The Integration of Neurography and EMG

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Uppsala, Sweden

Different or overlapping information

<table>
<thead>
<tr>
<th>Neurography</th>
<th>EMG</th>
</tr>
</thead>
<tbody>
<tr>
<td>myelin</td>
<td>+</td>
</tr>
<tr>
<td>cond block</td>
<td>+</td>
</tr>
<tr>
<td># axons</td>
<td>(+)</td>
</tr>
<tr>
<td>axonal degen</td>
<td>(+)</td>
</tr>
<tr>
<td>reinnervation</td>
<td>(+)</td>
</tr>
<tr>
<td>n-m junction</td>
<td>(+)</td>
</tr>
<tr>
<td>muscle unit</td>
<td>+</td>
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</tbody>
</table>

Neurography and EMG, the integration

<table>
<thead>
<tr>
<th>Condition</th>
<th>neurography</th>
<th>RNS</th>
<th>auton</th>
<th>EMG</th>
<th>SFEMG</th>
<th>other</th>
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<tbody>
<tr>
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<td></td>
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</tr>
<tr>
<td>GBS</td>
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<td></td>
</tr>
<tr>
<td>focal nerve lesion</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>- cervical</td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>- root/plexus</td>
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<tr>
<td>MND/MMN</td>
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</tr>
<tr>
<td>St p polio</td>
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<tr>
<td>MG</td>
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</tr>
<tr>
<td>myotonia</td>
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<td></td>
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<tr>
<td>other myasthenia</td>
<td></td>
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</tr>
<tr>
<td>pt/IBM</td>
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</table>

First choice
Complementary
Not necessary
Place of EMG

1. Ways to express EMG abnormality
2. MUP and IP analysis
3. Neurography and EMG, integration

What do we want to express

- Muscle membrane function - spontaneous
- Muscle fibre characteristics; diameter
- MU organisation
  - number of fibres
  - grouping
- N-M transmission
- # motor units
  - total
  - activation; pattern, fullness

Amplitude (Tib.ant.)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Amplitude Values</th>
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<td>Polymyositis</td>
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<tr>
<td>ALS</td>
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</table>

STD = 1.11
Mean = -1
N = 32,00

STD = 1.21
Mean = 1
N = 52,00
Irregularity Coefficient

\[ IR = \sum_{i=1}^{n} |y_i - y_c| / A \]

SIZE and IRREGULARITY of MUP

\[ SI = Size \text{ index} \]

\[ IR = \text{Irregularity Coefficient} \]
Mean values vs outliers

IR

myo

SIIR

Outliers (amplitude) below min

Outliers (duration) below min

Mean values vs outliers

Outliers (amplitude) above max

Outliers (duration) above max

Mean values vs outliers

amplitude
duration

amplitude
duration

amplitude
duration

amplitude
duration

amplitude
duration
SLIGHT MYOPAHY

MODERATE NEUROGENIC

CENTRAL WEAKNESS
Comparison between different QEMG parameters

<table>
<thead>
<tr>
<th>Parameter</th>
<th>MUP</th>
<th>T/A</th>
<th>freq</th>
<th>sEMG</th>
<th>MUNE</th>
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<tbody>
<tr>
<td>ampl</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
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<tr>
<td>dur (local size)</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
</tr>
<tr>
<td>n,mj</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>spont act</td>
<td>+</td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>MU size</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>MU #</td>
<td>+</td>
<td></td>
<td></td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>MU distr recruitment/fullness</td>
<td>+</td>
<td></td>
<td></td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>central drive, tremor</td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>fatigue</td>
<td>-</td>
<td>+</td>
<td></td>
<td>+</td>
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</tbody>
</table>

Neurography in muscle disorders

- Indications
  - concomitant neuropathy? (mitochondr, pm, paramalignancy, secondary entrapment)
  - use CMAP to assess muscle bulk
Neurography in MND/MMN

MND:
Exclude axonal neuropathy
Confirm normal SCS
Exclude MMN

MMN:
Demonstrate motor conduction block in individual motor nerves
Confirm normal SCS

EMG in pnp

EMG in pnp, MUP summary
EMG in pnp, jiggle + poly

EMG in St p polio

Small fiber testing

- Autonomic test (RR, SR)
- Epidermal nerve fiber density
- Thermotests
- Near nerve needle neurography
- Microneurography
- Special neurography methods
Other investigations for muscle

- CK
- Muscle biopsy
  - morphology
  - histochemistry
  - electromicroscopy
  - metabolic factors
- Genetic studies
- MRI
- CT
- Ultrasound

Other tests in MND

- **MUNE**
  - Reduced # MU should be assessed in MND, St p polio
    - electrical stimulation (incremental, dual stim sites, statistical)
    - voluntary (MUNIX)
- **TMS**
  - Excitability (threshold and PSTH)
  - CCT
  - TST
CTS, palmar inteross and 1st lumbrical

EMG in myotonia
- confirm myotonic discharges
- is EMG myopathic or not
- explore distribution (prox-dist)
- effect of temperature
- effect of activity

Neurography in St p polio
- No primary reason
- Atypical symptoms need further EDX
  - neuropathy (pnp, entrapment)
EMG in St p polio

- confirm neurogenic involvement
- find subclinical involvement
- assess degree of MU loss
- find other cause of symptoms:
  - entrapment, radiculopathy

Neurography in MG

- No primary reason for neurography
- Used when picture is atypical and when RNS and SFEMG are negative
- NOTE:
  - during any neurography low CMAPs should alert the examiner on nmj problems (remember to test facilitation in routine and in ICU)

SFEMG in MG

- assess increased jitter (same as jiggle in conc EMG)
- confirm normal FD
- not expected
  - increased FD (reinnervation)
  - normal jitter in 20/20 recordings
EMG in CTS

- EMG NOT necessary for the diagnosis per se. Neurographic methods are sensitive and specific.

- If EMG is used,
  - the question is to exclude roots; in Ext Carp Rad (C6) and EDC and Flex carp rad (C7)
  - in APB it may answer the question of amount of axonal lesion (but CMAP is usually better)

Autonomic tests, RR, SSR

- To assess involvement
  - in GBS may be vital
  - small fiber involvement
  - specific conditions, e.g. amyloidosis,

EMG in Musc Dysr

- Typical findings
  - spont activity
  - small polyphasic MUPs
  - early recruitment
  - dense or reduced IP (severity)

- Not expected
  - normal EMG - think of non dystrophic cond.
  - myotonia
Neurography in Musc dystr

• No primary reason for neurography
  If performed:
  • Expected findings
    – low motor ampl,
    – normal MCV
    – F waves low ampl, normal persistence
    – normal sensory ampl
  • Not expected
    – abnormal neurography (think of mitochondrial cond, paramalignant condition)

Neurography

– pathophysiology  demyelinating/axonal/CB
– fiber type sensory/motor/autonomic
– fiber size large/small
– distribution distal/proximal
– severity

Neurography in GBS

• demonstrate acute motor and sensory neuropathy
• demonstrate conduction block
• assess: severity, pathology, distribution
Neurography in root/plexus

- Sensory (with sensory symptoms)
  - normal distal amplitudes - root or CB anywhere
  - reduced distal amplitudes - axonal plexus involvement

- Motor (with weakness)
  - reduced distal amplitudes - axonal lesion
  - normal amplitudes - CB

Neurography in focal lesion

Motor symptoms:
- pathophysiology and severity
  - demyelinating or CB focal testing (SSS)
  - axonal SSS may not help, go to EMG

Sensory symptoms:
- low distal amplitudes go to other nerves, + EMG
- normal distal amplitudes find focus (if not, make SEP)

Neurography in CTS

- to assess:
- pathophysiology:
  - demyelination latency
  - axonal distal ampl
  - CB block across ligament
- fiber type
  - sensory/motor
- severity

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CTS severity

- very slight: only relative abnormality (other nerves; uln mot, uln sens, rad sens)
- slight: only sensory abnormality
- moderate: sens + motor
- severe: no sens resp, motor abnormality
- very severe: no responses

EMG in GBS

- **EMG in Early phase:**
  - No indication
  - MUNE (but only MUNIX which includes voluntary act)

- **EMG in Late phase:**
  - degree of axonal involvement
  - jiggles
  - IP
  - Macro

EMG in MG

- No indication in diagnostic work up
- If SFEMG is neg, EMG is indicated to find alternative diagnosis to MG
EMG in MND

• To confirm
  – generalized denervation
  – fasciculations
• To exclude myopathy

EMG in MMN

• To demonstrate focal/multifocal denervation

Neurography in myotonia

• NCS (motor and sensory conducton studies)
  usually not necessary when EMG has confirmed myotonia
• When myotonia is suspected, it is wise to start with EMG

RNS in MG

• Least sensitive method. If this is pos. and typical, MG is highly suspected.
  – proximal muscles
  – no treatment
  – warm muscle
• exclude (think of…)
  – LEMS, myotonia, Mc Ardle, cong MG
EMG in PM/IBM

- Expected positive findings
  - myopathy
  - spont. activity (fib, CRD) (th. paraspinals)

- Not expected
  - normal EMG
  - neurogenic pattern (except in end stage)
  - myotonia

EMG in focal nerve lesions

- Localize site
  - pure axonal focal lesion cannot be defined with neurography
  - root lesions (involvement of post rami= root, ant rami for segment)

- assess degree of axonal damage
- follow reinnervation (spont activity, conventional MUP parameters, jiggle, IP)

- MUNE/MUNIX

Why EMG in pnp

Not always necessary…but possible objectives are to:

- assess amount of axonal damage
  - long nerves

- assess dynamics
  - jiggle

- assess distribution
  - distal/prox
  - asymmetric

- exclude other reasons of symptoms
  - distal myopathy

- find clue to underlying condition
  - neurotonia
Neurography in GBS

- confirm MOTOR-sensory demyelinating pnp
- confirm conduction block (MCS, F persistence)
- assess site (prox-dist --antiMAG)
- assess amount of axonal involvement (CAMP ampl)
- autonomic involvement

- NOTE:
  - CB due to high temperature
  - nerve hypoexcitability

Distribution of conduction slowing

<table>
<thead>
<tr>
<th></th>
<th>proximal</th>
<th>even</th>
<th>distal</th>
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<tbody>
<tr>
<td>GBS</td>
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<td></td>
<td>(+)</td>
</tr>
<tr>
<td>CIDP</td>
<td>+</td>
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<tr>
<td>CMT1</td>
<td></td>
<td>+</td>
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</tr>
<tr>
<td>anti MAG</td>
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</table>


Conduction block in MMN

Stålberg
MRI in muscle disorders

Titinopathy (Udd)

Courtesy Torbergsen, Löseth

EDX PATTERN IN MYOTONIAS
Fournier et al, Ann Neurol. 2004

<table>
<thead>
<tr>
<th>Gen</th>
<th>Titinopathy</th>
<th>BI</th>
<th>Other</th>
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<th>Hyperplastic</th>
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<tbody>
<tr>
<td>Def</td>
<td>T101M / MLEC</td>
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<tr>
<td>Needle EMG</td>
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<td>Latent change</td>
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<tr>
<td>Sensitivity</td>
<td>100</td>
<td>83</td>
<td>63</td>
<td>83</td>
<td>84</td>
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Protocol - myotonia

Every 2 sec
50 sec
-3 times

10 sec act

Every 2 sec
50 sec
-3 times

5 min
interrupted act

Immediately and then every 45 min
EDX PROTOCOL IN MYOTONIAS
Fournier et al, Ann Neurol. 2004

Protocol
• EMG, myotonia and MUP analysis
• Neurophysiology:
  – CMAP at rest ADM and EDB
  – Decrement; pre-values, then 10 stim 3Hz
  – Short exercise, 10 sec;
    • Stim every 2 seconds for 50 sec
    • Repeat this 3 times, 60 sec apart
  – Long exercise, 5 min with brief pauses every
    30 sec
    • Pre-values
    • Test after 2 seconds
    • Test every min for 45 minutes