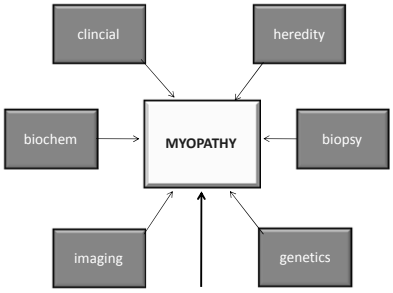
EMG in MYOPATHY

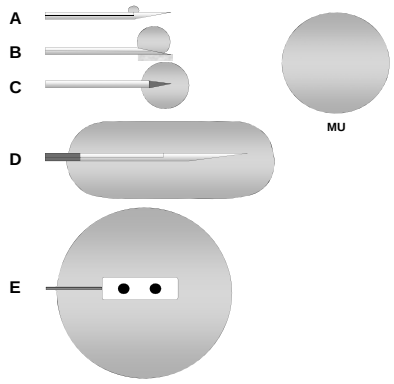
Which information do we get from EMG today?

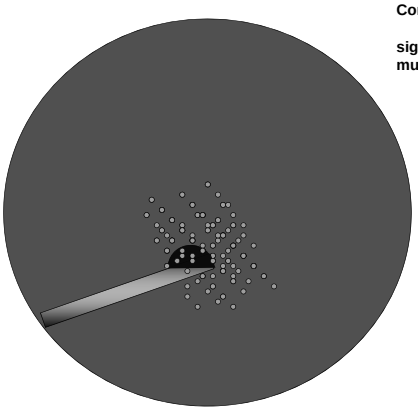
Erik Stålberg
Uppsala, Sweden



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**

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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**

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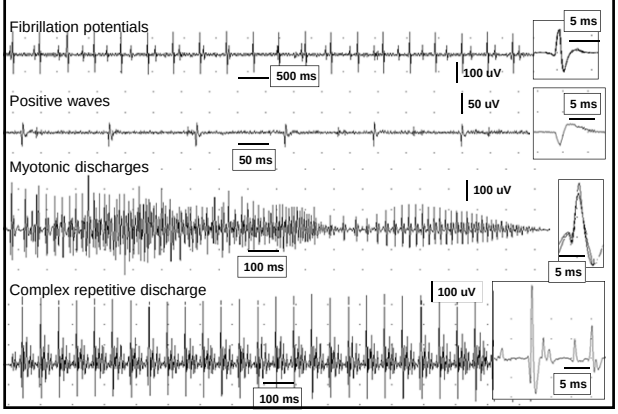
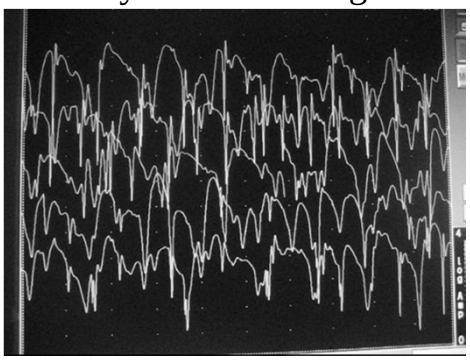
Spontaneous acitivity generated in the muscle fibre

(Stålberg,Daube 2003)

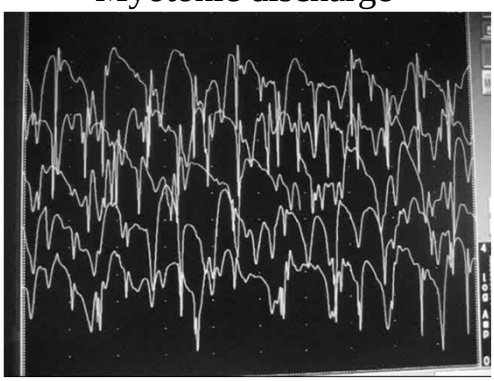


Myotonic discharge



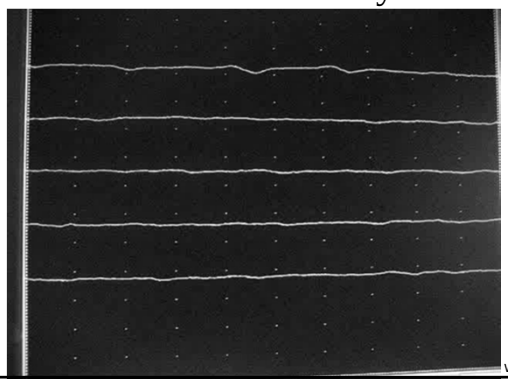
Avi

Myotonic discharge



wma

Myotonia; warm up after 1 minute of activity



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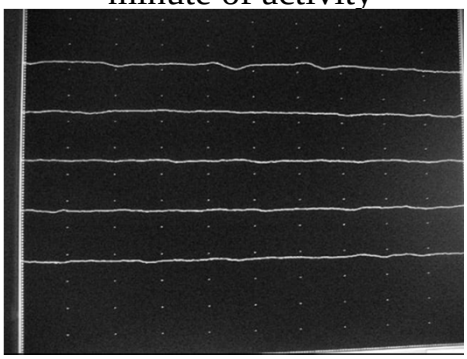

avi

CNEMG

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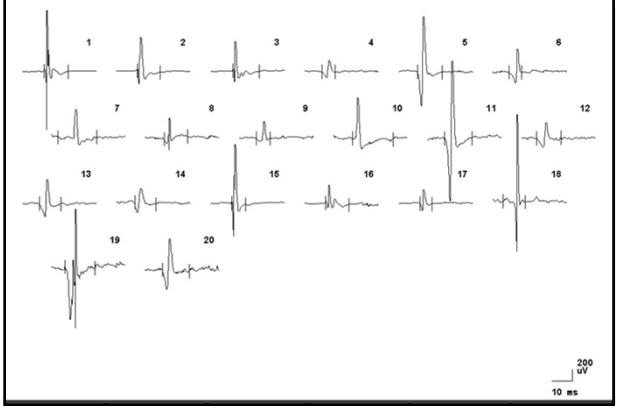
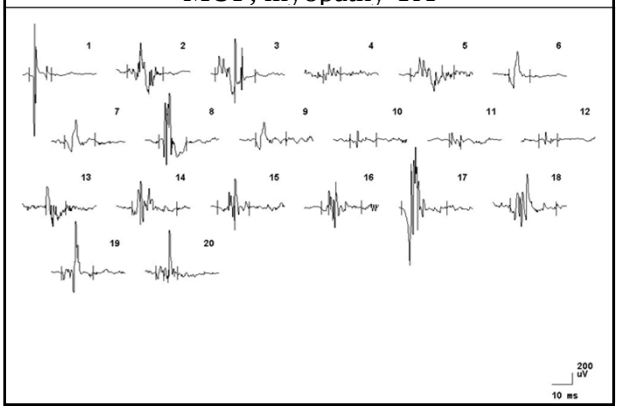
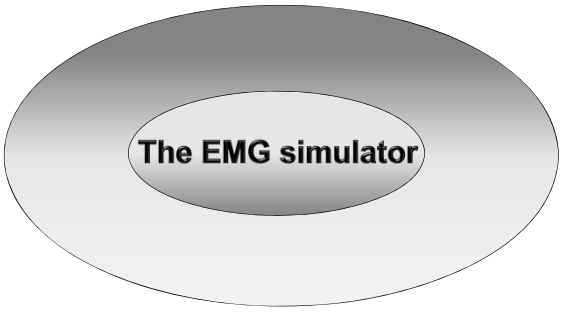
- **MUP** — **number of fibres in recorded area**
  fibre diameters
  n-m transmission
- IP — recruitment pattern
  total number of MUs at full effort

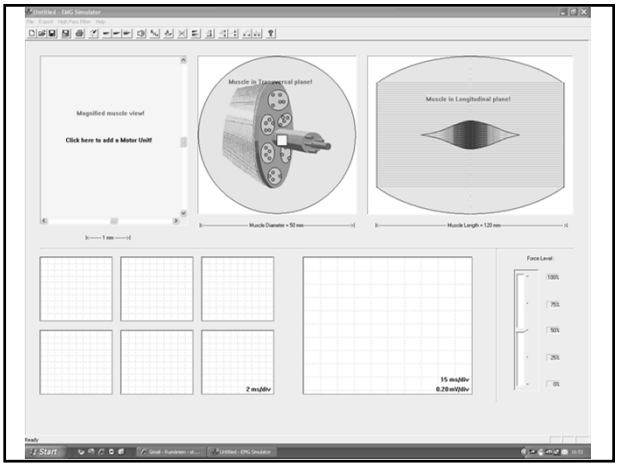
MUP, normal TA



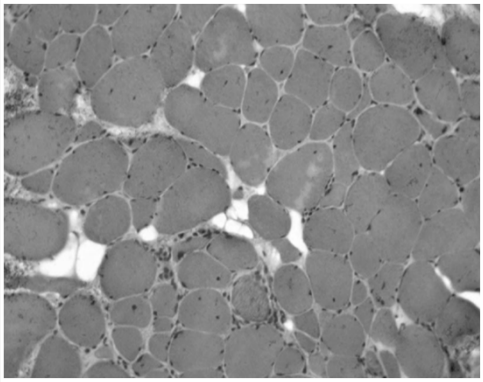
MUP, myopathy TA







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.

Courtesy Kallimo, 2010

CNEMG

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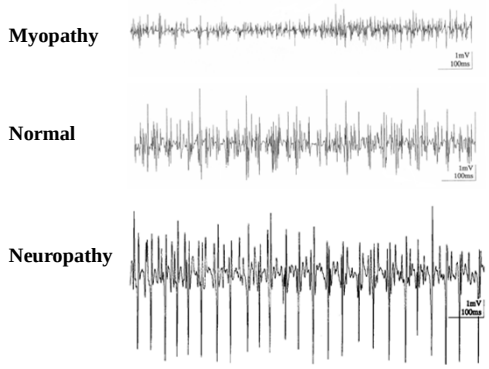
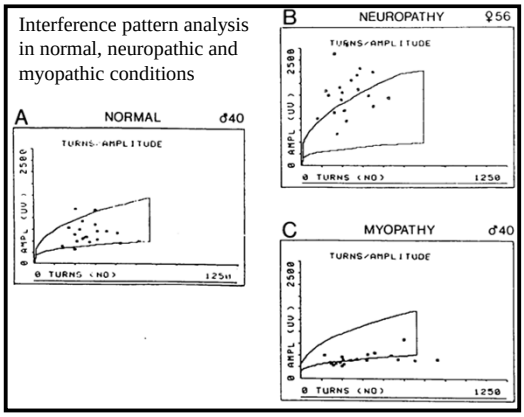
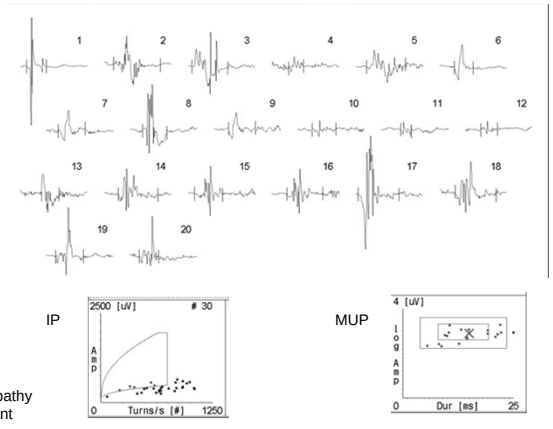
EMG - interference pattern



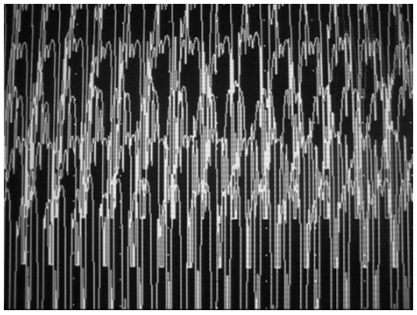
Interference pattern analysis in normal, neuropathic and myopathic conditions





Beyond conventional EMG

CRD in Pompe´s disease (CN rec)



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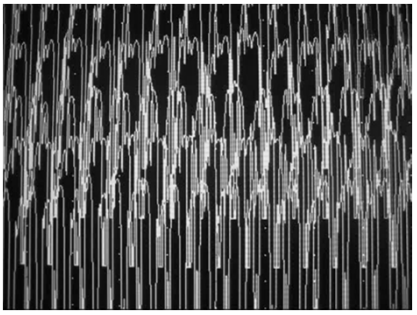
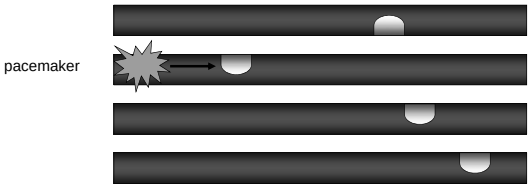
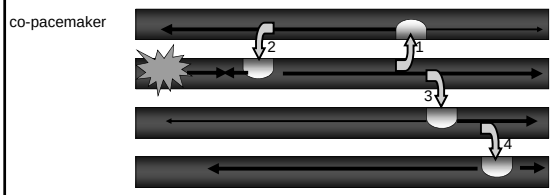


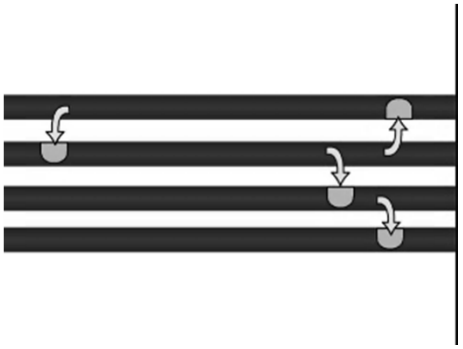
wma

Complex repetitive discharge, CRD

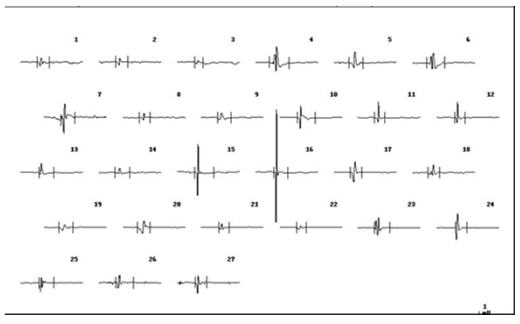


Complex repetitive discharge, CRD





Myopathy, EDB



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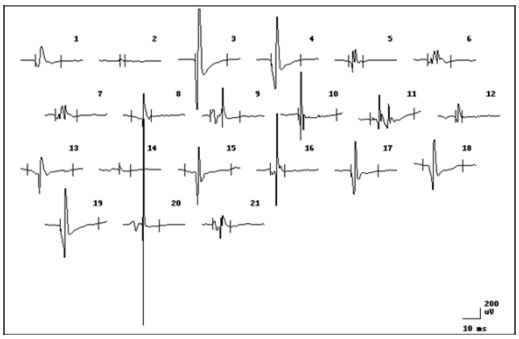
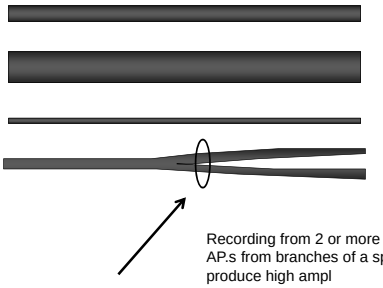
Hereditary distal myopathy (CN rec)



Lat vastus m

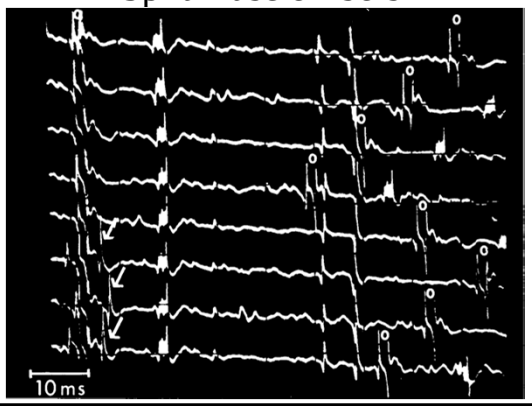
Muscle fibres in myopathy



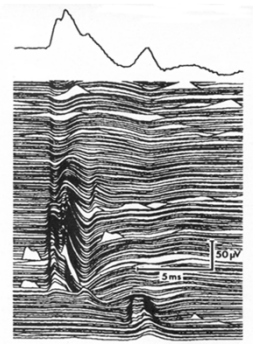
Recording from 2 or more still synchronous AP.s from branches of a split muscle fibre may produce high ampl

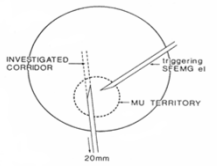
Stålberg

Split muscle fibers

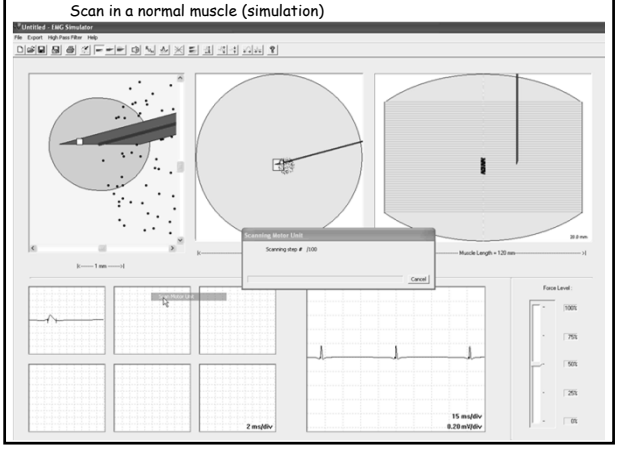


Scanning EMG

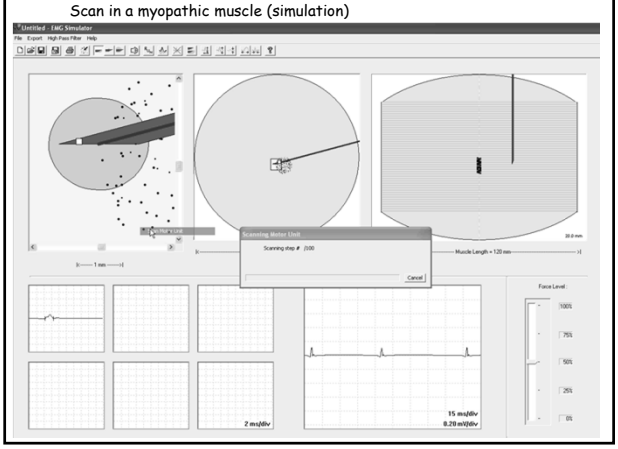




Scan in a normal muscle (simulation)



Scan in a myopathic muscle (simulation)



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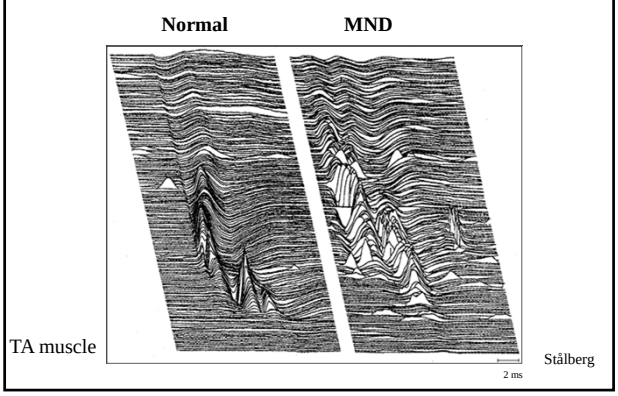

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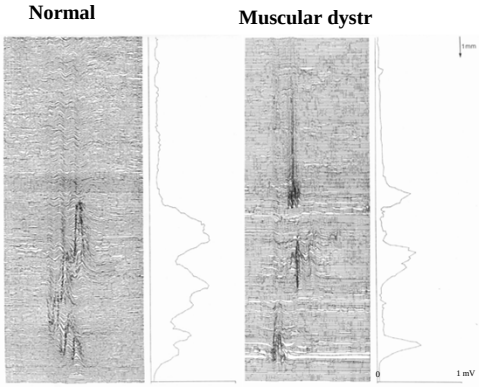
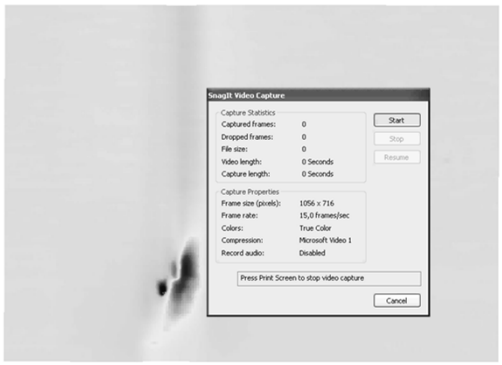
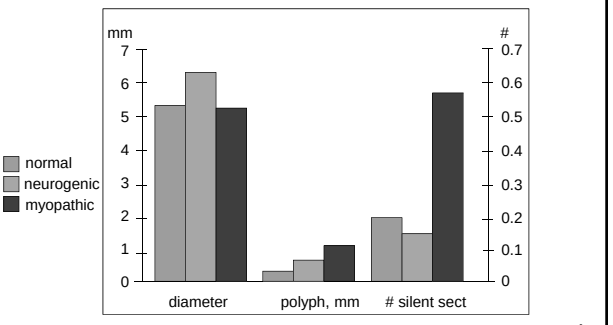




Scanning EMG

tibial anterior muscle



Dioszeghy, Stålberg

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 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?	Myopati a possibility (Udd)
Dist. ext weakness – radial n.?	Distal hereditary myopathy (Welander)

EMG may explain pathophysiological mechanisms

- CRD – SFEMG suggests ephaptic activation
- Myotonic weakness – decrement of single muscle fiber aps
- Hypokalemic paralysis – muscle fiber cond block (Zwartz)
- Painful myot (G1306A) giant psw due to ephases in fiber groups + longitud. cond block, channelopathy
- Biopsy: fiber type preponderance;
  - in cong. myop. - grouping? normal FD excludes reinnervation
  - in hypothyreosis - normal EMG = fiber type transformation

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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