14 Basic Electromyography:
Analysis of Spontaneous Activity

The recognition of abnormal spontaneous activity is one of the most important parts of the needle electromyography (EMG) examination. The presence of abnormal spontaneous activity on EMG can yield several key pieces of information. First, the distribution of abnormal spontaneous activity may indicate the neuroanatomic localization of the lesion. For example, in an isolated 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 only with specific disorders. For example, myotonic discharges are seen only in a few myopathies and in hyperkalemic periodic paralysis. Third, the degree or amount of spontaneous activity often helps to determine the severity of the lesion. Finally, the presence of abnormal spontaneous activity may 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 identification of any spontaneous activity can be achieved by either pattern recognition or analysis of the waveform. With experience, the characteristic sound and appearance of each waveform become easily recognizable. However, when first learning needle EMG or when encountering an unusual waveform, one must be able to systematically analyze the waveform according to the following attributes: (1) morphology, (2) stability, and (3) firing characteristics (Figure 14–1). Using this information, nearly every spontaneous waveform can be identified correctly.

Morphology

The source of a spontaneous discharge often can be identified by its distinctive morphology, including the size and shape of the potential (amplitude, duration, number of

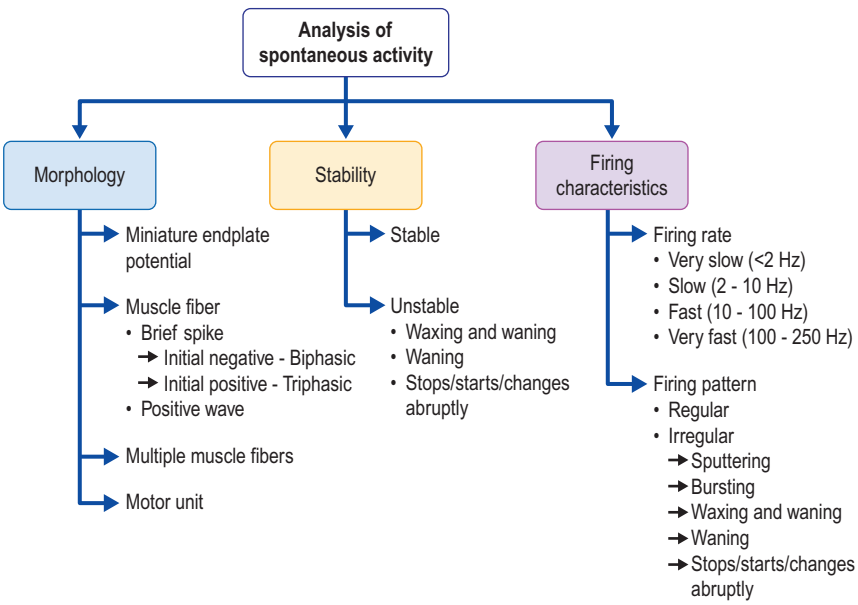

FIGURE 14–1 Algorithm for analysis of spontaneous activity.

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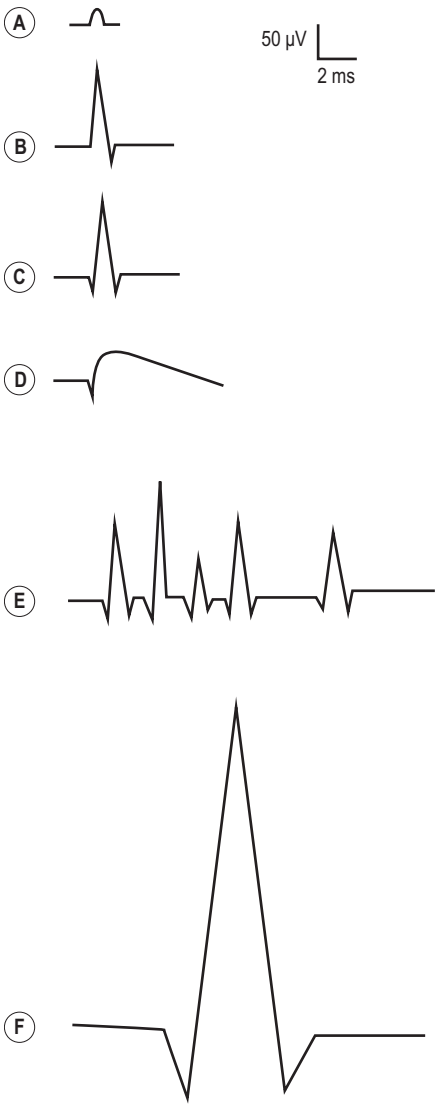

FIGURE 14–2 Spontaneous waveform morphologies. **A:** Miniature endplate potential (monophasic negative). **B:** Muscle fiber action potential, brief spike morphology. Triggered by needle-induced depolarization of a terminal nerve twig (initial negative, diphasic).
C: Muscle fiber action potential, brief spike morphology (initial positive, triphasic). **D:** Muscle fiber action potential, positive wave morphology (initial positive, slow negative). **E:** Multiple different muscle fiber action potentials linked together. **F:** Motor unit action

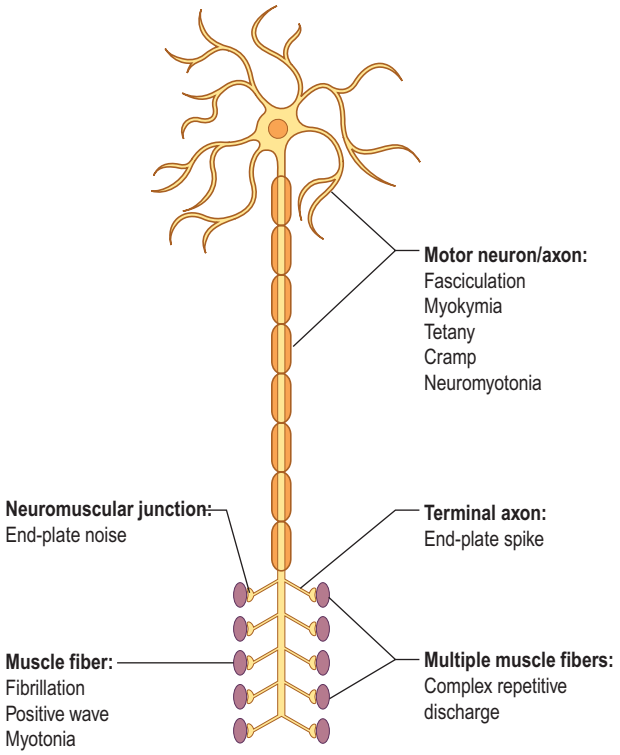

FIGURE 14–3 Spontaneous waveform source generators. Spontaneous activity originates from a variety of source generators. Each generator is associated with a specific morphology.

phases) and its initial deflection (Figure 14–2). By defining its source generator, the type of discharge usually can be identified. The source generators that must be differentiated include (1) the neuromuscular junction (NMJ), (2) a single muscle fiber, (3) the terminal axon twig, (4) a motor neuron/axon, and (5) multiple muscle fibers linked together (Figure 14–3, Table 14–1).

At the NMJ (i.e., endplate zone), miniature endplate potentials (MEPPs) occur spontaneously. They result from the normal spontaneous exocytosis of individual quanta of acetylcholine traveling across the NMJ, leading to a non-propagated, subthreshold endplate potential. If the EMG needle is near the endplate zone, MEPPs can often be recorded. They have a distinctive small amplitude and monophasic negative morphology (Figure 14–2A). These potentials are normal spontaneous discharges and are referred to as *endplate noise*.

When a muscle fiber depolarizes to threshold, a muscle fiber action potential (MFAP) is created. The MFAP can assume one of two basic morphologies, either a brief spike or a positive wave. The brief spike typically is 1 to 5 ms in duration, biphasic or triphasic, with a low amplitude (typically 10–100 µV). This brief spike morphology is seen most often when a muscle fiber depolarizes spontaneously, for example, in denervation, but it can also occur as the result of an individual terminal axon twig depolarizing and then propagating across the NMJ to create an MFAP. Attention to the initial deflection and to whether the brief spike is biphasic or triphasic often can help distinguish between the two (Figure 14–4). If the depolarization begins under the recording needle electrode, a biphasic potential is seen, wherein an initial negative peak is followed by a short positive phase (Figure 14–2B). This signifies that the needle is at the endplate zone, where the depolarization begins, and usually is the result of the EMG needle irritating the terminal nerve twigs near the endplate zone. A nerve twig action potential then leads to an MFAP, known as an *endplate spike*, which is a normal finding (see section on Endplate Spikes). The reason for the initial negativity is similar

Table 14–1. Spontaneous Activity

Potential	Source Generator/ Morphology	Sound on Loudpeaker	Stability	Firing Rate	Firing Pattern
Endplate noise	Miniature endplate potential (monophasic negative)	Seashell	–	20–40 Hz	Irregular (hissing)
Endplate spike	Muscle fiber initiated by terminal axonal twig (brief spike, diphasic, initial negative)	Sputtering, like fat in a frying pan	Stable	5–50 Hz	Irregular (sputtering)
Fibrillation potential	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 (occ. up to 30 Hz)	Regular
Positive sharp wave	Muscle fiber (diphasic, initial positive, slow negative)	Dull pops, rain on a roof or tick-tock of a clock	Stable	0.5–10 Hz (occ. up to 30 Hz)	Regular
Myotonic discharge	Muscle fiber (brief spike, initial positive, or positive wave)	Revving engine	Waxing/waning amplitude	20–150 Hz	Waxing/waning
Complex repetitive discharge	Multiple muscle fibers time-linked together	Machine	Usually stable; may change in discrete jumps	5–100 Hz	Perfectly regular (unless overdriven)
Fasciculation potential	Motor unit (motor neuron/axon)	Corn popping	Stable	Low (0.1–10 Hz)	Irregular
Doublets, triplets, multiplets	Motor unit (motor neuron/axon)	Horse trotting	Usually stable; may change in number of potentials	Variable (1–50 Hz)	Bursts of twos threes or a few potentials
Myokymic discharge	Motor unit (motor neuron/axon)	Marching soldiers	Usually stable; the number of potentials may change within the burst	1–5 Hz (interburst) 5–60 Hz (intraburst)	Bursting of the same individual motor unit potential
Cramp potential	Motor unit (motor neuron/axon)		Usually stable	High (20–150 Hz)	Interference pattern or one or more individual motor unit potentials
Neuromyotonic discharge	Motor unit (motor neuron/axon)	Pinging	Decrementing amplitude	Very high (150–250 Hz)	Waning
Rest tremor	Motor unit (motor neuron/axon)	Marching soldiers	Rising and falling amplitude	1–5 Hz (interburst)	Bursting – synchronous bursting of many different motor unit potentials

to that of the compound muscle action potential (CMAP) in motor nerve conduction studies, wherein the initial deflection is negative when the active recording electrode is properly placed over the motor endplate zone. Otherwise, brief spikes that occur from the spontaneous depolarization of a muscle fiber are associated with an initial positive, usually triphasic morphology. When the 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 (Figure 14–2C).

In addition to the brief spike, an MFAP can assume a positive wave morphology, with an initial brief positive phase followed by a long negative phase (Figure 14–2D). Both positive waves and initial positive, triphasic brief spikes are seen most often as denervating potentials, known as *positive sharp waves* and *fibrillation potentials*,

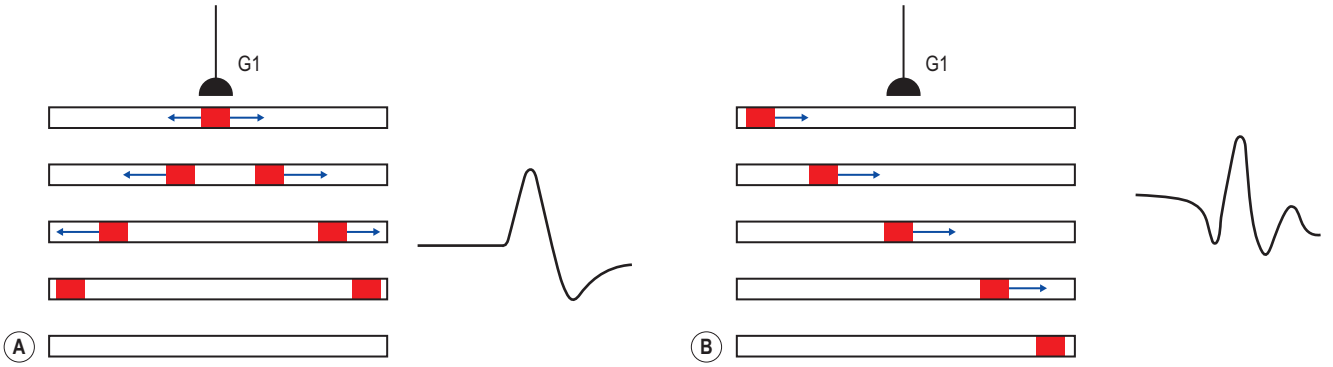

FIGURE 14–4 Waveform morphology and site of depolarization. **A:** Traveling depolarizing wave will create a biphasic potential if the waveform begins under the recording needle electrode (initial negative peak) and then moves away from the electrode (positive peak). An endplate spike shows this morphology. **B:** 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. Fibrillation potentials show this morphology. Endplate spikes are differentiated from fibrillation potentials by the absence of an initial positive deflection because the depolarization begins at the endplate.

respectively. However, it should not be surprising that myotonic discharges, which also originate in muscle fibers, have the same basic morphology as denervating potentials, either positive waves or brief spikes. This point exemplifies the important concept that morphology alone cannot be used to identify a potential. Although the morphology of a potential usually can be used to correctly identify its source generator, additional information regarding its stability and firing characteristics is needed to fully characterize and identify any potential (see later).

The next major category of spontaneous discharges is that which 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 (Figure 14–2F) known as a *motor unit action potential* (MUAP). Spontaneous discharges generated by the motor neuron or its axon include fasciculation potentials, doublets, triplets, and multiplets, myokymic discharges, neuromyotonic discharges, and cramp potentials, all of which lie along the spectrum of abnormal spontaneous MUAPs. They can be differentiated from each other, however, by their stability and firing characteristics (described in the following subsections). If the motor unit is normal, the MUAP morphology will be normal: typically two to four phases, 5 to 15 ms in duration, and variable amplitude depending on the needle position. If the motor unit is pathologic, the number of phases, duration, and amplitude of the MUAP may be abnormal. Differentiating a MUAP from a single MFAP usually is straightforward and typically can be done quite simply by analyzing its duration and amplitude.

The last distinctive waveform that must be recognized is that of time-linked individual muscle fibers, such as occurs in complex repetitive discharges. One might ask how this waveform differs from an MUAP, which also represents many muscle fibers linked together. The difference is that the muscle fibers in a motor unit fire more or less synchronously and, in almost every situation, summate to create a larger potential 5 to 15 ms in duration. In contrast, the multiple muscle fibers in a complex repetitive discharge fire consecutively and usually are discernible as individual spikes that are time linked together (Figure 14–2E).

Stability

Assessment of the stability of a waveform can be very informative. Nearly all spontaneous potentials are relatively stable in their morphology. If the morphology of the potential changes, note should be made of whether it waxes and wanes, wanes (decrements), or changes abruptly. MFAPs that wax and wane in amplitude are characteristically seen in myotonic discharges. Marked decrementing of an MUAP amplitude occurs in neuromyotonic discharges. Complex repetitive discharges typically are perfectly stable, but if additional loops or circuits drop in or out, the morphology may change abruptly in distinct or quantal jumps.

Firing Characteristics

After assessing the potential's morphology and stability, the electromyographer should look at the potential's firing characteristics, including the discharge pattern and firing rate. Note whether the pattern is regular or irregular. If it is regular, is it perfectly regular? Fibrillation potentials and positive sharp waves are more or less regular, but complex repetitive discharges are perfectly regular. If it is irregular, is it sputtering (e.g., endplate spikes), waxing/waning (e.g., myotonic discharges), or waning (e.g., neuromyotonic discharges)? Is there a bursting pattern (relative electrical silence between groups of discharges)? Such a pattern is characteristic of doublets and triplets as seen in tetany, and myokymic discharges. Note if the firing rate is very slow (<4–5 Hz). This is important because a slow firing rate signifies that the discharges cannot be voluntary. Voluntary activation of a motor unit has a firing frequency of at least 4 to 5 Hz. Any potential that fires more slowly than 4 to 5 Hz cannot be under voluntary control. Conversely,

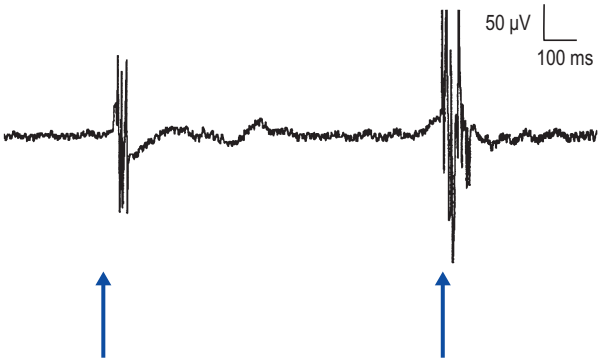

FIGURE 14–5 Normal insertional activity. Arrows show needle movement. With each needle movement, normal insertional activity is brief and usually lasts 300 ms or less. Increased insertional activity can be seen in both neuropathic and myopathic disorders. Note the long sweep speed.

extremely high firing rates are characteristic of neuromyotonic discharges, which can fire as fast as 150 to 250 Hz.

Table 14–1 summarizes the morphology, stability, and firing characteristics of the common spontaneous potentials seen during the needle EMG.

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 brief burst for several hundred milliseconds, known as insertional activity, which is a normal finding (Figure 14–5). The presence of insertional activity is important to the electromyographer to confirm that the needle is in muscle rather than fat or subcutaneous tissue. 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 waveform other than endplate potentials (see following section) 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.

SPONTANEOUS ACTIVITY: NORMAL

All spontaneous activity is abnormal, with the important exception of potentials that occur in the muscle endplate zone (i.e., the NMJ). Muscle endplate usually is found near the center of the muscle belly and often is encountered during routine EMG. Patients frequently perceive a deep, burning, unpleasant sensation when the needle is placed in the endplate zone. Two types of spontaneous activity occur here: endplate noise and endplate spikes. It is of utmost importance to properly identify these potentials so as not to mistake them for pathologic spontaneous activity.

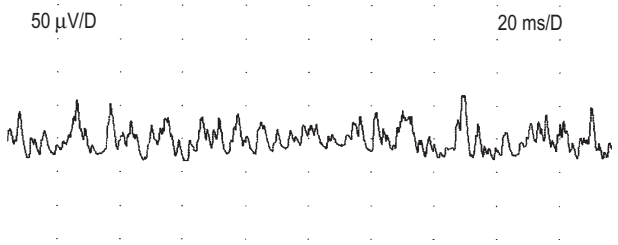

FIGURE 14–6 Endplate noise. Small, high-frequency, predominantly monophasic negative potentials, which are

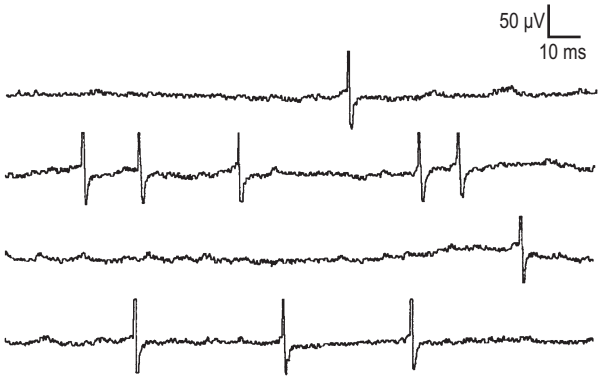

FIGURE 14–7 Endplate spikes. These result from needle-induced irritation of the terminal nerve twigs near the endplate zone. Note the initial negative deflection, brief duration, biphasic morphology, and the irregular, sputtering firing pattern, which differentiates them from fibrillation potentials.

Endplate Noise

These are low-amplitude, monophasic negative potentials that fire irregularly at 20 to 40 Hz and have a characteristic "seashell" sound on EMG (Figure 14–6). Physiologically, they represent MEPPs. They are recognized by their characteristic shape and sound and by their frequent association with endplate spikes (described in the next subsection).

Endplate Spikes ("Nerve Potentials")

Endplate spikes are MFAPs that fire irregularly up to a frequency of 50 Hz (Figure 14–7) and usually are seen along with endplate noise. They are biphasic, with an initial negative deflection, reflecting that the needle is at the site where the action potential is generated. 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.

Endplate spikes are thought to occur as a result of needle-induced irritation of a terminal nerve twig and the subsequent activation of a nerve action potential leading to an MFAP (Figure 14–8). Thus, the needle is necessary to create these potentials. This is in contrast to endplate noise (MEPPs), which occurs spontaneously, without the presence of an irritating source. In summary, endplate spikes

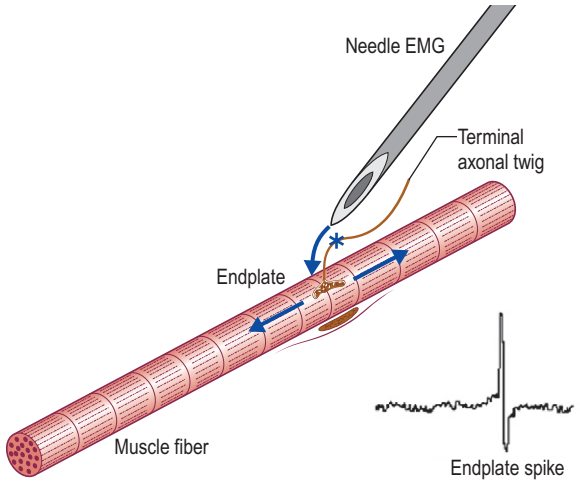

FIGURE 14–8 Generation of an endplate spike. Endplate spikes are thought to occur as a result of the EMG needle irritating a terminal nerve twig. This results in an action potential that runs down the terminal twig which subsequently results in a muscle fiber action potential (MFAP). The resulting waveform is biphasic and initially negative, signifying that the needle is right on top of where the MFAP originates.

only occur when a needle is in the muscle and close to the endplate zone, close enough to mechanically irritate nearby terminal nerve twigs.

SPONTANEOUS ACTIVITY: ABNORMAL MUSCLE FIBER POTENTIALS

Muscle is normally electrically silent outside of the endplate zone. Any persistent spontaneous activity outside of the endplate zone, usually defined as lasting longer than 3 seconds, is abnormal. Spontaneous activity may be ongoing when the needle is placed in the muscle or may be triggered by needle movement, voluntary muscle contraction, muscle percussion, or electrical stimulation.

Fibrillation Potentials

A fibrillation potential is derived from the extracellular recording of a single muscle fiber (Figures 14–9 and 14–10). These spontaneous depolarizations of muscle fibers are the electrophysiologic markers of active denervation. Although they typically are associated with neuropathic disorders (i.e., neuropathies, radiculopathies, motor neuron disease), they also may be seen in some muscle disorders (especially the inflammatory myopathies and dystrophies) and rarely in severe diseases of the NMJ (especially botulism). As they are generated in muscle fiber, fibrillation potentials are recognized by their single MFAP morphology: a brief spike with an initial positive deflection, 1 to 5 ms in duration, and low in amplitude (typically 10–100 μV). The firing pattern is very regular, with a rate usually 0.5 to 10 Hz, occasionally up to 30 Hz. In very chronic conditions (lasting >6–12 months), fibrillation potentials may become very tiny (<10 μV in amplitude) (Figure 14–11). On EMG, single fibrillation potentials sound like "rain on the roof." Although fibrillation potentials fire at a regular rate, they may slow down very gradually over several seconds before stopping.

FIGURE 14–9 Fibrillation potential. Spontaneous depolarization of a single muscle fiber. Note the initial positive deflection, brief duration, and triphasic morphology.

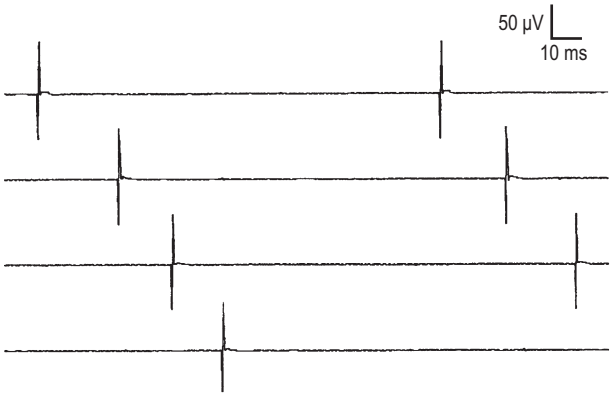

FIGURE 14–10 Fibrillation potential (rastered traces). Note the regular firing pattern, which helps identify the waveform as a fibrillation potential.

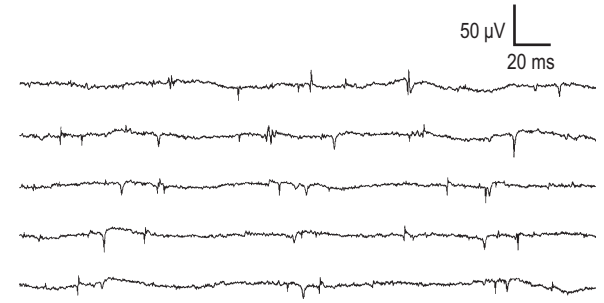

FIGURE 14–11 "Tiny" fibrillation potentials. In very chronic conditions (usually >6–12 months' duration), fibrillation potentials may become very small (<10 μV in amplitude). This trace is from a patient with a lumbar radiculopathy that had persisted for 2 years. Note several very small fibrillation potentials and positive sharp waves.

Positive Sharp Waves

Positive sharp waves have the same significance as fibrillation potentials: they are the spontaneous depolarization of a muscle fiber (Figure 14–12) and signify active denervation. Positive sharp waves have a brief initial positivity followed by a long negative phase. They sound like a dull pop because of their slow negative phase and long duration. The amplitude is variable (usually 10–100 μV, occasionally up to 3 mV). Like fibrillation potentials, they have a regular firing pattern, with a rate usually between 0.5 and 10 Hz, occasionally up to 30 Hz. This is a key point because voluntary motor unit potentials at a distance occasionally will have a positive wave morphology but can be distinguished by their lack of a regular firing pattern. Positive sharp waves usually are accompanied by fibrillation potentials, but they may be seen alone, sometimes early in denervation.

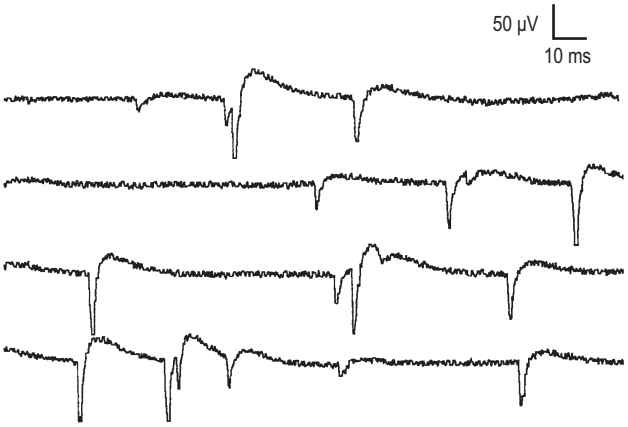

FIGURE 14–12 Positive sharp waves (rastered traces). Positive sharp waves have the same significance as fibrillation potentials: they represent the spontaneous depolarization of a muscle fiber. Note the initial positive deflection and the slow negative phase.

The mechanism by which a single muscle fiber action potential can assume either a fibrillation potential (i.e., brief spike) or a positive sharp wave morphology is not completely agreed upon. Fibrillation potentials occasionally will change to positive sharp waves with EMG needle movement, and vice versa (Figure 14–13). In order for a positive sharp wave to be formed, it is thought that the needle mechanically deforms a muscle fiber, rendering that segment of the membrane inexcitable. When a spontaneous depolarization arises distally in the muscle fiber, it can propagate toward the needle (creating the initial positive phase), but as it approaches the needle and the waveform begins to turn negative as expected, it cannot travel beyond the point of the mechanical deformation. The action potential then dissipates (Figure 14–13, right). This had been thought to be the most likely explanation for the generation of a positive sharp wave. This might also account for the fact that positive sharp waves are occasionally seen earlier than fibrillation potentials: the presence of the needle is required to help generate these positive sharp waves.

However, more recently, an additional explanation for positive sharp wave generation was proposed by Dumitru and colleagues. They postulated that positive sharp waves could originate, rather than terminate, at the recording needle electrode. In an eloquent series of experiments, two separate needle recording electrodes were placed along a single muscle fiber. By moving one needle and deforming the muscle membrane, spontaneous potentials could be generated. These were recorded by the needle near the origin of the spontaneous discharge, and then by the second

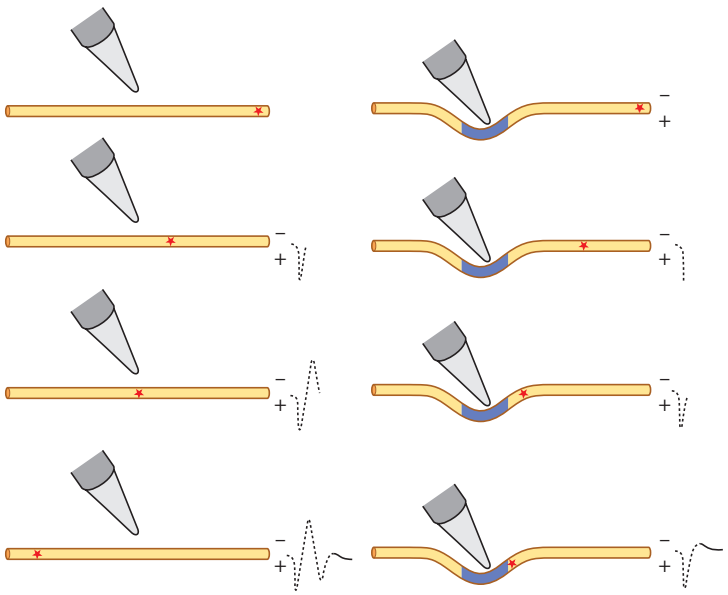

FIGURE 14–13 Generation of a fibrillation potential versus a positive sharp wave. **Left:** When the EMG needle is near a denervated fiber, the spontaneous firing of that fiber typically results in a triphasic brief spike potential (positive, negative, then positive) as the depolarization approaches, then is directly under, and then travels away from, the needle tip, respectively. **Right:** In the case of a positive sharp wave, the needle electrode mechanically deforms the fiber, which then makes the membrane electrically inexcitable at the segment (blue area). As the traveling depolarization wave approaches the needle, an initial positive wave is generated. With failure of conduction beyond the needle, the steep negative phase is aborted, and the waveform returns to baseline.
(Adapted with permission from Dimitru, D., 1989. Volume conduction: theory and application. In: Dimitru, D., (Ed.), Clinical electrophysiology: physical medicine and rehabilitation state of the art reviews. Hanley Belfus, Philadelphia.)

needle as the discharge propagated down the muscle fiber. When normal muscle fibers were studied, they demonstrated that normal insertional activity could result in local potentials that had either a positive sharp wave or brief biphasic spike (negative–positive) morphology. As these normal insertional activity potentials propagated down the muscle fiber and were recorded by the second needle, the potentials all had the morphology of a brief, triphasic spike (positive–negative–positive), the same as a fibrillation potential. However, when a denervated muscle fiber was studied, they demonstrated that when the needle deformed the muscle fiber membrane, a positive wave was recorded by that needle, but a time-locked fibrillation potential was recorded by the second needle at a distance down the same muscle fiber (Figure 14–14). Furthermore, if the first needle did not deform the muscle membrane, both needles recorded a fibrillation potential. Thus, the difference between whether positive sharp waves or fibrillation potentials were generated appeared to depend on whether or not the needle deformed the muscle fiber. The deformation of the muscle fiber by the needle is thought to create a "crushed zone," an area of membrane that cannot propagate action potentials but which remains connected to normal membrane on both sides. Near the crush zone, the extracellular recording of a spontaneous muscle fiber depolarization appears the same as its intracellular action potential (i.e., a positive depolarization).

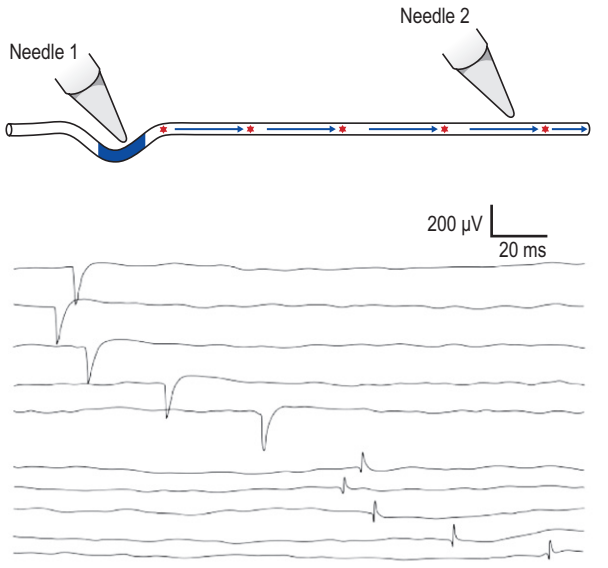

FIGURE 14–14 Additional explanation of positive sharp wave generation. **Top:** Two needle electrodes were placed along the same denervated muscle fiber. Needle 1 deformed the muscle fiber membrane and created a "crushed zone" effect. **Bottom, first 5 traces:** The spontaneous discharges of the muscle fiber recorded by Needle 1 all had a positive wave morphology. **Bottom, last 5 traces:** The corresponding time-locked potentials generated near Needle 1 had the morphology of fibrillation potentials when recorded at Needle 2.
(Adapted with permission from Dumitru, D., Martinez, C.T.J., 2006. Propagated insertional activity: a model of positive sharp wave generation. Muscle Nerve 34, 457–462.)

Regardless of whether the positive wave terminates or originates at the needle electrode, the take home message is that denervation results in both fibrillation potentials and positive sharp waves. They both represent the spontaneous firing of a single muscle fiber with an unstable resting membrane. The only difference between positive sharp waves and fibrillation potentials is that in the case of positive sharp waves, there is needle-induced deformation of the muscle membrane.

When positive sharp waves and/or fibrillation potentials are present in a muscle, they are conventionally graded on a scale from 0 to 4 as follows:

0 None present
+1 Persistent single trains of potentials (>2–3 seconds) in at least two areas
+2 Moderate number of potentials in three or more areas
+3 Many potentials in all areas
+4 Full interference pattern of potentials

Fibrillation potentials and positive sharp waves are the most common of all the abnormal spontaneous potentials, seen in a large number of common disorders (e.g., radiculopathy, entrapment neuropathies). With experience, their recognition becomes fairly straightforward, especially when one hears the characteristic "rain on the roof" sound. The exception is when one encounters the +4 grade of fibrillation potentials (Figure 14–15). In this situation, the screen is completely filled, and individual potentials cannot be seen. It is common to think at first that the patient is not relaxed and that the screen is filled with voluntary motor unit potentials. However, once the electromyographer is convinced that the patient is relaxed, one can still discern the sound pattern of "rain on the roof" when listening closely, but in this case it is a heavy downpour. This pattern is distinctly uncommon and is seen only in unusual situations where all or nearly all muscle fibers are denervated simultaneously. Most common among these situations are trauma (e.g., from nerve laceration) and infarction (e.g., from vasculitis).

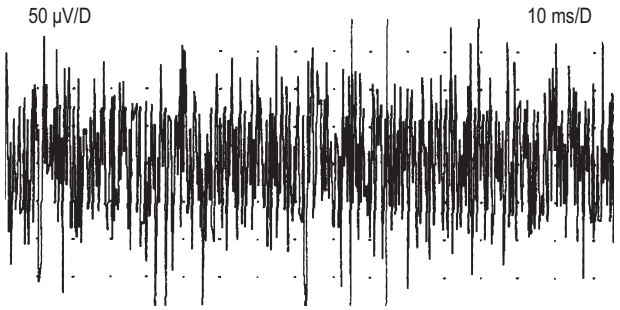

FIGURE 14–15 Grade +4 fibrillation potentials. The number of fibrillation potentials is so profuse that the screen is filled with a complete interference pattern. This pattern is distinctly uncommon and is seen only in unusual situations whereby all or nearly all muscle fibers are denervated simultaneously. Most common among these situations are trauma (e.g., from nerve laceration) and infarction (e.g., from vasculitis).

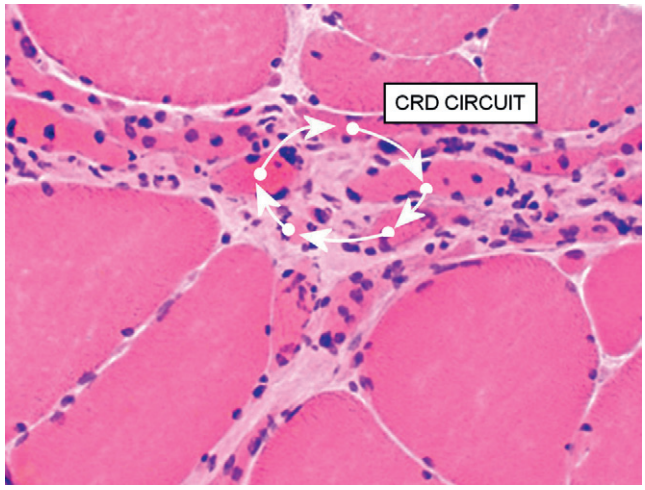

FIGURE 14–16 Pathophysiology of a complex repetitive discharge (CRD). A spontaneous depolarization occurs from ephaptic transmission from one denervated muscle fiber to an adjacent one. If the original pacemaker is reactivated, a circus movement is formed without an intervening synapse. In neuropathic conditions, the pathologic correlate is grouped atrophy, wherein denervated fibers lie next to other denervated fibers.

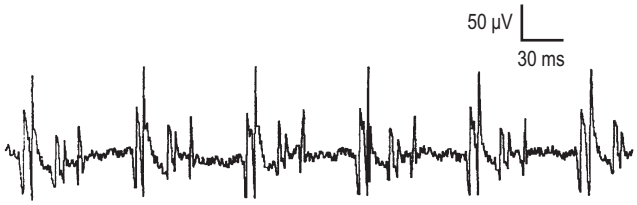

FIGURE 14–17 Typical complex repetitive discharge. Note the multiple spikes (each spike within the complex representing a different single muscle fiber) and the perfectly repetitive nature.

Complex Repetitive Discharges

Complex repetitive discharges (CRDs) are one of the most distinctive waveforms encountered on the needle EMG examination. They result from depolarization of a single muscle fiber followed by ephaptic spread to adjacent denervated fibers (i.e., direct spread from muscle membrane to muscle membrane). If the depolarization spreads in a circus movement whereby the original pacemaker muscle fiber is reactivated, a recurrent discharge develops (Figure 14–16). The morphology of a CRD is that of individual muscle fibers, discernible as individual spikes, that fire consecutively and are time linked together (Figure 14–17). On EMG, CRDs are recognized as high-frequency (typically 5–100 Hz), multi-serrated repetitive discharges with an abrupt onset and termination. These discharges usually occur spontaneously (e.g., when the pacemaker is a fibrillation potential) or following needle movement. Less frequently, they are triggered by a stimulated MUAP or by a voluntary MUAP.

CRDs are identical in morphology from one discharge to the next, creating a characteristic machine-like sound on EMG (Figure 14–18). They occur in both chronic neuropathic and myopathic disorders; they may arise in any

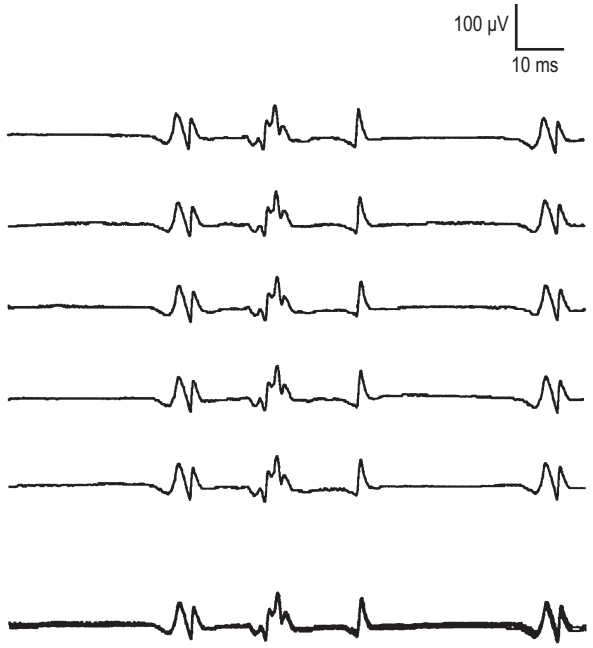

FIGURE 14–18 Complex repetitive discharge (CRD) triggered on a delay line (top five traces). Traces superimposed (bottom trace). Note the perfectly repetitive nature of a CRD. When superimposed, there is little or no jitter between successive potentials.

setting in which denervated muscle fibers lie adjacent to other denervated muscle fibers. Because muscle fibers from many different motor units intermix in normal muscle, CRDs do not usually occur in the acute setting because denervated muscle fibers do not lie adjacent to other denervated fibers in this setting. To create a setting whereby denervated muscle fibers lie adjacent to one another, in neuropathic conditions, there has to be denervation followed by reinnervation (i.e., fiber type grouping) and subsequent denervation (i.e., grouped atrophy). This situation may also occur in myopathic disorders associated with denervation (i.e., myopathies associated with necrosis or inflammation) or those associated with muscle fiber splitting.

Occasionally, individual phases or additional loops drop in and out, creating an abrupt change in frequency and sound (Figure 14–19). 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 once again becomes perfectly regular.

Because CRDs are generated by muscle fibers, they usually persist with NMJ blockade. On single-fiber EMG, there is a characteristic finding: abnormally low jitter. This occurs because the discharge spreads ephaptically from one muscle fiber to another; there is no intervening synapse, which would normally give rise to some jitter.

Myotonic Discharges

A myotonic discharge is the spontaneous discharge of a muscle fiber (similar to fibrillation potentials and positive

sharp waves) but is differentiated by its characteristic waxing and waning of both amplitude and frequency (Figures 14–20 and 14–21). The firing rate is generally between 20 and 150 Hz. An individual myotonic potential may have either a positive wave or a 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 other myopathies (acid maltase deficiency, polymyositis, myotubular myopathy), hyperkalemic periodic paralysis, and, rarely, in denervation of any cause. It is important to remember this last point: a single brief run of myotonic discharges may occur in any denervating disorder, although it is never the predominant waveform.

Myotonic discharges have a characteristic "revving engine" sound on EMG, due to the waxing and waning of amplitude and frequency. One of the most common pitfalls in the interpretation of needle EMG is to mistake myotonic discharges for acute denervation (i.e., fibrillation potentials and positive sharp waves). This error of interpretation occurs because both have the same basic morphology and both are generated in muscle fiber, and denervation potentials are common whereas myotonic discharges are uncommon in clinical practice. However, once the waxing and waning sound of myotonic discharges is recognized, the differentiation is easily made. More than one patient with myotonia congenita or myotonic dystrophy has been erroneously given the diagnosis of motor neuron disease based on the electromyographer's misinterpretation of myotonic discharges as widespread denervation potentials.

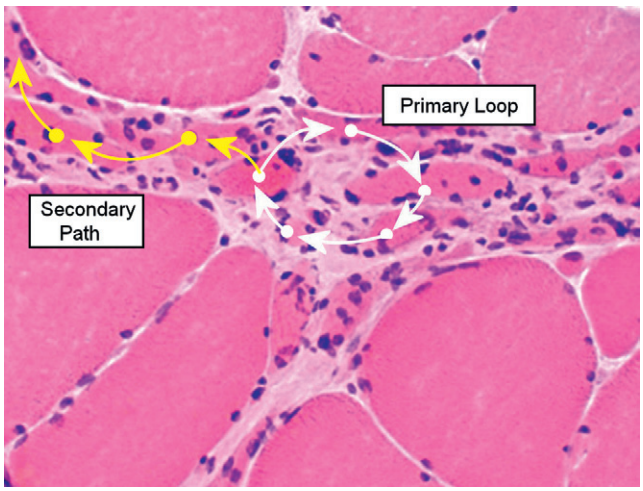

FIGURE 14–19 Complex repetitive discharges. These may change abruptly in frequency or number of potentials when extra paths or circuits drop in and out (note the secondary path compared with Figure 14–16).

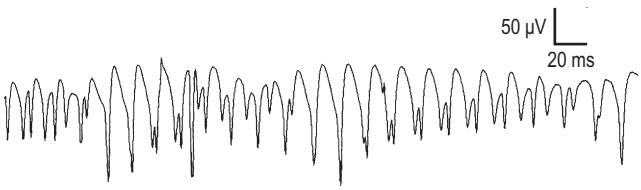

FIGURE 14–20 Myotonic discharge (spontaneous discharge). Note the waxing and waning of both amplitude and frequency.

SPONTANEOUS ACTIVITY: ABNORMAL MOTOR UNIT POTENTIALS

Fasciculation Potentials

A fasciculation potential is a single, spontaneous, involuntary discharge of an individual motor unit (Figure 14–22). Unlike voluntary motor unit potentials, fasciculation potentials generally fire very slowly and irregularly, usually less than 1 to 2 Hz. In contrast, voluntary motor unit potentials begin firing at 4 to 5 Hz when a patient is asked to slightly contract a muscle and cannot fire more slowly than this. Thus, potentials that fire more slowly than 4 to 5 Hz are not under voluntary control. The source generator of fasciculation potentials is the motor neuron or its axon, prior to its terminal branches. On EMG, fasciculation potentials usually have the morphology of simple MUAPs, or they can be complex and large if they represent a pathologic (i.e., reinnervated) motor unit. Despite the notorious association of fasciculations with diseases of the anterior horn cell, the actual site of origin of most fasciculations has been found to be distal in the axon.

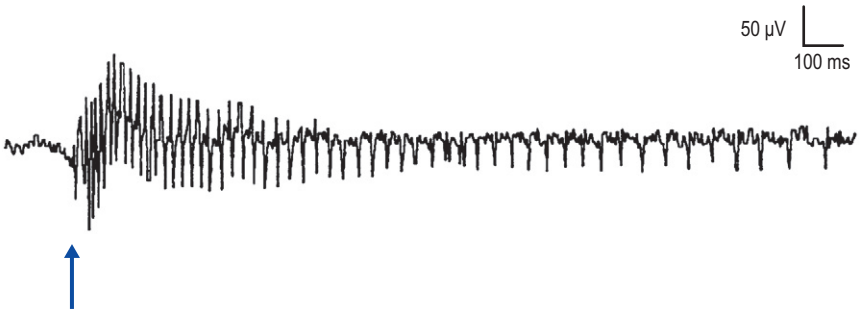

FIGURE 14–21 Myotonic discharge (needle induced). Arrow marks needle movement triggering the discharge. Myotonic discharges may occur spontaneously or be triggered by needle movement, voluntary contraction, or muscle percussion.

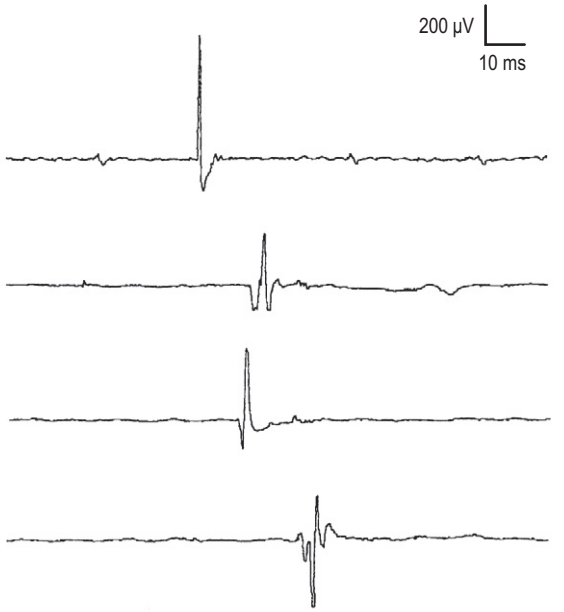

FIGURE 14–22 Fasciculation potentials (rastered traces). Each potential has the morphology of a motor unit potential. They are recognized by their morphology and irregular, slow firing pattern.

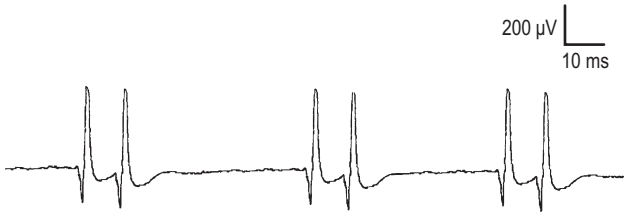

FIGURE 14–23 Doublets. Spontaneous discharges of motor unit action potentials that fire in groups of twos are known as doublets. Doublets often accompany fasciculations as well as groups of three potentials (triplets) or multiple potentials (multiplets). These potentials fundamentally have the same significance as fasciculation potentials and are seen in neuropathic conditions, but they also are characteristically seen in tetany from hypocalcemia.

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. Motor neuron disease, such as amyotrophic lateral sclerosis, is the best known. However, fasciculations can be seen in radiculopathies, polyneuropathies, and entrapment neuropathies. In addition, most normal individuals have some fasciculations, so-called *benign fasciculations*.

Distinguishing "benign" from "malignant" fasciculations on a clinical basis is nearly impossible. However, benign fasciculations are not associated with muscle weakness, wasting, or any abnormality of reflexes. In general, benign fasciculation potentials tend to fire faster and affect the same site repetitively (e.g., eyelid twitching), as opposed to fasciculation potentials in pathologic conditions such as motor neuron disease, which tend to be more random.

On EMG, fasciculation potentials have the sound of "corn popping": they are dull, irregular pops. Because fasciculation potentials usually are very slow, they can be easily missed on needle EMG if the electromyographer does not wait a sufficient amount of time with the needle in the muscle at rest. It is often said that the best way to look for fasciculation potentials is to place the needle in the muscle and then have the electromyographer take his or her hand off the needle and wait.

Doublets, Triplets, and Multiplets

Spontaneous MUAPs that fire in groups of two are known as *doublets* (Figure 14–23). When they fire in groups of three or multiple potentials, they are known as *triplets* and *multiplets*, respectively. These potentials have the same significance as fasciculation potentials: they represent the spontaneous depolarization of a motor unit or its axon. They are often seen with fasciculation potentials. In this situation, fasciculation potentials may be referred to as singlets. Doublets, triplets, and multiplets can be seen in any situation where fasciculation potentials occur (i.e., neuropathic conditions), but they also are characteristically seen in tetany from hypocalcemia (Figure 14–24). Tetany, which is the involuntary contraction of muscles associated with hypocalcemia, predominantly affects the distal muscles, with involuntary spasms affecting the hands and feet (carpopedal spasms). In the hands, a characteristic posture develops: adduction of the thumb and fingers, extension of the interphalangeal joints, and flexion of the metacarpal–phalangeal joints and wrist.

Myokymic Discharges

Electrically, myokymic discharges (Figure 14–25) are rhythmic, grouped, spontaneous repetitive discharges of the same motor unit (i.e., grouped fasciculations). The firing frequency within the burst typically is 5 to 60 Hz. The number of potentials within a burst varies widely and may change from burst to burst (Figure 14–26). The firing frequency between bursts is much slower (typically <2 Hz) and produces a marching sound on EMG. Changing to a longer sweep speed often makes it easier to recognize the bursting pattern of a myokymic discharge. Freezing the screen often makes it easier to recognize the presence of the same motor unit potential firing repetitively in bursts. The origin of myokymic discharges likely involves spontaneous depolarization of or ephaptic transmission along demyelinated segments of nerve.

Clinically, myokymia usually is recognized as continuous involuntary quivering, rippling, or undulating movement of muscle. The finding of myokymic discharges on EMG is very helpful in limiting the differential diagnosis (Box 14–1). Limb myokymia occurs in a variety of conditions but is most characteristically seen in radiation-induced nerve damage. The most typical situation occurs in a patient with a progressive plexopathy who has a prior history of cancer treated with radiation therapy. In this situation, the

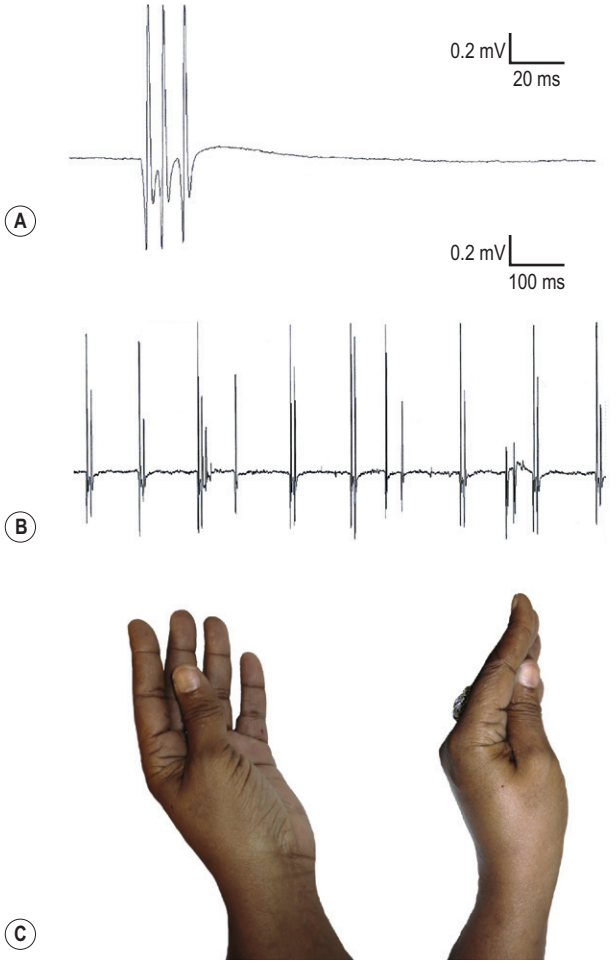

FIGURE 14–24 Tetany and carpopedal spasm. EMG traces and photo from a patient who developed hypoparathyroidism resulting in hypocalcemia following total thyroidectomy. This patient reported intermittent paresthesias in the fingers and toes, as well as around the mouth, with involuntary spasms of the hands. Routine nerve conduction and needle EMG were normal. When a blood pressure cuff was inflated above systolic blood pressure with the EMG needle in a distal hand muscle, paresthesias developed within a minute. Two minutes later, occasional doublets and triplets were present on EMG. **A:** Isolated triplet. This was followed by the firing of many singlets, doublets and triplets. **B:** The sweep speed has been increased to 100 ms/division. Note the irregular firing of doublets, triplets, and an occasional singlet. The hand then went into an involuntary spasm. **C:** Note the characteristic posture of carpopedal spasm associated with tetany when a blood pressure cuff was inflated above systolic pressure (Trousseau sign): adduction of the thumb and fingers, extension of the interphalangeal joints, and flexion of the metacarpal–phalangeal joints). When the blood pressure cuff was deflated, all the spontaneous EMG activity ceased within seconds. The Trousseau sign is provoked by making the limb ischemic with a blood pressure cuff and is useful both clinically and during EMG to demonstrate the potentials associated with tetany.

differential diagnosis often lies between relapse of the cancer with invasion of the plexus and a delayed radiation-induced plexopathy. The presence of myokymic discharges on needle EMG strongly supports a diagnosis of radiation plexitis rather than recurrent neoplastic invasion. Limb myokymia also occurs infrequently in radiculopathy, entrapment neuropathy, and spinal cord lesions associated with demyelination.

Box 14–1. Disorders Commonly Associated with Myokymic Discharges

Radiation injury (usually brachial plexopathy)
Guillain–Barré syndrome (facial)
Multiple sclerosis (facial)
Pontine tumors (facial)
Hypocalcemia
Timber rattlesnake envenomization

Occasionally seen in
- Guillain–Barré syndrome (limbs)
- Chronic inflammatory demyelinating polyneuropathy
- Nerve entrapments
- Radiculopathy

Facial myokymia characteristically occurs with brainstem lesions associated with multiple sclerosis, pontine gliomas, and vascular disease, but it can also be seen after radiation. In Guillain–Barré syndrome, facial myokymia may occur in 15% of patients, usually occurring early in the illness and remitting as the patient clinically improves.

Myokymia from peripheral nerve lesions can be provoked or enhanced by lowering serum ionized calcium with hyperventilation or with the use of acid–citrate–dextrose anticoagulant as is commonly given during plasma exchange. Administration of calcium can transiently decrease the generation of myokymic discharges.

Cramp Potentials

Cramps are painful, involuntary contractions of muscle that tend to occur when a muscle is in the shortened position and contracting. Surprisingly, cramp potentials are actually high-frequency discharges of motor axons and are not primarily a muscle phenomenon. EMG characteristically shows several normal appearing motor unit potentials firing repetitively and sometimes irregularly at high frequencies (usually 40–75 Hz) (Figure 14–27). Cramps may be benign (e.g., nocturnal calf cramps, post-exercise cramps) or may be associated with a wide range of neuropathic, endocrinologic, and metabolic conditions. Clinically, cramps may resemble the contractures that occur in several of the metabolic muscle diseases. However, the needle EMG of a cramp potential is quite different from that of a contracture, which typically is completely electrically silent (see Chapter 35).

Neuromyotonic Discharges

Neuromyotonic discharges are high-frequency (150–250 Hz), decrementing, repetitive discharges of a single motor unit that have a characteristic "pinging" sound on EMG (Figure 14–28). They have the highest frequency of any discharge. They are a rare phenomenon and represent the end of the spectrum of abnormal spontaneous activity generated in motor nerve (Figure 14–29).

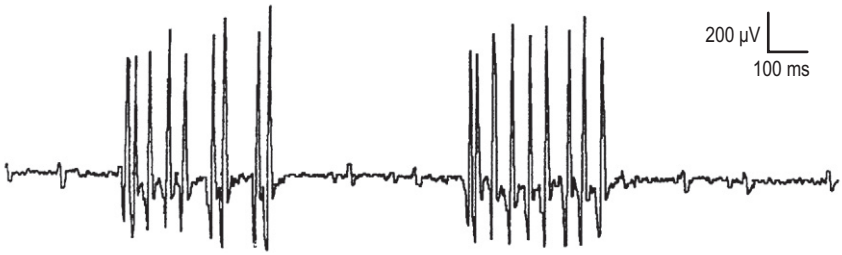

FIGURE 14–25 Myokymic discharges. Involuntary grouped repetitive discharges of the same motor unit action potential. Note the high-frequency pattern within the burst and the slow frequency between the bursts. Myokymia produces a marching sound on electromyogram.

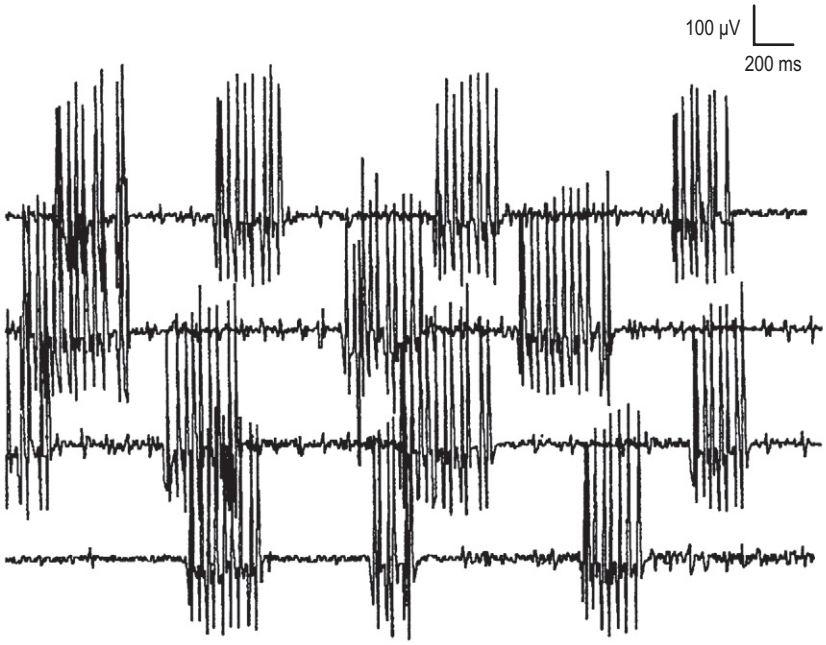

FIGURE 14–26 Myokymic discharges (rastered traces, long sweep speed). Note that the number of potentials within a burst may change from burst to burst.

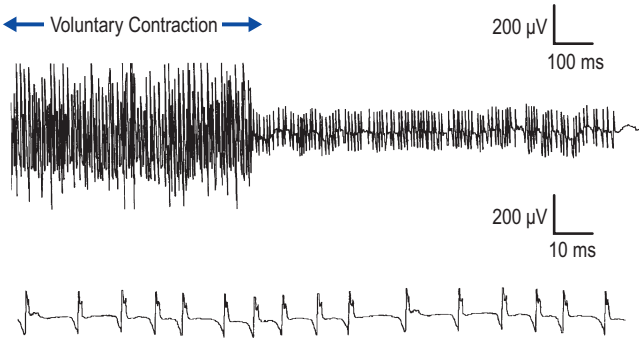

FIGURE 14–27 Cramp discharge. In the figure, the subject is actively contracting the muscle, followed by relaxation. The cramp discharge is seen in the relaxation phase **(upper trace)** following voluntary contraction. In the **lower trace,** the cramp discharge is expanded. Note that the cramp discharge is composed of the same MUAP firing quickly but slightly irregularly. Clinically, cramps are painful, involuntary contractions of muscle that tend to occur when a muscle is in the shortened position and contracting. Cramps are high-frequency discharges of motor axons, with the electromyogram characteristically showing motor unit action potentials (MUAPs) with a normal morphology firing repetitively and sometimes irregularly at high frequencies.

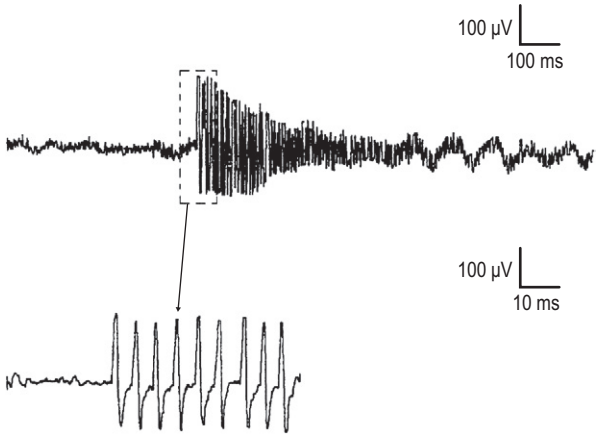

FIGURE 14–28 Neuromyotonic discharges. Spontaneous discharges of a single motor unit potential at very high frequencies (150–250 Hz). Note the decrementing response. **Inset:** Change in sweep speed identifies each potential as the same motor unit potential.

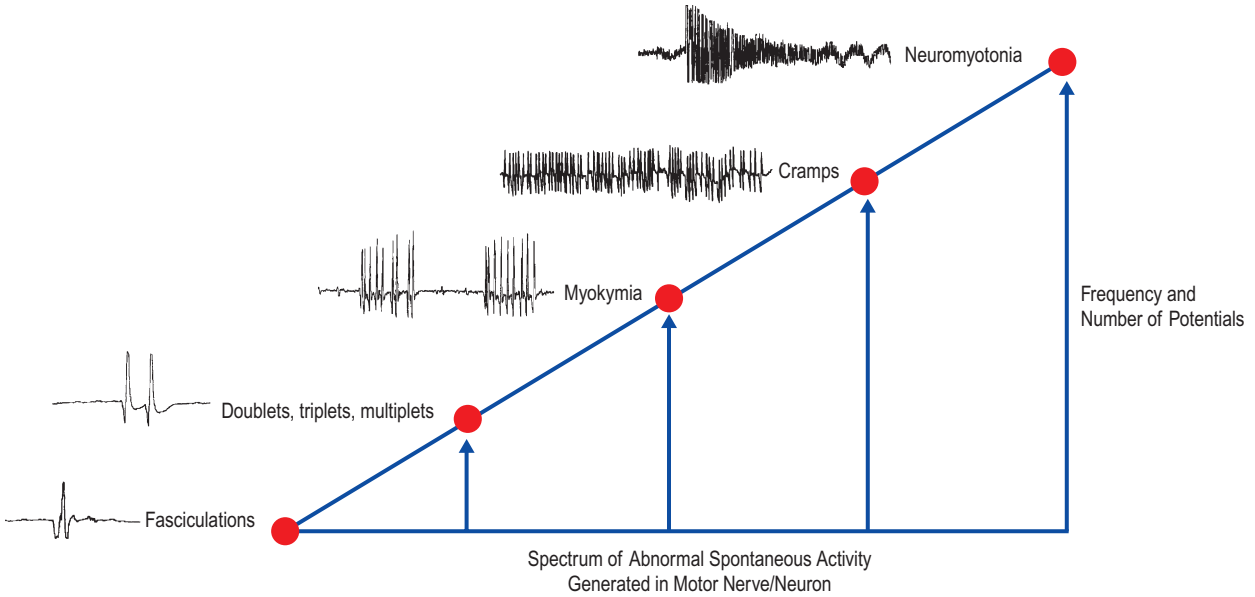

FIGURE 14–29 Spectrum of abnormal spontaneous activity generated in the motor nerve/neuron. Conceptually, it is useful to consider spontaneous activity that arises from the motor nerve/neuron to be on a spectrum. They all share the same basic morphology: that of a motor unit action potential. They are differentiated by their stability and firing characteristics. Often, these potentials accompany one another. For instance, cramp discharges and fasciculation potentials are commonly seen together.

Clinically, patients with neuromyotonia display generalized stiffness, hyperhidrosis, and delayed muscle relaxation after contraction. The delay in relaxation and improvement with repetitive use can be difficult to distinguish clinically from myotonia of muscle origin. However, in myotonia of muscle origin, direct muscle percussion may elicit myotonia, whereas this does not occur in neuromyotonia. Electrically, these syndromes are easily differentiated. Whereas myotonic syndromes are associated with spontaneous discharges of muscle fibers (with a positive wave or brief spike morphology), neuromyotonic disorders are associated with involuntary spontaneous discharges of motor neurons or their axons (with an MUAP morphology). In the neuromyotonic disorders, it is not unusual to see other spontaneous discharges that originate in the motor nerve, including fasciculation potentials and myokymic discharges.

Several lines of evidence suggest that these discharges are generated by peripheral motor axons. The activity persists during sleep, as well as during spinal or general anesthesia, and is abolished by curare. Progressively distal nerve blocks diminish the intensity of the spontaneous discharges. Phenytoin and carbamazepine frequently are helpful in reducing symptoms.

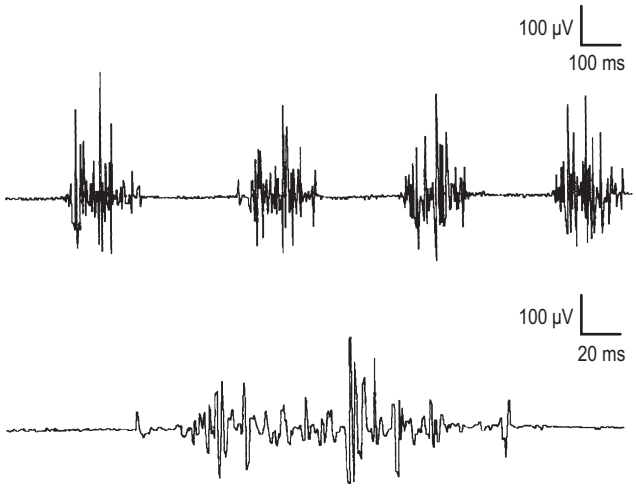

FIGURE 14–30 Rest tremor is recognized as a bursting pattern of motor unit action potentials (MUAPs) separated by relative silence **(top).** Because multiple different MUAPs overlap, polyphasia appears increased and individual MUAP morphology is difficult to assess **(bottom).**

The nomenclature of the neuromyotonic syndromes is complicated; they have been referred to as Isaac's syndrome, neuromyotonia, pseudomyotonia, neurotonia, normocalcemic tetany, and continuous muscle fiber activity. Although the neuromyotonic syndromes are rare, neuromyotonic discharges are seen most commonly in the syndrome of acquired neuromyotonia. There is now considerable evidence that this disorder is an autoimmune channelopathy, with the target antigen being a peripheral nerve voltage-gated potassium channel. An association with myasthenia gravis, thymoma, various malignancies, and inflammatory demyelinating polyneuropathies, among other conditions, has been reported. Some cases have been reported to improve with immunosuppressive therapy. Neuromyotonic discharges also may be seen in extremely chronic neuropathic diseases (especially old poliomyelitis and adult spinal muscular atrophy). Rare cases of familial neuromyotonia have been described, with the age of onset

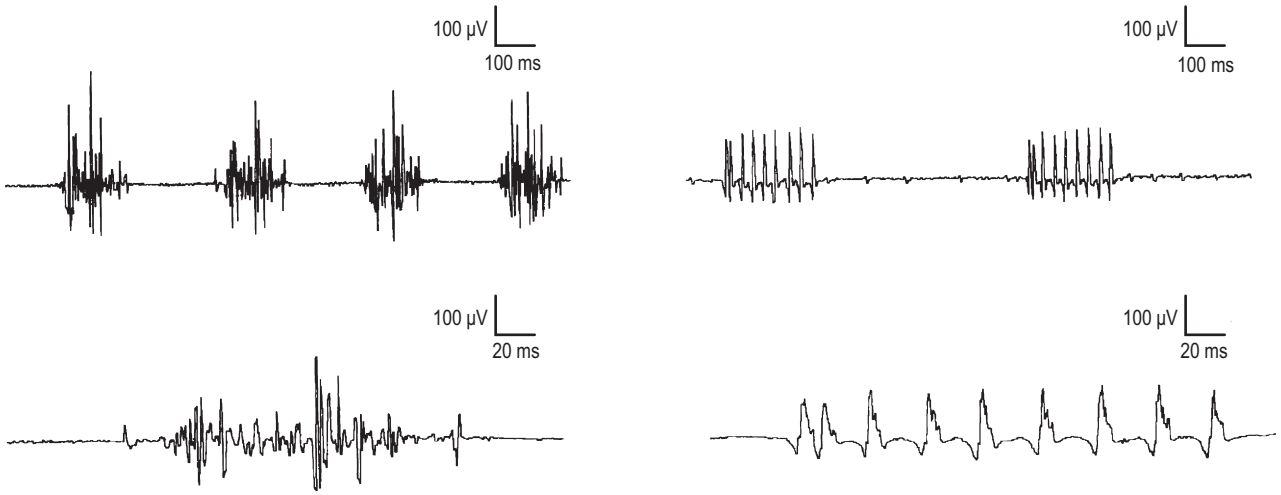

FIGURE 14–31 Tremor versus myokymia. When tremor occurs at rest **(left traces),** it may be mistaken for myokymic discharges. Myokymic discharges **(right traces)** also result in a bursting pattern. Myokymia can be differentiated from tremor by noting that the same motor unit action potential (MUAP) fires repetitively in a myokymic burst, compared with many different MUAPs firing simultaneously in tremor.

ranging from infancy to the eighth decade. It should be noted that neuromyotonic discharges are not seen in stiff-person syndrome, which is a central disorder of spinal interneurons, wherein involuntary firing of normal-appearing MUAPs is seen and for which diazepam is frequently helpful.

Rest Tremor

Although tremor, if present, usually occurs during voluntary contraction, it can complicate the interpretation of spontaneous activity on EMG if it is present at rest. Tremor is recognized as a synchronous bursting pattern of MUAPs separated by relative silence (Figure 14–30). As multiple MUAPs fire simultaneously, the morphology of individual MUAPs may be difficult to assess, and there appears to be increased polyphasia. When tremor occurs at rest (e.g., Parkinson's disease), the spontaneous bursting discharge may be mistaken for myokymic discharges. Although myokymic discharges and tremor both 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 (Figure 14–31). Also, if one freezes the screen and looks closely at the burst, one can see that the amplitude often will rise and fall in tremor, whereas it remains relatively unchanged in myokymia.

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15 Basic Electromyography: Analysis of Motor Unit Action Potentials

After assessment of insertional and spontaneous activity, the needle electromyography (EMG) examination moves on to the evaluation of motor unit action potentials (MUAPs). In a process similar to the analysis of spontaneous activity, MUAPs must be assessed for morphology (duration, amplitude, phases), stability, and firing characteristics. The pattern of MUAP abnormalities that emerges from this part of the examination usually will allow a determination of whether a disorder is primarily neuropathic or myopathic and often helps determine the time course (acute vs. chronic) and severity of the lesion. The assessment of MUAPs often is demanding and improves with the experience of the electromyographer over time. The task of evaluating MUAPs is made all the more difficult by the wide variation in what is considered a normal MUAP, depending on the muscle being studied and the age of the patient.

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 (NMJs) and muscle fibers. The extracellular needle EMG recording of a motor unit is the MUAP (Figure 15–1). The number of muscle fibers per motor unit varies greatly, from 5 to 10 in laryngeal muscles to a couple of thousand in the soleus. The transverse 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 because of the increase in individual muscle fiber size.

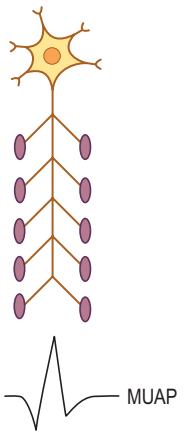

FIGURE 15–1 The motor unit. The basic component of the peripheral nervous system is the motor unit, defined as an individual motor neuron, its axon, and associated neuromuscular junctions and muscles fibers. The extracellular needle electromyography recording of a motor unit is the motor unit action potential (MUAP).

When a motor neuron depolarizes to threshold, a nerve action potential is generated and propagates down the axon. 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 in NMJ transmission times.

The "size principle" governs many of the properties of motor units (Figure 15–2). The size of the motor neuron is directly related to (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 (hence, the fastest conduction velocity), highest threshold to depolarization, and connections to type II, fast twitch muscle fibers. Conversely, the smaller motor neurons have smaller axons, less myelin sheath, slower conduction velocity, 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 thus from the smaller motor units that innervate type I muscle 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. A MUAP recorded just outside a muscle membrane is 1/10 to 1/100 the amplitude of the actual transmembrane potential and the amplitude decreases rapidly as the distance between the needle and the membrane increases. *The classification of an MUAP as normal, neuropathic, or myopathic rests on*

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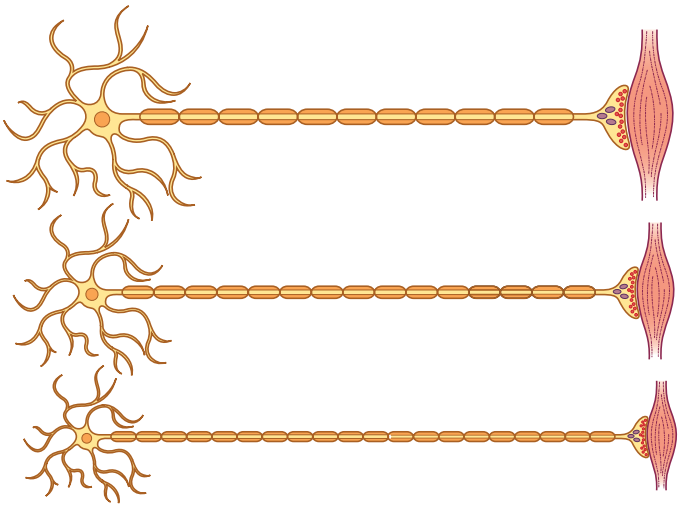


Neuron	Axon	Myelin	CV	Threshold	Fiber Type
Large	Large	Thick	Fast	High	Type II
Medium	Medium	Medium	Medium	Medium	Type I/II
Small	Small	Thin	Slow	Low	Type I

FIGURE 15–2 Size principle and motor unit properties.

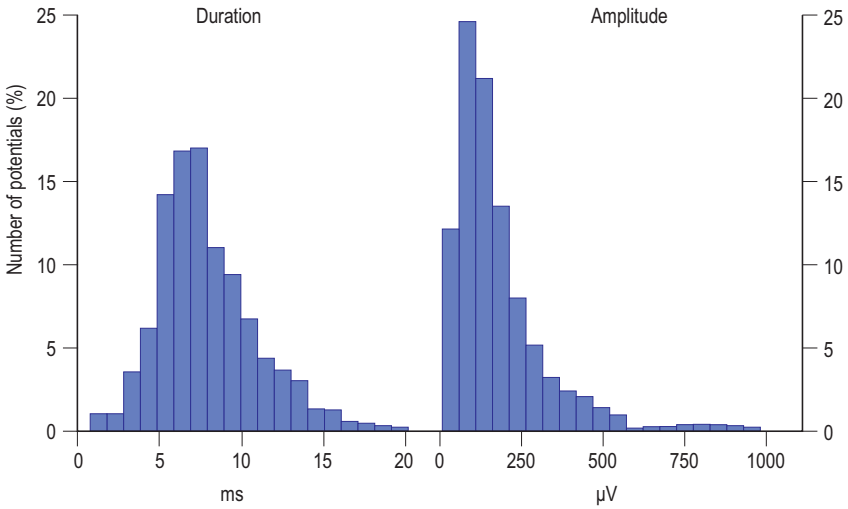


FIGURE 15–3 Range of normal motor unit action potential (MUAP) duration and amplitude. Histogram of MUAP duration and amplitude in the biceps brachii of a normal subject. Note that both MUAP duration and amplitude vary markedly in normal muscles, with small and large units in the same muscle. MUAP duration or amplitude should not be classified as abnormal based on one or two MUAPs but requires a mean of many motor units.
(Reprinted with permission from Buchthal, F., Guld, C., Rosenfalck, P., 1954. Action potential parameters in normal human muscle and their dependence on physical variables. Acta Physiol Scand 32, 200.)

no single finding. As is true of spontaneous activity, recorded MUAPs must be assessed for morphology (duration, polyphasia, amplitude), stability, and firing characteristics before any conclusions can be reached.

MORPHOLOGY

MUAP properties vary widely both within and between different muscles. Even within a muscle, there is a wide range of normal motor unit morphology, with MUAP size following a bell-shaped distribution curve (Figure 15–3). Due to this normal variability, normal values of MUAP morphology are based on the mean of many different MUAPs. The analysis of MUAP morphology can be performed on either a qualitative or a quantitative basis. To perform quantitative MUAP analysis, one must isolate 20 different MUAPs for each muscle being studied and measure their individual durations, amplitudes, and number of phases. From these values, the mean duration, amplitude, and number of phases are calculated and compared with a set of normal values for that particular muscle and age group. MUAP morphology varies depending on the muscle being studied and the patient's age. This is particularly true of MUAP duration (Table 15–1). 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 because of an increase in the size of muscle fibers during development. In addition, MUAP size is generally larger in older individuals, probably as the result of dropout of motor units from the normal effects of aging, leading to some compensatory "normal" reinnervation. The loss of motor units has been estimated to be approximately 1% per year, beginning in the third decade of life, which then increases rapidly after age 60.

Only by comparing mean MUAP morphology in each muscle studied to normal values for that particular muscle and age group can one determine whether the morphology is truly abnormal. Previously, quantitative MUAP analysis was tedious and time consuming. However, many modern

Table 15–1. Mean Motor Unit Action Potential Duration Based on Age and Muscle Group

Age of Subjects	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

ADM, abductor digiti minimi; BF, biceps femoris; EDB, extensor digitorum brevis; Gastroc, gastrocnemius; Per Long, peroneus longus; Quad, quadriceps; Tib Ant, tibialis anterior.

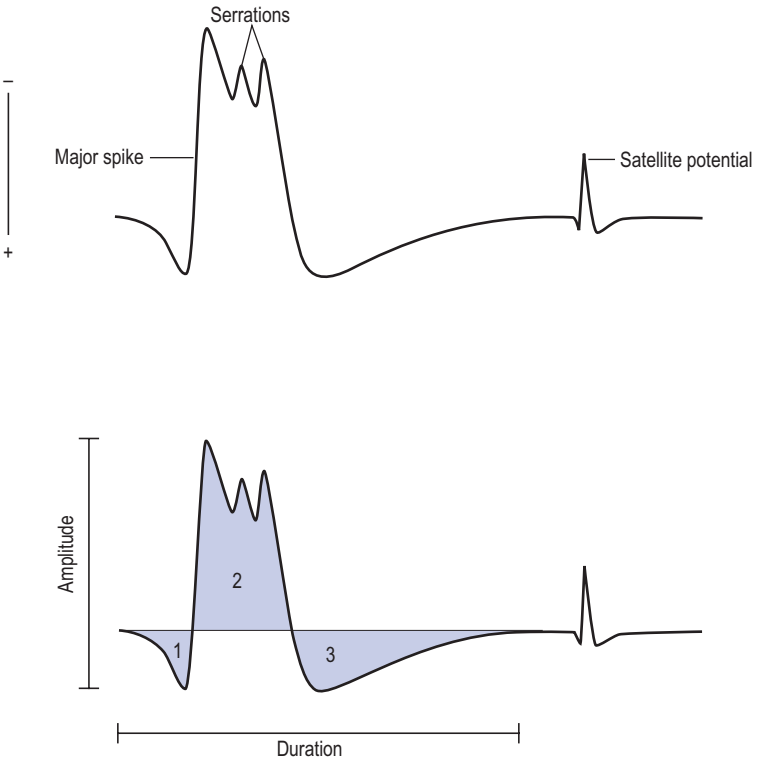


FIGURE 15–4 Motor unit action potential (MUAP) measurements. Duration is measured as the time from the initial deflection of the MUAP from baseline to its final return to baseline. It is the parameter that best reflects the number of muscle fibers in the motor unit. Amplitude reflects only muscle fibers very close to the needle and is measured peak to peak. Phases (shaded areas) can be determined by counting the number of baseline crossings and adding one. MUAPs are generally triphasic. Serrations (also called turns) are changes in direction of the potential that do not cross the baseline. The major spike is the largest positive-to-negative deflection, usually occurring after the first positive peak. Satellite, or linked, potentials occur after the main potential and usually represent early reinnervation of muscle fibers.

EMG machines now have programs that largely automate the procedure. With experience over time, however, the well-trained electromyographer usually can perform qualitative MUAP assessment with the same precision as can be achieved using quantitative methods. Essentially the same procedure is used. The needle is moved to several locations within the muscle until approximately 20 different MUAPs have been examined, qualitatively analyzed, and compared to the expected normal values for that particular muscle and age group.

Duration

MUAP duration is the parameter that best reflects the number of muscle fibers within a motor unit (Figure 15–4). Typical MUAP duration is between 5 and 15 ms. *Duration*

is defined as the time from the initial deflection from baseline to the final return of the MUAP to baseline. It depends primarily on the number of muscle fibers within the motor unit and the dispersion of their depolarizations over time. Dispersion in turn depends on the longitudinal and transverse scatter of endplates and on variations in terminal distances and conduction velocities. Duration lengthens as the number of fibers and the territory of a motor unit increase; it varies directly with age (increased age, increased duration) and inversely with temperature (decreased temperature, increased duration) and depends on the individual muscle being studied. Proximal and bulbofacial muscles in general have MUAPs of shorter duration. When performing EMG, it often is more rewarding to listen to the potential than to see it. This is especially true when evaluating MUAP duration, because *duration correlates with pitch*. Long-duration MUAPs (low frequencies) sound dull and thuddy, whereas short-duration MUAPs (higher frequencies) sound crisp and static-like. As the electromyographer gains experience, the sound of a long-duration versus a short-duration MUAP becomes unmistakable.

Polyphasia, Serrations, and Satellite Potentials

Polyphasia is a measure of synchrony, that is, the extent to which the muscle fibers within a motor unit fire more or less at the same time. This is a nonspecific measure and may be abnormal in both myopathic and neuropathic disorders. The number of phases can be easily calculated by counting the number of baseline crossings of the MUAP and adding one (Figure 15–4). Normally, MUAPs have two to four phases. However, increased polyphasia may be seen in up to 5 to 10% of the MUAPs in any muscle and is considered normal. The one exception is the deltoid, where up to 25% polyphasia may be normal. Increased polyphasia beyond 10% in most muscles and 25% in the deltoid is always abnormal. Through the speaker, polyphasic MUAPs are recognized as a high-frequency "clicking" sound.

Serrations (also called turns) are defined as changes in the direction of the potential that do not 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 (also known as *linked potentials* or *parasite potentials*) are interesting phenomena seen in early reinnervation. After denervation, muscle fibers often are reinnervated by collateral sprouts from adjacent intact motor units. The newly formed sprout often is small, unmyelinated or thinly myelinated, and therefore very 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 (Figures 15–5 and 15–6). These satellite potentials are extremely unstable (see section on Stability) and may vary slightly in their firing rate or may block and not fire at all

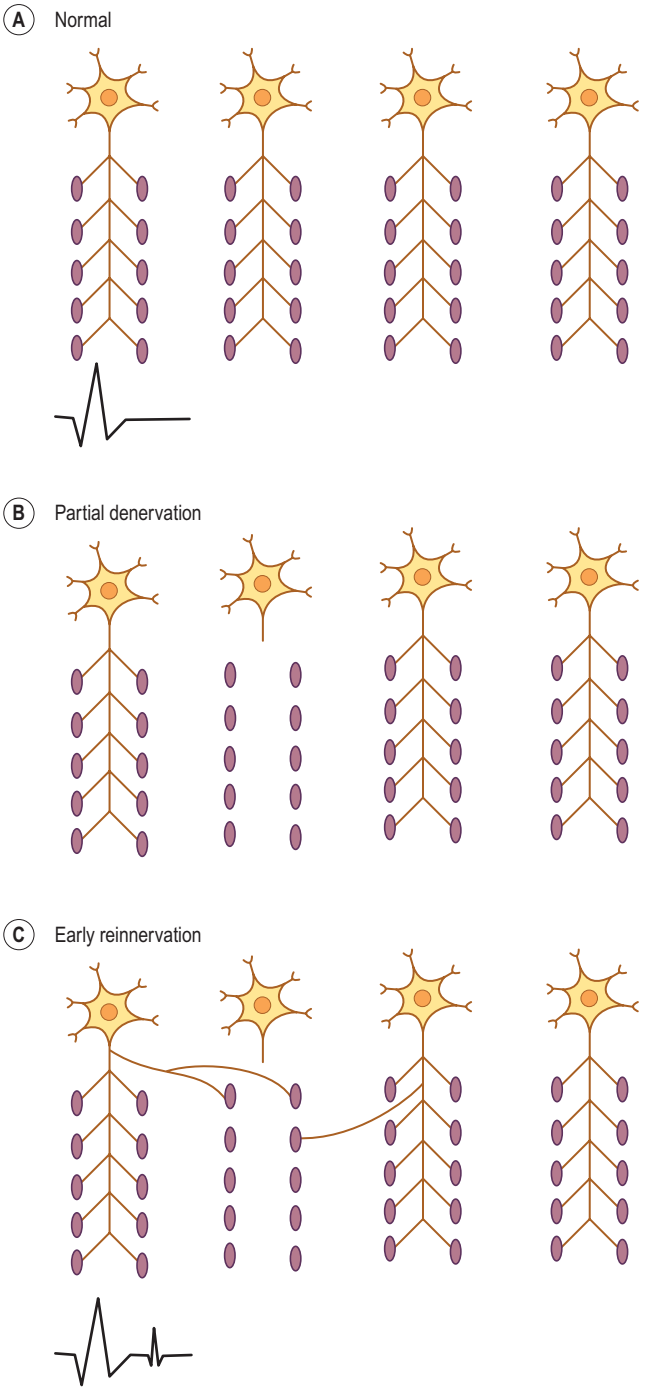


FIGURE 15–5 Collateral sprouting and satellite potentials. **A:** Normal state. **B:** Following partial denervation, the injured axon(s) undergoes wallerian degeneration. **C:** Reinnervation commonly occurs from sprouting by adjacent surviving axons. In early reinnervation, sprouts are small and thinly myelinated and conduct slowly. Because of the slow conduction time and increased distance, these reinnervated fibers initially occur as time-locked potentials (satellite potentials) trailing the main motor unit action potential (MUAP). As sprouts mature and conduct more quickly, the time-locked potentials are eventually incorporated into the main MUAP, resulting in an MUAP with increased amplitude, duration, and number of phases.

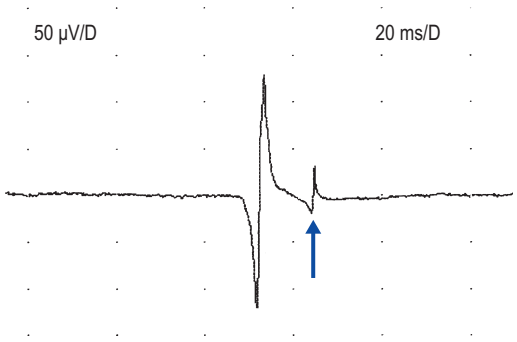


FIGURE 15–6 Satellite potential. Note in the tracing the small potential that is time locked to the main motor unit action potential (MUAP). This is a satellite potential, a sign of early reinnervation. After denervation, muscle fibers often are reinnervated by collateral sprouts from adjacent intact motor units. The newly formed sprout often is small, unmyelinated or thinly myelinated, and therefore very 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.

(Figure 15–7). Over time, the sprout matures, and the thickness of the myelin and consequently the conduction velocity increase. The satellite potential then fires more closely to the main potential and ultimately will become an additional phase or serration within the main complex. It is usually necessary to put the main MUAP on a delay line to appreciate a satellite potential and to demonstrate that it is time locked to the main potential.

Amplitude

MUAP amplitude varies widely among normal subjects. 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 (Figure 15–4). Amplitude is essentially a high-frequency response. Tissue between the needle and muscle fibers effectively acts as a high-frequency filter. Thus, unlike duration, most muscle fibers of a motor unit contribute little to the amplitude. *MUAP amplitude reflects only those few fibers nearest to the needle (only 2–12 fibers)*. Hence, amplitude is not as helpful as duration in judging motor unit size. Several factors are associated with increased amplitude, including (1) the proximity of the needle to the motor unit (Figure 15–8), (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. *Listening to the EMG, the amplitude of MUAPs is correlated not with pitch but with volume.*

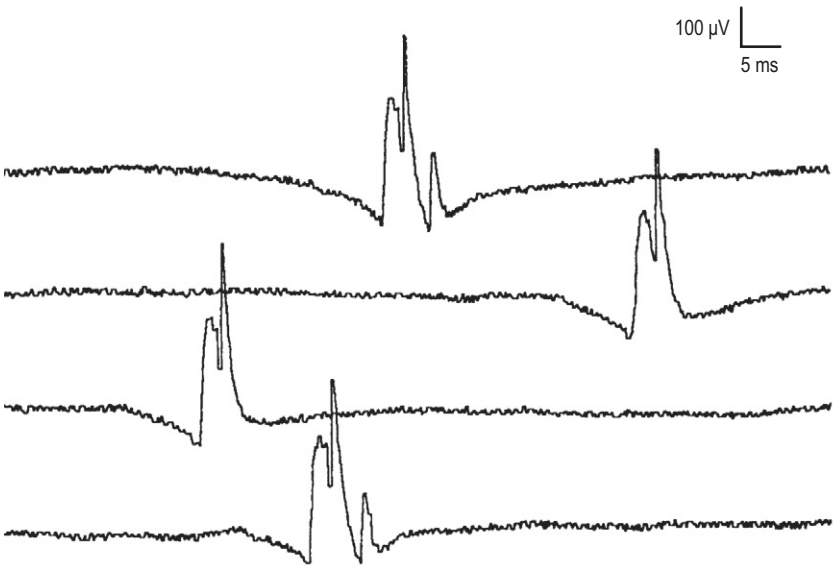
Major Spike

The major spike is the largest positive-to-negative component of the MUAP and usually occurs after the first positive peak (Figure 15–4). The major spike is the highest-frequency component of the MUAP. Because tissue acts as a high-frequency filter, as the needle is moved closer to the MUAP, the major spike increases in amplitude and its rise time shortens, indicating the proximity of the needle to the motor unit. MUAP parameters should be measured only when the needle is very close to the motor unit (Figure 15–9). When the needle is close to the motor unit, the MUAP becomes "sharp." The sharp sound represents the high-frequency component of the major spike, occurring when the major spike rise time is less than 500 µs, indicating proper needle placement.

STABILITY

MUAPs usually are stable in morphology from potential to potential. This stability is due to the fact that each time a nerve action potential is generated, there is normally effective transmission across the NMJs, and all muscle fibers of

FIGURE 15–7 Unstable satellite potential. Note the satellite potential in the first and fourth firing of the MUAP. However, the satellite potential is not present in the second and third firing of the MUAP. In early reinnervation, collateral sprouts attach to nearby denervated fibers which results in satellite potentials. However, the newly formed NMJs are immature, and do not always reach threshold, resulting in intermittent firing of the satellite potential. Eventually the satellite potential becomes incorporated into the main MUAP. This is the basis of unstable MUAPs that occur following reinnervation.



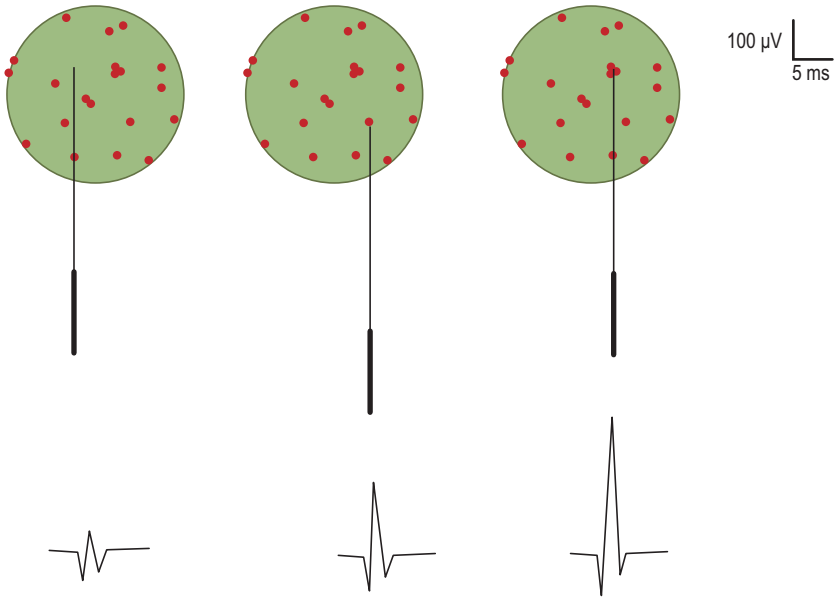


FIGURE 15–8 Relationship of motor unit action potential (MUAP) amplitude to needle position. Of all MUAP parameters, amplitude is most dependent on needle position. Only muscle fibers very close to the needle contribute to amplitude, as opposed to duration, wherein most muscle fibers contribute. Note change in amplitude as needle is moved to different locations within the same motor unit.

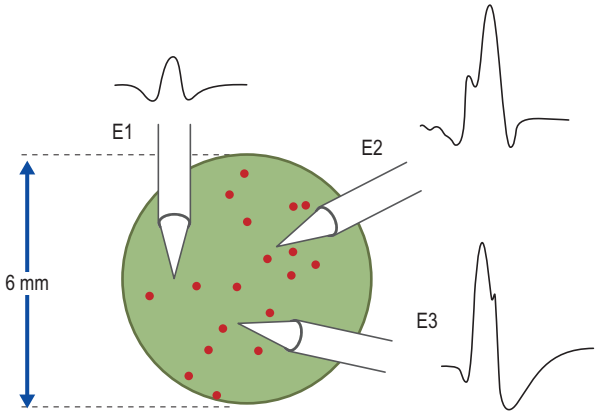


FIGURE 15–9 Motor unit action potential (MUAP) morphology and needle electromyography (EMG) position. The position of the EMG needle influences the morphology of the recorded MUAP. To properly assess MUAP parameters, the major spike must be as steep as possible, indicating the proximity of the needle to the motor unit. Note that needle electrode position E3 has the shortest major spike rise time and is the preferable position in which to assess the MUAP. Also note that although MUAP amplitude changes markedly with needle position (compare position E1 with E3), duration is relatively unaffected.
(From Dimitru, D., DeLisa, J.A., 1991. AAEM minimonograph #10: volume conduction. Muscle Nerve 14, 605. Reprinted by permission of Wiley.)

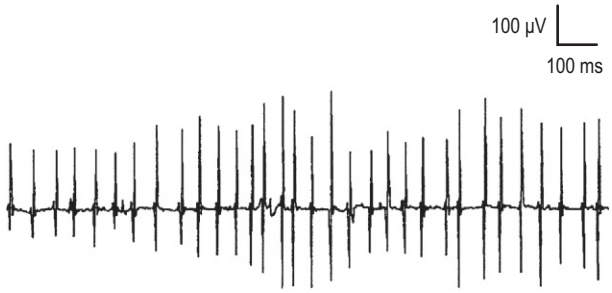


FIGURE 15–10 Unstable motor unit action potentials (MUAPs). MUAPs that change in either amplitude or number of phases from impulse to impulse are called unstable MUAPs. Unstable MUAPs occur in both primary neuromuscular junction (NMJ) disorders and disorders associated with new or immature NMJs, as commonly occur early in reinnervation. Note change in amplitude from potential to potential.

the motor unit fire. If there is impaired NMJ transmission, unstable MUAPs may result (Figure 15–10). Unstable MUAPs occur when individual muscle fibers either are blocked or come to action potential at varying intervals, leading to an MUAP that changes in configuration from impulse to impulse. There is a change between potentials in either the amplitude or the number of phases (or serrations), or both. Although unstable MUAPs always indicate unstable NMJs, they occur not only in primary disorders of the NMJ (e.g., myasthenia gravis, Lambert–Eaton myasthenic syndrome) but are also often seen as secondary phenomena in both neuropathic and myopathic disorders. Any disorder associated with denervation may demonstrate unstable MUAPs. During the process of early reinnervation, newly formed, immature NMJs often fail to conduct NMJ transmission faithfully. The result is variability in endplate transmission or intermittent blocking of transmission across some of the muscle fibers within a motor unit (Figure 15–7).

FIRING PATTERN (ACTIVATION, RECRUITMENT, INTERFERENCE PATTERN)

One of the most important and yet 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

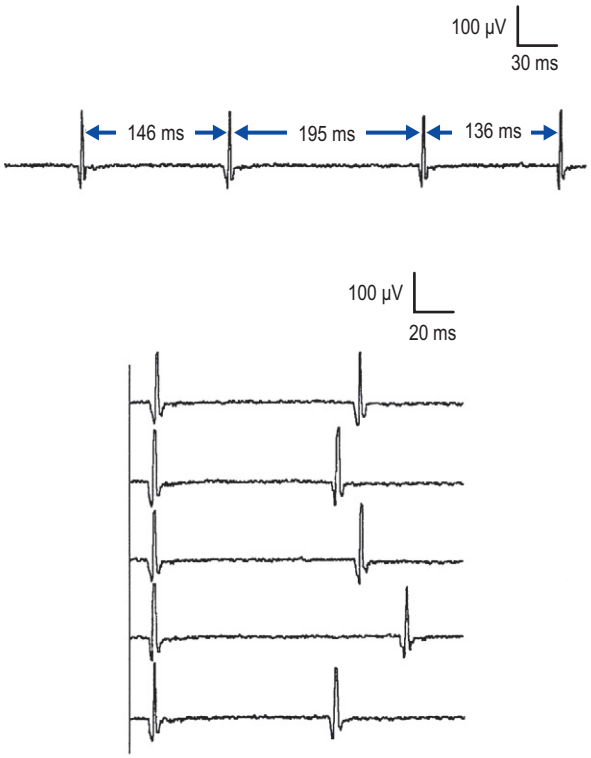


FIGURE 15–11 Motor unit action potential (MUAP) firing pattern. Normally, MUAPs fire in a semi-rhythmic pattern, with a slight variation in the interval between potentials. **Top:** Single voluntary MUAP firing at approximately 6 Hz. Note the variation in interpotential intervals. **Bottom:** Single voluntary MUAP placed on a delay line and rastered. First potential of each sweep triggers sweep. Note the variation between firing time of the next consecutive MUAP. The pattern is not quite regular (i.e., it is semi-rhythmic). This firing pattern is seen only with voluntarily activated MUAPs.

slight variation in the time interval between the same MUAP as it fires consecutively (Figure 15–11). This unique firing pattern helps to identify the potential as an MUAP under voluntary control, in contrast to various spontaneous waveforms that are not under voluntary control, and have other distinct firing patterns, such as fibrillation potentials and positive sharp waves, which are regular; complex repetitive discharges, which are perfectly regular or change abruptly; myotonic discharges, which have a waxing/waning amplitude; or fasciculation potentials, which are very slow and irregular.

During muscle contraction, there are only two ways to increase muscle force: either motor units can increase their firing rate (up to tetanic fusion frequency which is approximately 50 Hz), or additional motor units can fire (Figure 15–12). Normally, one increases force using a combination of these two processes, resulting in an orderly recruitment of motor units. With the smallest contraction, a single motor unit action potential normally begins firing semi-rhythmically at 4 to 5 Hz. Any potential that fires more slowly than 4 to 5 Hz cannot be an MUAP under voluntary control and must be a spontaneous potential. As one increases force, the first motor unit action potential increases its firing rate, and then a second motor unit action

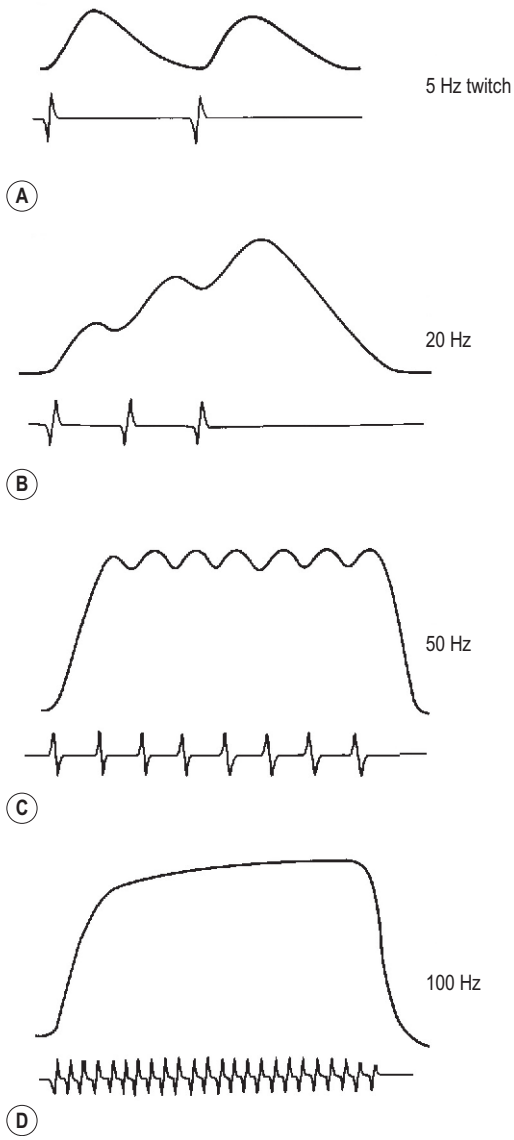


FIGURE 15–12 Relationship of force to firing frequency. **Top traces** of each pair show twitch forces; **bottom traces** show motor unit action potentials (MUAPs) firing at different frequencies. Note increased force with increased firing frequencies. To increase muscle twitch force, either motor units must fire faster or additional motor units must be added. Although the electrical MUAP lasts only 5 to 15 ms, the mechanical twitch lasts more than 100 ms. As MUAP firing rate increases, twitch forces summate. Force increases up to a frequency of approximately 50 Hz (tetanic fusion frequency). Near that frequency, the maximal overlap occurs between muscle myosin and actin filaments. Firing above that frequency may result in more even firing but does not appreciably change the amount of force generated.
(Adapted with permission from Kandel, E.R., Schwartz, J.H., Jessell, T.M. (Eds.), 1991. Principles of neural science, third ed. Appleton & Lange, Norwalk, CT.)

potential begins to fire, and so forth. This process continues, with the firing rate increasing and additional motor unit action potentials being recruited as force is increased. Normally, the ratio of firing frequency to the number of different MUAPs firing is approximately 5:1. Thus, by the time the first MUAP firing frequency reaches 10 Hz, a

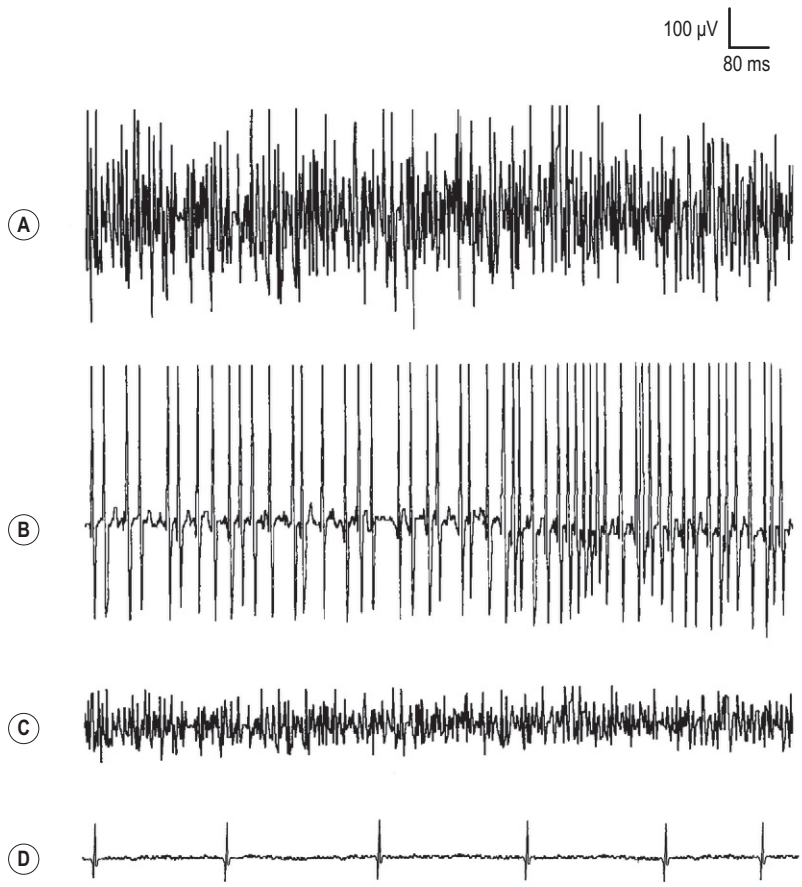


FIGURE 15–13 Interference patterns. **A:** Normal. **B:** Neuropathic. **C:** Myopathic. **D:** Central. In each trace, the patient is asked to contract maximally. In normal subjects, so many motor unit action potentials (MUAPs) fire during maximal contraction that differentiating individual motor unit action potentials is difficult. In neuropathic recruitment, a reduced number of MUAPs fire at a high frequency, resulting in an incomplete interference pattern (often referred to as the "picket fence" pattern, when only one MUAP is firing). In myopathic recruitment, although the number of MUAPs is normal, the interference pattern consists of short-duration, small-amplitude MUAPs, which fire with a small amount of force. In central disorders, the primary problem is the inability to fire faster (i.e., decreased activation); although the number of MUAPs is reduced, it is appropriate for the level of firing.

second MUAP should begin to fire; by 15 Hz, a third motor unit action potential should fire, and so forth. During maximal contraction, multiple MUAPs normally overlap and create an interference pattern in which no single motor unit action potential can be distinguished (Figure 15–13A). 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 (e.g., soleus), in which the maximal firing frequency is approximately 15 Hz.

One of the key questions to answer in assessing MUAPs is the following: *Are the number of different MUAPs firing appropriate for the firing rate*? That is, is the ratio of firing rate to MUAPs approximately 5:1? To answer that question, one must understand that increasing force depends on two processes: activation and recruitment. *Activation* refers to the ability to increase firing rate. This is a central process. Poor activation 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 unit action potentials as the firing rate increases. Recruitment is reduced primarily in neuropathic diseases, although rarely it may also be reduced in severe end-stage myopathy.

An incomplete interference pattern may be due to either poor activation or poor recruitment. Consider the two different incomplete interference patterns shown in Figure 15–14. In both cases, the patient has been asked to maximally contract the muscle of interest. In the first case (top trace), note that the same MUAP is firing rapidly at 30 Hz. Thus, although the firing rate is maximal, only one MUAP is seen firing at 30 Hz (30:1 ratio). In a normal muscle, by the time the firing rate reaches 30 Hz, one should see five or six different MUAPs firing (a ratio of approximately 5:1). Thus, in this case, the interference pattern is reduced because of decreased recruitment, but activation (firing rate) is normal. Decreased recruitment occurs when there has been loss of MUAPs, usually through axonal loss or conduction block. In the unusual situation of end-stage myopathy, if every muscle fiber of an MUAP is lost, the number of MUAPs also will effectively decrease, leading to reduced recruitment.

Contrast this with the pattern of the second patient (bottom trace), in which one also sees a single MUAP firing. In this case, however, the single MUAP is firing at 5 Hz. Thus, the firing rate (activation) is clearly submaximal, although the number of MUAPs firing (recruitment) is normal for the firing rate (a ratio of approximately 5:1). In this case, the interference pattern is reduced primarily because of decreased activation, but recruitment (i.e., the number of different MUAPs) is appropriate for the level of firing. The patient's weakness is reflected in the decreased activation of motor unit action potentials, judged by the

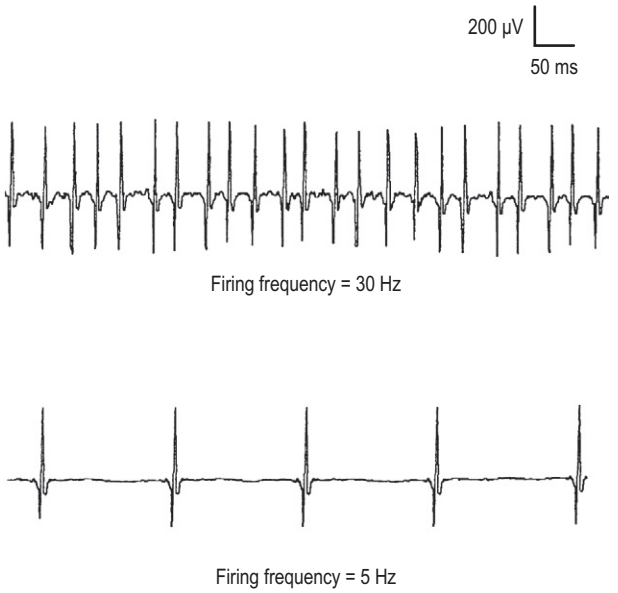


FIGURE 15–14 Incomplete interference patterns. In both traces, the patient is asked to contract the muscle maximally with the electromyography needle in place. The **top trace** demonstrates an incomplete interference pattern due to reduced recruitment. The **bottom trace** demonstrates an incomplete interference pattern due to reduced activation (see text for details).

submaximal sustained firing rate. This pattern of reduced activation may be seen if a patient cannot fully cooperate, perhaps because of pain, or has a CNS lesion (e.g., stroke, multiple sclerosis).

Of course, both decreased activation (i.e., an upper motor neuron disorder) and decreased recruitment (i.e., a lower motor neuron disorder) may be present in the same patient. This situation occurs most classically in amyotrophic lateral sclerosis, a disorder of both upper and lower motor neurons. More commonly, though, it occurs in patients with neuropathic disorders who also have difficulty moving a limb because of pain (e.g., abducting the hip in a painful L5 radiculopathy). In this latter situation, there is decreased recruitment due to loss of L5 nerve root fibers, and decreased activation due to pain.

The last concept to understand is "early recruitment." In diseases in which there is dropout of individual muscle fibers from a motor unit (e.g., myopathies 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 unit action potentials to generate a small amount of force*. On the monitor, many MUAPs will appear to fire almost simultaneously, with small amounts of force. Usually, only the electromyographer performing the study can assess early recruitment; it requires knowledge of how much force is being generated. To reiterate, early recruitment refers to the inappropriate (i.e., increased) number of MUAPs firing for the degree of force generated; it does not refer to the number of MUAPs firing for the level of activation or for the firing rate. An early recruitment pattern is typically seen in muscle disorders and in some disorders of the NMJ.

Many electromyographers will judge recruitment only during maximal contraction, by examining the interference pattern. However, not as well appreciated is that recruitment is more easily evaluated during moderate levels of contraction. Remember, the key question that must be answered is the same: Are the number of different MUAPs firing appropriate for the level of activation (firing rate)? If only one MUAP is seen firing at 15 to 20 Hz (medium level of activation), recruitment is decreased, regardless of the interference pattern. There is no need to increase the firing rate using maximal contraction in order to make this determination. Maximal contraction with the EMG needle in the muscle often is perceived as more painful by the patient, and is best avoided or at least minimized. Indeed, during maximal contraction, judging the relationship between the number of MUAPs firing and the firing rate can actually be more difficult.

PATTERNS OF MOTOR UNIT ABNORMALITIES

MUAP morphology and firing patterns usually can discriminate among the various disorders affecting the motor unit. No single parameter identifies an MUAP as myopathic, neuropathic, or associated with an NMJ disorder. Specific patterns of abnormalities in MUAP morphology and firing rate reflect whether the underlying disorder is (1) acute, chronic, or end stage; (2) neuropathic, myopathic, or associated with an NMJ transmission defect; and, if neuropathic, (3) whether the primary pathophysiology is axonal loss or demyelination (Table 15–2).

Neuropathic

Acute Axonal Loss

After an acute axonal injury to a nerve, the process of wallerian degeneration occurs in motor nerve fibers within the first 3 to 5 days, followed by denervation of the distal muscle fibers of the involved motor units. Reinnervation normally occurs as surviving nearby axons form sprouts that grow and eventually reinnervate the 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 (Figure 15–15). 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 the initial loss of motor units. Thus, in acute axonal loss lesions on needle EMG, there is a pattern of decreased recruitment of MUAPs with normal morphologies*. This pattern does not occur in slowly progressive or chronic conditions (e.g., most polyneuropathies). In those conditions, changes in MUAP morphology are always present by the time the patient

Table 15–2. MUAP Patterns and Pathophysiology

	MUAP Morphology			MUAP Firing Pattern	
	Duration	Amplitude	Phases	Activation	Recruitment
Acute neuropathic – axonal	NL	NL	NL	NL	↓
Chronic neuropathic – axonal	↑	↑	↑	NL	↓
Neuropathic – demyelinating (CV slowing alone)	NL	NL	NL	NL	NL
Neuropathic – demyelinating (conduction block)	NL	NL	NL	NL	↓
Early reinnervation after severe denervation (nascent units)	↓	↓	↑	NL	↓↓
Acute myopathic	↓	↓	↑	NL	NL/EARLY
Chronic myopathic	↓/↑	↓/↑	↑	NL	NL/EARLY
Myopathic – endstage	↓/↑	↓/↑	↑	NL	↓↓
NMJ disorders – increased jitter	NL	NL	NL	NL	NL
NMJ disorders – intermittent block	NL/↓*	NL/↓*	NL/↑*	NL	NL/EARLY
NMJ disorders – severe block	↓	↓	↑	NL	↓↓
CNS disorders	NL	NL	NL	↓↓	NL

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

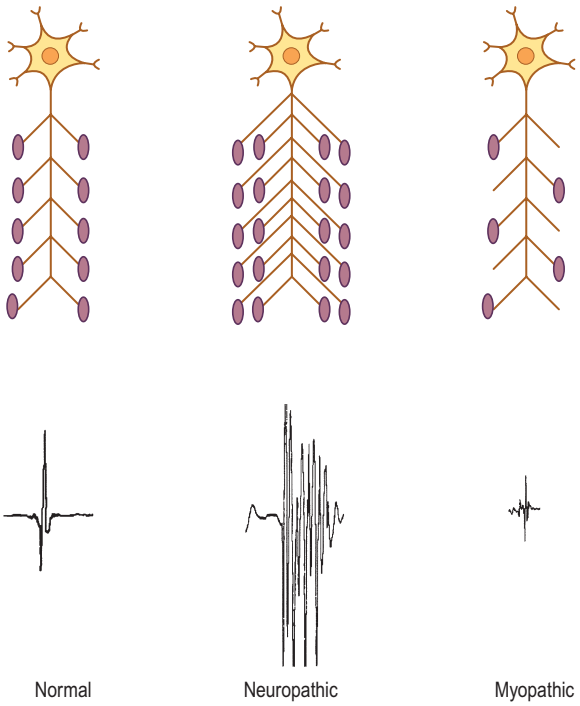


FIGURE 15–15 Motor unit action potential (MUAP) morphologies. Normal MUAPs have two to four phases. In chronic neuropathic lesions that occur after reinnervation, the number of muscle fibers per motor unit increases, resulting in long-duration, high-amplitude, and polyphasic MUAPs. In myopathies or in neuromuscular junction disorders with block, the number of functional muscle fibers in the motor unit decreases. This leads to short-duration, small-amplitude, and polyphasic MUAPs.

presents with symptoms. The acute neuropathic pattern associated with axonal loss characteristically occurs in the first several weeks after trauma, compression, or nerve infarction. The only other situation in which a similar pattern is seen is in pure demyelinating lesions with conduction block (discussed in section on Neuropathic: Demyelinating).

Chronic Axonal Loss

After axonal loss and denervation, the process of reinnervation can occur by one of two mechanisms. If there has been complete denervation, the only possible mechanism for reinnervation is axonal regrowth from the point of injury (see section on Early Reinnervation Following Severe or Complete Denervation). Typically, this regrowth is quite slow (no more than 1 mm per day) and may take months to years, depending on the length of the nerve. For the regrowth to occur, however, the anterior horn cells must remain intact. For example, the original nerve fibers can regrow following transection of a nerve, but not after poliomyelitis, which results in the death of anterior horn cells.

In contrast, in cases of partial or gradual denervation, reinnervation usually occurs through collateral sprouting by adjacent surviving motor units (Figure 15–5). As the number of muscle fibers per motor unit increases, MUAPs become prolonged in duration, with a high amplitude, and polyphasic. *These MUAP changes, in conjunction with decreased recruitment, are the hallmarks of reinnervated motor unit action potentials and nearly always imply chronic neuropathic disease (i.e., disorders of the anterior horn cell, nerve root, or peripheral nerve). Similar to other neuropathic conditions, during maximal contraction, the interference pattern will not be full, secondary to decreased recruitment*

of MUAPs (Figure 15–13B). Long-duration, high-amplitude, polyphasic MUAPs are never seen in acute conditions. When present, they always imply that the process has been present for at least several weeks and more often for months or years.

Demyelinating

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. *In pure demyelinating lesions, MUAP morphology remains normal.* If demyelination results in conduction velocity slowing alone, the nerve action potential will still reach the muscle, albeit more slowly, and the number of functioning motor units will remain normal. Accordingly, there will be no change in either MUAP morphology or recruitment pattern on needle EMG. If demyelination results in conduction block, however, the number of available MUAPs effectively decreases. Although the MUAP morphology remains normal, the firing pattern shows decreased recruitment. This pattern of reduced recruitment with normal MUAP morphology is seen only in demyelinating lesions with conduction block (e.g., some cases of Guillain–Barré syndrome, carpal tunnel syndrome) or in cases of acute axonal loss before enough time has passed for reinnervation to occur.

Myopathic

Acute

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 MUAPs of shorter duration and smaller amplitude (Figure 15–15). 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 (i.e., the number of anterior horn cells and axons) remains normal. Thus, the recruitment pattern remains normal for the level of activation. Because each motor unit contains fewer muscle fibers, however, it cannot generate as much force as a normal motor unit. To compensate, more MUAPs will fire than are normally needed for a certain level of force, resulting in early recruitment. The interference pattern will fill easily with a small amount of force from the patient (Figure 15–13C). *Consequently, the pattern associated with an acute myopathy is short-duration, small-amplitude, polyphasic MUAPs with normal or early recruitment.*

Chronic

In chronic myopathies, especially those with necrotic or inflammatory features (e.g., polymyositis, dystrophies), some denervation and subsequent reinnervation commonly occur. Consequently, long-duration, high-amplitude, polyphasic MUAPs can develop, although such MUAPs are most commonly seen in chronic neuropathic disease. In many chronic myopathies, two populations of MUAPs are often seen: both long-duration, high-amplitude, polyphasic MUAPs and short-duration, small-amplitude, polyphasic MUAPs, often in the same muscle. Rarely, only long, large, polyphasic MUAPs are seen. *The key to differentiating chronic myopathic from chronic neuropathic MUAPs is the assessment of the recruitment pattern.* In chronic myopathies, recruitment usually is normal or early. If an early recruitment pattern is not seen, at the very least the recruitment pattern appears better than what would be expected based on the chronic MUAP changes. In some cases of very 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), except for the recruitment pattern that appears "too good" for the apparent amount of reinnervation.

Endstage

In the very late stages of some dystrophies, periodic paralysis, and unusual, very chronic focal myopathies (e.g., inclusion body myositis), endstage muscle may occur. In such situations, the actual number of motor units may effectively decrease if every fiber of some motor units dies or becomes dysfunctional. The result is 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 endstage muscle from myopathy.

Early Reinnervation following Severe or Complete Denervation

Reinnervation most often occurs 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, at some point in time it will reinnervate some, but not all, of the original muscle fibers. At that point, the MUAP will be short duration, small amplitude, and polyphasic, similar to an acute myopathic motor unit action potential (Figure 15–16). Early reinnervated motor unit action potentials following severe denervation are known as *nascent motor units*. *The key factor that differentiates nascent motor unit action potentials from myopathic motor unit action potentials 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.* Although nascent motor units are uncommon, they emphasize that not all short-duration, small-amplitude, polyphasic MUAPs are myopathic.

Neuromuscular Junction Disorders

MUAP morphology and firing patterns in NMJ disorders depend on the severity of the disorder. If the NMJ

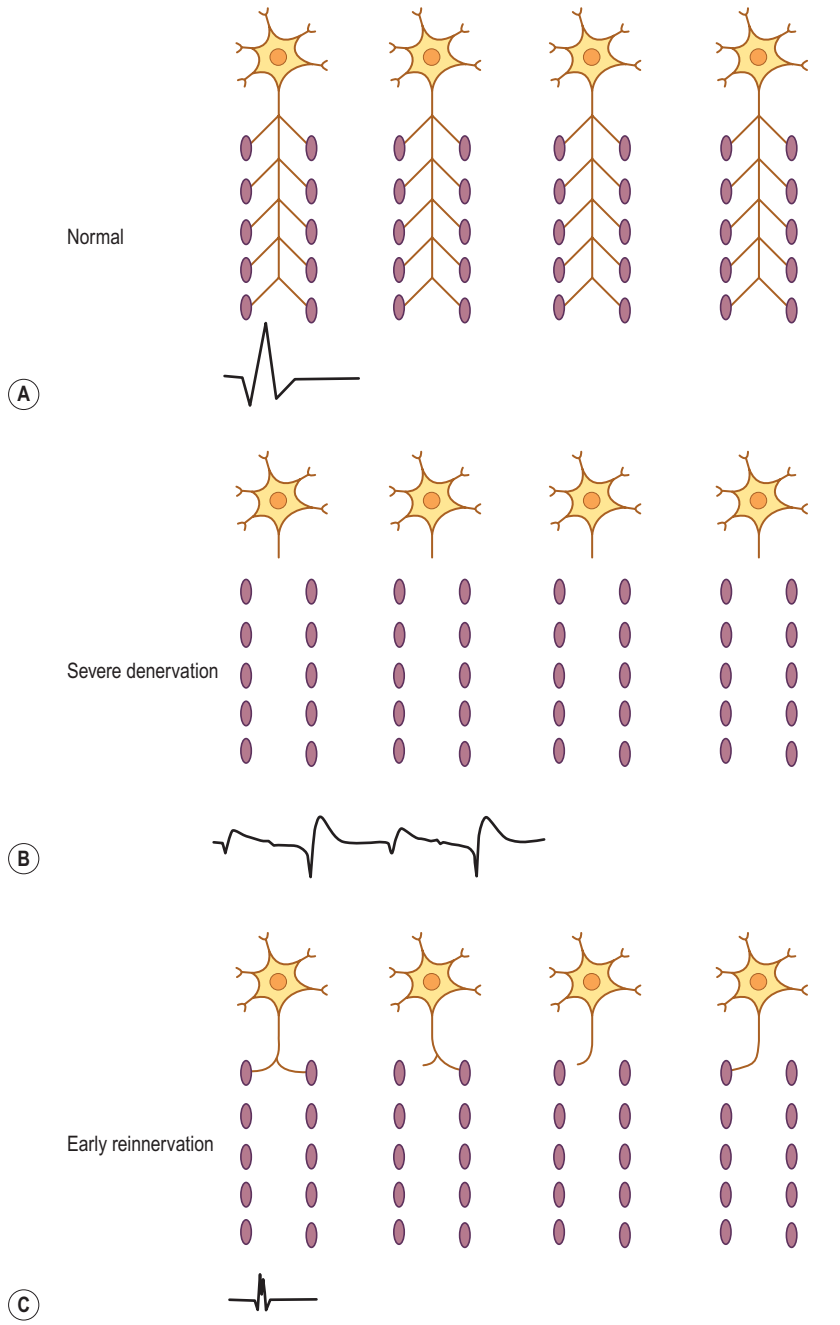


FIGURE 15–16 Nascent motor units. After a severe axonal loss lesion, wallerian degeneration occurs distal to the injury, resulting in denervation **B**. If there are no surviving nearby axons, reinnervation can occur only from axonal regrowth from the terminal stump. Early in this reinnervation process, there will be a point at which some but not all of the muscle fibers are reinnervated **C**. At that point, motor unit action potentials (MUAPs) will be short duration, small amplitude, and polyphasic, resembling myopathic units. Compare the nascent MUAP **C** with the normal MUAP **A**. Nascent MUAPs are differentiated from myopathic MUAPs by the reduced recruitment pattern compared with normal or early recruitment seen in myopathy.

disorder is mild, resulting in only slight variation of the firing of muscle fibers within the motor unit, 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. The morphology (amplitude, number of phases, or both) will vary from potential to potential. With greater and more persistent block, there will effectively be loss of individual muscle fibers within a motor unit. Thus, the MUAP will become short, small, and polyphasic, similar to a myopathic MUAP. Similarly, recruitment remains normal, or it may become early as each motor unit is able to generate less force. To reemphasize, both myopathies and severe NMJ disorders may result in short, small and polyphasic MUAPs with normal or early recruitment. 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 short duration, small amplitude, and polyphasic, but with decreased recruitment, reflecting the reduced number of available motor units. This latter unusual pattern also can be seen in endstage myopathy and in nascent motor units.

Central Nervous System Disorders

In CNS disorders, normally there is 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 unit action potentials rapidly (i.e., reduced activation). Thus, although the interference pattern is incomplete, with a reduced number of motor unit action potentials firing, the actual number of motor unit action potentials (i.e., recruitment) is appropriate for the reduced level of activation (Figure 15–13D).

Occasionally, other patterns may be seen with CNS disorders. In spinal cord lesions, motor units may be lost at the level of the lesion because of segmental loss of anterior horn cells. For example, in a C6 spinal cord lesion, denervation, reinnervation, and decreased recruitment of MUAPs may be seen in the C6-innervated muscles. In the weak lower extremities, however, only decreased activation, but not decreased recruitment, of MUAPs will be seen. In those muscles that receive partial innervation from C6 (e.g., pronator teres, C6–C7 innervated), there may be a combination of decreased recruitment and decreased activation of MUAPs.

Only rarely are other EMG abnormalities seen in CNS disorders. In some reported patients with multiple sclerosis, signs of denervation and reinnervation have been seen, presumably due to involvement of motor fibers as they leave 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.

Tremor may occur in some CNS disorders and can complicate the interpretation of both spontaneous activity (see Chapter 14) and MUAP morphology. Tremor is recognized as a bursting pattern of voluntary MUAPs separated by relative silence. When tremor occurs at rest (e.g., in Parkinson's disease), the spontaneous bursting discharges may be mistaken for myokymic discharges. Although both tremor and myokymia 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, whereas myokymia cannot be voluntarily influenced by the patient. Most tremors, however, worsen with activation. Because multiple MUAPs fire simultaneously in tremor, the morphology of individual MUAPs may be difficult to assess, and polyphasia may appear to be increased. In general, it is very difficult to accurately judge MUAP morphology, stability, or recruitment if the patient has a tremor when activating their muscles.

Lastly, persistent involuntary contraction can be seen during the needle EMG as the result of central disorders, including dystonia, stiff-person syndrome, and tetanus. In all of these disorders, MUAP morphology will be normal, and the EMG pattern will be one of involuntary persistent firing of MUAPs, characterized by delayed relaxation and co-contraction of muscles. Normally, individuals can easily relax their muscles and stop contracting. In these CNS disorders, however, this often is not possible. In addition, co-contraction of agonist and antagonist muscles occurs. Normally, antagonist muscles are quiet while agonists are contracting (e.g., the triceps is relaxed while the biceps is contracting and flexing the elbow). In dystonia, MUAP firing often actually increases in the antagonist muscle when the patient is instructed to move the agonist muscle (e.g., increased firing in the tibialis anterior when the patient plantar flexes the ankle) (Figure 15–17).

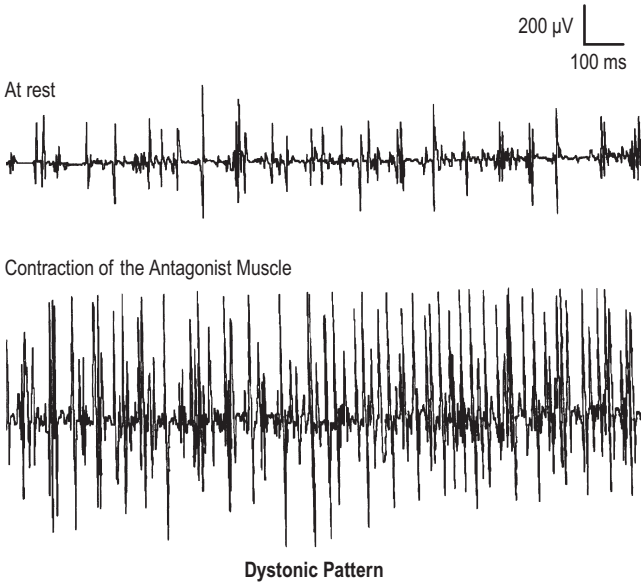


FIGURE 15–17 Dystonic firing pattern. Recording from the tibialis anterior muscle in a patient with dystonia. **Top trace:** At rest. Note the persistent firing of motor unit action potentials. **Bottom trace:** The patient is asked to plantar flex the ankle (i.e., activate an antagonist muscle). Note that the firing markedly increases. This pattern of co-contraction of agonist and antagonist muscles occurs in dystonia and other central nervous system disorders.

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