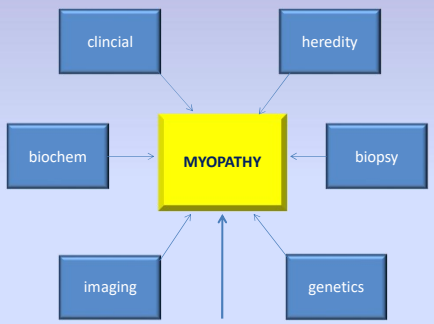
EMG in MYOPATHY

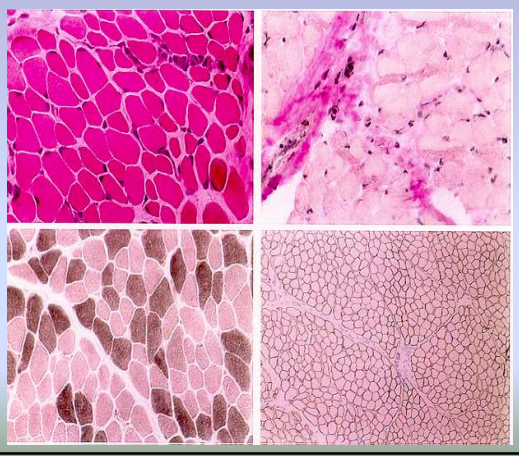
Which information do we get from EMG today?

Erik Stålberg
Uppsala, Sweden

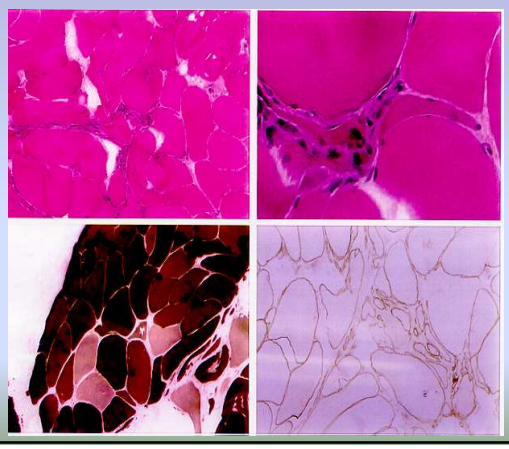
30



Fibre diam variation

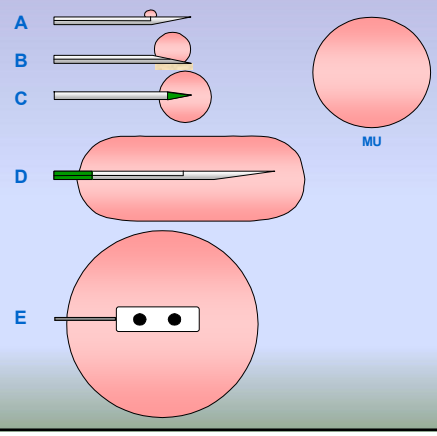


Myopathy



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

Electrodes



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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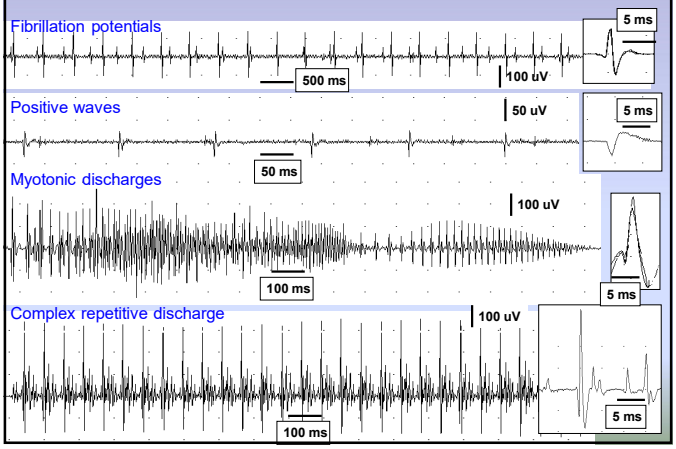
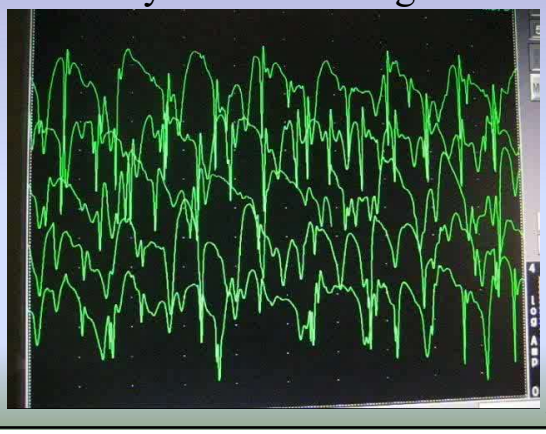
Spontaneous acitivity generated in the muscle fibre

(Stålberg,Daube 2003)

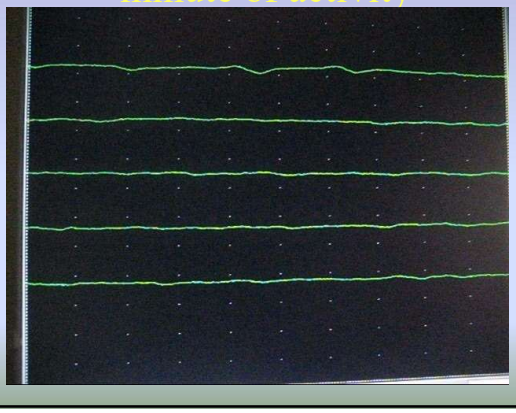


Myotonic discharge



wma

Myotonia; warm up after 1 minute of activity

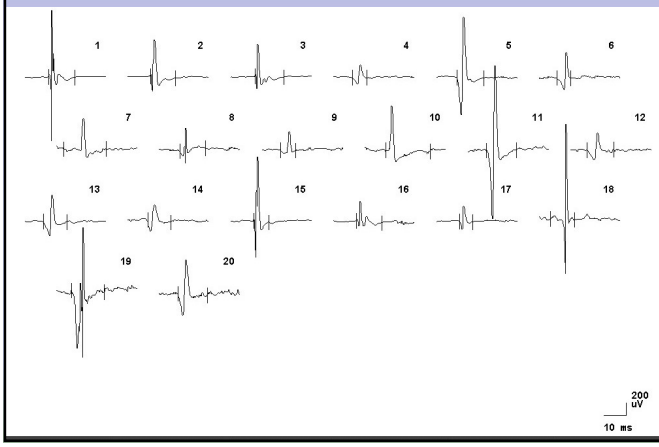


avi

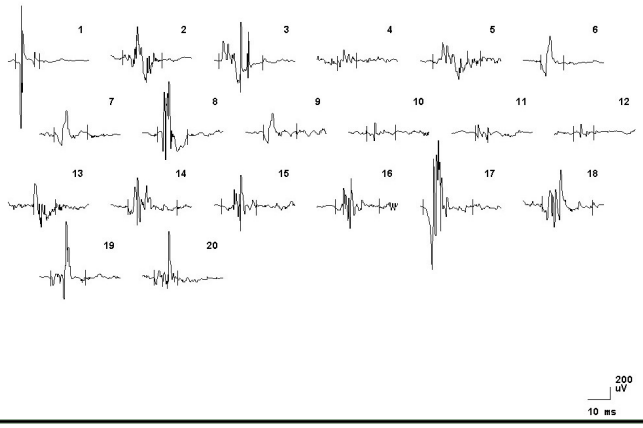
CNEMG

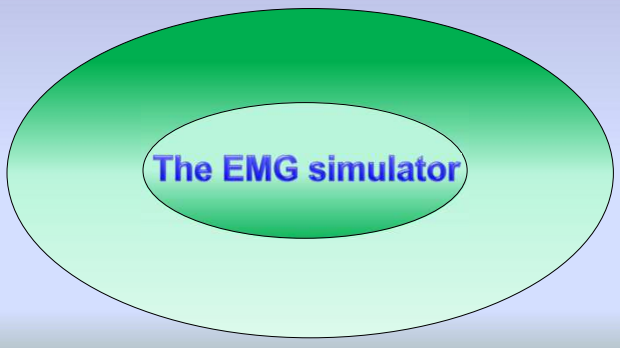
- At rest — denervation and spec spontaneous activity (myotonia, CRD, neuromyotonia)
- **MUP** — **number of fibres in recorded area**
  fibre diameters
  n-m transmission
- IP — recruitment pattern
  total number of MUs at full effort

MUP, normal TA

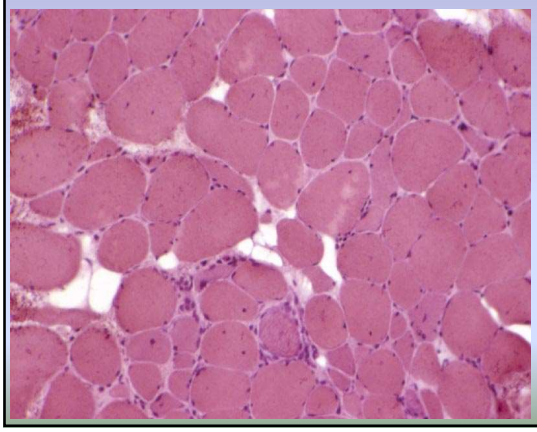


MUP, myopathy TA



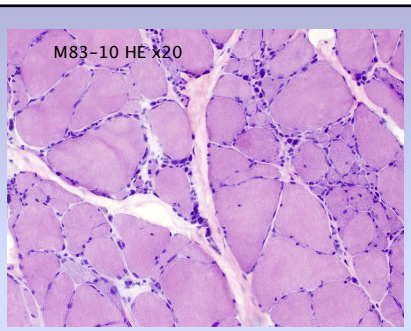


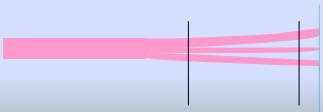
Emery-Dreifuss muscular dystrophy, X-linked type 1 (EDMD; emerinopathy)



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





longitudinal splitting

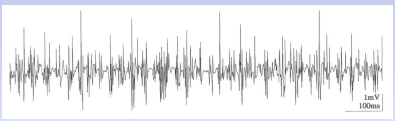
CNEMG

- 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

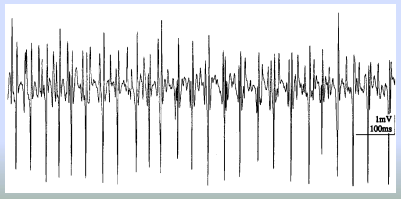
EMG - interference pattern

Myopathy

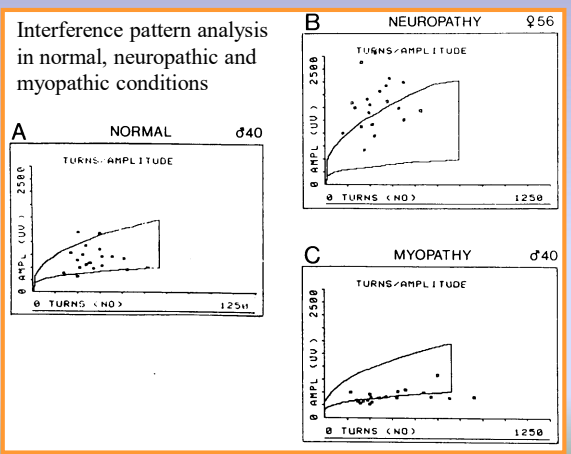
Normal



Neuropathy

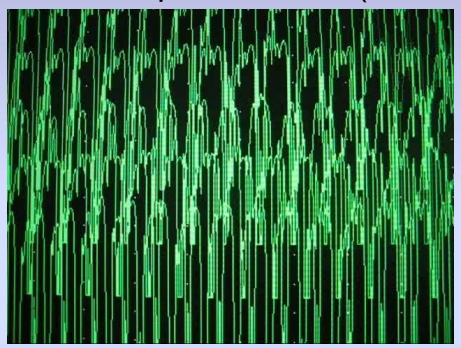


Interference pattern analysis in normal, neuropathic and myopathic conditions



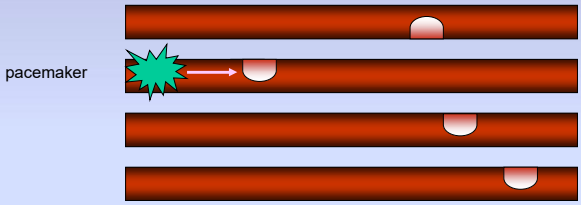
Beyond conventional EMG

CRD in Pompe´s disease (CN rec)

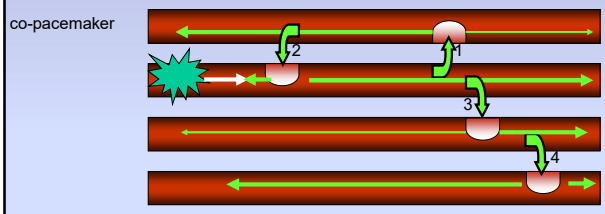


wma

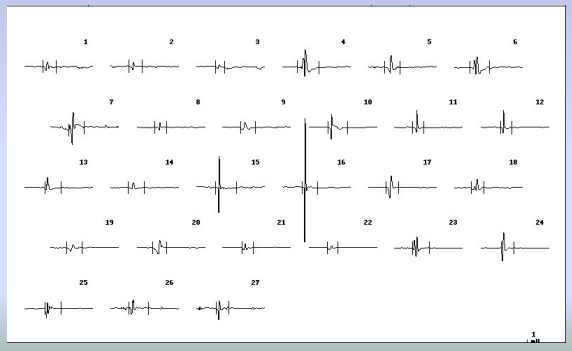
Complex repetitive discharge, CRD



Complex repetitive discharge, CRD



Myopathy, EDB



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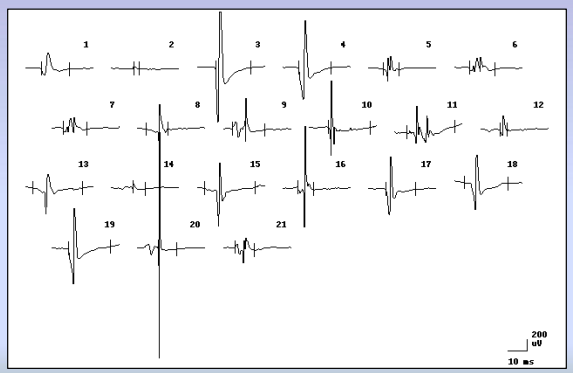
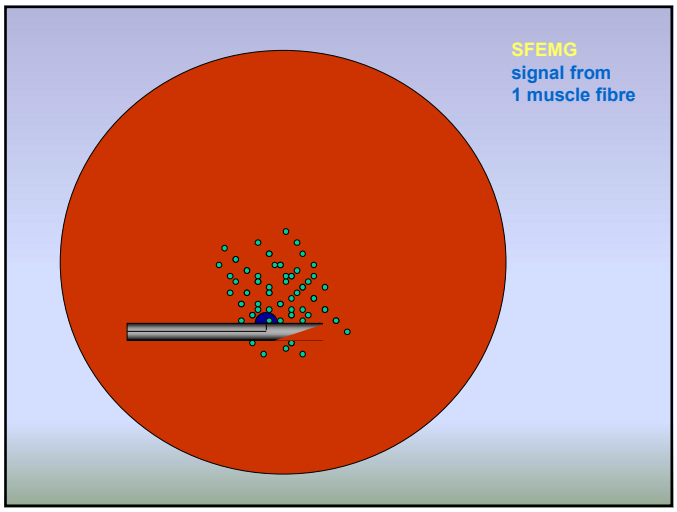
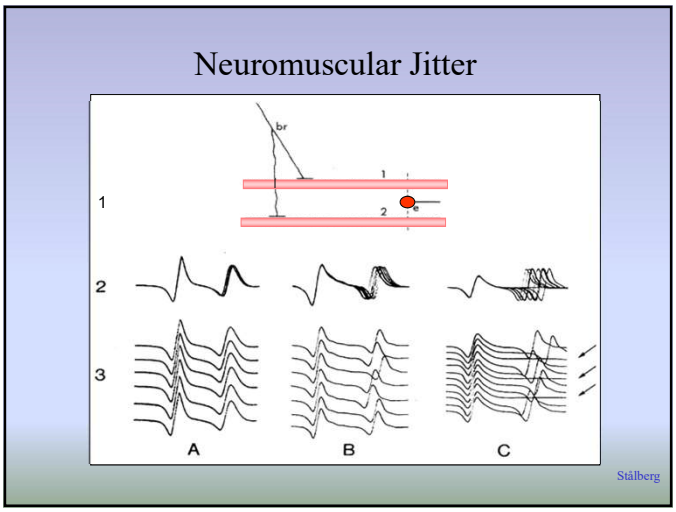
Hereditary distal myopathy (CN rec)

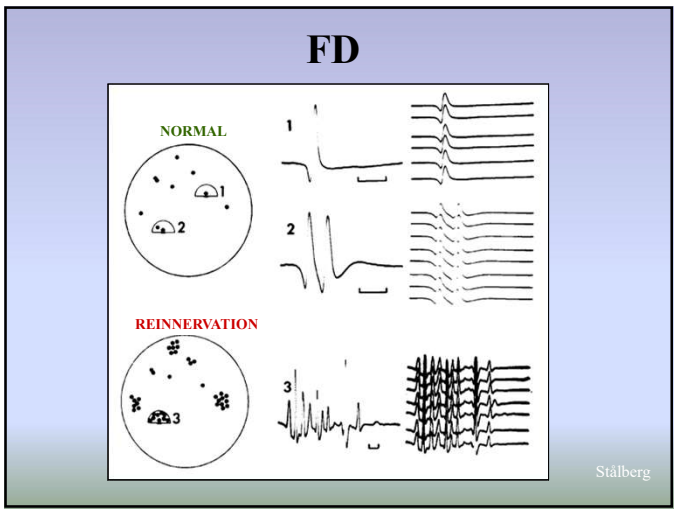


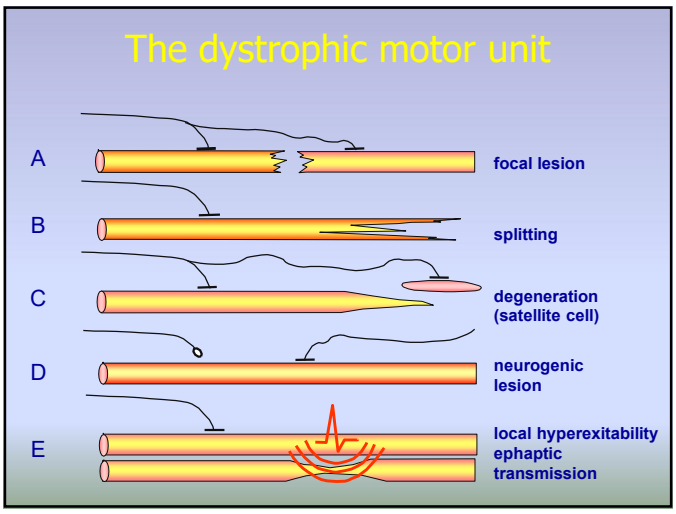
Lat vastus m

Other EMG methods

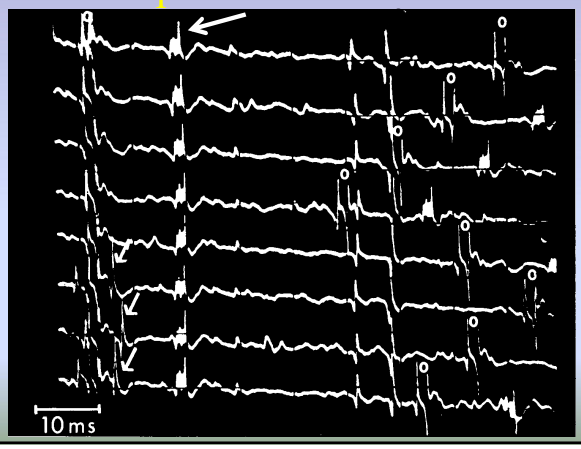

SFEMG
signal from
1 muscle fibre


Neuromuscular Jitter
br
1
2
e
1
2
3
A
B
C
Stålberg

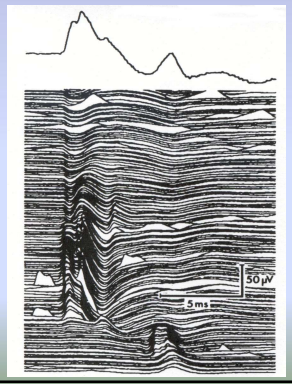

FD
NORMAL
1
2
REINNERVATION
3
Stålberg

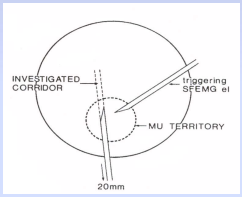

The dystrophic motor unit
A
focal lesion
B
splitting
C
degeneration
(satellite cell)
D
neurogenic
lesion
E
local hyperexitability
ephaptic
transmission

Split muscle fibers



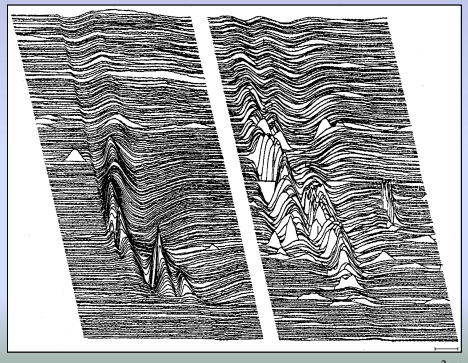
Scanning EMG





Scanning EMG

Normal **MND**

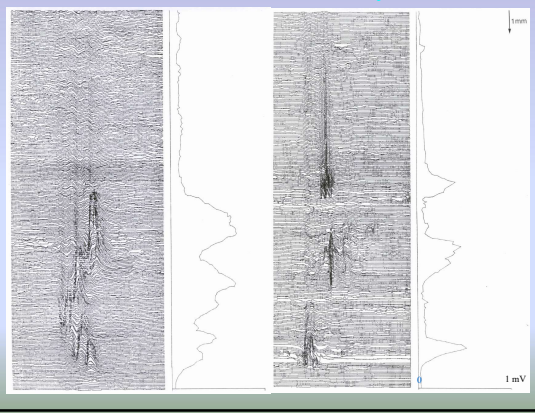


TA muscle

Stålberg

Scanning EMG

Normal **Muscular dystr**



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 MUPs — central

Indications for EMG and myopathy

- **Weakness/fatigue**
  - central
  - motor neurone
  - peripheral nerve; pnp, focal
  - muscle (nm-j, myopathy, periodic weakness)
- **Cramps**
  - myotonia, ben. fasc. syn., neurotonia, stiff p. syn
- **Pain**
- **ICU**
  - Critical illness…