Chapter 8
Electrodiagnostic Testing

Jeffrey A. Allen

Abbreviations

CMAP	Compound muscle action potential
CRD	Complex repetitive discharges
DRG	Dorsal root ganglion
EMG	Electromyography
Fibs	Fibrillation potentials
MUAP	Motor action unit potential
NCS	Nerve conduction studies
PSW	Positive sharp waves
SNAP	Sensory nerve action potential

Key Points

- A good electrodiagnostic study starts with a specific question and evolves as new information that is uncovered during the test.
- Failure to adequately warm a patient during NCS may result in erroneously slow conduction velocities or prolonged distal latencies.
- Although small motor and sensory amplitudes on NCS typically represent axon loss, other explanations include severe muscle or neuromuscular junction injury, distal conduction block, or submaximal nerve stimulation.
- In conduction block, both amplitude and area are reduced, whereas in temporal dispersion amplitude drops, but area is relatively preserved, and duration is prolonged. Both suggest focal nerve demyelination.
- Sensory responses are measured in μV and motor responses are measured in mV. The smaller sensory responses are technically more challenging to accurately record and require greater attention to detail in order to minimize external electric noise.

J.A. Allen (✉)
University of Minnesota Medical Center, Department of Neurology, Minneapolis, MN, USA
e-mail: jaallen@UMN.edu

D. Walk (ed.), *Clinical Handbook of Neuromuscular Medicine*,
https://doi.org/10.1007/978-3-319-67116-1_8

- Late response abnormalities are not specific for proximal nerve injury, but if distal recordings are normal and late responses are abnormal, the presumption is that the lesion is localized to a proximal segment.
- Dorsal nerve root lesions proximal to the dorsal root ganglion may cause sensory symptoms but have normal sensory nerve conduction studies.
- NCS assess large motor and sensory fibers. Polyneuropathies that affect only small fibers (small fiber polyneuropathy) may have normal nerve conduction studies.
- Reduced amplitude NCS usually indicate axon loss. Slow CV, prolonged latency, CB, and temporal dispersion usually indicate demyelination.
- Referencing available electrodiagnostic demyelinating guidelines can provide clarity when amplitude reductions and CV slowing are both present in a single nerve.
- End plate noise and spikes are normal and should not be confused with other pathologic forms of spontaneous activity
- Fibrillation potentials and positive sharp waves can be seen with both neurogenic and myopathic disorders that result in denervation or muscle membrane instability.
- Fasciculation potentials may be normal, benign, or part of a peripheral nerve disorder but are not seen in primary myopathic conditions.
- Large, long, polyphasic MUAPs are classically seen in chronic neurogenic disorders.
- Short, brief, polyphasic MUAPs are classically seen in myopathies.
- Reduced recruitment – neurogenic
- Early recruitment – myopathic

Introduction

Electrodiagnostic testing (nerve conduction studies (NCS) and electromyography (EMG)) is an essential neuromuscular tool [1]. Often called "an extension of the physical exam," NCS/EMG can help clinicians localize lesions, characterize pathophysiology, understand injury severity, and assess chronicity. The intent of this chapter is to highlight abnormalities encountered during routine electrodiagnostic testing and provide the clinician with a framework for how the findings might be interpreted.

Nerve Conduction Studies/Electromyography: Overview

A typical NCS/EMG study starts with a question. Often the referring provider will raise concern over a particular condition, e.g., neuropathy or ALS (Table 8.1). The electrodiagnostic physician uses this referral information and integrates it with a focused clinical history and physical examination taken at the time of the study.

Table 8.1 Common referral questions encountered during NCS/EMG testing

Localization	Disorder
Myopathy	Inflammatory myopathy
	Muscular dystrophy
	Muscle channelopathy
Neuromuscular junction	Myasthenia gravis
	Lambert-Eaton myasthenic syndrome
Focal neuropathies	Median nerve at the wrist
	Ulnar nerve at the elbow
	Peroneal nerve at the knee
Polyneuropathy	Length-dependent polyneuropathy
	Generalized polyneuropathy
	Mononeuropathy multiplex
	Sensory ganglionopathy
Nerve root	Cervical radiculopathy
	Lumbosacral radiculopathy
Plexopathy	Brachial plexopathy
	Lumbosacral plexopathy
Anterior horn cell	Amyotrophic lateral sclerosis
	Spinal muscular atrophy

This information is critical when determining the most appropriate nerves and muscles to be tested. NCS/EMG is a dynamic evaluation. Once the study begins, the choice of nerves and muscles tested evolves based upon the clinical question and in response to electrodiagnostic findings as they emerge [2].

Nerve Conduction Studies

The integrity of the peripheral nervous system can be explored with nerve conduction studies. Surface stimulation of a nerve with a small electrical impulse results in nerve depolarization, action potential generation, and propagation of action potentials in two directions from the point of stimulation. Recordings can then be made of the amplitude and latency of either action potential as it passes beneath a surface recording electrode. In the case of motor conduction studies, recordings can be made of the electrical activity generated by depolarization of muscle cells innervated by the stimulated nerve. Results of the NCS reflect the integrity and function of the myelin sheath and the axon.

Each patient evaluation may include the following recorded NCS responses:

- Motor
- Sensory
- Late responses (F wave and H reflex)

Nerve Conduction Study Procedures

Before the testing begins, the clinician should ensure that the examination room is clean, all supplies needed for the examination are readily available, and the patient is counseled on the procedures anticipated during the test. Temperature of the patient's hand, foot, or other involved areas should be recorded with a goal temperature of ≥32 °C in the upper extremity and ≥31 °C in the lower extremity. If needed, warming can be performed by submerging the affected limb in a warm water bath. *Inadequate warming will result in artifactually slow conduction velocities and a misdiagnosis of neuropathy* [3].

Electrodes are placed on the skin to record the response from a desired nerve. Electrical stimulation is delivered to the nerve so that the response can be recorded by the electrodes (Fig. 8.1). Current is initially provided at low submaximal levels (5 mA or less) and slowly increased until supramaximal stimulation is achieved, that is, until the amplitude of the response no longer increases with increases in stimulus intensity, indicating that all axons available have been depolarized. The number of stimuli should be minimized when possible. Evoked responses can be measured for latency, amplitude, duration, and conduction velocity (Table 8.2; Fig. 8.2).

Motor Nerve Conduction Studies

Motor NCS are performed by stimulating a motor nerve at one or more points along its course and recording the resulting compound muscle action potential (CMAP) with surface electrodes over the belly of the corresponding muscle. Each CMAP is a composite of all the muscle fiber action potentials of the nerve/muscle pair in question.

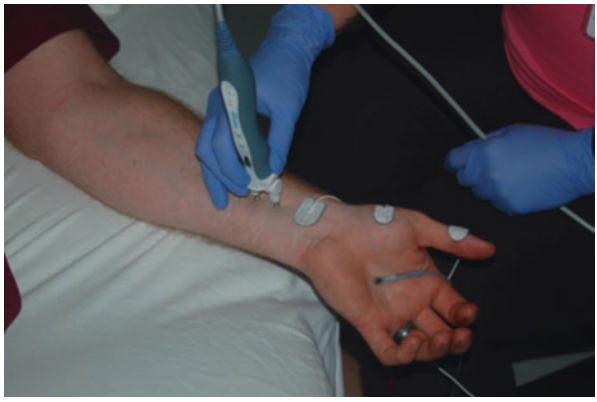

Fig. 8.1 Median motor nerve conduction study. Active recording electrode is over the abductor pollicis brevis muscle on the thenar eminence

Table 8.2 Parameters assessed during nerve conduction studies

Parameter	Unit	Measurement	Represents
Latency	ms	Distal latency: stimulus onset to initial response baseline deflection	Speed of fastest conduction fibers
		Peak latency: stimulus onset to midpoint of negative peak	Speed of slower conduction fibers
Amplitude	μV or mV	Baseline to negative peak	Number of muscle fibers or sensory fibers that depolarize
Conduction velocity	m/s	Calculated by dividing distance traveled by conduction time	Speed of fastest conduction fibers
Duration	ms	Initial baseline deflection to first baseline crossing (negative peak)	Synchronicity of muscle fiber depolarization

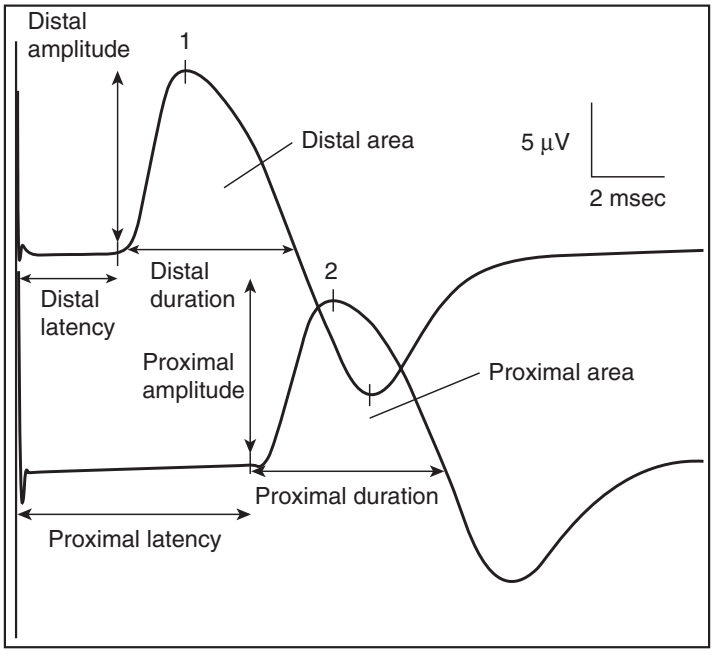

Fig. 8.2 Principal components of the CMAP. Conduction velocity is calculated by dividing distance traveled by conduction time (latency) between the onset of the proximal and distal responses

CMAP amplitude reflects the number of activated action potentials. A reduced CMAP amplitude indicates one of the following:

- Axon loss
- Distal conduction block
- Severe neuromuscular junction transmission failure
- Myofiber atrophy
- Submaximal nerve stimulation

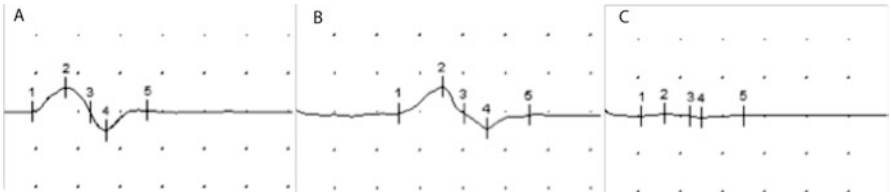

Fig. 8.3 (**a**) Normal motor conduction study (MCS) after distal stimulation; (**b**) MCS demonstrating prolonged latency due to demyelination; (**c**) MCS demonstrating reduced amplitude due to axon loss

While the CMAP itself does not indicate which of these is the reason, it can usually be inferred accurately from the findings of EMG, discussed below, and the clinical context. Because axonal neuropathy is far more common than the other pathologies listed, axon loss is the most common reason for low CMAP amplitude.

Latency reflects the speed of the fastest conducting nerve fibers (Fig. 8.3). Because latency measurements also include the time required for neuromuscular transmission and muscle fiber activation, latency measurements are not true assessments of velocity. For that reason, motor nerve conduction responses are typically obtained from at least two separate points along the nerve, and the motor nerve conduction velocity is then calculated using the distance between the points. Prolonged distal latency and slowed conduction velocity are findings that typically indicate demyelination, although mild slowing, typically >70% of the lower limit of normal, can also be seen with loss of the fastest-conducting axons [4].

Other important findings on motor NCS are *conduction block* and *temporal dispersion*. *Conduction block and temporal dispersion are findings that usually are indicative of an acquired demyelinating process*.

Conduction block is the failure of an action potential to conduct over a segment of nerve. Electrically this is demonstrated by showing a reduction in the recorded CMAP amplitude and area in a proximal stimulation site when compared to a distal stimulation site. While there is normally a small degree of amplitude attenuation with proximal stimulation, in most cases, pathologic conduction block is suspected when there is a reduction in proximal CMAP amplitude compared with distal CMAP amplitude by >30% and certainly when >50% in the absence of temporal dispersion, which is discussed below (Table 8.3) [5]. The degree of conduction block reflects the proportion of individual axons in which conduction is blocked.

Temporal dispersion refers to a pathologic increase in the duration of the CMAP. While normally the CMAP duration is modestly greater with proximal stimulation than distal stimulation, pathologic temporal dispersion reflects the fact that pathologic demyelination or remyelination can result in conduction velocity slowing without frank conduction block. When this occurs in all nerves to an equal degree, one would expect the CMAP duration to be unchanged but the conduction

Table 8.3 Electrophysiological criteria for conduction block

Conduction block	Definition
Definite[a]	Negative peak CMAP area reduction on proximal vs. distal stimulation of at least 50% whatever the nerve segment length (median, ulnar, and peroneal). Negative peak CMAP amplitude on stimulation of the distal part of the segment with motor CB must be >20% of the lower limit of normal and >1 mV and increase of proximal to distal negative peak CMAP duration must be ≤30%
Probable[a]	Negative peak CMAP area reduction of at least 30% over a long segment (e.g., wrist to elbow or elbow to axilla) of an upper limb nerve with increase of proximal to distal negative peak CMAP duration ≤30%
Probable[a]	Negative peak CMAP area reduction of at least 50% (same as definite) with an increase of proximal to distal negative peak CMAP duration >30%

Used with permission © John Wiley and Sons 2010 [5]
CMAP compound muscle action potential, *LLN* lower limit of normal
[a]Evidence for conduction block must be found at sites distinct from common entrapment or compression syndromes

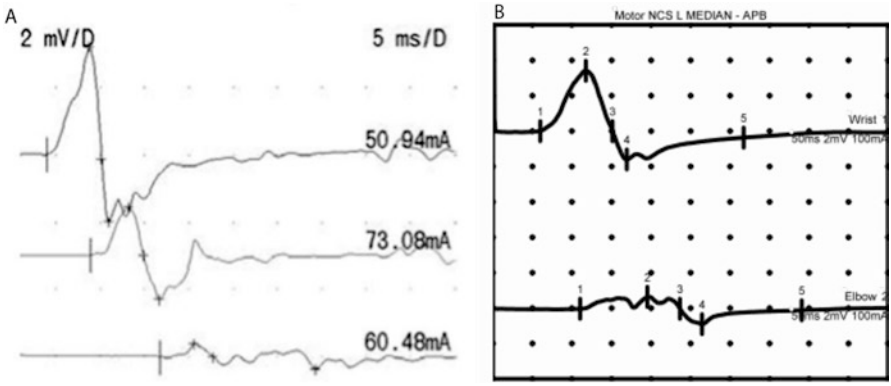

Fig. 8.4 Conduction block and temporal dispersion. (**a**) >50% amplitude decrement between the distalmost (top trace) and proximal (middle trace) stimuli, indicative of conduction block. In the most proximal (bottom trace) stimulus, marked temporal dispersion is noted. (**b**) Near-total conduction block and modest temporal dispersion are noted upon proximal stimulation (bottom trace)

velocity to be slowed. When this occurs in some axons more than others, the CMAP duration increases (pathologic temporal dispersion), because the range of conduction velocities, from some unaffected axons to others that are affected, broadens, and the components of the CMAP become desynchronized. If there is no associated conduction block, the area under the waveform remains essentially constant (Fig. 8.4).

Conduction block, temporal dispersion, and conduction velocity slowing that differ in severity between nerves and nerve segments are characteristic features of chronic inflammatory demyelinating polyneuropathy (CIDP). By contrast, in the demyelinating form of Charcot-Marie-Tooth (CMT1), conduction block and temporal dispersion are distinctly unusual, and the degree of conduction velocity slowing is typically uniform.

Sensory Nerve Conduction Studies

Sensory NCS are performed by depolarizing a nerve via electrical stimulation and recording the amplitude and latency of a resultant nerve action potential as it travels beneath an electrode at either a more proximal site along the nerve (*orthodromic* technique) or a more distal site (*antidromic* technique). The response recorded is known as the sensory nerve action potential (SNAP) [6].

There is no muscle or neuromuscular junction (NMJ) to navigate with sensory response recordings. As such, latency represents essentially the same information as conduction velocity, and the conduction velocity can be determined by simply dividing the latency between time of stimulation and onset of the recorded response by the distance between the two points. SNAP amplitudes are a reflection of the number of individual sensory fiber action potentials and are generally reduced in axon loss disorders. Latency (or conduction velocity) reflects the speed of conduction.

Recall that acquired demyelinating disorders (CIDP, its variants, and GBS) are associated with variable degrees of conduction slowing between axons, resulting in the phenomenon of temporal dispersion of the CMAP. Because the sensory nerve action potentials of individual axons are of relatively short duration, temporal dispersion of SNAPs leads to superposition of negative and positive phases of individual sensory axon potentials, resulting in *phase cancellation*. As a result, *while conduction velocity slowing, conduction block, and even temporal dispersion can be seen in sensory conduction studies, SNAP amplitudes are often reduced in acquired demyelinating disorders due to phase cancellation.* This is an important exception to the usual rule of thumb that axon loss results in low amplitudes, while myelin loss results in slow conduction.

Late Responses

Late responses (F waves and H reflexes) evaluate conduction along the entire course of the nerve, including the most proximal segments (Fig. 8.5) [7].

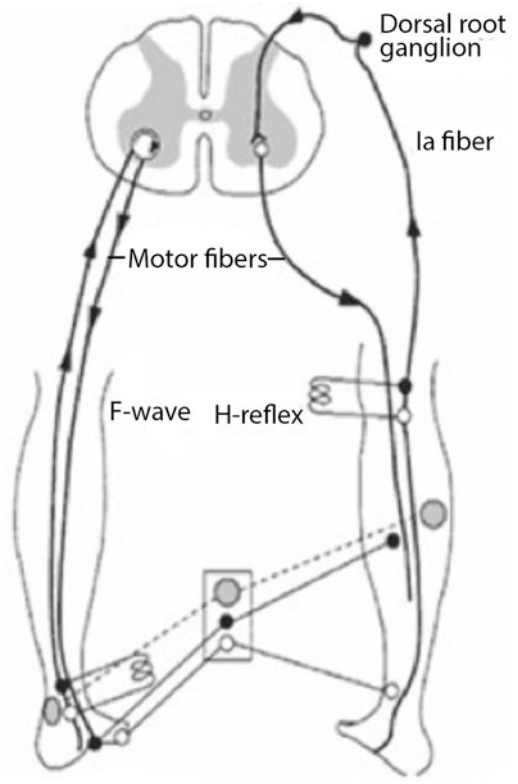

Fig. 8.5 Anatomy of late responses

F Wave

Stimulation of a motor axon causes propagation of the response in an antidromic direction (i.e., impulse propagation in a direction opposite to normal physiology) to the anterior horn cell. When the action potential reaches the anterior horn cell, in a small proportion of cells, an orthodromic (i.e., impulse propagation in the normal physiologic direction) motor response traverses the initial segment after the refractory period and travels back to the recording electrode. Ten to 20 stimuli for recording F-wave responses are routinely performed from an individual nerve/muscle pair. Unlike the CMAP, F-wave configurations and latencies vary due to a polysynaptic response in the spinal cord, where Renshaw cells inhibit impulses from traveling the same path each time. F waves are generated with supramaximal stimulation and can be evaluated for minimum F-wave latency, persistence (number of recorded responses per group of stimuli), and chronodispersion (difference between the shortest and longest F-wave latencies). Prolonged minimum F-wave latency and increased chronodispersion generally indicate myelin injury. If F-wave latency is prolonged and distal nerve conduction velocity is normal, the presumption is that the myelin injury affects the proximal segment. Reduced persistence can be seen with axon injuries, loss of anterior horn cells, or proximal sites of conduction block.

H Reflex

The tibial H reflex is the electrophysiologic equivalent of the Achilles reflex. An H reflex is initiated with a submaximal stimulus to the tibial or, less commonly, median nerve. The response travels orthodromically along the Ia afferent fibers to the spinal cord where it activates motor efferents. The orthodromic motor response travels back to the recording electrode (soleus muscle). Unlike the F wave, H-reflex morphology and latency remain constant with each stimulus at a given intensity. Like the F wave, prolonged latency generally indicates myelin injury. Absent H reflexes have little diagnostic utility as they can be seen with axonal injuries, proximal conduction block, or occasionally as a normal physiologic phenomenon.

Interpretation of Nerve Conduction Study Parameters

Axon loss is typically characterized by reduced motor and sensory response amplitude with normal or near normal conduction velocity and distal latency (Table 8.4). Demyelination is typically characterized by prolonged distal latency and slowed conduction velocity with normal or near normal amplitude. Conduction block and temporal dispersion also usually reflect focal or segmental peripheral nerve demyelination and are generally indicative of a demyelinating process. As previously noted, both amplitude loss and conduction velocity slowing may be concomitantly appreciated in the same nerve. Widely available electrodiagnostic demyelinating criteria can be referenced when the pathophysiologic significance of mild or moderate degrees of conduction velocity slowing is uncertain (Table 8.5) [4].

Table 8.4 Interpretation of motor nerve conduction studies

Motor nerve abnormality	Alternative explanation
Findings suggesting axon loss	
Amplitude reduction	Submaximal stimulation Distal conduction block Severe neuromuscular junction transmission failure Myofiber atrophy
Findings suggesting demyelination	
Distal latency prolongation	Reduced limb temperature Loss of fastest conducting fibers
Conduction velocity slowing	
Conduction block	
Temporal dispersion	

Table 8.5 Electrodiagnostic criteria for CIDP according to 2010 EFNS/PNS criteria

(1) Definite: at least one of the following:
(a) Motor distal latency prolongation ≥50% above ULN in two nerves (excluding median neuropathy at the wrist)
(b) Reduction of motor conduction velocity ≥ 30% below LLN in two nerves
(c) Prolongation of F-wave latency ≥30% above ULN in two nerves (≥50% if amplitude of distal negative peak CMAP <80% of LLN values)
(d) Absence of F waves in two nerves if these nerves have distal negative peak CMAP amplitudes ≥20% of LLN + ≥1 other demyelinating parameter[a] in ≥1 other nerve
(e) Partial motor conduction block[b]: ≥50% amplitude reduction of the proximal negative peak CMAP relative to distal, if distal negative peak CMAP ≥20% of LLN, in two nerves, or in one nerve + ≥1 other demyelinating parameter[a] in ≥1 other nerve
(f) Abnormal temporal dispersion (>30% duration increase between the proximal and distal negative peak CMAP) in ≥2 nerves
(g) Distal CMAP duration (interval between onset of the first negative peak and return to baseline of the last negative peak) increase in ≥1 nerve (median ≥ 6.6 ms, ulnar ≥6.7 ms, peroneal ≥7.6 ms, tibial ≥8.8 ms) + ≥1 other demyelinating parameter[a] in ≥1 other nerve
(2) Probable
Partial motor conduction block[b]: ≥30% amplitude reduction of the proximal negative peak CMAP relative to distal, excluding the posterior tibial nerve, if distal negative peak CMAP ≥20% of LLN, in two nerves, or in one nerve + ≥1 other demyelinating parameter[a] in ≥1 other nerve
(3) Possible
As in (1) but in only one nerve

Used with permission © John Wiley and Sons 2010 [4]
To apply criteria, median, ulnar, peroneal (stimulated below the fibular head), and tibial nerves on one side are tested. If criteria are not fulfilled, the same nerves are tested at the other side, and/or ulnar and median nerves are stimulated at the axilla and Erb's point. Temperatures are maintained to ≥33 °C palm and ≥30 °C external malleolus
[a]Any nerve meeting of any of the criteria (a–g)
[b]Conduction block is not considered in the ulnar nerve across the elbow and at least 50% amplitude reduction between Erb's point and the wrist is required for probable conduction block

Localization Using Nerve Conduction Studies

Nerve conduction studies are a collection of data from individual nerves. Hence, using NCS to localize or characterize pathology requires obtaining sufficient data to answer the referring provider's diagnostic question.

- If the question relates to a possible polyneuropathy, a sufficient number of nerve conduction studies from both sides and upper and lower limbs are needed to determine whether the problem is length dependent, non-length dependent, or multifocal.
- If the question relates to a possible single lesion of peripheral nerve, plexus, or root, nerve conduction studies referable to the structure in question are needed. If they are abnormal, nerve conduction studies of neighboring or contralateral structures determine whether the findings reflect a single lesion or a polyneuropathy.

- If NCS are abnormal, it is imperative to determine whether the findings are motor only, sensory only, or both.
- If NCS are abnormal, it is imperative to determine whether the primary process is axon loss or a disorder of myelin, using the principles discussed above.

In this way, NCS supplement the neurologic examination in establishing the distribution, modalities affected, and primary pathology in neurogenic disorders. Note that motor NCS can also be abnormal in disorders of muscle and neuromuscular transmission, diagnoses that become clear with EMG, RNS, and the clinical context.

Localization of pathology based on nerve conduction studies requires detailed knowledge of peripheral nervous system anatomy, including the course of individual named nerves through their corresponding plexus and nerve roots. Lesion localization based on nerve conduction studies is performed by recognizing NCS abnormalities with intersecting anatomical localizations as well as identification of focal abnormalities across discrete nerve segments. Detection of focal slowing or conduction block across a short segment of nerve indicates pathology at that site.

Sensory Symptoms with Normal Sensory Conduction Studies

The motor nerve cell body is within the spinal cord at the anterior horn cell, but the sensory cell bodies are in the peripheral nervous system at the dorsal root ganglion (DRG). As such, peripheral lesions proximal to the dorsal root ganglion may cause sensory symptoms in the corresponding area but will leave the peripheral nerve components structurally intact. *Hence, normal sensory nerve conduction studies in people with large-fiber-type sensory loss should alert the clinician to preganglionic localization. For example, patients with sensory symptoms due to radiculopathy have normal SNAP amplitudes, while patients with sensory symptoms due to axonal neuropathy typically have reduced SNAP amplitudes. Recall also that sensory NCS only evaluate large myelinated axons. Sensory NCS are, by definition, normal in small fiber neuropathy.*

Electromyography

Needle electromyography (EMG) refers to the recording of electrical activity of skeletal muscle using an intramuscular needle recording electrode [2]. The electrical characteristics of the muscle at rest and during activity can be interpreted by the electrodiagnostic physician. The muscles studied with EMG will vary depending upon the initial diagnostic query and the coalescence of information obtained by the physician during the electrodiagnostic test. *With each muscle sampled, spontaneous activity, motor unit potential morphology, and motor unit potential recruitment should be analyzed* (Table 8.6) [8].

Table 8.6 Parameters assessed during needle EMG

Parameter	Muscle activity	Observation
Spontaneous activity	Rest	Presence or absence of spontaneous discharges
MUAP morphology	Low-level activation	Size, duration, and complexity of the MUAPs
Recruitment	Mid- or high-level activation	Firing pattern and rate of MUAPs

EMG Procedures

After the skin is cleaned with an antiseptic solution (usually alcohol), a small disposable concentric needle electrode is inserted into the muscle. The electrode consists of a fine wire, which serves as the active electrode, surrounded by a cannula, which serves as the reference electrode. Unlike NCS, which record either nerve or compound muscle action potentials but are used principally as a test of nerve function, EMG directly records only the electrical potentials generated by depolarization of muscle cells. The electromyographer first assesses *spontaneous activity*, which represents muscle cell discharges at rest, and then assesses *volitional activity, or the configuration and firing patterns of individual motor unit action potentials (MUAPs),* which represent the near-simultaneous discharge of all muscle cells innervated by a single axon, and hence reflect the structure and function of a motor unit.

Note that the EMG electrode only registers electrical activity from a small surrounding area; hence, although the electromyographer does sample several areas by repositioning the needle, needle electromyography is subject to sampling error and can easily miss abnormalities if they are nonuniform and in a large muscle. Furthermore, some components of a motor unit may extend beyond the recording area of the electrode.

The electrical activity is amplified and transduced into both visual and auditory signals, so the examiner can both hear and see the electromyographic findings.

- *Spontaneous activity is assessed first* by moving the needle through the muscle with small quick movements. Multiple locations of the muscle should be sampled with a single-needle insertion. At each sampling site, the examiner pauses for several seconds and notes any abnormalities. Types of spontaneous activity are outlined below and in Table 8.7.
- *Motor unit action potentials (MUAP) are evaluated next.* With the needle in a fixed position, the patient minimally contracts the muscle until an action potential of a single motor unit is observed. Each MUAP is assessed for amplitude (peak to peak, mV), duration (ms), and number of phases (turns or baseline crossings). Multiple individual MUAPs should be analyzed.
- *Recruitment is assessed next* by asking the patient to increase the level of contraction (Fig. 8.6). The number and speed with which additional MUAPs appear define recruitment.

Table 8.7 Spontaneous activity during needle EMG

	Amplitude	Duration	Morphology	Rhythm	Sound	Interpretation
End plate noise	10–20 μV	0.5–1 ms	Simple	Irregular	Seashell	Normal
End plate spikes	100–300 μV	2–4 ms	Simple	Irregular	Popcorn	Normal
Fibrillation potentials and PSW	10–100 μV	1–5 ms	Simple	Regular at 1–50 Hz	Rain on a tin roof	Spontaneous muscle fiber contraction
Fasciculation	Often ≥200 μV	Often ≥10 ms	Simple or complex	Irregular at 0.1 to 10 Hz	Pop	Spontaneous single motor unit discharge
CRD	100 μV–1 mV	Variable	Complex	Regular at 20–150 Hz	Machine like	Ephaptic transmission between adjacent fibers
Myotonia	Waxing and waning	Waxing and waning	Simple	Irregular 20–150 Hz	Dive bomber	Muscle membrane instability
Myokymia	Variable	Variable	Simple or complex	Regular or irregular 5–60 Hz	March	Spontaneous or ephaptic motor nerve discharge
Neuromyotonia	Waning	Variable	Simple or complex	Irregular 150–250 Hz	Ping	Motor axon hyperexcitability

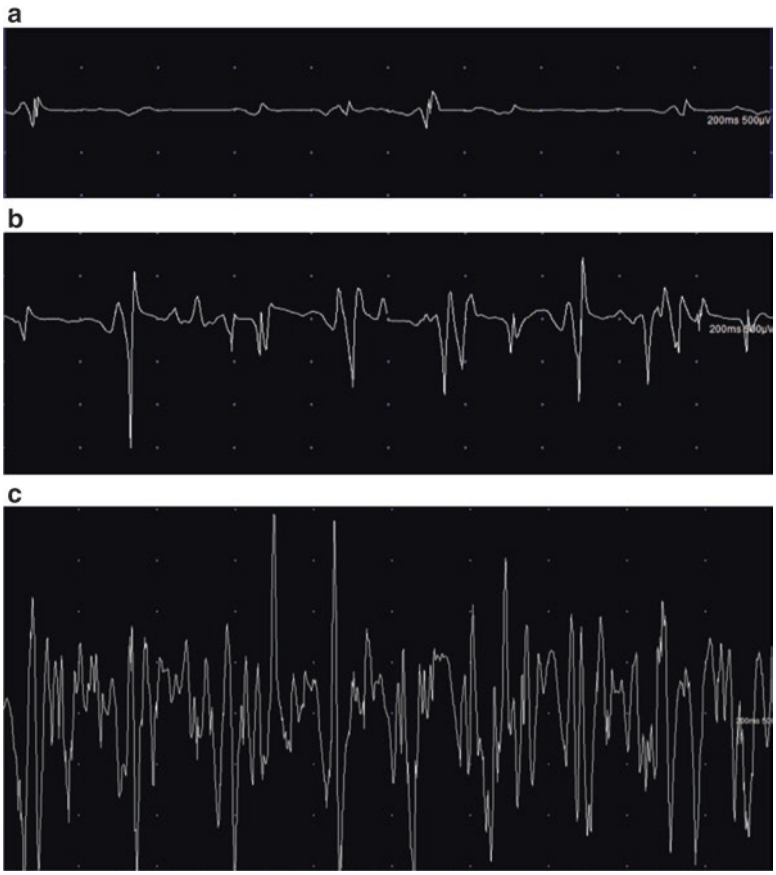

Fig. 8.6 Normal motor unit activation patterns at minimal (**a**), moderate (**b**), and full activation (**c**)

Spontaneous Activity

The term "spontaneous activity" applies to electromyographic discharges that can be recorded with the muscle at rest and is distinguished from recordings made from voluntarily activated motor units. Most are abnormal and have specific etiologic implications. By far the most common are fibrillation potentials and positive sharp waves. The following are the types of spontaneous activity.

Fibrillation Potentials and Positive Sharp Waves

Fibrillation potentials and positive sharp waves are action potentials of a single muscle fiber undergoing spontaneous depolarization. Although they have different names and appearances, their pathologic significance is essentially

indistinguishable. As noted they are by far the most common type of abnormal spontaneous activity. Fibrillation potentials and positive sharp waves *indicate either denervation, as occurs in axon loss, or loss of the integrity of muscle membrane, as occurs in many myopathies, such as muscular dystrophy or rhabdomyolysis.* By contrast they are not seen in neuropathies and myopathies with preserved neuromuscular junctions and muscle cell membranes, such as pure demyelination and muscle channelopathies. Fibrillation potentials and positive sharp waves do not develop until 2–3 weeks after axonal injury, so in the acute phase, it can be difficult to distinguish axon loss from conduction block on the basis of EMG alone.

Fibrillation potentials and positive sharp waves are scored as follows: 0 (none), 1+ (single train of potentials longer than 2 s in 2 areas), 2+ (moderate numbers in 3 or more areas), 3+ (many potentials in all areas), and 4+ (full interference pattern of potentials).

Fasciculation Potentials

Fasciculation potentials reflect a spontaneous discharge of a single motor axon and are the electrophysiologic signature of a clinical fasciculation. Because they reflect the simultaneous contraction of all muscle fibers innervated by a single axon, they are much larger than fibrillation potentials, which represent the contraction of only a single muscle cell. They often sound like a large "pop." Fasciculation potentials may be benign but also occur in disorders of axonal excitability, polyneuropathies, and anterior horn cell disorders.

Complex Repetitive Discharges (CRD)

CRDs are stereotyped, recurrent discharges of several adjoining muscle fibers. They have a constant frequency and a very distinct sound reminiscent of mechanical equipment. CRDs probably arise from ephaptic transmission between adjacent denervated fibers. CRDs are most commonly seen in chronic neurogenic disorders in which there has been denervation, reinnervation, and subsequent denervation.

Myotonic Discharges

Myotonic discharges are high-frequency trains of fibrillation-like potentials that vary in frequency and amplitude in a waxing and waning patterns. These regular variations result in a characteristic sound often likened to that of a "dive bomber," or the Doppler effect superimposed on the sound of a rapidly firing airplane engine. Myotonic discharges occur with disorders in muscle fiber membrane channels, including myotonic dystrophy and non-dystrophic channelopathies such as myotonia congenital and paramyotonia congenita.

Neuromyotonia

Neuromyotonia arises from hyperexcitability of a single peripheral motor axon. It is characterized by a very high-frequency discharge (100–300 Hz, faster than myotonic discharges) of a motor unit, partial motor unit, or single-fiber potentials. Clinical syndromes associated with electrical neuromyotonia include autoimmune Isaac's syndrome due to voltage-gated potassium channel antibodies.

Myokymia

Myokymia is a spontaneous, rhythmic, regular, or irregular discharge of groups of motor units that produce the clinical appearance of quivering in the muscles (grouped fasciculations). The discharges fire repetitively in single or grouped units at a uniform rate. They often sound like a group of soldiers marching. Myokymia can occur with chronic disorders of peripheral nerve and is most commonly seen in radiation nerve injury.

End Plate Noise and Spikes

End plate noise and spikes are normal spontaneous activity. The activity can be heard when the needle electrode is close to end plates. End plate noise has a characteristic "seashell" sound, whereas end plate spikes are irregular and sometimes sound like "popcorn." These reflect release of subthreshold quantities of acetylcholine across the neuromuscular junction and are the only type of spontaneous activity that is always normal.

Motor Unit Action Potential Morphology

Normal Motor Unit Action Potentials

MUAPs have a characteristic appearance. MUAP duration is defined as the time from initial deflection from baseline to the final return to baseline and is normally between 5 and 15 ms. Duration reflects the number of muscle fibers within a motor unit and the dispersion of fiber depolarizations over time. Short or brief duration MUAPs are classically seen in myopathic disorders, reflecting the presence of fewer functional muscle cells. Duration typically lengthens with increasing numbers of muscle fibers in a motor unit, such as when neurogenic reinnervation follows denervation. Duration lengthening can also be seen with increasing age and with decreased temperature. Audibly, duration is associated with pitch. Short duration MUAPs sound crisp and high pitched, like a sound from a small drum, while long-duration MUAPs sound dull and low-pitched, like a sound from a bass drum.

MUAP amplitude is generally between 200 μV to 2 mV, measured peak to peak, although it can vary depending on the muscle sampled. Amplitude represents only those few fibers closest to the needle, rather than the total number of fibers, and reflects muscle fiber density or muscle fiber diameter. That said, MUAP amplitude and duration are highly correlated, and both are increased most commonly due to increased motor unit size due to reinnervation. Increased MUAP amplitude can also be appreciated in chronic myopathies with muscle fiber hypertrophy. Audibly, amplitude is associated with volume. Larger amplitude MUAPs sound louder.

Polyphasia is a measure of muscle fiber firing synchronicity. MUAPs generally have ≤5 phases (baseline crossings). MUAPs from muscle fibers with poor synchronicity (muscle fibers firing at different times) have increased phases and turns (direction changes). This can occur with both neurogenic and myopathic disorders. Some degree of polyphasia is seen in normal muscle. Pathologic increased polyphasia requires at least 10% of the sampled muscle fibers to have >5 phases.

Abnormal Motor Unit Action Potentials

Changes in motor unit potential morphology can give clues to the underlying mechanism of injury and the duration of injury (Table 8.8). Generally speaking, high-amplitude, long-duration motor unit potentials with or without increased polyphasia develop after denervation and reinnervation (Fig. 8.7a). As part of the reinnervation process, collateral sprouting extends axons to denervated muscle fibers, effectively increasing the number of muscle fibers of that motor unit. More muscle fibers within a given motor unit lead to larger MUAPs with longer durations and increased polyphasia. In primary muscle disease, individual muscle fibers are injured, resulting in a reduced number of muscle fibers within any given motor unit. With fewer muscle fibers to contribute to the compound action potential, amplitudes become small and durations short (Fig. 8.7b). Polyphasia may increase as well. Rarely, chronic myopathies with muscle fiber hypertrophy can have a mixed appearance with both large and small MUAPs.

Motor Unit Recruitment

Recruitment refers to the successive activation of motor units with increasing strength of voluntary muscle contraction [9]. With minimal muscle activation, a single motor unit becomes activated with a firing rate of about 5 Hz. With increased voluntary effort (central drive), the single motor unit fires faster to increase force generation. When the firing rate increases to about 10 Hz, another motor unit is activated or *recruited*. As central drive increases further, the first MUAP increases firing to 15 Hz, the second to 10 Hz, and then another MUAP is recruited. This pattern

Table 8.8 Motor unit action potential morphology

Parameter	Represents	Normal	Pathologic increase when	Pathologic decrease when
Duration[a]	Number of muscle fibers in motor unit	5–15 ms	Neurogenic reinnervation after denervation	Myopathy
Amplitude[a]	Muscle fiber density or muscle fiber size	100 μV–2 mV	Neurogenic reinnervation; muscle fiber hypertrophy	Myopathy without muscle fiber hypertrophy
Polyphasia[a]	Synchronicity of muscle fiber firing	<5 phases	Early neurogenic reinnervation; chronic reinnervation; acute or chronic myopathy	No pathologic significance

[a]Additional variations may be seen depending on muscle selected, age, and temperature

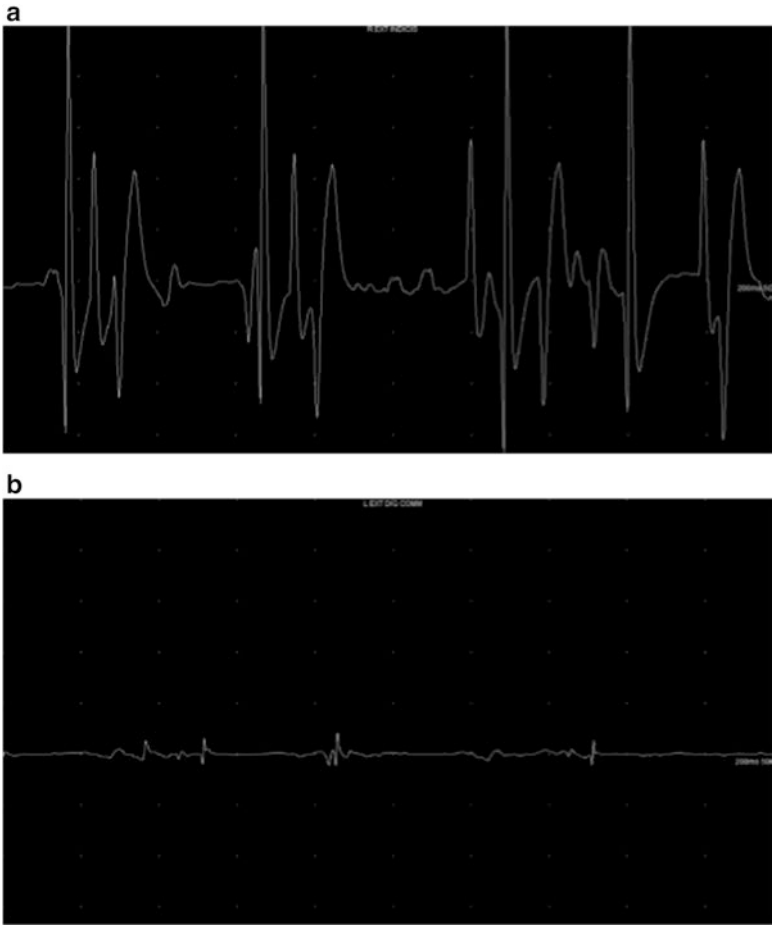

Fig. 8.7 (**a**) High-amplitude, long-duration, "neurogenic" motor unit potentials reflecting reinnervation; (**b**) low-amplitude, short-duration "myopathic" motor unit potentials

of recruitment is continued until MUAPs reach tetanic frequency of about 30–50 Hz, with the ratio of firing frequency to the number of different MUAPs firing remaining at approximately 5:1.

Neurogenic injuries (axon loss or conduction block) result in *reduced recruitment*. Reduced recruitment means that the firing rate of individual motor units is normal, but as central drive increases, there is failure to add more motor units. The result is that the electromyographer sees and hears only a few MUAPs that fire rapidly.

Myogenic injuries typically result in *early recruitment*. When there is a loss of individual muscle fibers, the motor unit generates less force. The motor units remain intact and so to increase power motor units must fire earlier than would be expected with a normal 5:1 recruitment ratio. Early recruitment means that the number of MUAPs firing is increased for the degree of force generated.

Interpreting Electrodiagnostic Findings

As indicated above, NCS identify the distribution, modalities affected, and pathophysiology of neurogenic disorders and will often demonstrate reduced CMAPs in myopathic disorders. EMG will usually reliably distinguish myopathic, acute neurogenic, and chronic neurogenic processes based upon the features of spontaneous activity and the configuration and firing pattern of voluntarily active MUAPs. When investigating a focal lesion such as a mononeuropathy or radiculopathy, the electromyographer will study several muscles that share root or nerve innervation and make an anatomic diagnosis based upon the common innervation pattern of affected muscles. The electromyographic features of common neuromuscular conditions are outlined in Table 8.9.

Summary

The key to a successful NCS/EMG study begins with understanding what question is being explored. The electrodiagnostic clinician can then plan a well-organized study that evolves with the findings as they emerge. Many abnormalities might be uncovered during an NCS/EMG study. An understanding of these abnormalities provides the clinician with an important foundation to determine localization, pathophysiology, severity, and chronicity.

Table 8.9 Interpreting electrodiagnostic findings

	Nerve conduction studies		Electromyography			
	CMAP amplitude	SNAP amplitude	Spontaneous activity (Fibs/PSW)	MUAP morphology	Recruitment	Comment
Hyperacute axon loss (<1 week)	Normal	Normal	Absent	Normal	Reduced	Before Wallerian degeneration
Acute axon loss (days-weeks)	Decreased	Decreased[a]	Absent	Normal	Reduced	Wallerian degeneration occurred, but fibs may take 2–6 weeks
Subacute axon (weeks-2 months)	Decreased	Decreased[a]	Present	Normal	Reduced	Denervation potentials present
Chronic axon loss with reinnervation (>2 months)	Decreased	Decreased[a]	Absent	Large, long	Reduced	MUAP remodeling
Myopathic	Normal or decreased	Normal	Increased or normal	Small, brief	Early	Variable depending on type and duration

[a]If postganglionic
Fibs fibrillation potentials, *MUAP* motor action unit potential, *PSW* positive sharp waves

References

1. Barboi AC, Barkhaus PE. Electrodiagnostic testing in neuromuscular disorders. Neurol Clin. 2004;22(3):619–41.
2. https://www.aanem.org/getmedia/65934187-d91e-4336-9f3c-50522449e565/Model-Policy.pdf.
3. Gutmann L. Pearls and pitfalls in the use of electromyography and nerve conduction studies. Semin Neurol. 2003;23(1):77–82.
4. Van den Bergh PY, Hadden RD, Bouche P, et al. European Federation of Neurological Societies/Peripheral Nerve Society guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society – first revision. Eur J Neurol. 2010;17(3):356–63.
5. Joint Task Force of the EFNS and the PNS. European Federation of Neurologic Societies/Peripheral Nerve Society guideline on management ofmultifocal motor neuropathy. Report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society--first revision. J Peripher Nerv Syst. 2010;15(4):295–301.
6. Wilbourn AJ. Sensory nerve conduction studies. J Clin Neurophysiol. 1994;11(6):584–601.
7. Fisher MA. H reflexes and F waves. Fundamentals, normal and abnormal patterns. Neurol Clin. 2002;20(2):339–60.
8. Daube JR, Rubin DI. Needle electromyography. Muscle Nerve. 2009;39(2):244–70.
9. Enoka RM. Morphological features and activation patterns of motor units. J Clin Neurophysiol. 1995;12(6):538–59.