You Make the Call !
Mastering EMG Waveform Recognition
Advanced

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Learning Objectives

After this session, you should be able to:

• Identify and understand the significance of uncommon spontaneous EMG waveforms
• Determine subtle changes in MUP recruitment and morphology changes in disease
**Firing Patterns of EMG Potentials**

*Regular*: linear change (fibrillation potential)

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*Regular*: no change (complex repetitive discharge)

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*Regular*: exponential change, *wax/wane* (myotonic)

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*Irregular*: (random change) (end plate spike)

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*Semi-Rhythmic*: (motor unit potential)

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*Bursts*: – Regular or semi-rhythmic (myokymic)
An unreported, distinctive type of increased insertional activity.

Wilbourn AJ.

**Abstract**
A previously unreported type of increased insertional activity, referred to as "Snap, Crackle, Pop" (SCP) because of its distinctive sound, is described. It consists of trains of potentials, of varying length, which follow normal insertional activity; the individual components of each train characteristically vary in amplitude, wave form, and interpotential interval. Earlier investigation showed that SCP is not artifact, and that it is associated with normal muscle biopsies. A retrospective search of 1,047 electromyographic studies was performed to yield 50 patients with SCP. Although the overall prevalence was 5%, SCP was seen 4 times more often in male than in female patients, and was most common in young, usually muscular, males. It was found more often in leg than arm muscles and was seen with the highest frequency in the medial gastrocnemius. It has no apparent clinical significance in itself, but SCP may be mistaken for more significant types of increased insertional activity.
• 16% of 49 pts with DM2 had minimal myotonic discharges with diffusely increased insertional activity (waning myotonic discharges)

• Increased insertional activity (brief runs of positive waves) may be seen in cases of mild myotonic dystrophy type 2

that can substantially limit the differential diagnosis of myopathy.\textsuperscript{7} MDs are classically defined as biphasic spikes or positive waves that fire repetitively at 20–80 Hz and wax and wane in amplitude and frequency. MDs may also be slow and difficult to distinguish from a waning positive wave.\textsuperscript{7,8} Waning MDs without a waxing component may be the only type of MD in patients with DM2.\textsuperscript{9}

Discovery of diffuse MDs may be an early clue to the diagnosis of a myotonic disorder; the presence of predominantly waning MDs may distinguish DM2 from DM1.\textsuperscript{9} However, the spectrum of myotonic disorders also includes rare patients without MDs, in whom a high clinical index of suspicion for DM2 and/or myotonic hyperkinesis is
Complex Repetitive Discharge
Ephaptic transmission from neighboring muscle fibers
Myokymic Discharges

Recurrent BURSTS (Regular or Irregular)
Doublets, triplets, multiplets
Firing rate within burst (5-150 Hz)*
# Myokymia

## Etiologies

<table>
<thead>
<tr>
<th>Focal</th>
<th>Radiation (cranial, plexus, single nerve)</th>
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<tbody>
<tr>
<td></td>
<td>Brainstem (multiple sclerosis, glioma, syringobulbia)</td>
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<tr>
<td></td>
<td>Mononeuropathy (CTS), radiculopathy (isolated muscles)</td>
</tr>
<tr>
<td></td>
<td>ALS (5% pts, more in cranial muscles)*</td>
</tr>
<tr>
<td><strong>Generalized</strong></td>
<td>Isaacs’ syndrome (VGKC)</td>
</tr>
<tr>
<td></td>
<td>Polyradiculopathy (AIDP, CIDP)</td>
</tr>
<tr>
<td></td>
<td>Episodic ataxia type 1 (VGKC (\alpha)-subunit gene KCNA1 mutation)</td>
</tr>
<tr>
<td></td>
<td>Rattlesnake envenomation</td>
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</table>

Neuromyotonic Discharges

- Spontaneous MUP
- Regular (wane)
- Very high frequency (200-300 Hz)
  - Continuous single MUP firing or
  - Discontinuous bursts

- Voltage-gated, potassium channel disorders
  - “Isaacs’ syndrome”, Morvan’s syndrome
27 yo healthy woman with 10 years of muscle “spasms”, cramps, and twitching in feet and lower legs.
Thoughts?
Temporal Course of MUP Changes
Acute Neurogenic Injury

- Normal
  - 1 minute: Reduced Recruitment
  - 1-2 months: Unstable Turns
  - 2-6 months: Polyphasic, Long duration
  - > 6 months: Long duration
Normal MUP Recruitment

Firing Rate (Hz)

Force

Mild

Moderate

MUP1

MUP2

MUP3

MUP4
Reduced Recruitment
MUP Firing Rate is TOO FAST

Assessing Recruitment

**Step 1**
Determine the maximum firing rate of any individual MUP?

**Step 2**
Determine how many different MUP are firing (sharp rise time)?

**Step 3**
Calculate the recruitment ratio
(max firing rate ÷ number of nearby MUP)

*Normal Recruitment Ratio < 5 (most limb muscles)*
(15 Hz: 3 MUP; 20 Hz: 4 MUP)

**Recruitment Frequency**
- Firing rate of 1st MUP when 2nd MUP begins to fire.
- NL < 11 Hz (exceptions: triceps, wrist/finger extensors, cranial muscles)
“Nascent” MUP

#1: Severe Loss of axons
- Markedly reduced # of MUP

#2: Minimal Collateral sprouting
- Loss of synchrony of fibers
- Few fibers in new MUP
- Low amplitude, short duration

Reduced recruitment
Short (or NL) Duration
Low amplitude
Polyphasic
Varying
Synkinesis
(Aberrant reinnervation – taking the wrong path)
Hemifacial Spasm

- Bursts (10 – 200 msec) of single or few MUP
- Variable interval between bursts (20 – 225 ms)
- High firing rate within burst (200-300 Hz)
Tetany EMG findings

- Doublets, triplets and multiplets
- Increase in frequency until a tonic spasm of flexor muscles ensue
CHARACTERISTICS AND SIGNIFICANCE OF DOUBLEDTS ON NEEDLE EMG

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Accepted 31 October 2016

ABSTRACT: Introduction: Voluntary doublets are electrophysiological phenomena thought to be associated with metabolic derangements or neuromuscular conditions. Methods: We prospectively studied 232 consecutive patients examined by a single examiner during routine electromyography (EMG) to determine the frequency of doublets in individual patients, specific muscles, neuromuscular conditions, electrolyte levels, and doublet characteristics. Results: Of 232 patients, 25 (10.7%) exhibited doublets. The mean age was 59 (52% men). Only 32 of 1,303 (2.5%) muscles exhibited doublets. Lower extremity and paraspinal groups represented 91% of muscles with doublets. Doublet frequency grouped by EMG diagnoses was: ALS (3 of 11; 27.1%), myopathy (3 of 10; 30.0%), axonal polyneuropathy (7 of 29; 24.1%), and no disease (7 of 109; 6.4%). There were no differences in serum electrolytes between doublet and matched subjects. Conclusions: Doublets occur in approximately 10% of patients, more commonly in lower extremity and paraspinal muscles, and are not correlated with a specific metabolic abnormality or neuromuscular condition.

Muscle Nerve 000:000–000, 2016

- Semi-rhythmic firing pattern (voluntary MUP)
- Similar spike morphology
- Slightly varying interspike interval, may be blocking

an identical MUP firing twice with a varying interspike interval and sometimes with blocking of the second spike. The digital EMG buffer of each doublet was saved and stored for off-line review.

Medical records were reviewed from all patients
Decreased Insertional Activity

- End stage muscle (neurogenic or myopathic)
- Ischemia
- Inexcitable muscle during channelopathy attack
- Technical
Hyperkalemic Periodic Paralysis

- Myotonic discharges
  - May disappear during attack
- NO fibrillation potentials, but appear with cooling
- Normal MUPs or mildly short duration MUPs – During attack reduced recruitment
- NCS – CMAPs normal between attacks (low during attack)
Slow Channel Congenital Myasthenic Syndrome

- **Autosomal dominant**
- **Due to a kinetic defect in the AchR**
  - Prolongs the endplate potential = repetitive CMAP
  - May cause spontaneous channel openings
  - With repetitive stimulation, successive prolonged EPPs progressively depolarize the membrane, producing depolarization block
  - $\text{Ca}^{2+}$ may accumulate in post-synaptic region, causing end plate myopathy
- **May present in adulthood**
- **Fluctuating, fatigable ptosis, ophthalmoparesis**
- **Neck, wrist and finger extensor weakness most prominent**

- **Repetitive CMAP 3-6 msec after M-wave, Decrements more**
- **No response to AchE inhibitors**

![Graph showing repetitive CMAP and the effects of stimulator settings]
Distinction between neoplastic and radiation-induced brachial plexopathy, with emphasis on the role of EMG

C. Michel Harper, Jr., MD; Juergen E. Thomas, MD; Terrence L. Cascino, MD; and William J. Litchy, MD

Table 6. Needle examination

<table>
<thead>
<tr>
<th>Type of plexopathy</th>
<th>Radiation-induced</th>
<th>Neoplastic</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. (%)</td>
<td>No. (%)</td>
<td></td>
</tr>
<tr>
<td>Muscles examined</td>
<td>315</td>
<td>534</td>
</tr>
<tr>
<td>Muscles examined per patient</td>
<td>9.0</td>
<td>9.7</td>
</tr>
<tr>
<td>Patients with</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fibrillation potentials</td>
<td>28 (80)</td>
<td>46 (85)</td>
</tr>
<tr>
<td>Paraspinal fibrillation potentials</td>
<td>7 (23)*</td>
<td>1 (2)†</td>
</tr>
<tr>
<td>Fasciculation potentials</td>
<td>11 (31)</td>
<td>8 (15)</td>
</tr>
<tr>
<td>MUP changes‡</td>
<td>35 (100)</td>
<td>55 (100)</td>
</tr>
<tr>
<td>Myokymic discharges§</td>
<td>22 (63)</td>
<td>2 (4)</td>
</tr>
</tbody>
</table>

Muscles with

| Fibrillation potentials             | 99 (31)           | 186 (35)   |
| Fasciculation potentials            | 26 (8)            | 20 (4)     |
| MUP changes‡                        | 219 (70)          | 281 (53)   |
| Myokymic discharges                 | 47 (15)           | 2 (0.4)    |

MUP Motor unit potential.

* Examined in 30 of 35 patients.
† Examined in 50 of 55 patients.
‡ Large amplitude, long duration, polyphasic, poorly recruited MUPs.
§ In patients with myokymia, 47 of 194 muscles examined (24%) had myokymic discharges.

Electrophysiologic studies are retrospectively reviewed for 55 patients with brachial plexopathy. The presence or absence of pain as the presenting mass on CT of the plexus, and presence of myokymic discharges on EMG are the cause of the brachial plexopathy. The distribution of weakness helps in distinguishing neoplastic from radiation-induced brachial plexopathy.

Table 7. Distribution of myokymic discharges*

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Number examined</th>
<th>Myokymic discharges No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pronator teres</td>
<td>34</td>
<td>13 (38)</td>
</tr>
<tr>
<td>Abductor pollicis brevis</td>
<td>24</td>
<td>7 (29)</td>
</tr>
<tr>
<td>Flexor pollicis longus</td>
<td>10</td>
<td>2 (20)</td>
</tr>
<tr>
<td>Extensor indicis proprius</td>
<td>10</td>
<td>2 (20)</td>
</tr>
<tr>
<td>Triceps</td>
<td>29</td>
<td>4 (14)</td>
</tr>
<tr>
<td>Biceps</td>
<td>33</td>
<td>4 (12)</td>
</tr>
<tr>
<td>Deltoide</td>
<td>27</td>
<td>3 (11)</td>
</tr>
<tr>
<td>First dorsal interosseous</td>
<td>34</td>
<td>3 (9)</td>
</tr>
<tr>
<td>Paraspinal</td>
<td>42</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Infraspinatus</td>
<td>26</td>
<td>1 (4)</td>
</tr>
<tr>
<td>Extensor carpi radialis</td>
<td>10</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>

* In muscles examined 10 or more times.
Iterative Discharge ("repetitive discharge")

“General term for the recurrence of an action potential with the same or nearly identical form. May refer to recurring potentials recorded in muscle at rest, during voluntary contraction, or in response to a single nerve stimulus.

See double discharge, triple discharge, multiple discharge, myokymic discharge, complex repetitive discharge, neuromyotonic discharge, and cramp discharge.”

AANEM Glossary of Terms, 2001
SHORT REPORT

MOTOR UNIT POTENTIAL INDUCED REPETITIVE DISCHARGES (MIRDS): DESCRIPTION OF AN UNUSUAL ITERATIVE DISCHARGE

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ABSTRACT: Introduction: Repetitive discharges may be recorded during nerve conduction studies (NCS) or during needle electromyography in a muscle at rest. Repetitive discharges that occur during voluntary activation and are time-locked to voluntary motor unit potentials (MUP) have not been described. Methods: Retrospective review of motor unit potential induced repetitive discharges (MIRDS) identified in the EMG laboratory. Characteristics of each MIRD, patient demographics, other EMG findings in the same muscle, and electrophysiological diagnosis were analyzed. Results: MIRDs were observed in 15 patients. The morphology and number of spikes and duration of MIRDs varied. The discharges fired at rates of 50–200 Hz. All but 2 patients had EMG findings of a chronic neurogenic disorder. Conclusions: MIRDs are rare iterative discharges time-locked to a voluntary MUP. The pathophysiology of MIRD is unclear, but their presence may indicate a chronic neurogenic process.


FIGURE 1. (A,B) Examples of MIRDs with a group of 2–4 different time-locked spikes in the RD. (C) Example of MIRD with a single spike firing recurrently in the RD. Asterisks (*) denote the triggering MUP.
Characteristics of Cramp Discharges

- **Rapid firing** (up to 40 Hz) (similar appearing to reduced recruitment)
- Avg discharge rate 14.5 Hz (6 – 25 Hz)*
- **Irregular** firing (higher interspike interval variability)*
- “Sputters” out

Juvenile Distal Segmental Muscular Atrophy (Hirayama disease)

- Focal motor neuron disease restricted to limited number of myotomes (C7-T1)
- Upper limb involvement is most frequent
  - Forearm and hand wasting
- Onset in 2nd or 3rd decade
- Male predominance (>80%)
- EMG: chronic neurogenic changes in C7-T1 distribution