Neuromuscular Disorders for the LTCF

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Outline

- Localization by Neuro Exam
- What are "neuromuscular" diseases?
  - Localization by anatomy
- Some overview of EMG / NCV testing
- Your common enemy: BACK / NECK pain
- Nerve disorders
- NMJ disorders
- Muscle disorders

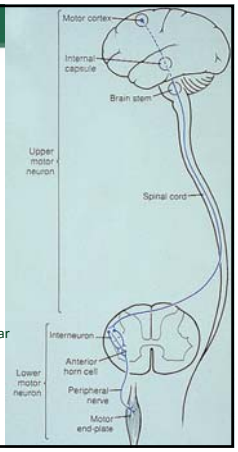
Using Neuro Exam to Sort Out Neurologic Symptoms

- Weakness
  - Proximal or distal?
  - Reflexes?
  - Fasciculations?
- Pain
  - Myalgias?
  - Neuropathic pain?
- Numbness
  - Dermatomal, sensory level, or hemibody?

Anatomy of the Motor System Overview

- Localize the Lesion!!
  - Motor Cortex
  - Modulators (*Extrapyramidal*)
    - Basal Ganglia
    - Cerebellum
  - Spinal Cord
  - Ant. Horn cell
  - Peripheral nerve
  - NMJ
  - Muscle

Neuromuscular



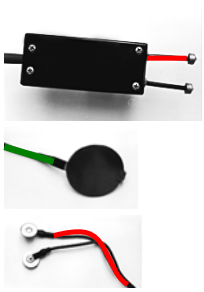
Clinical Assessment

- Diagnostic Testing
  - CK
  - Lumbar Puncture (if suspect GBS)
  - AChRAb (if suspect Myasthenia Gravis)
  - Electrodiagnostics
    - Needle EMG (if suspect myopathy, ALS)
    - Nerve conduction studies (if suspect GBS)
    - Repetitive nerve stimulation [Jolly] (if suspect MG)

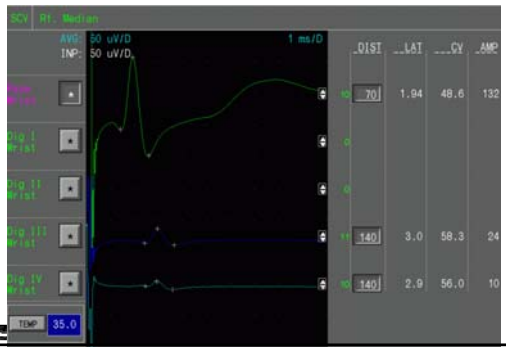
EMG/NCV Overview

Basic NCS Intro

- **Orthodromic** (with the normal direction of conduction) or **Antidromic** (opposite to direction of conduction)
- Motor NCS or Sensory NCS
- At UAB, we stimulate with the wand and record with recording electrodes
- Black is active, Red is reference, Green is ground



Basic NCS Intro

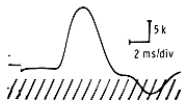


Basic NCS Intro

- Major parameters are AMPLITUDE and VELOCITY, which are recorded for multiple segments of each nerve tested
- Amplitude – summated size in mV of either the CMAP (motor) or SNAP (sensory)
- Velocity – determined by your measured length of segment being tested, divided by the latency (in ms) of the stimulus to the onset of the response

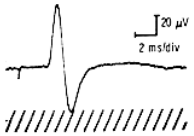
Motor NCS

- Stimulate over a nerve and record the motor response over the corresponding muscle
- Upper extremities
  - Median – record over APB
  - Ulnar – record over ADQ
- Lower extremities
  - Peroneal – record over EDB (or AT if atrophied) **Note, most common reason for isolated low amplitudes in peroneal n. is EDB atrophy**
  - Posterior Tibial – record over abductor hallucis



Sensory NCS

- Stimulate in the dermatome and record over the sensory nerve
- Upper extremities
  - Median
  - Ulnar
  - Radial
- Lower extremities
  - Sural

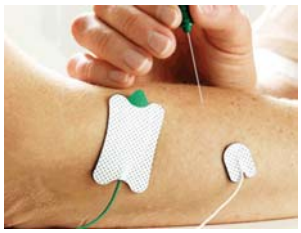


Commonly Tested Nerves

- Upper extremities
  - Median motor
  - Median sensory
  - Ulnar motor
  - Ulnar sensory
  - (sometimes) Radial sensory
- Lower extremities
  - Peroneal (motor)
  - Posterior tibial (motor)
  - Sural (sensory)

Basic EMG Intro

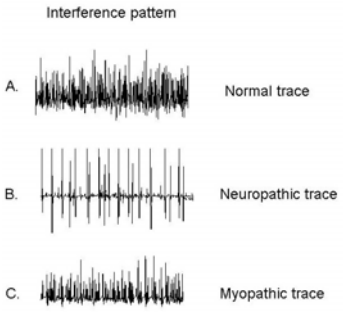
- Always physician performed (not a tech)



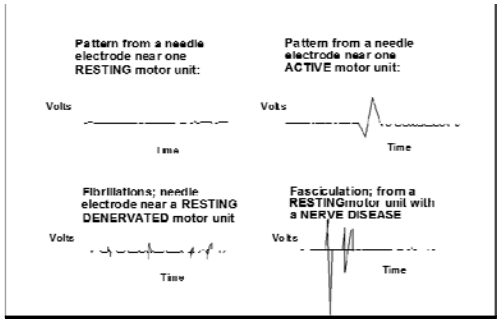
EMG Basics

- Clinician selects muscles to stick based on exam/DDx
- Typical Muscles to Test
  - Myopathy: vastus lateralis, deltoid
  - Radiculopathy: muscles in that myotome plus paraspinal muscles
  - Neuropathy: muscles in that myotome
- All muscles tested in 3 ways
  - Insertional activity
  - Activity with sustained mild contraction
  - Recruitment activity with maximal contraction

EMG



EMG



Primer for Back Pain / Neck Pain

LBP / Neck Pain General Principles

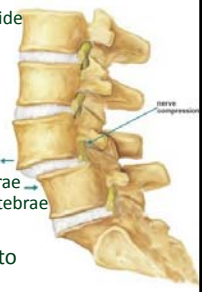
- Most common cause is osteoarthritis, whether low back or neck
  - Stiffness worst in AM, better as day goes on
- Due to age and medical comorbidities, many LTCF patients will not be surgical candidates
  - Focus on conservative management
- Want to avoid opiates in geriatric population if possible
  - Don't forget about use of topicals

LBP / Neck Pain with NO Neuro Dysfunction

- Routine DJD
  - Pain but non-radiating and no weakness/numbness
  - No need for diagnostic testing
    - At most, possibly flexion/extension plain films
    - NO need for MRI
  - **"Conservative Tx:"**
    - Scheduled NSAID's
    - Muscle relaxants
    - PT, modalities (acupuncture, chiropractic, TENS)
  - Acute flares: ESI/facet blocks, steroid dose packs

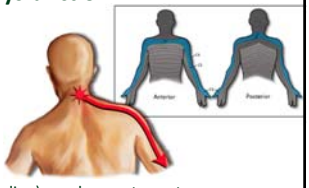
LBP / Neck Pain General Principles

- **Spondylosis**
  - Grinding/popping when turns side to side
  - Means a misalignment of vertebrae
  - Can confirm mal-alignment with X-ray
  - Tx: conservative management most appropriate
- Spondylolisthesis
  - Specific misalignment with one vertebrae slipped forward on top of adjacent vertebrae
- In general: failure of "conservative management" means time to refer to specialist before giving opiates



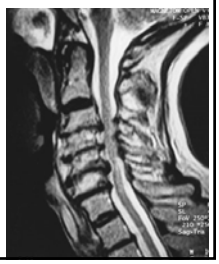
Neck Pain WITH Neuro Dysfunction

- Cervical Radiculopathy
  - Presentation: Neck pain radiating down arm
  - Numbness/weakness in distribution of nerve root that is pinched
  - Most common is C8/T1
  - Ulnar neuropathy and CTS (median) numbness stops at wrist but goes up arm in radic
  - Eval:
    - EMG/NCS to eval radic vs. neuropathy
    - MRI to confirm foraminal stenosis/nerve root impingement
  - Tx:
    - Previous plus gabapentin for neuropathic component, surgery if severe/refractory



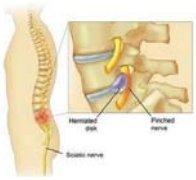
Neck Pain WITH Neuro Dysfunction

- Cervical Myelopathy
  - Presentation: Chronic neck pain with gait problems
  - Caused by disc compressing cervical cord
  - UMN problem so should have *brisk reflexes* +/- upgoing toes
  - **Gait** is spastic
  - Eval:
    - MRI Cspine to confirm level(s) of cord compression
  - Tx:
    - Surgical evaluation; if ineligible, then pain meds, PT/assistive devices



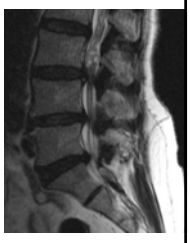
LBP WITH Neuro Dysfunction

- Lumbar radiculopathy
  - Presentation: LBP radiating down leg
  - Numbness/weakness in distribution of nerve root being pinched
  - Most common are L5/S1 and L4/5
  - S1 radic should have loss of ankle jerk, L4 radic may have loss of patellar reflex
  - Eval
    - EMG/NCS to confirm active nerve root impingement
    - MRI L-spine to eval level(s)/extent of foraminal stenosis / nerve root impingement
  - Tx:
    - Conservative mgmt plus gabapentin
    - Surgery if refractory



LBP WITH Neuro Dysfunction

- Lumbar spinal stenosis / Neurogenic Claudication
  - Presentation: LBP relieved by leaning forward/resting on an object such as walker/shopping cart; may have pain worsening with ambulation, relieved by rest
  - Commonly have complaint of **"legs giving out,"** which may cause falls
  - Eval
    - MRI L-spine to confirm level(s) of central canal stenosis
  - Tx:
    - Conservative mgmt
    - Surgery if refractory



DDx by Localization

- Anterior Horn Cell
  - Exam Features
    - Mixed UMN/LMN (Fasciculations, Atrophy, Increased reflexes)
    - Patchy weakness
    - Preserved sensation
  - Causes
    - Amyotrophic Lateral Sclerosis
    - Poliomyelitis or Postpolio syndrome
    - Infectious (Rabies, West Nile Virus)
    - Spinal Muscular Atrophy
    - Paraneoplastic motor neuronopathy

Motor Neuron Disease – ALS

- In geriatric population, painless foot drop is ALS until proven otherwise
- Life expectancy: 3-5 years from diagnosis
- Presentation: Weakness, brisk reflexes with atrophy/fasciculations, normal sensory
- Diagnostics: EMG**/NCV, often MRI Cspine to rule out cervical myelopathy
- Treatments
  - Riluzole – generic, no effect on daily function, increased life expectancy
  - Edaravone – IV monthly, improved rate of functional decline based on 6mo study

ALS Supportive Care in LTCF

- Weight loss predicts more rapid decline
  - Increased caloric intake
  - Early discussion about /placement of PEG
- Often will require home ventilator support
  - Often on BiPAP first before full ventilatory support
  - Early GOC discussion
  - Trying to avoid emergent intubation as wean to extubation very difficult
  - Can live for a very long time with home ventilator

DDx by Localization – Peripheral Nerve

- Exam Features
  - Prominent sensory loss, Distal weakness, Loss of reflexes
- Causes
  - Toxic (Alcohol, Thallium, Lead, Saxitoxin)
  - Metabolic (Diabetes)
  - Autoimmune (Guillain Barre Syndrome, Chronic Immune Demyelinating Polyneuropathy (CIDP))
  - Infectious (Lyme, Diphtheria)
  - Systemic diseases (SLE, PAN), Neoplastic
  - Drugs (vincristine, amiodarone, dapsone, phenytoin)
- Diagnostics
  - NCV will distinguish toxic/metabolic (axonal) vs. autoimmune/inflammatory (demyelinating)

Peripheral Neuropathy in LTCF

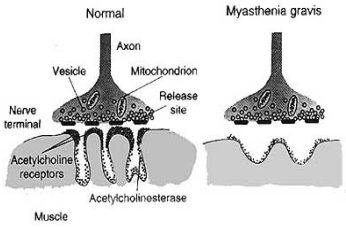
- Treatment
  - Remove underlying cause if possible
  - Symptomatic
    - Common meds like gabapentin, amitryptyline all problematic in geriatric population
    - TOPICALS! – lidocaine cream/patch, custom compounded creams
  - Supportive
  - Remember that neuropathies commonly cause gait problems in geriatric population
    - Falls at night or other low lighting

DDx by Localization

- Neuromuscular junction
  - Exam Features
    - Fatigable/Fluctuating weakness
    - Oculobulbar symptoms
    - Preserved sensation
  - Causes
    - Myasthenia Gravis
      - Cholinergic crisis
    - Lambert Eaton Syndrome
    - Toxins (Organophosphates, Botulism, Tick Paralysis, Black widow spider bite)
    - Iatrogenic neuromuscular blockade

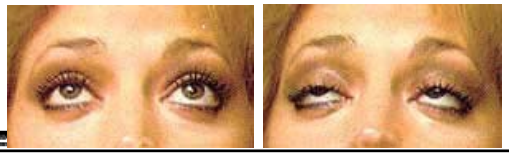
Myasthenia Gravis

- Autoimmune disorder at Neuromuscular Junction (NMJ) caused by Ab against ACh Receptor (post-synaptic)
- Remodeling of post-synaptic membrane with time



Myasthenia Gravis

- Clinical presentation
  - Bimodal (20's and 70's) age predominance
    - Much worse prognosis in geriatric population
  - Like all autoimmune disease, prone to flares/exacerbations
  - Symptoms: *Fatigable weakness*, especially oculobulbar involvement (diplopia, ptosis, dysarthria, dysphagia)
  - Sensation and Reflexes are NORMAL



Myasthenia Gravis

- <u>Diagnosis</u>
  - Serum AChRAb's – positive in 70% of cases
  - RNS shows *decremental* response to repetitive stimulation (95% sensitivity)
  - CK can be mildly elevated
  - LP, NCV, MRI of no utility
- <u>Drugs that Precipitate Acute Exacerbation</u>:
  - Quinolones, beta blockers, aminoglycosides, Ca chan blockers, narcotics, and paralytics (of course)
  - Therefore commonly presents with failure to wean post-operatively

MG in LTCF

- Be very careful about concomitant meds that can precipitate exacerbation (common scenario is UTI and prescribed a quinolone)
- Treatment principles
  - Pyridostigmine (Mestinon) is a staple but watch out for diarrhea – can use glycopyrrolate to counteract
  - Keeping out of crisis will require immunosuppression
    - Steroids or steroid-sparing (mycophenolate, azathioprine)
    - Many geriatric patients will need scheduled IVIG or PLEX, complicated to coordinate from LTCF

DDx by Localization

- Muscle
  - Exam Features
    - Proximal weakness +/- Aching muscle pain
    - Preserved sensation, Intact reflexes
    - Elevated CK
  - Causes
    - Congenital Muscular Dystrophies, Metabolic Myopathies (ie Acid Maltase Deficiency)
    - Polymyositis / Dermatomyositis
    - Drugs (statin, glucocorticosteroids, amiodarone)
    - Acute Rhabdomyolysis
    - Electrolyte Imbalances (hypokalemia, hypophosphataemia)
    - Critical illness myopathy

Myopathy in LTCF

- Proximal muscle weakness with normal reflexes (and no sensory involvement)
- As muscle damage progresses, will also have atrophy
  - Though atrophy may not be present early in the course
- In LTCF setting, most commonly acquired
  - Toxins – statin, steroids, etc.
  - Autoimmune – polymyositis, DM, IBM
    - Inclusion Body Myositis – weakness/atrophy in unusual pattern: distal arms (forearm wrist flexors) and proximal legs (quads)

Questions?

- Whew, that was a whirlwind!!
- Any questions?