscle | Nerve | Roots | Insertional activity | Fibrillations | Positive waves | Amplitude | Duration | Polyphasic | Recruitment pattern |
| Left tibialis anterior | Deep peroneal | L4-L5 | Normal | None | None | 3+ | 2+ | No | Reduced (mild) |
| Left first dorsal interosseous | Ulnar | C8-T1 | Normal | None | None | 3+ | 1+ | No | Reduced (moderate) |

## Answer

**A.** Results are consistent with axonal polyneuropathy

## Test Characteristics

Electrodiagnostic testing typically includes NCS and needle EMG. NCS involves application of an electrical stimulus to the skin above a nerve and use of surface electrodes to record waveform responses from individual sensory and motor nerves. The action potential generated after electrical stimulation represents the number of fibers depolarized. The amplitude, distal latency, and conduction velocity of an action potential help distinguish whether a neuropathy is axonal or demyelinating. Needle EMG involves

placement of a small recording needle electrode into a muscle to evaluate for abnormal insertional activity or spontaneous activity upon needle insertion and to assess voluntary motor units with volitional activation of the muscle. EMG can help distinguish neurogenic or myopathic weakness.[1]

Definitive assessment of the sensitivity and specificity of NCS for detection of polyneuropathy is difficult due to the lack of a criterion standard. A study of 313 patients with confirmed polyneuropathy 1 to 6 years after NCS testing reported that surface sural NCS recordings had a sensitivity of 49% and specificity of 100%.[2] Medicare reimbursement for electrodiagnostic testing, including both NCS and EMG, typically ranges from $190.48 to $371.26.[3] However, the extent of electrodiagnostic testing depends on a patient's distribution of symptoms, so costs may vary.

## Application of Test Results to This Patient

Electrodiagnostic testing may be used to diagnose polyneuropathy, which is typically characterized by distal and symmetric abnormalities of sensory and motor nerve function. Polyneuropathy affects approximately 15% of individuals in the US older than 40 years, with diabetes being the most common cause.[4] Electrodiagnostic testing can help delineate whether a neuropathy is axonal, which is characterized by direct injury to and loss of axons, or demyelinating, which involves damage to the myelin sheath surrounding nerve fibers. Axonal polyneuropathy typically results in low-amplitude or absent sensory or motor nerve responses on NCS findings, whereas demyelinating polyneuropathy presents with prolonged latencies or slowed conduction velocities. Certain abnormalities on needle EMG results in patients with polyneuropathy may suggest active or chronic denervation associated with the neuropathy.[5] An EMG test may also be helpful to evaluate for radiculopathy.[6]

Results of the NCS showed that the patient had an absent sural sensory response and decreased amplitudes of the peroneal motor and radial sensory responses (Table 1). These abnormalities are consistent with a severe sensory and motor polyneuropathy, characterized by axonal loss rather than demyelination, because the predominant abnormality is the low amplitude of the affected nerves rather than slowed conduction velocities. The patient's polyneuropathy appeared length-dependent, meaning the nerve endings in the feet were affected before the hands, as shown by the results of sural sensory and peroneal motor studies being substantially more abnormal than the ulnar motor and radial sensory studies. Results of the needle EMG showed changes in voluntary motor unit morphology and a reduction in the recruitment pattern of the voluntary motor units in the first dorsal interosseous and tibialis anterior muscles, consistent with a severe degree of chronic reinnervation. The patient's electrodiagnostic test findings, coupled with her abnormal foot appearance, raised the clinical suspicion for an underlying hereditary neuropathy.

## What Are Alternative Diagnostic Testing Approaches?

If the diagnosis of polyneuropathy is highly likely based on a patient's medical history (eg, diabetes, chemotherapy exposure), electrodiagnostic testing may not be necessary because it will not change the clinical management. If Charcot-Marie-Tooth disease or another hereditary sensory and motor neuropathy is strongly suspected, a patient with polyneuropathy may directly undergo genetic testing instead of proceeding first with electrodiagnostic testing.

## Patient Outcome

Due to her severe axonal polyneuropathy and abnormal foot morphology in the absence of other common causes for polyneuropathy, genetic testing for Charcot-Marie-Tooth disease was performed, which revealed a pathogenic variant in *MFN2* (Mitofusin 2). Variants in this gene are a common cause of axonal Charcot-Marie-Tooth disease[7-9] and may result in a mild and late-onset presentation of sensory and motor neuropathy.[10] Obtaining the diagnosis of Charcot-Marie-Tooth disease is important for appropriate genetic counseling, symptomatic treatment, and avoidance of additional, unnecessary tests.

### Clinical Bottom Line

- Electrodiagnostic testing consists of both nerve conduction studies and needle electromyography. The protocol for testing depends on the distribution of symptoms and neurological examination.
- Electrodiagnostic testing can determine the presence and severity of a polyneuropathy and whether it is primarily axonal or demyelinating.
- Electrodiagnostic testing may elucidate less common causes of polyneuropathy, such as hereditary neuropathies.


### ARTICLE INFORMATION

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