Degenerative Diseases

Parkinson's Disease

Huntington's

Multiple Sclerosis

Amyotrophic Lateral Sclerosis

Myasthenia Gravis

Restless Leg Syndrome

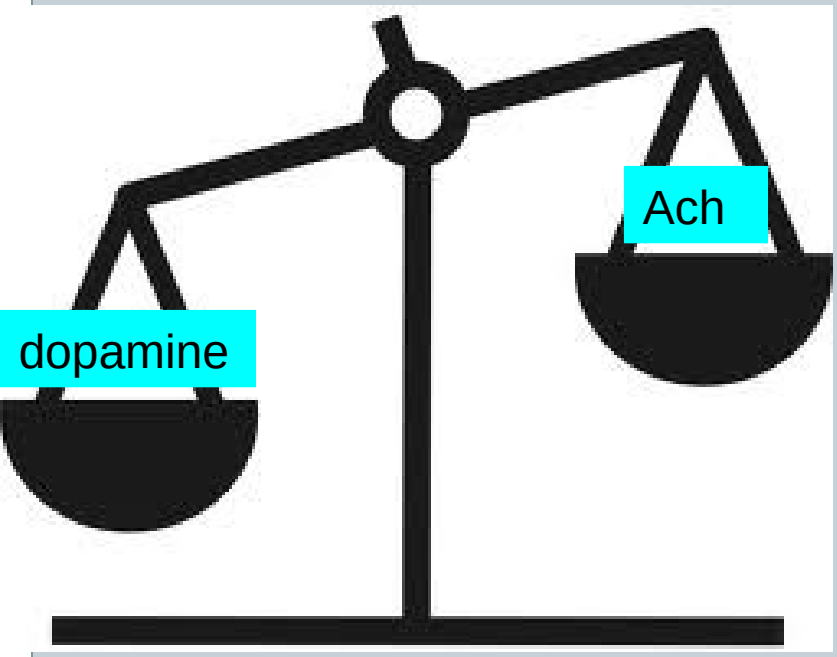
Parkinson's Disease

- Slowly progressive neurologic movement disorder
  - Initiation of movements slow down
  - Increased muscle tone
  - Tremors at rest
  - Gait disturbances
- Risk Factors
  - Men
  - Young onset PD rare but
    - Slower disease progression
  - Risk increases with age

Etiology

- Cause is unknown
  - Lack of dopamine in the brain
  - Degeneration of cells in midbrain that produce dopamine
  - Genetic/environmental factors could play a part?
- Parkinsonism
  - Tremors, Rigidity, Akinesia (Bradykinesia), Postural Instability set of symptoms

Pathophysiology



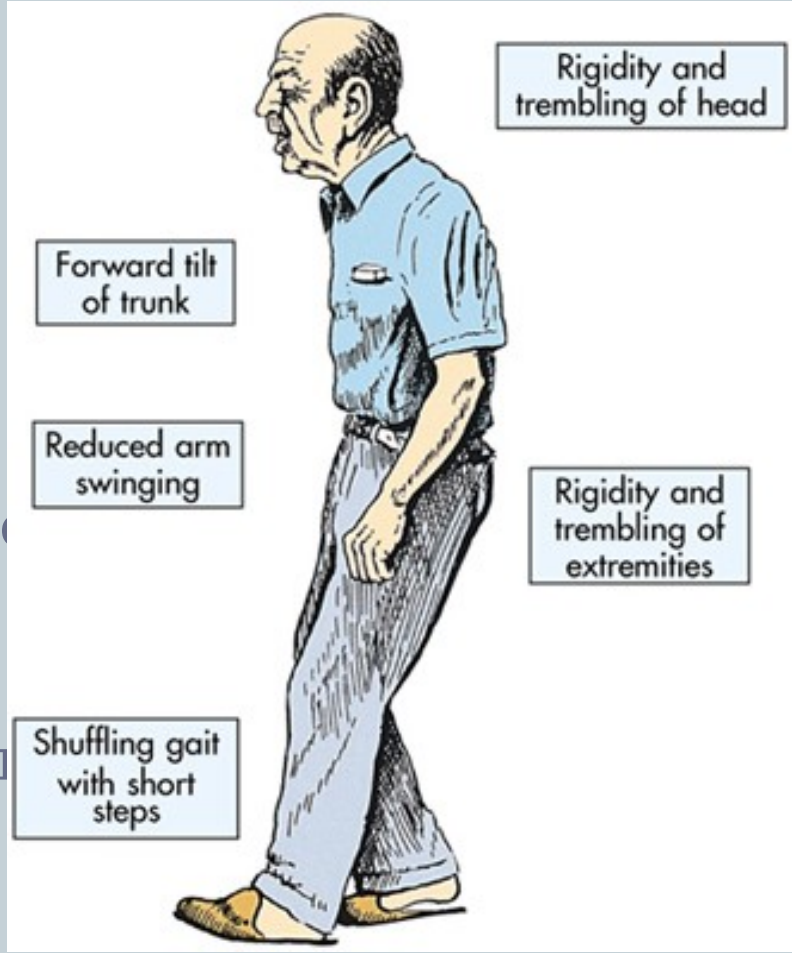
= Parkinson's Disease

- Degeneration of dopamine producing neurons
- Decrease in dopamine and increase in acetylcholine
  - Usually we have a balance of the two neurotransmitters
  - Now the scale is tipped
- Dopamine needed for
  - Posture control
  - Smooth voluntary movements

Clinical Manifestations: Cardinal

- Cardinal Signs
  - Tremor
  - Rigidity
  - Akinesia (bradykinesia)
  - Postural Instability
- Other
  - Psychiatric- depression/anxi
  - Pain, fatigue, memory loss
  - ANS s/s: sweats, flushing

  Constipation, urinary retentio
  - Sleep Disorders



Diagnosis & Medical Management

DIAGNOSIS

- Symptoms- is there TRAP?
- R/O other causes (stroke, brain tumor, drug use)
- Positive response to anti-parkinsonism meds

MANAGEMENT

- No cure
- Controlling symptoms is goal
- Maintain independence for as long as possible

Medication Management

- Sinemet=

Carbidopa/ Levodopa

  - Weeks to months before effects are seen
  - Works best for first few years, then could stop working. Wears off. Turns off some days/works others.
    - Teach to take at same time each day. Never stop abruptly

- Side Effects
  - Dyskinesia
    - Involuntary movements
  - On off syndrome

Medical Management

- Dopamine Agonists
  - Stimulate dopamine receptors to create more availability
    - Ropinirole, pramipexole, bromocriptine
    - Could affect blood pressure and cause orthostatic hypotension
- Anticholinergics
  - Block activity of Ach to create a better balance of the two neurotransmitters
    - Benztropine
- Antihistamines can control tremors
- Antivirals
  - Promote release of dopamine
    - Amantadine

**Avoid meds such as: metoclopramide, prochlorperazine which could block dopamine causing worsening imbalance

Nursing Diagnosis and Care

- Impaired Physical Mobility
- Risk for Falls
- Self-Care Deficits
- Impaired verbal communication
- Imbalanced nutrition: less than body requirements
  - Constipation
- Ineffective Coping
- R/F Aspiration

Huntington's Disease

- Progressive, degenerative brain disorder
- Results in excessive involuntary choreiform movements and mental deterioration (cognitive & psychiatric effects)
  - Writhing and twisting of face, limbs, body
  - Cognition, personality, emotions all affected
- Abnormal gene HTT identified
- Autosomal Dominant= 50% chance to pass to offspring when a parent is affected

Pathophysiology

Dopamine

GABA and Ach

=

Excess

Clinical Manifestations

- Chorea
- Facial Movements
  - Tic, grimace
- Speech
  - Slurred, hesitant, explosive
- Chewing/Swallowing difficulties
- Gait is disorganized
- Bladder/Bowel control lost
- Mental status changes
  - Cognition, personality, and emotional changes
  - Difficult for family

Diagnosis

- Clinical presentation
  - Characteristic symptoms
- Family History of Disease
- Genetic Marker

Management

- Medications
  - Treat movements
    - Xenazine decreases dopamine availability
    - Deutetrabenzine
    - Neuroleptics
  - Cognitive and Psychiatric Symptoms
    - Counseling and Psychiatric Meds
- Safety
- Nutrition
- Altered mentation and social interaction
- Collaborative Care
- Prognosis

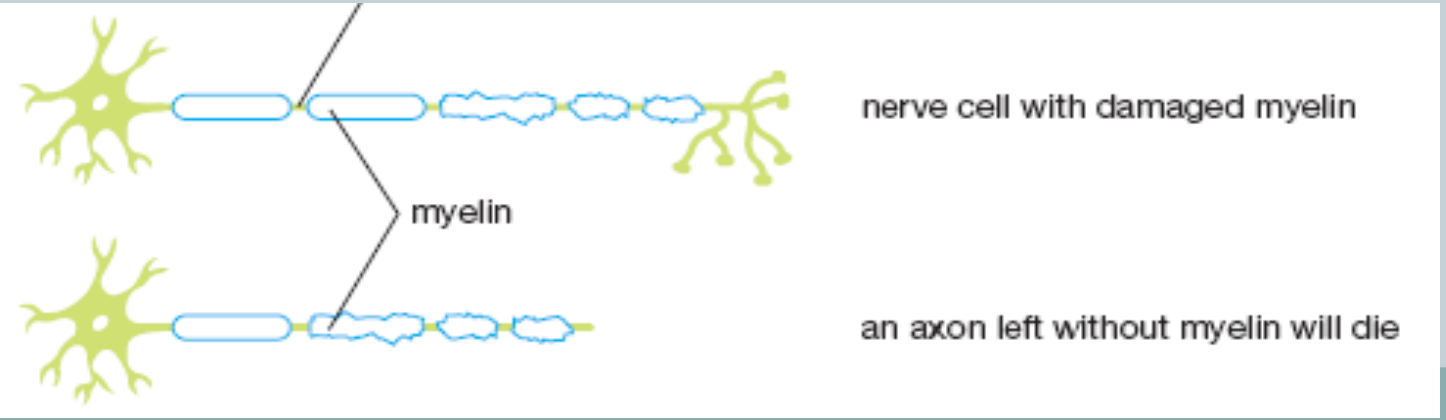
Multiple Sclerosis

- Chronic
- Progressive
- Degenerative
- Myelin sheath damage
- Affects the brain and/or spinal cord
- Nerve conduction issues are seen

- Onset 20-50 years of age
- Women 2x the risk
- Cause unknown
  - Autoimmune factors present though
- Autoimmune response leads to T-cells entering the circulation and attacking the body. These cells cause inflammation and demyelination of the nerves

Pathophysiology

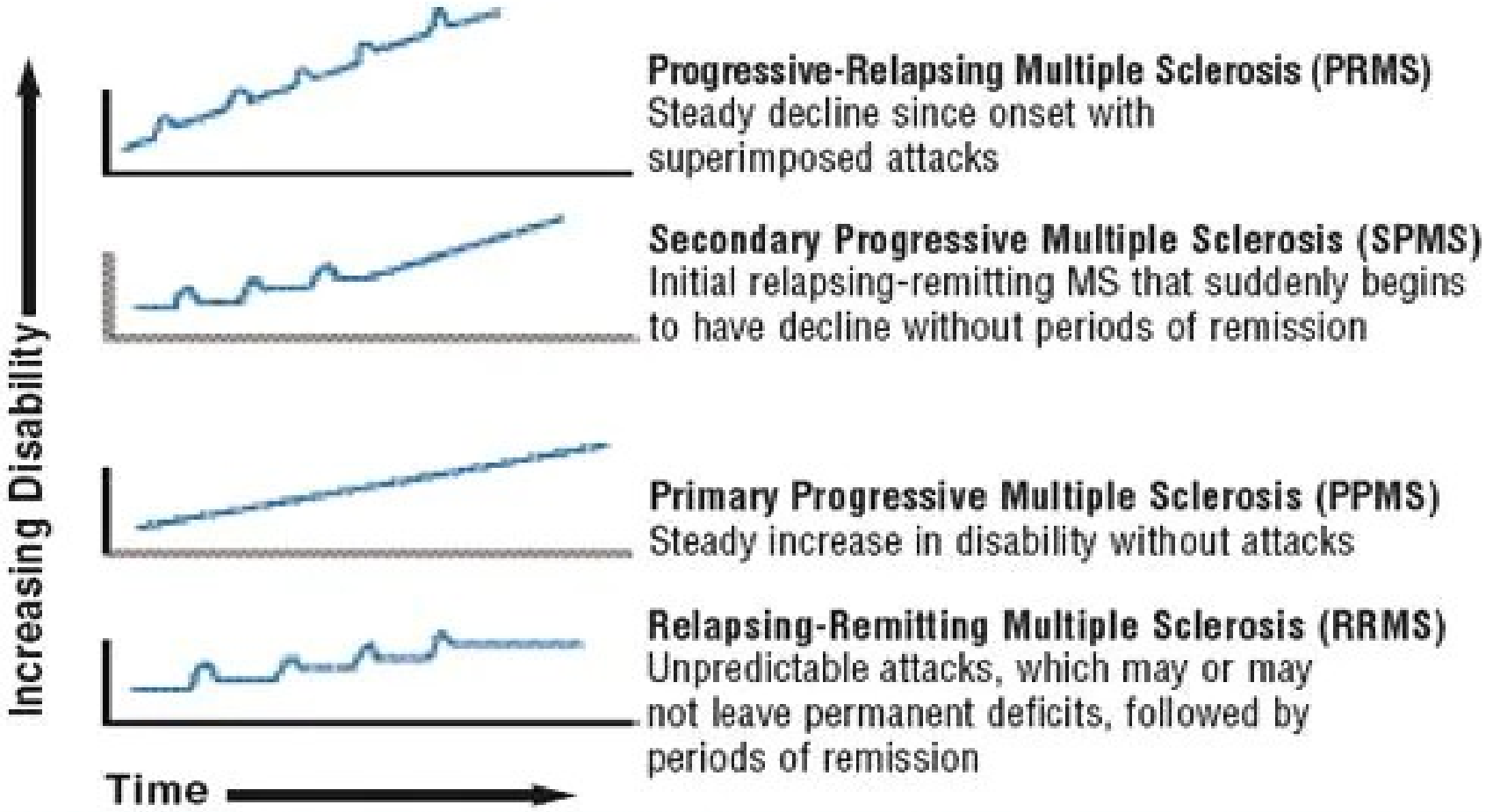
- Autoimmune response leads to:
  - chronic inflammation
  - myelin sheath damage
  - and scar formation



Signs and Symptoms

- Fatigue-often severe
- Impaired Movement
  - Weak limbs/heavy limbs
- Optic Nerve
  - Diplopia, blurry, color issues, blindness
- Acoustic Nerve
  - Tinnitus, hearing difficulty
- Dysarthria and Dysphagia
- Bowel/Bladder Dysfunction
  - Constipation, incontinence
- Sensory Disturbance
  - Paresthesia
  - Pain- Lhermitte's sign
- Cognitive/Emotional
  - Seen later in the disease
  - Decreased concentration/memory
  - Depression, anger

Categories/Classifications of MS

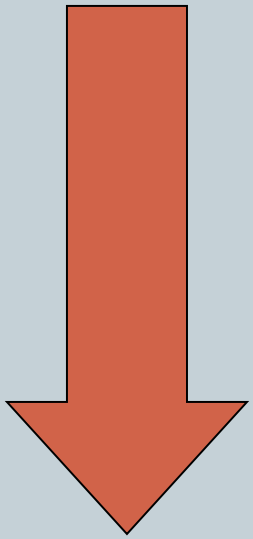


Diagnostics

- No definitive test
- Look at S+S
- CSF analysis via LP
  - Increased WBCs
  - Ig G bands- oligoclonal bands show autoimmune response happening in the CNS
- MRI-main dx tool to look for lesions/plaques/scarring through the CNS

Disease Modification: Interferons

- Interferons- Avonex, betaseron, rebif
  - Are natural protein that communicate in the immune system.
- Blocks T cells and immune response that causes inflammation
- Require injection
  - Rotate sites
- SE: flu like symptoms



Goal: Decrease in MS attacks or disease activity

Others

- Immunomodulators
- Anti- inflammatory
- Immunosuppressants
- A lot of medications available. See p. 1370 for various options.

- Meds have many side effects, potential adverse rxns
- Can lower immune system ability to fight infection
- Can cause cancer, liver dysfunction, neutropenia
- Meds do not cure, only potentially modifies course or delays course-delays exacerbations/relapses is the hope
- Drugs are very expensive

Acute Exacerbation Management

- Need fast acting medication to decrease edema and inflammation that leads to demyelination
  - IV admin
    - Steroids
    - Methylprednisone or Prednisone
- Discharged with instructions to taper down using oral prednisone
  - To prevent return of exacerbation

Management

- Urinary Retention
  - Cholinergics stimulate bladder contraction
- Spastic Bladder
  - Anticholinergic relax bladder
- Fatigue
  - Treat with CNS stimulants
  - Methylphenidate (Ritalin)
- Spasticity
  - Muscle relaxants

    - Diazepam
- Bradykinesia
  - Dalfampridine
- Pain and Paresthesia
  - Wide array of choices
  - Antidepressants/ anticonvulsants
- Cognitive changes
  - Therapy

Other Therapies

- Physical Therapy
  - Keep patient functional as possible
- Water Exercise
  - Buoyancy allows for easy functioning. Water therapeutic
- Nutrition Therapy
  - Low inflammatory diet
  - Vitamin D rich foods/supplements

Goals of Care

- Maximize neuromuscular function
- Maintain independence in ADL for as long as possible
- Optimize psychosocial well-being
- Adjust to the illness
- Reduce factors that precipitate exacerbations
  - Illness, trauma, immunizations, stress, extreme temperatures

Nursing Care

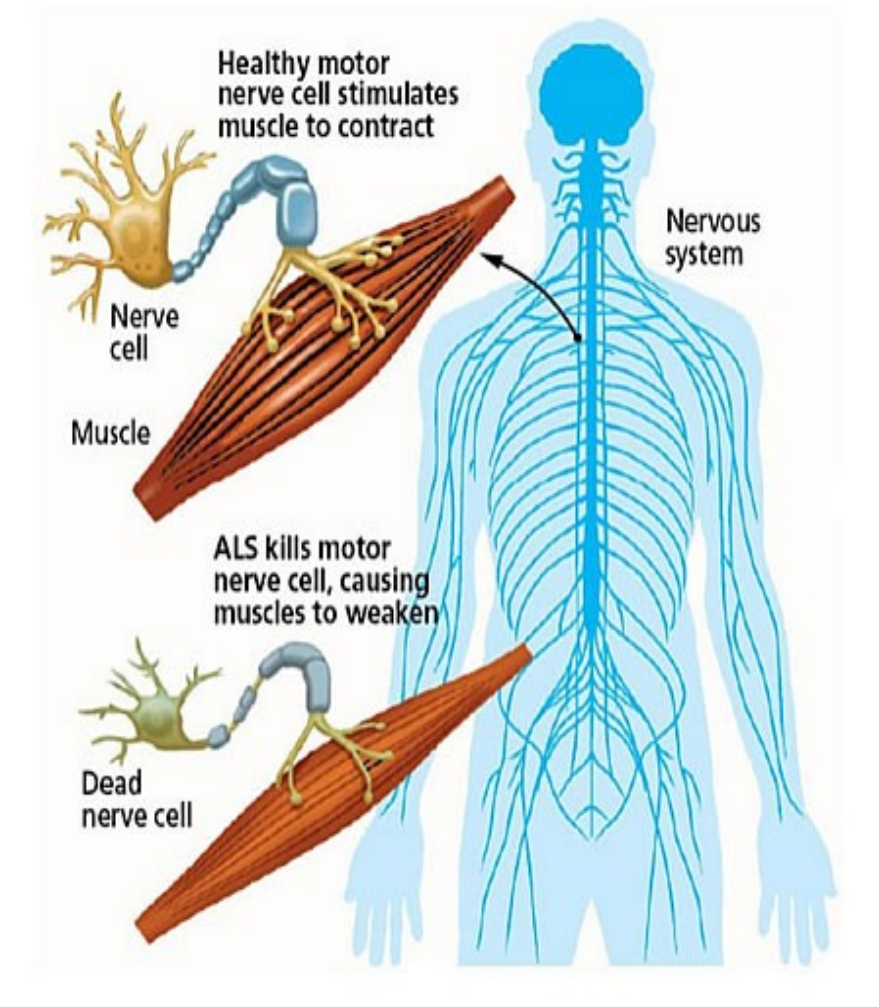
- Bladder Control
  - Medications that help spastic bladder
  - Self-cath may be needed for urinary retention
  - Void schedule
  - Teach S+S of a UTI
- Bowel Control
  - Fiber
  - Fluid
  - Juice, softeners, suppositories
- Coping
  - Support groups, resources

Amyotrophic Lateral Sclerosis

- Rare Progressive Neuromuscular Disorder
- Lou Gehrig's

- Age: 40-70
- Sex: Men 2x more
- Poor prognosis
  - 3-5 years average from onset to death

Definition



- Rapidly progressing
- Degenerative disease
- Upper & lower motor neurons
- Debilitating muscle weakness

Pathophysiology

- Cause is unknown- some theories exist
  - May be related to too much glutamate amino acid in the brain?
- Upper and Lower Motor Neurons are damaged
  - See spasticity (stiff muscles) with upper motor neuron damage and flaccidity with atrophy due to lower motor neuron damage
- ALS does not alter:
  - Sight, hearing, taste, smell, or touch
  - Intellect

Clinical Manifestations

- Early-progressive muscle fatigue, atrophy and weakness
  - Trouble with fine motor skills, tripping, dropping things, stumbling
- As disease progresses
  - Dysphagia and dysarthria
  - Other: pain, sleep disorders, worsening spasticity/hyperreflexia, drooling, constipation, reflux
  - Difficulty breathing
    - Dyspnea
    - Death usually related to respiratory complications and compromised respiratory function.

Diagnosis

- No specific test for ALS diagnosis
- H+P: Signs and symptoms
- EMG to evaluate muscle activity during rest/activity
- Muscle Biopsy
- Pulmonary Function studies

Medical Management

- No cure
- Riluzole (Rilutek)
  - Reduces damage to motor neurons by decreasing release of glutamate in the brain.
  - May slow progression
- Edaravone (Radicava)
  - Relieves effects of oxidative stress that could make ALS progress

Nursing Care

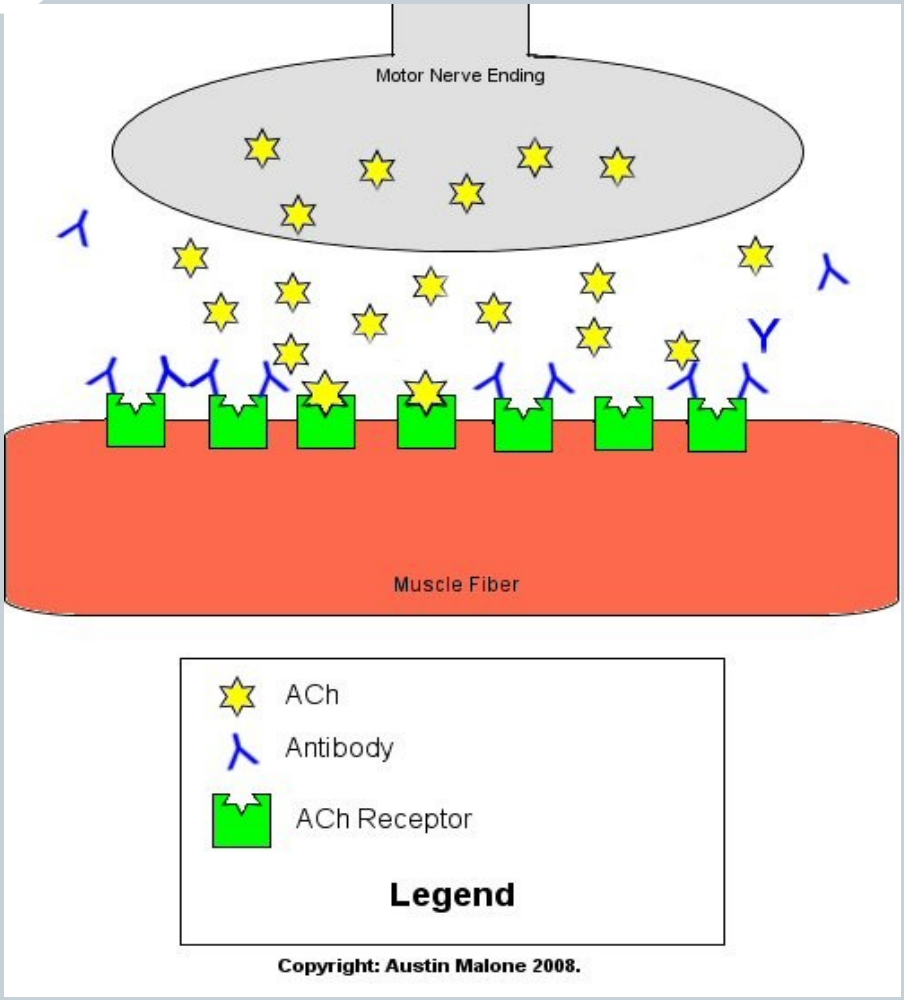
- Most managed at home
  - Support
  - Diversional activities
  - Avoid ill contacts- simple respiratory infection could cause death
  - Home equipment
- Hospitalized for
  - Dehydration, malnutrition, pneumonia, respiratory failure
- Nursing
  - ADL assistance
  - Airway
  - Living Will
- Collaborative Care
  - Nutritional, Occupational, Speech, and Physical

Myasthenia Gravis

- Autoimmune disorder
- Fluctuating muscle weakness
  - Course varies
  - Can have exacerbations
- Ach receptor antibodies
- Prevalence
  - 2x more in women in younger populations

Pathophysiology

- Antibodies are produced against ACH receptors
- This causes not enough ACH reaching the receptors which then causes an inability to stimulate normal muscle contractions
- Cause unknown



Clinical Manifestations

- Motor effects only
- Muscle fatigue
  - Strongest muscles in the AM
  - Most notable weakness in the PM
  - Improvement after rest occurs
  - Weakness to:
    - Muscles that move the eyes and eyelids
    - Muscles that are used to chew, breathe
    - Facial muscles- expression bland
    - Trunk, limbs, neck, hip, shoulder weakness can also occur

Diagnostic Tests

- Antibody Testing
  - Anti- Ach-R
  - Elevated in most patients with MG
- CT to evaluate thymus
  - Is it enlarged? A tumor?
  - May be r/t ACH antibody production
- Electromyography
  - To eval muscle function

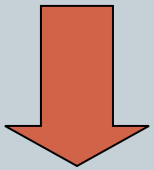
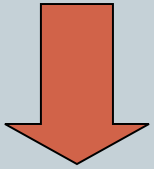
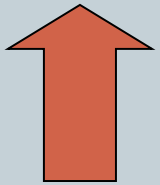
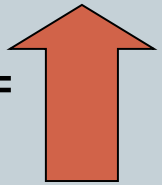
- Tensilon Test
  - Tensilon is anticholinesterase drug that prevents breakdown of the ACH neurotransmitters
  - More ACH is now available
  - Measure muscle strength
  - Give Med
  - Reassess muscle strength
  - Strength improvement = MG Dx

Antidote for Tensilon= Atropine

Treatment

- Anticholinesterase Drugs
  - First line of therapy- pyridostigmine most successful drug
  - Inhibit enzyme that breaks down Ach
  - Tailor dose and maintain schedule
- Corticosteroids and Immunosuppressant
  - Suppress immune response causing Ach-R antibodies
- Thymectomy
  - Thymus gland involved with Ach-R antibody production
- Plasmapheresis
  - Decrease circulating antibodies through plasma exchange
  - Temporary improvement-antibodies will return
- Teach to avoid triggers- illness, stress, trauma

Complications: Myasthenic and Cholinergic Crisis

-  med =  Ach receptor stim = myasthenic crisis
-  med =  Ach receptor stim = cholinergic crisis
- Concerns:
  - Aspiration
  - Respiratory insufficiency and infection

Myasthenic Crisis

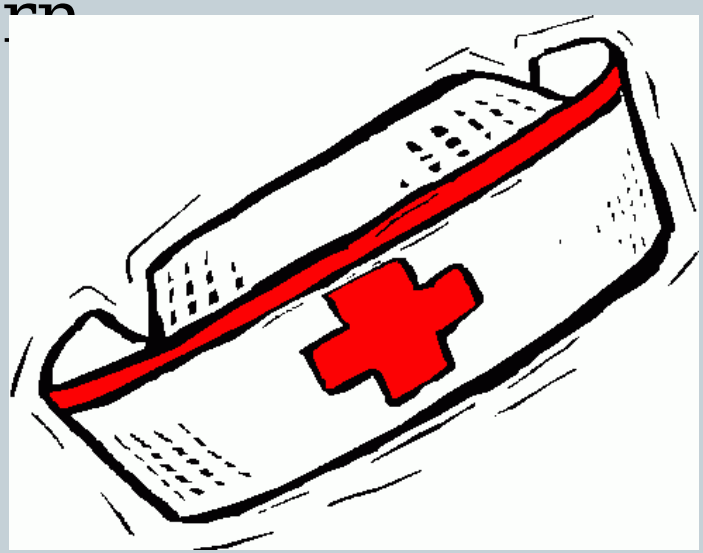
- Underdosage of anticholinesterase drugs
- Exacerbation of MG after a trigger, missed dose, or inadequate dose
- S+S
  - Ptosis, dyspnea, dysarthria, dysphagia
- Treatment
  - IV admin Tensilon (anticholinesterase)
  - Increases the ACH availability
  - Reverses myasthenic crisis

Cholinergic Crisis

- Overdose of anticholinesterase medication
- Too much stimulation of Ach causes muscle to stop responding completely
- S+S
  - Same symptoms as seen with myasthenic crisis PLUS…..
  - Pupil constriction (miosis), diarrhea, N/V, abdominal cramps, increased bronchial secretions, sweating, lacrimation, salivation
  - Weakness within 1 hour after ingestion of too much meds usually

Nursing Diagnoses

- Ineffective Breathing Pattern or Airway Clearance
- Activity Intolerance
- Imbalanced Nutrition
- R/F Injury: Eye
- Deficient Knowledge
  - Patient and Family Teaching



Restless Leg Syndrome

- complaints of paresthesia and motor changes to legs
- Can be unilateral or bilateral
- Symptoms
  - Numbness, tingling
  - Involuntary movements, restless movements
  - Worse at night
- Diagnostics
  - Look at S/S
  - Sleep study to r/o other issues
- Goal-decrease symptoms and improve sleep
- Conservative treatments
  - Avoid ETOH, caffeine, drugs
  - Exercise
- Medications
  - Parkinson's Meds
    - Carbidopa/Levodopa
  - Antiepileptics
    - Gabapentin
  - Opioids