Spontaneous activity in EMG

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Goals

- Generation of spontaneous activity in needle EMG
- Able to interpret clinical significance of the findings

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Outline 45 min

- Anatomy and physiology of the motor unit
- Spontaneous activity in muscle
  - Normal and abnormal
- Generated by the motor units
  - Fasciculation
  - Myokymia and neuromyotonia
- Generated by muscle fibres
  - Fibrillation potentials
  - Complex repetitive discharges
  - Myotonia

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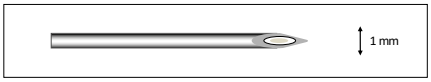
Concentric EMG electrode



- Introduced by Adrian and Bronk 1929
- Electrode 150 x 580 um elliptical shape
- Area of recording surface 0.07 mm^2

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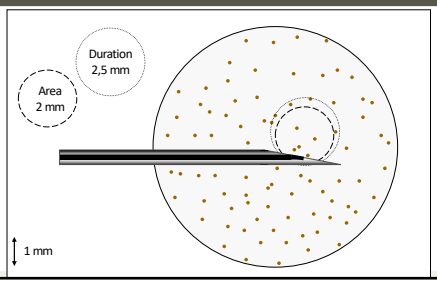
Concentric EMG electrode – uptake area



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Needle EMG procedure

- Spontaneous activity at rest
  - Insertional activity
  - Different kinds of spontaneous activity
- Recruitment at slight and strong contraction
  - Turns and amplitude analysis
- Motor unit potential analysis
  - Qualitative
  - Quantitative

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Insertional activity

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Moving the needle smoothly

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EMG procedure

- Insertional activity
  - Short, gentle needle movements in one direction
  - Retract needle to fascia and move in another direction in the same plane
  - 20-25 insertions

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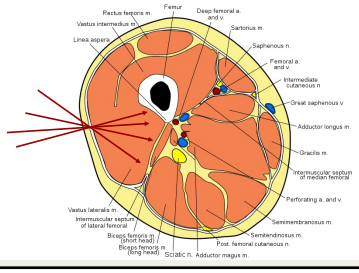

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Spontaneous activity

- *Activity in the muscle at rest*
- Some present constantly
- Some provoked by movement of electrode
  - Not truly spontaneous!
- Some types are normal
- Some types are abnormal

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Normal spontaneous activity

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

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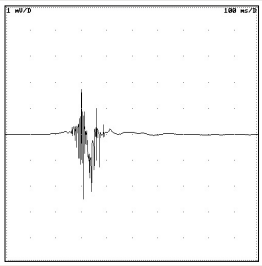
Insertional activity



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Insertional activity

- Muscle fiber action potentials provoked by the needle
- Duration <100 ms
- **Increased insertional activity not reliable parameter**
  - **Too subjective**

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Insertional activity

- Decreased
  - Muscle necrosis – no insertional activity
- Increased
  - In neuropathies before fibrillation potentials appear
  - Myopathy
  - Some muscles are quite lively
    - Calf muscles
    - Thenar muscles

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End-plate activity

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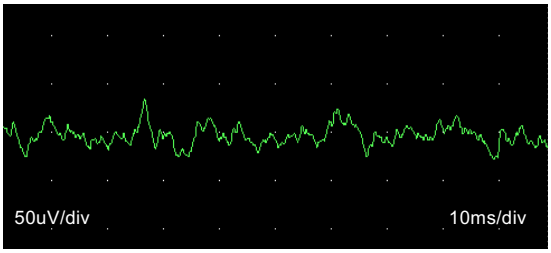
End-plate activity - monophasic



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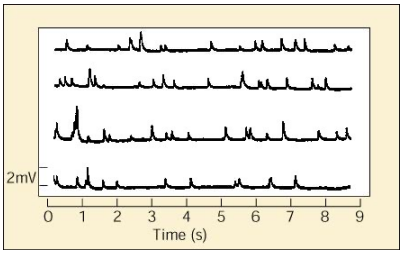
Miniature end-plate potentials



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End-plate activity - monophasic 1

- Spontaneous electric activity recorded at end-plates
- Low-amplitude (10-20 μV)
- Short-duration (0.5-1 ms)
- Monophasic (negative) potentials
- Dense, steady pattern
- Restricted to a localized area
- Exact frequency cannot be defined

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End-plate activity - monophasic 2

- Non-propagated potentials
- Miniature end-plate potentials recorded extracellularly
- *End-plate noise*
- *Seashell sound*

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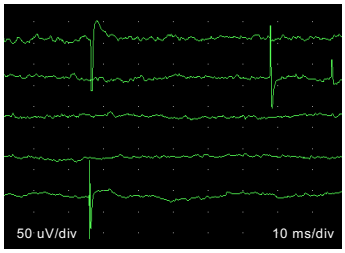
End-plate activity biphasic



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End-plate activity - biphasic

- Amplitude 100-300 μV
- Duration 2-4 ms
- Biphasic (negative-positive) spike potentials
- Occur irregularly in short bursts, high frequency (50-100 Hz)
- Restricted to a localized area within the muscle.
- Potentials generated by muscle fibers
- *Biphasic spike potentials, end-plate spikes*

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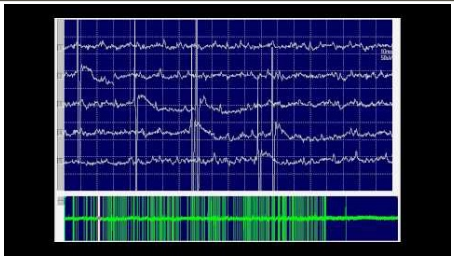
End-plate noise and end-plate spikes



https://www.youtube.com/watch?v=R9wMgcdVxGI

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Abnormal spontaneous activity

Motor units

- Fasciculation potentials
- Neuromyotonia
- Myokymia
- Cramps

Muscle fibers

- Fibrillation potentials
- Positive sharp waves
- Complex repetitive discharges
- Myotonic discharges

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Fasciculation potentials

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Fasciculation potentials (FP)

- Fasciculation = random spontaneous twitching of a motor unit
- Twitch may be seen under the skin
- Electric activity of fasciculation called **fasciculation potentials**

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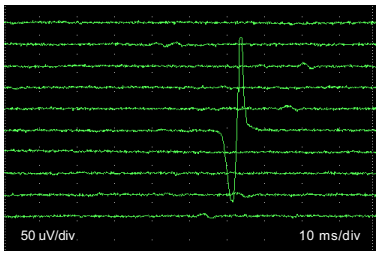
Fasciculation potentials



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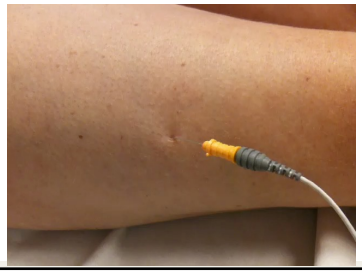
Recording fasciculation potentials



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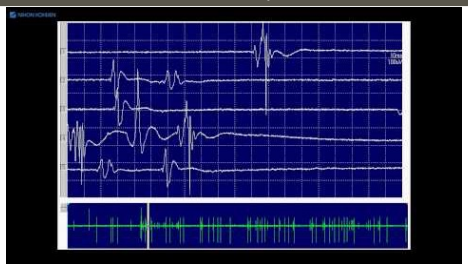
Fasciculation potentials



https://www.youtube.com/watch?v=6-3PP_S-Q8I&t=18s

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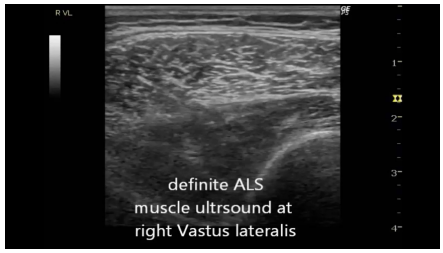
Fasciculation - ultrasound



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Generation of fasciculation potentials

- FPs generated in the motor neuron both centrally and peripherally
  - Initial axon hillock of lower motor neuron
  - Along the axon
    - Local anesthesia will not block all FPs in ALS
- May be generated by the upper motor neuron in ALS
- Benign FPs arise distally in the muscle

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Fasciculation potentials - significance

- Occur often in healthy subjects
  - Especially in intrinsic foot muscles
- Benign fasciculation
  - May be transient
  - Often permanent
- Neurogenic disorders
  - Chronic or inactive neuropathies (focal or polyneuropathies)
  - Motor neuron disease

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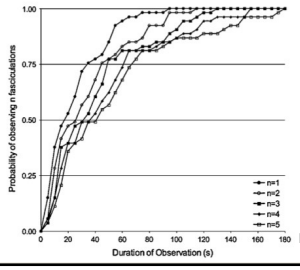
Detection time of fasciculation potentials in ALS



Kerry Mills JNNP 2011

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Fasciculation potentials in ALS

- Record activity for 90 sec
- ALS patients are often not aware of FPs
- Motor neuron excitability
  - Increased persistent sodium conductance
  - Reduced potassium conductance
  - Axonal hyperexcitability

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Sequence of EMG abnormalities in ALS

- Fasciculation potentials
  - Initially simple and stable
- Unstable MUPs with "jiggle"
- Fibrillation potentials
- Collateral reinnervation with large, complex MUPs
- In weak muscles
  - Fasciculation potentials large, >5 phases and unstable
  - Double discharges of fasciculation potentials

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Benign fasciculation

- FPs are simple and stable
- No doublet FPs
- No fibrillation potentials
- Normal MUPs

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Myokymia and neuromyotonia

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Definitions

Myokymia is undulating rippling of muscles

- Resemble worms crawling beneath the skin
- Usually focal in one part of the body
- May be generalized

Neuromyotonia is persistent muscle contraction

- Severe enough to cause deformity of hands
- Often facilitated by muscle contraction
- Usually generalized

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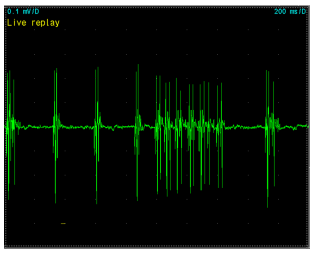
Myokymic discharges



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Myokymic discharges

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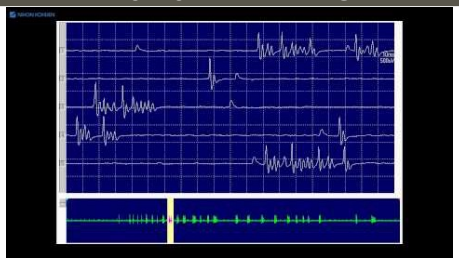
Myokymic discharges



https://www.youtube.com/watch?v=SXhmV3uYcT0

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Myokymic discharges

- Motor unit action potentials that fire repetitively and may be associated with clinical myokymia.
- Burst of single motor units at 5 - 150 Hz followed by a brief period of silence (up to a few seconds), the bursts are repeated at regular intervals
- Longer bursts followed by longer intervals
- Usually spontaneous, may be triggered by exercise
- Less commonly uniform firing rate (1-5 Hz)
- Clinically undulating spontaneous movements or contractions

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Myokymic discharges

- Generated in the motor axon
- Blocked by curare
- Spinal anesthesia has no effect
- Demyelination seems to be important

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Myokymic discharges

- Focal myokymia
  - Brachial plexopathy following radiation therapy
  - Facial myokymia
    - MS, pontine glioma, GBS, ALS, trigeminal neuralgia
- Generalized myokymia (= neuromyotonia)
  - Idiopathic or hereditary form
  - GBS, metabolic disorders

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Neuromyotonic discharges

- Bursts of *motor unit action potentials* at high rates (150-300 Hz) for a few seconds
- Often start and stop abruptly
- The amplitude of the potentials typically varies.
- Discharges may occur spontaneously or be initiated by needle movement, voluntary effort and ischemia or percussion of a nerve.
- Generated in the motor axon
- Continue during sleep
- Not blocked by local nerve blocks

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Neuromyotonia

- Muscle fiber activity manifested as continuous muscle stiffness
- The accompanying electric activity continuous
- Terms used to describe related clinical syndromes
  - Isaac's syndrome, Isaac-Merton syndrome

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Differences myokymia/neuromyotonia?

- Myokymia/neuromyotonia related with increased axon excitability
- Voltage gated potassium channels abnormal
  - Acquired (Isaac' s syndrome)
  - Genetic (Episodic ataxia with myokymia)
- Probably not meaningful to differentiate the two phenomena?

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Neuromyotonia Isacs syndrome

- Antibodies against K^+ channels
- May be a paraneoplastic phenomenon
- Generated in the axons
- Respond to Na^+ channel blockers
  - Phenytoin or carbamazapine

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Fibrillation potentials (FP)

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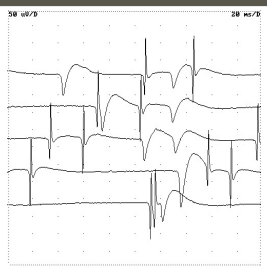
Fibrillation potentials and positive sharp waves



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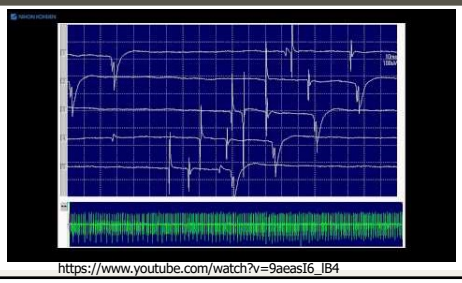
Fibrillation potentials and positive sharp waves



https://www.youtube.com/watch?v=9aeasI6_IB4

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Effects of denervation on muscle

- Sensitivity to acetylcholine increases x 100
- Decreased resting membrane potential
- New sodium channels develop after denervation
- Increased sodium conductance
- Acetylcholine receptor hypersensitivity not only cause of fibs
- Fibs require 2-4 weeks to develop, may occur after 8-10 days

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Effects of denervation on muscle fibers

- Muscles close to neuropathic lesion first to show FPs
- Steroids and cytostatic drugs suppress FPs
- Alpha - bungarotoxin and ischemia suppress FPs

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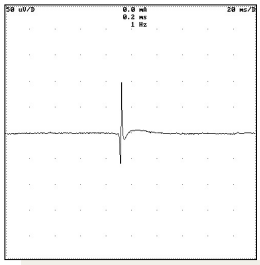
Fibrillation potential



- Biphasic spikes
  - Short duration (<5 ms)
  - Initial positive phase
  - Amplitude < 1 mV
- Single muscle fiber action potentials
- Occur spontaneously
- Triggered by the electrode
- Usually fire at a constant rate
  - A small proportion fire irregularly

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Fibrillation potentials

- Firing rate has a wide range (1-50 Hz)
- Often rate decreases just before cessation
- 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”

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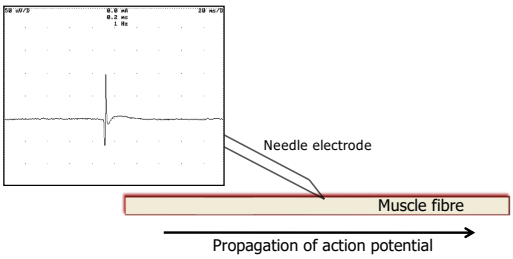
Fibrillation potential



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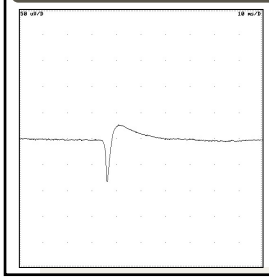
Positive sharp wave



- Biphasic spikes
  - Short duration (<5 ms)
  - Initial positive phase
  - Amplitude < 1 mV
- Single muscle fiber potential
- Usually triggered by the electrode
- FPs usually fire at a constant rate
  - A small proportion fire irregularly

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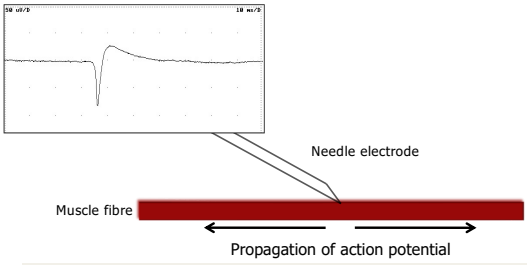
Positive sharp wave



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Fibrillation potentials – Positive sharp waves

- Lump together FPs and PSs
- Have identical significance
- No reason for separation
- Call them FPs
- PSs may be seen sooner after axonal nerve injury

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Timing of fibrillation potentials

- FIBs to develop 3-4 weeks after nerve injury
- In severe injuries fibs may be seen after 1-2 weeks
- Muscles close to injury show FIBs earlier

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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 occur in myopathies !!!!!

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Fibrillation potentials - clinical significance

- Sometimes in healthy muscle
  - < 1 out of 20 insertions
  - Muscle tissue is not static – constant regeneration
  - Distal foot muscles 25-30% of healthy adults
- Neuropathy
  - Acute or subacute
  - If initial lesion was severe, also in inactive disorder
- Myopathy
  - Actively progressing myopathies
- CNS lesions
  - Following stroke or CNS trauma

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Fibs in healthy paraspinal neck muscles

- <40 years 0%
- 40-60 8%
- >60 90%

J Clin Neurophys 2006:23:573-

ORIGINAL ARTICLES

Cervical Paraspinal Electromyography: Normal Values in 100 Control Subjects

R. Gilad, R. Dabby, M. Boaz, and M. Sadeh

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Fibrillation potentials – amplitude

- The amplitude is dependent on
  - Diameter of fibrillating muscle fiber
  - Old denervated muscle fibers are atrophic - small FP amplitude
- Very small FPs indicate an old lesion.

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Quantification: Uppsala and Turku

- Number of insertions with FIBs/10 insertions
  - Does not consider number of fibs at each insertion
  - Accurate and reproducible in mild cases (2-5/10)
  - Not accurately reproducible at levels 6-9/10

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Quantification: Mayo clinic

Grading	Characteristics
0	No fibrillation potentials
1+	Single trains in at least two sites
2+	Moderate number in at least 3 sites
3+	Many in all muscle regions
4+	Baseline obliterated with FPs

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Complex repetitive discharges
CRD

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Complex repetitive discharges

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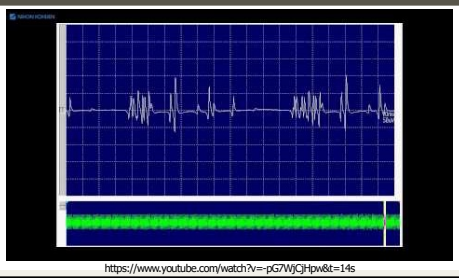
Complex repetitive discharges



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Complex repetitive discharges

- Polyphasic action potentials
- Start spontaneously or after a needle movement
- Abrupt onset and cessation
- Uniform frequency, shape
- Amplitude 100-1000 µV
- Frequency 5-100 Hz

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CRD - not recommended terms

- Pseudomyotonic discharge
- Bizarre high frequency discharge
- Bizarre repetitive discharge
- Bizarre repetitive potential
- Near constant frequency trains

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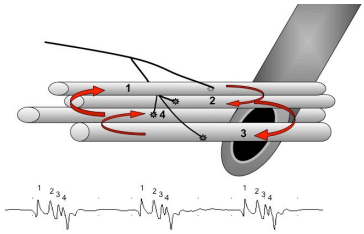
Generation of complex repetitive discharges



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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

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Myotonic discharges

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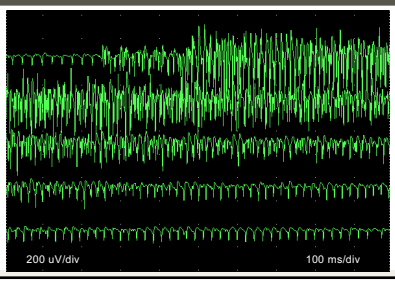
Myotonic discharges



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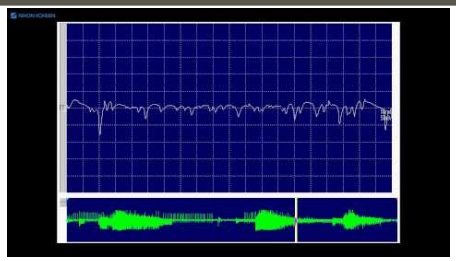
Myotonic discharges



https://www.youtube.com/watch?v=vh2e_Y4lxX8

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Myotonic discharges

- Repetitive discharge
  - 20 to 80 Hz
  - ***Waxing and waning amplitude and frequency***
  - FPs
  - PSs

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Myotonic discharges

- Repetitive discharges of single muscle fibers
- Recorded after
  - Needle insertion
  - Voluntary muscle contraction
- Amplitude and frequency ***must both wax and wane***
- Sounds like “starting a motorcycle”

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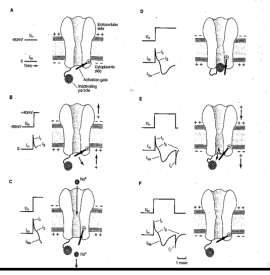
Na+ channel function



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Myotonic disorders

- Progressive myopathy and myotonia
  - Myotonic dystrophy type 1 and type 2
- Main symptom myotonia
  - Myotonia congenita, Myotonia fluctuans
- Other myotonias
  - Paramyotonia congenita
  - Paraneoplastic myotonia
- Periodic paralysis
  - Hyperkalemic periodic paralysis

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Spontaneous joy of skiing

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