CHAPTER 27

Degenerative Diseases of the Central Nervous System*

Glen Gillen

Key Terms
- Degenerative neurological diseases
- Progressive neurological diseases
- Multiple sclerosis (MS)
- Demyelination
- Exacerbations
- Remission
- Parkinson’s disease (PD)
- Bradykinesia
- Rigidity
- Dementia
- Amyotrophic lateral sclerosis (ALS)
- Alzheimer’s disease (AD)
- Task segmentation

Chapter Objectives
After studying this chapter, the student or practitioner will be able to do the following:

1. Describe four degenerative diseases.
2. List the signs and symptoms of these diseases.
3. Understand the focus of occupational therapy related to the treatment of degenerative diseases.
4. Describe the precautions that must be observed in the treatment of these diseases.
5. Recognize and describe the occupational therapy interventions used to treat clients with degenerative diseases across the various stages of the disease.

Introduction and General Concerns

Degenerative neurological diseases cause progressive pathological changes in the central nervous system (CNS). These changes result in loss of functioning in one or more of the following areas: sensation, motor action and control, and cognition. The diseases addressed in this chapter are multiple sclerosis (MS), Parkinson’s disease (PD), amyotrophic lateral sclerosis (ALS), and Alzheimer’s disease (AD). Although no cures exist for these diseases, their debilitating effects can be partially ameliorated with effective medical and rehabilitative management. The role of occupational therapy (OT), in the treatment of degenerative neurological diseases is to assist in managing symptoms, maintaining function, and optimizing quality of life. Because of the nature of the diseases included in this chapter, compensation/adaptation is a common method used to maximize function.

Because of the progressive and sometimes unpredictable course of the diseases that will be discussed below, the occupational therapy assistant (OTA) in conjunction with the occupational therapist must consider the following factors that influence management of this population:

- A probable decline in performance in areas of occupation requires planning ahead. In terms of assessment, periodic reevaluations are necessary to monitor performance. Interventions such as prescription of adaptive devices and/or durable medical equipment should take into account the length of time the equipment will be usable.
- Inconsistent performance in areas of occupation throughout the day due to fatigue issues or cognitive changes, as in delirium in the evening or night (“sundowning”).
- Impact of the diagnosis on the family related to role changes, ability to provide care, or potential loss of a loved one.
- Limited medical interventions for some diseases.
- Psychological issues related to coping with a progressive disease including coping with end-of-life issues, fear related to probable future decline of function, anxiety, depression, etc.

Working with those presenting with progressive neurological diseases is complex and challenging. The focus of interventions varies based on the stage of the disease as well as the context in which the OT practitioner is interacting with the client (e.g., acute care vs. long-term care vs. home).

*Phyllis Ber contributed large portions of this chapter to the first edition of this book.
A client-centered approach is recommended for these reasons: “Client-centered practice is an approach to providing occupational therapy which embraces a philosophy of respect for, and partnership with, people receiving services. Client-centered practice recognizes the autonomy of individuals, the need for client choice in making decisions about occupational needs, the strengths clients bring to a therapy encounter, the benefits of client-therapist partnership, and the need to ensure that services are accessible and fit the context in which a client lives.”

Law et al. and Pollack suggest that the therapy practitioners who implement this approach to evaluation consider advice listed in Box 27-1.

With these strategies the OT process becomes more focused and defined; patients become immediately empowered; the goals of therapy are understood and agreed upon; and an individually tailored treatment plan may be established. The Canadian Occupational Performance Measure is a standardized tool that uses a client-centered approach to allow the recipient of treatment to identify performance areas of difficulty, rate the importance of each area, and rate his or her satisfaction with current performance. It is a particularly useful tool for this population because of the multiple and extensive problems experienced in performance of areas of occupation. In addition, it can be used with family members and caregivers to help prioritize interventions.

In general, after the evaluation is completed, the occupational therapist should have a thorough understanding of the client’s strengths and weaknesses. In collaboration with the client, the therapist establishes realistic goals with the overall purpose of increasing or maintaining the patient’s present functional status. Long- and short-term goals are addressed. General treatment goals for those living with progressive neurological diseases include the following:

- Maximize the client’s ability to engage in meaningful occupations despite disease progression.
- Prevent secondary complications such as decubitus ulcers, contractures, pain, or injury.
- Maximize quality of life.
- Teach clients strategies to self-manage the effects of the disease process (e.g., medication management, dealing with fatigue, coping strategies, etc.).
- Teach caregivers safe and effective ways to provide assistance to the client while not undermining the client’s abilities.

The success of the OT program depends somewhat on the patient’s perceived sense of accomplishment and a positive attitude. The progressive nature of these diseases requires that treatment goals be established in small enough increments so that the patient is assured of some measure of progress.

Because of the progressive and chronic nature of the diseases included in this chapter, clients in the latter stages of the disease process may require substantial care beyond what is possible within the client’s social support system. Placement in a long-term care facility is indicated when the client and family can no longer cope with advancing symptoms in the home. Nursing homes offer (1) comprehensive rehabilitation programs; (2) contracture prevention programs (via positioning, splinting, and range of motion [ROM]); (3) decubitus ulcer prevention and treatment; (4) adjunctive medical interventions such as tube feedings; (5) bowel/bladder management strategies; (6) suctioning of respiratory secretions; (7) palliative care; (8) counseling programs; and (9) the 24-hour supervision necessary for patient safety. In addition, the nursing home will have specific durable medical equipment that may be required to assist those who require substantial physical assistance, such as mechanical lifts, electric hospital beds, pressure-relieving mattresses, and so on.

All clients with a progressive disease can benefit from periodic home evaluations. The foci of the home evaluation are described in the following discussion.

Any hazardous conditions that could trigger a fall should be eliminated. Special care should be taken to remove all throw or scatter rugs. Bathroom mats should be removed. Doorway thresholds should be even with the floors. On the outside of the home, gravel or cobblestone walkways represent a safety hazard.

Assist in the evaluation of necessary assistive devices or durable medical equipment. In the bathroom, grab bars, a raised toilet seat with a safety frame, and a shower chair or tub transfer bench are recommended. In the bedroom, a sturdy chair with armrests should be used while dressing. A raised bed with a firm mattress and a trapeze over the bed also help with bed mobility. A bedside commode or urinal should be considered when the patient makes frequent nighttime trips to the bathroom.

Consider simple home modifications. In the living room, the patient should be advised not to sit in deep, low chairs. The preferred chair has firm cushions, a straight back, and padded armrests. The height and depth of the chair should allow the patient to maintain feet on the floor with knees flexed at 90°. For those patients who cannot get out of the chair independently, automatic lift chairs are available. In the kitchen, commonly used items should be placed so that excessive bending and reaching are not required. The patient’s
Multiple Sclerosis

MS is thought to be an autoimmune disease that affects the CNS, including the brain, spinal cord, and optic nerves. Myelin, the tissue surrounding and protecting the nerve fibers of the CNS, helps nerve fibers conduct electrical impulses. In MS, myelin is lost in multiple areas (demyelination), leaving scar tissue known as sclerosis. These damaged areas are also known as plaques or lesions. The two processes of demyelination and plaque formation impede the transmission of nerve impulses to and from the brain. Depending on the area(s) that develop plaques, various signs and symptoms occur.

Epidemiology

Facts and figures are maintained related to MS. Key facts to consider include the following:5,22,31

- Typical age of people diagnosed with MS ranges from 20-50.
- More common in women (2-3:1).
- Not hereditary but has a probable genetic aspect.
- 1/1000 risk of developing MS in the general population.
- More common in Caucasians than Hispanics or African-Americans.
- More prevalent in higher latitudes.
- Approximately 400,000 are living with MS in the United States, with 1 to 2 million patients worldwide.

Disease Course

MS may follow various courses related to progression. The following four courses have been identified:5,22,31

1. Relapsing/Remitting: Characterized by acute attacks with full or partial recovery. Between attacks the disease does not progress. Eighty-five percent of those initially diagnosed with MS present with this course.
2. Secondary Progressive: Initially, clients follow a relapsing/remitting course, which is followed by progression at a variable rate. Of those initially diagnosed with a relapsing/remitting course, 50% develop secondary progressive within 10 years and 90% within 25 years.
3. Primary Progressive: Progressive disability without remission from the onset of the disease. Relates to 10% of those diagnosed with MS.
4. Progressive Relapsing: Progressive from the onset with clear acute relapses. Relates to 5% of those diagnosed.

Impact on Client Factors

The signs and symptoms of MS vary depending on the areas of the CNS that have been affected by demyelination and plaque formation. Each patient will present with a unique set of clinical manifestations depending on the locations of the lesions and the stage of the disease process. Symptoms directly caused by demyelination include fatigue, visual disturbances, cognitive disturbances1 (slowed processing, memory loss, decreased sustained attention, poor abstract reasoning, impaired problem solving, decreased verbal fluency, impairment of executive functions, impaired visuospatial skills), affective disturbances2,42,43 (depression, bipolar disorders, lability, euphoria, antisocial behavior), sensory changes (numbness, tingling, pain), loss of postural control, tremor, dysphagia, heat intolerance, spasticity, weakness, and bowel/bladder dysfunction.

Medical Management

The treatment of MS focuses primarily on alleviating the patient’s symptoms5,22,31 through, for example, use of antidepressants or antispasmodics. High-dose corticosteroids are often given for acute exacerbations. Immunomodulators are used to reduce the number of relapses and limit the development of new lesions. Typical drugs include Avonex®, Betaserone®, Copaxone®, and Rebif®. The OTA can assist the physician by reporting changes in the patient’s behavior and physical status.

During the chronic stages of MS, medical management of the disease may include catheterization for urinary dysfunction, tube feeding for swallowing disorders, and nerve blocks or surgical release of tendons for treatment of severe contractures.

Occupational Therapy Management

The patient with MS offers a challenge to the OT practitioner. The disease follows a variable and unpredictable course that changes the patient’s functional ability from morning to night, from day to day, and over the course of the disease. These changes demand regular adjustments to the patient’s therapy program.

Precautions

The functional status of an MS patient may be affected by a variety of factors such as stress, heat, pain, fatigue, and exacerbations.44 Considerations include the following:

- Avoiding over-fatigue
- Awareness of room temperature; cooler is generally better
- Using heat modalities with caution because of both sensory loss and possible heat intolerance
- Being aware of fluctuations in level of independence. For example, a client who uses a sliding board to transfer may require only supervision in the morning but may require physical assist by afternoon

walker can be fitted with a bicycle basket to make it easier to carry objects.

More substantial home modifications may be necessary. Examples of substantial modifications include the installation of an elevator, stair-glide, stall shower, or ramp.

Accessibility issues may need to be addressed for those clients using a device for gait or wheeled mobility.
• Guarding against soft tissue injury secondary to sensory loss (e.g., sharp objects, hot water)
• Monitoring for safety issues secondary to loss of postural control and/or cognitive impairment

**Evaluation**
The initial evaluation process for the MS patient sets the tone and prepares both the patient and the OT practitioner for the treatment sessions to follow. Principles of a client-centered assessment are necessary because of the multiple areas of occupation that may be adversely affected. It is important that client and caregivers determine which occupations should be addressed first. In addition, the evaluation reveals the areas amenable to remediation (such as deconditioning) and those for which compensatory techniques should be taught (such as long-standing memory loss). Because the patient may have cognitive deficits or may be experiencing anxiety or stress, the practitioner must be careful to explain the reason for the evaluation and to describe what will occur during the evaluation. Rest periods may be required if the patient becomes fatigued. If necessary, the evaluation may be broken up into two or more sessions. At the conclusion of the evaluation process, the therapist and the patient should agree on the goals for the OT program. The practitioner should emphasize that the OT program will be directed toward the improvement and/or maintenance of meaningful occupations—that is, those occupations that the client wants to do, needs to do, or has to do to return home.

The evaluation includes assessment of client factors, areas of occupation, and quality of life issues. Client factors include strength, muscle tone, sensation, coordination, joint ROM, endurance, balance, and cognitive functions.

Performance in areas of occupation including basic and instrumental activities of daily living (ADL, IADL), work, and play/leisure must be objectively evaluated. The ADL evaluation may be administered by an experienced OTA who has demonstrated competency in these evaluation techniques. The coordination and interpretation of the evaluation results is the role of the occupational therapist.

**Interventions**
The treatment methods selected for MS clients are determined by individual goals for the client and are guided by the client’s clinical presentation.

**Improving Participation via Fatigue Management**
Fatigue is reported as a limiting factor for 40% to 70% of those living with MS. Multiple factors may cause a loss a function secondary to fatigue, including co-morbid medical conditions, psychological issues such as anxiety and stress, disrupted sleep, poor trunk stability, movement disorders, and the environment (increased ambient temperature). MS-related fatigue has been characterized as primary and secondary fatigue.

It is hypothesized that primary fatigue is due to the disease process itself: cortical damage, conduction blocks from demyelinated motor pathways, increased energy demands for muscle activation, and increased energy demands from co-contraction of agonist and antagonists. In MS, secondary fatigue may be due to deconditioning, respiratory muscle weakness, and pain. Interventions that may counteract fatigue and result in improved occupational functioning include cooling via use of a cooling garment, energy conservation techniques, and aerobic conditioning. Schwid et al examined the use of a liquid cooling garment worn 1 hour per day for 1 month. Subjects reported less fatigue during the month of daily cooling, and cooling therapy was associated with objectively measurable but modest improvements in motor and visual function as well as persistent subjective benefits.

Both Mathiowetz et al and Vanage et al have documented the effectiveness of a group format energy conservation program for people living with MS. A six-session, 2-hour-per-week energy conservation course taught by occupational therapists for groups of 8 to 10 participants resulted in less fatigue impact, increased self-efficacy, and improved quality of life. A variety of energy conservation techniques may be used with the MS population (Box 27-2).

Aerobic training has been demonstrated to improve overall fitness and decrease fatigue in the MS population. Petajan et al tested a program of 30 minutes of combined upper extremity (UE) and lower extremity (LE) ergometry plus 5 minutes of warm up and cool down (40 minutes total) three times per week. This program resulted in multiple benefits, including decreased fatigue. The OTA must work closely with the occupational therapist to determine the correct intensity and duration of any exercise program with this population.

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**Box 27-2**

**Energy Conservation Techniques Used by Patients with Multiple Sclerosis**

- Pacing
- Successful work/rest ratio
- Use electronic aids as needed
- Flexible home and work schedules
- Recognition of fatigue warning signals
- Successful use of compensatory strategies
- Acceptance of a request for assistance
- Home/work modifications
- Appropriate ambulatory aids
- Power mobility aids (power wheelchair or scooter)
- Control of spasticity
- Improved trunk control
- Techniques to control tremor
- ADL assistive devices
- Durable medical equipment
- Heat control
- Pharmacologic interventions
Improving Participation via Control of Tremors and Movement Disorders

Movement disorders such as tremor and ataxia are common problems for people living with MS. Compensatory strategies appear to be the most successful for controlling movement disorders and improve performance despite their presence (Box 27-3).20,21,29

Improving Participation via Cognitive Compensations

Decreased cognitive functioning significantly impedes the rehabilitation process because the patient is less able to store and receive new information. Cognitive changes may be present throughout the disease and fluctuate with time of day, task difficulty, and environmental distractions; cognitive function may worsen with fatigue.3 Short- and long-term memory deficits contribute to confusion and agitation. New information should be presented to the patient simply and repetitively. Consistency in the therapy program as to day, time, and modalities helps to orient the patient and minimize frustration.

Compensatory strategies seem to be the most effective intervention, as remediation of cognitive deficits (e.g., memory drills) has little research support (Box 27-4).

Improving Participation via Managing Sensory Deficits

The patient with MS may have a variety of sensory and perceptual disorders. In most cases, therapists aim to make the patient and family aware of the problem and to teach the patient to use compensatory techniques. Loss of tactile sensation, especially for stereognosis, interferes with performing fine motor tasks such as buttoning, managing utensils, money manipulation, computer use, and writing. If there is no visual loss the patient may be able to compensate visually. Adaptations such as built up handles, use of Velcro, etc. may help the client compensate.

Patients with loss of pain and temperature sensation need to be cautioned to avoid situations that could cause burns or other injuries, particularly kitchen and bathroom activities. On the other hand, patients may present with pain secondary to a variety of causes such as decubiti, muscle imbalance, overuse of a particular movement pattern, postural malalignment, etc. A variety of conservative techniques may be used to decrease pain and include deep breathing, visualization, biofeedback, correcting muscle imbalance, correcting postural alignment, transcutaneous electrical nerve stimulation (TENS), cryotherapy (cooling), and adapting relevant occupations to decrease the use of compensatory movement patterns.

Visual deficits such as double vision, blurred vision, decreased acuity, and nystagmus may make even simple ADL difficult. Patients can be taught compensatory techniques such as covering one eye (full or partial visual occlusion) to minimize double vision or using devices for those with low vision such as magnifying glasses, large-print books, and audio-books. Consider the following intervention guidelines when working with clients with visual loss:9

- Magnification/enlargement via magnifying devices, large print, and so on

Box 27-3

Interventions Related to Ataxia and Tremor20,21,29

- Orthotics/splinting (wrist support, thumb support via opponens splints, cervical collar)
- Using the environment for stability (e.g., leaning on the work surface, high back chairs)
- Adaptive devices (Dycem®, long straw, suction devices, built-up handles)
- Assistive technology (speaker phone, adapted mouse/keyboard)
- Weights (wrist weights, weighted gloves, weighted devices, etc.) may be effective for subtle movement disorders.
- Posture/position of activity focused on trunk support
- Minimize the number of joints moving simultaneously during activities
- Keep upper extremities stabilized against the trunk
- Keep the elbow on the work surface (propping)
- Prevent a calm/focused emotional state
- Control fatigue
- Adapt activities to eliminate the need to reach into space
- Decrease effort
- Decrease fine motor coordination demands
- Experiment with both slow and fast movements
- Provide exercise for proximal (trunk/scapula) stability
- Pharmacologic management

Box 27-4

Cognitive Strategies Used by Patients with Multiple Sclerosis

- Use memory aids: timers, watch alarms, personal data assistants, reminder lists, memory books, Post-it notes, computerized organizers, dictation systems.
- Allow extra time for task completion and processing.
- Decrease environmental distractions.
- Avoid multi-tasking.
- Schedule difficult cognitive tasks during periods of high energy.
- Avoid fatigue.
- Delegate responsibilities.
- Solve problems aloud.
- Check work for accuracy.
- Keep organized and avoid clutter.
- Determine which (visual or auditory) processing system is most effective.

- Color contrast—for example, using white dishes on a blue placemat
- Decreasing background clutter to increase clarity
- Increased and task-focused illumination. Consider task-specific lighting based on where clients cook, manage medication, and so on.

Improving Participation via Strengthening and Endurance Training

Patients with MS are taught by the OTA to plan their day and pace themselves accordingly (see Box 27-1). Planning and
energy conservation help the patient budget strength and endurance to meet daily needs. Important activities, including exercises, are done in the morning or after a scheduled nap or rest period. Graded resistive exercises increase the strength of key muscle groups and should be targeted to specific muscle groups that are task-specific. To increase endurance, emphasis should be placed on increasing repetitions rather than increasing weights. The benefits of aerobic exercise for this population have been documented in terms of improved fitness and quality of life.31 The OTA should work closely with the occupational therapist to determine the correct duration, frequency, and intensity of exercise as to avoid over-fatigue.

Patients with severe loss of muscle strength sometimes can perform functional activities with the help of devices that substitute for weak muscles. For example, the OTA may encourage the patient to use an overhead suspension sling or mobile arm support to increase independence.

**Contracture Prevention and Treatment**

Patients with severe weakness that prevents full active range of motion (AROM) as well as those with spasticity have the potential to develop soft tissue contractures. Contracture is prevented by deliberate and regular limb movement; active movement is preferred over passive when possible. Moving the patient through complete ROMs—and not just the middle ranges—is essential. Therapists must determine what a full ROM is for each individual patient; therefore age-related factors must be considered. A joint that moves or is moved through its full ROM via engagement in daily activities develops almost no deformities. A program of AROM and passive range of motion (PROM) combined with a terminal stretch at least twice per day is recommended if contracture is beginning to develop. Low-load prolonged stretch via splints or positioning must be used if a contracture has developed. During the terminal stretch the proximal body part should be well stabilized.

Whenever possible, the patient should perform self-ranging techniques. PROM of each joint through the full range must be done daily. The occupational therapist may be responsible for teaching the family or a nursing assistant the techniques of ROM. The occupational therapist may administer inhibition techniques (such as icing or positioning to decrease tone before PROM) to patients with severe spasticity. Splints to maintain range or provide sustained stretch may be indicated to treat or prevent contractures of the elbow, wrist, ankle, or hand.

**Activities of Daily Living**

Independence in ADL promotes self-esteem and quality of life.32 OT may provide assistive devices and adaptive techniques to enable MS patients to be safer, more independent, and more efficient in both ADL and IADL. Cups with lids, scoop dishes, and adapted utensils facilitate eating. Long-handled shoe horns, reachers, sock aids, and elastic shoe laces are just some of the devices that can increase independence and ease of dressing. The OTA needs to work with the patient to evaluate the effectiveness of equipment and provide training on an individual basis. As the disease progresses, modifications in equipment and techniques will be needed. A severely disabled patient may require an environmental control unit (ECU) to operate lights, television, or radio with a simple switch. The OT practitioner may be asked to determine which body part should activate the switch and what type of switch should be used.

**Communication**

In MS, both written and verbal communication skills may be affected. The occupational therapist often works closely with the speech therapist to devise a method of improving the patient’s communication skills. Patients with severe deficits may be given augmentative communication devices such as communication boards or computers that speak for them. The OTA may provide equipment that enables the patient to use these devices, such as splints for UE stabilization or head or mouth pointers. Adaptive writing devices (built-up and weighted pens, pen holders, and magnetized wrist stabilizers) compensate for decreased coordination and weakness. Special computer keyboards and large-button and speech-activated telephones facilitate independent communication for patients with motor deficits.

**Seating and Wheeled Mobility**

Many patients with MS will require the use of a wheelchair for mobility as their disease progresses. The primary considerations in recommending a wheelchair for a MS patient are the following:

- Overall endurance
- Trunk control
- LE strength, coordination, sensation, and endurance
- UE strength, coordination, sensation, and endurance
- Disease prognosis

Patients with sufficient UE strength and overall endurance should have a lightweight, high-strength manual wheelchair if they are to propel their own wheelchairs. Patients with a rapidly progressive type of the disease may require wheelchairs with reclining backs or a tilt-in-space frame to compensate for diminished trunk control. In some cases an electric wheelchair or electric scooter may be indicated. Modifications to the wheelchair that are helpful with the MS population include oblique rim projections, lateral supports, solid back and seat inserts, head positioners, and brake extensions. Training in wheelchair mobility and transfer techniques may be taught as part of the patient’s OT program. In a long-term care setting, MS patients with poor alignment and no wheelchair mobility skills may be positioned in geri-chairs for proper alignment and comfort.

Proper positioning of the patient both in and out of the wheelchair is under the scope of the OT department in many facilities. MS patients are at high risk for decubitus ulcers because of their diminished sensation, incontinence, and poor ability to reposition themselves. Pressure-relieving cushions and mattresses should be provided to all MS patients. Patients with sufficient strength should be taught and
include the following: 2, 43
characteristics of those who cope well with MS (and other diagnoses) and (less commonly) antisocial behavior. Assets and characteristic is higher for people living with MS than for the general population or for people with other neurological diagnoses. Other affective characteristics include bipolar disorder, lability, and (less commonly) antisocial behavior. Assets and characteristics of those who cope well with MS (and other diagnoses) include the following: 39

• Support
• Connectedness
• Sense of humor
• Spirituality
• Openness

Clinicians may assist in fostering adaptive coping strategies such as active coping, seeking emotional support, seeking instrumental support, positive reframing, planning, humor, acceptance, and religion. 39

The OTA can offer emotional support to both the patient and family. The patient should be encouraged to maintain a daily schedule of activity. Referral to MS support groups sponsored by the local MS Society may be helpful in dealing with the daily stress of having a chronic progressive disease. Additional support and information are available online at the reference and health sites and in chat groups.

The members of the rehabilitation team, including the OTA, have an obligation to help the family and the patient adjust to the disease and to provide encouragement, emotional support, training, and exercise programs. OT can help the MS patient function as productively as possible within the limits of the disease.

Leisure Skills
Leisure skills provide mental and social stimulation and substitute avocational interests for vocational skills. The OTA can assist the patient by stressing the importance of activity and by recommending adaptive devices such as card holders and shufflers and adapted board games. The patient should be encouraged to manage time effectively, planning for social events with naps and limited exercise on days when evening activities are scheduled. 36

Work Skills
MS patients may be able to maintain their ability to work during various stages of the disease or during remission, with changes to their schedule and adaptive devices to increase ease of work tasks. Worksite modifications for wheelchair accessibility may be required. The OTA may assist the occupational therapist in conducting the vocational evaluation, which may be used to determine a worker’s ability to continue the job, and in making recommendations for adaptive equipment.

Psychosocial Issues
The patient and his or her family typically greet a diagnosis of MS with shock, denial, and anger. However, in some cases, the patient may feel some relief in finally knowing that the symptoms were not psychosomatic. As the disease progresses, the client may become depressed. 31, 42, 43 Incidence of depression is higher for people living with MS than for the general population or for people with other neurological diagnoses. Other affective characteristics include bipolar disorder, lability, and (less commonly) antisocial behavior. Assets and characteristics of those who cope well with MS (and other diagnoses) include the following: 39

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Parkinson’s Disease
PD is a slow, chronic, progressive disease of the nervous system that was first described by James Parkinson in 1817. PD is characterized by four cardinal signs: resting tremor, rigidity in skeletal muscle, bradykinesia, and postural instability. Pathology is characterized by degeneration in dopaminergic pathways in the basal ganglia, particularly in the substantia nigra. The function of the substantia nigra is to produce dopamine, the neurotransmitter that transports signals to motor control areas such as the caudate and putamen. In PD, dopaminergic neurons deteriorate at a fast rate and the amount of dopamine that is produced decreases, resulting in initial impairments. When signs and symptoms are noticed, 80% of the dopaminergic neurons have already deteriorated. Diagnosis is made by the presence of at least two of the cardinal signs as well as the client’s response to levodopa, a dopamine precursor. 8, 46

The symptom complex of PD is termed parkinsonism. Not all clients with parkinsonism have PD. Besides the formal diagnoses of PD other pathologies that result in parkinsonism include drug-induced parkinsonism (i.e., parkinsonism has been associated with the use of antipsychotics), progressive supranuclear palsy, corticobasal degeneration, multiple system atrophy, and vascular parkinsonism (multiple small strokes). 46

Epidemiology
At least 1 million Americans, or 1% of older adults, are living with diagnosed PD. Approximately 40,000 individuals are diagnosed each year, and thousands of people live with the disease undiagnosed. Approximately 15% of those living with PD are diagnosed before age 40, although the average age of diagnosis is 60. 8, 46, 61

Cause
The cause of PD is not clear. Researchers hypothesize that causes include a combination of genetic and environmental factors. Approximately 15% to 25% of people living with PD have a relative with the disease. Risk of developing PD is increased twofold to threefold if a first-degree relative is affected. Those who sustain serious and recurrent traumatic brain injuries (e.g., professional boxers) may develop a form of PD, as may those living in rural conditions and those exposed to herbicides, pesticides, or some synthetic narcotic agents. Further research continues to attempt to establish the cause of the disease. 8, 46

Impact on Client Factors
PD normally affects the client’s motor systems first and foremost. Examples include bradykinesia (slowness of movement), resting tremor, pill-rolling, tonal changes such as cog-wheel rigidity (rhythmic/jerky/ratchet-like resistance to passive movement), loss of gross and fine motor coordination, loss of coordination for writing (presenting as
micrographia—small and crowded writing style), and decreased facial expression (mask face).

Gait and balance are also affected.31 The client develops a stooped posture and loose arm swing during gait. Episodes of freezing51—sudden difficulty in walking through doorways or making turns—are experienced during gait activities. Postural dysfunction and rigidity result in typical gait patterns that are characterized by a stooped forward posture; a slow, shuffling gait; or a festinating gait that includes small, fast steps, which propel the patient forward with ever-increasing speed. The patient has difficulty in stopping the forward motion. During standing activities the client may experience retropulsion (falling backwards).

As the disease progresses, the patient experiences problems with oral musculature, resulting in drooling, dysphagia, and monotone speech with low volume. The patient may demonstrate disorders in bowel and bladder control. Depression is common in this population.35 Dementia and memory deficits are documented as well.4,18,49 Signs and symptoms are progressive and may be staged for the purposes of developing OT interventions (Box 27-5).26 All of the above impairments result in decreased participation and performance in areas of occupation.

Medical Management

Although no cure for PD exists, medical management concentrates on relieving the symptoms of the disease primarily through medication and, less often, through surgical techniques. Levodopa is the most widely prescribed medication to manage the symptoms of PD and is usually prescribed in a combined carbidopa-levodopa formula called Sinemet. The OTA should be aware that clients being managed with drugs such as Sinemet will fluctuate in motor and functional status during “on and off” periods. It is helpful to work with clients during periods of both optimal and nonoptimal drug responses to have a full overview of the client’s functioning throughout the day. Level of assistance required can vary greatly depending on the timing of drug administration.15

Dopamine agonists also commonly used with this population include Parlodol, Permax, Mirapex, and Requip. All dopamine drugs may cause nausea and decreased blood pressure; therefore clients should be monitored closely. In older adults, dopamine agonists may result in hallucinations. The OTA should report any adverse reactions to the physician as soon as possible.

It is common in the early stages of the disease for the neurologist to prescribe milder medications such as the dopamine agonists or amantadine and add levodopa only when further symptom control is needed. Similarly, during the early stages of the disease, the physician may prescribe anticholinergic drugs, which reduce rigidity and tremor. In addition to antiparkinsonian drugs, medication for treatment of depression, nutritional supplements, and pain medication may be prescribed.35

If patients do not respond well to medications, surgical options include deep brain stimulation, pallidotomy, thalamotomy, and others.67 Like medication management, surgical interventions are used to manage symptoms, not to cure the disease. Specific symptoms that may be managed via surgical interventions such as deep brain stimulation include tremor, bradykinesia, and rigidity. Loss of balance, gait-freezing episodes, speech deficits, and postural deficits are not amenable to surgery.

Occupational Therapy Management

Participation restriction and activity limitations are substantial problems for those living with PD. Decreased performance in areas of occupation may result from multiple client factors impacted adversely in this population. In addition, self-imposed limitations may be a factor as well. Those living with PD may limit their involvement in life situations because of fear of falling and concerns related to continence, drooling, or the amount of time and energy required to participate in meaningful occupations. OT is usually initiated when those living with PD present with a decreased ability to participate in daily life.19,45

Precautions

The parkinsonian patient has diminished postural control and may present with orthostatic hypotension as well; therefore risk for falls is significant. Care must be taken during ambulation and transfers. Doorways, elevators, crowds, and surface changes may trigger freezing behavior.31 The patient may have dysphagia and should not be offered food or drink unless the clinician is sure of the client’s ability to swallow. The patient’s tendency toward immobility (coupled with incontinence) increases the risk of pressure ulcers. Patients need to be encouraged to stand or reposition themselves frequently and will benefit from pressure-relieving wheelchair cushions and mattresses.

Evaluation

The occupational therapist will perform the OT evaluation, focusing on functional performance level related to work,
Interventions
Evidence-based reviews of rehabilitation strategies including OT interventions have demonstrated a positive effect on both client factors and areas of occupation. In addition, they have demonstrated the need for further research to solidify the evidence base for OT intervention. In general, reviews have concluded that people living with PD can effectively learn new tasks and improve their functional performance through focused practice of meaningful tasks.

Improving Participation in Areas of Occupation
Patients with PD benefit from learning strategies designed to increase efficiency, safety, and independence. Strategies may be learned via graded task-specific practice and may include the use of adaptive devices and environmental modifications.

The patient should attend physical therapy for gait training as a foundation for functional mobility. The occupational therapist can supplement this training by using verbal cues to remind the patient to stand erect, lift the feet, and follow the prescribed gait pattern. Bed mobility skills, transfer training, and wheelchair mobility skills should be taught by the OTA if indicated. As the patient’s ambulation status declines, a wheelchair may be required. The patient should be advised to purchase a lightweight wheelchair. Oblique rim projections, a pressure-relieving cushion, elevating swing-away leg rests, and reclining backs should be considered.

PD often makes it difficult for patients to pursue their hobbies and interests. Adaptive devices for playing cards and board games, gardening, and doing crafts are available through various vendors. The OTA may have to encourage the patient and family to develop new interests.

Motor Skills/Prevention of Deformities
The increased rigidity and tendency toward immobility put the PD patient at great risk for contracture development and general deconditioning. The PD patient requires a daily home exercise program for AROM and stretching as well as clinic appointments in which the exercise program can be closely supervised by the OTA or occupational therapist. The frequency of the therapy is determined by the physician in consultation with the occupational therapist. AROM exercises may be done individually or in a group setting. Passive and/or active stretching exercises are indicated to maintain flexibility. Typical muscle groups and individual muscles that become limited and are a focus for stretching include:
- Hip flexors
- Knee flexors
- Gastrocnemius
- Pectoralis major and minor
- Anterior trunk/neck musculature

Box 27-6
Examples of Modifications in Activities of Daily Living for People Living with Parkinson’s Disease

<table>
<thead>
<tr>
<th>Feeding</th>
<th>Grooming</th>
<th>Bathing</th>
<th>Toileting</th>
<th>Writing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skills may be improved with weighted utensils (for very subtle tremors), scoop dishes, Dycem®, long straws, rocker knives, and cups with lids. Because of the control required for this task, meals should be timed with peak medication effects. See Box 27-3 for suggestions related to tremor control during feeding. Consider safe swallow strategies such as small bites/sips, alternating solids/liquids, staying upright after a meal, and avoiding taking thin liquids by straw. Food consistencies and the thickness of liquids should be considered.</td>
<td>Tasks are simplified with electric toothbrushes, electric razors, and hands-free hair dryers. Bimanual oral care and shaving may be helpful. Suction brushes and soap holders may increase ease of grooming. Grooming tasks should be performed while seated. See Box 27-3 for suggestions about controlling tremors that interfere with grooming.</td>
<td>Bathing can be performed safely and more independently using long-handled brushes, “soap on a rope,” soap pumps, and bath mitts. Durable medical equipment such as a bath bench or seat, grab bars, and a handheld shower will decrease fall risks and increase independence as well. No-slip bath mats (inside and outside of the tub) should be considered. Sliding doors should be replaced with curtains.</td>
<td>Ability is enhanced and made safer via raised toilet seats, 3:1 commodes, or toilet frames in conjunction with grab bars. Bedside commodes, male/female urinals, or condom-style catheters for men may be options for nighttime toileting needs.</td>
<td>May be enhanced by rhythmical writing programs, lined paper, or built-up pens. Printing may be easier than cursive writing. Word processors may be an option for some.</td>
</tr>
</tbody>
</table>
In the later stages of the disease, splinting may be indicated to maintain joint ranges and skin integrity. Verbal prompting and use of visual cues (sitting patient in front of mirror) can be used to promote improvement in posture. Encourage the patient to take deep breaths, and offer breathing exercises if indicated.

Clinicians can use visual, tactile, and auditory cues to help patients initiate movement. Auditory cues should be short, firmly spoken commands such as "stop," "step up," etc. Rhythmic music and counting can also help to initiate movement. Auditory commands coupled with counting are especially helpful in teaching the patient transfer techniques.

Graded resistive exercises and gross motor activities, particularly sports activities, are used to develop strength and general mobility. Functional fine motor tasks, such as jewelry making, manipulating money, and picking up small objects may assist in developing and maintaining hand function and coordination. These tasks can be graded by changing the size of the objects. The clinician should monitor and record the time it takes for the patient to complete the task assigned. Hand-strengthening modalities include repetitions with hand grippers and therapeutic putty exercises.

### Communication

Parkinsonian patients often develop a monotone, low-volume speech. OT practitioners can increase the benefits of speech therapy by providing breathing and postural exercises. Diminished blinking responses and disturbances of the ocular muscles impair the patient's ability to read. Large-print books and audiobooks are useful with these patients. Computers and word processors offer an alternative for patients who have difficulty writing. Felt-tip markers are easier to use than are regular pens. A signature stamp is helpful in the workplace. Cordless and automatic dialing telephones simplify communication.

### Psychosocial Issues

The person with PD typically tends to withdraw from society because of embarrassment, difficulty in mobility, and depression. The patient and family need a daily schedule that encourages exercise, outside activity, and social contacts. Information and support groups for patients with PD and their families are available through local chapters of the Parkinson’s Disease Foundation. The National Parkinson Foundation and the online computer network for PD also offer advice and education. Group counseling and day treatment programs provide emotional and social outlets.

### Advanced Parkinsonism

In the latter stages of the disease, patients have severe deficits in communication, mobility, swallowing, and cognition. Social isolation becomes a serious problem. Secondary complications, such as decubitus ulcers, aspiration pneumonia, fractures from falls, and contractures may arise. The OTA may help to prevent some of these problems. Use of a pressure-relieving mattress and cushion for the wheelchair, proper head alignment and use of a flow control cup at meals, daily PROM, and splinting are interventions that should be explored with this population. Group activities at the patient’s cognitive level help minimize the social isolation often experienced by those with advanced parkinsonism.

### Amyotrophic Lateral Sclerosis

ALS, or Lou Gehrig’s disease, is a progressive disease characterized by the degeneration of the motor neurons in the anterior horn cells of the spinal cord, brain stem, and corticospinal tracts. Classic ALS involves both the upper and lower motor neurons with onset in the bulbar area or limbs. After diagnosis and the onset of symptoms, the average life span is two to five years.

### Epidemiology

ALS is the most common form of motor neuron disease. Men are twice as likely to have the disease as women. The usual age for diagnosis of ALS is between the ages of 40 and 60. People of all races and ethnic backgrounds are affected. Approximately 30,000 Americans live with ALS, and an estimated 5000 are diagnosed each year.

### Cause

The cause of ALS is unknown. The majority of cases (90% to 95%) occur randomly (sporadic), whereas 5% to 10% of cases are considered familial. The pattern of inheritance requires only one parent to carry the gene that causes the disease. Approximately 20% of familial cases result from a gene defect. Environmental factors such as exposure to toxins or infectious agents are also being researched.

### Impact on Client Factors

ALS affects voluntary muscles. Because ALS involves both the upper and lower motor neurons, motor involvement includes both spasticity and stiffness (upper motor neuron) and weakness, low tone, and atrophy (lower motor neuron). In addition, bulbar signs such as speech deficits, swallowing difficulties, and respiratory involvement occur. Eye muscles; external sphincters controlling bowel and bladder management; the five senses; and the heart, liver, and kidneys are usually spared. Early symptoms of the disease include difficulty walking and/or picking up objects, and/or performing fine motor tasks. The number and side of the limbs affected vary from person to person. The client complains of weakness and stiffness. There is atrophy of the intrinsic muscles of the hands. The client exhibits hyperactive reflexes and fasciculations (twitching) that can be observed under the skin. Complaints of cramping are common. The weakness spreads to other muscle groups relatively quickly and involves all the limbs and the neck and trunk muscles. Eventually the client’s muscles become flaccid, resulting in severe disabilities (i.e., requiring total care) in performance in areas of occupation. Although ALS is
primarily a disease of the motor system, emerging evidence suggests the presence of some cognitive involvement similar to that of frontal and temporal lobe dementia processes.\textsuperscript{1,2,4,6} Signs may include decreased judgment and decision making.

Because of the severe pattern of weakness that emerges, ALS clients may consider themselves prisoners of their bodies. As the disease progresses, clients, families, and the team must decide whether or not ventilator support will be administered as respiratory muscles fail. Death generally results from respiratory complications.\textsuperscript{37}

**Medical Management**

There is no cure for ALS. Treatment is primarily palliative. The Food and Drug Administration has approved the drug Rilutek to slow progression of the disease; it can prolong survival up to several months. The drug may also extend the period before ventilator support is needed.\textsuperscript{47}

Medication is also prescribed to control uncomfortable symptoms and improve quality of life through control of muscle spasms and pain, minimization of drooling, and treatment of depression. Respiratory and swallowing problems may require tracheotomy and gastrostomy procedures. Frequent suctioning to clear the airway may be needed. Because of the patient’s compromised respiratory system, care should be taken to avoid exposure to respiratory infections.

**Occupational Therapy Management**

The role of OT with the ALS patient is to enable the client to adapt and to maintain the maximal level of functioning throughout the course of the disease as well as to assist care providers with the necessary skills to safely and effectively assist with daily care issues.\textsuperscript{6,11,65}

The initial OT evaluation establishes a baseline of functional abilities and limitations related to areas of occupation, ROM, muscle strength and tone, pain, and chewing and swallowing abilities. In addition, information regarding the home layout is obtained. Frequent reevaluation of the patient’s status is required as the disease progresses. Evaluation procedures include standardized evaluations, interviews, and performance-based observations. The ALS Functional Rating Scale\textsuperscript{82} is used throughout the country to monitor disease progression in this population.\textsuperscript{17,23} It measures the following areas: speech, salivation, swallowing, handwriting, cutting food, dressing and hygiene, turning in bed, walking, climbing stairs, dyspnea (shortness of breath), orthopnea (difficulty sleeping), and respiratory insufficiency. The OTA may assist in the evaluation of occupational performance areas.

**Interventions**

**Improving Participation in Areas of Occupation**

Intervention will vary depending on the stage of the disease. Early symptoms may include loss of fine motor coordination and hand weakness. Assistive devices such as built-up utensils and writing devices, Dycem\textregistered{}, suction devices, scoop dishes, plate guards, key holders, and devices to open containers may help improve function. Early balance changes due to weakness (i.e., foot drop) may necessitate adaptive ambulation devices or braces (e.g., an ankle foot orthotic) to prevent falls and improve upright function. The OTA should focus on using ambulation devices in functional situations such as kitchen activities and train the patient in relation to daily living problems such as transporting and carrying items.\textsuperscript{6} Walker baskets or trays may be useful. Energy conservation techniques should be taught in the early stages of the disease.

As the disease progresses and UE weakness continues to progress, further adaptations are needed. Functional splints may be necessary to maintain the ability to eat, use a computer, write, use a communication device or ECU, or turn pages. Typical splints that are used to maintain or increase function in this population are wrist extension splints, short or long opponens splints, universal cuff, or dorsal wrist extension splints with a universal pocket (Figure 27-1). In addition, head and neck stability may be compromised, thus requiring a cervical collar. Splints may be used in conjunction with an overhead suspension sling or deltoid aid to compensate for proximal weakness (Figure 27-2).

Trunk and/or LE weakness will impact the client’s functional mobility skills. Transfers may be made easier for both clients and caregivers by teaching the use of a sliding board or mechanical lift.

Depending on financial means, home modifications such as ramps, elevators, stair-glides, and ceiling lifts may be necessary. Similarly, bathroom modifications and equipment such as roll-in showers, rolling commodes, pocket doors, offset hinges, removal of glass shower doors, and/or adapted bath seats help maintain independence, increase safety, and facilitate caregiver assistance.

**Motor Skills and Prevention of Deformities**

When using gentle therapeutic exercises with this population, the OT practitioner should take care to avoid fatiguing the patient. General conditioning and mild aerobic exercises are recommended. PROM is provided when the patient cannot move the joint actively to the end of the range; PROM may be used in conjunction with positioning and splints to prevent contracture and control spasticity. Relaxation and deep breathing exercises should be taught. If exercise is an important occupation for the client, it should be done over several short periods throughout the day, rather than in one long session, so as to avoid fatigue. Exercise will not result in increased muscle bulk but may improve aerobic function, decrease fatigue, or help control depression. In the latter stages of the diseases more intensive passive stretching and ranging and splinting (i.e., resting hand splints) are required to prevent contracture development.\textsuperscript{41}

**Communication**

The trauma associated with the loss of verbal communication skills is compounded by the fact that the patient retains significant mental capacity. The OT and speech therapy departments need to work jointly to establish an alternative
Degenerative Diseases of the Central Nervous System

Assistive Technology

Initially the patient may be able to write. A communication board, an electronic communication aid, and a computer with a voice module offer other alternatives. The OTA may be involved in positioning the patient and fabricating the UE equipment and switches that enable use of these devices. It is important that the patient be able to call for help. Call bells that can be controlled with any part of the body with a minimal amount of pressure are available and will become necessary when the patient can no longer speak.

Mobility and Positioning

In the early stages of the disease, the patient will continue to ambulate with the help of a cane or walker but eventually will require a wheelchair. Although initially the patient may have sufficient strength to sit and propel a standard wheelchair, the progressive nature of the disease will inevitably necessitate

Figure 27-1 Typical splints used to increase or maintain function in those living with ALS. The splints provide stability for unstable joints secondary to weakness and place the wrist or hand in a functional position. A, Wrist extension splint. B, Short opponens splint. C, Long opponens splint. D, Universal cuff. The universal cuff can be used in conjunction with a wrist support as well. (A-C From Fess EE, Gettle KS, Philips CA et al: Hand and upper extremity splinting: principles and methods, ed 3, St. Louis, 2005, Mosby; D Courtesy Sammons Preston, Bolingbrook, IL.)

Figure 27-2 Overhead suspension sling used for those with proximal upper extremity weakness to enhance self-feeding, computer use, etc. (From Gillen G, Burkhardt A: Stroke rehabilitation: a function-based approach, ed 2, St. Louis, 2004, Elsevier.)
a reclining or tilt-in-space motorized wheelchair. For financial reasons, it may be wise to plan ahead when consulting on a wheelchair purchase. Proper positioning of the head may require a cervical collar and head support. Lateral supports to the wheelchair may have to be supplied in the latter stages of the disease. Pressure-relieving cushions are indicated. Local ALS care centers may be helpful in terms of procuring equipment via a lending program or suggesting ways to obtain financial assistance.

**Psychosocial Issues**

Reactive depression and anxiety disorders are common among those living with terminal diseases such as ALS. Medication and counseling may help ameliorate depression. The rehabilitation team needs to provide psychological support to help in coping with the devastating effects of the disease. Independence in meaningful activities may increase the patient’s self-esteem in addition to improving functional status. Support groups or individual counseling for clients and their families should be recommended.

**Alzheimer’s Disease**

Dementia is not a disease itself but rather a set of symptoms that accompanies a disease. Dementia refers to the loss of mental function in two or more areas such as language, memory, visual abilities, spatial abilities, or judgment that is severe enough to impact daily life. Many diseases cause dementia including multi-infarct dementia, PD, Huntington’s disease, Pick’s disease, Creutzfeldt-Jakob disease, etc. Other conditions such as AIDS, traumatic brain injury, and tertiary syphilis may result in symptoms of dementia. The most common known cause of irreversible dementia in adults is AD. Also known as senile dementia of the Alzheimer type (SDAT), this disease was first described by Dr. Alors Alzheimer in 1907. AD is a progressive, slow deterioration of brain tissue that results in decreased cognitive abilities, including deficits in language and memory, disturbances in the sense of time and place, personality changes, and emotional instability.

**Epidemiology**

AD affects approximately four and one half million Americans. By the year 2050, because of aging of the general population, the number of individuals living with AD could range from 11 to 16 million. One in ten Americans has a family member with AD, and one in three Americans knows somebody with the disease. In addition, one in ten individuals over 65 years old is affected, as are nearly half of those over the age of 85. Rarely, inherited forms of the disease can affect individuals in their 30s and 40s.

A person with AD will live an average of eight years from the diagnosis but can also live up to twenty years after initial diagnosis. After diagnosis, individuals with AD survive about half as long as those of a similar age without dementia.

**Cause**

The cause of AD remains unclear. Potential causes and risks for AD include the following:

- Increasing age. The likelihood of developing AD doubles every five years after age 65.
- Family history of the disease. The likelihood of developing AD increases as the number of family members with AD increases.
- Familial AD as been linked to specific genes. Most familial AD occurs before age 60.
- Variations in certain genes are being studied to determine whether they make some individuals more or less susceptible to the disease.

**Signs and Symptoms**

Early warning signs of AD include memory loss, difficulty performing familiar tasks, problems with language (i.e., as in word finding or word substitutions), disorientation, impaired judgment, decreased abstract thinking, misplacing items, mood or behavior changes, personality changes, and loss of initiative.

Various staging systems have been developed to document the disease progression from early stages to middle stages to late stages (Box 27-7). Understanding the stage of AD will assist in setting goals and planning treatment.

**Medical Management**

Treatment of the patient with AD is primarily symptomatic. No cure or treatment will slow the progression of the disease. Medications are available to treat some of the behavioral and psychiatric manifestations of the disease but are most effective in conjunction with behavioral interventions and environmental modifications. Examples of medications that may be used include Celexa, Prozac, Paxil, and Zoloft to ameliorate decreased mood and irritability. For anxiety and restlessness, Ativan or Serax may be useful, and for management of hallucinations or delusions Seroquel, Risperdal, Clozaril, and so on may be beneficial.

**Occupational Therapy Management**

The role of OT in the treatment of patients with AD varies according to the stage of the disease but aims to maximize independence and to provide guidance and support to the family.

**Occupational Therapy Evaluation**

The OT evaluation of the patient with AD establishes a baseline related to performance in areas of occupation and in cognitive, motor, and sensory skills. This evaluation is used not only to establish treatment guidelines and goals but also may help to determine the level of care and supervision necessary for the patient. Additionally, the evaluation will help in determining the timing and type (physical, gestures, verbal) of cues that improve performance. Information from the evaluation may assist the family in
making the difficult decision of whether to maintain the patient in the home or to seek placement in an assisted living or long-term care facility.

Evaluation of cognition should be done through formalized assessment tools. Motor functioning should be evaluated by assessing active and passive ROM, coordination, balance, transfer skills, and praxis. The speech-language pathologist and the physical therapist may provide additional information to supplement the OT evaluation. Self-care evaluation includes observation of the patient’s ability to feed, dress, groom, and toilet. In the early stages of the disease the OT evaluation may also include the patient’s ability to perform housekeeping and work-related tasks.10,16

Leisure skills, including the patient’s ability to continue to participate in previously enjoyed hobbies, should also be assessed. A thorough history should be obtained from the patient’s family before evaluation.

The OT evaluation is performed by the occupational therapist. The OTA may be asked to complete the occupational performance part of the evaluation or to assist in determining the patient’s perceptual and sensory skills. The occupational therapist coordinates the findings of the evaluation and determines the recommendations and treatment goals for the patient.

**Interventions**

**Activities of Daily Living**

In the middle stages of AD the patient begins to demonstrate difficulty performing simple ADL. Frequent reminders to initiate a task—such as “wash your face” or “put on your shirt”—may be required. As the disease progresses, the patient has problems sequencing multi-step tasks and benefits from help with breaking the tasks down into one-step segments. This process, known as task segmentation, guides the patient to complete ADL tasks with the verbal prompting of the therapist and also helps to train and refocus the task at hand. Instead of saying, “Wash your face,” the patient is instructed, “Pick up the wash cloth. Put the wash cloth in the water. Now put some soap on the wash cloth. Wash around your mouth.” These instructions are offered in a calm, reassuring manner. Positive feedback in the form of praise is provided after each step. Physical prompts (hand-over-hand assistance) may also be given if the patient permits it. The OT should instruct patients’ caregivers in these task segmentation methods and may need to remind them of the importance of allowing patients to perform as much of their own daily care as possible.

A patient with AD typically demonstrates poor frustration tolerance. It is important to avoid situations that may trigger catastrophic reactions. Activities should be analyzed and graded to ensure that the patient has sufficient physical and cognitive skill to perform the task requested.

The patient’s physical deterioration may contribute to safety problems in the home. Bathroom safety devices should be prescribed if indicated. The family should be advised of hazards in the home or the patient’s environment that need to be removed or safeguarded. Scatter rugs, power tools, electric appliances, medications, stairways, windows, balconies, household cleansers, smoking materials, and the kitchen stove are just some of the items that represent a serious potential hazard to the patient.

Mealtimes can be difficult for both the patient and the caregiver. Confused patients are often messy, fussy eaters. Adaptive eating devices such as scoop dishes, spill-proof cups, and built-up utensils may be helpful. If the patient no longer uses utensils it may be easier to have the caretaker provide finger foods than to try to force use of a fork. The patient may demonstrate swallowing difficulties. The occupational therapist can instruct the caregiver in feeding techniques for the dysphagic patient, including changing the food consistency, maintaining the head in neutral position, putting small amounts of food on the spoon, ensuring that the previous mouthful of food has been swallowed before offering the next spoonful, encouraging swallowing with verbal prompting and facilitation techniques, using a thickening agent in liquids, and using a flow control cup.

**Box 27-7**

**Stages of Alzheimer’s Disease**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 1: No cognitive impairment</td>
<td>Subjective complaints of memory loss or word finding, not detected on formal examination.</td>
</tr>
<tr>
<td>Stage 2: Very mild cognitive decline; subjective complaints</td>
<td></td>
</tr>
<tr>
<td>Stage 3: Mild decline: word-finding deficits, naming objects, misplacing objects, decrease in planning.</td>
<td></td>
</tr>
<tr>
<td>Stage 4: Moderate cognitive decline: decreased knowledge of recent events, decreased ability to perform higher-level mental calculations, decreased memory for personal information, inability to participate in complex tasks, socially withdrawn.</td>
<td></td>
</tr>
<tr>
<td>Stage 5: Moderately severe cognitive decline: major memory gaps, assistance needed for complex daily living tasks, confusion related to orientation, inability to perform simple calculations, still knows own name and names of spouse and children, needs help picking out clothing based on season and weather.</td>
<td></td>
</tr>
<tr>
<td>Stage 6: Severe cognitive decline: memory worsens, personality changes emerge, extensive help required for ADL, occasionally forgets name of spouse, decreased dressing ability, dysfunction of sleep/wake cycle, assistance required for toileting, incontinence, delusions, hallucinations occur, compulsion/repetition of behaviors.</td>
<td></td>
</tr>
<tr>
<td>Stage 7: Very severe cognitive decline: does not respond to environment, mutism, inability to control movement, requires feeding assist, loses ability to walk and sit without assistance, dysphagia, skeletal muscle rigidity.</td>
<td></td>
</tr>
</tbody>
</table>

**Mild/Early Stage Alzheimer’s Disease**

**Moderate/Mid Stage Alzheimer’s Disease**

**Severe/Late Stage Alzheimer’s Disease**

The patient’s physical deterioration may contribute to safety problems in the home. Bathroom safety devices should be prescribed if indicated. The family should be advised of hazards in the home or the patient’s environment that need to be removed or safeguarded. Scatter rugs, power tools, electric appliances, medications, stairways, windows, balconies, household cleansers, smoking materials, and the kitchen stove are just some of the items that represent a serious potential hazard to the patient.
Environmental Design
The OTA can help the caregiver structure the environment to help maximize the patient’s functioning. Eliminating clutter in the environment helps to minimize confusion. Contrasting colors make it easier for the patient to differentiate an object from the background. Simple changes such as eliminating the condiments from the kitchen table and ensuring contrast of color between the plate and table (and the plate and the food) simplify mealtime. Contrasting the color of the toilet seat and bowl with the bathroom floor helps to aid in toileting. Minimizing the amount of furniture and maintaining traffic areas free of obstacles decrease the risk of falls in the home. Gates and locks on stairways and doors may need to be installed for safety. Signs identifying the patient’s room and bathroom may be helpful. The OT clinic should also incorporate some of these design features. 16, 30, 50, 58

Day Care and Group Activities
Programs at adult day care facilities offer the caregiver a respite from the daily pressure and stress of taking care of a loved one with AD. These programs offer a variety of group activities within a structured environment, striving to provide the patient with positive social opportunities. Group activities, whether offered in a day care setting or as part of the activity program in a long-term care facility, may be administered by OT clinicians. The patients often enjoy music-based activities, simple and familiar games, and crafts. Teaching new activities is not recommended. Also enjoyed are sensory stimulation, reminiscing games, and pet therapy visits. Childish activities that may be demeaning to the patient should be avoided. The occupational therapist working in this area should consult the numerous books on activity programs for the dementia patient for further suggestions.

Reality Orientation
Patients with AD become less oriented as the disease progresses. Formal daily orientation programs to review the patient’s name, the date, the weather, and the location may be helpful. In addition, the patient should always be addressed by name, and all staff should introduce themselves and tell the patient their function regularly as if they have just met. The practitioner should regularly review the names of the patients’ close family members and show them their rooms, the dining room, etc. Clocks and calendars should be clearly displayed. Memory books may also be used. 27

Exercise Programs
Exercise, whether offered as a group or individual activity, is important to maintain strength, coordination, and ROM. Simple group calisthenics can be done in a standing or sitting position. The addition of rhythmic music helps to keep the patient involved in the activity. Parachute activities and ball, scarf, or balloon tosses are good group exercise activities. Dancing and walking are activities that the caretaker can easily do with the patient. If the patient cannot participate in group exercise programs or if he or she has joint contractures, the patient should be involved in a daily PROM program. It is recommended that exercise programs be offered at the same time each day to help establish a routine.

Psychosocial Issues
Patients with AD may demonstrate a variety of behavior problems including agitation, physical aggression, depression, inappropriate sexual behaviors, “stealing,” paranoia, and hallucinations. The patient may receive some form of medication for these problems but behavioral interventions are frequently preferable to chemical interventions that may have serious side effects. 66 The OT practitioner should always approach the patient while using a calm, reassuring voice. He or she should not become angry or argue with the patient and should redirect and refocus the patient to a different topic. Clear instructions on how to behave or what is acceptable should be offered. Using the name of the patient’s spouse in the request often helps to achieve the desired behavior (e.g., “Mary wants you to take a bath now.”). Reducing the amount of stimulation, noise, or unstructured time sometimes helps to decrease agitation and antisocial behavior. 56 The patient should be seen in OT when the clinic is relatively quiet and distractions are minimal.

In the final stages of the disease, the patient most often requires long-term and skilled nursing care. The patient may receive OT services in the form of nursing rehabilitation programs for ROM, positioning recommendations, splinting to prevent contractures or maintain ROM and skin integrity, and sensory stimulation.

Throughout the course of the disease, the family will need guidance and support to help cope with the effect of the AD on the patient and the family structure. 27 The nature of the disease demands changes and may necessitate role reversals in the family. The OTA and other team members should be prepared to provide support for these difficult transitions. Families may also seek advice from the Alzheimer’s Disease and Related Disorders Association and from online chat groups and informational services.

Summary
This chapter has focused on four degenerative diseases: MS, PD, ALS, and AD. Although other neurological diseases also result in progressive loss of function, the OT practitioner will find that the treatment goals and interventions for the diseases outlined in this chapter will be helpful in treating all patients with degenerative disorders.

Managing this population is difficult and can take an emotional toll on clients, therapists, and caregivers. Because of the multitude of problems encountered in this population, close collaboration between the occupational therapist and the OTA is necessary. In addition, all people living with these diagnoses can benefit from an interdisciplinary approach for optimal outcomes.
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CASE STUDY

Peter

Peter is a 30-year-old loan officer who enjoys movies, fishing, eating out, and concerts. He was recently engaged to be married. Peter has a five-year history of multiple sclerosis. One week ago he suffered an exacerbation of symptoms. He is motivated, alert, and oriented x 3. He is able to follow complex directions but lacks concentration on tasks as presented. His speech is understandable. He often loses his train of thought.

Motor function: Peter demonstrates 4/5 strength in both UEs. The greatest limiting factor is fatigue. His LEs are graded as 2/5 on manual muscle testing (MMT).

Sensory: UE sensation is intact, absent light touch, and pinprick is evident from his navel level and below.

Postural control: Peter requires close supervision for static sitting and moderate assist for all weight shifting and reaching activities beyond his arm span. He falls laterally and posteriorly while seated.

Neurobehavioral deficits: Peter’s greatest complaint is loss of short-term memory and feeling “disorganized.” He also reports feeling “blue and tired.” His greatest fear is that this exacerbation will limit his ability to work. In addition, the occupational therapist notes that he presents with deficits related to sustained attention and short-term memory.

ADL: Peter requires moderate assist for dressing, bathing, and toileting. In addition, all sliding board transfers require minimal assistance. He feels that his dependency in these areas is “humiliating.” Peter has just been transferred to the rehabilitation unit, and you will be the primary OT practitioner working with him. Peter’s desire is to return home and to work as soon as possible.

1. Reread the above material. Underline and star the problems of most concern to the patient.
2. Place problems in order of priority and select the first three. State a short-term goal for each. Describe one or more treatment methods the OTA could use to address this goal.
3. Describe the type of wheelchair and the wheelchair features that would be appropriate for Peter at this time.
4. Describe how you would respond to Peter’s desire to return to work.

Selected Reading Guide Questions

1. List the precautions that need to be observed when treating a patient with MS.
2. Briefly describe the three clinical signs associated with PD and explain how these signs affect the treatment process.
3. Discuss the psychosocial aspects of treating patients with degenerative diseases.
4. Describe how assistive technology can be used to increase the functional level of the patient with ALS.
5. Describe the treatment goals associated with each of the diseases discussed in this chapter: MS, PD, ALS, and AD.
6. Describe the ways the OTA can help the MS patient cope with fatigue.
7. Explain how the OTA can use adaptive devices to promote independence in feeding skills for patients with degenerative diseases.
8. Describe the techniques used by the OT practitioner to promote proper positioning and the prevention of decubitus ulcers in patients with degenerative diseases.
9. Describe the role of the OTA in the treatment and prevention of contractures with patients with MS, PD, ALS, and AD.
10. List some of the environmental changes that the OTA may recommend in the home of a patient with AD.

References


