**Chapter 26: Degenerative Diseases of the Central Nervous System**

**Degenerative Neurologic Diseases:** cause progressive pathologic changes in the CNS resulting in loss of function in one or more areas like sensation, motor control, and cognition.

* Multiple Sclerosis (MS), Parkinson’s Disease (PD), Amyotrophic Lateral Sclerosis (ALS), and Alzheimer’s Disease (AD) are examples

Client center approach is recommended & occupational therapy practitioners must consider:

* Probable decline in function and occupation requires planning ahead
* Inconsistent performance in areas of occupation in day can be caused by fatigue or cognition changes
* Impact of family and care givers
* Limited medical interventions for some diseases
* Psychological issues related to coping with progressive disorder
* With a client centered approach, the patients become more empowered & aware of all factors of treatment
* General treatment goals for progressive neurologic disorders are: maximize client’s ability to engage in meaningful occupations, prevent secondary complications, maximize quality of life, teach strategies to self-manage the effects of disease, and educate caregivers of safe and effective ways to provide assistance
* Home evaluations include looking at hazardous conditions, potential assistive devices if necessary, and more home modifications if needed.

**Multiple Sclerosis:** an autoimmune disease that affects the CNS (brain, spinal cord, and optic nerves) and demyelination (body attacks myelin) and plaque formation impede the transmission of nerve impulse to and from the brain appropriately.

* Epidemiology: typical age is 20-50 but can vary in all ages
  + More common in women and not hereditary but probable genetic impact
  + 1/750 chance developing MS in gen. population, rises to 1/40 w/family history
* 4 Disease Courses: Relapsing/remitting which is acute attacks w/full or partial recovery. Between attack the disease does not progress
  + Secondary Progressive: clients start in relapse course but then followed by a progression at a variable rate. 50% people develop secondary progressive Within 10 yrs and 90% within 25 yrs
  + Primary progressive: progressive disability without remission from onset of disease (10% of people with MS)
  + Progressive Relapsing: progressive from onset w/clear acute relapses (5% ppl)
* Impact on client factors: depends on what area of CNS was affected and how severely
  + Usually symptoms include fatigue, visual & cognitive disturbances, mental disturbances, sensory changes, loss of postural control, dizziness, tremor, dysphagia, weakness, and bowel/bladder dysfunction
* Medical Treatment: main focus is to alleviate patient symptoms
  + High dose of corticosteroids are given for acute exacerbations
  + Immuntomodulators are used to reduce number of relapses
  + May require medical management for catheterization, surgery, or feeding tube
* Occupational Therapy Management: usually changes demand regular adjustments
  + Precautions: patient may be affected by stress, heat, pain, fatigue, & more
    - Avoid over fatigue
    - Be aware of room temp (cooler is better)
    - Use heat modalities with caution
    - Be aware of fluctuations with level of independence
    - Guard against soft tissue injury secondary to sensory loss (sharps)
    - Awareness of all impairments that can be affected
* Evaluation: eval process sets the tone and prepares all involved for treatment
  + Eval includes assessments of client factors, areas of occupation, and quality of life
    - Client factors: Strength, ROM, muscle tone, sensation, coordination, endurance, balance, vision, and cognitive functions
    - Areas of occupation: mainly ADL, IADL, work, and play/leisure
* Interventions: Improving participation via fatigue management is vital because 75% to 95% of MS patients experience fatigue and 50% to 60% fatigue is their chief complaint
  + Primary Fatigue due to the disease process (cortical damage, biological factors, ect)
  + Secondary fatigue may be due to conditioning, respiratory muscle weakness, & pain
  + Treatment is two-step process: first to eliminate any secondary causes of fatigue (treatment for coexisting diseases like depression, improving sleep patterns, and energy conservation) and second step is managing primary fatigue.
    - Energy conservation techniques are pacing, use of electronic aids, flexible home and work schedules, home/work modifications, heat control, improved trunk control, power mobility aids, and more
  + Interventions related to ataxia and tremors: orthotics/splinting, using the environment for stability, adaptive devices, control fatigue, decrease effort, provide exercise for proximal stability, adapt activities to eliminate the need to reach
  + Cognitive strategies: use of memory aids, avoid multitasking, keep organized avoiding cutter, solve problem aloud, & allow extra time for task completion
  + Improving participation via managing sensory deficits include a variety of compensatory strategies, avoiding harmful situations, and ADL training
    - Factors to consider with vision loss are: make interventions large print, color contrast, decrease background clutter, and increase illumination on task
  + Strengthening and endurance training include resistive training, slings or devices to assist with muscle weakness, or ROM/MMT exercises
  + Contracture prevention: usually patients with severe weakness that prevents full AROM and with spasticity have risk of developing contractures
    - AROM and PROM, splints, and inhabitation techniques to prevent spasticity
  + ADL’s: help with affective of equipment and education for different techniques
  + Communication: OT works with ST to devise method to improve comm. Skills
  + Seating and wheeled mobility considerations are overall endurance, trunk control, LE strength, UE strength, and disease prognosis
  + Leisure and work skills: plan for social outings, worksite modification, and adaptive equipment in home and at work
  + Psychosocial issues: characteristics of those who cope well with MS include support, connectedness, sense of humor, spirituality, and openness

**Parkinson’s Disease**: slow, chronic, progressive disease of nervous system and is characterized by resting tremor, rigidity in skeletal muscle, bradykinesia, and postural instability.

* Pathology is degeneration in dopaminergic pathways in basal ganglia, and substantia nigra
  + When signs and symptoms are noticed 80% of neurons are already deteriorated
* Epidemiology: average age of diagnoses is 60, 15% of people with PD is before 40
* Cause: Not clear but researchers think its genetic and environmental factors (15%-25% of people with PD have a relative with the disease)
* Impact on Client Factors: Tremors are involuntary movements of hand, rigidity is resistance to movement, and bradykinesia s slowness of movement all cause issues w/life.
  + Gait and balance are affected, stooped-forward posture, shuffled walk, and no stop.
  + With progression comes problems with oral musculature, drooling, dysphagia, ect.
* Medical Management: Levodopa is common medication for PD or medications involving dopamine.
* Occupational Therapy Management: precautions involve safety with ambulation and transfers. Immobility should be addressed & pt should be encouraged to stand or reposition themselves.
* Evaluation: Functional performance of ADL, IADL, flexibility, strength, movement, standing, sitting, balance, coordination, and cognition is necessary
* Interventions: Participation in areas of occupation include bed mobility skills, transfer training, and wheelchair training
  + Motor skills/prevention of deformities: AROM and stretching focusing on muscles like hip flexors, knee flexors, calf muscle, pec major and minor, and more
* Communication: OTA’s help with providing breathing and postural exercises
* Psychosocial issues: group counseling and emotional and social outlets are recommended
* Advanced Parkinsonism is when the patients have severe deficits in communication, mobility, swallowing, and cognition. Symptoms management is necessary

**Amyotrophic Lateral Sclerosis:** ALS is a progressive disease characterized by the generation of motor neurons in the anterior horn cells of spinal cord, brainstem, and cortical spinal tracts.

* Muscles weaken & atrophy. Person becomes unable to breathe independently
* Some individuals may experience problems with memory and executive functions
* Epidemiology: men are 20% more likely. Usual age is 40-70. Some cases are 20-30’s.
* Cause: unknown cause but majority of cases (90%-95%) are random whereas 5%-10% of cases are considered familial. 20% of familial cases are a gene defect
* Impact on Client factors: symptoms include difficulty walking, difficulty picking up objects, performing fine motor skills. The number and side of limbs affected varies from person to person.
  + Weakness and stiffness in intrinsic hand muscles. Hyperactive reflexes & twitching
  + Weakness, overtime, spreads to all muscles & become flaccid w/severe disability
* Medical Management: No cure for ALS just treatment to manage symptoms
* Occupational Therapy management: enable patient to adapt and maintain max level of functioning and assist care providers with necessary skills to assist ALS patient
* Interventions: Improving participation in areas of occupation- interventions will vary depending on the stage of the disease.
  + Early symptoms include loss of fine motor coordination and hand weakness
    - Assistive devices for writing and eating are useful
    - OTA should focus on using ambulation devices in functional situations
  + As disease progress UE weakness continues and further adaptation are necessary
    - Home modifications like ramps and bathroom modifications to maintain independence, increase safety, and facilitate caregiver assistance
  + Motor skills and prevention of deformities: water aerobics, resistive training, and gently PROM are interventions for the occupational therapy practitioner
  + Communication: OT and speech work together and OT may provide communication assistive devices or position patient to use switches or call bells
  + Assistive technology: computers and environmental control units can be used
  + Mobility and positioning: Proper head positioning is important and lateral supports of wheelchair are necessary
  + Psychosocial issues: depression and anxiety disorders are common

**Alzheimer’s Disease:** is not a disease itself but refers to a set of symptoms. Dementia refers to loss of mental function in two or more areas like language, memory, visual abilities, spatial abilities, or judgement. AD is progressive, slow deterioration of brain tissue.

* Epidemiology: 1 in 10 has a family member with AD. 1 in 10 people older than 65 are affected. Rarely, inherited disease can affect people in their 30s and 40s. Person will live an average of 8 years after diagnoses but can live up to 20 years
* Cause: Increasing age (likelihood doubles every 5 years after age 65), family history of disease, genetics, and specific genes linked to AD.
* Signs and Symptoms: Memory loss, difficulty performing familiar tasks, problems with language, disorientation, impaired judgement, decreased abstract thinking, and more.
* Medical Managements: No cure will slow progression of disease, so treatment is symptomatic. Medications include Celexa, Prozac, Zoloft, and more.
* Occupational Therapy management: Evaluation of cognition should be done through formalized assessment tools. Motor functioning, self-care evaluation, and ability to perform specific skills should be evaluated.
* Interventions: ADL’s can be managed with the help of an OTA or OT. Task segmentation is breaking activities up into different steps to help with sequencing.
  + Patient may need verbal prompting or cue cards to remember hygiene
  + Patient may need help with feeding techniques and using adaptive equipment
* Environmental design: the OTA can help the caregiver structure the environment to help maximize the patients functioning
  + Eliminate clutter, signs to identify rooms, and visual cues
* Day care and group activities: programs offer care givers a respite from daily care of AD
* Reality Orientation: patients with AD become less oriented as disease progresses. Formal daily orientation groups review the patient’s name, date, weather, and location.
* Exercise Programs: it’s important to maintain strength, coordination, and ROM whether it’s with a group or individually
* Psychosocial issues: patients with AD may demonstrate variety of behaviors like agitation, aggression, depression, inappropriate sexual behaviors, paranoia, & hallucinations.