Rapid MUAP Quantitation

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INTRODUCTION

Electromyography (EMG) is used to study the electrical activity of muscles to assist in the diagnosis of neuromuscular diseases. During each section of the examination, normal or abnormal signals may be recorded which represent any underlying function or dysfunction of the muscle fibers or nerves that supply the muscle being examined. A major component of the needle EMG examination is the assessment of voluntary motor unit action potentials (MUAPs) which reflect the morphologic changes that occur within the motor units (MUs) in a muscle. The MUAP changes are used to detect an underlying neuromuscular disorder, distinguish between neuropathic or myopathic processes, and assess chronicity of the disease.

The most important tasks of a clinical electrodiagnostic (EDX) physician include recognition, quantitation, and interpretation of MUAPs. This depends on the EDX physicians’s skills in the areas of pattern recognition and rapid quantitation. Pattern recognition is the basic skill that allows for the identification of a waveform as a MUAP that can distinguish MUAPs from fibrillation potentials or endplate spikes. The skill of rapid quantitation is used to efficiently determine whether the parameters of MUAPs are normal or abnormal. Pattern recognition and rapid quantitation are equally important in mastering clinical EMG and are skills that can be learned simultaneously.

PATTERN RECOGNITION

Each potential that is recorded during EMG may be characterized by its: 1) firing pattern (regular, semi-rhythmic, irregular), 2) configuration (triphasic, biphasic, polyphasic, etc.), and 3) auditory sound. Pattern recognition allows for identification of the firing pattern for each discharge and is most useful in the recognition of each type of waveform. Pattern recognition is learned by associating a sound with a name when hearing them together many times. The skill of pattern recognition allows human auditory systems to categorize the wide range of EMG signals that occur in normal and diseased muscle.

The skills of auditory pattern recognition form the basis of learning the major distinct patterns of firing of EMG discharges:

- **Semi-rhythmic** – recurring in orderly, but not precise intervals
- **Regular** – recurring at precisely defined intervals that may be identical, or change slowly or rapidly, or to change in linear or exponential manners
- **Irregular** – recurring in random intervals with no predictability
- **Burst** – groups of discharges firing at one interval in the burst, with the burst recurring at slower intervals.
RAPID QUANTITATION

Quantitation refers to placing a numerical value on a measured parameter. Nerve conduction studies (NCSs) quantitate amplitudes, distal latencies, and conduction velocities of nerve and muscle responses to an applied electrical stimulus. During needle EMG, estimation of the different parameters of MUAPs is compared with normal values in determining whether a muscle is “normal” or “abnormal.” In many cases, determination of whether a MUAP is normal or abnormal is made by a general estimation (e.g. “these MUAPs sound normal” or “those MUAPs sound long”). In actuality, when the EDX physician is making these decisions, he or she is performing an aspect of rapid quantitation – possibly without even realizing it!

In contrast to pattern recognition, quantitative assessment of voluntary MUAPs provides information that has specific clinical value in the assessment of neuromuscular diseases. It would be ideal to have formal, quantitative measures of each of the parameters of the MU that is assessed during needle EMG examinations, just as is done for NCSs. The limitations of current EMG equipment, and the time required to accomplish such measurements preclude this for routine EMG. A number of methods have been published for formal quantitation of MUAPs, including manual and automated measurement of the MUAP parameters as will be described (i.e., individual MUAP measurement, turns-amplitude measurement, and frequency analysis). While these continue to be developed, none are sufficiently efficient to be used for routine clinical EMG examinations. Formal quantitative EMG is rarely performed.

However, the quantitation of MUAP parameters in most clinical settings can efficiently be performed by “rapid quantitation”. A skilled EDX physician can accomplish this by applying the well-defined techniques of “rapid quantitation” EMG, after learning how to make estimates of each of the parameters of the MU that is assessed during needle EMG examinations, just as is done for NCSs. The limitations of current EMG equipment, and the time required to accomplish such measurements preclude this for routine EMG. A number of methods have been published for formal quantitation of MUAPs, including manual and automated measurement of the MUAP parameters as will be described (i.e., individual MUAP measurement, turns-amplitude measurement, and frequency analysis). While these continue to be developed, none are sufficiently efficient to be used for routine clinical EMG examinations. Formal quantitative EMG is rarely performed.

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Rapid quantitation EMG requires the following steps to be taken separately for voluntary EMG activity:

- One to three individual MUAPs from a single area of muscle are recorded, with a stable needle, and a display as outlined above.
- The rate of firing is determined by the free-running sweep or from an automated measurement (triggering on a single MUAP) for each of the potentials.
- The number of MUAPs is compared with the rate of firing of the potentials to determine recruitment.
- The MUAP parameters (duration, amplitude, phases, turns, stability) of each of the potentials with a rise time less than 0.5 ms are estimated from their sound and appearance on the triggered sweep.
- With no change in activation, the needle is moved to additional areas of muscle (0.5 mm), where the steps above are repeated.
- Recordings in different areas are repeated until a minimum of 20 to 30 potentials have been recorded. The findings at each location are mentally averaged for the locations tested in the muscle.
- The averaged measurements of recruitment and MUAP parameters are recorded for a muscle before proceeding to the next muscle.

When these steps have been mastered, the EDX physician will be able to make each of the measures with over 90% accuracy and do so in 2 minutes or less per muscle.

Mastery requires learning the following steps:

- The activation of MUAPs is maintained at a fixed level so that there are one to three potentials firing at each location in the muscle (fewer as learning begins; more as learning proceeds).
- The sound of the potentials at each location in the muscle must be auditorily monitored for 3 to 5 s as the potentials are recorded.
- An estimate of the average firing rate, number of potentials firing, and of the other MUAP parameters of each active potential is made by ear from the sound, and written down.
- The MUAPs that have been estimated are stored on the free running and triggered sweep.
- The actual firing rates, numbers of potentials, and parameters of the potentials are made from the storage scope and compared with the estimates.
- Estimates are corrected and the measurements are repeated until the estimates are within 90% of those measured.
RAPID QUANTITATION OF MUAP PARAMETERS

MUAPs are the electrical discharges of MUs under voluntary control and are identified by the semi-rhythmic pattern of firing which can be altered by patient effort. The following parameters of MUAPs can be rapidly quantitated, and each parameter reflects the underlying changes of the MUs within the muscle being examined.

- Rise time
- Duration
- Amplitude
- Phases / turns
- Recruitment
- Stability

MUAP parameters should be estimated at the same level of contraction where individual potentials can be readily distinguished with flat baselines between them. Duration and amplitude estimates should be made only where potentials are recorded with a rapid rise time (sharp, clicking sound). This is best done at a gain of 200 µV/cm and sweep speeds of both 1 s (50 ms/div) for pattern and rate, and of 50 ms (10 ms/cm) for configuration.

The following are the parameters of MUAPs that can be rapidly quantitated:

### Rise Time

- Time from maximal positive deflection to negative deflection.
- Reflects the distance of the recording electrode to the muscle fiber. The closer to the fiber, the shorter (more rapid) rise time.
- Rise time should be less than 0.5 ms (recording from fibers within 0.5 mm) when assessing MUAPs.

### Duration

The duration of a single MUAP is the time from the initial (often slow unless at the end-plate zone) shift of the potential away from the baseline until its final return to the baseline (also often slow). This duration is much longer than the spike component of the potential. The durations of MUAPs in a muscle have a normal distribution that can be described by a mean and standard deviation. The normal values differ from muscle to muscle and also vary with age and temperature. Average duration increases by 35% from infancy to adulthood (biceps) and by 65% (abductor digiti minimi), probably due to dispersion of the endplates when the muscles become larger in size. Mean duration increases by 10% per degree decrease in temperature, and the percent of polyphasic potentials also increase with decreased temperature.

When assessing the durations of MUAPs in a muscle, the overall assessment is based on the interpretation of 20 to 30 different MUAPs, averaging the durations of each of the MUAP measured, and comparing the mean duration with those of normal or published data. The mean values published by Buchthal (which exclude long polyphasic units) are the accepted normal:

1. Normal - A mean duration for the MUs in a muscle within two standard deviations of the mean values determined for the same muscle in normal individuals of the same age group (20 or more units measured) and no more than 10% of the units outside the normal range for that muscle. For example, for a muscle in which the mean published duration is 10 ms, the normal range of duration is 8 to 12 ms.
2. Short Duration - Short duration MUAPs are those in which the mean duration of the 20 to 30 MUAPs measured is less than a value two standard deviations below the mean for that muscle in normal individuals. Alternatively, if more than 10% of the potentials are of shorter duration than the lower limit of the normal range for that muscle, the muscle is considered to have abnormally short duration.
MUAP. This abnormality may be described as: (1) Increased proportion of short duration potentials; (2) Scattered, few occasional, moderate numbers of or many short duration potentials; or (3) Localized areas of short duration MUAPs. While short duration MUAPs may be described as “spiky,” it is always best to specify the mean and range of durations, or give some estimate of the number or percentage of short duration MUAPs seen in the total population. It should be based on the study of at least 20 MUAPs in different areas of the muscle.

Short duration MUAPs are indicative of a process in which there has been a loss of muscle fibers in the MU. These are commonly seen in myopathies, but may also occur in neuromuscular junction disorders and early reinnervation following a severe neurogenic lesion (“nascent MUAPs”).

3. Long Duration – Long duration MUAPs are those in which the mean duration of the 20 to 30 MUAPs measured is greater than a value two standard deviations above the normal mean, or if more than 10% of MUAPs are longer duration than the upper limit of the normal range. The description of this change should use grades similar to that used for short duration potentials, and should include an estimate of the proportion of long duration potentials in the population. It should be based on the study of at least 20 MUAPs in different areas of the muscle.

Long duration MUAPs are reflective of an increased number of muscle fibers innervated by the anterior horn cell/axon, and is characteristic of reinnervation following a neurogenic disorder.

The amplitude of the MUAP reflects the sum of the action potentials of only a few muscle fibers that are closest to the tip of the recording electrode, rather than the area of the fibers in the MU. Thus, the amplitude is less indicative of the size of the MU than the duration.

The amplitude of a single MUAP remains constant throughout its period of firing. It is a function of the distance of the needle electrode from the MUAP. Therefore, it is more variable for any single MU. Because of these difficulties, acceptable normal values for amplitudes of MUAPs are not available. Amplitude is the parameter that is least able to be quantitated rapidly, since there is minimal recognizable auditory change with different amplitude sizes. In other words, a high amplitude MUAP will sound essentially the same, only louder, than a smaller amplitude MUAP of the same duration.

### Amplitude

<table>
<thead>
<tr>
<th>Amplitude</th>
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<tbody>
<tr>
<td>- Positive peak to negative peak</td>
</tr>
<tr>
<td>- Sum of the action potentials of muscle fibers closest to the recording electrode. Usually made up of less than 15 muscle fibers (typically 1 to 8).</td>
</tr>
<tr>
<td>- Determined by the diameter of muscle fibers, number of muscle fibers, and temporal distribution of action potentials closest to the recording electrode.</td>
</tr>
<tr>
<td>- May have several peaks due to asynchronous firing of potentials.</td>
</tr>
<tr>
<td>- Larger amplitudes may reflect increased fiber density of muscle.</td>
</tr>
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</table>

### Number of Phases or Turns

A phase of an MUAP is defined as the number of baseline crossings plus one. Turns are defined as potential reversals without crossing the baseline. Both reflect the synchrony of firing of the muscle fibers in the MU. When fibers in a MU fire less synchronously, increased turns and phases develop. This may occur in early reinnervation in a neurogenic process or in myopathic disorders. MUAPs are typically triphasic with an initial positivity followed by a negative spike and then a slow positivity. They may be more complex, having four to six phases in normal muscle. Potentials with more than four phases are referred to as “polyphasic”, and are found in different normal muscles in varying proportions (5% to 15%).

1. Normal - A muscle contains no more than the accepted percentage of polyphasic units for that muscle in a patient of that age.
2. Increased Numbers of Polyphasic - A greater than normal proportion of polyphasic potentials for that muscle. Usually graded by percentage.
3. Highly Polyphasic - The occurrence of potentials having greater than six separate phases. The number of these as a proportion of the total number of motor unit potentials should be estimated.

Caution - Estimates of the numbers of polyphasic MUAPs should be made with minimal to moderate contractions since with strong contractions the superimposition of different MUs may give the appearance of polyphasic units, particularly in the presence of tremor.

<table>
<thead>
<tr>
<th>Phases / Turns</th>
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</thead>
<tbody>
<tr>
<td>• Phases = changes in direction of the potential which cross the baseline (equals baseline crossings plus one)</td>
</tr>
<tr>
<td>• Turns = changes in direction of the potential greater than 50 μV that do not cross the baseline</td>
</tr>
<tr>
<td>• Both are indicators of synchrony of firing of potentials. With decreased synchrony (loss of muscle fibers in myopathy, collateral sprouting in neuropathy), there is increase in number of spikes (either turns or phases)</td>
</tr>
<tr>
<td>• Normal potentials have three or four phases. Less than 15% of MUAPs should have more than four phases (polyphasic).</td>
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Recruitment

The number of MUs in a muscle may be considered in two ways. The first is the total number of MUs that could be fired if the anterior horn cell pool received adequate input. The second is the actual number of MUs that are activated when a patient attempts a voluntary contraction. The first of these is more pertinent in assessing the presence or absence of disease involving the lower motor neuron (MN) and is called recruitment. The second is quite variable and changes with the patient's cooperation, the strength of the muscle, pain, and the presence or absence of disease of the upper MN.

The recruitment of MUAPs, as reported in the EMG report, should estimate the total number of MUs available for activation and not the actual number which the patient fires. The total number available can be best assessed by the pattern of recruitment of MUAPs with increasing voluntary effort. In a normal muscle, as the effort exerted increases, each MUAP that is already firing will increase its frequency. As it does so, additional MUAPs will begin to fire (be recruited). This pattern of recruitment can be characterized objectively by the recruitment frequency - the frequency of firing of any single MUAP when the next MUAP is recruited. It can also be described by the number of units firing at specific frequencies.

In the presence of a lower MN disease with loss of the anterior horn cell or peripheral motor axon, the recruitment frequency will increase (i.e. MUAPs will fire more rapidly before additional MUs are recruited). Conversely, the rate of firing of MUAPs already firing will be unduly fast for the number of MUAPs that have been activated. By using the pattern of recruitment, it is possible to assess the normality of the number of MUs in the muscle with mild, moderate, or maximal effort on the part of the patient.

Although there have been few studies that have identified the normal recruitment frequencies for each muscle, for most muscles, a general guideline in assessing recruitment is the “Rule of 5”. This guideline suggests that at a low level of activation of motor unit potentials (with approximately 3-4 MUAP firing), the electromyographer should (1) identify the number of MUAP firing, (2) determine the firing rate of the fastest firing MUAP, and (3) calculate the ratio of firing rate: # MUAP firing. This ratio (firing rate of a MUAP / number of individual), should be 5 or less. For example, when the firing frequency of an initial MUAP reaches 10 Hz, a second MUAP should begin to fire (10 Hz / 2 potentials). When the frequency reaches 15 Hz, 3 MUAPs should be firing (15 / 3 = 5). With loss of MUs as in neurogenic lesions (see below), MUAPs fire faster with a reduction in number of units firing (e.g. initial MU fires at 20 Hz before the second MUAP begins firing – 20 Hz / 2 = 10). This indicates a reduction in recruitment.

1. Normal - The pattern of recruitment is normal for that muscle, with adequate numbers of MUAPs being recruited for the frequency of firing present. If maximal effort can be obtained, a full interference pattern is seen.
2. Reduced Recruitment - A higher recruitment frequency or a smaller number of MUAPs recruited for any given rate of firing than is expected from that muscle. This should not be used to describe patients who fire relatively few MUAPs because of pain, strong muscles, upper MN lesions, or poor cooperation. Since they fire few potentials, the potentials can be expected to fire slowly with a normal pattern of recruitment.
3. Poor Activation - A normal recruitment pattern and normal recruitment frequency, but with relatively few MUAPs firing. These potentials fire slowly, but the actual recruitment frequency of additional potentials is normal. This occurs in upper MN disorders, poor cooperation, pain, an excessively strong muscle, or two joint muscles, such as the gastrocnemius. It is not evidence of lower MN disease.
4. Increased Number in Proportion to Force - The occurrence of large numbers of MUAPs with normal recruitment frequency and normal patterns of recruitment, but with minimal effort. This must be graded in proportion to the force exerted, since the patterns of firing are entirely normal. It is the only estimate described
which requires a consideration of the force exerted by the muscle. It is evidence of disease involving the muscle directly.

**Recruitment**

- The orderly addition of MUAPs firing as the rate of individual MUAPs firing increases.
- Normal muscle – increasing voluntary effort causes an increase in the rate of firing of individual MUAPs and the initiation of the discharge of additional MUAPs.
- Individual MUs begins firing at 4 to 5 Hz and the second unit is recruited when the first unit is firing at 8 to 10 Hz.

**Reduced recruitment**

- Reduced recruitment occurs in disorders where a smaller muscle population is recruited to generate the same force. This may be seen in disorders such as myasthenia gravis or Lambert-Eaton myasthenic syndrome.

**Rapid Recruitment**

- Rapid recruitment occurs in disorders where a larger muscle population is recruited to generate a stronger force. This may be seen in disorders such as myopathy or neurogenic disorders.

**Poor activation**

- Poor activation occurs in disorders where the muscle fibers are activated at a slower rate. This may be seen in disorders such as MN disease or myopathies.

**Stability**

Each time a normal MUAP fires, the morphology of the waveform remains identical. The stability of a MUAP is due to maintenance of the integrity of neuromuscular transmission at each nerve terminal. Therefore, each time a MU fires, all of the innervated muscle fibers within that MU are depolarized and the summated action potential from all of those fibers is recorded. When neuromuscular transmission fails, such as in diseases of the neuromuscular junction or in immature collateral nerve sprouts in neurogenic disorders, MUAPs become “unstable” and their morphology varies with each firing. Varying MUAP can be recognized by the “bongo drum” sound during the needle EMG examination. Formal quantitation of the degree of MUAP variation requires single fiber EMG (assessing jitter and blocking), although the degree of variation can be estimated by rapid quantitation.

**Fibrillation Potentials**

Fibrillation potentials are the MUAPs of single muscle fibers firing spontaneously in the absence of innervation of that muscle fiber. Fibrillation potentials occur in disorders of either muscle or nerve dysfunction, such as polyneuropathies, mononeuropathies, radiculopathies, MN disease, or myopathies. Fibrillation potentials fire with a regular rate, although a linear slowing of firing rate may occur. Typical fibrillation potential configurations include biphasic and monophasic spike potentials, or triphasic, initially positive spike potentials firing slowly and regularly (sound like the “ticking of a clock”).

Quantitation of fibrillation potentials is rarely necessary. In most cases, grading of the density of fibrillations (e.g. from 1+ to 4+) is sufficient to estimate the degree of denervation of the muscle.
Fasciculation Potentials

A spontaneously occurring, single discharge having the general configuration of a MUAP is a fasciculation. Fasciculation potentials fire in an irregular pattern, and may occur only occasionally or continuously. The size, shape, configuration, and duration of the discharge will not enter into the consideration of whether it is called a fasciculation. However, the waveform and its pattern of occurrence should be described. If such single, spontaneously occurring discharges are complex in appearance or have an unusual pattern of recurrence, these should be specifically described.

Fasciculation potentials may be rapidly quantitated by counting the number occurring per minute.

Complex Repetitive Discharges

Complex repetitive discharges (CRDs) are spontaneously occurring waveforms generated by ephaptic spread of muscle fiber action potentials in a “circuit” along neighboring muscle fibers. CRDs appear polyphasic due to the time-locked firing of multiple muscle fiber action potentials, recur at variable rates of 10 to 150 Hz, and remain uniform in firing frequency, shape, and amplitude. The pattern of firing is regular, although they start and stop abruptly and may show a sudden change in character during the discharge. They often sound like a “motor boat”. CRDs may be seen in chronic or longstanding neuropathic or myopathic disorders, and are due to ephaptic activation of groups of several adjacent muscle fibers following reinnervation.

Quantitation of complex repetitive discharges is rarely necessary. However, rapid quantitation can be used to identify the firing rate of a discharge.

Myotonic Potentials

Monophasic, biphasic, or triphasic individual muscle fiber potentials occur at 20 to 80 per s with gradually increasing and/or decreasing amplitudes and rates. The firing pattern is regular, although gradually changes occur exponentially, and the discharges are usually induced by needle movement or other mechanical stimulus. Myotonic discharges sound like a “dive-bomber”. They are seen in myopathies characterized by abnormalities in the sodium or chloride channel on muscle membranes, such as myotonnic dystrophy, myotonia congenita, hyperkalemic periodic paralysis, and some inflammatory myopathies.

Quantitation of myotonic discharges is rarely necessary. However, rapid quantitation can be used to identify the firing rate of a discharge.

Neuromyotonia

Monophasic or biphasic potentials occurring at a very high frequency (over 100 per s), of gradually decreasing amplitude. It may be spontaneous, induced by needle movement, or voluntary action. They often sound like an “Indy 500 racecar”. Quantitation of neuromyotonic discharges is rarely necessary. However, rapid quantitation can be used to identify the firing rate of a discharge.

Myokymic Discharges

Regular recurrence of bursts of potentials at relatively constant intervals of 0.5 to 10 s. The potentials within the burst may have any of the characteristics of MUAPs, but fire at rapid rates of 30 to 80 per s. The number of potentials in a burst may vary from 2 to 50. These are involuntary discharges. They often sound like “marching soldiers”. Quantitation of myokymic discharges is rarely necessary. However, rapid quantitation can be used to identify the firing rate within a burst and between bursts of a discharge.

**EVOLUTION OF NEEDLE EMG FINDINGS IN ACUTE NEUROGENIC DISORDERS**

MU configuration is altered in disease states. In neurogenic disorders, the loss of MUs results in reduced recruitment, and the collateral sprouting of remaining axons produces long duration, high amplitude, and polyphasic MUAPs. In myopathic processes, loss of muscle fibers within MUs leads to reduction in amplitude and duration of MUAP. Abnormal MU variation is also seen in some neurogenic and myopathic processes, in addition to neuromuscular junction disorders. By using rapid quantitation to assess the multiple parameters of MUAPs, determination of the chronicity of a neurogenic process can be made.

<table>
<thead>
<tr>
<th>Time Interval</th>
<th>Spontaneous Potential</th>
<th>Recruitment Stability</th>
<th>Duration</th>
<th>Turns/Phases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Day 1 to 10</td>
<td>Normal</td>
<td>Reduced</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Day 10 to 15</td>
<td>Increased</td>
<td>Reduced</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Day 15 to 25</td>
<td>Fibrillations</td>
<td>Reduced</td>
<td>Unstable</td>
<td>Normal or mildly increased</td>
</tr>
<tr>
<td>Day 25 to 60</td>
<td>Fibrillations</td>
<td>Reduced</td>
<td>Stable</td>
<td>Polyphasic</td>
</tr>
<tr>
<td>Day 60 to 180</td>
<td>Normal or residual fibrillations</td>
<td>Reduced</td>
<td>Unstable or Stable</td>
<td>Increased</td>
</tr>
<tr>
<td>After Day 180</td>
<td>Normal</td>
<td>Reduced</td>
<td>Stable</td>
<td>Normal or residual polyphasia</td>
</tr>
</tbody>
</table>