Disclosures

• None
Goals and Objectives

- To provide a concise, high-yield overview of a variety of neuromuscular diseases
- Understand common clinical presentations of neuromuscular disease
- Review electrodiagnostic findings in several neuromuscular diseases
- Systematically diagnose diseases common to a neuromuscular clinic
Case 1

- A 64 year old woman had problems with muscle stiffness “since birth”
- After seconds of activity any muscle used becomes stiff
- Cold temperatures or emotional stress also cause muscle stiffness
- 1 sister and 1 son have identical symptoms
- Action myotonia with eye closure, smiling, hand grip; diffuse percussion myotonia
Case 1
Case 1

• Serum CK is normal
• Routine nerve conduction studies are normal
Short Exercise Test

**Stimulus Site**

| A1: BASE1  | Lat: 3.0 | Dur: 7.0 | Amp: 12.6 | Area: 47.3 | Temp: 32.1 
|-----------|---------|---------|-----------|------------|---------
| A2: BASE 2 | 3.0     | 7.1     | 14.9      | 47.2       | 32.1    |
| A3: IMM POST E | 3.6 | 5.7 | 9.6 | 32.8 | 32.0 |
| A4: 10 SEC | 5.3     |         |           | 0.6        | 31.9    |
| A5: 20 SEC | 3.4     | 5.7     | 1.9       | 4.7        | 31.9    |
| A6: 30 SEC | 3.3     | 5.5     | 3.0       | 8.5        | 31.9    |
| A7: 40 SEC | 3.3     | 5.6     | 3.3       | 9.5        | 31.9    |
| A8: 50 SEC | 3.2     | 5.9     | 3.7       | 11.2       | 31.9    |
# Short Exercise Test

<table>
<thead>
<tr>
<th>Stimulus Site</th>
<th>Lat (ms)</th>
<th>Dur (ms)</th>
<th>Amp (mV)</th>
<th>Area (mVms)</th>
<th>Temp (°C)</th>
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Short Exercise Test

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Short Exercise Test

Amp 1: 2-10kHz
Temp: 33.8 °C

Stimulus Site
- A1: 30 MIN
- A2: 35 MIN
- A3: 40 MIN
- A4: 45 MIN
- A5: 50 MIN
- A6: 55 MIN
- A7: 60 MIN
- A8: 65 MIN

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<th>Dur (ms)</th>
<th>Amp (mV)</th>
<th>Area (mVms)</th>
<th>Temp (°C)</th>
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<td>A1: 30 MIN</td>
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</table>
Needle Examination

- Discharges in all limb muscles at rest, accentuated upon activation
- MUPs obscured by discharges
Diagnosis?
Case 2

- 50 year old woman with 15 years of muscle stiffness
- Initially in hands only, now diffuse
  - Tongue and jaw, eyelids, diffuse extremity
- Hand weakness
- Difficulty swallowing solids, liquids “take longer to go down”
- Chronic constipation, cataracts
- 3 miscarriages, 1 preterm birth
- Mother possibly mild stiffness, 2 of 4 daughters affected, 1 granddaughter severely affected since birth
Case 2
Case 2
Case 2
Case 2
Case 2

• Serum CK is normal
• Routine nerve conduction studies are normal
Needle Examination - FDI

50 uV  Amp 1: 20-10kHz  10 ms
Diagnosis?
Case 3

- 50 year old woman presents with 1 month of left hand paresthesias
  - Seen in followup for a L ICA distribution stroke 5 months prior
- Hyperlipidemia, h/o smoking, cocaine use, OCPs
- Baclofen, diazepam, Cymbalta, gabapentin, Prilosec, ropinirole, simvastatin, demerol, ASA

Current exam
- NIHSS 2
- -1 R deltoid, biceps, triceps, knee flexors and extensors, plantar flexors; -2 R hip flexors; -3 R dorsiflexors
- Normal exam otherwise
Case 3

• LUE EDX ordered to evaluate for carpal tunnel syndrome versus cervical radiculopathy
L Median Motor
L Median F-wave
L Median Sensory
L Ulnar Motor

[Graph showing nerve conduction velocity test results]

Stimulus Site:
- A1: Elbow
- A2: Wrist

Segment:
- Elbow-Wrist: 341 ms (Dist), 5.6 ms (Diff), 61 ms (CV)
- Wrist-Abductor digiti minimi: 65 ms (Dist), 2.5 ms (Diff), no response

Values:
- Duration: 0.1 ms
- Rate: 0.5 Hz

Parameters:
- Level: 0.0 mA
- Dur: 0.1 ms
- Single
- Average: Off
- Sig. Enhancer: Off
- Recording Site: Abductor digiti minimi

Comments:
- Display: Average
- Replicate
- Rollback
- Stim: Recurrent
- Signal Enhancer: Off
- Amp Control: All

Note: The graph and table data are typical for nerve conduction studies to assess motor function along the ulnar nerve pathway.
L Ulnar F-wave
L First Dorsal Interosseus
L APB
L Pronator Teres
L Biceps Brachii
L Tibialis Anterior

50 uV  Amp 1: 20-10kHz  50 ms
Summary

Nerve conduction studies:

<table>
<thead>
<tr>
<th>Stimulate (record)</th>
<th>Amplitude</th>
<th>Velocity</th>
<th>Distal Latency</th>
<th>F-wave Latency</th>
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<tr>
<td></td>
<td>R</td>
<td>L</td>
<td>NL</td>
<td>R</td>
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<tr>
<td>Ulnar, m (hypothenar)</td>
<td>8.4</td>
<td>61</td>
<td>(&gt;51)</td>
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<td>Ulnar, s anii (fifth)</td>
<td>34</td>
<td>69</td>
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<tr>
<td>Median, m (thenar)</td>
<td>9.3</td>
<td>52</td>
<td>(&gt;48)</td>
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<tr>
<td>Median, s anii (index)</td>
<td>11</td>
<td>49</td>
<td>(&gt;56)</td>
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EMG:

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<tr>
<th>Muscle</th>
<th>Inc</th>
<th>Fib</th>
<th>Fasc</th>
<th>MUP</th>
<th>NL</th>
<th>Act</th>
<th>Rec</th>
<th>Duration</th>
<th>Amplitude</th>
<th>Phases</th>
<th>%</th>
<th>Turns</th>
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<td>Vastus medialis*</td>
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<tr>
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</table>

Remark: * brief myotonic discharges
Diagnosis?
Case 4

- 18 year-old AHWM
- Muscles “lock up” following unclear time of activity for about the past year
  - Nonpainful
- Relaxes after “a second”
  - Whole-body fatigue afterwards
- If continues to exert himself, develops weakness of the exerted muscles
- Some tightness exerting muscles after rest
- No effects of different foods
- ? Problems with leg and neck stiffness, handgrip stiffness, eye closure stiffness
- No myoglobinuria, cramps, fasciculations
- No sensory, coordination, gait, cognitive abnormalities
Case 4

• No family history
  • 21 year-old brother without similar problems
• Product of full-term gestation, no perinatal difficulties
• Developed normally, able to perform on level with peers
• High school basketball and baseball standout
• ROS, PMHx, SHx otherwise unremarkable
Case 4

- CK, prolactin, thyroid function, ESR, RF, CRP, ANA, CMV, Lyme normal
- Parvo B19 and EBV IgG positive
- EMG May, 2009 – profuse fibrillation potentials, positive sharp waves “consistent with an inflammatory myopathy”
  - Prednisone 20 mg per day no benefit
- Left vastus lateralis muscle biopsy June, 2009 – normal
- Thought possibly to be a side effect minocycline, but was discontinued with no benefit
- EMG June, 2009 – normal RNS, fibrillation potentials and positive sharp waves consistent with a “myopathy”
Neurological Exam

• No EOM, eye closure, grip, percussion myotonia
• Grip weaker with repeated testing
• Reduced UE MSRs
• Otherwise normal exam
EMG

Nerve conduction studies:

<table>
<thead>
<tr>
<th>Stimulate (record)</th>
<th>Amplitude R</th>
<th>L</th>
<th>NL</th>
<th>Velocity R</th>
<th>L</th>
<th>NL</th>
<th>Distal Latency R</th>
<th>L</th>
<th>NL</th>
<th>F-wave Latency R</th>
<th>Est.</th>
<th>L</th>
<th>Est.</th>
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<tbody>
<tr>
<td>Ulnar, m ( hypothenar )</td>
<td>11.3</td>
<td>(&gt;6)</td>
<td></td>
<td>61</td>
<td>(&gt;51)</td>
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<td>Median, s anti (index)</td>
<td>36</td>
<td>(&gt;15)</td>
<td></td>
<td>62</td>
<td>(&gt;56)</td>
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<td>44</td>
<td>(&gt;41)</td>
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<td>6</td>
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EMG:

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<tr>
<th>Muscle</th>
<th>Ins. Act.</th>
<th>Spont Fib</th>
<th>Fasc</th>
<th>MUP NL</th>
<th>Recruitment Act</th>
<th>Rec</th>
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Right upper and lower extremity nerve conduction studies were normal. A short exercise test studying the ulnar motor nerve following 10 seconds of exercise showed no decrement or post-exercise myotonic potentials.

Concentric needle examination performed in selected right upper and lower extremity muscles showed myotonic discharges as well as scattered iterative discharges and fibrillation potentials in all muscles examined. Voluntary motor units appeared normal.

Interpretation:

Abnormal study. There is electrophysiological evidence for a diffuse myotonic disorder,
Left Vastus Lateralis
Diagnosis?
Myotonic Disorders

- Group of disorders characterized by:
  - Stiffness
  - Muscle pain
  - Sometimes weakness
  - Symptoms may be episodic or constant
Myotonic Disorders

• Dystrophic
  • Myotonic Dystrophy Type 1
  • Myotonic Dystrophy Type 2

• Nondystrophic
  • Myotonia congenita
  • Paramyotonia congenita
  • Potassium aggravated myotonia (“sodium channel myotonia”)

• Periodic Paralyses
  • Hyperkalemic Periodic Paralysis
  • Hypokalemic Periodic Paralysis *not* a myotonic disorder
Categorization

- Myotonia
  - DM1
  - DM2
  - MC
  - PAM
- Myotonia and periodic paralysis
  - PMC
  - HyperPP
- Periodic paralysis
  - HypoPP
Myotonic Discharges (MT)

- Spontaneous discharge
- Muscle fiber origin
- Brief spike, initial positive, or positive wave morphology
  - Classically waxing/waning amplitude and firing pattern
    - Can be waning only or short bursts
      - Easily confused with short runs of positive waves
Myotonia & Myotonic Discharges

Clinical and Electrical
• DM1
• DM2
• MC
• PMC
• PAM
• HyperPP

Electrical without clinical
• Acid maltase deficiency
• Myofibrillar myopathy
• Myotubular myopathy
• Hypothyroidism
• Toxic (fibrates, statins, colchicine, chloroquine)
• Inflammatory myopathies
• Amyloid
• Chronic denervation
Rep Stim Interpretation

• Immediate fall in CMAP following exercise and repair if present are specific (but not necessarily sensitive) for myotonic disorders (but not a specific syndrome); repair is prolonged in PMC

• Return to normal is usually in <2 minutes for all forms other than PMC, in which it may take up to 90 minutes

• Look for postexercise myotonic potentials (PEMPs) in MC, PMC, PAM
Rep Stim in PMC

Before exercise
A

After exercise
C

B

D

EDX Approach to Myotonic Disorders – Routine CNE

- Consider facial muscles
- Look for proximal or distal predominance
- Myopathic MUPs *in rested muscle* occur in DM1, DM2, AR MC
- MT in MC are *often* shorter in duration, more variable in rate and amplitude, higher frequency
- MT in PMC are *often* lower frequency and appear following exercise
Additional CNE

- In clinically involved muscle
- Up to 20 forceful contractions
  - Changes in MT, loss of MUPs, fib’s
- In PMC, MT appear or increase, MUPs drop out, fib’s may appear
- In other forms, MT lessen and MUPs do not change
Short Exercise Test (SET)
• Ulnar/ADM (immobilized), record supramax CMAP
• Record CMAP every 5 minutes at rest
• Maximum voluntary contraction for 5-10 seconds
• Record CMAP immediately
  • If decrement, continue to record CMAP every 10 seconds until at baseline (usually 1-2 minutes)
• If decrement occurs after exercise, repeat several times to assess for decrement habituation
• Look for PEMPs
Prolonged Exercise Test (PET)

- Ulnar/ADM (immobilized), record supramax CMAP
- Record CMAP every 5 minutes at rest
- Maximum voluntary contraction for 2-5 minutes, resting every 15 seconds for 3-4 seconds
- Record CMAP immediately, then every 1-2 minutes for 40-60 minutes afterward or until there is no further decline in CMAP (can go to every 5 minutes after 5 minutes)
  - Decrement = (max CMAP – min CMAP)/max CMAP x 100
  - >40% abnormal
- In PP, immediate post-exercise CMAP may be larger, then have slow decline
PET

Myotonic Dystrophy Type 1 - Electrophysiology

• NCS usually normal
  • Distal CMAPs may be low

• Prominent distal MT
  • Waxing-waning
  • More easily elicitable than in DM2 (except TFL/VL)

• Distal and facial myopathic MUPs, early recruitment

• No change with cooling

• SET:
  • CMAP drop immediately after exercise with rapid recovery (<2 minutes)
  • Decrement habituates
Myotonic Dystrophy Type 1 - Pathology

- Number of otherwise nonspecific features on biopsy
- Typical findings
  - Increased internal nuclei
  - Highly atrophic fibers
  - Type 1 fiber atrophy
  - Ring fibers
  - Type 2 fiber hypertrophy, fibrosis also common
Myotonic Dystrophy Type 1 - Pathology
Myotonic Dystrophy Type 2

- NCS normal
- Predominant UE distal, LE distal and proximal, and paraspinal MT
  - Waning only is possible
- Proximal myopathic MUPs, early recruitment
- No change with cooling or on SET
Myotonia Congenita

- NCS normal
- Generalized MT with needle movement or contraction
- Mildly myopathic MUPs with early recruitment in Becker’s AR MC
- Cooling may produce longer duration, more easily detected MT
- SET:
  - CMAP drop immediately after exercise with rapid recovery (<2 minutes), habituates
  - Larger drop, less habituation in AR MC (mutation dependent decrement)
  - PEMP
- 10 Hz rep stim may produce more decrement (>40%) than SET when warm in AR MC, cooling may increase decrement
Myotonia Congenita

Paramyotonia Congenita

- NCS normal
  - CMAPs may decrease in amplitude and area, increase in duration in cold
- Generalized MT (less easily elicited than MC), possible distal predominance
- Cooling:
  - Produces fibrillation potentials
  - Cooling <20 degrees C, MT disappears → paralysis (inexcitable muscle) and long-lasting silent contracture until warmed (may be delayed)
- SET:
  - CMAP drop immediately after exercise with prolonged recovery (up to 1 hour), drop worsens if repeated
  - May need to cool to elicit decrement, including with rep stim
  - PEMP
- PET:
  - May have severe decrement
Paramyotonia Congenita

Potassium Aggravated Myotonia

- NCS normal
- Proximal and distal MT
- Normal MUPs
- ? Effect of cooling, SET, PET
  - No CMAP changes
<table>
<thead>
<tr>
<th></th>
<th>Myotonic Dystrophy</th>
<th>Myotonia Congenita</th>
<th>Paramyotonia congenita</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Nerve conduction studies</strong></td>
<td>Low distal CMAP in DM1 (variable)</td>
<td>Normal</td>
<td>Normal CMAP falls with cooling</td>
</tr>
<tr>
<td><strong>Fibrillation potentials</strong></td>
<td>Prominent</td>
<td>Rare</td>
<td>Present Worsens with cooling</td>
</tr>
<tr>
<td><strong>Myotonic discharges</strong></td>
<td>Longer trains, Slower firing rate</td>
<td>Shorter trains, Faster firing rate</td>
<td>Short trains</td>
</tr>
<tr>
<td><strong>Warm-up phenomenon</strong></td>
<td>Yes</td>
<td>Yes</td>
<td>No (worse with repeated contractions)</td>
</tr>
<tr>
<td><strong>Motor unit potentials</strong></td>
<td>Short duration, polyphasic (Distal &gt; Proximal in DM1)</td>
<td>Normal or mildly short</td>
<td>Normal or short</td>
</tr>
<tr>
<td><strong>Short Exercise Test</strong></td>
<td>DM1: immediate CMAP drop after exercise with repair &lt;2 minutes</td>
<td>Immediate CMAP drop after exercise with repair &lt;2 minutes, PEMPs</td>
<td>Immediate CMAP drop after exercise with prolonged recovery (may need to cool to elicit), PEMPs</td>
</tr>
</tbody>
</table>

*CMAP = Compound Muscle Action Potential*
Case 1
Case 2
Case 3

<table>
<thead>
<tr>
<th>Stimulus Site</th>
<th>Lat.1 ms</th>
<th>Lat.2 ms</th>
<th>Amp. uV</th>
<th>Area uV/m</th>
<th>Temp °C</th>
</tr>
</thead>
<tbody>
<tr>
<td>A1: Wrist</td>
<td>3.6</td>
<td>4.9</td>
<td>11</td>
<td>5</td>
<td>38.4</td>
</tr>
<tr>
<td>A2: Elbow</td>
<td>8.4</td>
<td>9.5</td>
<td>8</td>
<td>5</td>
<td>38.3</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Segment</th>
<th>Dist mm</th>
<th>Diff ms</th>
<th>CV m/s</th>
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</thead>
<tbody>
<tr>
<td>Wrist-Index</td>
<td>130</td>
<td>3.6</td>
<td></td>
</tr>
<tr>
<td>Elbow-Wrist</td>
<td>235</td>
<td>4.8</td>
<td>49</td>
</tr>
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</table>
Likely Diagnosis?
Case 4
Diagnosis?