Partial Denervation
Complete Denervation
No MUAPs
WAYS PSWs DIFFER FROM FIBRILLATION POTENTIALS

- PSWs can be recorded sooner after PN injury
- Generalized PSWs can be seen in absence of fibrillation potentials ("EMG disease")
- PSWs may be seen in distal muscles of clinically normal subjects
- PSWs without fibrillation potentials may be seen following muscle trauma
- PSWs may be seen alone in demyelination
Myopathy
Neuropathy

ELECTROMYOGRAPHY

- Severity estimate
- Time estimate
- Combined with clinical findings, can be used to estimate prognosis
VALUE OF CLINICAL ELECTROMYOGRAPHY

Normal
Abnormal
  • Distribution
    Generalized - proximal, distal
    Localized - peripheral nerve, nerve root, plexus
  • Time of Onset
  • Severity
  • Neuropathy/Myopathy
  • Improving/Progressing
Overview

- Introduction
- Pathophysiology of Radiculopathy
- Electrophysiologic exam of Radiculopathy
- Cervical Radiculopathy
- Lumbar Radiculopathy
- Recent Literature
- Practice Questions

Why do we EMG?

- “Extend” the physical exam
- Confirm suspected clinical diagnosis
- Rule out competing diagnoses (peripheral neuropathy, plexopathy)
- Characterize the localization and pathologic mechanism of the disease process
- Assess extent/severity of disease process
- Assess for subclinical disease
- Assess for acuity/chronicity of disease process
- Assist with determining prognosis/ response to specific treatment interventions

Pathophysiology of Radiculopathy

- Sensory only>Motor and Sensory> Motor only
- Root compressions always occur proximal to the dorsal root ganglion
Electrophysiologic evaluation of radiculopathy

• Nerve conduction studies
• Late responses
  • F waves
  • H reflex
• Somatosensory Evoked Potentials (SEP)
• Motor Evoked Potentials (MEP)
• Needle Electrode Exam (NEE)

Nerve conduction studies

• Purpose:
  • Assess for peripheral neuropathy
  • Assess severity of radiculopathy
• Recommendations (AANEM Practice Parameter):
  • 1 motor
  • 1 sensory
• What is the expected finding (if any) in a motor nerve conduction study in the setting of radiculopathy?

H-reflex

• Purpose:
  • Assess for peripheral neuropathy
  • Supplement an evaluation for S1 radiculopathy
• Latency or amplitude?
  • >1.0-1.8 ms side to side difference
  • >50% side to side amplitude difference
• Caveat: poor sensitivity (Dillingham ref)
• Absent in only 8% of healthy 60-88 year olds in one study
• Benefit: assess sensory pathways that are neglected with needle electrode exam
• Downsides:
  • Only assess S1 roots
  • Can be normal in radiculopathy when fibers involved in the reflex are spared
  • Abnormal H reflex can reflect many other peripheral conditions
  • H reflex may not return following S1 nerve root injury

F-waves

• What:
  • Antidromic stimulation of motor nerves
• What assessed: motor nerve patency
• What is measured:
  • Minimal latency
  • Mean latency
  • Chronodispersion
  • Size
  • Persistence
F-waves

- Purpose:
  - Screen for polyneuropathy
  - Supplement radiculopathy eval (controversial)

- Caveat: Low sensitivity for radiculopathy (13-69%)
  Abnormal segment “diluted” in normal axon.

Somatosensory Evoked Potentials

- What:
  - stimulation over mixed or cutaneous nerve or dermatome
  - record over peripheral nerve, spine, scalp
  - Generally, not used for radiculopathy evaluations

Motor Evoked Potentials

- Nerve root stimulation
- Transcranial magnetic stimulation

- Essentially, nerve conduction studies with proximal stimulation over spine or nerve roots

- Not in wide clinical use

Needle Electrode Exam

- Abnormal Spontaneous Activity
  - Acute denervation
  - Positive sharp waves
  - Fibrillation potentials
  - Complex repetitive discharges
  - Myotonic discharges
Needle electrode Exam: MUAP analysis

- What is normal?
  - Buchthal studies defined normal ranges of amplitudes and duration

- Polyphasia
  - <20% polyphasics considered normal

- Amplitude
  - Dependent on type of needle electrode (monopolar/concentric)

- Recruitment
  - Reduced recruitment

Needle Electrode Exam

- How many muscles?

<table>
<thead>
<tr>
<th>Source</th>
<th># muscles</th>
<th>Sensitivity (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>AANEM, 1999</td>
<td>50-71%</td>
<td></td>
</tr>
<tr>
<td>Dillingham, 2000</td>
<td>4 w/o PSP</td>
<td>38-75</td>
</tr>
<tr>
<td></td>
<td>4 w/PSP</td>
<td>77-90</td>
</tr>
<tr>
<td></td>
<td>5 w/o PSP</td>
<td>50-77</td>
</tr>
<tr>
<td></td>
<td>5 w/PSP</td>
<td>84-91</td>
</tr>
<tr>
<td></td>
<td>6 w/o PSP</td>
<td>62-79</td>
</tr>
<tr>
<td></td>
<td>6 w/PSP</td>
<td>87-93</td>
</tr>
<tr>
<td></td>
<td>8 w/PSP</td>
<td>73-80</td>
</tr>
</tbody>
</table>

Needle electrode exam

- How many muscles?
  - 6, including paraspinals
  - "In order to avoid harm, 6 in the leg and 6 in the arm" - Dillingham

- Which muscles?
  - In the suspected root:
    - One proximal
    - One distal
  - Two different peripheral nerves
  - In the nerve root above suspected level
  - In the nerve root below suspected level
  - Paraspinals

Cervical Radiculopathy
Cervical Radiculopathy

- C7>C6>C8>C5
- H reflex: FCR
- CMAPs for prognosis:

<table>
<thead>
<tr>
<th>Root</th>
<th>Nerve conduction Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>C6</td>
<td>Infraspinatus</td>
</tr>
<tr>
<td>C7</td>
<td>Lateral head triceps</td>
</tr>
<tr>
<td>C8</td>
<td>Pronator quadratus</td>
</tr>
<tr>
<td>T1</td>
<td>Abductor digiti minimi</td>
</tr>
</tbody>
</table>

Upper extremity myotomal chart

Lumbar Radiculopathy

- L5 > S1 > L4
- H- reflex: gastroc/soleus

Lower extremity myotomal chart
Electrophysiologic evaluation of radiculopathy - timing of study

- Week 1:
  - Reduced recruitment
  - Prolonged H-reflex latency
  - "early polyphasic"

- Week 2:
  - Positive waves in paraspinals
  - Possibly decreased CMAP amplitude

- Week 3:
  - Abnormal findings in limb muscles

Practice Question #1

- A 35-year-old gentleman presents to your clinic with pain radiating from the right low back to the right lateral leg and dorsal foot. Physical exam reveals weakness in right great toe extension. Which needle electrode study would be most appropriate?

  - RFEM, ATIB, PTIB, MGAS
  - ATIB, PTIB, MGAS, RFEM, SHBF, LGAS
  - ATIB, PTIB, MGAS, VMED, TFL, PSP
  - RFEM, ADD, ATIB, PTIB, MGAS, LGAS, SHBF

Practice Question #2

- Your favorite local football hero presents to your office with a stinger sustained on the field over the weekend (4 days ago). His coach requests urgent Edx evaluation. What finding may yield helpful information at this point?

  - CMAP amplitude
  - H reflex latency
  - Recruitment in limb muscles
  - Spontaneous denervation in the paraspinals

Resources

- AAEM Minimongraph #32: The Electrodiagnostic Examination in Patients with Radiculopathies
Other works cited

- "Brachial Plexus Logical Schematic" by Chris Talbot - Own work. Licensed under CC BY-SA 3.0 via Wikimedia Commons - http://commons.wikimedia.org/wiki/File:Brachial_Plexus_Logical_Schematic.svg#mediaviewer/File:Brachial_Plexus_Logical_Schematic.svg

- "Lumbar plexus" By Gray822_es.svg: *Gray822.png: Gray derivative work: Ninovolador (talk) derivative work: Mcstrother (Gray822_es.svg) [CC BY 3.0 (http://creativecommons.org/licenses/by/3.0)], via Wikimedia Commons
Traumatic and Entrapment Neuropathies

Arthur Rodriguez, MD, MS
Emeritus Associate Professor, Rehabilitation Medicine
University of Washington School of Medicine

Frequency of Peripheral Nervous System Trauma

- Of patients admitted to Level 1 trauma centers:
  - 2-3% have peripheral nerve injuries
  - 2-3% have brachial plexus injury
- Of those with PNS injuries, 60% have TBI
- Of those with traumatic brain injury
  - 10-30% have PNS injuries

Nerves Most Often Affected

- Upper limb > Lower limb
- Upper Limb
  - Radial
  - Ulnar
  - Median
- Lower Limb
  - Sciatic
  - Peroneal

Classification of Nerve Injuries (Seddon)

- Neurapraxia
- Axonotmesis
  - Sunderland subdivides axonotmesis into 3 anatomically based categories depending on the degree of intraneural disorganization
- Neurotmesis
Neurapraxia
- Comparatively mild injury
- Motor and sensory loss
- No axonal (Wallerian) degeneration
- Nerve conducts normally distally
- Focal demyelination and/or ischemia
- Recovery within hours to a few months

Axonotmesis
- Commonly seen in crush injuries
- Axon and myelin sheaths are broken
- Surrounding stroma partially intact
- Wallerian degeneration occurs
- Recovery depends upon axonal regrowth, internal disorganization, and distance to muscle

Neurotmesis
- Nerve is completely severed, or so scarred (endoneurium and perineurium) that regrowth does not occur
- Sharp injury, traction and intraneural injection
- Prognosis is very poor without surgery

Classifying the Nerve Injury
- Neuropraxia
  - Distal CMAP and CNAP maintained
- Axonotmesis/Neurotmesis
  - CMAP and CNAP drop in rough proportion to degree of axon loss
  - Drop is complete by day 9 for CMAP and day 11 for SNAP
Mixed Lesions
(axon loss and conduction block)

- Percentage of axon loss best estimated by distal CMAP
- Percentage of conduction block by examining loss of amplitude from stimulation above and below the lesion

Needle EMG in Neurapraxia

- Reduced recruitment
  - (Increased recruitment ratio >7)

Needle EMG in Axonotmesis/Neurotmesis

- Length-dependent onset of fibrillations and positive sharp waves
  - Proximal muscles 10-14 days
  - Distal muscles 3-4 weeks
  - Fibrillation amplitudes decrease over time
- Sensitive indicator of axon loss, but does not quantify
- Beware of mixed lesions
- Beware of muscle trauma

Timing of the Electrodiagnostic Study Depends on the Question

- 7-10 days for localization and sorting neurapraxia from axonotmesis
- 3-4 weeks for most diagnostic information
- 3-4 months for detecting reinnervation
Localization of Nerve Injuries

- Focal slowing of conduction
  - Only with demyelination or conduction block
  - Not seen in pure axonal lesions
- SNAP amplitude
  - Helps with pre- vs post-ganglionic lesions
  - Normal SNAP in presence of complete denervation usually indicate root avulsion
  - Reduced amplitude indicates some post-ganglionic axon loss

Localization using SNAP’s

- Upper trunk
  - Median to thumb
  - Lat. Antebrachial Cutaneous
- Middle trunk
  - Median to long finger
- Lower trunk
  - Ulnar to small finger
  - Dorsal Ulnar Cutaneous

Localization of Root vs Plexus using paraspinal EMG

- Denervation suggests pre-ganglionic lesion
- Cannot differentiate between complete and incomplete lesions due to segmental overlap

Estimation of Prognosis using the CMAP Amplitude

- Much data comes from the study of facial nerve lesions
- Comparing CMAP on involved to uninvolved side:
  - > 30% - excellent outcome
  - 10-30% - good but incomplete recovery
  - <10% - poor recovery (insufficient overlap of intact axons for optimal terminal reorganization)
Immediate Surgical Reconstruction

- Sharp lacerations
- Complete nerve section
- Nerve ends are intact
- Minimal local tissue trauma

Delayed Surgical Reconstruction

- Nerve continuity unclear
- Natural recovery could be better than surgery
- Wait to see if there is clinical or EMG evidence of reinnervation
- Operate on those without ongoing recovery
- Usually intervene by 6 months to prevent end organ deterioration

Top 10 Entrapments

1. CTS
2. CTS
3. CTS
4. Ulnar Elbow
5. Radial Spiral Groove
6. Anterior interosseous
7. Ulnar Wrist
8. Radial Wrist
9. Fibular Knee
10. Tarsal Tunnel Syndrome
The best electrodiagnostic testing for CTS should be:

- In descending order of priority:
- Specific (few false positives)
- Sensitive (few false negatives)
- Reliable (get the same results on repeat testing)
- Resistant to Temperature Effects
- Efficient

Median motor conduction in CTS

- Prolonged distal latency
  - less sensitive than sensory prolongation
- Motor amplitude
  - low amplitude may indicate axon loss or conduction block
  - palm stimulation unreliable
- Martin-Gruber anastamosis

Repeated Measures in CTS

There are 3 good sensory tests for CTS
The problem is how to interpret them! More tests = more false positives

Combined Sensory Index (CSI)

Add 3 sensory latency differences

- Median – radial thumb difference
- Median – ulnar ring finger difference
- Median – ulnar mid-palmar difference

= CSI

Sensory Reference Values

<table>
<thead>
<tr>
<th>Reference Value</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Palm: Med - Uln</td>
<td>≤ 0.3</td>
</tr>
<tr>
<td>Ring: Med - Uln</td>
<td>≤ 0.4</td>
</tr>
<tr>
<td>Thumb: Med - Rad</td>
<td>≤ 0.5</td>
</tr>
<tr>
<td>CSI</td>
<td>≤ 0.9</td>
</tr>
</tbody>
</table>
Combined Sensory Index (CSI)

- Improved Sensitivity in mild CTS
- High Specificity
- Improved test-retest reliability

Sensitivity and Specificity of CSI

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>MidPalm</td>
<td>≤ 0.3</td>
<td>70%</td>
<td>97%</td>
</tr>
<tr>
<td>Ring</td>
<td>≤ 0.4</td>
<td>74%</td>
<td>97%</td>
</tr>
<tr>
<td>Thumb</td>
<td>≤ 0.5</td>
<td>76%</td>
<td>97%</td>
</tr>
<tr>
<td>CSI</td>
<td>≤ 0.9</td>
<td>83%</td>
<td>95%</td>
</tr>
</tbody>
</table>

**Results: Palmdiff**

![Median Ulnar Mid-Palmar](image)

**Results: CSI**

![CSI Test - Retest](image)
Reliability

<table>
<thead>
<tr>
<th>Test</th>
<th>Spearman Rho</th>
</tr>
</thead>
<tbody>
<tr>
<td>MidPalmar</td>
<td>.74</td>
</tr>
<tr>
<td>Ring</td>
<td>.67</td>
</tr>
<tr>
<td>Thumb</td>
<td>.75</td>
</tr>
<tr>
<td>CSI</td>
<td>.95</td>
</tr>
</tbody>
</table>

Effect of temperature on latency

- 8 cm Median: 0.06 msec/degree Celsius
- 10 cm Median: 0.11 msec/degree Celsius
- 14 cm Median: 0.14 msec/degree Celsius
- All Latency Diff: not affected by temperature

Single Test vs. CSI

With very small or very large differences
CSI may not be necessary
Perform Needle EMG?

- Debated
- My practice:
  - Signs or symptoms of cervical radiculopathy
  - History of Trauma
  - Abnormal Median Motor Response

Ulnar Neuropathy at the Elbow

Motor Conduction Studies

- Elbow should be flexed
- Stimulate at wrist, below elbow, above elbow, and axilla
- At least 10 cm across elbow is advocated
- NCV < 48 m/s is likely slowing
- Less useful to compare with forearm NCV
- Beware of Martin Gruber!

Ulnar Neuropathy at the Elbow

FDI recording

- Active over bulk of the muscle
- Reference proximally over the dorsal CMC joint of the thumb
- (Don't place over index finger MCP.)
- 2 Channel recording
How much slowing is abnormal?

- Segmental Difference
  - Difference of 11 - 15 m/s often used
  - Assumes normal forearm conduction
  - But the forearm doesn’t stay normal, with axon loss!

- Absolute CV
  - < 48 m/s used
  - Does not depend on assuming forearm conduction is normal
  - Better sensitivity!

Comparison of Methods
(Specificity = 95% for all, Shakir & Robinson 2004)

<table>
<thead>
<tr>
<th>Method</th>
<th>Reference</th>
<th>Value m/s</th>
<th>Sensitivity</th>
</tr>
</thead>
<tbody>
<tr>
<td>ADM CV</td>
<td></td>
<td>48</td>
<td>80%</td>
</tr>
<tr>
<td>FDI CV</td>
<td></td>
<td>49</td>
<td>77%</td>
</tr>
<tr>
<td>ADM Difference</td>
<td></td>
<td>10</td>
<td>51%</td>
</tr>
<tr>
<td>FDI Difference</td>
<td></td>
<td>15</td>
<td>38%</td>
</tr>
</tbody>
</table>

Ulnar Motor Nerve Conduction

Median Nerve Stimulation
Wrist and Elbow
**Ulnar Neuropathy at the Elbow**

**Motor Conduction Studies**
- Inching studies may pick up more mild cases
- Stimulate at 2 cm increments along nerve
- > 0.7 msec latency change over 2 cm is suspicious
- More convincing if accompanied by sudden change in amplitude or shape

**“Inching” Technique**
- Avoid excessive stimulation intensity
- Move stimulator following submaximal stimulation to localize the nerve path prior to supramaximal stimulation

**Sensory Conduction Studies**
- Distal sensory amplitude is reduced early.
  - Reflects loss of sensory axons
  - Compare with the opposite side
- Don’t over-interpret drop in sensory amplitude from proximal stimulation
  - Expect 50% drop from wrist to elbow
  - Temporal dispersion and phase cancellation
### Ulnar Neuropathy at the Elbow

**Needle EMG**

- May show abnormalities in Ulnar distribution
  - even in a few cases with normal NCS
- FCU and FDP are often not involved
  - rarely, branches come off proximal
  - intraneural topography spares these branches

### Ulnar Neuropathy at the Wrist

- Ganglions
- Rheumatoid Arthritis
- Lacerations
- Fractures
- Occupational
  - Pipe cutters
  - Metal polishers
  - Mechanics

### Ulnar Neuropathy at the Wrist

- Mixed Motor and Sensory (30%)
- Pure Motor (52%)
- Pure Sensory (18%)
- Dorsal Ulnar Cutaneous Branch

### Anterior Interosseus Lesions

- Supplies FPL, FDP (digits 2 & 3), PQ; No sensory fibers
- Clinical Presentation:
  forearm pain, weakness of FPL FDP
difficult to isolate PO
- EMG of affected muscles is abnormal
  - beware of Martin Gruber (50% from AIN)
  - beware of FDS supply from AIN (30%)
- NCS - conventional studies normal
- **Etiology**
  - anomalous muscles
  - neuralgic amyotrophy
  - partial higher median neuropathy
**High Radial Nerve Lesions**

**Etiology**
- almost always traumatic
- crutch palsy (axilla, triceps often weak)
- Saturday night or honeymoon palsy (triceps usually spared)

**EMG and NCV**
- EMG of triceps, brachioradialis, forearm extensors most useful.
- Radial SNAP reduced in amplitude
- Motor studies may show focal slowing or conduction block, but these are not optimal.

**Electrodiagnostic Exam**
- Needle EMG is most useful
  - Work down radial nerve
    - Triceps
    - (Anconeous same as branch to medial triceps)
    - Brachioradialis (important for prognosis)
    - ECR
    - EDC
    - ECU
    - EIP
  - Non radial muscles by same roots

**Superficial Radial Nerve Lesions**
- Reduced sensation in radial distribution
  - pain is often most disabling
- Etiologies include:
  - wristwatch, handcuffs, casts
  - laceration during IV or deQuervain’s surgery
- Only electrodiagnostic finding is abnormal SNAP.
Fibular Neuropathy

- There is no more Peroneal Nerve
  - Anatomists killed it
  - Worried about confusion with perineum
  - If you leg and perineum confused, you have big problems

- Important to record from Tibialis Anterior
  - EDB has no useful function
  - EDB is often absent in Fibular Neuropathy
  - Tib Anterior more helpful for prognosis

Fibular Motor NCS

- Record EDB and Tibialis Anterior
  - provides confirmatory results
  - helpful when EDB response is absent

- Stimulate
  - ankle, below fibular head, lateral popliteal fossa
  - beware of volume conduction from tibial nerve

- Inching across fibular head often helpful

Recording Site for Tibialis Anterior

- Active over motor point
- Reference distal over tendon
  - Not over muscle
- Better for prognosis and localization
- Can do 2 channels

Beware of Accessory Fibular Nerve

- Originates from Superficial Fibular Nerve
- Supplies Lateral Head of EDB
- Fibular Motor NCS to EDB
  - small CMAP at ankle
  - larger CMAP at fib head
  - CMAP present behind lateral malleolus
Fibular Nerve EMG

- Deep Fibular Distribution
  - Tibialis Anterior, EHL
- Superficial Fibular Distribution
  - Peroneus Longus
- Tibial Distribution
  - Gastroc, Soleus (to rule out higher lesion)
- Short Head of Biceps Femoris
  - to rule out higher lesion

Nerve Conductions for TTS
Are there more techniques than patients?

- Motor latencies to AH and ADQP
- Sensory latencies from 1st and 5th toes
  - surface recording
  - near nerve recording
- Mixed nerve latencies with plantar stim
  - recording over medial malleolus
- Variations of the above

Nerve Conduction in TTS
Galardi et al 1994 (14 Pts, 12 Nmls)

<table>
<thead>
<tr>
<th>Technique</th>
<th>Sensitivity</th>
<th>Specificity</th>
</tr>
</thead>
<tbody>
<tr>
<td>CMAP latency</td>
<td>22%</td>
<td>100%</td>
</tr>
<tr>
<td>SNAP Med Plantar</td>
<td>93%</td>
<td>92%</td>
</tr>
<tr>
<td>SNAP Lat Plantar</td>
<td>100%</td>
<td>83%</td>
</tr>
<tr>
<td>CNAP Med Plantar</td>
<td>64%</td>
<td>100%</td>
</tr>
<tr>
<td>CNAP Lat Plantar</td>
<td>71%</td>
<td>100%</td>
</tr>
</tbody>
</table>

“Causes” of Tarsal Tunnel Syndrome

- Post-traumatic fibrosis
- Ganglion
- Tumor
- Fracture
- Thrombophlebitis / varicosities
- Anomalous muscle - FDAL
- Rheumatoid arthritis
- Joint hypermobility / hyperpronation
### MRI in Tarsal Tunnel Syndrome

**33 cases Kerr et al 1991**

- Varicosities 8
- FHL Tenosynovitis 6
- Normal 6
- Fracture / soft tissue injury 5
- Mass lesions 5
- Fibrous scar 2
- Abductor Hallucis Hypertrophy 1
- 17 / 19 confirmed surgically

### Other Foot Neuropathies

- Medial plantar neuropathy behind the navicular tuberosity (jogger’s foot)
  - Needle exam of the flexor hallucis brevis
- Lateral plantar neuropathy near the insertion of the plantar fascia (Baxters neuropathy)
  - Needle exam of the ADQ
  - Motor latency to the ADQ

---

Thanks for being entrapped in this lecture for so long!
Peripheral Neuropathies

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ACTING ASSISTANT PROFESSOR
UNIVERSITY OF WASHINGTON MEDICAL CENTER

Objectives

• Definition
• Neurophysiology
• Evaluation of polyneuropathies
  • Cases
  • Summary of Pearls

PERIPHERAL NEUROPATHY

Definition

• **Peripheral Neuropathy:**
  • Deranged function and structure of peripheral MOTOR, SENSORY, and/or AUTONOMIC neuron
  • Involving the entire neuron or selected level

• **Polyneuropathy:**
  • Dysfunction or disease of many or all peripheral nerves

Terminology

- Neuronopathy:
  - Cell body is affected
- Peripheral Nerve Lesions:
  - Segmental Demyelination
  - Wallerian Degeneration
  - Axonal degeneration: dying back
- Axonal:
  - Loss of amplitude
- Demyelinating:
  - Increased distal latency
  - Prolonged conduction velocity
  - Conduction block
  - Temporal dispersion
Peripheral Neuropathy Evaluation

- History
  - Temporal course
  - Fiber type affected
  - Hereditary vs. Acquired (immune vs. exposure to neurotoxic agents)

- Physical exam – consider EDX as an extension of your exam
  - Pattern of the polyneuropathy
  - Underlying pathophysiology

Temporal Course

- Acute: less common
  - AIDP
  - Illness: Porphyria
  - Medications (ex. Vincristine)
  - Toxins (ex. Arsenic)
  - Vasculitis

- Chronic: most common form of presentation

Temporal Course

- Acute/Subacute Axonal PN
  - EMG: very acute
    - No denervation
    - Normal-appearing MUAPs with reduced recruitment
    - Similar to Demyelinating PN

- Very rare cases:
  - Porphyria
  - Vasculitis
  - Guillain–Barré syndrome axonal variant

Fiber Type

- Motor
  - Weakness: distribution
  - Hypo/Areflexia
  - Fasciculations
  - Cramps

- Sensory
  - Large Fibers: deficits in joint position and vibration
  - Ataxia

- Small Fibers: deficits in temperature and pain
Fiber type

- Autonomic
- Hypo/hypertension
- Urinary retention
- Changes in sweat pattern

Pearls of EDX

- Pure sensory symptoms: sensory neuronopathy
  - Acute/Subacute: paraneoplastic syndrome, post-infectious process, Sjögren's syndrome, or pyridoxine (B₆) intoxication
  - Chronic: Friedreich's ataxia
  - Pure small-fiber polyneuropathy: HIV, Diabetes, Amyloidosis, ETOH, Fabry’s dz, etc.
  - EDX: no changes if pure small-fiber involvement

Diabetes

- Numerous peripheral nervous system manifestations
  - Isolated mononeuropathies of cranial nerves (e.g., facial palsy)
  - Intercostal nerves (known as diabetic thoracoabdominal neuropathy)
  - Distal sensorimotor polyneuropathy
    - On EMG: polyradiculoneuropathy
    - Autonomic polyneuropathy or a small-fiber sensory polyneuropathy
    - Diabetic amyotrophy

Practical Approach to EDX

- Simplified NCS protocol
  - Sural Sensory
  - Peroneal motor nerve conduction study

- Most sensitive for detecting symmetrical polyneuropathy
Pattern of presentation

• Most common: distal, symmetric
• length dependent
• stocking-glove distribution
• Symptoms first present in the toes and then progress up the leg
• When the polyneuropathy reaches the upper calves, the fingertips become involved as well

Pattern of presentation

• Pearl of edx:
• Asymmetry is a key finding:
  • Mononeuropathy multiplex pattern: important to recognize (Hint: Vasculitis)
  • Superimposed radiculopathy or entrapment neuropathy
  • Variant of CIDP

Underlying Pathophysiology

• Axonal
• Demyelinating
• Mixed
• Axonal is the most common form of PN

• Borderline cases:
  • CMAP amplitudes: markedly reduced secondary to axonal loss
  • Conduction velocity: slowing secondary to severe axonal loss
  • dropout of the fastest-conducting fibers
  • How to differentiate Primary demyelinating vs. severe axonal PN?

Key Findings in Demyelination

• Prolonged distal latency
  • (>130% of the upper limit of normal)
• Slowed conduction velocity
  • (usually <75% of the lower limit of normal)
• Conduction block
• Temporal dispersion
• Prolonged or absent late responses (>130% of the upper limit of normal)
Conduction Block

- Definite: greater than 50% drop in CMAP amplitude with less than 15% prolongation of CMAP duration
- Or greater than 50% drop in CMAP area
- Or more than 30% drop in CMAP amplitude over a short segment without significant increased duration

Conduction Block

- Possible: 20-50% drop in amplitude with less than 15% prolongation of duration
- Or 20-50% drop in CMAP area
- Except for the tibial nerve

Temporal Dispersion

F-waves

- Antidromic motor and orthodromic motor
- Not a true reflex
- Activate about 1-5% of the muscle fibers
- Mild diffuse and/or proximal pathology affecting the nerve can be detected early
**AIDP**

- Most common form on North America
- Immune-mediated
- Rapidly progressive
- Predominantly motor polyneuropathy
- May lead to bulbar and respiratory compromise
- Autonomic instability
- ~65% patients have some form of illness 1-3 weeks prior to onset of sxs
  - C. Jejuni (Anti-GM1 ab)
  - Progression: 2-4 weeks
  - 50% reach nadir at 2 wks.
  - 85% by 3 wks.

**Labs:**
- Elevated CSF protein
- No cells
- Anti-GM1 ab

**Poor prognosis:**
- Age > 50-60 y/o
- Distal CMAP < 10-20% of normal
- Mechanical ventilation
- Treatment:
  - PLEX vs. IVIG
  - No indication for PLEX followed by IVIG
  - Rehab Training
  - Improvements in functional status and cardiopulmonary capacity

**AIDP: EDX**

- First few days: All NCS may be normal
- First changes in AIDP:
  - Delayed, absent, or impersistent F responses
  - Proximal demyelination → AIDP often starts at the root level as a polyradiculopathy
- Later: prolonged distal latencies, segmental demyelination (conduction block and temporal dispersion)

**CIDP**

- > 6 weeks
- Chronic progressive or relapsing polyneuropathy
- Clinically it may resemble AIDP
- Areflexia or hyporeflexia is the rule
- Treatment:
  - Corticosteroids
  - IVIG
  - PLEX
Case Presentation #1

• 21 y/o male with history of right UE weakness > LE weakness, no sensory deficits
• History?
• PE?
• EDX findings
• Treatment

HMSN (CMT)

• Genetics are heterogeneous
• CMT1, CMT3 (Dejerine-Sottas), CMT4, and some forms of CMTX: demyelinating
• CMT2, CMT5, CMTX: axonal
• CMT1A: demyelinating and the inheritance is autosomal dominant
  • 70–80% of all CMT1 cases: duplication PMP22 gene
  • Pearl: Slowing is uniform in all nerves, without evidence of temporal dispersion or conduction block

Case Presentation #2

• 19 y/o RHM, otherwise healthy, who woke up with a right wrist drop
• History?
• Physical Exam?
• Would you do an edx?
• Genetics?
Neuropathy associated with malignancy

• Direct result of the malignancy
• Secondary to the treatment: Chemotherapy and/or Radiation therapy
• Paraneoplastic syndrome
• Indirect effects of chronic illness
• Infection
• Unrelated underlying medical condition

Chemotherapy and PN

• Peripheral neuropathy is the most common clinical neurologic syndrome caused by chemotx
• Some degree tolerable
• Effect on quality of life (QOL) should not be underestimated

Chemotherapy-Induced Peripheral Neuropathy (CIPN) can be a limiting factor in rehabilitation
• Most of the agents reviewed are neurotoxic in a dose-dependent manner

Chemotherapy and PN

• Examples: Vinca alkaloids, Taxanes, Platinum agents, suramin, etc.
• Taxanes can cause neuropathy after a single dose
• Cisplatin can cause coasting: increasing symptoms for weeks after the drugs have been stopped
• Suramin: may present with AIDP

PN in Monoclonal Gammopathies

• ~10% of patients with idiopathic PN
• Presence of monoclonal gammopathy should lead to w/u:
  • Amyloidosis
  • Multiple myeloma
  • Osteosclerotic myeloma
  • Waldenström’s macroglobulinemia
  • Cryoglobulinemia
  • Leukemia
  • Lymphoma
Further Work-up after EDX*

• Highest yield laboratory tests:
  • Blood Glucose Tolerance Test
  • Vitamin B-12 with metabolites of cobalamin (methylmalonic acid with or w/o homocysteine)
  • SPEP with IFE

• Genetic Tests:
  • Highest yield: guided by the clinical phenotype, inheritance pattern, and EDX features
  • Classical hereditary neuropathy phenotype:
    • CMT1A: duplication/deletion PMP22
    • CMTX: Cx32 (GJB1)
    • CMT2: MFN2

Further Work-up after EDX*

• For Autonomic Neuropathy
  • Should be considered in the evaluation to document autonomic nervous system involvement
  • May be considered in the evaluation of patients with suspected distal SFSN
  • Currently available autonomic tests can provide indices of cardiovagal, adrenergic, and postganglionic sudomotor function

Further Work-up after EDX*

• Skin Biopsy
  • Small fiber sensory neuropathy
  • Intra-epidermal nerve density fiber
  • Quantify the myelinated Aδ fibers and the unmyelinated C fibers

Further Work-up after EDX*

• Nerve biopsy:
  • No recommendations can be made regarding the role of nerve biopsy in determining the etiology of DSP
  • Inflammatory diseases
  • Vasculitis
  • Sarcoidosis
  • Infectious disease
  • Amyloid Infiltration

*AANEM Practice Parameter: Distal Symmetric Polyneuropathy*
Summary

• Polyneuropathy
  • Temporal course: Acute vs. Chronic
  • Type: Demyelinating vs. Axonal
  • Acquired vs. Hereditary
  • High yield studies for w/u
  • Rehabilitation options
ELECTROMYOGRAPHY IN MOTOR NEURON DISEASE

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Associate Professor
Department of Neurology
2015

Outline

- Amyotrophic lateral sclerosis
- Nerve conduction studies/Electromyography
- Differential diagnosis
- Other motor neuron syndromes
  - Spinal muscular atrophy
  - Poliomyelitis

Background

- Prevalence 2 to 7 per 100,000
- Incidence 1.4 per 100,000
- Male:female ~1.6:1
- Forms/presentations
  - amyotrophic lateral sclerosis
  - progressive bulbar palsy
  - progressive muscular atrophy
  - primary lateral sclerosis
    - <1/2 of pure PLS maintain diagnosis @ mean 8 years*

*Gordon PH, March 2006

Familial forms

- 10% of all ALS
- SOD1 15-20%
- C9ORF72 40%
  - hexanucleotide repeat expansion in noncoding gene region
  - associated with FTD and TDP-43 inclusions
- TARDP 1-4%
- FUS 1-5%
Table 1 Genes known to carry ALS-causing mutations

<table>
<thead>
<tr>
<th>Gene</th>
<th>Location</th>
<th>Inheritance</th>
<th>Familial Percentage</th>
<th>Poss protein function</th>
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<td>1p36</td>
<td>AD</td>
<td>4</td>
<td>RNA metabolism</td>
</tr>
<tr>
<td>C9orf72</td>
<td>9p21</td>
<td>AD</td>
<td>7</td>
<td>Ubiquitination, autophagy</td>
</tr>
<tr>
<td>VCP</td>
<td>9p13</td>
<td>AD</td>
<td>1</td>
<td>Proteasome, vesicle trafficking</td>
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<td>QTPN</td>
<td>19q13</td>
<td>AR and AD</td>
<td>&lt;1</td>
<td>Vesicle trafficking</td>
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<td>FUS</td>
<td>16p11</td>
<td>AD and AR</td>
<td>4</td>
<td>RNA metabolism</td>
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<td>OPTN</td>
<td>10p13</td>
<td>AR and AD</td>
<td>&lt;1</td>
<td>Cytoskeletal dynamics</td>
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<tr>
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<td>12</td>
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<td>Xp11</td>
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<td>&lt;1</td>
<td>Proteasome</td>
</tr>
</tbody>
</table>

Values represent the percentage of ALS explained by each gene in populations of European ancestry. References are provided in the main text. AD, autosomal dominant; AR, autosomal recessive; XD, X-linked dominant; DENN, differentially expressed in normal and neoplasia.

Areas affected
- Upper motor neuron (UMN)
- Lower motor neuron (LMN)
- In same limb
- ALS criteria

Upper motor neuron (UMN)
- Weakness
- Increased tone
- Increased deep tendon reflexes
- Reduced speed of fine motor movements
  - Finger taps in 10 sec
  - Foot taps in 10 sec
- Pseudobulbar signs

Table 1. Summary of Revised El Escorial Criteria and Awaji Criteria for ALS Diagnosis: Differences Between Awaji Criteria and El Escorial Recommendations and the Revised El Escorial Criteria (Awaji House 1999).
**Lower motor neuron (LMN)**

- Weakness
- Atrophy
- Fasciculations
- Cramps
- Decreased deep tendon reflexes

**Atrophy – LMN sign**

**Other findings**

- Creatine kinase (CK) – elevated in 50% of patients to 2-3 x normal
  - UW lab normal value 30-285 mg/dL
  - 2-3 x normal = 570-855
- Involves select degeneration of motor cells in spinal cord, brainstem and to a lesser extent, cortex

**Pathology**

Degeneration of anterior horn cells results in denervation of muscle fibers

Collateral sprouts from surviving motor neurons reinnervate the affected motor units

Denervation atrophy with fiber type grouping
Diagnosis of ALS

- Clinical diagnosis-
  - In the absence of a biological marker to establish diagnosis, electrodiagnostics (EDX) play a critical role in the establishing the diagnosis and severity
- NCS/EMG
  - Supports lower motor neuron component
  - Excludes other etiologies
    - SUCH AS?

Nerve conduction studies

- Ulnar motor
  - Recording ADM
  - Stimulation up to axilla +/- Erb’s point
- Peroneal motor
  - Recording EDB
  - Stimulation to knee
- Sensory nerve action potentials: ulnar and sural
- Late responses – F waves
- Contralateral studies in those with upper motor neuron signs lacking

Nerve conduction

- Compound muscle action potentials-
  - normal or reduced amplitude
- Normal or only slightly reduced speed
- Minimal “demyelinating” features
  - Prolonged distal latencies (<125% ULN)
  - Reduced conduction velocities (>80% LLN)
  - Prolonged F wave latencies (<125% ULN)
- Normal sensory nerve studies

Axonal loss versus demyelination
Needle electromyography

- Fibrillation potentials and positive sharp waves widespread —
  - diffuse denervation
- Fasciculation potentials
  - motor neuron irritability in this setting, but non-specific
- Large and small fibrillation potentials
  - recent and chronic denervation

Source of spontaneous activity
Fasciculation potential

Distribution of spontaneous activity in ALS patients

Electromyography (EMG) approach

- **Limb** -
  - 3 limbs
  - Proximal and distal muscles
  - Muscles with different nerve innervation
  - Muscles with different root innervation
- Exclude structural pathology – i.e., severe cervical/lumbar spondylosis as etiology
  - Thoracic (or abdominal)
  - Cranial nerve-innervated
Non-limb EMG

- Thoracic paraspinal muscles
  - 2-3 segments
  - Avoid T11-12
  - Abnormal in 78%

- Craniobulbar
  - one pathological EMG finding in (91.7%) with both frontalis and orbicularis oris together.
  - 83.3% for frontalis alone
  - 75% orbicularis oris alone


EMG - bulbar

- Bulbar
  - At least one
  - Tongue, masseter, sternocleidomastoid, facial muscles, (trap)
  - Shorter duration MUAPs
  - Higher firing frequency

Motor unit action potentials

- Reduced in number
- **Recruit** poorly
- **Rapid** discharge
- Reduced interference pattern
- Large amplitude
- Polyphasic

Denervation/reinnervation
EMG criteria

- 3/4 regions involved
  - Bulbar
  - Cervical
  - Thoracic
  - Lumbosacral
- Active –
  - positive sharp waves
  - Fibrillation and fasciculation potentials,
- Chronic –
  - Reduced number MUAPs
  - increased amplitude/duration motor unit action potentials (MUAPs)

Diagnostic classification

- No. of Muscles Affected by Region:
  - Clinical and laboratory evidence: presence of 3 or more MUAPs in different muscles and nerves
  - Clinically definite ALS is defined by clinical and laboratory evidence: presence of 2 or more MUAPs in 3 regions, or the presence of UMN and LNN signs in at least 2 regions.
  - Clinically probable ALS is defined by clinical and laboratory evidence: presence of UMN and LNN signs in at least 1 region, or UMN signs in 1 region and LNN signs in 1 region.
  - Clinically possible ALS is defined when clinical or laboratory signs of UMN and LNN dysfunction are found in only 1 region.
  - Clinically definite ALS is defined when clinical or laboratory signs of UMN and LNN dysfunction are found in at least 2 regions.

Revised ALS Criteria:

- UMN signs: Muscles fail to move (are flaccid or spastic) and have exaggerated reflexes (hyperreflexia).
- LNN signs: Muscles become weak and atrophy (loss of muscle mass).

Muscles Affected:

- Bulbar: Face, tongue, pharynx, larynx
- Cervical: Arms, shoulders, neck
- Thoracic: Trunk, chest
- Lumbosacral: Legs, hips, buttocks

Abnormal Motor Unit Potentials (MUAPs):

- Fibrillation: Brief, spontaneous, high-frequency potentials.
- Fasciculation: Sustained, low-frequency potentials.
- Positive Sharp Waves: Short, high-amplitude potentials.

Normal Motor Unit Potentials:

-均正常

Interference pattern

- Normal MUAPs
- Neurogenic MUAPs
- Myogenic MUAPs

Reinnervation

- Normal MUAPs
- Neurogenic MUAPs
- Myogenic MUAPs
Motor unit number estimate (MUNE)
- Progressively stronger electrical shocks to nerve result in stepwise increase in amplitude of evoked compound muscle action potential (CMAP)
- Each addition = recruitment of one motor unit
- Ratio between full response from max stimulation to average increment
- Noninvasive
- Mean number differs per muscle e.g., 200 for EDB, 250 or 340 for thenar muscles

Typical cases
- Asymmetric
- Multifocal
- Involves – active and chronic
  - more than 2 muscles
  - innervated by different nerves
  - and different spinal roots
  - in at least 3 limbs (or 2 limbs and cranial)

Differential diagnosis
- Rarely
  - Lead intoxication
  - Multifocal motor neuropathy *conduction block
  - Lower motor neuron only –
    - hexosaminidase deficiency – autosomal recessive
    - Immune-mediated disease such as multifocal acquired motor axonopathy (MAMA)
  - Cervical spondylosis *root only
  - Inclusion body myositis *myopathic units

Multifocal motor neuropathy (MMN)
- Slowly progressive
- Begins distally
- Fasciculation/cramp uncommon
- <45 years
- Male:female 2:1
- More nerve than neuron distribution
- Weakness > atrophy
- Normal to decreased DTRs
MMN
- Partial conduction block

Pearls
- Most common presentation – one hand first
- Fasciculations clinically and electrophysiologically
- Start in affected area
- Include bulbar (eg genioglossus if symptomatic)
- Include thoracic paraspinals

CRDs unusual – seen with more chronic process
- Myotomal – should not spare individual nerves in the same myotome
  - Eg, if C8 innervated median muscle is abnormal, a C8 innervated ulnar muscle should also be abnormal
- Chronic myopathies, particularly inclusion body myositis
  - active denervation with long-duration, high-amplitude polyphasic MUAPs, but recruitment is usually normal or early
- Decreased activation may be seen secondary to upper motor neuron dysfunction

Summary
- “On the whole, the EMG picture of classic ALS is one of”
  - Partial denervation,
  - reinnervation,
  - decreased activation and decreased recruitment of MUAPs
  - in multiple muscles innervated by different nerves and myotomes
OTHER MOTOR NEURON SYNDROMES

Spinobulbar muscular atrophy
- Kennedy’s disease
- Onset 3rd-5th decade
- X-linked, some sporadic
- Muscle cramps with exercise
- Proximal muscles then bulbar
- Distal muscles affected later

Spinal muscular atrophy
- Prevalence of 1 in 6000 live born infants
- Majority inherited – Recessive
- Linked to locus 5q13 in > 95% of patients
- Werdnig-Hoffman – most severe resulting in death in 2 years
- Kugelberg-Welander
  - Adolescent or adult onset
  - Slowly progressive
  - Symmetrical proximal weakness
  - Lack bulbar and long-tract signs
  - Positive family history
  - DNA analysis – survival motor neuron

Kennedy’s disease
- Onset 3rd-5th decade
- X-linked, some sporadic
- Muscle cramps with exercise
- Proximal muscles then bulbar
- Distal muscles affected later
- Rest or contraction fasciculations of face, chin
- Reflexes hypoactive
- Gynecomastia in most
- Diabetes, infertility
- Mild CK elevation
- Expansion of a trinucleotide repeat (CAG) on androgen receptor gene
- NCS/EMG – chronic denervation >
Postpoliomyelitis

- Nerve conduction studies
  - Normal sensory studies; motor latencies & CV
  - Reduced amplitude compound muscle action potentials

- EMG
  - Spontaneous activity
    - Ongoing denervation
    - Chronic reinnervation
  - Fibrillation and fasciculation potentials not prominent
  - Motor unit action potentials
    - High amplitude, long duration motor unit action potentials
    - Reduced recruitment
<table>
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<tr>
<th>Nerve</th>
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<th>Amplitude</th>
<th>CV</th>
<th>F wave lat</th>
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<td>0.7/0.5</td>
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<td>2.9</td>
<td>50</td>
<td>48</td>
<td>31.7</td>
<td></td>
</tr>
</tbody>
</table>

**Muscle and side Fibs Fasci Poly Amp (mV) Dur (ms) Recruitment Comm**

- **Tibialis anterior R/L**: 2+ 1+ Rare INC+ 7-14 Mild reduced dc rlx
- **Gastrocnemius (Med).R/L**: 2+ 1+ Rare INC+ 7-14 Mild reduced dc rlx
- **Vastus lateralis R**: - - - .6-2.5 7-14 Mild reduced insr
- **Hamstring (lat).R**: - - - .6-2.2 INC Very reduced pain
- **Vastus medialis L**: - - - .6-2 7-10 Mild reduced
- **1st dorsal interosseous-R**: 4+ 1+ INC INC Very reduced
- **1st dorsal interosseous-L**: - - - .6-1.6 INC INC 3+ INC MCS
- **Pronator teres R**: 2+ 1+ Rare INC INC Mod reduced
- **Extensor digitorum communis-R**: 3+ 1+ - .6-2.2 7-14 Mod reduced
- **Triceps brachii-R/L**: - - - .6-2.2 7-12 Mild reduced
- **1st dorsal interosseous-L**: - - - .6-1.6 INC INC 3+ INC MCS
- **Pronator teres L**: 1+ - - .6-1.6 7-12 Severe
- **Extensor digitorum communis-L**: - - - .6-2.2 7-14 Severe
- **Biceps brachii-R/L**: - - - .6-2.2 7-12 Moderately reduced
- **Triceps brachii-R**: 1+ - Rare INC INC Mild reduced
- **1st dorsal interosseous-L**: - - - .6-1.6 INC INC 3+ INC MCS
- **Pronator teres L**: - - - .6-1.6 7-12 Severe
- **Extensor digitorum communis-L**: - - - .6-2.2 7-14 Severe
- **Pectoralis major-R/L**: 1+ - - .6-1.6 7-12 Severe
- **Genioglossus-L**: ? - - .6-1.6 7-12 Normal
- **Lumbar paraspinals-R**: 1+ - - .6-1.6 7-12 Severe
- **Thoracic paraspinals-R**: 2+ - - .6-1.6 7-12 Severe
- **Cervical paraspinals R**: ? ? ? no rlx

**YES – ALS!**
References

Neuromuscular Junction Testing

Neurophysiology

Electrodiagnostic Evaluation

Clinical Application

Summary

Motor Unit

Cell body
myelin
Nodes of Ranvier
synapse
NMI
Neuromuscular Junction

- Presynaptic
- Motor Nerve Terminal
- Post-synaptic
- Endplate
- Clefts

- The chemical neurotransmitter: \textbf{ACh}
- Packaged vesicles in the presynaptic terminal: quanta
- Each quantum: \sim 10,000 molecules of ACh

Presynaptic Site

- Nerve Action Potential
- VGCCs activated
- Influx Ca++
- Release of ACh

Neuromuscular Junction

- Pre-synaptic terminal
- Quanta
- Post-synaptic membrane (muscle)
- AChR
- Action Potential
Presynaptic Site

- Quanta:
  - Primary (immediately available store): 1000 quanta
  - Secondary (mobilization store): 10,000 quanta
  - Tertiary (reserve store): more than 100,000 quanta

Neuromuscular Junction

- ACh diffuses across the synaptic cleft
- Binds to ACh receptors → on the postsynaptic muscle membrane
- Postsynaptic membrane
  - Junctional folds (increasing the surface area of the membrane)
  - AChRs clustered on the crests of the folds

Post-synaptic Membrane

- Binding of ACh to AChRs opens Na+ channels → local depolarization (endplate potential (EPP))
- Size of the EPP is proportional to the amount of ACh that binds to the AChRs
- ACh broken down by Acetylcholinesterase
- EPP depolarizes the muscle membrane → all-or-none muscle fiber action potential
- EPP always rises above threshold
- Safety Factor: amplitude of the EPP above the threshold value needed to generate a muscle fiber action potential

Post-synaptic Membrane

- Resting State
- Random release of quanta producing the miniature end plate potentials (MEEP)
- Sea shell noise
EMG correlation

- MEEP (miniature end plate potential):
  - Initial deflection: negative
  - Amplitude: 10-50 µV
  - Rhythm: irregular
  - Rate: high frequency (150 Hz)

- Endplate Spikes:
  - Initial deflection: negative
  - Amplitude: 100-300 µV (<1 mV)
  - Rhythm: irregular
  - Rate: 50-100 Hz

ELECTRODIAGNOSTIC STUDIES

Slow RNS

- RNS (2–3 Hz) – normal physiology
  - ACh quanta are progressively depleted
  - EPP falls in amplitude, but because of the normal safety factor, it remains above threshold
  - After the first few seconds, the secondary store begins to replace the depleted quanta

Rapid RNS

- RNS (10–50 Hz) - normal physiology
  - Depletion of quanta from the presynaptic terminal is counterbalanced by:
    - mobilization of quanta from the secondary store
    - accumulation of calcium
  - Ca++ accumulates in the presynaptic terminal → increased release of quanta → higher EPP → end result is the same as with any other EPP above threshold: an all-or-none muscle fiber action potential
  - Effect of Mg++ → Opposite to Ca++ → Competes with Ca++
Exercise Testing

• Voluntarily contraction at maximum force
• Motor units fire at their maximal firing frequency → typically 30 to 50 Hz
• We use it as it demonstrates many of the same effects as rapid (30–50 Hz) RNS
• Both result in higher-amplitude EPPs

Post tetanic facilitation

• Voluntary contraction 10-20 seconds
• Or stimulation at 50 Hz for 30 seconds
• Improvement of CMAP amplitude in MG after exercise
• Post tetanic exhaustion is less well understood

Lower Temperature...

• EPP duration and amplitude increases
• AChR remain open longer
• AChE slow down
• Safety factor increases

RNS is pathological states

• If safety factor is reduced...
• Slow RNS → depletion of quanta → drop the EPP below threshold → absence of a muscle fiber action potential
RNS is pathological states

- In conditions where baseline EPP is below threshold (fiber action potential is not generated)
- Rapid RNS may increase the number of quanta released, resulting in a larger EPP, so that threshold is reached

Neuromuscular Junction

**Pre-synaptic pathologies**

**Post-synaptic pathologies**

Presynaptic

- Blocking Agents
  - LEMS
  - Botulism
  - Mg++/Ca++ deficiency
  - Aminoglycosides
  - Procainamide

Enhancing Agents

- Ca++
Postsynaptic

- Blocking:
  - Myasthenia Gravis
  - Organophosphate poisoning
  - Aminoglycosides
  - Procainamide
  - Xs neostigmine

- Enhancing:
  - Edrophonium
  - Neostigmine
  - Succinylcholine

Myasthenia Gravis

- Characterized by weakness and fatigability
- Reduction of AChR
- Usually sporadic
- Hyperplasia thymus: 60-80% MG pts
- Generalized form: ~80% ab positive
  - 20% ab negative: about 50% are MUSK ab positive

Myasthenia Gravis

- Complement binds to the Antibody-AChR complex
- Membrane-attack complex (MAC) forms on the membrane
- The post-junctional membrane is destroyed

http://neuromuscular.wustl.edu/mtime/modulation.htm#complement
RNS in MG

- Decrement between the first and fourth potentials
- After the fourth potential, the decrement is not as marked and forms a “U shape”
- Decrement begins to improve when the mobilization store begins to resupply the immediately available store
- Muscles to target for test:
  - ADM/Trap/Nasalis or Frontalis

Single Fiber EMG

SFEMG

LEMS

- Paraneoplastic or autoimmune
- ~3% of patients with small cell CA of lung will develop LEMS
- Over ½ of patients with LEMS will have tumor
- Ab to VGCC in 85% pt
  - CA++ entry to presynaptic terminal is decreased
NCS in LEMS

- Important: LOW CMAP amplitudes in NCS
- > 100% increase in amplitude after short exercise test/ Rapid RNS
- Seen in all muscles

Botulism Toxin

Summary
Myopathies

Arthur Rodriguez, MD, MS
Emeritus Associate Professor
Rehabilitation Medicine
University of Washington

Myopathy - Symptoms

- Proximal Weakness
  - arising from chair, stair climbing
  - brushing hair
  - lifting head off pillow
- Fatigue
- Atrophy
- Muscle Pain

Myopathy History

- Other Medical History
  – connective tissue disease, cancer
- Family History
- Toxic Exposure
- Statin Therapy

Myopathies - Signs

- Strength
  – proximal weakness (mostly)
  – scapular winging
  – neck, spine weakness
- Gait
  – Gower’s Sign
  – excessive lordosis
  – genu recurvatum
  – Trendelenburg sign
Myopathies - Signs

- Myotonic Dystrophy
  - facial weakness, frontal balding, temporalis muscle wasting
  - percussion myotonia
- Dermatomyositis
  - rash

Myopathies - Signs

- What should not be seen in pure myopathies?
  - Sensation - usually normal
  - Reflexes - usually preserved early on
  - Fasciculations - not seen

Myopathies - Laboratory Tests

- Serum Creatine Kinase
  - upper normal varies from 200 - 500
  - depends upon lab, gender, race
  - can see up to ~1000 in denervating diseases
  - over 1000 suggests muscle disease
- AST, LDH, aldolase can also be elevated
  - less sensitive than CK
  - also elevated in liver disease

Role of Electrodiagnosis

- Confirmation
- Exclusion
- Localization
- Severity
- Pathophysiology
- Prognosis/Response to therapy
Electrodiagnostic Approach to Myopathies

- Sensory Nerve Conduction
  - should be normal
  - if abnormal, consider other disease process

- Motor Nerve Conduction
  - velocity should be near normal
    - if not, consider peripheral nerve disease
  - amplitude can be reduced in myopathies
    - but also in axonal neuropathies, NMJ disease

Electrodiagnostic Approach to Myopathies

- If NMJ disorder is suspected, then do repetitive stimulation studies.
  - Usually normal in myopathies
  - Some myotonic conditions do have a decrement

Normal MUAP

At Rest
Myopathies - Spontaneous Activity

Fibrillation Potentials

Complex Repetitive Discharge

Complex Repetitive Discharges
Complex Repetitive Discharge

- Seen in chronic myopathies or neuropathies
- Due to ephaptic transmission between muscle fibers. Pacer cell.
- Similar to cardiac re-entry phenomenon
- Constant discharge, sudden on-off
- Sounds like machinery

Myopathies - Other Spontaneous Activity

- Myotonia
  - originate from single muscle fibers
  - look like fibrillations or positive sharp waves
  - due to abnormal Cl conductance
  - wax and wane in frequency and amplitude
  - sound like dive bomber or revving motorcycle

Myotonia in Action
Tora Tora Tora

Myotonia
Myopathic MUAPs

- Reduced Motor Unit Territory
  - fewer muscle fibers per motor unit
  - temporal dispersion along muscle fibers
  - less force per MUAP
- On EMG, one sees
  - small amplitude, short duration
  - polyphasic
  - early recruitment

Recruitment: The Orderly Activation of Motor Units to Increase Muscle Tension

- Spatial
- Temporal
Measuring MUAPs

- **Duration**
  - most reliable
  - more difficult to measure
- **Amplitude**
  - easy to measure
  - depends upon needle position
- **Phases**
  - non specific

Quantitative EMG

- Best way to measure duration
- Concentric Needle
- 2 Hz - 10 kHz filters
- 20 different average MUAPs
- exclude satellites
- get mean duration

Interference Pattern Analysis

- Limb Girdle Muscular Dystrophy
- Vastus Medialis
- Biceps brachii

Specificity of EMG

- EMG can be diagnostic of myopathy but is rarely specific as to type of myopathy
  - exceptions exist, e.g. myotonia
- Specific diagnosis usually dependent upon combination of clinical presentation, lab data, biopsy, and EMG.
**Hereditary Myopathies**

- **Duchenne and Becker**
  - normal motor and sensory NCS
  - fibs and psw’s (Duchenne > Becker)
  - small MUAPs
  - early recruitment
  - some abnormalities in carriers, but not sufficient for reliable identification

- **Limb Girdle Muscular Dystrophy**
  - a number of distinct entities grouped together
  - normal motor and sensory NCS
  - fibs and psw’s
  - mixture of small and normal MUAPs
  - +/- early recruitment

- **Facioscapulohumeral Dystrophy**
  - normal motor and sensory NCS
  - small amplitude CMAPs from atrophied muscles
  - fibs and psw’s less prominent
  - small MUAPs
  - early recruitment
  - may initially present asymmetrically

- **Myotonic Dystrophy**
  - normal motor and sensory NCS
  - small amplitude CMAPs from atrophied muscles
  - decrements to repetitive stimulation
  - fibs and psw’s (distal > proximal)
  - myotonia (distal > proximal)
  - small MUAPs (not in myotonia congenita)
  - early recruitment
  - may be associated with a polyneuropathy
<table>
<thead>
<tr>
<th>Hereditary Myopathies-Mitochondrial Myopathies</th>
<th>Hereditary Myopathy-Myotubular Myopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>• A group of myopathies with both maternal mitochondrial or mendelian inheritance</td>
<td></td>
</tr>
<tr>
<td>• Often multi system disease</td>
<td></td>
</tr>
<tr>
<td>• Often ragged red fibers on trichrome stain</td>
<td></td>
</tr>
<tr>
<td>• Often with ophthalmoplegia (confused with Myesthenia)</td>
<td></td>
</tr>
<tr>
<td>• EMG findings are usually minimal with early recruitment and short duration, low amplitude MUAP’s</td>
<td></td>
</tr>
<tr>
<td>• Infantile x linked severe form</td>
<td></td>
</tr>
<tr>
<td>• Juvenile autosomal recessive form</td>
<td></td>
</tr>
<tr>
<td>• Milder autosomal dominant</td>
<td></td>
</tr>
<tr>
<td>• EMG- polyphasic low amplitude MUAP,s fibs and pos sharp waves and CRD,s (the only congenital myopathies with spontaneous activity)</td>
<td></td>
</tr>
<tr>
<td>• Myotonic like discharges may suggest myotonic dystrophy</td>
<td></td>
</tr>
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</table>

<table>
<thead>
<tr>
<th>Inflammatory Myopathies</th>
<th>Polymyositis - Dermatomyositis</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Idiopathic</td>
<td></td>
</tr>
<tr>
<td>– polymyositis, dermatomyositis, inclusion body myositis</td>
<td></td>
</tr>
<tr>
<td>• Infectious</td>
<td></td>
</tr>
<tr>
<td>– HIV, Influenza, Hep B, Hep C, other viruses</td>
<td></td>
</tr>
<tr>
<td>• Bacterial (Strep, Staph, Yersinia)</td>
<td></td>
</tr>
<tr>
<td>• Fungal</td>
<td></td>
</tr>
<tr>
<td>• Parasites (Toxo, Trichinosis, Cestodes - tapeworms)</td>
<td></td>
</tr>
<tr>
<td>• Proximal &gt; Distal Weakness, muscle pain</td>
<td></td>
</tr>
<tr>
<td>– dysphagia, dyspnea, arrhythmias</td>
<td></td>
</tr>
<tr>
<td>• Increased CK, usually 5 - 50 fold increase</td>
<td></td>
</tr>
<tr>
<td>– SGOT, SGPT, LDH, aldolase also increased</td>
<td></td>
</tr>
<tr>
<td>• Biopsy - endomysial inflammation, segmental necrosis</td>
<td></td>
</tr>
<tr>
<td>• Dermatomyositis (a vasculitis) - heliotrope rash</td>
<td></td>
</tr>
</tbody>
</table>
Polymyositis - Dermatomyositis

- Needle EMG demonstrates
  - psw’s and fibs, proximal > distal muscles
  - paraspinals most sensitive (thoracic good to test)
  - most patients have them
  - reflect severity of inflammation
  - reduced after steroids
- CRDs
  - typical “myopathic” MUAPs, early recrt.
  - EMG one side, biopsy mod involved contralateral muscle

Inclusion Body Myositis

- Usually >50y/o, M>F
- Weakness proximal = distal
  - finger and wrist flexors, knee extensors
  - may present asymmetrically
- CK only mildly increased (<10 x normal)
- Less responsive to any treatment-a degenerative rather than immune disorder
- EMG similar to DM-PM but, less psw’s & fibs, mixed large and small MUAPs.

IBM Patients Mimicking ALS

- 9/70 IBM patients initially diagnosed with ALS in Columbia University series (Dabby R. et al., Archives of Neurol, 2001)
- Fasciculation potentials in 7 and long duration MUAPs seen in 8
- Quantitative motor unit analysis helped confirm myopathy in 4/5 patients restudied

Critical Illness Myopathy

- Probably more common cause of ICU weakness than Critical Illness Polyneuropathy
- More likely in patients who receive steroids or non-depolarizing NMJ blockers
- Severe generalized weakness over several days
- Recovery occurs slowly over several months
Critical Illness Myopathy

- Normal SNAPs (unless CIP co-exists)
- Small or absent CMAPs
- Diffuse fibs/psw’s
- Short duration, small MUAPs expected
- Difficult to recruit
- Direct muscle and nerve stimulation both show small responses (research tool)

Myopathy - Summary

- Important to complete thorough H&P
- Examine one side
- Do proximal muscles
- Specific diagnosis depends upon clinical history, lab values, biopsy, genetic testing and EMG

Summary

- Normal SNCV, Possibly small CMAP’s in weak muscles, Normal RNS
- Early recruitment in weak muscles
- Short duration MUAP’s when complex, polyphasic MUAP are excluded
- Fibs/PSW’s most characteristic of inflammatory myopathies, inclusion body myositis, critical illness and a few metabolic and congenital myopathies.

Summary

- Expect occasional larger amplitude, polyphasic MUAP’s and occasional late components.
- Myotonic like discharges and myotonia in the inflammatory myopathies, myotonic dystrophy, myotubular myopathy, hyperkalemic periodic paralysis and chloroquine myopathy
Summary

- Pattern of EMG changes may suggest the etiology (i.e. predominant involvement of deep forearm flexors in IBM, myotonic dystrophy)
- Sensory nerve conduction abnormalities uncommon but suggest a specific cause (e.g. IBM, alcoholic, critical illness, or paraneoplastic) or unrelated neuropathy
- Mixed neurogenic and myopathic changes on needle EMG also suggestive of IBM, myofibrillar myopathies, and other specific causes

Question

- You find fibrillations in a patient in whom you are evaluating for possible myopathy. You start thinking that:
  a. This isn’t a myopathy
  b. This is steroid myopathy
  c. This is polymyositis
  d. This is more likely a neuropathy

Question

- In a patient with critical illness myopathy, motor nerve conduction would most likely show:
  a. Marked slowing in CV
  b. Prolonged distal latency
  c. Reduced CMAP amplitude
  d. Increased temporal dispersion
SUMMARY OF SPONTANEOUS POTENTIALS

MINIATURE ENDPATE PLATE POTENTIALS
"ENDPLATE NOISE"
"MEPP"

ID - NEGATIVE
DUR - 0.2-1 MSEC
AMP - 10-100 uV
RA - 150 Hz
RHY - IRREGULAR
DS - NORMAL
O - PREJUNCTIONAL

ENDPLATE POTENTIALS
"EPP"

ID - NEGATIVE → POSITIVE
DUR - 2-4 MSEC
AMP - 100-300 uV
RA - 50-100 Hz
RHY - IRREGULAR
DS - NORMAL
O - PREJUNCTIONAL

FIBRILLATIONS

ID - POSITIVE → NEGATIVE
DUR - 0.5-5 MSEC
AMP - 50-1000 uV
RA - 1-50 Hz
RHY - REGULAR
DS - ABNORMAL
O - PREJUNCTIONAL

<table>
<thead>
<tr>
<th>ID</th>
<th>DUR</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>ABSENCE</td>
</tr>
<tr>
<td>1+</td>
<td>EQUIVOCAL</td>
</tr>
<tr>
<td>2+</td>
<td>PERSIST OVER 1 SEC IN 2 AREAS</td>
</tr>
<tr>
<td>3+</td>
<td>PERSIST OVER 2 SEC IN 3 AREAS</td>
</tr>
<tr>
<td>4+</td>
<td>INTERMITTENT IN ALL AREAS</td>
</tr>
<tr>
<td>5+</td>
<td>CONTINUOUS IN ALL AREAS</td>
</tr>
</tbody>
</table>

POSITIVE WAVES

ID - POSITIVE → NEGATIVE
DUR - 5-100 MSEC
AMP - UP TO 1 mV
RA - 2-50 Hz
RHY - REGULAR
DS - ABNORMAL
O - PREJUNCTIONAL

REPEATED DISCHARGES

a) Myotonia

ID - BIPHASIC: POS. NEG.
MONOPHASIC: POS.
DUR - POS.-NEG. SPIKE=5 MSEC.
POS.=5-20 MSEC
RA - 20-80 Hz
RHY - WAX & WANE IN RATE & AMP
O - POSTJUNCTIONAL
DS - ABNORMAL

b) Complex & Other

ID - POSITIVE OR NEGATIVE
CAN LOOK LIKE:
Normal MUP
Polys
Fibs
+ Waves
RA - 1-100 Hz
RHY - REGULAR
DS - ABNORMAL
O - PREJUNCTIONAL

c) Myokymia

ID - NEGATIVE
DUR - SAME AS MUP
AMP - 1) GROUPS AT 20-60 Hz OR
2) CONTINUOUS 1/10 sec.-10/sec.
RA - 2/3 SEC
RHY - REGULAR
DS - NORMAL OR ABNORMAL
O - PREJUNCTIONAL

FASICULATIONS

ID
DUR
SAME AS MUP
ONLY 10-20 % POLY
AMP
RA - 2/min. - 2/sec.
RHY - USUALLY IRREG.
DS - BASED ON "COMPANY THEY KEEP" - ABNORMAL
O - PREJUNCTIONAL

GRADING

<table>
<thead>
<tr>
<th>ID</th>
</tr>
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<tbody>
<tr>
<td>0</td>
</tr>
<tr>
<td>+</td>
</tr>
<tr>
<td>1+</td>
</tr>
<tr>
<td>2+</td>
</tr>
<tr>
<td>3+</td>
</tr>
<tr>
<td>4+</td>
</tr>
</tbody>
</table>

ARTIFACTS

60 CYCLE
DEFFECTIVE NEEDLE
AUDIOFEEDBACK
BUMPING NEEDLE
WANDERING BASELINE

* ID = INITIAL DEFLECTION
DUR = DURATION
AMP = AMPLITUDE
RA = RATE
RHY = RHYTHM
DS = DIAGNOSTIC SIGNIFICANCE
O = ORIGIN
SPONTANEOUS POTENTIALS

Normal: End plate activity
1) MEPP
2) EPP
3) EP Spikes

Abnormal:
1) Fibrillations
2) Positive Waves
3) Repetitive discharges
   a) Myotonia
   b) Low and high frequency
   c) Myokymia
4) Fasiculations
5) Continuous
   a) Neuromyotonia
      (Isaacs syndrome)
   b) Stiff man syndrome
      (Moersch, Waltman)
   c) Cramps

Artifacts

General Characteristics
Normal potential
Initial negative deflection
are irregular

Abnormal
Initial positive deflection
are regular
**MEPP** *(Miniature end-plate potentials)*

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Initial deflection</th>
<th>Duration</th>
<th>0.2-1 msec</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amplitude</td>
<td>10-100 uV</td>
<td>Rate</td>
<td>150 sec.*</td>
</tr>
<tr>
<td>Rhythm</td>
<td>Irregular</td>
<td>Diagnostic significance</td>
<td>Normal</td>
</tr>
<tr>
<td>Origin</td>
<td>Prejunctional</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Some believe these potentials occur closer to 20-40 Hz. Moreover, a single source of MEPP generate these potentials at 3-5/sec.*

**Physiology:**
The source of an MEPP is a single quanta of ACH, which releases 10,000 molecules of ACH. Since about 100 quanta of ACH are needed to generate a propagated potential, the MEPP's are non-propagated.

**Differential:**
Background voluntary contraction.
**EPP (Endplate Potentials) and EP Spikes**

**Characteristics:**
- Initial deflection: Negative positive
- Duration: 2-4 msec.
- Amplitude: 100-300 uV
- Rate: 50-100 Hz
- Rhythm: Irregular
- Diagnostic Significance: Normal
- Origin: Prejunctional

**Physiology:**
1) EPP classically described by Katz are non-propagated. There is a rapid decay of the potential a short distance from the endplate. This is due to subliminal depolarization of the endplate but of greater magnitude than MEPP.

2) Endplate spikes are propagated and due to:
   1) Needle irritation of the muscle fibers or small intramuscular nerves (Eisen pg. 28 AAEE Course A 1984)
   2) Synchronization of MEPP of a significant magnitude to generate a propagated potential (Eisen p. 28)
   3) Final possibility is they originate from intrafusal muscle fibers (Partanen et. al. Neurology 1983; 33:1039-1043).

**Diagnostic Significance:**
1) Normal but
2) EP activity absent in totally denervated muscle.
3) Increase in partially reinnervated muscle (Buchthal in Culp and Ochoa p. 647)

**Differential:**
Voluntary motor unit potentials (MUP) do not depolarize at the rapid rates and are not as irregular as EPP. Moreover, MUP disappear when the needle is moved a fraction. Moreover, EPP do not disappear with contraction of antagonists.

EPP are sometimes difficult to differentiate from fibrillations but this will be covered later.
FIBRILLATIONS

Characteristics:
- Initial deflection
- Duration
- Amplitude
- Rate
- Rhythm
- Diagnostic Significance
- Origin

*Rate - Grading
- Absence
- Equivocal
- Persist over 1 sec. present in 2 areas
- Moderate number in equal to or > 3 areas Discontinuous in all areas
- Continuous in all areas

**Regularity
- Buchthal says 1/2 fibs regular and the other 1/2 are irregular.

Differential:
- i.e. helpful hints
- Differentiate fibrillations from EPP:
  - Some EPP's have an initial positive deflection because they are recorded away from the endplate zone. Secondly, coaxial needle electrodes may record EPP's from the shaft of the electrodes and they would then have an initial positive deflection. But a potential is likely to be an EPP if:
    1) The initial positive deflection disappears with the slightest movement of the electrode
    2) MEPP's are found very close to the potential
    3) The rate is fast
    4) The rhythm is irregular

Also, a potential is likely to be a fibrillation even if the initial deflection is negative and:
- 1) The initial deflection becomes positive with a slight movement of the electrode
- 2) The potential is 0.5 to 2 msec. in duration.
- 3) The rate is 1-10 Hz
- 4) Rhythm is regular
## POSITIVE WAVES

### Characteristics:
- **Initial deflection**
- **Duration**
- **Amplitude**
- **Rate**
- **Rhythm**
- **Diagnostic Significance**
- **Origin**

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Value</th>
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<tbody>
<tr>
<td>Positive</td>
<td>5-100 msec.</td>
</tr>
<tr>
<td>Usually &lt; 30 msec.</td>
<td></td>
</tr>
<tr>
<td>Initial deflection</td>
<td>usually &lt; 5 msec.</td>
</tr>
<tr>
<td>Up to 1 mV</td>
<td></td>
</tr>
<tr>
<td>usually 100-400 μV</td>
<td></td>
</tr>
<tr>
<td>2-50 Hz</td>
<td></td>
</tr>
<tr>
<td>Regular</td>
<td></td>
</tr>
<tr>
<td>Abnormal</td>
<td></td>
</tr>
<tr>
<td>Postjunctional</td>
<td></td>
</tr>
</tbody>
</table>

### Physiology:
Positive waves originate adjacent to depolarized, damaged area of a muscle fiber.

### Grading:
- **Absence** 0
- **Equivocal** ±
- **Persist over 1 sec.** +1
- **Present in 2 areas** +1
- **Moderate number in equal to or > 3 areas** +2
- **Discontinuous in all areas** 3+
- **Continuous in all areas** 4+
REPETITIVE DISCHARGES

High and low frequency complex repetitive discharges

Characteristics:
Initial deflection
Duration
Amplitude
Rate:
   Low frequency 1-10/sec.
   High frequency 10-100
Rhythm: Regular
Origin: Postjunctional

Like a polyphasic motor unit potential*

*Physiology:
These potentials could occur in a split muscle fiber, the components of which share a common membrane. They arise postjunctionally; one branch of the split muscle fiber depolarizes an adjacent branch of the same split muscle fiber. This is called ephaptic conduction.

Diagnostic Significance:
Most frequently seen in chronic diseases where fiber splitting becomes prominent.

Motor neuron disease
Paraspinals in chronic radiculopathies
Acid Maltase Deficiency
Myopathies

Differential:
These potentials are not myotonic. Myotonic wax and wane.
MYOTONIA

Characteristics:
- Initial deflection
- Biphasic spike:
  - Positive → Negative
- Needle induced → Monophasic:
  - Positive waves
  - Positive → Negative
  - Spike = 5 msec.
  - Positive = 5–20 Hz
- Duration
- Rate
- Rhythm
- Wax and wane in rate and amplitude

Diagnostic Significance
- Myotonia
- Congenita (Thomsens)
- Dystrophy (Steinert)
- Paramyotonia
- Dermatomyositis
- Muscle trauma
- Hyperkalemia
- Hyperthyroidism
- Acid Maltase deficiency
- Diazocolesterol

Myotonic discharges are not a consistent feature

Origin
- Postjunctional
FASCICULATIONS

Characteristics: Initial deflection Duration Amplitude

Rate - see below*
Rhythm - Usually irregular
Diagnostic significance:** (Abnormal but this is based on the company they keep)

Origin - Prejunctional

*Rate i.e., Grading Fasciculations:
1+ = 2 areas; 2-10/min.
2+ = many areas; 10-15/min.
3+ = all areas; 50-100/min.
4+ = all areas; >100/min.

**Diagnostic Significance:
ALS
Primary muscular atrophies
Syringomyelia

Root lesions
Peripheral neuropathies

Thyrotoxic myopathy
Polymyositis

Physiology: Origin from the anterior horn cell and more distal axon.

ARTIFACTS

60 cycle
Needle artifact
Audio feedback
Bumping the needle
Wandering baseline
STUDY STRATEGIES

Karen Wooten, M.D.
March 17, 2015

original slides by Rina Reyes

CERTIFICATION

“As part of the requirements for certification by the ABPMR, candidates must demonstrate satisfactory performance in an examination conducted by the Board covering the field of PM&R.”

- Certification Booklet of Information
- www.abpmr.org
- As of 2014: 11,433 diplomates

2014 Data

- Part I- 479 candidates (395 1st time)
  - 78% overall pass rate
  - 91% pass for first time candidates
  - 31% pass for repeat takers
- Part II- 386 candidates (335 1st time)
  - 80% overall pass rate
  - 86% for first time candidates
  - 55% for repeat takers

SAE exam results are a generally a good predictor of board scores
2013 Pass Rates: Subspecialty Exams

<table>
<thead>
<tr>
<th>Subspecialty</th>
<th>1st Time</th>
<th>Repeat</th>
<th>MOC/Other</th>
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</thead>
<tbody>
<tr>
<td>SCI</td>
<td>86%</td>
<td>50%</td>
<td>74%</td>
</tr>
<tr>
<td>Pain</td>
<td>99%</td>
<td>100%</td>
<td>82%</td>
</tr>
<tr>
<td>Pediatrics</td>
<td>100%</td>
<td></td>
<td>90%</td>
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<tr>
<td>Sports Med</td>
<td>60%</td>
<td>40%</td>
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<tr>
<td>Hospice/Palliative</td>
<td>ABPMR 76%</td>
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<tr>
<td>Neuromuscular</td>
<td>ABPMR 37%</td>
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<tr>
<td>Brain Injury</td>
<td>pending</td>
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</table>

2014 MOC Data

Time-limited certificates since 1993; valid for 10 years
- 97% First time pass rate
- 510 examinees
- Four components (evolving frequently):
  - Professional standing (maintain license)
  - Lifelong learning & Self-assessment: (300 CME/10yr)
  - Cognitive expertise: 160 7s, computer based testing
  - Practice Performance Project

PART I

- Questions “are designed to test the candidate’s knowledge of basic sciences and clinical management as related to PM&R and will be in the form of objective testing.”
- Certification Booklet of Information
- Examination Content Areas Outlined in appendix

PART I – August 17, 2015

- Designed to show your fund of knowledge
- 7 hours total with two 3-hour blocks
- 325 multiple-choice questions; closed book
  - 165 + 160
- Local computer-based exam (Pearson VUE Professional Centers-
  http://www.pearsonvue.com/abpmr/)
- Demo tutorial available online/pre-test (excludes exam time)
- You can look at any question in the 3 hour block, but not between sessions
PART I

- Questions-review pamphlet from the ABPMR for structure (a-d multiple choice)
- Content Areas- Look at website for exam outline and target weights
- Questions written at the level of a rehab textbook

PART I

- Different version each year
- Scaled scores, based on prior (1998) exam
- Scores are structured to “reveal, by content area, candidates’ strengths and weaknesses”

PART I EXAM OUTLINE

Two Independent Content Domains

- Type of Problem/Organ System
  - 30% Neurologic Disorders
  - 32% Musculoskeletal Medicine
  - 5% Amputation
  - 8% Cardiovascular and Other Systems
  - 15% Rehab Problems & Outcomes
  - 10% Basic Sciences
- Focus of Question/Patient Management
  - 31% Patient Evaluation & Diagnosis
  - 15% Electrodiagnosis
  - 32% Patient Management
  - 10% Equipment & Assistive Technology
  - 12% Applied Sciences

Part 1 Preparation

- Review topic list
  - Ensure that you have had exposure to each one
  - Strategize based on topic weights
- Read a general textbook (Braddom or DeLisa); “Board Review” Books (Cuccurullo); none specifically endorsed by ABPMR
- SAE to survey your strengths/weaknesses and practice answering multiple choice questions
- Study guides
- Study groups, Review Course
- Evaluate your program’s historical areas of strength, weakness
- Expect multimedia (images, tables, graphs, video)
Part 1: General Test-taking Strategies

- Look for salient points of question
- Read and understand the true question or final sentence carefully (e.g., correct vs. incorrect answer)
- Look for lead-in statement in stem
- Clarifies the task
- Qualifiers (next, immediately, initially)
- Caution with answers containing absolutes or exclusionary words

Part 1: General Test-taking Strategies

- Try answering the question without looking at the answers
- Look for mutually exclusive answers to cut down the potential options
- Time management—go through all the questions and answer the easy ones first

Other Clues: Use only if you’re stuck!

- Uneven length of options
- Cues of grammar
- Non-random or illogical placement of options

Note: Multilevel review guards against this possibility

PART II: Purpose

- “Candidates will be expected to present, in a concise, orderly fashion, evidence of their proficiency in the management of various clinical conditions within the field of PM&R. During the oral examination, the examiner will ask questions about diagnostic procedures, therapeutic procedures, and patient management.”

- Certification Booklet of Information
### PART II

- "It is the intention of the ABPMR to use the oral examination to assess different areas of physiatric competence than the Part I exam."
- NOT A KNOWLEDGE TEST
- "A candidate must have competency in medical knowledge to address these questions, but the vignettes were not designed to ask for a recitation of facts."
  - "Diplomate News, Summer 2005, Vol. 12, No. 1

### PART II: Enhancements 2005

Candidate presented with at least 3 standardized clinical case vignettes developed by the Oral Examination Committee.

17 possible categories; vignette may include more than 1 category

### PART II: VIGNETTES

- Examiners extensively trained on the vignettes and scoring system.
- Independently rated on each skill
- Statistically adjusted for the severity of examiners, difficulty of vignettes, difficulty of clinical skills
VIGNETTES

- Examiner presents brief case description
- Further details provided as relevant or elicited
- Opportunity to demonstrate clinical skills, integration, apply professional knowledge, interactive skills, standards
  - Part II Certification Examination Information for Candidates, 2009 Exam (abpmr.org)
- Learn to think out loud, be able to discuss your rationale for management, thinking process
  - *Acquiring, not losing, points*

PART II: VIGNETTES
Test Areas/Clinical Skills

Independently rated:
- Patient Care:
  1) Data acquisition
  2) Problem solving
  3) Patient evaluation & management
- 4) Systems-based practice: knowledge of practice and delivery systems,
- 5) Interpersonal and communication skills: professionalism; potentially role-playing

PART II SCORE

- Composite, scaled scores 1-10 (mean in 2005 was 5.96)
- Pass/Fail determined by performance in five domains, not on content knowledge of specific clinical disorders.

PART II: THE EXAM

- Brief, written description of case
- Examiner guides you through 5 skills by asking general questions, adding further details
- Responses should be efficient, focused, but demonstrate your thinking process
### PART II: SKILL AREAS

<table>
<thead>
<tr>
<th>Data Acquisition</th>
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<tbody>
<tr>
<td><strong>Goal:</strong> elicit appropriate information to accurately generate differential diagnosis and management</td>
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<thead>
<tr>
<th>Problem Solving</th>
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<tr>
<td>Integrate medical knowledge, data</td>
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<tr>
<td>Prioritize rehab goals and medical issues</td>
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<td>Differential diagnosis, leading diagnosis</td>
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<td>Explain why R/O or R/I.</td>
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<td>Expound upon why</td>
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<td>What diagnostic information you need</td>
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<td>Use of Evidence Based Medicine (EBM)</td>
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<td><strong>Goal:</strong> Organized approach to data collection; propose reasonable leading diagnosis</td>
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<th>Patient management</th>
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<tr>
<td>Medical management</td>
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<tr>
<td>Prioritize medical vs. rehabilitation goals</td>
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<tr>
<td>Rx: exercises, modalities, DME</td>
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<tr>
<td>Diagnostic/therapeutic injections</td>
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<tr>
<td>Use of EBM</td>
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<tr>
<td>Comprehensive, therapeutic care plan, F/U</td>
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<td>Health promotion, disease prevention</td>
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<td><strong>Goal:</strong> Efficiently develop informed, appropriate treatment plan at right time.</td>
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<th>Systems-Based Practice</th>
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<tr>
<td>Practice and delivery systems</td>
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<tr>
<td>Integrating management into larger system of care</td>
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<tr>
<td>Outcomes</td>
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<tr>
<td>Risks/benefits</td>
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<tr>
<td>Limitations</td>
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<tr>
<td>Costs of resources, resource utilization</td>
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<tr>
<td>Patient advocacy</td>
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<tr>
<td>Consultation of other services</td>
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<td>Team management</td>
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<tr>
<td>Quality of care</td>
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<tr>
<td><strong>Goal:</strong> Make appropriate decisions regarding resource utilization, requesting consultations; considered risks, patient safety, care quality; advocates appropriately for patient needs</td>
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<th>Patient safety</th>
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<tr>
<td>Risk Management</td>
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<td>IP Rehab candidacy</td>
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PART II: SKILL AREAS
- Interpersonal and Communication Skills
  - Patients, families, other providers
  - Compassion, listening skills, sensitivity
  - Key: get feedback about your communication style; use patient-centered language
  - Goal: Respond with sensitivity, sound ethics, professionalism; be able to provide appropriate explanation to patient/family; provide opinion to consultants, effectively communicate with staff

PART II: Strategies
First questions usually open-ended case presentation:
(want to know your thought process)
- Show that you know how to take a thorough history and physical
- Think out loud- “I am interested in this… I would do this and check such and such because…”
- Elaborate on differential diagnosis
- Need to be precise and clear when drawing conclusions

PART II
- Not a normal conversation
- Taught to behave neutrally
- Examiner takes notes
- They may cut you off
- May have observers
- Grooming, behavior, communication skills are important
- Okay to take notes

PART II
- How to deal with uncertainty. You may not know the “right answer” or there may be no “right answer.” Tell the examiner your thought process. Who would you turn to for help?
- Avoid wild guesses
## PART II

- **Preparation-**
  - Attend a review course
  - Archives Study Guides for the last 5+ years: excellent guides and should be reviewed
  - Mock Orals
  - ABPMR website for Oral Exam Demonstration Video
  - Evaluate program strengths, weaknesses
  - Maintain breadth of knowledge

## 4 MOC Components

- **Professional Standing**
  - Must hold valid, unrestricted license to practice medicine

- **Cognitive Expertise: Computer Based Testing**
  - 160 questions/5-hour block
  - Fundamental knowledge, practice-related knowledge, knowledge of the practice environment (quality assurance, safety, ethics, professionalism, legal and economic issues)

### 4 MOC Components

- **Lifelong Learning and Self-assessment:**
  - 300 Category I credits/10 year MOC cycle
  - At least 50% PM&R or related areas

- **Self-assessments**
  - 4 now, 8 if recert on or after 2012
  - Self-Directed Physiatric Education Program Study Guide and SAE-P (AAPM&R) every March
  - AANEM EDX or Neuromuscular SAE
  - ABMS Patient Safety Foundations Module

- **Practice Performance**
  - Clinical Care Practice Improvement Project (PIP)
    - Select appropriate area for self-improvement within practice setting
    - Lead a team in developing a project to improve an element of practice: need outcomes demonstrated
  - AAPM&R PIP
    - Logical framework w/ forced progression
    - Following process based on EBM
    - Currently LBP, CVA, DVT, Osteoporosis
    - Minimum 5 wks to complete
  - ABMS Patient Safety Improvement Program
MOC Study Strategies

- Standard sources already mentioned
- Maintain broad knowledge base
- Review MOC Exam Outline
  - [www.abpmr.org](http://www.abpmr.org)
  - subjects are weighted
- Read *Preparing for Your Computer-Based Exam*