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

- 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

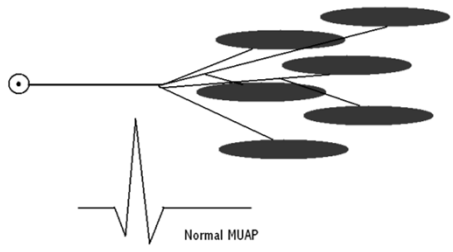
Electrodiagnostic Approach to Myopathies

- 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

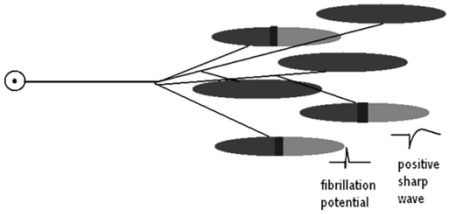
- If NMJ disorder is suspected, then do repetitive stimulation studies.
  - Usually normal in myopathies
  - Some myotonic conditions do have a decrement

Normal MUAP



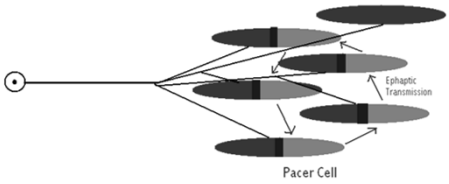
At Rest

Myopathies - Spontaneous Activity

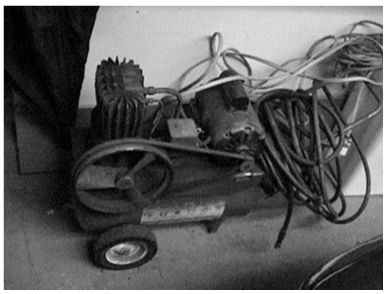


Fibrillation Potentials

Complex Repetitive Discharge



Complex Repetitive Discharges



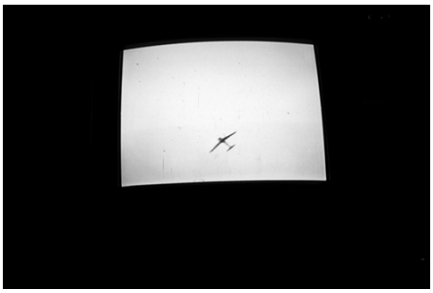
Complex Repetitive Discharge

- 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

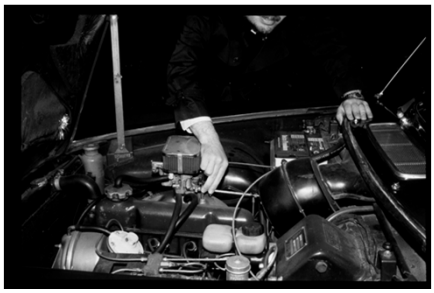
Myopathies - Other Spontaneous Activity

- 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

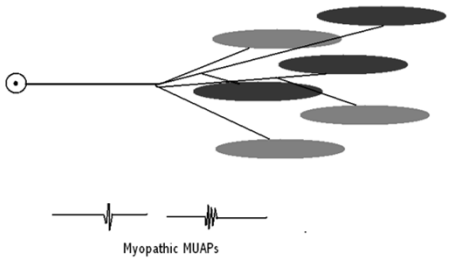
Myotonia in Action Tora Tora Tora

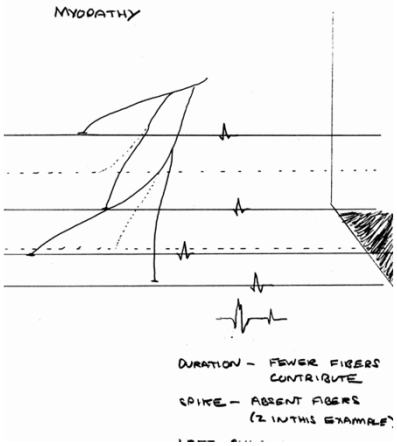


Myotonia



Myopathic MUAPs





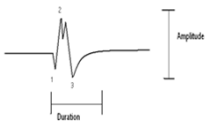
Myopathic MUAPs

- 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



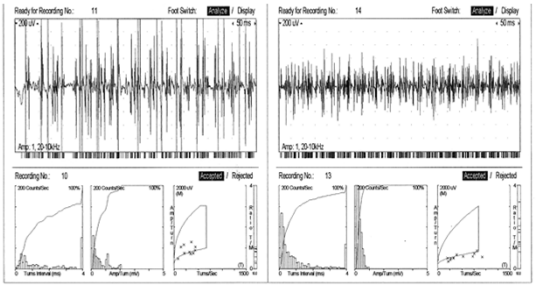
- Duration
  - most reliable
  - more difficult to measure
- Amplitude
  - easy to measure
  - depends upon needle position
- Phases
  - non specific

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

Hereditary Myopathies

- 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

Hereditary Myopathies

- 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

Hereditary Myopathies

- 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- Mitochondiral 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 opthalmoplegia (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 inflamatory 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 paraneoplasic) 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