Myopathies

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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

<table>
<thead>
<tr>
<th>Condition</th>
<th>Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myotonic Dystrophy</td>
<td>- facial weakness, frontal balding, temporalis muscle wasting</td>
</tr>
<tr>
<td></td>
<td>- percussion myotonia</td>
</tr>
<tr>
<td>Dermatomyositis</td>
<td>- rash</td>
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### Myopathies - Signs

<table>
<thead>
<tr>
<th>Condition</th>
<th>Signs</th>
</tr>
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<tbody>
<tr>
<td>What should not be seen in pure myopathies?</td>
<td></td>
</tr>
<tr>
<td>Sensation</td>
<td>- usually normal</td>
</tr>
<tr>
<td>Reflexes</td>
<td>- usually preserved early on</td>
</tr>
<tr>
<td>Fasciculations</td>
<td>- not seen</td>
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</table>

### Myopathies - Laboratory Tests

<table>
<thead>
<tr>
<th>Test</th>
<th>Details</th>
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<tbody>
<tr>
<td>Serum Creatine Kinase</td>
<td>- upper normal varies from 200 - 500</td>
</tr>
<tr>
<td></td>
<td>- depends upon lab, gender, race</td>
</tr>
<tr>
<td></td>
<td>- can see up to ~1000 in denervating diseases</td>
</tr>
<tr>
<td></td>
<td>- over 1000 suggests muscle disease</td>
</tr>
<tr>
<td>AST, LDH, aldolase</td>
<td>- also elevated in liver disease</td>
</tr>
<tr>
<td></td>
<td>- less sensitive than CK</td>
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### 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
Hereditary Myopathies-
Mitochondrial Myopathies

- A group of myopathies with both maternal mitochondrial or mendalian inheritance
- Often multi system disease
- Often ragged red fibers on trichrome stain
- Often with ophthalmoplegia (confused with Myesthenia)
- EMG findings are usually minimal with early recruitment and short duration, low amplitude MUAP’s

Hereditary Myopathy-
Myotubular Myopathy

- Infantile x linked severe form
- Juvenile autosomal recessive form
- Milder autosomal dominant
- EMG- polyphasic low amplitude MUAP,s fibs and pos sharp waves and CRD;s (the only congenital myopathies with spontaneous activity)
- Myotonic like discharges may suggest myotonic dystrophy

Inflammatory Myopathies

- Idiopathic
  - polymyositis, dermatomyositis, inclusion body myositis
- Infectious
  - HIV, Influenza, Hep B, Hep C, other viruses
- Bacterial (Strep, Staph, Yersinia)
- Fungal
- Parasites (Toxo, Trichinosis, Cestodes - tapeworms)

Polymyositis - Dermatomyositis

- Proximal > Distal Weakness, muscle pain
  - dysphagia, dyspnea, arrhythmias
- Increased CK, usually 5 - 50 fold increase
  - SGOT, SGPT, LDH, aldolase also increased
- Biopsy - endomysial inflammation, segmental necrosis
- Dermatomyositis (a vasculitis) - heliotrope rash
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 asymetrically
• 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 conductions would most likely show:
  a. Marked slowing in CV
  b. Prolonged distal latency
  c. Reduced CMAP amplitude
  d. Increased temporal dispersion