Clinical Neurophysiolgy: Electromyography & Nerve Conductions

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Question 1

A 78 year old female with history of breast cancer presents with left arm weakness. Exam reveal weakness of left shoulder abduction and forearm flexion. EMG is notable for myokymic discharges. What is the most likely mechanism of injury?

1. C5/C6 radiculopathy due to herniated disc
2. Post-radiation plexopathy
3. Metastatic invasion of the brachial plexus
4. Left axillary neuropathy

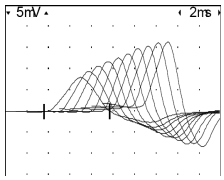
Question 2

A 34 year old male presents with two weeks of right foot weakness. Examination is notable for weakness of right ankle dorsiflexion and inversion. Plantarflexion and eversion are normal. What are the most likely findings on the nerve conduction study?

1. Absent right superficial peroneal SNAP response
2. Absent right sural SNAP response
3. Conduction block across the fibular head
4. Spontaneous activity in the lumbar paraspinal muscles

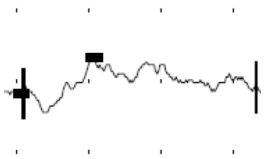
Question 3

- A 67 year old male present with complaint of slowly progressive weakness. Repetive stimulation reveals the following. What is the most likely cause of his weakness?
  1. Presynaptic blockade of ACh release
  2. Presynaptic blockade of calcium channels
  3. Postsynaptic blockade of ACh receptors
  4. Failure of AChRs to congregate at the NMJ



Question 4

- A 38 year old female presents for progressive difficulty walking and distal numbness. Proximal stimulation of the right tibial nerve results in the following CMAP. What is the most likely diagnosis?
  - 1. HSMN I (CMT)
  - 2. HSMN II
  - 3. Vitamin B12 polyneuropathy
  - 4. CIDP

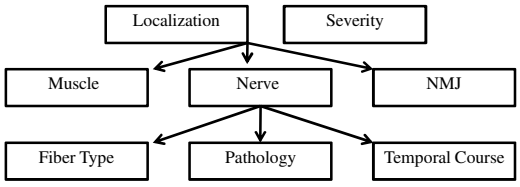


Clinical Neurophysiolgy

- Basic Tenets:
  - 1. EMG/NCS is *an extension* of the history of physical examination….it confirms your suspicions
  - 2. You should never be "surprised" by the findings
  - 3. If the study does not match your clinical index of suspicion, something is wrong with your EMG (likely technical)
  - 4. Your "answer" should be clinically relevant

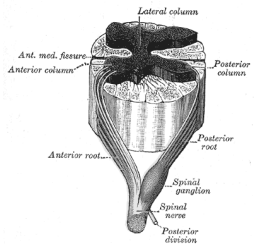
Clinical Neurophysiology

Goals of the EDX



Goals of EDX: Localization

- EMG/NCS is a test of the *Peripheral Nervous System*
  - Neuron (DRG, AHC)
  - Nerve Root
  - Plexus
  - Nerve
    - Motor, Sensory, Mixed
    - Large/Small Fiber
  - NMJ
  - Muscle



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Goals of EDX: Fiber Type

Myelinated	Diameter	C.V. m/s
Cutaneous afferents	6-12 μm	35-75
Muscle afferents	12-21μm	80-120
Muscle efferents	6-12 μm	35-75
Unmyelinated		
Afferents to the DRG	0.2-1.5 μm	1-2

1. The large myelinated fibers are the fibers measured in nerve conduction studies
2. Neuropathies that preferentially affect small fibers may have NORMAL nerve conduction studies

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Goals of EDX: Pathology

- Pathologic Responses to Injury:
  - 1. Neuronopathy (Motor or Sensory)
  - 2. Axonal Transection with Wallerian Degeneration
  - 3. Axonal Loss
  - 4. Demyelinating

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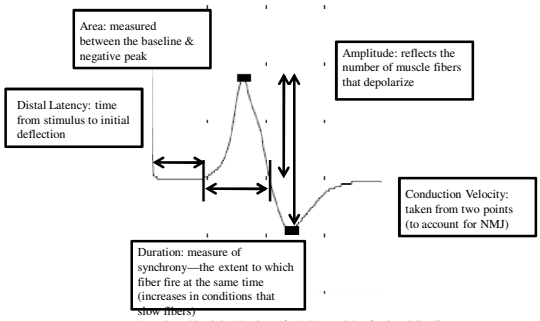
Nerve Conductions: CMAP



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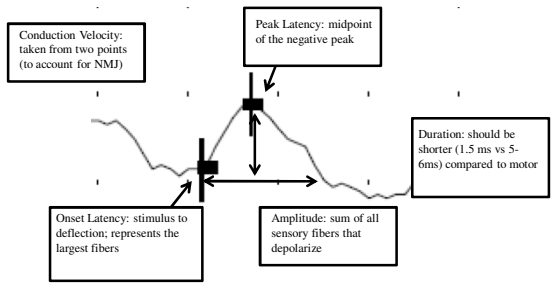
Nerve Conductions: SNAP



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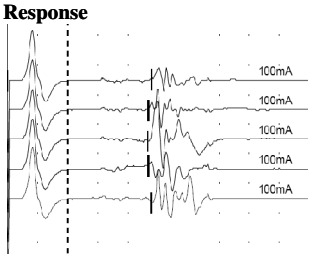
Nerve Conductions: Late Responses

F (Foot) Response

- Antidromic response towards the spinal cord
- Travels to the *anterior horn cell* and backfires a small population of AHCs
  - Represents 1-5% of all muscle fibers
  - *Pure motor*….it is *not* a reflex arc



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Nerve Conductions: Basic Patterns

Neuropathic Patterns

- Axonal Loss
  - *Reduced amplitude* is the primary abnormality associated with *axonal loss*
  - Conduction velocity & distal latency are normal (*)
    - CV should *never* be less than 75% of normal
    - DL *do not* exceed 130% of normal
- Demyelination
  - Associated with *marked slowing* of CV
    - Slower than 75% of the lower limit of normal
    - Arms: <35 m/s
    - Legs: <30 m/s
  - Distal latencies *markedly prolonged* (>130%)

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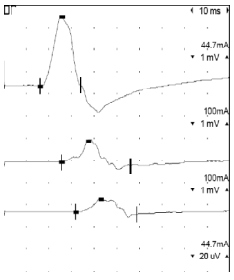
Nerve Conductions: Basic Patterns

Neuropathic Lesions

- Conduction Block
  - Occurs in *acquired demyelination* (and is *not seen* in hereditary demyelinating conditions)
  - Drop in CMAP amplitude or area of *at least* ≥20% and/or increase in duration of ≥15%



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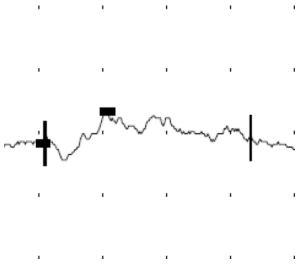
Nerve Conductions: Basic Patterns

Neuropathic Lesions

- Temporal Dispersion
  - Implies *demyelination*
  - Slower fibers lag behind the faster ones
    - More prominent with sensory fibers than motor normally
    - More prominent with proximal stimulation
  - Can cause the *amplitude* to drop by >50% without true conduction block
    - Drop in *area* of >50% *always* signifies conduction block



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Demyelinating Neuropathies

Hereditary

- Charcot Marie Tooth I
- Dejerine-Sottas (HSMN III)
- Refsum Disease (HSMN IV)
- HNPP
- Metachromatic Leukodystrophy
- Krabbe Disease
- Adrenoleukodystrophy
- Cockayne Syndrome
- Niemann-Pick Disease
- Cerebrotendinous xanthomatosis

Acquired

- AIDP/Guillian-Barre
- CIDP
  - HIV
  - MGUS (IgM)
  - Anti-MAG
  - Osteosclerotic myeloma
  - Waldenstrom macroglobulinemia
- MMN with CB (GM_1 Ab)
- Diptheria
- Toxic (amiodarone, arsenic)

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Demyelinating Neuropathies

AIDP

- First changes are delayed, absent or impersistent F-waves (reflecting proximal demyelination)
- Prolonged DL, conduction block/temporal dispersion are common
  - Present in 50% by 2 weeks and 85% by 3 weeks
- Motor abnormalties (90%) are more common than sensory (often normal)
  - "Sural sparing" despite median/ulnar SNAP involvement
- No denervation, normal MUAP morphology, reduced recuitment
  - Myokymia *may* be present

CIDP

- >8 weeks
- Prolonged DL, slowing of CV, conduction block/temporal dispersion
- Nerve conductions are often asymmetric
- Secondary axonal changes are the rule
- Needle examination with spontaneous activity, large MUAPs and reduced recruitment

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Common Sources of Error

- Temperature
  - The most important/common source of error
  - Cooler temperatures *slow conduction velocity* and *increase amplitude*
  - CV slows by 1.5-2.5 m/s for every 1° C drop in temperature and DL prolongs by 0.2 m/s
    - UE: 32° C
    - LE: 30° C

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Common Sources of Error

- Age
  - Peripheral myelination is complete by age 3-5 and CVs reach "adult" values
  - After age 60, CV decreases 0.5-4.0 m/s per decade
  - SNAP amplitudes may drop by 50% by age 70

Common Sources of Error

- Height
  - Taller individuals often have slower CVs
    - Nerves taper in size with length (velocity is proportional to diameter)
    - Distal limbs are cooler
  - Most relevant in the measurement of late responses (F-waves)

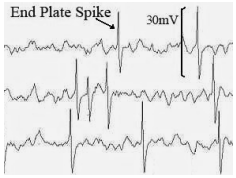
Common Sources of Error

- Stimulation Errors
  - Submaximal Stimulation
    - The largest fibers have the highest threshold for stimulation and thus are evoked last
    - May cause falsely prolonged DL and slowed CV
  - Co-Stimulation of Adjacent Nerves
    - Can lead to spuriously large amplitudes
    - Wave form may change

Electromyography

Spontaneous Activity

- End Plate Noise
  - Miniature end-plate potentials that occur spontaneously at the NMJ
  - Appear as small, monophasic negative potentials
  - "Hissing" or "seashell" noise
- End Plate Spikes
  - Brief spike morphology & irregular pattern
  - Initial deflection is *negative*
  - "Fat in the frying pan"



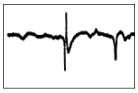
	EPS	Fib
Deflect	Neg	Pos
Firing	Irregular	Regular
Sound	"Frying"	"Rain"

Electromyography

Spontaneous Activity

- Fibrillations
  - Spontaneous discharge of one muscle fiber
  - Indicate *acute* or *active* denervation
  - Initial positive deflection
  - Regular rate
  - "Rain on the roof"
    - I think it sounds more like a metronome

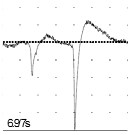
Occurrence of Fibrillations
1. Neuropathic Disorders
2. Inflammatory Myopathies
3. Muscular dystrophies
4. Botulism



Electromyography

Spontaneous Activity

- Positive Waves
  - Spontaneous depolarization a muscle fiber
  - Signify active denervation
  - Regular pattern
  - "Dull pop"



Grading
0. None present
1+. Persistent single trains (>2-3s) in at least two areas
2+. Moderate number or potentials in ≥3 areas
3+. Many potentials in all areas
4+. Full interference pattern (can occur with trauma, vasculitis and/or infarction)

Electromyography

Spontaneous Activity

- Complex Repetitive Discharges
  - Depolarization of a single muscle fiber followed by the ephaptic spread to adjacent denervated fibers
  - Seen in chronic neuropathic & myopathic disorders
    - Can occur in myopathies associated with denervation, necrosis and inflammation
    - Do *not* occur in acute settings
  - "Machine-like" sound

Electromyography

Spontaneous Activity

- Myotonic Discharges
  - Spontaneous discharge of a muscle fiber
  - Can have similar wave form to p-waves (brief positive spike)
  - Waxing and waning pattern (vs regular sound of CRDs)
  - "Revving engine" sound

Conditions Associated with Myotonic Discharges
1. Myotonic Dystrophy
2. Myotonia Congenita
3. Paramyotonia Congenita
4. Myopathies
   1. Acid maltase deficiency
   2. Polymyositis
   3. Myotubular myopathy
5. Hyperkalemic periodic paralysis
6. Colchicine myopathy
7. Cyclosporine toxicity

Electromyography

Spontaneous Activity

- Fasciculations
  - Spontaneous involuntary discharge of a single motor unit
  - Associated with diseases of the anterior horn cell and proximal root
  - "Corn popping" sound
- Doublets & Triplets
  - Similar to fascics ("singlets")
  - Characteristically seen in *tetany from hypocalcemia*

Benign Fasciculation Syndrome

- Most common in the eyelids, arms, legs and feet
  - The tongue *can be* affected
  - No pathologic findings on exam
- Muscle cramping and pain *may* occur
  - AAN Guideline for Symptomatic Treatment for Muscle Cramps:
    - Quinine derivatives should be avoided for *routine use* (can be considered on an individual trial)
    - Vitamin B complex, Naftidrofuryl, and calcium channel blockers are *possibly* effective

Electromyography

Spontaneous Activity

- Myokymic Discharges
  - Essentially grouped fasciculations (repetitive discharges of the same motor unit)
  - "Marching sound"
    - I think it sounds more like bursts of machine gun fire
    - Rest tremor can have a *similar* sound that is more continuous

Conditions Associated with Myokymia

1. Radiation induced nerve injury (esp. brachial plexus injury following radiation therapy for breast cancer).
2. Facial myokymia occurs in GBS, multiple sclerosis and pontine tumors
3. Can be provoked by lowering serum ionized calcium

Electromyography

Spontaneous Activity

- Neuromyotonia
  - High frequency repetitive discharges of a single motor unit
  - Rare
  - "Pinging" sound

Clinical Presentation Associated with Neuromyotonia

1. Generalized stiffness, hyperhidrosis and delayed muscle relaxation after contraction
2. Direct muscle percussion *does not* elicit myotonia (unlike myotonic dystrophy)
3. Activity persists during sleep and during surgery
   1. Abolished by curare
4. Neuromyotonic discharges are most commonly seen in acquired myotonia
   1. Autoimmune condition associated with voltage-gated potassium channels
   2. Can be seen in chronic polio & SMA

Electromyography: MUAP Analysis

- Three major components:
  - Morphology
  - Stability
  - Firing characteristics

Electromyography: MUAP Analysis

- Morphology
  - Duration
    - Reflects the number of muscle fibers within a unit
      - Increases as the number of fibers and the territory of a unit increases
    - Correlates with pitch: long durations sound dull & thuddy, short durations sound crisp (like static)
  - Polyphasia
    - Measure of "synchrony"—the extent to which muscle fibers within a unit fire at the same time
    - Calculated by the number of times the baseline is crossed (normal is 2-4)
    - Polyphasic units have a "clicking" sound
  - Amplitude

Electromyography: MUAP Analysis

- Stability
  - MUAPs should appear stable in morphology from potential to potential
  - Unstable MUAPs occur when individual muscle fibers are blocked
  - Indicates an unstable NMJ
    - Can occur in neuropathic, myopathic or NMJ disorders

Electromyography: MUAP Analysis

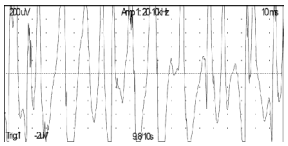
- Firing Characteristics
  - Muscles can increase force by:
    - Increasing the firing rate of motor units
    - Firing additional motor units
  - During maximum contraction, multiple units fire creating an interference pattern, which depends on:
    - Activation: the ability to increase the firing rate
    - Recruitment: the ability to add units as the firing rate increases
    - In a healthy person, 5-6 different MUAPs should be present when fully activated (firing rate of 30-50 Hz)

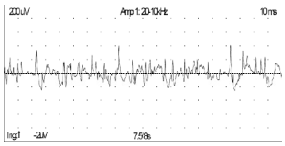
Electromyography: MUAP Analysis

	Duration	Phases	Amplitude	Stability	Activation	Recruitment
Acute axonal	Normal	Normal	Normal	Normal	Normal	**Reduced**
Chronic Axonal	**Prolonged**	**Polyphasic**	**Increased**	Normal	Normal	**Reduced**
Demyelinating	Normal	Normal	Normal	Normal	Normal	Normal
Demyelinatng + Cond. nlock	Normal	Normal	Normal	Normal	Normal	**Reduced**
Acute myopathy	**Short**	**Polyphasic**	**Small**	Normal	Normal	**Early**
Chronic myopathy	**Prolonged**	**Polyphasic**	**Small**	Normal	Normal	**Reduced**

Electromyography: MUAP Analysis

Neuropathic **Myopathic**





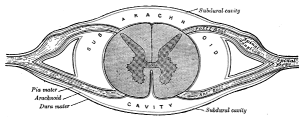
Reduced recruitment of large amplitude MUAPS

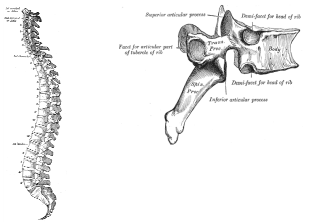
Early recruitment of small, polyphasic units

Specific Patterns

Radiculopathy

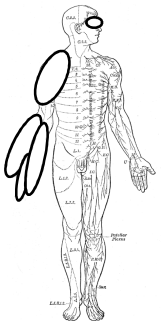
- Nerve conductions are usually *normal*
  - With chronic lesions there may be motor axonal loss
  - Sensory fibers are almost always spared (the lesion is *proximal* to the DRG)
- Neuropathic abnormalities in a myotomal distribution
  - There may be preferential involvement of fascicles





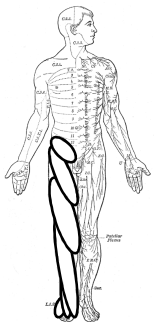
Radiculopathy

- C5: deltoid, supra & infraspinatus; lateral upper arm sensory loss
- C6: biceps, brachioradialis, ECR; thumb/index sensory loss
- C7: triceps, FCR, finger extensors; middle finger
- C8: intrinsic hand muscles; medial hand sensory loss; can be associated with a Horner's Syndrome



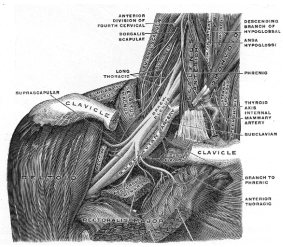
Spinal Cord—Compressive

- L2: Hip flexion weakness (iliopsoas), anterior thigh numbness
- L3: Knee extension (quadriceps); medial thigh numbness
- L4: Knee extension, foot dorsiflexion (Tib anterior, posterior); medial leg numbness
- L5: Great toe dorsiflexion (EHL); numbness of the great toe
- S1: Plantar flexion; digit 5 & lateral foot



Plexopathy

- Plexus lesions *usually* result in abnormal SNAP responses
  - Side-to-side difference of >50% in amplitude
  - Medial antebrachial and lateral antebrachial
- Motor studies are *less useful*
- Needle examination
  - Assessment of the most proximal muscles is important to evaluate root vs plexus
  - Absent CMAPs, profuse denervation and absent MUAPs is an ominous sign for recovery
    - ☞ Surgical exploration may be considered



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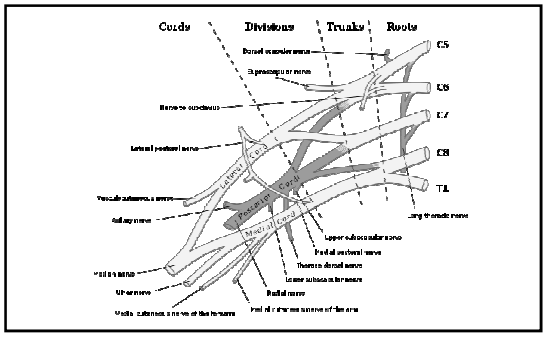
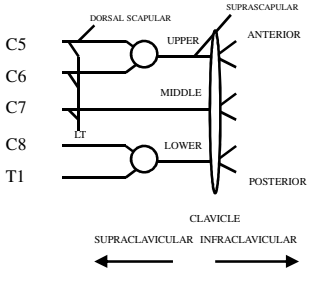
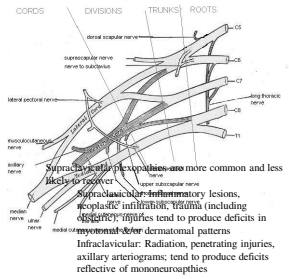
Brachial Plexus



Brachial Plexus

ROOTS TRUNKS DIVISIONS





Supraclavicular plexopathies are more common and less likely to recover

Supraclavicular: Inflammatory lesions, neoplastic infiltration, trauma (including obstetric), injuries tend to produce deficits in myotomal &/or dermatomal patterns

Infraclavicular: Radiation, penetrating injuries, axillary arteriograms; tend to produce deficits reflective of mononeuroapthies

Brachial Plexopathy

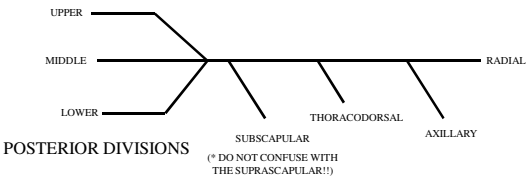
- Disorders of the Brachial Plexus
  - Neuralgic amyotrophy (Parsonage-Turner)
    - Immune-mediated inflammatory process
      - One of the most-common plexopathies (other than trauma)
    - Acute onset shoulder and arm pain followed by weakness and sensory loss
      - 65% of pts have pain at onset; in 90% it is sudden and intense; lasts days to weeks
      - Pts keep the arm in an elbow-flexion/shoulder adduction position
      - Weakness may lag behind pain by weeks (2 or more)
      - Can be bilateral (30%)
    - No specific treatments have been proven to be helpful
      - Steroids may reduce pain but do not alter the disease
      - Significant improvement in 36% at 1 year, 75% at 2 and 89% at 3
      - Recurrence in 1-5%
  - Hereditary Neuralgic Amyotrophy
    - AD inheritance; recurrent brachial plexopathy
    - Typically painful (vs HNPP which is usually painless)

Brachial Plexopathy

- Disorders of the Brachial Plexus
  - Radiation Plexopathy
    - Symptoms occur from 1 month to 18 years after exposure
    - May occur in up to 9% of pts
    - Risks include: higher dose of radiation, # ports, chemotherapy, axillary node dissection
    - Myokymic discharges on EMG are characteristic
      - Occur in 63% of pts
      - Do not occur in neoplastic plexopathies
        - 2' local spread; breast, lung, lymphoma
        - Presence of a Horner's and high signal on T2 favor neoplastic vs. radiation-induced
    - No specific treatment proven
      - Anticoagulation may be helpful

Brachial Plexus

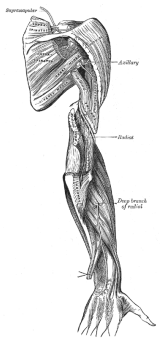
- Posterior Cord (The "STAR")



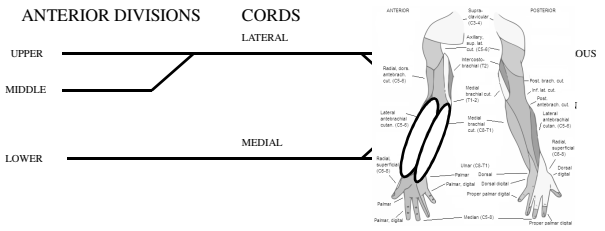
- Cords are named in relation to the axillary artery

Mononeuropathies

- Radial Neuropathy
  - Most commonly compressed at the spiral groove (spares the triceps); can also be compressed at the Arcade of Frohse (formed by the supinator muscle; PIN)
  - Muscles innervated:
    - Radial Nerve (Main Trunk)
      - Triceps
      - Brachioradialis
      - ECR Longus
      - ECR Brevis
    - Posterior Interosseous
      - Supinator
      - EDC
      - ECU
      - EPL
      - EPB
      - APL
      - EIP
      - EDM

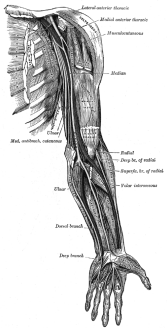


Brachial Plexus



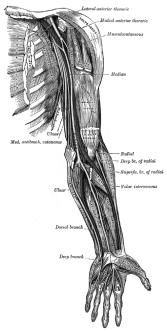
Mononeuropathies

- Musculocutaneous N.
  - Can be damaged with shoulder dislocation & weight lifting (including "carpet carrier neuropathy"
  - Muscles Innervated
    - Biceps brachii
    - Brachialis



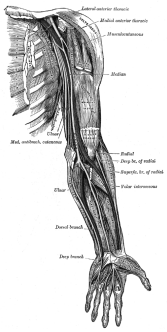
Mononeuropathies

- Median Nerve
  - CTS is the most common entrapment neuroapthy
  - Muscles Innervated
    - Pronator
    - FCR
    - Anterior Interosseous
      - FDP
      - FPL
      - Pronator Quadratus
    - Lumbricals I & II
    - Opponens Pollicis
    - APB
    - FPB



Mononeuropathies

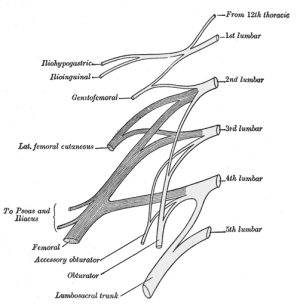
- Ulnar Nerve
  - Commonly entrapped at the elbow (cubital tunnel) and wrist (Guyon's canal)
  - Muscles innervated:
    - FCU
    - FDP
    - ADM
    - Interossei
    - Lumbricals



Lumbosacral Plexopathy

Lumbar Plexus

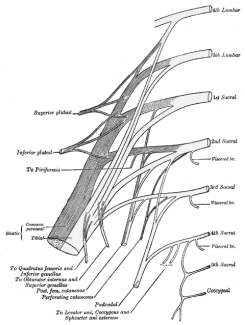
- Two Major Nerves:
  - Femoral (L2-L4)
    - Iliopsoas
    - Quadriceps
    - Pectineus
    - Sartorius
    - Saphenous (sensory)
  - Obturartor
    - Obturator externus
    - Adductor Longus
    - Adductor Magnus
    - Adductor brevis
    - Gracilis



Lumbosacral Plexopathy

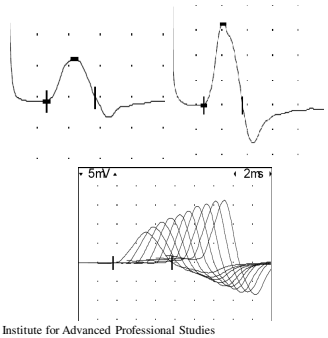
Lumbar/Sacral

- Sciatic
  - Tibial Division
    - Semitendinous
    - Semimembranous
    - Biceps femoris, long head
  - Peronal Divisino
    - Biceps femoris, short head



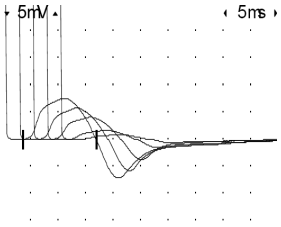
Neuromuscular Junction

- Pre-Synaptic
  - Characterized by facilitation of CMAP amplitudes post-exercise & high-frequency (30Hz)
  - LEMS: >100% facilitation
    - Baseline CMAPs are usually borderline low
  - Botulism: 30% facilitation
    - P-waves/fibs are common



Neuromuscular Junction

- Post-Synaptic
  - Baseline, then 1-minute of exercise, followed by RNS at 1 minute intervals for 3-4 minutes
  - >10-20% decrement after exercise
  - Needle exam may reveal unstable MUAPs



Myopathies

Distal Myopathies

1. Myotonic Dystrophy Type 1 (DM1)
2. FSH Muscular Dystrophy
3. Scapuloperoneal Muscular Dystrophy
4. Inclusion Body Myositis

Myopathies with Denervating Features

1.Polymyositis
2.Dermatomyositis
3.Inclusion body myositis
4.Sarcoid Myopathy
5.Duchenne's/Beckers
6.FSH
7.EDMD
8.Oculopharyngeal MD
9.Acid maltase deficiency

Steroid Myopathy

•Risk increases with use
•Typically proximal
•Abnormal spontaneous activity is *usually* not seen—which may help differentiate from undertreated polymyositis

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Question 1

A 78 year old female with history of breast cancer presents with left arm weakness. Exam reveal weakness of left shoulder abduction and forearm flexion. EMG is notable for myokymic discharges. What is the most likely mechanism of injury?

1. C5/C6 radiculopathy due to herniated disc
2. Post-radiation plexopathy
3. Metastatic invasion of the brachial plexus
4. Left axillary neuropathy

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Question 1

- Answer: Post-radiation plexopathy
- Myokymic discharges are virtually pathognomonic for post-radiation plexopathy.

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Question 2

A 34 year old male presents with two weeks of right foot weakness. Examination is notable for weakness of right ankle dorsiflexion and inversion. Plantarflexion and eversion are normal. What are the most likely findings on the nerve conduction study?

1. Absent right superficial peroneal SNAP response
2. Absent right sural SNAP response
3. Conduction block across the fibular head
4. Spontaneous activity in the lumbar paraspinal muscles

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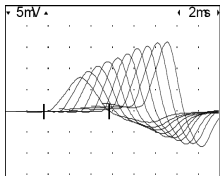
Question 2

Answer: Spontaneous activity in the lumbar paraspinal muscles

This most likely represents a lumbar radiculopathy (L4-L5). Weakness of ankle inversion occurs with involvement of the tibialis posterior (tibial innervated, L4-L5). The peroneus longus (peroneal innervated, L5-S1) everts the foot.

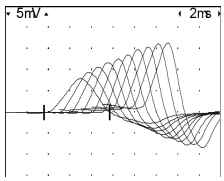
Question 3

- A 67 year old male present with complaint of slowly progressive weakness. Repetive stimulation reveals the following. What is the most likely cause of his weakness?
  1. Presynaptic blockade of ACh release
  2. Presynaptic blockade of calcium channels
  3. Postsynaptic blockade of ACh receptors
  4. Failure of AChRs to congregate at the NMJ



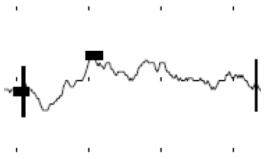
Question 3

- Answer: Presynaptic blockade of calcium channels
- The patient's clinical history and repetitive nerve stimulation are most consistent with Lambert-Eaton myasthenic syndrome, which is typically associated with a P/Q calcium channel antibody. With LEMS, there is >100% facilitation of CMAP amplitudes on RNS.



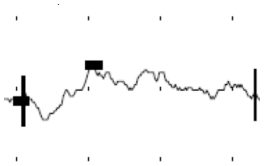
Question 4

- A 38 year old female presents for progressive difficulty walking and distal numbness. Proximal stimulation of the right tibial nerve results in the following CMAP. What is the most likely diagnosis?
  - 1. HSMN I (CMT)
  - 2. HSMN II
  - 3. Vitamin B12 polyneuropathy
  - 4. CIDP



Question 4

- Answer: 4. CIDP
- The image is consistent with temporal dispersion, which is only seen is acquired demyelinating conditions.



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