INTRODUCTION

In the clinical setting of a suspected peripheral neuropathy, the history and neurologic examination provide information about general features but cannot define the nature of the pathologic changes and the clinical features may underrepresent the distribution of nerve involvement or time course. Electrodiagnostic tests allow for a more detailed characterization of a neuropathy. Electrodiagnostic tests include nerve conduction and needle electromyographic (EMG) studies. Reliable interpretation of the data assumes recognition of potential technical issues and overall must be within the context of the clinical information. A systematic electrodiagnostic approach is based on an understanding of basic nerve anatomy and physiology and how pathologic changes affect electrodiagnostic data, and this foundation will be reviewed. Electrodiagnostic findings in prototypic axonal and demyelinating neuropathies are presented. These principles are put together as a diagnostic strategy to aid in planning informative tests and interpreting outside studies.

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PERIPHERAL NERVE ANATOMY

A nerve consists of individual nerve fibers of different types bundled together and can be divided along several lines: somatic and autonomic fibers, motor and sensory fibers, large and small fibers. Each fiber consists of an axon insulated by segments of myelin, which is thick and tightly wrapped for large myelinated fibers and thin and loosely wrapped for small unmyelinated fibers. The functional and electrodiagnostic implications of different nerve fiber diameters and their degree of myelination is varied in nerve fiber conduction velocities. Myelinated fibers have faster velocities as a result of saltatory conduction (30–60 m/s), whereas unmyelinated fibers conduct relatively slowly (<1 m/s). Routine nerve conduction studies assess exclusively larger myelinated nerve fibers, as the contributions from smaller myelinated and unmyelinated fibers to the recorded signal are by comparison minimal. Special tests can assess these fibers but are not commonly performed and rarely help with characterization of common neuropathies.

Named (large) peripheral nerves contain all fiber types (large and small fiber somatic, autonomic, and motor and sensory nerves) before branching in the end organs, where somatic motor and sensory nerves separate and are accessible for study. Sensory nerves contain 3000 to 6000 fibers. Motor nerves frequently innervate a group of muscles, such as the thenar or hypothenar groups, and 100 to 300 motor nerve fibers innervate muscle(s) in the groups. However, each motor axon branches into 500 to 800 terminal branches within a muscle and each branch innervates a muscle fiber.

Another anatomic feature of motor nerves is the motor unit, which is the axon and its terminal branches. Terminal branches and the muscle fibers they innervate are distributed over a circular area within a muscle with a diameter of 5 to 10 mm. With more than 100 motor units innervating a muscle, the territories of approximately 20 motor units overlap at any given site in the muscle.

PRINCIPLES OF NERVE CONDUCTION PHYSIOLOGY

During nerve conduction studies, the entire nerve is electrically activated. Sensory and motor responses are recorded separately by the position of electrodes over sensory nerves or over muscles. The sensory response is recorded as the sensory nerve action potential (SNAP) and the motor response as the compound muscle action potential (CMAP). The SNAP represents the sum of single nerve fiber action potentials. The SNAP waveform shape is determined by the arrival times of nerve fiber action potentials, the duration of the action potentials, and the degree of phase cancellation. Nerve fiber action potential waveforms are biphasic (negative-positive) with a duration of approximately 1 millisecond. Sensory nerves conduct over a range of approximately 25 m/s. The arrival of slower conducting nerve action potentials positions their negative peaks at the time of the positive portions of the faster conducting action potentials, resulting in significant phase cancellation and a lower SNAP amplitude than might be expected from the simple algebraic addition of the nerve fiber action potentials (Fig. 1). With greater conduction distances, the effect of phase cancellation is marked and the SNAP amplitude decreases greatly with longer conduction distances. The shape of the CMAP is affected by the same elements. However, motor nerves activate muscle fibers whose action potentials are approximately 6 milliseconds in duration. Further, motor nerves conduct over a smaller range of conduction velocities, approximately 15 m/s, and the slower arriving muscle fiber potentials are minimally affected by phase cancellation (see Fig. 1). Accordingly, the CMAP amplitude is higher and decreases little with longer conduction distances.
Another difference between sensory nerve fibers and muscle fibers is their diameters: sensory nerve fibers that contribute to the SNAP have diameters of 8 to 12 \( \mu \text{m} \), whereas muscle fibers that contribute to the CMAP have diameters of 30 to 70 \( \mu \text{m} \). The amplitudes of individual action potentials are proportional to the diameter of the nerve fibers.

Fig. 1. Effects of normal temporal dispersion on the SNAP and the CMAP. **Left,** Actual waveforms from ulnar nerve study (stimulating at the wrist, A1, and up the arm to Erb point, A5) are shown for sensory (A) and motor (B) nerve conduction. **Right,** Model of summation of action potentials. Sensory nerve action potentials have an approximately 1-millisecond-duration waveform and conduct over an approximately 25 m/s range, leading to marked phase cancellation and relative low amplitude when nerve fiber waveforms are summed in the SNAP. Muscle fiber action potentials have an approximately 6-millisecond-duration waveform and conduct over an approximately 15 m/s range, leading to much less phase cancellation and a relative high amplitude when muscle fiber waveforms are summed in the CMAP.
fiber; SNAP amplitude (normal range) is 6 to 90 μV, whereas CMAP amplitude (normal range) is 2000 to 15,000 μV, or 2 to 15 mV.

Nerve stimulation is accomplished by passing current between anode and cathode electrodes (which are 3–4 cm apart) on the skin over the nerve. Current flows from the anode to the cathode with hyperpolarized beneath the anode and depolarization under the cathode (the site of impulse initiation). With sufficient current, all axons are activated to produce a maximal SNAP or CMAP response. The cathode must be placed closest to the recording site, for if the anode is placed closest, 2 situations occur. Because the distance between the electrodes is 3 to 4 cm, the intended measured distance between recording and stimulating electrodes will be approximately 3 to 4 cm greater, leading to artificially longer distal latencies or slowed conduction velocities. The other is the phenomenon of anodal block that can cause partial conduction block along the nerve caused by hyperpolarization of some axons under the anode, leading to artificially lower SNAP and CMAP amplitudes.

Nerve conduction studies yield 2 types of metrics: the amplitude of the evoked response and a set of timing measures (Fig. 2). SNAP and CMAP amplitudes are roughly proportional to the number of axons conducting between the stimulating and recording electrodes. With progressive axonal loss, both responses lose amplitude but the CMAP amplitude drops less than the SNAP amplitude because of the effects of collateral reinnervation of motor nerves (to be discussed later). Timing measures generally reflect activity of the fastest myelinated fibers and include distal latency (conduction over set distances), F-wave latency (conduction over twice the whole length of motor nerves), and conduction velocity (over segments of nerve). The duration of the CMAP waveform (negative peak duration) provides an estimate of the spectrum of more slowly conducting myelinated fibers.

Nerve fiber conduction velocity is slowed at lower temperatures in a linear manner. However, the effects of temperature are more apparent with sensory than with motor nerves. With lower nerve temperatures, sensory nerve action potentials are longer in duration, resulting in less phase cancellation and larger SNAP amplitudes. Although muscle fiber action potentials also lengthen in duration, the effect on CMAP amplitude is negligible. For both, the lower nerve temperature slows conduction, approximately 2 m/s/°C. Correction factors can be applied when limb temperature is low, but the factors are approximate and diagnostic interpretation uncertainties can be avoided by warming limbs to approximately 31°C.

![Fig. 2. CMAP waveform metrics, applicable primarily to the CMAP.](image-url)
**PRINCIPLES OF NEEDLE EMG**

Needle EMG records electrical activity from muscle fibers to assess the integrity and architecture or arrangement within the muscle. The electrical motor unit recorded by the EMG needle is called the motor unit action potential (MUAP) and represents only a portion of the anatomic motor unit, because the electrical uptake area of the electrode is less than 1 to 2 mm in diameter. Thus, the MUAP includes only about 7 to 15 fibers of an anatomic motor unit, for both concentric and monopolar electrodes. The MUAP waveform varies with slight needle movement based on the intimate proximity of the needle tip to 1 to 3 muscle fibers of the anatomic motor unit that contribute most to the waveform amplitude and shape.

The EMG study is performed in 2 stages. The first stage is assessment for the presence of abnormal spontaneous muscle fiber activity when the muscle is at rest, and the second is assessment of MUAPs during voluntary activation of the muscle. The presence of abnormal spontaneous activity is assessed at high EMG screen display sensitivity or gain, 20 to 50 μV per division. The presence of positive waves and fibrillation potentials indicates denervation and, in the context of a peripheral neuropathy, denervation from axonal loss (neurogenic denervation). Note that the waveform differences between positive waves and fibrillation potentials depend on the relationship between the electrode tip and the spontaneously active muscle fiber and thus are of equal clinical significance. The presence of very low amplitude abnormal spontaneous activity, less than approximately 50 μV, suggests very long-standing and slowly progressive denervation.

There are about overlapping 20 motor units in the electrode’s uptake area, and during voluntary muscle activation the number of active motor units increases from zero at rest toward the 20 with increasing effort. Of note, most routine EMG studies are performed at low levels of activation so that individual MUAPs can be observed. Accordingly, during low levels of activation, the electrode records from 3 to 5 MUAPs close to the needle. At low levels of muscle activation, the recruitment pattern of MUAPs can be assessed for MUAP discharge frequencies. The EMG screen display sensitivity or gain is set at 200 μV per division and sweep speed at 10 milliseconds per division. If the screen has 10 horizontal divisions, the beam sweep time will be 10/s: thus, it is rare for a single MUAP to consistently discharge at 20 Hz (twice/sweep), and observation of several MUAPs discharging at rates greater than 20 Hz support motor unit loss. MUAP waveforms can be inspected for increased amplitude by determining if they exceed 2000 to 3000 μV, an approximate upper limit of normal for distal muscles. MUAP complexity in terms of polyphasia (>4 phases) or polyturn (>5 turns) can be assessed by a trigger and delay line, which permits individual MUAPs to be electronically captured and spread out. It is to be noted that up to 10% of motor units can be polyphasic and excessive emphasis should not be placed on occasional complex motor units.

The goal of these observations is to estimate pathophysiologic changes. In the context of a neuropathy, abnormal spontaneous activity supports denervation of muscles. The remaining motor nerve fibers sprout collateral branches to reinnervate orphaned muscle fibers and the anatomic motor increases in muscle fiber density but not in area. Reinnervated muscle fibers no longer discharge spontaneously, but abnormal spontaneous activity usually persists for 2 reasons. There is subsequent ongoing denervation from the neuropathy and the degree of collateral reinnervation is limited, all of which leads to permanently orphaned (denervated) muscle fibers. With reinnervation, the increased density of the anatomic motor unit is reflected in larger and more complex MUAPs. In neuropathies that are progressing relatively
rapidly, there will be ongoing denervation, marked by abnormal spontaneous activity, mildly decreased recruitment, and complex MUAPs. In very slowly progressive neuropathies, there is sufficient time for maximal reinnervation; positive waves and fibrillation potentials from permanently orphaned muscle fibers will be of very low amplitude; and the motor units will be few in number and of very high amplitude, but not complex, because there has been sufficient time for polyphasia and polyturns to simplify. Thus, when MUAPs are observed whose amplitudes are 5 to 10 times normal (10,000–20,000 mV), consideration should be given for a hereditary neuropathy, the most slowly developing form of neuropathy. The extent of denervation (distal-proximal gradient) can be assessed by identifying the most proximal muscle with abnormal needle EMG findings.

LIMITS OF NORMAL

As with most biologic data, “normal limits” are derived from distributions of values from subjects who have no apparent disease. The pool of subjects should include a wide spectrum of ages and body sizes (height). How limits of normal are set is controversial, but most laboratories use upper and lower limits (ULN of distal latency and F-wave latency, LLN for amplitude and conduction velocity) set at 2 to 3 SDs, but data for the different nerve conduction metrics may not be normally distributed and other limits such as confidence intervals may be more appropriate. Although it is preferable for each laboratory to gather its own normal data, this is rarely done, and most limits are handed down. An obscure origin of a laboratory’s normal values seems unscientific, but values are generally similar. Some metrics are influenced by body habitus and, although difficult to take into account precisely, should be part of the general considerations. Thin fingers in women can result in very high amplitude digital SNAP amplitudes, and vice versa for thick fingers. Conduction velocities can be reduced and F-wave latencies in particular can be long in very tall individuals, and vice versa in very short patients. Thus, ULN and LLN should be viewed as references and not absolute values and should be interpreted in the overall clinical context. Nerve conduction metrics in the elderly are especially challenging, and although good sural responses can be obtained in the very elderly, absent responses may not be abnormal.

ELECTRODIAGNOSTIC MANIFESTATIONS OF NEUROPATHY

Nerve conduction tests help in distinguishing 3 basic conditions of peripheral nerves. The first state is normal conduction, seen when most nerve fibers and axons are intact. The second situation is axonal injury, seen when the primary injury occurs to axons. The third case is loss of myelin, seen with demyelination, which generally occurs at multiple focal sites along a nerve. This causes variation in nerve action potential propagation resulting in slowing of conduction velocities or slowing to zero (conduction block). Slowed conduction velocity causes abnormal dispersion of arrival times of nerve action potentials at the recording electrode (abnormal temporal dispersion). Of note, there can be mixed patterns with primary demyelination and secondary axonal loss. Finally, there may be slowed conduction from reversible metabolic causes with no obvious damage to myelin.

Normal Conduction

SNAP and CMAP waveforms are influenced by normal temporal dispersion: SNAP amplitude decreases markedly over greater distances, whereas CMAP amplitude decreases minimally and the CMAP negative peak duration increases only minimally.
Normal conduction is defined as values within the laboratory limits of normal. Distal latency, F-wave latency and conduction velocity are influenced by limb length and limb temperature.

**Axonal Injury**

Loss of axons disconnects fibers from their receptors (sensory nerves) or muscles (motor nerves). Not all fibers may be affected, and the remaining unaffected fibers conduct normally, or all fibers may be affected in severe neuropathies. Generally, axonal loss occurs at the distal ends of fibers, a process called "axon dying-back." This results in reductions in both SNAP and CMAP amplitudes. The SNAP amplitude is especially sensitive to axon loss because of the lack of compensatory collateral reinnervation. Essentially, there is a linear relationship between the number of axons lost and the amplitude of the SNAP, and with loss of greater than 50% of axons, the amplitude may be unrecordable. The CMAP amplitude, in contrast, is less sensitive to early axonal injury because of the supportive effects of collateral reinnervation. With mild denervation, collateral reinnervation preserves the number of innervated muscle fibers and the CMAP amplitude remains high, but with either further axonal loss or a very rapid rate of loss, greater numbers of muscle fibers remain denervated and the CMAP amplitude decreases. The effects of collateral sprouting can, in some cases, maintain CMAP amplitude at greater than the LLN until greater than 80%
of axons are lost. When the CMAP is within normal limits, the needle EMG can detect the effects of motor unit enlargement and verify that axonal loss has occurred.

With axonal loss, each remaining nerve fiber conducts at its innate speed. Thus, measures of timing (distal latency, F-wave latency, and conduction velocity) are reduced only to the extent of loss of large axons (see Fig. 3). The limits of slowing in axonal neuropathy can be empirically assessed by reviewing data from patients with amyotrophic lateral sclerosis (ALS), a disorder characterized by reduced numbers of axons with no predilection to axon size: distal latency and F-wave latency are rarely longer than 125% of the ULN, and conduction velocity is rarely slower than 75% of the LLN.11 Temporal dispersion, as measured by negative CMAP peak duration, is largely unaffected in axonal neuropathies. Overall, with moderate to major axonal loss, the SNAP response will be absent and thus provide no information about conduction velocity.

**Demyelination**

Damage to myelin affects nerve conduction at multiple sites along nerve fibers and nerve roots, resulting in varying degrees of slowed conduction in affected fibers. Thus, distal latency and F-wave latency are prolonged, and conduction velocity is slowed. The increased variability of nerve fiber conduction velocities results in greater degrees of phase cancellation within the SNAP and CMAP waveforms, causing low-amplitude responses (see Fig. 3). The sites of demyelination are generally not uniformly distributed along the length of a nerve, and thus the effects of multifocal demyelination can best be demonstrated by measurements over longer nerve conduction distances. Abnormal temporal dispersion (and secondary axonal loss) can markedly reduce SNAP amplitude, frequently to zero, and thus motor responses are more robust and more commonly used to assess conduction velocity and abnormal temporal dispersion. Conduction of individual nerve fibers may be infinitely slowed (conduction block), which can contribute to the reduction in the response amplitude. The degree of temporal dispersion can be measured in the CMAP waveform by the ratio of the amplitude or area of the proximal to the distal response (P:D ratio); however, the P:D ratio can also be affected by conduction block of axons between the 2 stimulation sites (Fig. 4). A more direct measure of temporal dispersion is the CMAP negative peak duration value, comparing values from the proximal to the

![Fig. 4](image_url). Changes in actual CMAP waveforms comparing normal with slowed conduction velocities (20 m/s), highlighting differences between acquired primary demyelinating neuropathy (eg, CIDP) and hereditary neuropathy (eg, Charcot-Marie-Tooth type 1). Waveforms in CIDP show effects of abnormal temporal dispersion, whereas in Charcot-Marie-Tooth, the waveforms show effects of uniform slowing and normal temporal dispersion.
distal response; normally, the negative peak duration increases by less than 10% even over long distances (CMAP from Erb point stimulation compared with wrist stimulation; see Figs. 1 and 4).

There is one neuropathy that is an exception with slowed conduction velocity without abnormal temporal dispersion. In Charcot-Marie-Tooth type 1, the CMAP amplitude does not markedly decrease between distal and proximal stimulation sites (see Fig. 4). This is because of uniform changes to myelin along nerve fibers. Conduction can also be mildly slowed by metabolic causes that do not affect myelin structurally, and diabetic neuropathies frequently show this pattern.

There have been many efforts to design sets of nerve conduction criteria that can distinguish between primary axonal and primary demyelination in terms of the degree of slowing. The degree of demyelination in such neuropathies is variable: when mild, there will be only small prolongations in distal and F-wave latency and slowing of conduction velocity that may be within (or just beyond) normal limits. When demyelination is more marked, there will be slowing greater than expected for the degree of axonal loss, and the amount of slowing can be referred back to timing values from patients with ALS. The published criteria are complex to apply, and a more simple set of guidelines, based on the degree of slowing in ALS, is given in Table 1. These guidelines are not intended to be strict or exclusive but to assist deciding whether there is a demyelinating component in a neuropathy.

**Focal Conduction Block**

Conduction block refers to blockage of a large number of axons over a short segment of nerve. Focal conduction block occurs most frequently at sites of entrapment (median nerve at the wrist, carpal tunnel syndrome; ulnar nerve at the elbow, tardy ulnar palsy; peroneal nerve at the knee) resulting in mononeuropathies. In the context of peripheral neuropathies, focal conduction block is sought at sites unaffected by entrapment. The pathologic condition may represent focal demyelination (or alteration to myelin at the node of Ranvier) or block by other mechanisms such as alteration or block of membrane ion channels. Focal conduction block can be demonstrated by showing loss of CMAP amplitude across the site of block and normal amplitudes along nerve segments above and below the site. The blocking mechanism can be very specific with block only of motor axons with sensory axons.

**Table 1**

<table>
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<tr>
<th>Electrodiagnostic guidelines that can be applied to help identify primary demyelination show greater slowing than expected for the degree of axonal loss for motor nerves</th>
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<td>Distal latency</td>
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<td>Conduction velocity</td>
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<td>F-wave latency</td>
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<td>CMAP negative peak duration (measured at return to baseline of last negative peak)</td>
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<td>P:D CMAP negative peak duration ratio</td>
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Recording 2 or more values beyond limits raises the question of a primary demyelinating neuropathy. Limits of slowing based on data from patients with pure axonal neuropathy (ALS).

unaffected (multifocal motor neuropathy with conduction block)\textsuperscript{15} or with block of both motor and sensory fibers (Lewis-Sumner syndrome).\textsuperscript{16} Although the electrodiagnostic hallmark of conduction block is reduction in the CMAP amplitude or area across the site of block, the effects of abnormal temporal dispersion can also produce CMAP amplitude reductions as a result of increased phase cancellation that may be misinterpreted as conduction block. Pure focal conduction block has been defined as a greater than 50% reduction of CMAP area (area is less affected by abnormal temporal dispersion than is amplitude) across the site of block without an increase in negative peak CMAP duration (<20\%).\textsuperscript{17}

**DESIGNING AN ELECTRODIAGNOSTIC STUDY**

Patients may be initially seen in a clinic, where a detailed history is obtained and a neurologic examination is performed. However, patients are frequently referred to the EMG laboratory for electrodiagnostic tests only. Under these circumstances, a brief and focused history and examination are required to properly design and interpret the studies.\textsuperscript{3}

**Patient History**

A pertinent history and examination allow for a rational selection of electrodiagnostic tests.\textsuperscript{18} Referrals may be for complaints that do not accurately represent the true clinical issue. Important topics to review and specific points to clarify include (1) time course: acute, chronic, or insidious; (2) symptoms: sensory (including numbness or pain) or motor (including cramps, fasciculations, or weakness); (3) distribution: distal legs, distal and proximal limbs, or single nerve; (4) medical history: such as diabetes or collagen vascular diseases; (5) family history: family members with neuropathy, high arches, or hammertoes; and (6) medications: neurotoxic chemotherapeutic drugs or others. A focused neurologic examination should confirm the clinical suspicion for the presence and distribution of sensory loss and weakness. Tendon reflexes are helpful because they are usually absent, at least distally (Achilles reflex), in axonal neuropathies and more diffusely absent or reduced in demyelinating neuropathies.

**Study Design**

At the conclusion of the clinical evaluation, there should be a good notion of the nature of the neuropathy. The electrodiagnostic study should be designed to confirm what is expected from the history and answer questions that may not be evident from the history and examination. Specific issues include (1) type of nerves involved (motor, sensory, or both); (2) degree of involvement (mild, moderate, or severe); (3) anatomy (which nerves are involved); (4) underlying pathologic conditions (axonal, demyelinating, or both); and (5) time course (acute, ongoing, or slowly progressive).

It is argued here that there is benefit to designing an electrodiagnostic study test by test for each patient, as opposed to applying a predetermined study protocol. With the former approach, data from each nerve or muscle should be predicted. If the predicted result is confirmed, the process of confirming the clinical diagnosis is proceeding on course. If there is an unexpected result, a technical error is sought or the clinical diagnosis is questioned and reviewed. With the latter approach, data are reviewed at the end of the study. As such, it may be hard to make clinical sense of disparate findings, and technical errors may escape detection at a time when they could be corrected.


**Study Strategies**

**Sensory nerve conduction**
Assess the most distal sensory nerve first, usually the sural or superficial peroneal nerve. Most peripheral neuropathies have a greater effect on sensory than motor nerves and distal than proximal nerves. If the response is absent, to exclude a technical cause, study the contralateral nerve. This will document symmetry, although the history and examination will support symmetry clinically without the need to document symmetry electrodiagnostically for every nerve. If sensory responses are absent in the legs, a sensory nerve in the arm should be assessed for the overall extent (distribution) of axonal loss, and it is reasonable to choose the ulnar instead of the median sensory nerve, because it will not be affected by common focal conduction abnormalities at the wrist.

**Motor nerve conduction**
Assess the longest motor nerve next, usually the peroneal motor recording from the extensor digitorum brevis, before the tibial nerve; the tibial nerve is less informative because the amplitude is normally relatively large and small reductions resulting from axonal loss will not be detected. If motor responses are absent in the legs, motor nerves in the arms should be studied, and when a primary demyelinating neuropathy is suspected, the ulnar nerves should be tested with stimulation up to the axilla. Motor nerves will provide information about primary axonal versus primary demyelinating neuropathies by inspection of the distal latencies, F-wave latencies, and conduction velocities and the presence of abnormal temporal dispersion. The amplitude of the tibial response stimulating at the knee usually shows a marked loss of amplitude, and the P:D amplitude ratio from this nerve is not a reliable measure of abnormal temporal dispersion or conduction block. The F-wave response should be assessed after 10 maximal stimuli for minimum latency. Response amplitudes and timing values should be compared for fulfillment of guidelines for primary demyelination (see Table 1) when suggested by the history and examination.

**Needle EMG**
The needle EMG study will provide information on the presence of subtle denervation, proximal extent, and chronicity. Assessing the anterior tibialis muscle is useful because it is usually affected to a mild degree and hence provides full information, whereas the extensor digitorum brevis muscle represents a very distal muscle that may be very atrophic. Assessment for abnormal spontaneous activity is to determine whether there is mild motor nerve axonal loss, especially when the CMAP is mildly reduced. Assessment of recruitment and MUAP amplitude and complexity is helpful to determine the time course: moderately reduced recruitment and moderately increased MUAP amplitude and complexity support ongoing denervation, whereas markedly reduced recruitment, markedly increased MUAP amplitude, but simple waveforms support a very longstanding neuropathy.

**Electrodiagnostic Report Summary and Interpretation**
The electrodiagnostic report should contain sufficient historical and examination features to provide a context for the interpretation. A summary should not repeat in prose nerve conduction and EMG data tables but should present the salient points that will be used in the overall interpretation, in particular technical or physical issues that influence the data. The overall interpretation can include information from the history and examination and must make sense internally (data internally consistent with the conclusions) and clinically (consistent with examination findings, past medical
and family histories, and laboratory findings). Overall, the interpretation can be most informative if the study is designed to answer questions formulated from the history and examination with assessment of each metric to determine consistency.

REVIEWING OUTSIDE ELECTRODIAGNOSTIC STUDIES

When reviewing an outside electrodiagnostic study, one should not rely on the interpretation but instead review the data and make your own conclusions and then compare the 2 interpretations. It must be kept in mind that there may be technical issues not recognized and inappropriate interpretations and numbers, and waveforms should be reviewed when provided. In reading the nerve conduction and EMG data tables, it is most efficient and informative if one reads the study as if one were performing the study; that is, look at the data in the order suggested earlier (steps 1–3).

CLINICAL AND ELECTRODIAGNOSTIC FEATURES OF PROTOTYPIC NEUROPATHIES

Primary Axonal Neuropathies

This represents the most common group of polyneuropathies (Fig. 5). Axonal neuropathies are typically slowly progressive but also may have an insidious onset that suggests a hereditary neuropathy. The distribution of axonal loss is length dependent with longest nerves affected first and progression to shorter nerves over time. Clinically, patients describe either a loss of function with reduced or loss of sensory perception and poor balance or positive symptoms with discomfort in the feet. Over time, these symptoms progress proximally as a stocking unrolls, and in the extreme can involve upper extremity sensation as a long glove unrolls, and occasionally in a shieldlike distribution across the abdomen. Foot muscle atrophy is evident in the thinness of the feet, and in hereditary forms, with elevation of the arch and hammering of the toes. Weakness of toe movements and ankle dorsiflexion may not be

Fig. 5. Primary axonal neuropathy (primary axonal neuropathy). The distribution of sensory loss is demonstrated, with mild involvement in legs in stocking distribution loss, more severe involvement in legs and arms in stocking-glove distribution loss, and very severe involvement with shield distribution loss.
Tendon reflexes are usually absent, at least at the ankle. Electrodiagnostic findings are greatest in distal leg nerves and may not be evident early on in distal arm nerves. SNAP amplitude is decreased earlier and to a greater degree than CMAP amplitude. Distal latency, F-wave latency and conduction velocity are mildly affected (see Figs. 3 and 5) and values are within 75% of the LLN or 125% of the ULN. Needle EMG will be more sensitive to mild degrees of denervation when the CMAP amplitude is within the LLN, and the greatest EMG changes will be in most distal muscles. In very chronic forms, such as hereditary neuropathies, there will be expected absent SNAP responses and absent or low amplitude CMAP responses indistinguishable from other causes, but the clue to chronicity will be in the EMG result, which will show markedly reduced motor unit recruitment with very large amplitudes but simple waveforms in distal muscles. Clinical and electrodiagnostic findings are usually symmetric in axonal neuropathies of any cause.

**Primary Demyelinating Neuropathies**

This group is less common but potentially treatable. The distribution of demyelination is diffuse with multiple foci along nerves and nerve roots, and thus both distal and proximal nerves are affected clinically and electrically (Fig. 6). Clinically, patients describe numbness and tingling with a distal emphasis but also along the proximal portion of the limbs. Weakness also has a distal emphasis but can be demonstrated in proximal muscles. The time course can be rapid (within days), as in acute inflammatory demyelinating polyradiculoneuropathy (AIDP), or slow (over months), as in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP). Tendon reflexes are usually uniformly absent, but at least absent in the legs.

**Fig. 6.** Primary demyelinating neuropathy (primary demyelinating neuropathy). The distribution of sensory loss resulting from multifocal demyelination involving nerve and spinal root fibers is shown. Sensory loss involves both legs and arms, both distally and proximally, with distal predominance.
Electrodiagnostic findings are greatest when longer lengths of nerve are studied, and abnormalities are found in leg and arm nerves but more severely in leg nerves. SNAP amplitudes may be markedly reduced and frequently absent because of distal abnormal temporal dispersion and conduction block. CMAP amplitude is usually reduced to distal stimulation and may be markedly reduced to more proximal stimulation caused by multifocal demyelination and conduction block (see Figs. 3 and 4). Distal and F-wave latencies are prolonged and conduction velocities slowed. With relatively severe primary demyelinating neuropathies, these values will be markedly slowed, greater than seen in axonal neuropathies, but with more mild degrees of severity, values may overlap with the limits of primary axonal neuropathies. This represents a challenge if electrodiagnostic criteria are not met. A practical approach is to consider the possibility of a demyelinating neuropathy when one or more nerves has values supporting slowing greater than expected for primary axonal neuropathy (see Table 1). This may result in false positive demyelinating neuropathies with erring on the side of a possibly treatable neuropathy.

**Mixed Primary Axonal and Primary Demyelinating Neuropathies**

Several neuropathies may contain a mixture of demyelination and axonal loss. When demyelination is the primary process, axonal loss occurs frequently as a secondary pathologic process. Thus, absent responses in more severe demyelinating neuropathies may be in part the result of secondary axonal loss. Most examples of primary demyelinating neuropathies, AIDP and CIDP, include varying degrees of secondary axonal loss, and responses can be absent in leg nerves. The needle EMG will help determine the degree of axonal loss with the presence of abnormal spontaneous activity and the degree of reduced MUAP recruitment and complexity. Diabetes and uremia are the most common causes of mixed axonal and demyelinating neuropathy, and findings are reduced SNAP and CMAP amplitudes, similar to findings in primary axonal neuropathies, but the slowing of conduction velocities is not as substantial as expected in a primarily demyelinating neuropathy.

**Focal Conduction Bock Neuropathies**

Multifocal motor neuropathy with conduction block is characterized by focal conduction block over short nerve segments away from entrapment sites and in a mononeuropathy multiplex pattern. There is a potential technical issue in identify this neuropathy in that there may be inadequate activation of axons at the proximal stimulation site leading to a falsely low CMAP response. There is a physiologic issue in that mild reductions in amplitude to proximal stimulation can occur from abnormal temporal dispersion leading to greater phase cancellation, mimicking conduction block. This can be assessed by considering the negative peak duration values: in conduction block, there will be a greater than 50% loss of CMAP amplitude with a less than 15% increase in negative peak duration. In classic multifocal motor neuropathy, sensory conduction across the site is normal, but in the Lewis-Sumner syndrome, there is involvement of sensory nerves at sites of motor block.

**SUMMARY**

Electrodiagnostic testing follows from the history and is an extension of the neurologic examination. When clinical assessment implicates a peripheral neuropathy, the goal of electrodiagnostic testing is to more fully characterize the neuropathy in terms of the distribution (motor, sensory, symmetric, or asymmetric), extent of a neuropathy (symmetric, legs, or arms), and time course (very chronic or ongoing). Of greatest
importance is that electrodiagnostic testing should help identify the type of underlying pathologic condition (primary axonal, primary demyelinating, or conduction block). Once this is accomplished, the differential diagnosis narrows and rational laboratory testing can be ordered or treatment trials initiated.1

REFERENCES