EMG in MYOPATHY

Which information do we get from EMG today?

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1. Background to typical “myopathic” EMG findings
2. Situations where EMG gives “unique” information

Electrodes

Conc EMG signals from 2-15 muscle fibres

CNEMG

- At rest: denervation and spec spontaneous activity (myotonia, CRD, neuromyotonia)
- MUP: number of fibres in recorded area
  - fibre diameters
  - n-m transmission
- IP: recruitment pattern
  - total number of MUs at full effort
Spontaneous activity from the muscle

FINDING
- fibrillation potentials, psw
- myotonic discharges
- CRD
- myokymic discharges
- myogenic extra discharges

QUANTIFY AS
- #/10 recording sites
- or +, ++, ++++, ++++
- few
- moderate
- abundant
- or spontaneous or after provocation

Spontaneous activity from the nerve

FINDING
- neuromyotonic discharges
- myokymic discharges
- muscle cramps
- fasciculations
- neurogenic extra discharges

QUANTIFY AS
- #/10 recording sites
- or +, ++, ++++, ++++
- Few (per time unit)
- moderate
- abundant
- or spontaneous or after provocation

Spontaneous activity generated in the muscle fibre (Stålberg, Daube 2003)

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

Myotonic discharge

Myotonia; warm up after 1 minute of activity
Myopathy, Stålberg

Myotonia; warm up after 1 minute of activity

CNEMG

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MUP, normal TA

MUP, myopathy TA

The EMG simulator
Emery-Dreifuss muscular dystrophy, X-linked type 1 (EDMD; emerinopathy)

Mild to moderate dystrophic changes:
- Fiber size variation, a few necrotic fibers, central nuclei,
- Increase of fibrous connective tissue and fat between myofibers.

CNEMG
- At rest: denervation and spec spontaneous activity (myotonia, CRD, neuromyotonia)
- MUP: number of fibres in recorded area, fibre diameters, n-m transmission
- IP: recruitment pattern, total number of MUs at full effort

EMG - interference pattern

Myopathy

Normal

Neuropathy

Interference pattern analysis in normal, neuropathic and myopathic conditions

Beyond conventional EMG
Hereditary distal myopathy (CN rec)

Muscle fibres in myopathy

Recording from 2 or more still synchronous APs from branches of a split muscle fibre may produce high ampl

Split muscle fibers

Scanning EMG

Scan in a normal muscle (simulation)

Scan in a myopathic muscle (simulation)
EMG in general diagnostic workup in neuromuscular conditions

- Gives a quick multidimensional information about the condition
- Myopathy- nmj- neuropathy
- Spontaneous activity
- Distribution, severity
- Biopsy guidance

Ultrasound, CT, MRI, genetics important complements

Sensitivity/specificity of EMG in Myopathies

- **Sensitivity** (abnormal vs normal):
  - depends on type of myopathy:
    - Duchenne, myositis........ 90-99%
    - Metabolic myopathy ........may be very low

- **Specificity** (classification):
  - EMG usually not specific in separating subgroups
 EMG combined with other findings gives a clue

- Myopathy + Neuropathy;
  - think of mitochondrial dysfunction,
  - malignancy

- Normal EMG in clinical myopathy;
  - think of metabolic myopathy

- Performance/EMG discrepancy;
  - Weakness + full EMG pattern myopathy
  - Weakness + normal EMG central

 EMG may explain pathophysiological mechanisms

- CRD –
- Myotonic weakness –
- Hypokalemic paralysis –
- Painful myot (G1306A)
- Biopsy: fiber type preponderance;
  - in cong. myop. - grouping?
  - in hypothyreosis -

SFEMG suggests ephaptic activation decrement of single muscle fiber aps
- muscle fiber cond block (Zwartz)
- giant paw due to ephases in fiber groups
- longitud. cond block, channelopathy

- Biopsy: fiber type preponderance;
  - in cong. myop. - grouping?
  - in hypothyreosis -

normal FD excludes reinnervation
- normal EMG = fiber type transformation

 EMG gives "unique" information

| Fatigue: | MG |
| Muscle pain: | Promm (myot + myop. in EMG) |
| Muscle "cramps": | EMG silent = RMD (Torbergsen) |
| Unspec distal movements: | Neuromyotonia – |
| IBM: | EMG can be performed in "any" muscle, also where biopsy is uncommon |
| Muscle disease? | EMG sometimes gives "specific" findings (myotonia, Pompe) |
| Bilat per. atrophy – CMT2? | Myopathy a possibility (Udd) |
| Dist. ext weakness – radial n.? | Distal hereditary myopathy (Welander) |

 Indications for EMG

- Weakness/fatigue
  - central
  - motor neurone
  - peripheral nerve; pnp, focal
  - muscle (nm-j, myopathy, periodic weakness)

- Numbness
- Cramps
  - myotonia, ben. fasc. syn., neurotonia, stiff p. syn
- Pain
- ICU
  - Critical illness...

 Indications for EMG and myopathy

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