Testing
Neuromuscular transmission

Erik Stålberg

The neuromuscular junction

Howard in Stålberg, 2003
Multiple targets at the NMJ

Tests for MG

- **CLINICAL**
  - History
  - Tests
  - fatique, Tensilon, ice test

- **EMG**
  - Rep nerve stimulation
  - slow-fast, postactivation facilitation/exhaustion

- **INTRACELL REC**
  - Needle-EMG SFEMG
  - shape variability jitter

- **STAPEDIUS REFLEX**
- **OCULOGRAPHY**
- **TONOMETRY**
- **ACHR ANTIBODIES**

**Intracellular recordings,**
- action potentials not shown

**Intracellular recordings,**
- schematic with APs
Schematic explanation to the myasthenic decrement

All or none response of individual motor end-plates

CMAP representing the sum of above

Myasthenic disorders

- Non-familial
  - Autoimmun MG (post)
  - LEMS (pre)
  - Toxins, drugs (pre or post)

- Congenital syndromes
  - presynaptic, synaptic, postsynaptic

Myasthenic disorders

- Myasthenia gravis
  - reduced AChR
  - antibodies to AChR (85%)

- Seroneg MG
  - normal ACHR density
  - anti-MUSK antibodies in 2/3

- LEMS
  - reduced release of Ach
  - antibodies to presynaptic Ca-channels
  - autonomic symptoms
  - malignancy in 65%
Decrement protocol

- **normal**
- **MG**
- cholinergic crisis
- **LEMS**

Decrement at rest
(pat referred as ?MG, result normal)
5% confidence limits

<table>
<thead>
<tr>
<th>Muscle</th>
<th>n</th>
<th>decrement</th>
</tr>
</thead>
<tbody>
<tr>
<td>ADM</td>
<td>66</td>
<td>-3,4</td>
</tr>
<tr>
<td>Anconeus</td>
<td>255</td>
<td>-3,8</td>
</tr>
<tr>
<td>Deltoid</td>
<td>92</td>
<td>-4,8</td>
</tr>
<tr>
<td>Nasalis</td>
<td>239</td>
<td>-4,9</td>
</tr>
<tr>
<td>Trapezius</td>
<td>208</td>
<td>-4,0</td>
</tr>
</tbody>
</table>

**Protocol**

- 3 Hz, 10 stimuli
- immobilize the muscle
- max stim strength, 125%
- test at: rest after 10 sec of act and after 1 minute

**Parameters to analyse**

- initial amplitude
- decrement
- amplitude after activity (postactivation facilitation)
- decrement after activity

**Rep.nerve stimulation: considerations**

- distal/proximal muscle
- rest/fatigue
- on/off treatment
- cold/warm
- stim. frequency
- muscle fixation
Muscles to test

- Generalized MG
  - Deltoideus
  - Trapezius
  - Anconeus
  - Nasalis
- Bulbar MG
  - Nasalis
  - Anconeus
- Ocular MG
  - RNS is quite insensitive
  - Nasalis
  - Start with SFEMG jitter

Bulbar MG
- Nasalis
- Anconeus
- Trapezius

Is there?
- myasthenia
- good/ bad effect of AchE inhib’s
- cholinergic overdose
- LEMS
- McArdle, myotonia

Decrement in 2 proximal muscles

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Decr %</th>
<th>Decr %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Deltoideus</td>
<td>-6%</td>
<td>-7%</td>
</tr>
<tr>
<td>Trapezius</td>
<td>-2%</td>
<td>-4%</td>
</tr>
</tbody>
</table>

Mean decrement 24.8
Mean amplitude 9.0

Repetitive nerve stimulation
Anconeus muscle

<table>
<thead>
<tr>
<th>3 Hz</th>
<th>3 Hz</th>
<th>3 Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td>-2%</td>
<td>-2%</td>
<td>-7%</td>
</tr>
</tbody>
</table>

2 ms/D 2.5 ms/D
**Repetitive nerve stimulation in a patient with severe MG**

Rest, 3 Hz 10 stim

Directly after 20 s act

Post-act facilitation

0 sec after 20 s activation

1 min after activation

3 Hz

-76%

-80%

3 Hz

-18%

-26%

3 Hz

-75%

-78%

**Facilitation after exercise in LEMS**

*Tim and Sanders, M&N, 1994*

**Facilitation with 20 Hz stimulation in LEMS**

*Tim and Sanders, M&N, 1994*
Jitter analysis (SFEMG, CNE)

Single fiber action potentials

Intramuscular stimulation and SFEMG recording
Measuring jitter with Concentric Needle electrodes

Erik Stålberg
Uppsala, Sweden

Four types of EMG electrodes

Recording surface: width (µm), length (µm), area (mm²)

- Monopolar: 680, 240, 0.240
- Concentric: 150, 580, 0.070
- Concentric, facial: 80, 300, 0.019
- SFEMG: 25, 25, 0.0005

Three types of EMG electrodes

- CN, 0.070 mm²
- SFEMG, 0.0005 mm²
- CN, 0.019 mm²
Three types of EMG electrodes

SFEMG

Jitter with conc needle electrode

Jitter SF vs Conc, mixed diag

#>10 values, total mtrf; n=92

non-MG

MG

reinn
Jitter with CNE

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Mean MCD limit µs</th>
<th>Individual data limit µs</th>
</tr>
</thead>
<tbody>
<tr>
<td>GO vol</td>
<td>31</td>
<td>45</td>
</tr>
<tr>
<td>GO stim</td>
<td>27</td>
<td>36</td>
</tr>
<tr>
<td>Frontalis vol</td>
<td>28</td>
<td>38</td>
</tr>
<tr>
<td>Frontalis stim</td>
<td>21</td>
<td>28</td>
</tr>
<tr>
<td>Ext dig vol</td>
<td>30</td>
<td>43</td>
</tr>
<tr>
<td>Ext dig stim</td>
<td>24</td>
<td>35</td>
</tr>
</tbody>
</table>

Stålberg et al. Multicenter study, 2015

% positive results from a total of 291 patients

<table>
<thead>
<tr>
<th>Group</th>
<th>SFEMG</th>
<th>Decrement</th>
<th>Stapedius reflex</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ocular</td>
<td>85</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td>EDC</td>
<td>59</td>
<td>31</td>
<td>68</td>
</tr>
<tr>
<td>Mild generalized</td>
<td>96</td>
<td>68</td>
<td>89</td>
</tr>
<tr>
<td>Mod-severe generalized</td>
<td>100</td>
<td>68</td>
<td>89</td>
</tr>
<tr>
<td>Remission</td>
<td>62</td>
<td>0</td>
<td>83</td>
</tr>
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Stålberg Sanders 1981

Sensitivity of Diagnostic Tests in MG

- 550 patients with acquired MG
- All tests performed before immunotherapy or thymectomy
- Ocular myasthenia (OMG)
  - weakness only in ocular muscles
- Generalized MG (GMG)
  - weak in any non-ocular muscle

Sensitivity of initial tests

550 untreated MG patients

% Abnormal
**EDX in MG; start with jitter analysis if available**

Jitter analysis → **normal** → STOP

Logics: we have ‘never’ seen normal jitter and abnormal RNS

Jitter analysis → **abnormal** → RNS

Why RNS in this case: to see specific patterns (LEMS, cong MGs, other disorders, to follow over time)

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**RNS if jitter analysis is unavailable**

Why RNS

- To see MG profile of decrement
- To see specific patterns (LEMS, cong MGs, other disorders,
- To follow over time

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**Some links**

Sfemg.info (SFEMG meetings videos)