



**40**  
YEARS OF ADVANCING  
EMERGENCY CARE  
1968 - 2008

### **Life-Threatening Weakness: Strengthen Your Diagnostic Skills**

Acute weakness is a common complaint of patients who present to the ED. The speaker will focus on such neurologic causes of life-threatening weakness, such as Guillain-Barré, myasthenia gravis, and botulism. Key elements of the neurologic examination and diagnostic tests that are helpful in identifying weakness syndromes will be discussed. Diagnostic tips based on deficit location and clinical presentation will be shared.

- Identify severe or life-threatening weakness syndromes.
- Discuss essential elements of the neurologic examination for diagnosing the cause of acute peripheral weakness.
- Discuss the diagnostic workup and treatment of life-threatening causes of weakness.

WE-232  
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McCormick Place - Lakeside Building

(+)No significant financial relationships to disclose

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**Life Threatening Weakness**  
*Strengthen your diagnostic skills*

Eric D. Katz, MD, FACEP  
Program Director, EM Residency  
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**CC: Weakness**

- What percentage of our patients do you think have a chief complaint of weakness?

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**Types of Weakness**

- Localized
- General
- Fatigue
- Breathlessness
- Paralysis
- Distal vs. proximal
- Gait change, grip strength

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## One problem

- Define normal
  - Age?
  - Health status?
  - Comorbidities

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## Characterization

- Rapidity of onset
- Symmetry
- Progression
- Pattern (for example, distal vs. proximal)

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## By the end of this lecture, you will

- Be able to rapidly and reliably identify three major causes of life threatening weakness
- Understand the necessity of rapid identification and treatment
- Understand the emergent management of these life threatening causes of weakness

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### Case #1

- 25yo Japanese male presents with 10 hours of paresthesias in his fingers and heaviness in his legs.
- BP 195/112 HR 122 RR 18 Sat 97%
- 4/5 B ankle flex/ext, 4+/5 B knee flex/ext
- Can't get patellar reflexes but brachioradialis intact
- Bilateral upper extremities grossly normal
- CN intact

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### Guillan Barre Syndrome

- Heterogenous grouping of immune mediated processes generally characterized by:

*Progressive*

*Symmetric and*

*Ascending weakness*

The most common cause of acute flaccid paralysis

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### Trick number one

- If they have DTR's, they don't have Guillan-Barre.

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### Onset

- Rapid progression
- Polyneuropathy
- MAY BE VARIABLE

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### Cause

- Immune mediated axonal demyelination
  - Antibodies to lipopolysaccharides created which cross react with surface layers of gangliosides
- Recovery only occurs after Remyelination.
- Thought to be triggered by recent illness or comorbidities.

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### Prognosis

- 85% will recover fully
  - Usually in 6-12 months
  - 7-15% have persistent neurological dysfunction
  - Mortality <5% in tertiary care centers
  - Rarely recurrent

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### The stuff in the wastebasket...

- 40% of GBS is AIDP subtype
  - (90% in US, Europe and developing world)
  - Acute demyelinating polyneuropathy
  - 40% positive of *Campylobacter*
  - Remyelinates reliably

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### The stuff in the wastebasket...

- Acute motor-SENSORY axonal neuropathy (AMSAN)
  - Variant which affects sensory nerves as well
  - Adult onset
  - Worse recovery, muscle wasting
- Acute motoraxonal neuropathy (AMAN)
  - Predominantly peds
  - One third HYPERreflexic
  - Two thirds positive for *Campylobacter*

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### Miller-Fisher Variant

- RARE
- Classic triad:
  - Ataxia*
  - Areflexia*
  - Ophthalmoplegia*
- Acute onset
- 1-3 month recovery time

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### Typical GBS History

- 2-4 weeks after benign URI or GI illness
- Weakness in legs
  - Possibly dysesthesias in hands
- Moves upwards (symmetrically) over hours to days
- Can involve respiratory muscles
- Can involve cranial nerves, even if ascending type
- Worst at 2 weeks, plateau's for 4 weeks, resolves

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### Sensory

- Paresthesias usually stay in hands and feet
- Pain in largest muscle groups – with movement
- Loss of proprioception/vibration, pain, light touch

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### Autonomic Dysfunction

- Tachy or bradycardia
- Postural hypotension OR wide BP fluctuations
- Constipation
- Flushing
- Hypersalivation
- Anhydrosis
- Tonic pupils

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## Respiratory function

- 35% will need mechanical ventilation
  - Prediction?
    - Symptoms
    - Exam?
    - CXR?
    - ABG?
    - Pulse oximetry?
    - NIF?

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## NIF

- What is normal?
- What indicates a need for intubation?
- How good are your nurses?

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## Labs

- Not very helpful
- Some recommend a wide spectrum of antibody titers
  - No real effect on clinical course
- Check an HIV test
- Look for SIADH
- Stool for *C. jejuni*
- LP for protein/bands

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## ED management

- Assessment of airway stability
- Good neuro exam – focusing on motor strength and reflexes
- Reevaluation q1-2 hours until out of department
- Treatment of hypertension with short acting meds ONLY due to fluctuations
- Watch for arrhythmias – treat normally
- Role of IVIG?

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## Case #2

- **28 yo Caucasian female presents with diplopia, garbled speech and trouble swallowing.**
- HR 110 Temp 38.2 BP 126/72 RR 24 Sat 96%
- Trouble smiling
- Eyes tearing, slight drool, dry axilla, N/V/D
- Weak cough
- Rales and rhonchi in RLL
- Extremities normal
- Reflexes normal

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## Myasthenia Gravis

- What is it?
- Antibodies against acetylcholine (ACh) receptors
  - Reduces total number of receptors

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**Cardinal Features**

- Symmetric
- Progressive
- Generalized vs. bulbar muscles
- Often chronic and recurrent
- Painless
- Reflexes PRESERVED

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**Characteristic sequence of weakness**

1. Eyes
2. Extra-ocular muscles (ptosis, diplopia)
3. Facial Muscles (expression, speech)
4. Pharyngeal (mastication, deglutition)
5. Neck/proximal limbs (usually not involved)

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**Severe exacerbations**

- Slack facial muscles
- Unable to support head
- Nasal Voice
- Limp body
- Loss of gag reflex

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## Possible presentations

1. Respiratory Distress
  - Ventilation
  - Clearance of bronchial secretions
    - Rales, rhonchi or wheezing
2. Cholinergic crisis (SLUDGE)

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## Trick #2

- Myasthenia always gets worse with repetitive muscle use and gets better after rest
- We'll talk about the edrophonium test

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## Other ED tricks

- Ice packs
  - Patients with ptosis
  - 2 minute application
  - 80% sensitivity
- Pyridostigmine for milder cases
- Antipyretics to lower temp

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## Etiology

- Idiopathic
- ACh tries to bind to receptors, but fewer and fewer are available over time (progressive)
- ONLY affects striated muscle
- 75% have thymic disease (useful in treatment) but actual trigger is unknown

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## Characteristics

- 0.5-14.2/100,000 people
- Asians slightly more common
- Females > males (3:2)
- Peak ages:
  - Female: 20-30
  - Male: >50

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## Morbidity/Mortality

- 1980's: 30-70% mortality
- 2000's: near-normal life expectancy
- Aspiration due to weakened musculature
- Falls
- Side effects of medications
- Myasthenic crisis, if recognized, mortality <5%

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### **Predictors of Crisis**

- Prior diagnosis
- Temperature, emotional state
- MEDICATIONS:
  - Antibiotics
  - Antidysrhythmics
  - Miscellaneous
- Thyroid abnormality (hyper OR hypo)

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### **Myasthenia meds**

- Which looks like a crisis? Too much or too little?
  - BOTH!
  - Myasthenic crisis vs. cholinergic crisis

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### **Labs / Imaging**

- ABG
- Cultures
- Otherwise, nothing specific
  
- CT chest for thymoma
  - Check TFT's before iodine

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### ED Tricks

- Tensilon (edrophonium)
    - Useful for diagnosis
    - Differentiates myasthenic from cholinergic crises!
    - Side effects: bradycardia, heart block, asystole!
1. Stabilize airway
  2. 1 mg edrophonium
  3. If no effect or adverse reaction, 3mg
  4. If no effect or adverse reaction 5mg

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### ED tricks

- Watch for RAPID decompensation
- Low threshold for airway stabilization
  - Often resistant to succinylcholine
  - Use higher doses
  - Often prolonged activity of succinylcholine
  - Non-depolarizing agents preferred

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### Case #3,4,5 and 6

- A family comes in with presentations of diplopia, dysarthria, dysphagia. One has a sore throat, another has difficulty lifting his head. Two have dilated, unreactive pupils. Gag reflexes are diminished or absent.
- VS normal
- CN's show multiple deficits. One has neck weakness.
- All have normal sensation
- All are fully alert

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## Botulism

- Paralytic disease
- Caused by toxins from *Clostridium botulinum*
  - Found in soil and marine life worldwide
- Toxin HIGHLY potent
- Heat stable at 100°C but not 120°C

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## 6 forms

1. Food-borne
2. Wound
3. Infant
4. Adult intestinal
5. Injection
6. Inhalational

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## Foodborne

- 1000 cases/year worldwide
- Multiple food sources
- 94% are from home processed foods
  - Most of the rest are from mishandling of packaged
- Onset from 6h-10d (36h) of ingestion
  - Rare patients with colonization for 1-2 months

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## Infantile

- 2% mortality
  - May be underestimated due to involvement with SIDS
- Risks
  - High birthweight, older mothers, educated parents
  - Constipation

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## Wound

- Handful of cases each year
- Most in California
- Overwhelming majority involve skin popping

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## Pathology

- Toxin binds to presynaptic terminals of PERIPHERAL cholinergic synapses
- Internalized
- Within the cell, it lyses proteins responsible for ACh release
- Permanent
  - Sx resolve when the axon makes another terminal

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### Classic presentation

- Symmetric
- Descending motor paralysis
- Involves cranial, spinal and peripheral nerves

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### ED Tricks

- They have no CNS change so they are alert
- They have no sensory changes
  
- Afebrile unless coinfection
- Normal or slow heart rate unless hypotensive

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### The Dozen D's

- Dry mouth
- Diplopia
- Dilated pupils
- Droopy eyes
- Droopy face
- Diminished gag
- Dysphagia
- Dysarthria
- Dysphonia
- Difficulty lifting head
- Descending paralysis
- Diaphragmatic paralysis

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### Other symptoms

- Sore throat
- Nystagmus
- Ataxia
- Paresthesias (14%)
- Paralytic ileus
- Constipation (severe)
- Urinary retention
- Orthostatic hypotension

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### Morbidity/Mortality

- 7-10% mortality
  - Doubles in geriatrics
  - Decreases with prompt antitoxin use
  - Death usually due to poor recognition
  - Underlying infection increases mortality
- Usually require 6-8 weeks of vent support then 1-2 years of respiratory rehabilitation

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### The other types

- Infant
  - 1.3% mortality
  - 5% relapse
  - 1-2 week progression and 2-3 before recovery
- Wound
  - 10% mortality
  - Longer medical care

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### **Labs**

- Identification is key but can't wait for micro.
- Get 15 cc serum, 50g feces and gastric aspirate (if NGT in place)
  - Send to state health department

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### **Antitoxin**

- Antitoxin can bind toxin in blood, not once it is in the nerve
- Equine!
- Dose: "One vial"
- Antitoxin administration IV
  - For wound botulism, debride wound then inject antitoxin. Consider HBO.
- Measure FVC and NIF

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### **BabyBIG**

- Human Botulinum Immune Globulin
- For infant botulism
- Have to get from state health services

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### So our big three

- Guillan-Barre
- Myasthenia Gravis
- Botulism
  
- All rare
- All have potential for “saves”
- All have specific tricks for rapid identification

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### End of conference learning

1. Neuromuscular diseases, while rare, have high fatality rates. The biggest predictor of survival is rapid identification.
2. Careful neuro exam will identify all three of these diagnoses reliably and quickly
3. Monitor neuromuscular disease patients aggressively for respiratory failure and coinfection.

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**Thank you for your time!**

Have a safe flight home!

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