Degenerative Disorders

Parkinson's disease

I. Chronic, progressive neurodegenerative disorder characterized by slowness in the initiation and execution of movement, increased muscle tone, tremor at rest, and gait disturbance.
   A. Risk Factors- affects men more than women & 85% of patients over age 50.

II. Etiology
   A. Cause unknown but thought to be due to: Lack of dopamine in the brain due to degeneration of dopamine producing neurons causing an imbalance of dopamine and acetylcholine.
   B. Parkinsonism set of symptoms: (TRAP)- **t**remor, **r**igidity, **a**kinesia, and **p**ostural instability

III. Pathophysiology
   A. Decreased dopamine levels due to destruction of dopamine producing cells in the substantia nigra located in the mid brain

IV. Clinical Manifestations
   A. Onset is gradual, course-chronic, prolonged course and is initially unilateral
   B. Cardinal signs- TRAP
      1. **T**remors- Resting tremors of tongue, diaphragm, lips, jaw; Pill rolling tremor, handwriting. Tremors can be more noticeable at rest and aggravated by stress or concentration
      2. **R**igidity: increased tone of the limbs and resistance to passive motion
         a. Cogwheeling- during passive motion of a joint.
      3. **A**kinesia- loss of control or absence of control (Bradykinesia) especially which is slowed movement; difficulty initiating movement, affects automatic movements
      4. **P**ostural Instability- lose balance easily

V. Other symptoms and complications
   A. Common symptoms
      1. depression, anxiety, apathy, pain, fatigue, short term memory loss, sweating, flushing, orthostatic hypotension, urinary retention, constipation, sleep disorders
   B. As disease progresses dementia, hallucinations or psychosis can occur

VI. Diagnostics
   A. Clinical findings- history and clinical features. Presence of TRAP
   B. Positive response/improvement with Anti-parkinsonian medications

VII. Medical Management

A. Treatment: control symptoms and maintain independence
    1. Antiparkinsonian drugs to restore balance of dopamine and acetylcholine

B. Carbidopa/Levodopa (Sinemet)
    1. Levodopa is converted to dopamine in the basal ganglia and Carbidopa prevents enzymatic break down of Levodopa allowing for more Levodopa to reach the brain which then converts to dopamine
    2. Side Effects
        a. Dyskinesia
        b. on/off syndrome or “wearing off” phenomenon

C. Dopamine agonists- mimic dopamine by stimulating actual dopamine receptors
    1. less dyskinesia and on/off syndrome
    2. ropinirole (Requip), pramipexole (Mirapex), bromocriptine (Parlodel) may then be used or in combination with Sinemet or alone
    3. orthostatic hypotension- watch out for orthostatic hypotension with bromocriptine especially after the 1st dosage

D. Anticholinergics- benztropine (Cogentin)-blocks acetylcholine creating better balance between acetylcholine and dopamine

E. Antihistamines can be used to help control tremors

F. Antiviral- amantadine (symmetrel) increases dopamine release

VIII. Surgical Management- provides relief but is not curative

A. Deep Brain Stimulation- Electrode in the brain used to stimulate diseased portion. Reduces tremors and uncontrolled movements. Is reversible and adjustable. Preferred method

IX. Collaborative Care

A. Medication Timing- keep consistent levels daily

B. Impaired Physical Mobility: increase strength, coordination, reduce rigidity, prevent contractures with daily exercise, PT, special walking techniques, teach how to initiate movements more easily and safely

C. Risk for Falls

D. Self-Care Deficits

E. Impaired verbal communication

F. Imbalanced nutrition: less than body requirements

G. Risk for Aspiration

H. Ineffective Coping

Huntington's Disease

- Progressive, degenerative brain disorder that results in involuntary movements and mental deterioration.

- Disease Characteristics
  - Men and Women equally affected, usual onset age 30-50
  - Course is chronic and progressive- no cure

- Genetic disorder- autosomal dominant disorder
  - Abnormal gene identified
    - With each pregnancy an affected parent has a 50% of passing gene to offspring
- Pathophysiology
  - HD abnormal gene causes:
    - A deficiency in the neurotransmitters GABA and Ach
    - Excess dopamine availability in HD- Leads to symptoms that are opposite of parkinsons

- Clinical Manifestations
  - Most prominent clinical features are: chorea and mental cognitive/psych decline
  - Chorea:
    - Jerky, brisk, and purposeless movements, Involuntary abnormal movements
    - As disease progresses writhing and twisting of the face, limbs and body can occur.
  - Facial Movements: tics/grimacing
  - Speech: slurred, hesitant, explosive
  - Chewing/Swallowing: difficulty
  - Gait: disorganized
  - Bladder/Bowel: control can be lost
  - Cognition, personality, and emotions are all affected.
    - Poor judgment, memory loss, decreased attention span, delusions, anger, dementia
- Diagnosis- presentation of symptoms, family history, genetic testing
- Management
  - No cure
  - Medications-symptom control
    - Abnormal movements
      - Tetrabenazine (Xenazine) or Deutetrabenazine (Austedo)
      - Neuroleptics: haloperidol (Haldol), risperidone (Risperdal)
    - Cognitive and Psychiatric symptoms
      - SSRI's: sertraline (Zoloft) and paroxetine (Paxil)

Nursing Care

- Safety
  - Padding-
  - Ambulation assistance
- Nutrition
  - Need higher calorie intake per day

- bite sized food, blended foods- r/f aspiration

❖ Altered mentation and social interaction
- orient patient upon awakening
- medical identification bracelet
- recruit and train volunteers

❖ Collaborative Care
- Medical, nursing, psychological, social, occupational, speech, rehabilitation services
- Home care assistance, day-care centers, respite care and eventually long-term care

Multiple Sclerosis

❖ Chronic, progressive, degenerative, disorder of the CNS with demyelination of nerve fibers
❖ Onset- usually ages 20-50 but can happen to younger or older, Women-affected more than men
❖ Cause is unknown- research for viral, immune, genetic involvement
❖ Some debate concerning precipitating factors of the disease
- Infection, Physical injury, Emotional stress, Pregnancy

❖ Pathophysiology
- Autoimmune response leads to chronic inflammation, myelin sheath damage by demyelination, and gliosis (scar formation) which disrupts nerve transmission in the CNS. Brain and/or Spinal Cord can be affected.
- Demyelination causes nerve transmissions to be slowed.
- Scar tissue causes plaques which interrupt nerve transmission
- Permanent loss of function could occur if axons become continually damaged

❖ Onset
- Vague, intermittent, symptoms vary depending on the location of damage and plaques

❖ Manifestations:
- Fatigue-common, severe, disabling
- Impaired Movement
  - Limbs-can feel weak or heavy, early symptom. Numbness & tingling may be present
  - As disease progresses- Stiffness, Gait problems-poor balance
- Optic Nerve: Visual disturbances- distortion of red/green colors, blindness in 1 eye
- Acoustic Nerve: Auditory Disturbances: Tinnitus or loss of hearing
- Dysarthria, Dysphagia
- Bowel/bladder function loss possible
- Sensory Disturbance- Paresthesia, pain
- Cognitive and emotional problems: decreased ST memory, concentration, and emotional stability

❖ Classifications are characterized by flares (exacerbations), remission, and disease progression
- Relapsing Remitting
  - Acute, unpredictable attacks with distinct remissions. Symptoms then improvements
- Secondary Progressive
  - Begins as relapsing remitting then definite progression with or without relapses.
- Primary Progressive

- Steadily worsening status with no attacks
- Progressive relapsing
  - Continued progression from onset with clear acute relapses, with or without full recovery.

❖ *Nice to know fact: Pregnancy*
- MS has no real negative effect on pregnancy, labor, delivery, or lactation. Some women report improvement of symptoms during pregnancy likely due to the hormone changes

❖ Diagnostics
- No definitive tests, using mostly history and clinical S&S
- Lumbar Puncture: CSF changes or IgG bands (oligoclonal bands) indicating autoimmune response
- MRI- main diagnostic tool to look for white matter lesions in brain/spinal cord (MS plaques)

❖ Drug Therapy- Early, continuous therapy most recommended
- Medications may reduce # & size of plaques, frequency, and duration of relapses
- Interferons (Avonex, Betaseron, Rebif, Copaxone)
  - Action-regulates immune system, prevents T cells from causing inflammation
  - Administration-all injections and should rotate sites between injections
  - SE: flu like symptoms, sun sensitivity
- Others: Fingolimod (Gilenya), Teriflunomide (Aubagio), dimethyl fumarate (Tecfidera)- immunomodulators, immunosuppressants and anti-inflammatory PO meds. Other meds via IV infusions are also available.

❖ Acute exacerbation or relapse treatment
  - decrease edema & inflammation at site of demyelination- PO prednisone
  - Methylprednisolone (solu-Medrol) IV course followed by PO taper

❖ Symptomatic Treatment
  - Urinary Retention- Cholinergics
  - Spastic Bladder- Anticholinergics (Ditropan)
  - Fatigue- Methylphenidate (Ritalin), modafinil (Provigil)
  - Spasticity- Muscle relaxants (diazepam)
  - Bradykinesia- Dalfampridine (Ampyra)

❖ Other therapies
- Physical therapy/ speech therapy- improves spasticity and coordination
- Water exercise-buoyancy to allow for more functioning
- Nutritional Therapy- anti-inflammatory diet

❖ Goals of Care: maintain function, independence, well-being, reduce exacerbations

❖ Nursing Implications
- Education
- Bladder & Bowel control
- Coping

Amytrophic Lateral Sclerosis

I. Rapidly progressive, neurologic disorder with degeneration of upper and lower motor neurons that leads to progressive and eventually debilitating muscle weakness

II. Pathophysiology

A. ALS effects both upper and lower motor neurons

1. Upper motor neurons- degenerate and stop communicating which leads to spasticity of the muscles (stiffness)
2. Lower motor neurons- can't produce or transport impulses from nerves which leads to flaccidity and atrophy of the muscles

B. ALS does not affect: Intellect, cognition

III. Clinical Manifestations

A. Manifestation progression can include weakness spreading to multiple muscles.

B. Early symptoms: progressive muscle weakness and atrophy

C. As advances: dysphagia, dysarthria, pain, sleep disorders, spasticity, drooling, constipation, reflux, respiratory difficulties (many will die due to resp complications)

IV. Diagnosis

A. History and Exam: look at the signs/symptoms.

B. EMG to look at muscle activity or a Muscle biopsy

C. Abnormal pulmonary function studies- will show weakened respiratory muscles

V. Medical Management

A. No cure but there is symptomatic treatment medication:

1. Riluzole (Rilutek)- glutamate antagonist that may slow progression of ALS
2. Edaravone (Radicava)- relieves oxidative stress

VI. Nursing Care

A. At home- emotional/psychological support- patient cognitively ok, but body wasting away

B. Acute care setting- manage dehydration, malnutrition, pneumonia, and respiratory failure

C. Nursing care in the acute care setting: ADLs with assist, Airway maintenance, DNR/DNI?

D. Collaborative Care

1. Nutritional Consult
2. Occupational Therapy
3. Speech Therapy
4. Physical Therapy

Myasthenia Gravis

I. Autoimmune disease characterized by muscle fatigue and weakness from inadequate Ach receptor stimulation due to ACh receptor antibodies that attack acetylcholine receptors.

A. Course: Slow and chronic with periods of:

1. Remission, exacerbations, stabilization

II. Physiology of Normal muscle contraction:

1. Nerve impulse arrives at a nerve ending, releases chemical called acetylcholine (Ach)
2. Ach travels across synaptic cleft and attaches at receptor sites, becoming activated by Ach and then causing muscle contraction once enough sites have been activated.

III. Pathophysiology of MG

A. Antibodies are produced against ACH receptor sites. This results in a fewer number of acetylcholine receptor sites so the acetylcholine molecules can't attach to these receptor sites and stimulate normal muscle contraction. Cause-unknown

IV. Clinical Manifestations

A. Primary feature-fluctuating weakness of skeletal muscles

B. Muscles affected: Those used to move the eyes, eyelids, chew, swallow, speak, breathe

C. Can also have trunk, shoulder, limb, neck weakness

D. Exacerbations can be precipitated by: emotional stress, trauma, pregnancy and menses temp changes, secondary illness, hypokalemia, some medications

V. Diagnostics

A. Lab Test: Anti-Ach R

B. Thymus abnormalities- enlarged thymus gland or tumor

C. EMG – progressive stimulation leads to decreased muscle function

D. Tensilon Test

1. IV admin of Tensilon – prevents breakdown of Ach
2. increases Ach availability
    a. assess muscle strength, give med, re-assess
    b. Atropine= antidote in case of adverse reaction cardiac arrythmias

VI. Treatment- No cure

A. Anticholinesterase drugs-

1. pyridostigmine (Mestinon) anticholinesterase drug

2. Action: prevents destruction of Ach by inhibiting the enzyme acetylcholinesterase, thus enhancing the availability of Ach and impulse transmission across the muscle junction
3. Maintain schedule of dosages

B. Corticosteroids and Immunosuppressants
   1. Prednisone which suppresses immune response that causes antibody production

C. Thymectomy- removal of the thymus gland can be shown to help some

D. Plasmapheresis – plasma exchange to remove/decrease receptor antibodies

E. IV Immunoglobulin G antibody can give short term improvement

F. Avoid triggers that can cause exacerbations: stress, trauma, illness, hypokalemia

VII. Complications: Myasthenic Crisis and Cholinergic Crisis

A. myasthenic crisis: insufficient medication causes not enough ACh

B. cholinergic crisis: excessive medication causes too much ACh

C. Myasthenic Crisis
   1. exacerbation when there was a trigger, failure to take med or inadequate/late dose
   2. Ptosis, Dyspnea, Dysarthria, Dysphagia- can result in respiratory insufficiency
   3. Treated - Anticholinesterase Tensilon for quick action to increase the Ach availability

D. Cholinergic Crisis
   1. Exacerbation when there is overdose, too much medication.
   2. Can get muscle fasiculations, sweating, excessive salivation, constricted pupils along with some muscle weakness
   3. Treated with IV atropine

VIII. Nursing Care

A. Ineffective Breathing Pattern or Ineffective Airway Clearance

B. Activity Intolerance

C. Imbalanced Nutrition

D. Risk for Injury: Eye Care

E. Deficient Knowledge: Patient and Family Teaching: Airway maintenance

Restless Leg Syndrome

1. Unilateral or bilateral lower extremity condition in which patients experience parasthesias and motor changes to one or both legs
2. Statistics: Risk factors- women, elderly, family history
3. Pathophysiology

a. Alterations in iron metabolism and the dopamine neurotransmitter system is thought to be the patho. There is some dysfunction with neurotransmitter dopamine, which controls movements.

4. Types
    a. Primary-cause is unknown
    b. Secondary- metabolic abnormalities from iron deficiency, renal failure, and polyneuropathy
5. Signs and Symptoms
    a. Vary in intensity and frequency
    b. Sensory Symptoms
        i. Paresthesia-described as bugs creeping up the legs
        ii. Discomfort progresses to pain
        iii. Timing-during inactivity-especially in evening and while sleeping
        iv. Relief with movement
    c. Motor Symptoms
        i. Voluntary restlessness and involuntary movements
6. Diagnostics
    a. History and S/S
    b. Criteria:
        i. Urge to move legs
        ii. Unpleasant leg sensations with rest or inactivity
        iii. Relief of urge to move and sensory Sx with movement
        iv. Symptoms worse in evening or at night
        v. No other condition contributing to these symptoms
    c. Sleep study to rule out other disorders
    d. Blood Tests to rule out other disorders causing secondary RLS
7. Nursing Care
    a. Conservative Treatments-establishing regular sleep pattern, exercises, avoiding ETOH and caffeine or any drugs that can impair sleep
    b. Medications
        i. Parkinson's Disease Medications
            1. Sinemet (carbidopa/levodopa)
            2. pramipexole (mirapex)
            3. Ropinorole (Requip)
        ii. Antiepileptics
            1. gabapentin
        iii. Opioids-for those who fail other treatments