APPROACH TO WEAKNESS

WEAKNESS IS A SUBJECTIVE TERM

THE KEY DIFFERENTIATION IS WHETHER THE PATIENT HAS ACTUAL, QUANTITATIVE MUSCLE WEAKNESS!

PATHOPHYSIOLOGY:

- NEUROMUSCULAR WEAKNESS:
  - Muscle contraction is the result of a series of events:
    - UMN begin in motor cortex → descend in lateral corticospinal tract (on opposite side to point of origin in the brain) → UMN synapses on LMN in anterior horn → LMN carries signal to muscle bundle → LMN releases acetylcholine in to synaptic clef → depolarises motor endplate
  - NEUROMUSCULAR WEAKNESS CAN BE CAUSED BY LESIONS OR DERANGEMENTS AT ANY LEVEL ABOVE
- NON-NEUROMUSCULAR WEAKNESS:
  - Multiple possible causes

DIFFERENTIAL DIAGNOSIS:

Non-neuromuscular

Critical: Hemodynamic instability
- Myocardial infarction
- Arrhythmia
- Severe infection/sepsis
- Respiratory failure
- Hyperkalemia

Emergent

- Acute anemia
- Dehydration
- Metabolic disorder
  - Hypothyroidism
  - Diabetes
- Electrolyte imbalance

Other

- Fatigue
- Psychiatric (anxiety, depression)
- Rheumatologic (fibromyalgia; SLE)
- Malignancy
- Renal or hepatic disease
- Metabolic disease
- Alcoholism and other toxin-related disease
- Malingering

Neuromuscular

Critical: Potential for respiratory compromise
- Rabies
- Botulism
- Tetanus
- Organophosphate poisoning
- Myasthenia gravis crisis

Emergent

- Guillian-Barré syndrome
- Transverse myelitis
- Impingement syndromes
- Spinal cord infarction
- Electrolyte imbalance

Other

- Lambert-Eaton syndrome
- ALS
- Paraneoplastic syndrome
- Diphtheria
- Porphyria
- Drugs and toxins
- Tick paralysis
- Poliomyelitis

- **CLEAR HISTORY IS VITAL**
  - Time course
  - Presence or absence of:
    - Bladder/bowel dysfunction
    - Sexual dysfunction
    - Altered sensation
    - Muscle pain or spasm
  - Medications → think electrolyte imbalance
  - Infective symptomatology → in elderly patients "weakness" may the only symptom!
- **EXAMINATION:**
  - Assess ability to maintain airway if severe weakness present
  - Systematic/thorough motor exam
  - Consider normal pressure hydrocephalus in those who have simultaneous incontinence and decreased cognitive function
  - DIFFERENTIATE BETWEEN UMN AND LMN PROBLEM

Table 11-3 Physical Examination: Localizing Neuromuscular Lesions

LOCATION OF LESION	DEEP TENDON REFLEXES	MUSCLE TONE	PLANTAR REFLEXES	STRENGTH
Upper motor neuron	Increased	Normal (Increased/spastic as disease progresses)	Upgoing	Weak/paralysis
Lower motor neuron	Decreased or absent	Decreased/flaccid (may see fasciculations)	Normal or absent	Weak/paralysis
Neuromuscular junction	Normal or decreased	Decreased/flaccid	Normal or absent	Variable weakness pattern
Muscle	Normal or decreased	Decreased/flaccid	Normal or decreased	Constant/progressive Proximal > distal

CHARACTERISTIC PRESENTATION OF THOSE WITH NEUROMUSCULAR WEAKNESS:

Table 11-1 Neuromuscular Diseases: A Brief Description

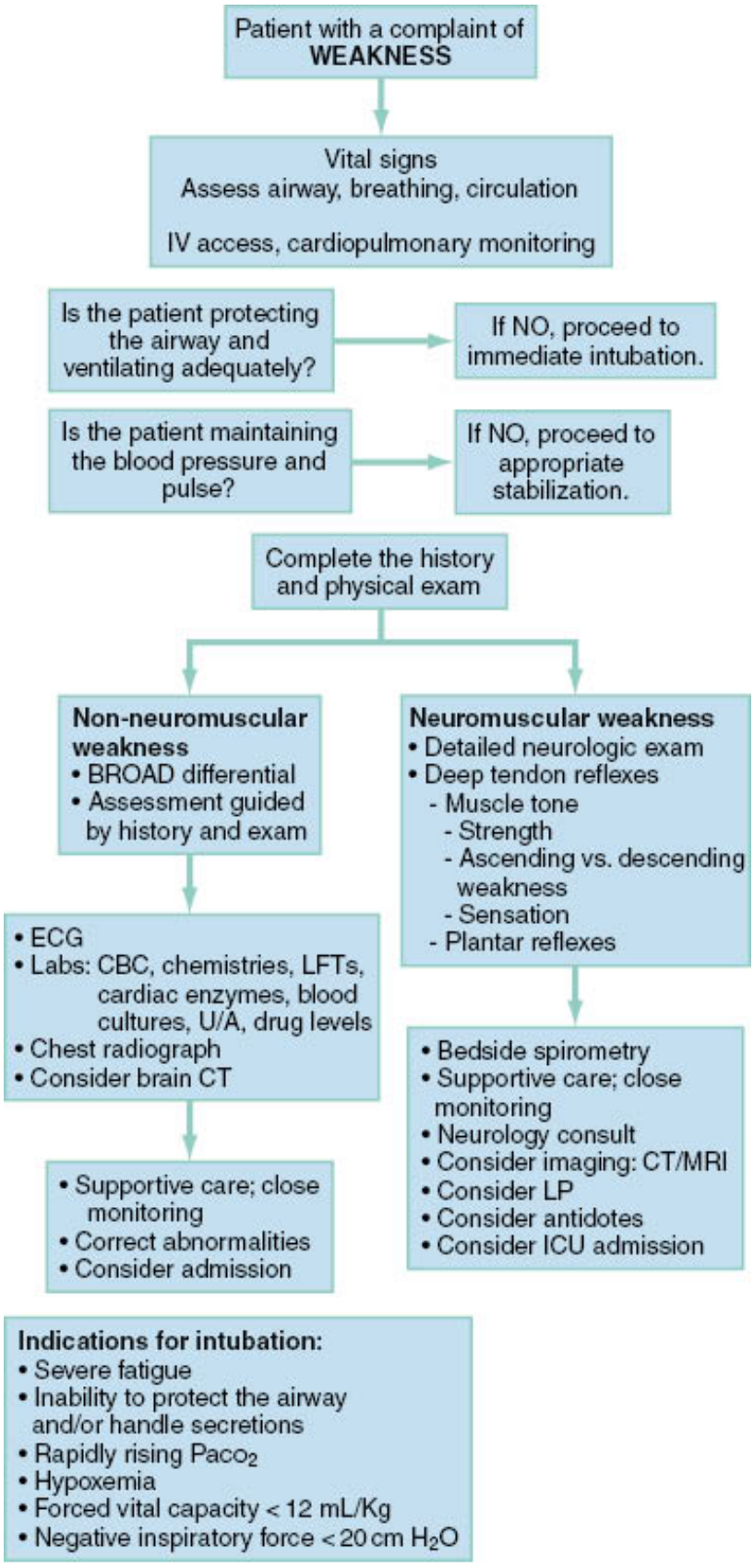
DISEASE	MECHANISM	HISTORICAL FEATURES/EXAM FINDINGS	ED MANAGEMENT
Botulism Toxin 12–72 hr postingestion	Neurotransmission Toxin prevents release of acetylcholine at the neuromuscular junction	Ingestion of contaminated canned goods 50% have GI symptoms Postural hypotension Diplopia, blurred vision, ptosis, facial weakness, dysphagia, respiratory compromise, then limb weakness	Supportive care, ICU admission Notify Health Dept/CDC Trivalent antitoxin (May try guanidine hydrochloride, facilitates release of acetylcholine from nerve endings; anticholinesterase drugs not helpful)
Myasthenia Gravis Idiopathic	Neurotransmission Decreased number of functioning acetylcholine receptors	Mild infection may exacerbate symptoms Fluctuating weakness; easy fatigability of voluntary muscles; cranial nerves involved with ptosis and diplopia in >25%; normal pupillary responses; normal sensation; normal reflexes Improves with rest May have a coexisting thymoma (CXR, chest CT)	Supportive care, ICU admission Neurology consult Edrophonium/neostigmine test Bedside spirometry Measure serum acetylcholine receptor antibody levels Tx: Anticholinesterase drugs—neostigmine; pyridostigmine
Organophosphate/ Carbamante Poisoning Immediate–3 wk	Neurotransmission Cholinergic crisis from inhibition of acetylcholine Neuropathy (weeks after exposure)	History of insecticide exposure Gastrointestinal symptoms, agitation, miosis, paralysis, diaphoresis, muscle weakness, bradycardia Cramping muscle pain, distal numbness and paresthesias, progressive muscle weakness; decreased reflexes; can develop flaccid/ wasted leg muscles	Decontamination Supportive care, ICU admission Atropine Pralidoxime (2-Pam)
Tetanus Toxin 3 wk 10–60% fatality	Neurotransmission Toxin interferes with release of inhibitory transmitters including GABA; results in motor nerve hyperactivity	Immunization status History of cutaneous infection Trismus, laryngospasm, painful muscle spasms and rigidity (opisthotonos), autonomic instability	Supportive care, ICU admission Débridement of wounds Tetanus immunoglobulin Penicillin for the infection High-dose benzodiazapines Neuromuscular blockade
Tick Paralysis Toxin 2–7 days Rocky Mountain wood tick and the American dog tick are most common in the USA	Neurotransmission Toxin reduces motor neuron action potential and the action of acetylcholine	History of outdoor activities/tick bite Progressive, ascending, flaccid weakness over several hours may lead to respiratory failure; may present as acute ataxia without muscle weakness; decreased or absent reflexes; ophthalmoplegia and bulbar palsy can occur	Removal of the embedded tick (look at the hairline/in the scalp) Supportive care Full recovery if tick removed; 10% fatality if not recognized
Ciguatoxin Toxin 12–24 hr; neurologic symptoms can last months	Neuropathy Toxin causes cell membrane excitability and instability	History of ingestion of large, tropical fish Diarrhea, abdominal pain, nausea, and vomiting are followed by painful paresthesias, ataxia, altered hot/cold perception, myalgias, bradycardia, and hypotension Rarely, death occurs through respiratory failure	Supportive care, ICU admission Atropine for bradycardia Hydration IV mannitol can be helpful
Diphtheria Toxin 2 wk–3 mo after infection	Neuropathy Lower motor neuron	Immunization status History of throat infection with pseudomembrane; cutaneous infection Palatal weakness, impaired pupillary responses, generalized sensorimotor polyneuropathy; respiratory failure; motor weakness of the proximal muscle groups and extending distally	Supportive care, ICU admission Equine diphtheria antitoxin Erythromycin or penicillin G for 14 days to halt toxin production, treat localized infection and prevent transmission of organisms Immunization

Table 11-1 Neuromuscular Diseases: A Brief Description—cont'd

DISEASE	MECHANISM	HISTORICAL FEATURES/EXAM FINDINGS	ED MANAGEMENT
Gullian-Barré Syndrome Idiopathic 1–4 wk 75% recover 5% fatality	Neuropathy Lower motor neuron Immune-mediated polyneuropathy Multiple variants	May have a history of infection; viral infection; *Campylobacter jejuni* in 15–40% Symmetrical ascending motor neuropathy; decreased/absence reflexes; mild sensory involvement; autonomic dysfunction; can progress to respiratory compromise	Lumbar puncture: CSF with elevated protein but normal WBC Bedside spirometry Plasmapheresis and IVIG Consider ICU admission Neurology consult
Transverse Myelitis Idiopathic, postinfectious, autoimmune Rapid onset (hours–days)	Neuropathy Upper motor neuron Axonal demyelination	Loss of spinal cord functions with symptoms depending on the level of the lesion; thoracic is most common Acute, focal back pain; distal muscle weakness; abnormal sensation; urinary retention/loss of bowel control; muscles may be flaccid; decreased or absent reflexes initially Differentiate from spinal cord compression, trauma or infarct; may be the first sign of multiple sclerosis	Supportive care, ICU admission if C-spine level for respiratory support Spine radiograph to evaluate for boney lesion Stat MRI/CT myelogram Decompress bladder
Electrolyte Imbalance	Myopathy	History of nausea/vomiting/diarrhea History of renal failure, alcohol dependence, new medication Ascending symmetric muscle weakness with normal to diminished reflexes	ECG Electrolyte panel: Na, K, Cl, PO_4, Ca, and Mg Renal function Correct the abnormality; close hemodynamic monitoring
Polymyositis Autoimmune	Myopathy	History of connective tissues disorders or cancer Progressive at a variable rate; muscle weakness and wasting; ascending pattern with proximal limb and girdle muscle involvement; muscle pain; dysphagia; respiratory difficulty; can have an erythematous periorbital and eyelid rash (dermatomyositis)	Elevated CPK, rhabdomyolysis rare Normal ESR Supportive care Corticosteroids

INITIAL MANAGEMENT AND APPROACH:

- In a patient with neuromuscular weakness, the respiratory drive is PRESERVED, but the ability to ventilate adequately may be impaired
- WARNING SIGNS OF WORSENING RESPIRATORY STATUS:
    - Inability to lift head
    - Ineffective cough
    - Alteration of voice
    - Difficulty controlling secretions
- AVOID SUCCINYLCHOLINE when progressive denervation syndrome is thought likely due to likelihood of hyperkalaemia


Patient with a complaint of WEAKNESS
Vital signs
Assess airway, breathing, circulation
IV access, cardiopulmonary monitoring
Is the patient protecting the airway and ventilating adequately?
If NO, proceed to immediate intubation.
Is the patient maintaining the blood pressure and pulse?
If NO, proceed to appropriate stabilization.
Complete the history and physical exam
Non-neuromuscular weakness
• BROAD differential
• Assessment guided by history and exam
Neuromuscular weakness
• Detailed neurologic exam
• Deep tendon reflexes
- Muscle tone
- Strength
- Ascending vs. descending weakness
- Sensation
- Plantar reflexes
• ECG
• Labs: CBC, chemistries, LFTs, cardiac enzymes, blood cultures, U/A, drug levels
• Chest radiograph
• Consider brain CT
• Bedside spirometry
• Supportive care; close monitoring
• Neurology consult
• Consider imaging: CT/MRI
• Consider LP
• Consider antidotes
• Consider ICU admission
• Supportive care; close monitoring
• Correct abnormalities
• Consider admission
Indications for intubation:
• Severe fatigue
• Inability to protect the airway and/or handle secretions
• Rapidly rising $Paco_2$
• Hypoxemia
• Forced vital capacity < 12 mL/Kg
• Negative inspiratory force < 20 cm H_2O

SPECIAL SITUATIONS:

- MYASTHENIC CRISIS:
    - Refers to rapid worsening of neuromuscular function with respiratory compromise
    - TRIGGERS:
        - INFECTION (~30%)
        - Change in medications
            - Dose of anticholinesterase
            - Tapering of steroids
            - Use of:
                - Aminoglycosides
                - Quinolones
                - β-blockers
        - Metabolic derangement
        - Physical stress
        - IDIOPATHIC (1/3)
- CHOLINERGIC CRISIS:
    - Excess of cholinesterase inhibitor medications that produces flaccid muscle paralysis and generalised weakness