**Spontaneous activity in EMG**

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**Spontaneous activity**

- Activity present in the muscle at rest
- Spontaneous activity may present constantly
- Spontaneous activity may be provoked by movement of the electrode
- Some types of activity is normal
- Some types are abnormal

**Normal spontaneous activity**

- Insertional activity
- End-plate spikes
- End-plate noise

**Insertional activity**

- Burst of muscle fiber action potentials provoked by the movement of the needle electrode
- Duration <100 ms
- Not a reliable parameter for abnormality
**Insertional activity**

- Decreased
  - Muscle necrosis
- Increased
  - Neuropathy often before fibrillation potentials appear
  - Myopathy
  - Some healthy muscles
    - Calf muscles
    - Thenar muscles

**End-plate activity**

**End-plate activity monophasic**

**Miniature end-plate potentials**

**End-plate activity - monophasic 1**

- Spontaneous electric activity recorded with a needle electrode close to muscle end-plates
- Low-amplitude (10-20 μV), short-duration (0.5-1 ms), monophasic (negative) potentials that occur in a dense, steady pattern and are restricted to a localized area of the muscle
- Because of the multitude of different potentials occurring, the exact frequency, although appearing to be high, cannot be defined

**End-plate activity - monophasic 2**

- These non-propagated potentials are probably miniature end-plate potentials recorded extracellularly
- This form of end-plate activity has been referred to as end-plate noise or seashell sound
**End-plate activity biphasic**

- Moderate-amplitude (100-300 μV), short-duration (2-4 ms), biphasic (negative-positive) spike potentials that occur irregularly in short bursts with a high frequency (50-100 Hz), restricted to a localized area within the muscle.
- These propagated potentials are generated by muscle fibers excited by activity in nerve terminals.
- These potentials have been referred to as *biphasic spike potentials, end-plate spikes*, and, incorrectly, *nerve potentials*.

**Abnormal spontaneous activity**

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<th>Muscle fibers</th>
<th>Motor units</th>
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**Fasciculation potentials**

**Recording fasciculation potentials**
Fasciculation potentials

- Random spontaneous twitching of a group of muscle fibers or motor unit. This twitch may produce movement of the overlying skin, mucous membrane or digits.
- The electric activity is called fasciculation potentials.

Generation of fasciculation potentials

- Abnormal spontaneous action potentials generated in the motor neuron.
- Fasciculation potentials are generated both centrally and peripherally in the motoneuron.
  - Initial axon hillock of the axon.
  - Local anesthesia does not block fasciculation in ALS.
- Muscle has been implicated in benign fasciculation.
- Also the upper motor neuron has been suggested.

Fasciculation potentials - significance

- Normal in distal intrinsic foot muscles, any age.
- Benign fasciculation:
  - May be short lasting.
  - Sometimes permanent.
- Neurogenic disorders:
  - Usually chronic or inactive.
  - Motor neuron disease.

Myokymia

- Myokymic discharges

- Myokymic discharges
<table>
<thead>
<tr>
<th>Myokymia - definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Motor unit action potentials that fire repetitively and may be associated with clinical myokymia.</td>
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<tr>
<td>- Two firing patterns have been described</td>
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<tr>
<td>- Brief repetitive firing of single units for a brief period (up to a few seconds) at a uniform rate (2-60 Hz) followed by a brief period of silence (up to a few seconds)</td>
</tr>
<tr>
<td>- Less commonly uniform firing rate (1-5 Hz)</td>
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<tr>
<td>- Clinically undulating spontaneous movements or contractions of the muscle</td>
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<td>- Probably generated in the motor axon</td>
</tr>
<tr>
<td>- Blocked by curare</td>
</tr>
<tr>
<td>- Spinal anesthesia has no effect</td>
</tr>
<tr>
<td>- Demyelination seems to be important</td>
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<tr>
<td>- Focal myokymia</td>
</tr>
<tr>
<td>- Brachial plexus lesions following radiation therapy</td>
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<tr>
<td>- Facial myokymia</td>
</tr>
<tr>
<td>- MS</td>
</tr>
<tr>
<td>- Pontine glioma</td>
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<tr>
<td>- GBS, ALS, trigeminal neuralgia</td>
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<tr>
<td>- Generalized myokymia (=neuromyotonia??)</td>
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<tr>
<td>- Idiopathic or hereditary form</td>
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<td>- GBS, metabolic disorders</td>
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<td>- Bursts of motor unit action potentials with originate in the motor axons firing at high rates (150-300 Hz) for a few seconds, and which often start and stop abruptly.</td>
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<tr>
<td>- The amplitude of the response typically wanes.</td>
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<tr>
<td>- Discharges may occur spontaneously or be initiated by needle movement, voluntary effort and ischemia or percussion of a nerve.</td>
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<tr>
<td>- These discharges should be distinguished from myotonic discharges and complex repetitive discharges</td>
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<td>- Clinical syndrome of continuous muscle fiber activity manifested as continuous muscle rippling and stiffness</td>
</tr>
<tr>
<td>- The accompanying electric activity may be intermittent or continuous</td>
</tr>
<tr>
<td>- Terms used to describe related clinical syndromes are continuous muscle fiber activity, Isaac syndrome, Isaac-Merton syndrome</td>
</tr>
</tbody>
</table>
Neuromyotonia Isacs syndrome

- Antibodies against K⁺ channels
- May be a paraneoplastic phenomenon
- Generated in the axons
- Respond to phenytoin or carbamazepine

Fibrillation potentials and positive sharp waves

Effects of denervation on muscle fibers

- Sensitivity to acetylcholine increases x 100
- Decreased resting membrane potential
- New sodium channels develop after denervation
- Increased sodium conductance
- Require usually 2-4 weeks to develop, may be seen after 8-10 days

Effects of denervation on muscle fibers

- Muscles close to local nerve lesion are first to show fibrillation potentials
- Steroids and cytostatic drugs suppress fibrillation potentials
- α - bungarotoxin and ischaemia suppress fibrillation potentials, acetylcholine receptors play a role
- Acetylcholine receptor hypersensitivity is not the sole cause

Fibrillation potential

- The electric activity associated with a spontaneously contracting (fibrillating) muscle fiber
- Action potential of a single muscle fiber
- The action potentials may occur spontaneously or after movement of the needle electrode
- The potentials fire at a constant rate
  - A small proportion fire irregularly
- Potentials are biphasic spikes of short duration (<5 ms) with an initial positive phase and a peak-to-peak amplitude of less than 1 mV
Fibrillation potential

† Firing rate has a wide range (1-50 Hz) and often decreases just before cessation of an individual discharge.

† A high-pitched regular sound is associated with the discharge of fibrillation potentials and has been described in the old literature as "rain on a tin roof”

Fibrillation potentials - 2

Positive sharp waves 1

† A biphasic, positive-negative potential
† Initiated by needle movement
† Recurring in a uniform, regular pattern at a rate of 1-50 Hz; the discharge frequency may decrease slightly just before cessation of discharge
† The initial positive deflection is rapid (<1 ms), its duration is usually less than 5 ms, and the amplitude is up to 1 mV
† The negative phase is of low amplitude, with a duration of 10-100 ms.

Positive sharp wave

Positive sharp waves 2

† Positive sharp waves can be recorded from the damaged area of fibrillating muscle fibers.
† The positive sharp waveform is not specific for muscle fiber damage
† Its configuration may result from the position of the needle electrode. The electrode triggers an action potential propagating away from the electrode

Positive sharp wave
Denervation activity

- This term has been used to describe fibrillation potentials and positive sharp waves.
- The use of this term is discouraged because fibrillation potentials may occur in myopathies.

Fibrillation potentials - clinical significance

- Occur rarely in healthy muscle
  - One out of 20 insertions
- Neuropathic disorders
  - Acute or subacute
  - If initial lesion was severe, also in inactive
- Myopathic disorders
  - Active myopathies
- CNS
  - Following stroke or CNS trauma

Quantification: Uppsala

- Number of insertions/10 with fibrillation potentials or positive sharp waves
  - Does not take into account the number of fibrillation potentials at each insertion
  - Accurate and reproducible in mild cases (2-5/10)
  - Not so reproducible at levels 6-9/10

Quantification: Mayo clinic

<table>
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<th>Grading</th>
<th>Characteristics</th>
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<tr>
<td>0</td>
<td>No fibrillation potentials</td>
</tr>
<tr>
<td>1+</td>
<td>Single trains in at least two sites</td>
</tr>
<tr>
<td>2+</td>
<td>Moderate number in at least three or more muscle areas</td>
</tr>
<tr>
<td>3+</td>
<td>Many in all muscle regions</td>
</tr>
<tr>
<td>4+</td>
<td>Baseline obliterated with fibrillation potentials</td>
</tr>
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Complex repetitive discharges

Complex repetitive discharges
Complex repetitive discharges

- Polyphasic or serrated action potentials that may start spontaneously or after a needle movement.
- Uniform frequency, shape, and amplitude, with abrupt onset, cessation, or change in shape.
- Amplitude ranges from 100 μV to 1 mV and frequency of discharge from 5 to 100 Hz.
- Bizarre high frequency discharge, bizarre repetitive discharge, bizarre repetitive potential, near constant frequency trains, pseudomyotonic discharge and not recommended.

Significance of CRD

- Rarely observed in healthy subjects.
- Myopathies:
  - Polymyositis
  - Muscle dystrophies
- Neuropathies:
  - May be seen in most neuropathies
  - Chronic
- CRD is a non-specific abnormality.

Myotonic discharges

- Repetitive discharge at rates of 20 to 80 Hz are of two different types:
  - Biphasic (positive negative) spike potentials less than 5 ms in duration resembling fibrillation potentials
  - Positive waves of 5 to 20 ms duration resembling positive sharp waves.
Myotonic discharges
- Both potential forms are recorded after needle insertion, after voluntary muscle contraction or after muscle percussion, and are due to independent, repetitive discharges of single muscle fibers
- The amplitude and frequency of the potentials must both wax and wane to be identified as myotonic discharges
- This change produces a characteristic musical sound due to corresponding change in pitch, which has been likened to the sound of a “starting motor cycle”

Myotonic disorders
- Progressive myopathy and myotonia
  - Myotonic dystrophy type 1
  - Myotonic dystrophy type 2 (Proximal myotonic myopathy)
- Main symptom myotonia
  - Myotonia congenita (Thomsen and Becker)
  - Myotonia fluctuans
- Other myotonias
  - Chondrodystrophic myotonia
  - Paramyotonia congenita
  - Paraneoplastic myotonia
- Periodic paralysis
  - Hyperkalemic periodic paralysis

Myotonic dystrophy - Pathophysiology
- Patients with mainly myotonia tend to have normal resting membrane potential
- Patients with severe dystrophy have reduced resting membrane potential
- Cl- conductance varies from low to low normal
- K- conductance normal
- Patch clamp studies have shown abnormal inactivation of Na+ channels
- In myotonic muscular dystrophy, abnormal muscle Na+ currents underlie myotonic discharges (Mounsey et al 1995).

Na+ channel function

Myotonic dystrophy - Pathophysiology
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Further reading

Channelopathies
- Chloride channel
  - Myotonia congenita (Thomsen)
  - Myotonia congenita (Becker)
  - Myotonia levi
- Calcium channel
  - Hypokalemic periodic paralysis
  - Myasthenic syndrome
- Sodium channel
  - Myotonia fluctuans
  - Paramyotonia congenita
  - Hyperkalemic periodic paralysis
  - Myotonia permanens
  - Acetazolamide responsive myotonia
- Potassium channels
  - Neuromyotonia
Game over