Neuromuscular Physical Examination

Megan has a Myopathy

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Disclosures
Dr. Pestronk: Receives research support from the NIH, Muscular Dystrophy Association, Neuromuscular Research Fund; Acceleron, AnnJi, Argenx, Cytokinetics, Biogen, Fulcrum, Genzyme, Idera, Sanofi & Ultragenyx. Owns stock in Johnson & Johnson; Directs the Washington University Neuromuscular Clinical Laboratory which performs Antibody testing & NM pathology; Off-label treatments discussed.

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Neuromuscular Physical Examination
Alan's Anecdotal Advice

- General
  - Observe: Face & Gait
- Motor
  - Qualitative
  - Quantitative
- Sensory
  - Vibration: Quantitative
  - Joint Position
  - Pin
- Reflexes
  - Tendon
  - Plantar

Vesalius 1555

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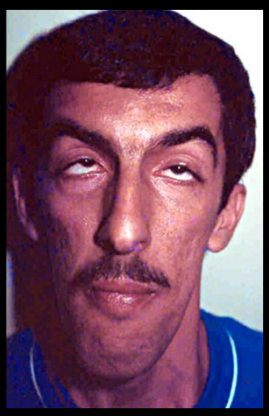
Neuromuscular Physical Examination

Motor Evaluation: Observation

- Face
  - Morphology
    - ⇒ Dysmorphism: MG DDx
      Onset: Intra-Uterine
      Cause: Reduced movements + Weakness, Ptosis
    - ⇒ Pattern of Weakness



Face: Ptosis
Elongated; Small Jaw

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Neuromuscular Physical Examination

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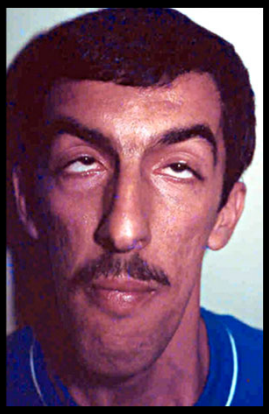

Face: Ptosis
Elongated; Small Jaw

Myasthenia gravis: Congenital

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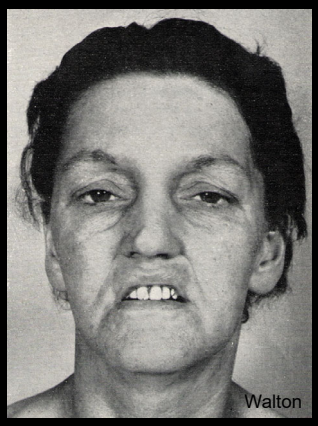
Neuromuscular Physical Examination

Motor Evaluation: Observation

- Face
  - Morphology
    - ⇒ Dysmorphism
    - ⇒ Pattern of Weakness
      Acquired MG



Myasthenia Gravis: "Snarl"

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Neuromuscular Physical Examination

Motor Testing: Qualitative; Confrontational

- Face
  - Strength testing: Active
    - ⇒ Orbicularis Oculi: Pull Eyelids apart
      NM Utility: MG status
      Active Immune MG: 98% weak
    - ⇒ Orbicularis Oris
    - ⇒ "Puffed Cheek" test

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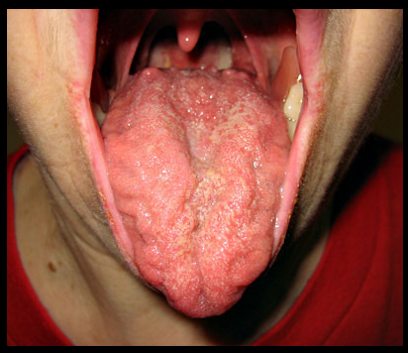
Neuromuscular Physical Examination

Motor Testing: Qualitative; Focused

- Tongue
  - Strength
    - ⇒ Confrontation
      Push against in Cheek



AAA syndrome
Tongue Atrophic & Weak

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Neuromuscular Physical Examination

Motor Testing: Qualitative; Focused

- Tongue
  - Strength: Confrontation
  - Other
    - ⇒ Speed
      Utility: Slow + Associated Jaw movement in
      ALS with bulbar/UMN involvement

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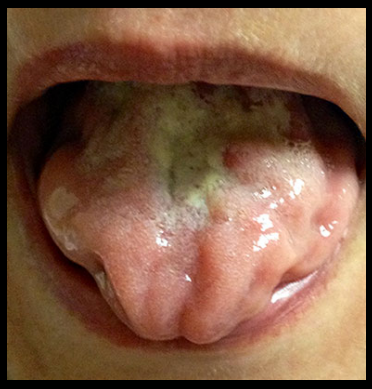
Neuromuscular Physical Examination

Motor Testing: Qualitative; Focused

- Tongue
  - Strength: Confrontation
  - Other
    - ⇒ Speed
    - ⇒ Bulk
    - ⇒ Spontaneous activity



ALS + Atrophy

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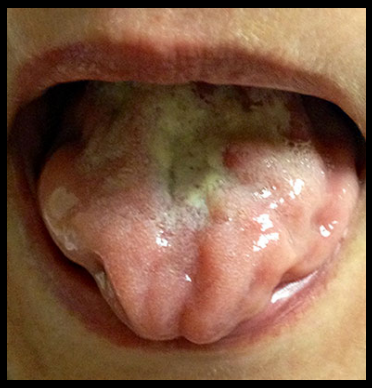
Neuromuscular Physical Examination

Motor Testing: Qualitative; Focused

- Tongue
  - Strength: Confrontation
  - Other
    - ⇒ Speed
    - ⇒ Bulk
    - ⇒ Spontaneous activity
      Fasciculations



ALS + Atrophy: SOD1 Mutation

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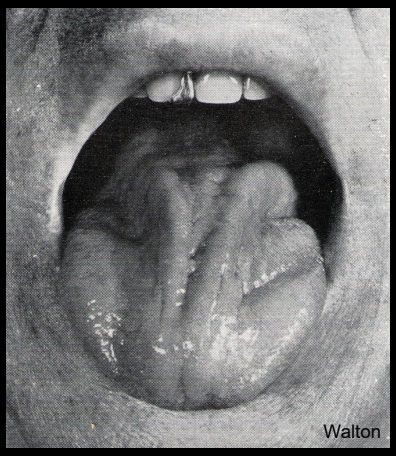
Neuromuscular Physical Examination

Strength Testing: Qualitative; Focused

- Tongue
  - Strength: Confrontation
  - Other
    - ⇒ Speed
    - ⇒ Bulk
    - ⇒ Spontaneous activity
    - ⇒ Shape



MG Tongue: Triple-Furrowed

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Neuromuscular Physical Examination

Strength Testing: Qualitative; Confrontational

- Strength testing: Trunk & Limbs
  - Principles
    - ⇒ Directly test: "Drift" is a 2º phenomenon
    - ⇒ Leverage & Position: Maximize
      Be able to overcome patient's effort
      Joint position: 90º
    - ⇒ Efficiency: Test a minimum set of muscles that provides maximal information
    - ⇒ Assess
      Initial effort: Break
      Weak or Not: No 5- or 4+
      Symmetry: Compare sides at same time, or sequentially
    - ⇒ Differential Diagnosis: Muscle vs Joint vs Pain Disorders

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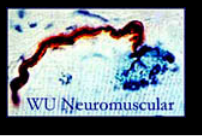
Neuromuscular Physical Examination

Strength Testing: Qualitative; Focused

- Strength testing
  - Neck (Trunk; Axial)
    - ⇒ Anterior weak: Common
    - ⇒ Posterior weak: Limited DDx
      ALS
      Immune Myopathy: BCIM
      Myasthenia Gravis

Gowers: Head ptosis in ALS



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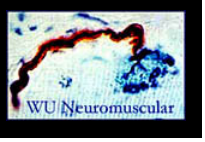
Neuromuscular Physical Examination

Strength Testing: Qualitative; Focused

- Upper Body, Proximal (Muscle; Nerve; Roots)
  - Trunk: Arm External Rotation (Infraspinatus; Suprascapular; C5-6)
    Always weak in: FSHD
  - Proximal: Arm Flexion (Deltoid +; Axillary +; C5-6)
  - Mid Arm: Elbow Flexion (Biceps; Musculocutaneous; C5-6)
  - Mid Arm: Elbow Extension (Triceps; Radial; C7)



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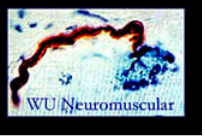
Neuromuscular Physical Examination
Strength Testing: Qualitative; Focused

- Upper Body, Distal (Muscle; Nerve; Roots): Test all 3 nerves
  - Finger Extension (Extensor Digitorum; Radial; C7)
  - Finger Abduction (1st Dorsal interosseous; Ulnar; C8-T1)
  - Thumb Abduction (Abductor Pollicis Brevis; Median; C8-T1)
  - Hand grip
    - ⇒ Usually weak in: Inclusion Body Myositis



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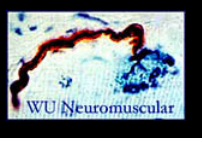
Neuromuscular Physical Examination
Strength Testing: Qualitative; Focused

- Lower Body, Proximal (Muscle; Nerve; Roots)
  - Trunk & Proximal: Standing from seated position
  - Proximal: Thigh
    - ⇒ Flexion (Psoas; Lumbar plexus; L2)
    - ⇒ Adduction: Acid Maltase Deficiency
  - Mid Leg: Knee Extension (Quads; Femoral; L3-4)
  - Mid Leg: Knee Flexion (Hamstrings; Sciatic; S1)



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Neuromuscular Physical Examination

Strength Testing: Qualitative; Focused

- Lower Body, Distal (Muscle; Nerve; Roots)
  - Distal: Ankle Dorsiflexion (Anterior Tibial; Deep Peroneal; L4)
  - Distal: Ankle Inversion (Posterior Tibial; Tibial; L5)
  - Very Distal: Toe Dorsiflexion (Peroneal; L5)

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Neuromuscular Physical Examination

Strength Testing: Quantitative

Hand-Held Dynamometer

12% (of normal) change in average of 6 muscles = $p < 0.05$

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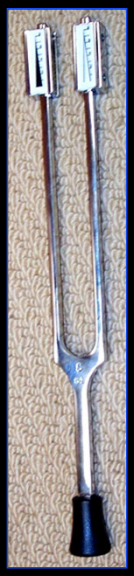
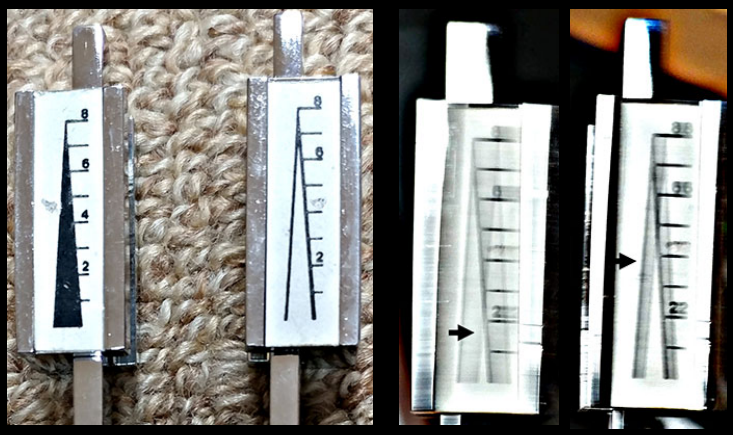
Neuromuscular Physical Examination



Sensory: Large Axon, Vibration Sensation



Rydel-Seiffer Tuning Fork

"Tell me when the Buzzing Stops."

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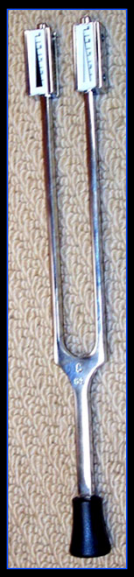
Neuromuscular Physical Examination



Sensory: Large Axon, Vibration Sensation

- R-S Tuning fork type
  - "No Screws"; Heavy
- Rapid
- Quantitative
  - Useful for: Children; Proximal; Arms
- Reproducible
- Physiological correlate
  - SNAP amplitude

Rydel-Seiffer Tuning Fork

"Tell me when the Buzzing Stops."

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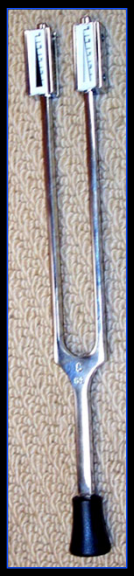
Neuromuscular Physical Examination



Sensory: Large Axon, Vibration Sensation

- Methods
  - 1st: Hold Stem between Thumb & 2 fingers
  - Then: Rest Proximal Tuning Fork Arm between Thumb & Index finger
- Interpretation
  - Number at which "Buzzing stops"
  - Normals (Toes)
    - ⇒ Children: ≥ 6
    - ⇒ Adults over 50 years: ≥ 3

Rydel-Seiffer Tuning Fork

"Tell me when the Buzzing Stops."

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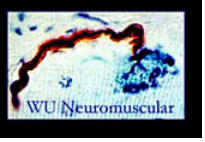
Neuromuscular Physical Examination

Large Axon: Joint Position

- Method
  - Hold digit with 2 fingers at sides
- Interpretation
  - Joint position sense ↓ ≥ Vibration ↓ : Suggests Lesion at
    - ⇒ Dorsal Root Ganglion
    - ⇒ Posterior Column: Look for pin level on trunk
    - ⇒ Axonopathy: Very severe



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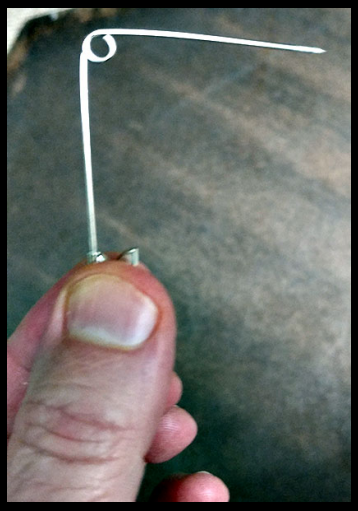
Neuromuscular Physical Examination

Small Axon: Pin Sensation



Hold: Pin by head

Scratch: Don't Stick
- No puncture wounds
- Reproducible stimulus

Move: Distal to Very Proximal
- Ask after 1st stimulus: "Is this sharp?"
- Gradient point: Often very proximal
- Physical sign: Patient "startle" or jump

Pin Gradient
- 94% specificity for Small Axon Loss on Skin Biopsy

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Neuromuscular Physical Examination

Tendon Reflexes

- Use: A correct tool
  - Hammer with
    - ⇒Moment of Inertia: Reproducible
    - ⇒ Soft Cushion Top
  - Good
    - ⇒ Queen Square
    - ⇒ Tromner
  - Bad
    - ⇒ Taylor

Gowers
Doing several things wrong

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Neuromuscular Physical Examination

Tendon Reflexes

- Employ: Good technique
  - Joint at 90º
  - Hit yourself, not the patient
    - ⇒ Place: Thumb on tendon
    - ⇒ Stretch: Muscle tendon
    - ⇒ Hit: Your thumb
  - Strike in a Specific direction
    - ⇒ To stretch the muscle

Gowers

Doing several things wrong

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Neuromuscular Physical Examination

Tendon Reflexes (DTRs)

Gowers

- DTRs to Test
  - Biceps (C5)
  - Pronator (C6)
  - Triceps (C7)
  - Quadriceps (L4)
  - Ankle (S1)
- Bonus test: Direct Muscle Percussion
  - Positive: Percussion induces muscle contraction > DTR
  - If present when DTR absent → Suggests Denervation

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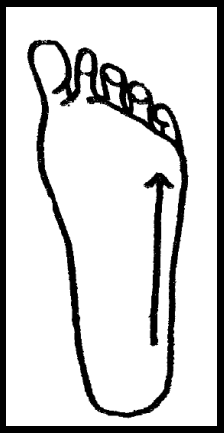
Neuromuscular Physical Examination

Plantar Reflex

- Use: A correct tool
  - Two Keys
- Employ: Good technique
  - Scratch: Lateral side of foot
  - Slow movement: 5 seconds
  - Don't need to hurt patient



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Neuromuscular Physical Examination

- Plantar Reflexes to Test
  - Plantar-Chaddock with 2 keys
    - ⇒ Sensitivity: Fair-Good
    - ⇒ No prominent pain or tickle
    - ⇒ Good for children
    - ⇒ May be normal in: ALS
  - Bing: Pin prick, or scratch, on dorsum of 1st toe
    - ⇒ Most Specific: No-one normally withdraws into a Pin
    - ⇒ Sensitivity: Low
- Alternative test for UMN: Foot tap, Repetitive
  - Positive: Slowed motion
  - Sensitivity: Higher for UMN lesions than Plantar Stimulation

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Neuromuscular Web Site

https://neuromuscular.wustl.edu

Instagram

pestronka2

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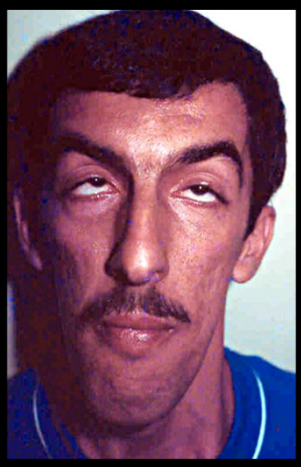
Neuromuscular Disorders: Clinical Evaluation

- Description of illness
  - Source
    - ⇒ History
    - ⇒ Examination
  - Processing info: Disease Patterns
    - ⇒ Anatomic: Regions involved
    - ⇒ Functional defects
    - ⇒ Time course
    - ⇒ Distinctive features



Face: Weak; Elongated
Myasthenia gravis: Congenital

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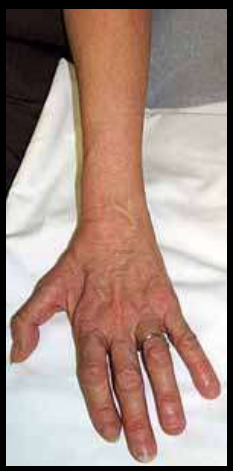
Neuromuscular Disorders
Clinical Patterns: Distinctive Features

Anatomic

- Arms vs. Legs vs. Cranial
  - Common: Legs early in disease course
  - Unusual: Arms only at disease onset



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Neuromuscular Disorders
Clinical Patterns: Distinctive Features

Anatomic

- Proximal vs. Distal
  - Common
    - ⇒ Proximal: Myopathy; Neuronopathy
    - ⇒ Distal: Axonal neuropathy
  - Uncommon: Very proximal
    - ⇒ Early respiratory failure
    - ⇒ Posterior neck weakness

Posterior neck weakness

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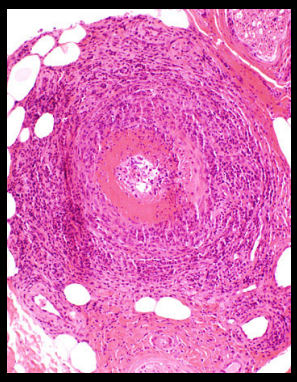
Neuromuscular Disorders
Clinical Patterns: Distinctive Features

Anatomic

- Symmetric vs. Asymmetric
  - Asymmetric myopathies
    - ⇒ Rarely treatable
  - Asymmetric neuropathies
    - ⇒ Nerve biopsy often useful to define cause
    - ⇒ Often treatable



Vasculitis

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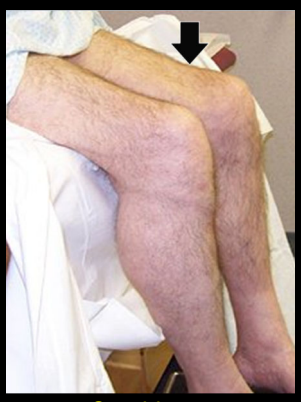
Neuromuscular Disorders
Clinical Patterns: Distinctive Features

Anatomic

- Focal regions involved
  - Short differential diagnosis
  - Examples
    - ⇒ Quadriceps or Adductor weakness
    - ⇒ Respiratory failure
    - ⇒ Posterior neck weakness



Quadriceps wasting

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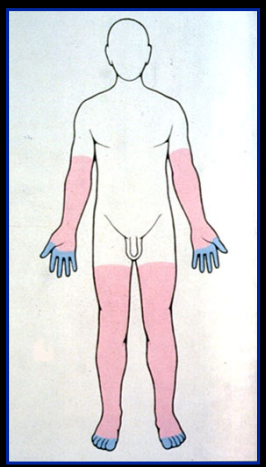
Neuromuscular Disorders
Clinical Patterns: Distinctive Features

Functional: Selective involvement

- Motor
  - Weakness
  - Muscle size: Large or Small
  - Abnormal movement
- Sensory
  - Loss: Small or Large axons
  - Gain: Pain; Paresthesias
- Autonomic



Stocking-Glove sensory loss

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Neuromuscular Disorders
Clinical Patterns: Distinctive Features

Temporal

- Long term: Course
  - Acute: Days to Weeks
  - Chronic: Months to Years
  - Episodic
  - Hereditary
- Short term
  - Minutes to Hours
  - "Fatigue"

- Onset age
  - Pediatric
    - ⇒ Neonate
    - ⇒ Child
  - Adult
    - ⇒ 20 to 60 years
    - ⇒ > 60 years

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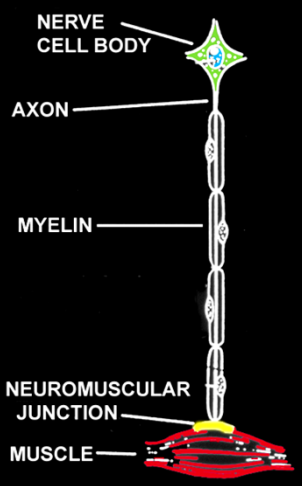
Neuromuscular Disorders: Clinical Evaluation

- Disease Patterns: Utility
  - Anatomic Localization
    - ⇒ Cell Body
    - ⇒ Axon
    - ⇒ Myelin
    - ⇒ Neuromuscular Junction
    - ⇒ Muscle
  - Differential Diagnosis
    - ⇒ Generate: Limited set of possibilities



Anatomic Localization

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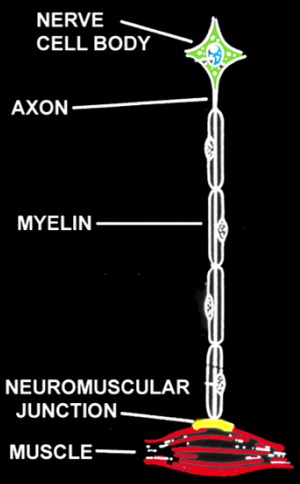
Neuromuscular Disorders: Evaluation



Anatomic Differential Diagnosis

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Neuromuscular Disorders
Myopathy

- Clinical features
  - Weakness
    - ⇒ Proximal
    - ⇒ Constant
  - Muscle size
    - ⇒ Early: Normal or Increased
    - ⇒ Late: Atrophy or Increased
  - Sensory exam & Tendon reflexes: Normal
- Laboratory changes
  - Creatine Kinase (CK) or Aldolase: High
  - EMG: Myopathic

Becker
Muscular Dystrophy

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Neuromuscular Disorders
Neuropathy, Axonal

- Clinical features
  - Weakness
    - ⇒ Anatomy: Distal > Proximal
      Asymmetric or Symmetric
    - ⇒ Constant
  - Muscle size: Atrophy
  - Sensory loss: Distal (Stocking-Glove)
  - Tendon reflexes: Reduced distally (Ankles)
  - Variable: Symmetric vs Asymmetric

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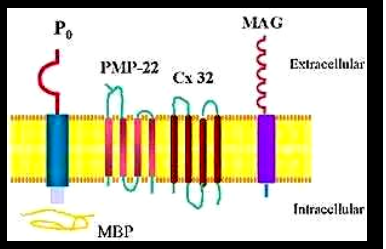
Neuromuscular Disorders
Neuropathy, Demyelinating

- Clinical features
  - Weakness
    - ⇒ Distal + Proximal
    - ⇒ Constant
    - ⇒ Symmetric (Most)
  - Muscle size: Normal
  - Sensory loss: Distal, Mild
  - Tendon reflexes: Reduced diffusely
    - ⇒ Out of proportion to weakness



Myelin proteins

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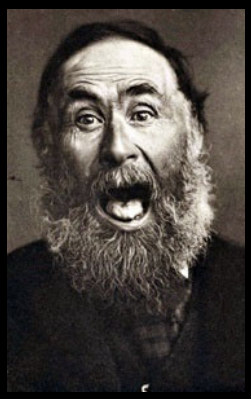
Neuromuscular Disorders
Nerve Cell Body Disorders (Neuronopathy)

- Clinical features
  - Selective modality loss: Motor; Sensory
  - Distribution
    - ⇒ Proximal + Distal
    - ⇒ Cranial
    - ⇒ Not length dependent
    - ⇒ Often asymmetric
  - Motor
    - ⇒ Weakness
    - ⇒ Atrophy
    - ⇒ Fasciculations
  - Course: No improvement with treatment



Tongue:
Wasting & Weakness

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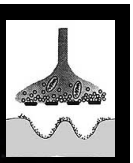
Neuromuscular Disorders
Neuromuscular Junction Disorders



- Clinical features
  - Weakness
    - ⇒ Proximal + Distal + Bulbar
    - ⇒ Variable: Fatigue
  - Sensory & Tendon reflexes: Normal
  - Course
    - ⇒ Chronic disease
    - ⇒ Rapid changes with physical activity or treatment

Rabbit with experimental
Myasthenia gravis

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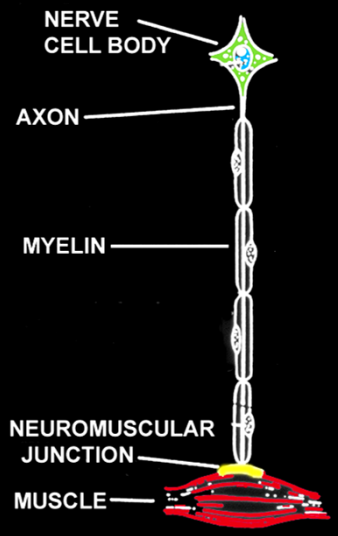
Neuromuscular Disorders: Evaluation

- Description of disease
  - History
  - Examination
- Localization of disease process
  - Clinical
  - Serum biomarkers: CK & Aldolase
  - Electrophysiology: EMG & NCV
- Identification of specific disease
  - Blood testing: Antibodies; Genetics
  - Biopsy: Nerve; Muscle



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Final outcome of evaluation: Diagnosis & Treatment

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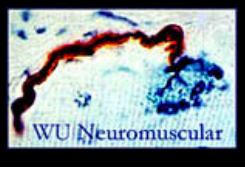
Clinical Approach to Neuromuscular Disorders

- Gather data: Patient interaction
  - History
  - Exam
- Process & Organize data
  - Describe clinical syndrome (Concise)
  - Order lab tests
- Localize lesion
- Identify: Diagnosis
  - Differential
  - Specific
  - Treatment modalities



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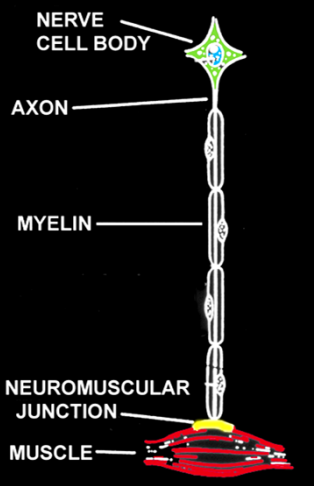
Neuromuscular Disorders: Evaluation

Description of disease: History & Exam

Localization of disease process

- Clinical features
- Laboratory features
  - Creatine Kinase or Aldolase high: Muscle
  - Electrophysiology
    - ⇒ Nerve conduction studies
    - ⇒ Electromyography (EMG)
  - Pathology: Muscle or Nerve
  - Anatomy: MRI; Ultrasound



Anatomic Differential Diagnosis

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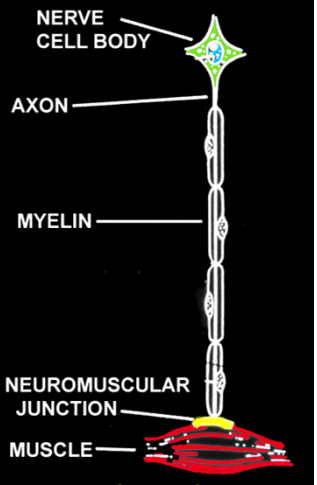
Neuromuscular Disorders: Evaluation

Description of disease: History & Exam

Localization of disease process

- Clinical features
- Laboratory features
  - Muscle enzymes (Serum)
  - Electrophysiology
    - ⇒ Nerve conduction studies
    - ⇒ Electromyography (EMG)
  - Pathology: Muscle or Nerve
  - Anatomy: MRI; Ultrasound



Anatomic Differential Diagnosis

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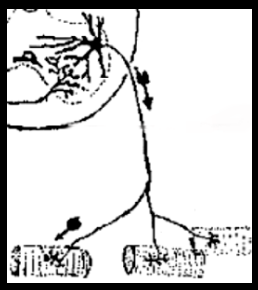
Neuromuscular Disorders
Electrodiagnostic testing

- Nerve conduction testing
  - Stimulate nerve maximally
  - Record evoked action potentials
    - ⇒ Motor: Muscle potentials (CMAP)
    - ⇒ Sensory: Nerve potentials (SNAP)



Motor unit

Compound Motor Action Potential (CMAP)
Electrical activity of all muscle fibers stimulated by nerve

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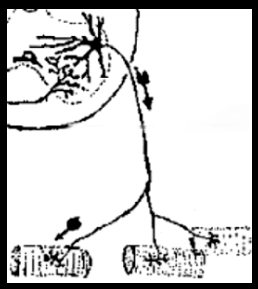
Neuromuscular Disorders
Electrodiagnostic testing

- Nerve conduction testing
  - Stimulate nerve maximally
  - Record evoked action potentials
  - Measure: Size & Velocity
    - ⇒ Small size: Axon loss; Myopathy
    - ⇒ Slow velocity; Cond block: Myelin damage
    - ⇒ Axon loss not length dependent: Cell body



Motor unit

Compound Motor Action Potential (CMAP)
Electrical activity of all muscle fibers stimulated by nerve

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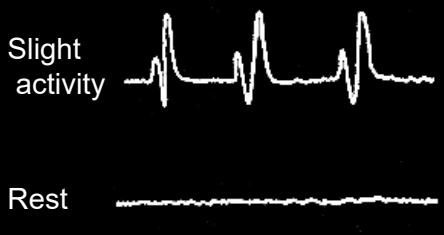
Neuromuscular Disorders
Electrodiagnostic testing

- Electromyography (EMG)
  - Electrical activity detected by needle inserted into muscle
  - Potentials detected: Motor units & Single muscle fibers

Normal EMG: Motor unit size normal



Motor unit potentials
Amplitude: Normal
Duration: Normal

Spontaneous activity
None

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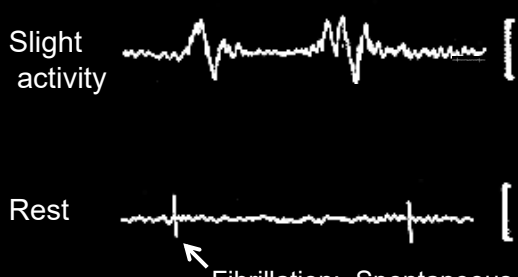
Neuromuscular Disorders
Myopathy: Electromyography

Myopathic EMG: Motor unit size small



Motor unit potentials
Amplitude: Small
Duration: Brief

Spontaneous activity
Occasional fibrillation

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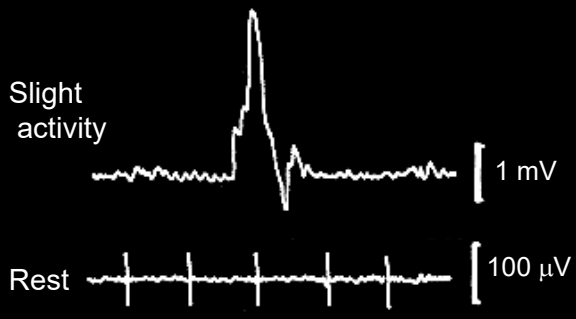
Neuromuscular Disorders
Axonal neuropathy: Electromyography

Neuropathic EMG: Motor unit size large



Motor unit potentials
- Amplitude: Large
- Duration: Long

Spontaneous activity
- Fibrillations

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