



Needle electromyography Fundamentals, normal and abnormal patterns

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Nerve conduction studies and needle electromyography (EMG) are the most common electrophysiologic tests utilized to evaluate patients with suspected neuromuscular disorders. Both tests must be individualized, based on the clinical findings and differential diagnosis, and modified as the tests proceed. With needle EMG, almost every muscle in the body can be studied. However, this is neither practical for the electromyographer nor desirable for the patient. For each study, a balance must be reached between studying a sufficient number of muscles to reach or exclude a diagnosis and the patient's ability to tolerate the exam; most patients tolerate the exam well, with minor discomfort, when performed skillfully.

The needle EMG is the more challenging part of the electrophysiologic exam. Knowledge of anatomy and physiology is required for a successful study, as are sound EMG technique and good patient rapport. Two competing influences make the needle EMG study especially demanding: First, many of the abnormalities on the needle study are subtle. At the same time, however, the range of normal findings is quite large and varies with age and the muscle being studied. Although the basics of the needle study, such as needle placement and recognition of certain types of abnormal spontaneous activity, can usually be learned in a short time, recognition of many of the uncommon and subtle needle EMG findings often take years to master. This

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article focuses on the fundamentals of performing the routine needle EMG examination and interpreting the findings.

Needle EMG examination

Needle electrode and equipment

In addition to the EMG machine, an EMG needle and cable, ground electrode, and gloves are necessary to perform the needle EMG study. The ground electrode is applied to the limb being studied to ensure safety and to suppress noise. Disposable gloves must always be worn to prevent the transmission of blood-borne infections between the patient and the electromyographer. The EMG needle is connected to a cable and then plugged into the EMG machine. Either a concentric or monopolar EMG needle can be used (Fig. 1). When measuring an electrical potential, including the potentials measured during the needle EMG study, voltage is the difference

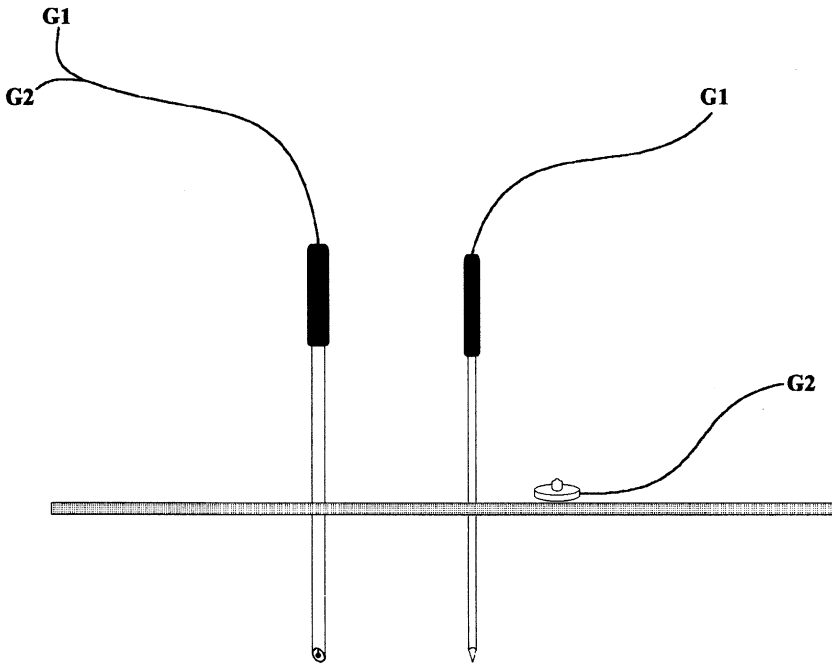


Fig. 1. EMG Needle electrodes. Concentric needle (*left*) containing the active (G1) and reference (G2) electrodes. Monopolar needle (*right*) is teflon coated with the exposed tip serving as the active electrode (G1); an additional surface disk electrode is needed as a reference electrode containing the active electrode (G1) and the reference electrode (G2). (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

between an active and reference-recording electrode. The concentric needle contains both the active and reference electrodes. The shaft of the needle serves as the reference electrode; the active electrode is a very small wire that runs through the center of the needle and is exposed at the needle tip. In the monopolar montage, the monopolar needle is Teflon-coated, with its exposed tip serving as the active recording electrode. An additional surface disk electrode is required as the reference electrode.

Both concentric and monopolar needles record electrical signals well from muscle. However, there are small differences between the two types of needle when recording motor-unit action potentials (MUAPs). With a concentric needle, MUAP amplitude is slightly smaller and the major spike rise time shorter than the potential obtained with a monopolar needle. Otherwise, there are no appreciable differences between the two in the recorded waveforms. The concentric needle is easier to use, as it does not require an additional reference electrode. However, the monopolar needle has the advantage of a smaller caliber and sharper point, and may be slightly less painful and easier for patients to tolerate. The major disadvantage of the monopolar needle is the need for an additional reference electrode. Because the reference electrode must be placed close to the active electrode, it must be moved from location to location with each muscle sampled. In addition, because the active electrode is an intramuscular needle and the reference is a surface disk, there is a much greater likelihood of electrode impedance mismatch and increased electrical noise.

Procedure

For each muscle studied, one must first identify the needle insertion point, and instruct the patient on how to properly activate the muscle. Once a muscle has been selected for study, the needle insertion point is located by identifying the proper anatomic landmarks. Second, while palpating for muscle movement, the patient is asked to activate and relax the muscle several times. Once muscle location is properly identified and palpated, the patient is asked to relax; this reduces the level of pain. Inserting a needle into a contracted muscle is much more painful than putting a needle into a relaxed one. Third, the needle is quickly inserted into the muscle, and the patient instructed to activate the muscle slightly, in order to confirm needle location. Sharp MUAPs with minimal contraction indicate a properly inserted needle. If sharp MUAPs are not indicated, the needle should be adjusted. The procedure should be repeated if sharp MUAPs are not indicated. One should not proceed until it is certain the needle has been inserted into the correct muscle.

Once correct needle placement is established, the first part of the examination is to assess insertional and spontaneous activity, with the muscle at rest. This is usually done with the sensitivity set at 50 microvolts (μV) per division, because spontaneous discharges are low amplitude. Five to ten brief insertions are performed looking for increased insertional activity and

spontaneous discharges at rest. Muscle is normally quiet at rest, with the exception of the potentials seen near the endplate zone. When the needle is quickly moved through muscle, there is a brief burst of muscle fiber potentials, known as insertional activity, which typically lasts no longer than 300 milliseconds after the needle has stopped moving. Increased insertional activity is defined as any activity, other than endplate potentials, that lasts longer than 300 milliseconds after brief needle movement. If the activity persists beyond 3 seconds, it is termed spontaneous activity, which can be normal or abnormal.

Once insertional and spontaneous activity has been characterized, the needle is left in place, and the analysis turns to the evaluation of MUAPs. The sensitivity is changed to 200 μ V per division. MUAPs are typically much larger than most abnormal spontaneous activity and therefore require the change in sensitivity. To analyze MUAPs, the patient is asked to slowly and evenly contract the targeted muscle. MUAPs are difficult to interpret in patients with uneven muscle contraction, especially those with a tremor.

With the patient minimally activating the muscle, the needle is gently moved until the MUAPs become sharp, that is, louder and crisper. As the needle moves closer to the MUAP, there is less intervening tissue to attenuate and filter the potential. Thus, the closer the needle to the MUAP, the higher the amplitude and the shorter the major spike rise time. It is at this point that the MUAP can be properly evaluated. MUAPs are assessed for duration, amplitude, and number of phases (see later). In addition, the number of MUAPs and their relationship to the firing frequency (recruitment and activation pattern) are also determined. As the patient slowly increases force, both the firing frequency and the number of MUAPs normally increase. After MUAPs are assessed at one location, the needle is moved slightly within the muscle to a different site, and the process is repeated. Several different MUAPs are analyzed at each site.

Insertional and spontaneous activity

Insertional activity

The needle EMG examination of each muscle begins with the assessment of insertional activity. When a needle is quickly moved through muscle, muscle fibers depolarize in a normal brief burst for several hundred milliseconds (ms), known as *normal insertional activity*. At least four to six brief needle movements are made in four quadrants of each muscle to assess insertional activity. Needle movement resulting in any abnormal waveform that lasts longer than 300 ms indicates increased insertional activity. Increased insertional activity may be seen in both neuropathic and myopathic conditions. In rare conditions, where muscle has been replaced by fat and fibrous connective tissue, insertional activity may actually be decreased.

The role of spontaneous activity in the needle EMG examination

The ability to recognize and identify abnormal spontaneous activity is one of the most important parts of the needle EMG examination. The presence of abnormal spontaneous activity on an EMG can yield several key pieces of information. First, the distribution of abnormal spontaneous activity helps determine the neuroanatomic localization of the lesion. For example, in a radiculopathy, denervation potentials are restricted to muscles in the same myotome. Second, the type of spontaneous activity often provides specific diagnostic information. Certain types of spontaneous activity are associated with specific disorders. For example, myotonic discharges are seen only in a few conditions, such as myotonic dystrophy or myotonia congenita. Third, the degree or amount of spontaneous activity often helps to assess the severity of the lesion. Finally, the presence of abnormal spontaneous activity might yield information regarding the time course of the lesion. For example, in a radiculopathy, several weeks must pass before fibrillation potentials are seen in the limbs.

Analysis of spontaneous activity

The ability to recognize spontaneous activity improves with experience. However, careful analysis of any spontaneous waveform can usually lead to its correct identification. Each waveform should be analyzed for morphology, stability, and firing characteristics [1]. Practically, every spontaneous waveform can be properly identified through use of this information.

Morphology

The source of a spontaneous discharge can often be discerned by its morphology, including the size and shape of the potential and its initial deflection (Fig. 2) [1]. The source generators that must be differentiated include: neuromuscular junctions (NMJ), single muscle fibers, terminal axon twigs, motor neuron and axons, and linked multiple muscle fibers.

At the NMJ, miniature endplate potentials (mepps) occur spontaneously. They result from the normal spontaneous exocytosis of individual quanta of acetylcholine traveling across the neuromuscular junction, leading to a non-propagated, subthreshold endplate potential. If the EMG needle is near the endplate zone, mepps can often be recorded [2,3]. They have a distinctive small amplitude and monophasic negative morphology.

When a muscle fiber depolarizes to threshold, a muscle-fiber action potential (MFAP) is created. An MFAP can assume one of two basic morphologies, either a brief spike or a positive wave. The brief spike is typically from 1 to 5 milliseconds in duration, biphasic or triphasic, with low amplitude (typically, 10–100 μ V). Brief spike morphology is commonly seen when muscle fibers depolarize spontaneously (eg, denervation), but can also occur through individual terminal axonal twig depolarizing followed by

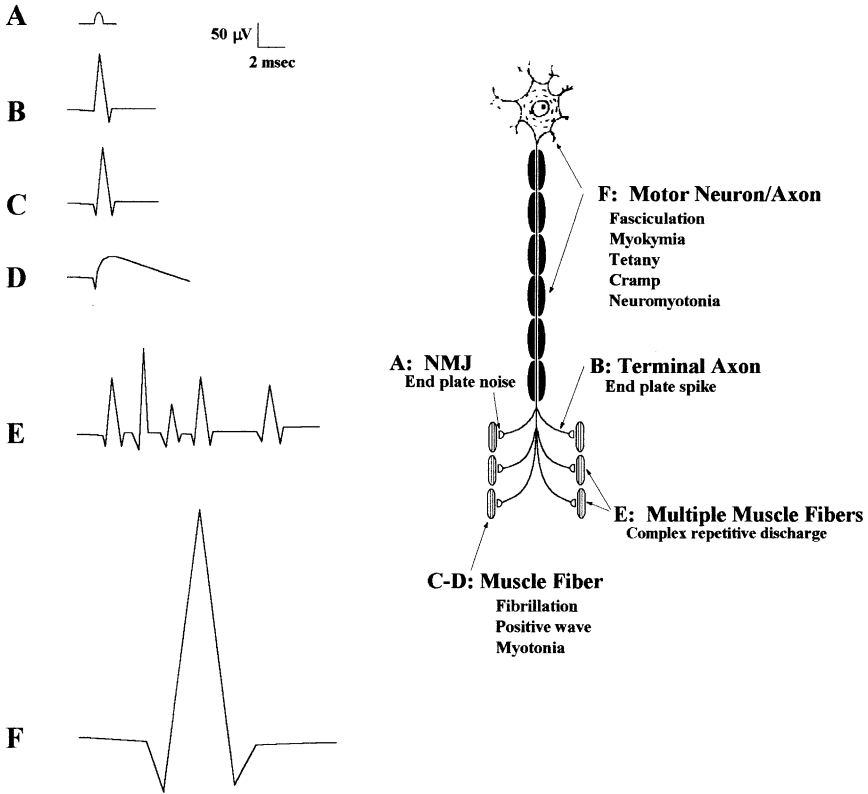


Fig. 2. Spontaneous waveform generators and morphologies. (A) Neuromuscular junction (NMJ). Miniature endplate potential (monophasic negative). (B) Terminal axon. Brief spike (initial negative, diphasic). (C) Muscle fiber action potential—brief spike morphology (initial positive, triphasic). (D) Muscle fiber action potential—positive wave morphology (initial positive, slow negative). (E) Multiple muscle fibers (linked, multiple brief spikes). (F) Motor neuron/axon—motor-unit action potential. (Note the longer duration and higher amplitude compared to the muscle fiber potentials A–E.) (Adapted from Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

propagation across the NMJ, which creates an MFAP. Attention to the initial deflection of the potential and whether the brief spike is biphasic or triphasic can often help distinguish between the two (Fig. 3). If depolarization begins under the recording needle electrode, a biphasic potential is seen, with an initial negative peak followed by a short positive phase. This signifies that the needle is at the endplate zone, where the depolarization begins, and is usually the result of the EMG needle irritating terminal nerve twigs near the endplate zone. Nerve twig action potential leads to an MFAP known as an endplate spike, which is a normal finding (see later). Otherwise, brief spikes occur from spontaneous depolarization of muscle fibers and are

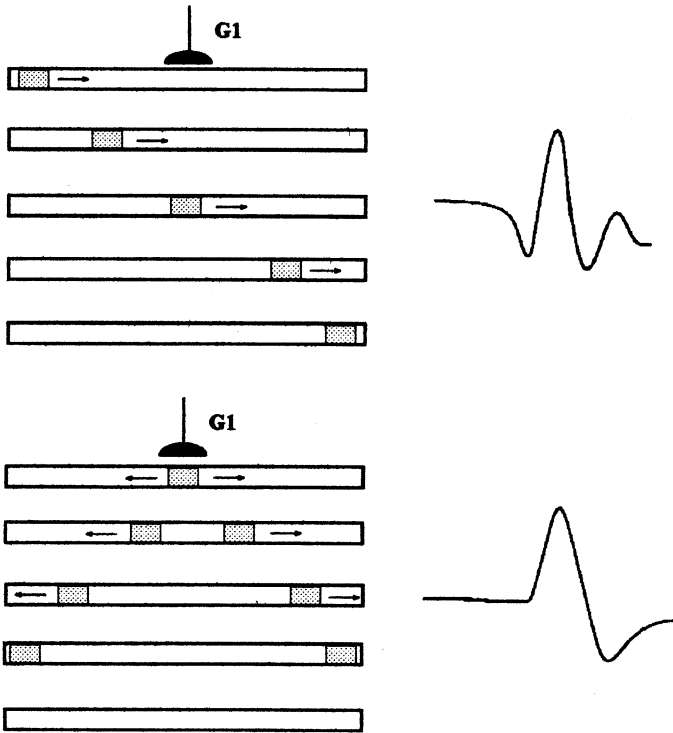


Fig. 3. Waveform morphology and site of depolarization. (*Bottom*) Trace shows a traveling depolarizing wave creates a biphasic potential if the waveform begins under the recording needle electrode (initial negative peak) and then moves away from the electrode (positive peak). This morphology occurs with endplate spikes. (*Top*) Trace shows that if the waveform begins at a distance from the needle, there is an initial positive deflection as it moves toward the needle, followed by a negative phase as it moves beneath the needle, and then a final positive deflection as it travels away. This morphology occurs with fibrillation potentials. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

associated with an initial positive, usually triphasic morphology, which is an abnormal finding. When a depolarization begins at a distance from the needle, there is an initial positive deflection as it moves toward the needle, followed by a negative phase as it moves beneath the needle, and then a final, positive deflection as it moves away from the needle.

In addition to the brief spike, an MFAP can also assume a positive wave, biphasic morphology with an initial brief positive phase followed by a long negative phase. Both positive waves and initial positive, triphasic brief spikes are most often seen as denervating potentials, which are known as positive sharp waves and fibrillation potentials, respectively. However, myotonic discharges, which also originate in muscle fibers, have the same basic morphology as denervating potentials—either brief spikes or positive waves.

This emphasizes the important concept that morphology alone cannot be used to identify a potential. Although the morphology of a potential can usually be used to identify its source generator, additional information regarding stability and firing characteristics is needed to fully characterize and identify the potential (see later).

The next major category of spontaneous discharges arises from motor neurons or their axons. Any discharge that occurs as a result of the spontaneous depolarization of a motor neuron or its axon (prior to its terminal branches) leads to a potential with the morphology of a motor unit, known as a motor-unit action potential (MUAP). Spontaneous discharges generated by the motor neuron/axon include fasciculations, tetany, myokymic discharges, and neuromyotonic discharges and cramps, which all lie along the spectrum of abnormal spontaneous MUAPs. They can be differentiated from each other, however, by their stability and firing characteristics (see later). If the motor unit is normal, then the MUAP morphology will typically have two to four phases, duration of 5–15 milliseconds, and variable amplitude depending on the needle position. If the motor unit is pathologic, the number of phases, the duration, and the amplitude may change. Differentiating an MUAP from a single MFAP is usually straightforward and can typically be done quite simply by analyzing a waveform's duration and amplitude.

The last distinctive waveform that must be recognized is that of time-linked individual muscle fibers, which occurs in complex repetitive discharges. Although a MUAP also contains many individual muscle fibers, the muscle fibers in a motor unit fire more or less synchronously, and in almost every situation summate to create a large potential, 5 to 15 milliseconds in duration. In contrast, multiple muscle fibers in a complex repetitive discharge fire consecutively, and are usually discernible as individual spikes that are time-linked together.

Stability

Assessment of the stability of any spontaneous waveform is essential. Most spontaneous potentials are relatively stable in their morphology. However, some waveforms may wax and wane, decrease, or change abruptly. MFAPs that wax and wane in amplitude are characteristically seen in myotonia. A marked decrement of a MUAP occurs in neuromyotonic discharges. Complex repetitive discharges are typically stable, but if additional loops or circuits drop in or out, the morphology may change in distinct or quantal jumps.

Firing characteristics

After assessing the potential's morphology and stability, attention turns to its firing characteristics, including the discharge pattern and firing rate. One should note if the pattern is regular or irregular. Many types of irregular firing may be seen, including sputtering (endplate spikes), waxing/waning

(myotonic discharges), waxing (neuromyotonic discharges), and bursting (tetany and myokymic discharges). Equally important is the approximate firing rate. For instance, some potential typically fire slowly (eg, fasciculations), whereas others fire quickly (eg, 150–300 Hertz (Hz), in the case of neuromyotonic discharges).

Table 1 summarizes the morphology, stability, and firing characteristics of the common spontaneous potentials seen during the needle EMG.

Spontaneous activity generated near the neuromuscular junction

Muscle is normally electrically silent outside of the endplate zone. All spontaneous activity is abnormal with the important exception of potentials that occur in the endplate region (ie, the NMJ). Muscle endplate is usually found near the center of the muscle belly and is often encountered during routine EMG [4]. Patients frequently perceive a deep burning sensation when the needle is placed in the endplate region. Two types of spontaneous activity occur: endplate noise and endplate spikes. It is essential to properly identify these potentials (described in the following paragraphs), so as not to mistake them for abnormal spontaneous activity.

Endplate noise (Fig. 4) Endplate noise potentials are low-amplitude, monophasic, negative potentials that fire irregularly at 20 to 40 Hz, have a characteristic sea shell sound on EMG, physiologically represent mepps, and are recognized by their characteristic shape and sound and frequent association with endplate spikes.

Endplate spikes (Fig. 4)

Endplate spikes are brief spikes that fire irregularly up to a frequency of 50 Hz. Endplate spikes are biphasic with an initial negative deflection, reflecting that the needle is at the site where the action potential is generated, and are usually seen along with endplate noise [5]. They are thought to occur as a result of needle-induced irritation of the terminal nerve twigs, which then causes nerve-twig action potentials and then MFAPs. They have a cracking, buzzing, or sputtering sound on EMG. The key features that differentiate endplate spikes from fibrillation potentials, which are also brief spikes, are their initial negative deflection and their highly irregular firing rate.

Spontaneous activity generated from muscle fibers

Fibrillation potentials (Fig. 5)

Fibrillation potentials are electrophysiologic markers of denervation [6,7]. Although they are typically associated with neurogenic disorders, they may also be seen in muscle disorders (especially inflammatory myopathies and muscular dystrophies), and rarely in severe diseases of the NMJ (especially botulism). Fibrillation potentials are recognized as brief spikes with an

Table 1
Spontaneous activity

Potential	Source generator/ morphology	Sound on loudspeaker	Stability	Firing rate	Firing pattern
Endplate noise	mepp (monophasic negative)	Sea shell	—	20–40 Hz	Irregular (hissing)
Endplate spike	Muscle fiber initiated by terminal axonal twig (brief spike, diphasic, initial negative)	Sputtering fat in a frying pan	—	5–50 Hz	Irregular (sputtering)
Fibrillation	Muscle fiber (brief spike, diphasic or triphasic, initial positive)	Rain on a tin roof or tick-tock of a clock	Stable	0.5–10 Hz (occasionally up to 30 Hz)	Regular
Positive sharp wave	Muscle fiber (diphasic, initial positive, slow negative)	Dull pops, rain on a tin roof, or tick-tock of a clock	Stable	0.5–10 Hz (occasionally up to 30 Hz)	Regular
Myotonia	Muscle fiber (brief spike, initial positive, or positive wave)	Revolving engine	Waxing/waning	20–150 Hz	Waxing/waning
CRD	Multiple muscle fibers time-linked together	Machine	Usually stable, may change in discrete jumps	5–100 Hz	Perfectly regular (unless overdriven)
Fasciculation	Motor unit (motor neuron/axon)	Corn popping	—	Low (0.1–10 Hz)	Irregular
Myokymia	Motor unit (motor neuron/axon)	Marching soldiers	—	1–5 Hz (interburst) 5–60 Hz (intraburst)	Bursting
Cramp	Motor unit (motor neuron/axon)	—	—	High (20–150 Hz)	Interference pattern or several individual units
Neuromyotonia	Motor unit (motor neuron/axon)	Pinging	Decrementing	Very high (150–250 Hz)	Waning

Abbreviations: CRD, complex repetitive discharge; mepp, miniature endplate potential.

(Adapted from: Preston DC, Shapiro BE. Electromyography and neuromuscular disorders. Boston: Butterworth-Heinemann; 1998; with permission.)

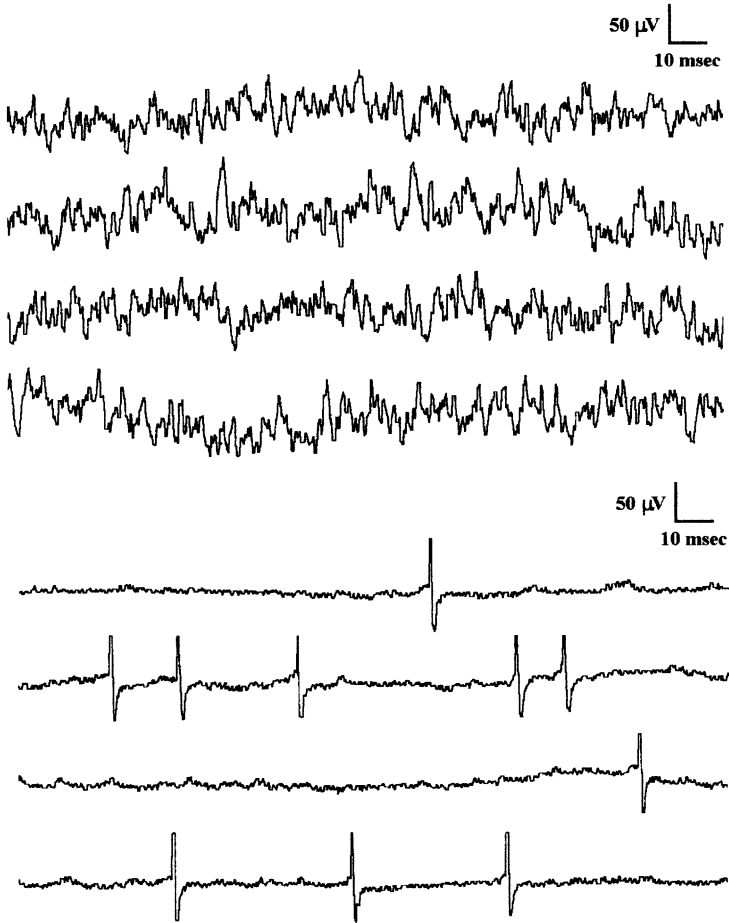


Fig. 4. Endplate noise (*top traces*) and endplate spikes (*bottom traces*). (Adapted from Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

initial positive deflection, duration of 1 to 5 milliseconds, and low amplitude (typically, 10–100 μV). Their firing pattern is regular, with a rate usually of 0.5 to 10 Hz. In the most chronic conditions (> 6–12 mo), fibrillation potentials may become very small (<10 μV in amplitude). On EMG, single fibrillation potentials often sound like rain on the roof. Although fibrillation potentials fire at a regular rate, they may slow down gradually over several seconds before stopping.

Positive sharp waves (Fig. 5)

Positive sharp waves have the same significance as fibrillation potentials: they occur in denervation and represent spontaneous depolarizations of

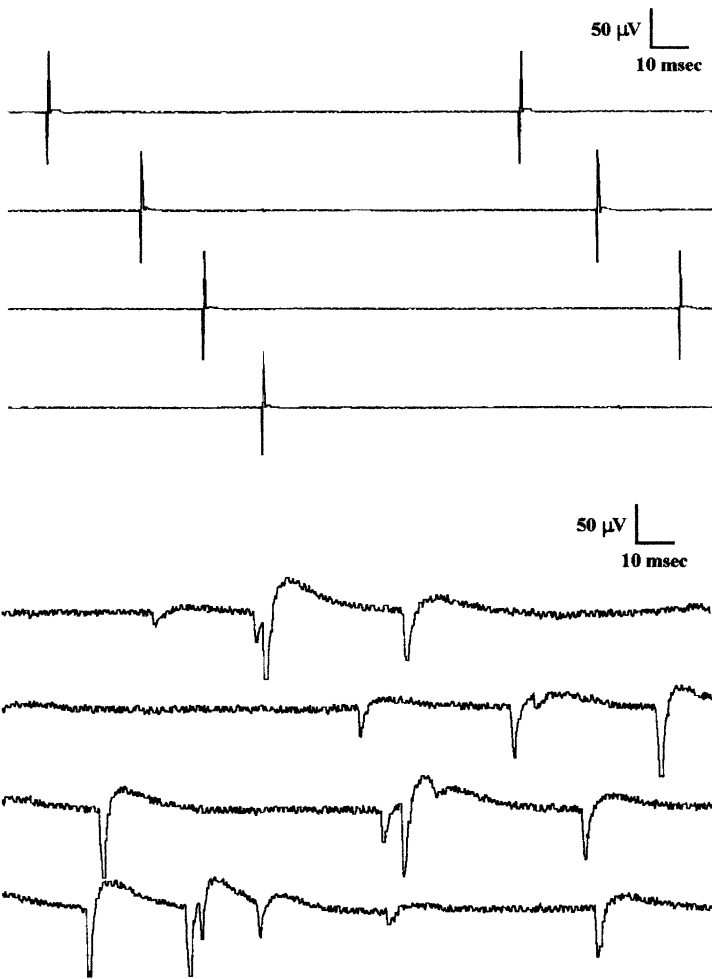


Fig. 5. Fibrillation potentials (*top traces*) and positive sharp waves (*bottom traces*). (Adapted from Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

single muscle fibers. Positive sharp waves have a brief initial positivity followed by a long negative phase, and sound like a dull pop. The amplitude is variable (usually 10–100 μV , occasionally up to 3 mV). Like fibrillation potentials, their firing pattern is regular. This is a key point, because voluntary MUAPs at a distance occasionally have positive wave morphology but can be differentiated by their lack of a regular firing pattern. Positive sharp waves are usually accompanied by fibrillation potentials but may be seen alone, sometimes early in denervation. The mechanism by which MFAPs take on two distinctive morphologies, either a brief spike or a positive wave, involves the actual EMG needle (Fig. 6) [8]. Probably, the needle mechanically

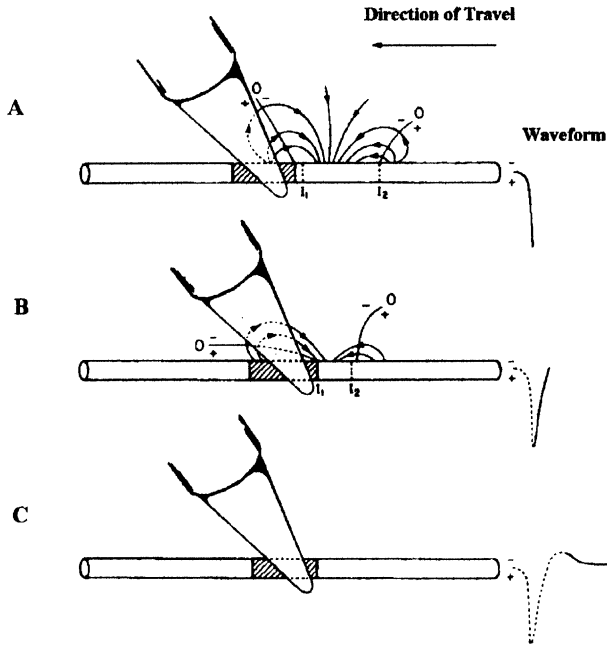


Fig. 6. Generation of a positive wave. The cross-hatched area is deformed by the needle electrode, which becomes electrically inexcitable. As the traveling depolarization wave approaches, an initial positive wave is generated (A–B). With failure of conduction beyond the needle, the steep negative phase is aborted and the waveform returns to baseline (C). Reprinted with permission from Dimitru D. Volume conduction: theory and application. (In: Dumitru D, editor. *Clinical electrophysiology: physical medicine and rehabilitation state of the art reviews*. Philadelphia: Hanley Belfus; 1989; with permission.)

deforms an irritable muscle fiber, thereby rendering part of the membrane electrically inexcitable. When an action potential arises at a distance down the fiber, it can propagate toward the area deformed by the needle but not beyond it, resulting in the positive wave morphology. Supporting this hypothesis is that fibrillation potentials can change to positive sharp waves with needle movement.

Positive sharp waves and fibrillation potentials conventionally are graded from 0 to 4 (0, none present; +1, persistent single trains of potentials [>2 –3 s] in at least two areas; +2, moderate numbers of potentials in three or more areas; +3, many potentials in all areas; +4, full interference pattern of potentials).

Complex repetitive discharges

Complex repetitive discharges (CRDs) result from the depolarization of a single muscle fiber followed by ephaptic spread to adjacent denervated fibers (Fig. 7) [9–11]. If a circus movement is created whereby the original pacemaker muscle fiber is reactivated, a recurrent discharge develops. These

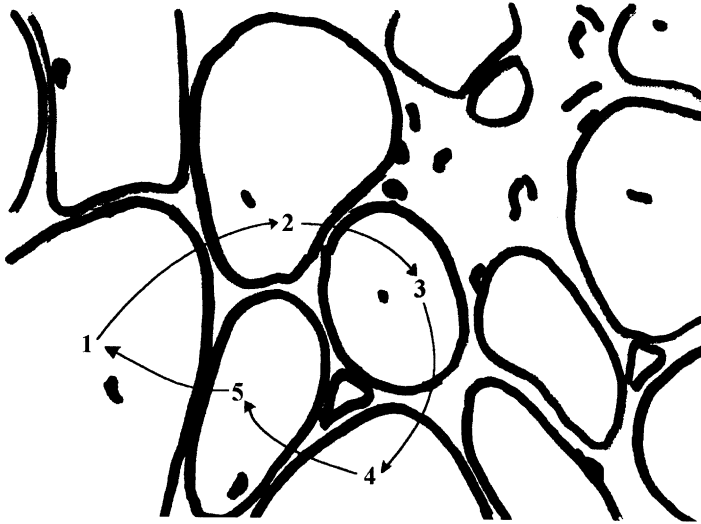


Fig. 7. Pathophysiology of a complex repetitive discharge. Complex repetitive discharges occur from ephaptic transmission from one denervated muscle fiber to an adjacent one. If the original pacemaker is reactivated, a circus movement is formed (1–5) without an intervening synapse. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

discharges usually occur spontaneously or after needle movement. CRDs are recognized on EMG as high frequency (typically 20–150 Hz), multiserated, repetitive discharges with an abrupt onset and termination (Fig. 8). Occasionally, individual phases or additional loops drop in and out, creating an abrupt change in frequency and sound. In rare cases, if the pacemaker is overdriven by another discharge, the CRD may be irregular. As soon as the overdriving pacemaker frequency falls below the inherent frequency of the CRD, the CRD again becomes regular.

CRDs are identical in morphology from one discharge to the next, creating a machine-like sound on EMG. They occur in both chronic neuropathic and myopathic disorders and may arise in any setting where denervated fibers lie adjacent to one another. In neuropathic diseases, this occurs where denervation is followed by reinnervation and subsequent denervation (ie, the pathologic equivalent of grouped atrophy). This situation may also occur in myopathic disorders associated with denervation/reinnervation (eg, inflammatory myopathies) or with muscle fiber splitting.

Myotonic discharges

A myotonic discharge is the spontaneous discharge of a muscle fiber, similar to fibrillation potentials and positive sharp waves. However, it is differentiated from fibrillation potentials and positive sharp waves by the waxing and waning of both amplitude and frequency (Fig. 9) [12]. The firing rate

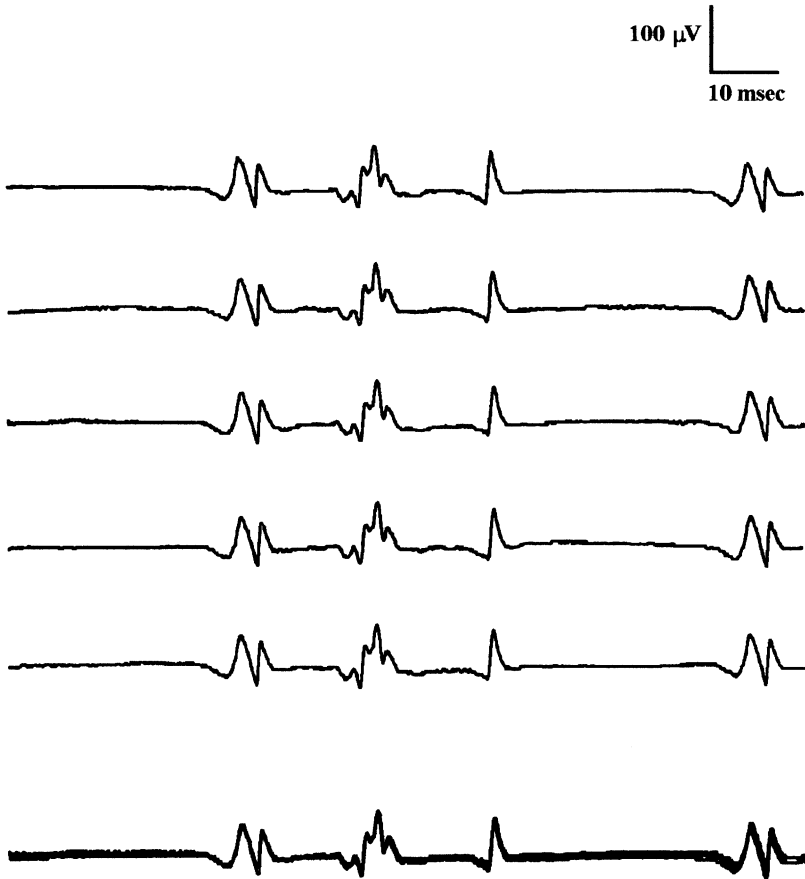


Fig. 8. Complex repetitive discharge (CRD). In the top five traces, note the multiple spikes (each spike within the complex representing a different single muscle fiber) and the repetitive nature. CRD triggered on a delay line. The bottom trace shows all traces superimposed. Note the repetitive nature of a CRD. When superimposed, there is little or no jitter between successive potentials. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

is generally between 20 to 150 Hz. An individual myotonic potential may have either a positive wave or brief spike morphology (identifying the source generator as a muscle fiber). Myotonic discharges are characteristically seen in myotonic dystrophy, myotonia congenita, and paramyotonia congenita. They may also occur in some metabolic, inflammatory, or congenital myopathies (eg, acid maltase deficiency, polymyositis, or myotubular myopathy); in some forms of hyperkalemic periodic paralysis; and may occur rarely in denervation of any cause. A single brief run of myotonic discharges may occur in many denervating disorders; however, this is never the predominant waveform.



Fig. 9. Myotonic discharge. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

Spontaneous activity generated from motor neurons

Fasciculation potentials (Fig. 10)

A fasciculation is a single, spontaneous, involuntary discharge of an individual motor unit [7]. Unlike a voluntary motor unit, fasciculations generally fire slowly and irregularly, usually between 0.1 to 10 Hz, whereas voluntary MUAPs fire initially at 4 to 5 Hz. The source generator is the motor neuron or its axon, prior to its terminal branches. On EMG, fasciculations usually have the morphology of simple MUAPs, or can be complex and large if they represent pathologic motor units. Despite the association of fasciculations with diseases of the anterior horn cell, the actual site of origin of most fasciculations is distal in the axon [13].

Clinically, fasciculations are recognized as individual brief twitches that seldom result in significant movement of a joint. Fasciculations are associated with numerous disease processes affecting the lower motor neuron, which amyotrophic lateral sclerosis is the most well known. However, fasciculations can be seen in many other disorders, including radiculopathies, polyneuropathies, and entrapment neuropathies. In addition, most normal individuals have occasional fasciculations, so-called benign fasciculations.

Differentiating benign from malignant fasciculations on a clinical basis is difficult. Benign fasciculations are not associated with muscle weakness, wasting, or any abnormality of reflexes. In general, benign fasciculations tend to fire faster and affect the same site repetitively (eg, eyelid twitching), as opposed to fasciculations in pathologic conditions that tend to be more random.

Doublets, triplets, multiplets (Fig. 10)

Spontaneous MUAPs that fire in groups of two, three, or multiple potentials are known as doublets, triplets, and multiplets, respectively. These potentials fundamentally have the same significance as fasciculation potentials: they represent the spontaneous depolarization of a motor unit or its axon. Doublets, triplets, and multiplets can be seen in any situation where fasciculation potentials occur (ie, neuropathic conditions), but are also characteristically seen in hypocalcemia. If hypocalcemia results in tetany, the distal muscles are predominantly affected, with involuntary spasms affecting the hands and feet (carpedal spasms). In hands, a characteristic posture

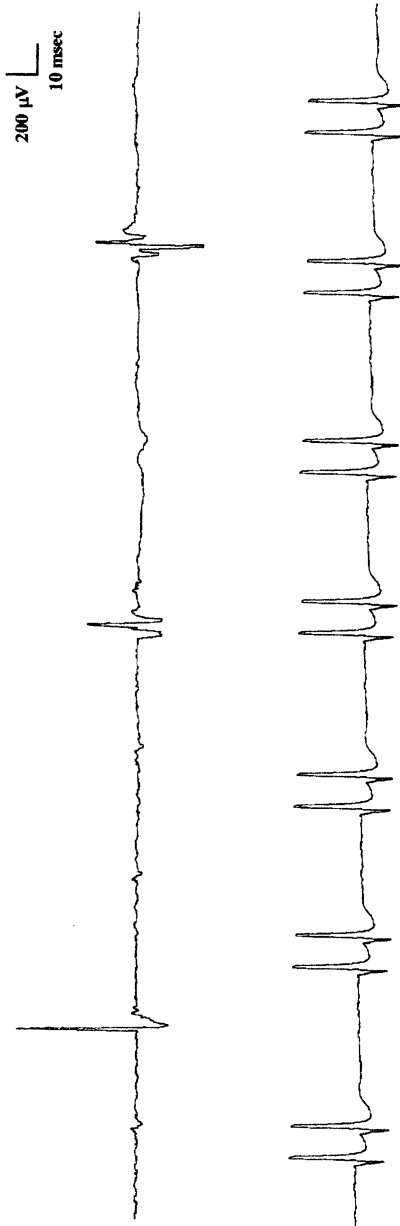


Fig. 10. Fasciculations (*top trace*) and doublets (*bottom trace*).

develops: adduction of the thumb and fingers, extension of the interphalangeal joints, and flexion of the metacarpal-phalangeal joints and wrist. On needle EMG, doublets, triplets, and multiplets are characteristically seen during tetany.

Myokymic discharges

Myokymic discharges are bursting, repetitive discharges of the same MUAP (Fig. 11). The firing frequency within the burst is typically 5 to 60 Hz, with the number of potentials within a burst varying and sometimes changing from burst to burst. The firing frequency between bursts is much slower, typically less than 2 Hz, and produces a marching sound on EMG. The bursting pattern of a myokymic discharge is more easily recognized if the sweep is changed to a longer sweep speed. Myokymic discharges are thought to arise from spontaneous depolarization or ephaptic transmission along demyelinated segments of nerve.

Clinically, myokymia is recognized as continuous involuntary quivering, rippling, or undulating movement of muscle. The finding of myokymia on EMG narrows the differential diagnosis to a limited set of disorders [1,14] (Display Box 1).

Cramp discharges

Clinically, cramps are painful, involuntary contractions of muscle, which tend to occur when a muscle is in the shortened position and contracting. Electrically, cramps are high-frequency discharges of MUAPs, thus marking them as a nerve rather than as a primary muscle phenomenon [1,15,16]. EMG shows either a full interference pattern of MUAPs with a normal morphology, or several MUAPs firing repetitively and sometimes irregularly at

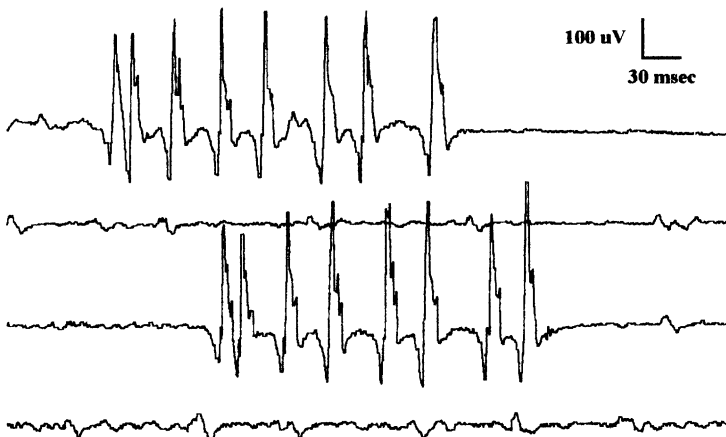


Fig. 11. Myokymic discharges (*rastered traces*). Note the high-frequency pattern within the burst and the slow frequency between the bursts.

Display Box 1**Disorders commonly associated with myokymic discharges**

Radiation injury (usually brachial plexopathy)

Guillain-Barré syndrome (facial)

Multiple sclerosis (facial)

Pontine tumors (facial)

Hypocalcemia

Timber rattlesnake envenomation

Occasionally seen in

Guillain-Barré syndrome (limbs)

Chronic inflammatory demyelinating polyneuropathy

Nerve entrapments

Radiculopathy

From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.

high frequencies (usually 40–60 Hz; Fig. 12). Cramps may be benign (eg, nocturnal calf cramps, postexercise cramps) or can be associated with a wide number of neuropathic, endocrinologic, and metabolic conditions. Clinically, cramps may resemble the contractures that occur in several of the metabolic muscle diseases. However, whereas the needle EMG of a cramp consists of rapidly firing MUAPs, a contracture is typically electrically silent.

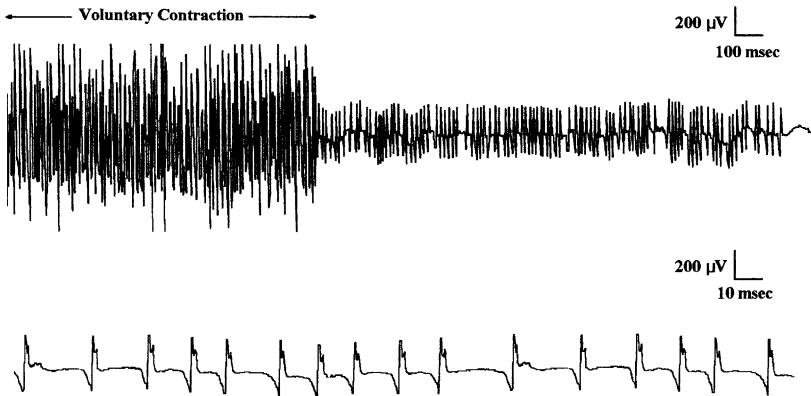


Fig. 12. Cramp discharge. (*Top trace*) The subject is voluntarily contracting strongly, which is followed by a cramp discharge. (*Bottom trace*) During the subject's cramp, EMG shows one or several motor units firing repetitively and sometimes irregularly at high frequencies (usually 40–60 Hz). (From Preston DC, Shapiro BE. *EMG waveforms—video companion to electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1999; with permission.)

Neuromyotonic discharges

Neuromyotonic discharges are high-frequency (150–250 Hz) repetitive discharges of a single MUAP [17], which marks them as a neuropathic phenomenon. They characteristically wane in amplitude and frequency, which results in a pinging sound on EMG (Fig. 13). These discharges are rare and are seen either in chronic motor neuron diseases (eg, poliomyelitis and adult spinal muscular atrophy) or in syndromes of continuous motor-unit activity (CMUA). The nomenclature of the syndromes of CMUA is complicated. These disorders have been described as Isaac's syndrome, neuromyotonia, pseudomyotonia, neurotonia, normocalcemic tetany, and continuous muscle-fiber or motor-unit activity [18]. They share clinical features of generalized stiffness, hyperhidrosis, delayed muscle relaxation, fasciculations, and myokymia [19–21]. Some cases are familial. However, there is increasing evidence that many cases of acquired neuromyotonia or CMUA may have an autoimmune etiology, with the target antigen being peripheral nerve potassium channels [22].

The delay in relaxation and improvement with repetitive use seen in neuromyotonia may be difficult to distinguish clinically from myotonia, which originates in muscle. Electrically, however, the neuromyotonic syndromes are easily differentiated from the muscle myotonias. Whereas the myotonic syndromes are associated with the spontaneous discharges of muscle fibers, the neuromyotonic disorders are associated with involuntary spontaneous discharges of motor units. Other motor neuron/axon discharges often

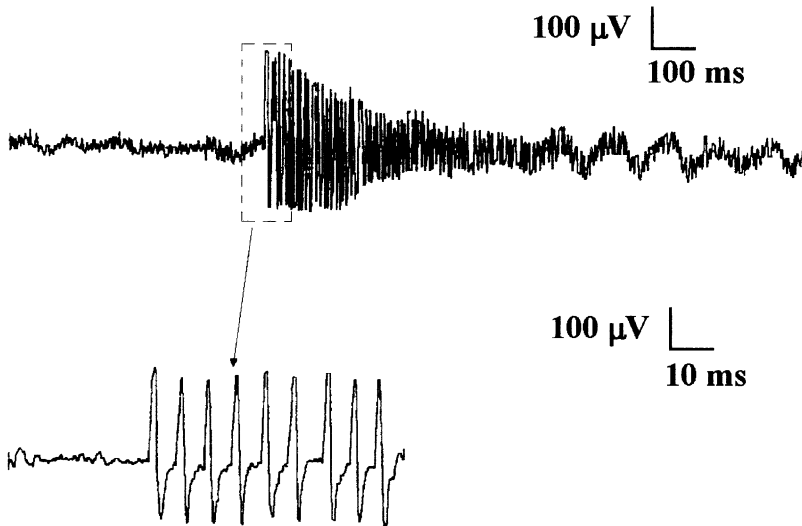


Fig. 13. Neuromyotonic discharges. Enlarged section of top trace shows change in sweep speed which identifies each potential as the same motor unit. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

accompany neuromyotonic discharges, especially fasciculations and myokymic discharges.

Voluntary MUAPs

Following the assessment of insertional and spontaneous activity, the needle EMG examination moves on to the evaluation of voluntary MUAPs. Similar to the analysis of spontaneous activity, MUAPs are assessed for morphology, stability, and firing characteristics. The pattern of MUAP abnormalities usually allows a determination of whether a disorder is primarily neuropathic or myopathic, and often helps determine the time course (acute versus chronic) and severity of the lesion.

Physiology

The basic component of the peripheral nervous system is the motor unit, defined as an individual motor neuron, its axon, and associated neuromuscular junctions and muscle fibers [23]. The extracellular needle EMG recording of a motor unit is the MUAP [5,24–26]. The number of muscle fibers per motor unit varies greatly, from 5 to 10 in laryngeal muscles to hundreds in the soleus. The territory of a motor unit usually ranges from 5 to 10 mm in adults, with many motor-unit territories overlapping with one another. Because of this overlap, two muscle fibers from the same motor unit rarely lie adjacent to each other. Transverse motor-unit territory increases greatly with age, doubling from birth to adulthood, mostly due to the increase in individual muscle fiber size.

When a motor neuron depolarizes to threshold, a nerve action potential is generated and propagates down the axon [27]. Under normal circumstances, this results in all muscle fibers of the motor unit being activated and depolarizing more or less simultaneously. Any variability between muscle fiber depolarization times is due to differences in the length of the terminal axons and NMJ transmission times.

The *size principle* governs many of the properties of motor units [28]. The size of the motor neuron is directly related to the following: (1) the size of the axon, (2) the thickness of the myelin sheath, (3) the conduction velocity of the axon, (4) the threshold to depolarization, and (5) the metabolic type of muscle fibers that are innervated. The larger motor neurons have larger axons, with the thickest myelin sheath (ie, fastest conduction velocity), and the highest threshold to depolarization and connections to type II, fast-twitch muscle fibers. Conversely, the smaller motor units have smaller axons, less myelin sheath, slower conduction velocity, a lower threshold to depolarization, and, in general, connections to type I, slow-twitch muscle fibers. Thus, with voluntary contraction, the smallest motor units with the lower thresholds fire first. As contraction increases, progressively larger

motor units begin to fire. The largest type II motor units fire with maximum contraction. During routine needle EMG, most MUAPs analyzed are the smaller, low-threshold motor units that innervate type I muscle fibers. (Note: this explains the lack of EMG findings in steroid myopathy, which characteristically affects type II fibers.)

During the needle EMG examination, each MUAP recorded represents the extracellular compound potential of the muscle fibers of a motor unit, weighted heavily toward the fibers nearest to the needle [29]. A MUAP amplitude recorded just outside a muscle membrane is 1/10th to 1/100th of the actual transmembrane potential, and decreases rapidly as the distance between the needle and the membrane increases [30]. The classification of a MUAP as normal, neuropathic, or myopathic rests on no single finding. MUAPs must be assessed for morphology (duration, polyphasia, amplitude), stability, and firing characteristics before any conclusions can be reached.

Normal findings

Morphology

For every muscle, MUAP morphology is assessed for duration, amplitude, and number of phases (Fig. 14). However, there is a wide range of normal motor-unit morphology, with large, medium, and small motor units present within each muscle (Fig. 15). Therefore, to determine normal versus abnormal, the mean duration, amplitude, and number of phases are compared with a set of normal values for that particular muscle and age group [16,27]. MUAP morphology also varies depending on the muscle being studied and the patient's age (Table 2). This is found most consistently for MUAP duration. In general, MUAPs in proximal muscles tend to be shorter in duration than those in more distal muscles. MUAP size in adults is larger than in children, primarily due to an increase in the size of muscle fibers during development. In addition, MUAP size is generally larger in older individuals, probably because of drop-out of motor units from the normal effects of aging [24].

Duration. MUAP duration reflects the number of muscle fibers within a motor unit. Typical MUAP duration is 5 to 15 milliseconds [5,25]. Duration is defined as the time from the initial deflection from baseline to the final return of the MUAP to baseline (see Fig. 14). It depends primarily on the number of muscle fibers within the motor unit and the dispersion of their depolarizations over time. Duration lengthens as the number of fibers and the territory of a motor unit increases; it varies directly with age, inversely with temperature, and depends on the individual muscle being studied. Proximal and cranial muscles have shorter duration MUAPs. When performing EMG, it is often more rewarding to listen to the potential than to see it. This is especially true when evaluating MUAP duration, as duration correlates well with pitch. Long-duration MUAPs (low frequencies)

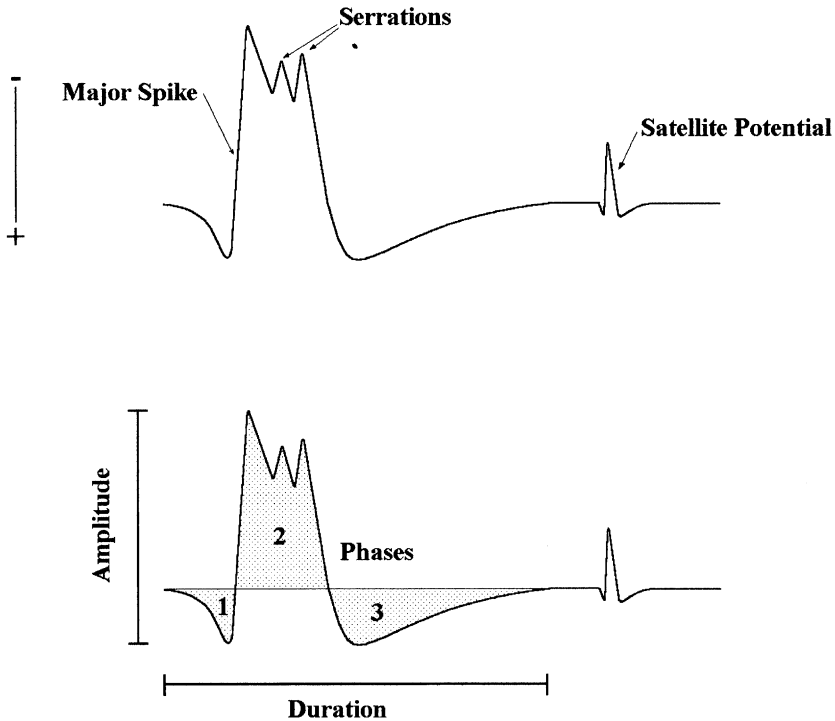


Fig. 14. Motor-unit action potential (MUAP) measurements. The three phases are shown in shaded areas. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

sound dull and thuddy, and short-duration MUAPs (higher frequencies) sound crisp and sharp.

Amplitude. MUAP amplitude varies widely among normals. Most MUAPs have an amplitude greater than 100 μV and less than 2 mV. Amplitude is generally measured from peak to peak of the MUAP (see Fig. 14). Unlike duration, most muscle fibers of a motor unit contribute little to the amplitude [26,30,31]. MUAP amplitude reflects only those few fibers nearest to the needle [29]. Several factors are associated with increased amplitude, including the following: (1) the proximity of the needle to the motor unit, (2) increased number of muscle fibers in a motor unit, (3) increased diameter of muscle fibers (i.e., muscle fiber hypertrophy), and (4) more synchronized firing of the muscle fibers. The amplitude of MUAPs is correlated not with pitch but with the volume, when listening to the EMG.

Polyphasia/serrations/satellite potentials. Polyphasia is a measure of synchrony (i.e., how well muscle fibers within a motor unit fire at the same time) [24]. This is a nonspecific measure and may be abnormal in myopathic and

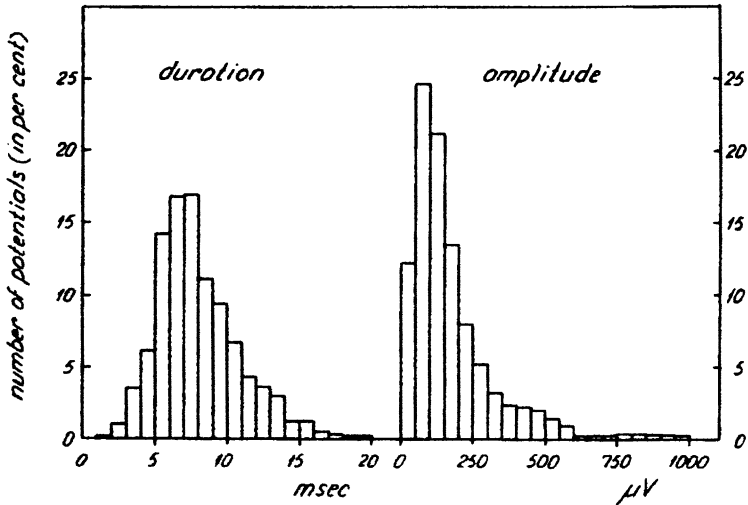


Fig. 15. Range of normal motor-unit action potential (MUAP) duration and amplitude. Histogram of MUAP duration and amplitude in the biceps brachii of a normal subject. Note: both MUAP duration and amplitude vary markedly in normal muscles with small, medium, and large units in the same muscle. MUAP duration or amplitude should not be classified as abnormal based on one or two MUAPs, but requires a mean of many units. (From Buchthal F, Guld C, Rosenfalck P. *Acta Physiol Scand* 1954; with permission.)

neuropathic disorders. The number of phases can easily be calculated by adding 1 to the number of baseline crossings of the MUAP (see Fig. 14). Normally, MUAPs have two, three, or four phases. However, up to 10% of the MUAPs in a muscle may have increased polyphasia, which is considered normal. Note that in the deltoid, up to 25% polyphasia may be normal. Increased polyphasia beyond 10% in most muscles and 25% in the deltoid is abnormal. Polyphasic MUAPs have a high-frequency clicking sound on EMG. Serrations (or turns) are defined as a change in the direction of the potential that does not subsequently cross the baseline. Increased polyphasia and serrations have similar implications, indicating less synchronous firing of muscle fibers within a motor unit. Often, a serration can be changed into an additional phase with needle movement. Satellite potentials (or linked potentials, parasite potentials) are seen in early reinnervation. Following denervation, collateral sprouts from adjacent intact motor units often reinnervate muscle fibers. The newly formed sprout is often small, thinly myelinated, and, therefore, slowly conducting. Because of the slow conduction time and increased distance, reinnervated muscle fibers are seen as time-locked potentials that trail the main MUAP. These satellite potentials are extremely unstable (see later) and may vary slightly in their firing rate, or may block and not fire at all. Over time, the sprout matures and the thickness of the myelin, and consequently the conduction velocity, increases. The satellite

Table 2
Mean MUAP duration based on age and muscle group

Age, yrs	Arm muscles					Leg muscles					
	Deltoid	Biceps	Triceps	Thenar	ADM	Quad, BF	Gastroc	Tib ant	Per long	EDB	Facial
0-4	7.9-10.1	6.4-8.2	7.2-9.3	7.1-9.1	8.3-10.6	7.2-9.2	6.4-8.2	8.0-10.2	6.8-7.4	6.3-8.1	3.7-4.7
5-9	8.0-10.8	6.5-8.8	7.3-9.9	7.2-9.8	8.4-11.4	7.3-9.9	6.5-8.8	8.1-11.0	5.9-7.9	6.4-8.7	3.8-5.1
10-14	8.1-11.2	6.6-9.1	7.5-10.3	7.3-10.1	8.5-11.7	7.4-10.2	6.6-9.1	8.2-11.3	5.9-8.2	6.5-9.0	3.9-5.3
15-19	8.6-12.2	7.0-9.9	7.9-11.2	7.8-11.0	9.0-12.8	7.8-11.1	7.0-9.9	8.7-12.3	6.3-8.9	6.9-9.8	4.1-5.7
20-29	9.5-13.2	7.7-10.7	8.7-12.1	8.5-11.9	9.9-13.8	8.6-12.0	7.7-10.7	9.6-13.3	6.9-9.6	7.6-10.6	4.4-6.2
30-39	11.1-14.9	9.0-12.1	10.2-13.7	10.0-13.4	11.6-15.6	10.1-13.5	9.0-12.1	11.2-15.1	8.1-10.9	8.9-12.0	5.2-7.1
40-49	11.8-15.7	9.6-12.8	10.9-14.5	10.7-14.2	12.4-16.5	10.7-14.3	9.6-12.8	11.9-15.9	8.6-11.5	9.5-12.7	5.6-7.4
50-59	12.8-16.7	10.4-13.6	11.8-15.4	11.5-15.1	13.4-17.5	11.6-15.2	10.4-13.6	12.9-16.9	9.4-12.2	10.3-13.5	6.0-7.9
60-69	13.3-17.3	10.8-14.1	12.2-15.9	12.0-15.7	13.9-18.2	12.1-15.8	10.8-14.1	13.4-17.5	9.7-12.7	10.7-14.0	6.3-8.2
70-79	13.7-17.7	11.1-14.4	12.5-16.3	12.3-16.0	14.3-18.6	12.4-16.1	11.1-14.4	13.8-17.9	10.0-13.0	11.0-14.3	6.5-8.3

Abbreviations: ADM, abductor digiti minimi; BF, biceps femoris; EDB, Extensor digitorum brevis; MUAP, motor-unit action potential; Quad, quadriceps; Tib ant, tibialis anterior.

(From Buchthal F, Rosenfalck P. Action potential parameters in different human muscles. *Acta Psych Neurol Scand*, copyright 1955, Munsgaard International Publishers. Copenhagen, Denmark; with permission.)

potential then fires more closely to the main potential and will ultimately become an additional phase or serration within the main complex.

Stability

MUAPs are usually stable in morphology from potential to potential. This stability is because each time a nerve action potential is generated, there is normally effective transmission across the NMJ, and all muscle fibers of the motor unit fire. If there is impaired NMJ transmission, unstable MUAPs may result. Unstable MUAPs occur when individual muscle fibers are either blocked or come to action potential at varying intervals, leading to an MUAP that changes in configuration from impulse to impulse. Either the amplitude and/or number of phases or serrations changes between potentials. Although unstable MUAPs always indicate unstable NMJs, they may occur not only in primary disorders of the NMJ (eg, myasthenia gravis), but are often seen as a secondary phenomenon in neuropathic and myopathic disorders. Any disorder associated with denervation may demonstrate unstable MUAPs.

Firing pattern

One of the most difficult tasks for the electromyographer is the assessment of firing pattern and its relationship to the number of MUAPs. MUAPs normally fire in a semi-rhythmic pattern—that is, there is slight variation in the time interval between the same consecutive MUAP (Fig. 16). This unique firing pattern helps identify the potential as an MUAP under voluntary control. When an individual is asked to slowly activate a muscle, a single motor unit begins firing semi-rhythmically at 4 to 5 Hz [32,33]. As force is increased, the first motor unit increases its firing rate followed by a second motor unit firing, and so forth. This process continues, with the firing rate increasing and additional motor units being recruited, as force is increased. *Normally, the ratio of firing frequency to the number of different MUAPs firing is approximately 5 to 1 [2].* Thus, when the firing frequency of the first MUAP reaches 10 Hz, a second MUAP should begin to fire; by 15 Hz, a third unit should fire, and so forth. During maximal contraction, multiple MUAPs normally overlap and create an *interference pattern* in which it is difficult to discern individual MUAPs. For most muscles, the maximal firing frequency is 30 to 50 Hz. Important exceptions include quick ballistic contractions, in which the firing frequency may transiently reach 100 Hz, and muscles that are predominantly slow twitch (eg, soleus), in which the maximal firing frequency is approximately 15 Hz.

When assessing MUAP firing pattern, two key parameters must be determined: activation and recruitment. *Activation* refers to firing rate and represents a central process. Poor activation (ie, low firing rate) may be seen in diseases of the central nervous system (CNS) or as a manifestation of pain, poor cooperation, or functional disorders. *Recruitment* refers to the ability to add motor units as the firing rate increases (Fig. 17). Recruitment is

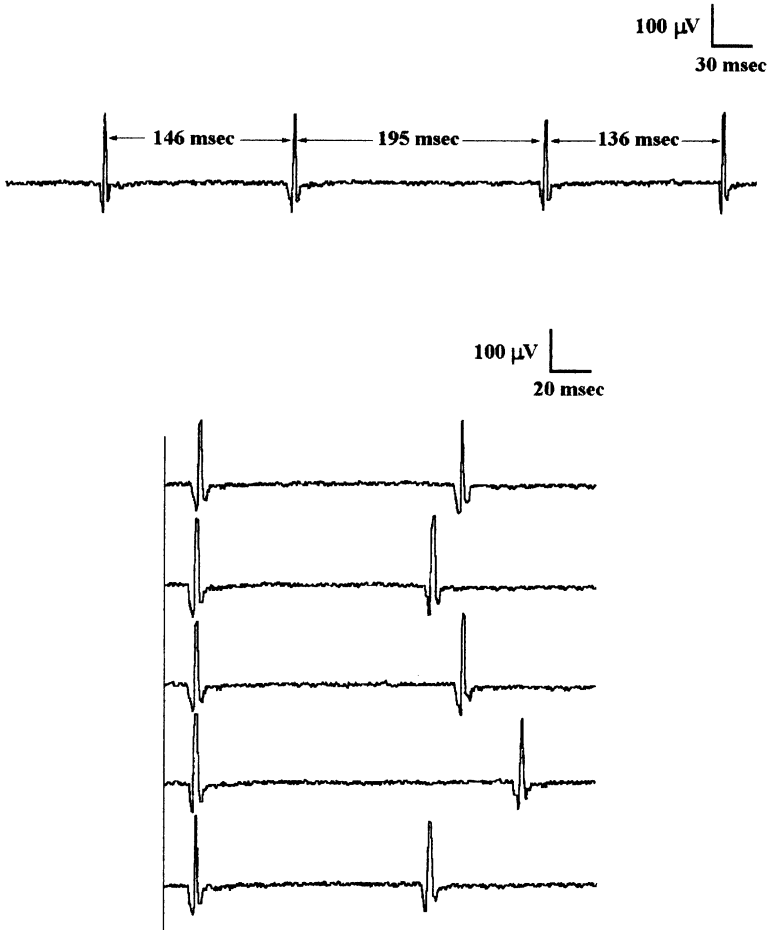


Fig. 16. Motor-unit action potential (MUAP) firing pattern. (*Top trace*) Single voluntary MUAP firing at ≈ 6 Hz. Note variation in interpotential intervals. (*Bottom traces*) Single voluntary MUAP placed on a delay line and rastered. First potential of each trace triggers the sweep. Note the variation between firing time of the next consecutive MUAP. The pattern is not quite regular (ie, semi-rhythmic). This firing pattern is only seen with voluntarily activated MUAPs. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

reduced primarily in neurogenic diseases, and rarely, in severe endstage myopathy. The key question to answer in assessing recruitment is: Are the number of different MUAPs firing appropriate for the firing rate? Or, is the ratio of firing rate to number of MUAPs approximately 5 to 1? It is essential to appreciate that an incomplete interference pattern may be due to either poor activation or poor recruitment (Fig. 18). Many electromyographers judge recruitment only during maximum contraction, by examining the interference pattern. However, recruitment is more easily evaluated during

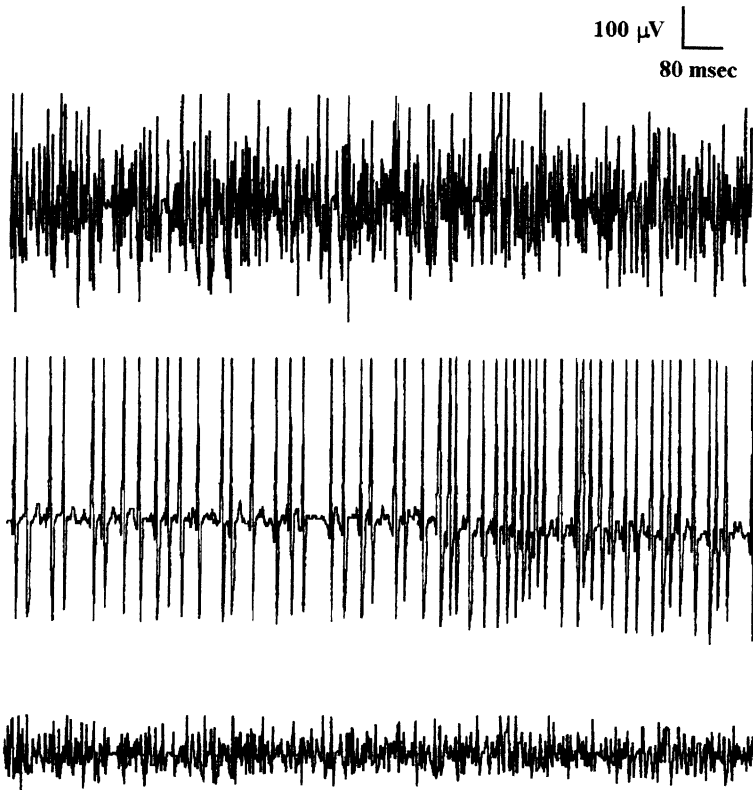
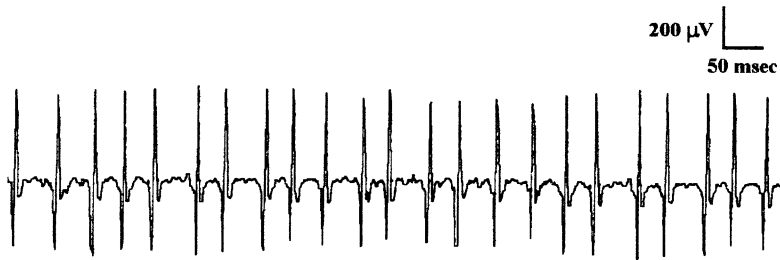


Fig. 17. Interference patterns. (*Top*) Normal trace. (*Middle*) Neurogenic trace. (*Bottom*) Myopathic trace. In each trace, the patient is asked to maximally contract. (From Preston DC, Shapiro BE. Electromyography and neuromuscular disorders. Boston: Butterworth-Heinemann; 1998; with permission.)

moderate levels of contraction. For instance, if only one MUAP is firing at 15 to 20 Hz (medium level of activation), then recruitment is decreased, regardless of the interference pattern. There is no need to increase the firing rate using maximal contraction, in order to assess recruitment. Furthermore, maximal contraction with the EMG needle in the muscle is often perceived as more painful and is best avoided or minimized.

The final concept to understand when assessing MUAP firing pattern is that of early recruitment. In diseases where individual muscle fibers drop out from a motor unit (eg, myopathies, periodic paralysis or NMJ diseases with block), the motor unit becomes smaller and subsequently can generate less force. Because each motor unit generates less force, many motor units must fire to generate even a small amount of force. This is known as early recruitment, which refers to the inappropriate firing of many motor units to generate a small amount of force. On the screen, many MUAPs appear to fire almost simultaneously when the patient is asked to contract the muscle



Firing Frequency = 30 Hz



Firing Frequency = 5 Hz

Fig. 18. Incomplete interference patterns. In both traces, the patient is asked to maximally contract their muscle with the needle EMG in place. The top trace demonstrates an incomplete interference pattern due to reduced recruitment. The bottom trace demonstrates an incomplete interference pattern due to reduced activation. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

minimally. Usually, only the electromyographer who is performing the study can assess early recruitment, since this judgement requires knowledge of how much force is being generated.

Patterns of MUAP abnormalities

The morphology and firing pattern of the MUAP can usually discriminate among the various disorders affecting the motor unit. No single parameter identifies an MUAP as myopathic, neuropathic, or associated with an NMJ disorder. Rather, specific patterns of abnormalities in MUAP morphology and firing pattern reflect whether the underlying disorder is (1) acute, chronic, or end stage; (2) neuropathic, myopathic, or associated with an NMJ transmission defect; or (3) if neuropathic, whether the primary pathophysiology is axonal loss or demyelination (Table 3).

Acute neuropathic disorders—axonal loss

Following an acute axonal injury, Wallerian degeneration occurs within the first week, followed by denervation of muscle fibers of the involved motor units [34]. Reinnervation normally occurs as surviving nearby axons

Table 3
MUAP patterns and pathophysiology

	MUAP morphology			MUAP firing pattern	
	Duration	Amplitude	Phases	Activation	Recruitment
Acute neuropathic—axonal	Normal	Normal	Normal	Normal	↓
Chronic neuropathic—axonal	↑	↑	↑	Normal	↓
Neuropathic—demyelinating (CV slowing)	Normal	Normal	Normal	Normal	Normal
Neuropathic—demyelinating (conduction block)	Normal	Normal	Normal	Normal	↓
Early reinnervation after severe denervation (nascent units)	↓	↓	↑	Normal	↓↓
Acute myopathic	↓	↓	↑	Normal	Normal/early
Chronic myopathic	↓/↑	↓/↑	↑	Normal	Normal/early
Myopathic—end stage	↓/↑	↓/↑	↑	Normal	↓↓
NMJ disorders—mild	Normal	Normal	Normal	Normal	Normal
NMJ disorders—intermittent block	Normal/↓*	Normal/↓*	Normal/↑*	Normal	Normal/early
NMJ disorders—severe block	↓	↓	↑	Normal	↓↓
CNS disorders	Normal	Normal	Normal	↓↓	Normal

Abbreviations: CNS, central nervous system; MUAP, motor-unit action potential; NMJ, neuromuscular junction; ↑, increased; ↓, decreased; ↓/↑, may be decreased and/or increased; ↓↓, usually markedly decreased; *, may vary from potential to potential (unstable MUAPs).

(From Preston DC, Shapiro BE. Electromyography and neuromuscular disorders. Boston: Butterworth-Heinemann; 1998; with permission.)

form sprouts, which grow and eventually reinnervate denervated fibers. When this occurs, the number of muscle fibers in the reinnervated MUAP is larger than normal, leading to an MUAP with increased duration, amplitude, and number of phases. However, this process takes time, usually many weeks to months. In the acute setting, MUAP morphology remains normal. The only abnormality seen on EMG in an acute neuropathic lesion is a decreased recruitment pattern in weak muscles, due to loss of motor units. The acute neuropathic pattern associated with axonal loss characteristically occurs in the first several weeks following trauma, compression, or nerve infarction. The only other situation in which a similar needle EMG pattern occurs is with a relatively pure demyelinating lesion with conduction block (see later).

Chronic neuropathic disorders—axonal loss

Following axonal loss and denervation, the process of reinnervation occurs by one of two mechanisms. If there has been complete denervation, then the only possible mechanism for reinnervation is through axonal regrowth from the point of injury (see later discussion on early reinnervation following severe or complete denervation). In contrast, if there is partial or gradual denervation, which is the more common scenario, reinnervation usually occurs through collateral sprouting by adjacent surviving motor

units. As the number of muscle fibers per motor unit increases, MUAPs become prolonged in duration, with a high amplitude, and increased polyphasia (Fig. 19). These changes in MUAP configuration, in conjunction with decreased recruitment of MUAPs, are the hallmarks of reinnervated motor units, and nearly always imply chronic neuropathic disease. Long-duration, high-amplitude, polyphasic MUAPs are never seen acutely. When present, they always imply that the process has been present for at least several weeks, and more often months or years.

Neuropathic disorders—demyelinating lesions

Loss of axons results in denervation and ultimately reinnervation with resultant changes in MUAP morphology. If, however, the pathology is purely or predominantly demyelinating, the underlying axon remains intact. Thus, there is neither denervation nor subsequent reinnervation. Consequently, in pure demyelinating lesions, MUAP morphology remains normal. If demyelination results in conduction velocity slowing alone, the number of functioning motor units remains normal. In this case, there will be no change in either MUAP morphology or recruitment pattern. How-

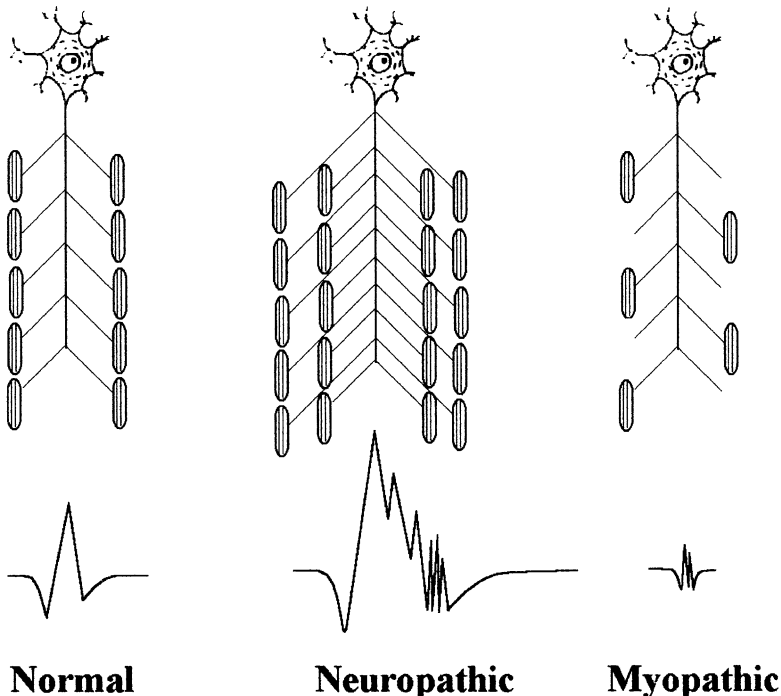


Fig. 19. Motor-unit action potential morphologies. (From Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders*. Boston: Butterworth-Heinemann; 1998; with permission.)

ever, if demyelination results in conduction block, then the number of available MUAPs effectively decreases. In this situation, the firing pattern shows decreased recruitment, although MUAP morphology remains normal. This pattern of reduced recruitment with normal MUAP morphology is seen only in demyelinating lesions with conduction block or in cases of acute axonal loss before enough time has passed for reinnervation to occur.

Acute myopathic disorders

In myopathies, the number of functioning muscle fibers in a motor unit decreases. Because there are fewer muscle fibers per motor unit, this results in shorter-duration and smaller-amplitude MUAPs [35–37] (see Fig. 19). In addition, there is less synchronous firing, and consequently polyphasia, of MUAPs, due to dysfunction of the remaining muscle fibers. However, the actual number of functioning motor units remains normal. Thus, the recruitment pattern remains normal for the level of activation. However, because each motor unit contains fewer muscle fibers, it cannot generate as much force as a normal motor unit. To compensate, more MUAPs will fire than normally needed for a certain level of force, resulting in early recruitment. Consequently, the pattern associated with an acute myopathy is short-duration, small-amplitude, polyphasic MUAPs, with normal or early recruitment.

Chronic myopathic disorders

In chronic myopathies, especially those with inflammatory or necrotic features (e.g., polymyositis, muscular dystrophies), some denervation and subsequent reinnervation commonly occurs. Consequently, long-duration, high-amplitude, polyphasic MUAPs can develop, although such MUAPs are most commonly seen in chronic neuropathic disorders. In many chronic myopathies, two populations of MUAPs are seen, often in the same muscle: long-duration, high-amplitude, polyphasic MUAPs in combination with short-duration, small-amplitude, polyphasic MUAPs. Rarely, only long, large, polyphasic MUAPs are seen. The key to differentiating chronic myopathic from neuropathic MUAPs is the assessment of recruitment pattern. In chronic myopathies, recruitment is usually normal or early. If an early recruitment pattern is not seen, then, at the least, the recruitment pattern appears better than what would be expected from the chronic MUAP changes. In some cases of the most chronic myopathy (especially inclusion body myositis), the EMG pattern may resemble that of active motor neuron disease (fibrillation potentials; long-duration, high-amplitude, polyphasic MUAPs), with the exception of recruitment.

Myopathic disorders—end stage

In the latest stages of some muscular dystrophies, in periodic paralysis, and in unusual, very chronic focal myopathies (eg, inclusion body myositis),

end-stage muscle may occur. In these situations, if every muscle fiber of some motor units is damaged, the actual number of motor units may effectively decrease. This results in an unusual pattern of reduced recruitment of short-duration, small-amplitude, polyphasic MUAPs, either alone or in combination with long-duration, high-amplitude, polyphasic MUAPs. Although decreased recruitment nearly always signifies neuropathic disease, the rare exception arises in end-stage muscle from myopathy.

Reinnervation following severe denervation

Reinnervation occurs most frequently from collateral sprouting by adjacent surviving motor units. If there is severe or complete denervation, with no nearby surviving axons, the only possible mechanism for reinnervation is regrowth of the axon from the site of injury. As the axon regrows, it will eventually reinnervate some, but not all of the original muscle fibers. At that point, the MUAP will be short-duration, small-amplitude, and polyphasic, similar in morphology to an acute myopathic motor unit (Fig. 20). Early reinnervated motor units following severe denervation are known as nascent motor units. The key factor that differentiates nascent motor units from myopathic motor units is the recruitment pattern. Nascent MUAPs are always seen in the context of markedly reduced recruitment, whereas myopathic MUAPs are seen in the context of normal or early recruitment.

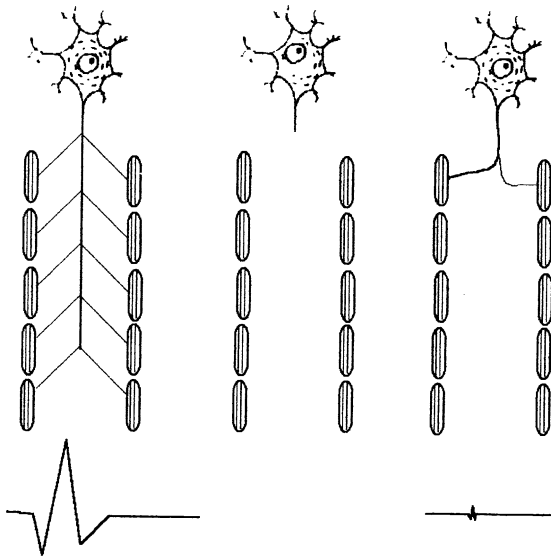


Fig. 20. Nascent motor units. (Left) Normal trace. (Middle) Following a severe axonal lesion, Wallerian degeneration occurs distal to the injury resulting in denervation. (Right) Trace shows that if there are no surviving nearby axons, reinnervation can occur only from axonal regrowth from the terminal stump.

Neuromuscular junction disorders

MUAP morphology and firing patterns in neuromuscular junction disorders depend on the severity of the disorder. If the NMJ disorder is mild, both the morphology and recruitment of the MUAP will be normal. If the disorder is more severe, resulting in the intermittent blocking of some muscle fibers within the motor unit, the MUAP will become unstable. In this situation, the morphology (amplitude and/or the number of phases) will vary from potential to potential. With greater and more persistent block, individual muscle fibers within a motor unit are effectively lost. In this case, the MUAP becomes short, small, and polyphasic, similar to a myopathic MUAP. Similarly, recruitment remains normal, or may become early as each motor unit can generate less force. Finally, in cases of severe NMJ block, such as botulism, all the fibers in some motor units may be blocked, effectively resulting in the loss of motor units. In these cases, the remaining MUAPs are of short duration, small amplitude and polyphasic, but with decreased recruitment, which reflects the reduced number of available motor units. This uncommon pattern can also be seen in end-stage myopathy and in nascent motor units.

CNS disorders

In CNS disorders, there is normally no loss of anterior horn cells, and accordingly, no denervation or reinnervation. MUAP morphology and recruitment remain normal. On needle EMG, weakness is demonstrated as the inability to fire motor units rapidly (ie, reduced activation). Thus, although the interference pattern is incomplete, with a reduced number of motor units firing, the actual number of motor units firing (ie, recruitment) is appropriate for the reduced level of activation.

Occasionally, other patterns may be seen with CNS disorders. In spinal cord lesions, motor units may be lost at the level of the lesion, due to segmental loss of anterior horn cells. For example, in a C6 spinal cord lesion, denervation, reinnervation, and decreased recruitment may be seen in the biceps, deltoid, and other C6 innervated muscles. In the lower extremities, however, only decreased activation will be seen, and recruitment remains normal. Muscles partially supplied by the C6 root (eg, the pronator teres with C6-7 innervation) may show both decreased recruitment and activation. Only rarely are other EMG abnormalities seen in CNS disorders. In some patients with multiple sclerosis, signs of denervation and reinnervation may be seen, presumably from involvement of motor fibers leaving the anterior horn cell in the spinal cord prior to exiting and becoming motor roots. Whether EMG abnormalities can be seen in other CNS disorders, especially stroke, remains controversial. Stroke patients are susceptible to entrapment and compression palsies because of poor mobility, which more often explains any EMG abnormalities.

Last, tremor may occur in some CNS disorders, which can complicate the interpretation of both spontaneous activity and MUAP morphology. Tre-

mor is recognized as a bursting pattern of voluntary MUAPs separated by relative silence. As multiple MUAPs fire simultaneously, the morphology of individual MUAPs may be difficult to assess, and polyphasia appears increased. When tremor occurs at rest (eg, Parkinson's disease), the spontaneous bursting discharge may be mistaken for myokymia. Although both myokymia and tremor result in a bursting pattern of MUAPs, the major difference is that in myokymia the same MUAP fires repetitively in a burst, whereas in tremor, the burst is composed of many different MUAPs. In addition, most patients can voluntarily alter their tremor by changing their limb position or action, as opposed to myokymia, which cannot be voluntarily influenced by the patient.

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