Neuro Rehab- Module 3: Multiple Sclerosis and other Neuromuscular Diseases

Course Description:
Using a problem-solving approach based on clinical evidence, the text written by Darcy Umphred, PT, PhD, FAPTA, Neurological Rehabilitation, 6th Edition covers the therapeutic management of people with functional movement limitations and quality of life issues following a neurological event. It reviews basic theory and covers the latest screening and diagnostic tests, new treatments, and interventions commonly used in today's clinical practice. This edition includes the latest advances in neuroscience, adding new chapters on neuroimaging and clinical tools such as virtual reality, robotics, and gaming.

Module 3: Multiple Sclerosis and other Neuromuscular Diseases cover chapter 17 and 19
Chapter 17: Neuromuscular Diseases
Chapter 19: Multiple Sclerosis

Methods of Instruction:
Online course available via internet

Target Audience:
Physical Therapists, Physical Therapy Assistants, Occupational Therapists, Occupational Therapy Assistants and Athletic Trainers.

Educational Level:
Intermediate

Prerequisites:
None

Course Goals and Objectives:
At the completion of this course, participants should be able to:
1. Recognize the demographics of patients with neuromuscular diseases
2. Differentiate between the stages of Amyotrophic Lateral Sclerosis
3. Recognize the common physical findings in Bulbar Amyotrophic Lateral Sclerosis
4. Identify appropriate exercise interventions for patients with ALS according to disease stage
5. Recognize the medical prognosis for patients with Guillain-Barre Syndrome
6. Identify goals for the care of patient with GBS
7. Differentiate between functional transitions in patients with muscular dystrophy
8. Recognize appropriate therapeutic interventions for patients with GBS
9. Recognize the incidence and prevalence of Multiple Sclerosis
10. Differentiate between types and clinical characteristics of Multiple Sclerosis
11. Recognize the areas of symptom management including new approved drugs
12. Identify general exercise guidelines for levels of disability with MS
13. Recognize evidence-based interventions for specific problems in patients with MS

Criteria for Obtaining Continuing Education Credits:
A score of 70% or greater on the written post-test
DIRECTIONS FOR COMPLETING
THE COURSE:

1. This course is offered in conjunction with and with written permission of Elsevier Science Publishing.
2. Review the goals and objectives for the module.
3. Review the course material.
4. We strongly suggest printing out a hard copy of the test. Mark your answers as you go along and then transfer them to the actual test. A printable test can be found when clicking on “View/Take Test” in your “My Account”.
5. After reading the course material, when you are ready to take the test, go back to your “My Account” and click on “View/Take Test”.
6. A grade of 70% or higher on the test is considered passing. If you have not scored 70% or higher, this indicates that the material was not fully comprehended. To obtain your completion certificate, please re-read the material and take the test again.
7. After passing the test, you will be required to fill out a short survey. After the survey, your certificate of completion will immediately appear. We suggest that you save a copy of your certificate to your computer and print a hard copy for your records.
8. You have up to one year to complete this course from the date of purchase.
9. If you have a question about the material, please email it to: info@advantageceus.com and we will forward it on to the author. For all other questions, or if we can help in any way, please don’t hesitate to contact us at info@advantageceus.com or 405-974-0164.
OBJECTIVES

After reviewing this chapter the student or therapist will be able to:

1. Describe the basic pathology and medical treatment of amyotrophic lateral sclerosis, Guillain-Barré syndrome, and Duchenne muscular dystrophy.
2. Describe the current goals and interventions for each condition.
3. Describe the “safe” exercise windows related to disuse atrophy and exercise (overwork) damage.
4. Be able to apply intervention concepts discussed in this chapter to other neuromuscular diseases.

Amyotrophic lateral sclerosis (ALS), commonly known in the United States as Lou Gehrig disease, is a relentless, degenerative, terminal disease affecting both upper and lower motor neurons. Massive loss of anterior horn cells of the spinal cord and the motor cranial nerve nuclei in the lower brain stem results in muscle atrophy and weakness (amyotrophy). Demyelination and gliosis of the corticospinal tracts and corticobulbar tracts caused by degeneration of the Betz cells in the motor cortex result in upper motor neuron symptoms (lateral sclerosis).

In considering the movement dysfunction associated with these diseases, strength and endurance are most affected, with flexibility deficits resulting from these. All three disorders decrease a person’s ability to generate force in the affected muscles, with weakness as a primary symptom. Loss of muscle strength can lead to speech, swallowing, and respiratory difficulties along with functional limitations. Fatigue is another primary deficit, although the neurogenic disorders tend to result in central fatigue (deficit in ability to recruit motor units) as opposed to the peripheral fatigue of the myopathies (deficit in ability of muscle fibers to contract forcefully). Secondary movement problems include loss of range of motion (ROM) in immobile muscles and joints, and pain or muscle spasms. Adaptability, the ability to sense obstacles or changes in the environment and change the course of a movement in response, may be affected with the sensory loss in GBS but is not typically a problem in ALS or DMD.

AMYOTROPHIC LATERAL SCLEROSIS
Pathology and Medical Diagnosis

ALS, commonly known in the United States as Lou Gehrig disease, is a relentless, degenerative, terminal disease affecting both upper and lower motor neurons. Massive loss of anterior horn cells of the spinal cord and the motor cranial nerve nuclei in the lower brain stem results in muscle atrophy and weakness (amyotrophy). Demyelination and gliosis of the corticospinal tracts and corticobulbar tracts caused by degeneration of the Betz cells in the motor cortex result in upper motor neuron symptoms (lateral sclerosis).

The cause of ALS is unknown; however, numerous theories have been proposed. Ninety percent of the cases of ALS are sporadic without a known genetic component; however, most neurodegenerative diseases are now thought to be related to complex protein misfolding disorders. The latest research suggests that ALS and other neurodegenerative disorders are related to TDP-43 proteinopathy. Approximately 5% to 10% of the cases seem to have a complex genetic basis coded on ALS1 through ALS8 and other mutations that are associated with frontal lobe dementias. Twenty percent of genetic causes of ALS are thought to be related to mendelian mutations in the superoxide dismutase–1 (SOD1) gene (ALS1). Other factors considered in the genesis of ALS are vascular endothelial growth factors, toxicity leading to motor neuron death, oxidative stress and mitochondrial dysfunction related to microglial inflammation, and environmental factors.
The differential diagnosis for ALS is extensive. The possibility of cervical or lumbar spondylosis, syringomyelia, multiple sclerosis, primary lateral sclerosis, and diseases associated with lower motor neuron pathology, among other diagnoses, needs to be excluded before the diagnosis of ALS is made. Currently, no single laboratory test is available to confirm a diagnosis of ALS, although creatine phosphokinase levels are elevated in approximately 70% of patients and tend to be higher in patients with limb onset ALS than bulbar onset. Genetic testing to identify the mutations in the Cu,Zn SOD1 gene is available when a family history of ALS is present. Other laboratory tests, such as identification of biochemical markers in the blood and cerebrospinal fluid, are used to exclude other neurological diseases. Electromyography (EMG) and nerve conduction studies can be helpful to confirm the presence of widespread lower motor neuron disease without peripheral neuropathy or polyradiculopathy. Neuroimaging studies are used to rule out conditions that may have clinical signs similar to those of ALS.

Because of the absence of clear laboratory markers of ALS, the clinical diagnosis must be made on the basis of recognition of a pattern of observed and reported symptoms of both upper and lower motor neuron disease and persistent declines in physical functions supported by inclusionary and exclusionary diagnostic testing. Because of the overlap of symptoms with other neuromuscular disorders, misdiagnosis is not uncommon.

ALS is the most common form of motor neuron disease, with an incidence of approximately three to five cases per 100,000 persons. Mean age at onset is 57 years, with two thirds of patients aged 50 to 70 years old at time of onset. Men are affected approximately 1.3 to two times more frequently than are women, although the differences are less with late onset of disease (ages 70+).

Clinical Presentation

The World Federation of Neurology (WFN) has developed suggested diagnostic criteria (suspected, possible, probable, and definite) for patients with ALS entering clinical research trials. Essentially, a patient with "definite" ALS must show concomitant upper motor neuron and lower motor neuron signs in three spinal regions or in two spinal regions with bulbar signs. Either upper or lower motor signs must also be evident in other regions of the body. Exclusionary criteria are oculomotor nerve pathway abnormalities (the oculomotor nerve is spared in ALS), significant movement disorder patterns, sphincter control problems, the presence of sensory and autonomic nervous system (ANS) dysfunction, and cognitive deterioration. (Refer to the WFN ALS website [www.wfnals.org] section on ALS education for up-to-date criteria used for clinical studies.)

Although a consistent diagnostic criterion for ALS has been the absence of sensory involvement, some evidence exists that there is a progressive functional deficit in sensation, perhaps related to ongoing immobility.

Similarly, cognitive deficits are considered exclusionary criteria for an ALS diagnosis. However, a small subgroup of patients with both familial and sporadic forms of ALS has been identified as having concomitant evidence of frontotemporal dementia (FTD), showing lower scores on executive cognitive functions, word finding, and phrase length. A combination of ALS and FTD suggests a common cause may be possible. Because of these findings, therapists should be aware of the possibility of cognitive deficits in their patients with ALS, manifested as a decrement in executive skills such as planning and organization and language problems. Such patients may have more difficulty following through on medication and therapeutic recommendations, and their families may need more support. Unassociated with overall cognitive impairment, some deficits in action knowledge as opposed to object knowledge have been noted in patients with ALS, correlating with atrophy in the motor and premotor cortex. Specific cognitive deficits, therefore, may be more common than previously noted.

The earliest clinical markers heralding ALS are fasciculations (especially unequivocal fasciculation in the tongue), muscle cramps, fatigue, weakness, and atrophy. During initial diagnostic visits, patients frequently report to their physicians a profound sense of fatigue or the loss of exercise tolerance. Ninety percent of patients report weakness occurring in a striated muscle or group of muscles. Because the onset of ALS is insidious, most patients are not aware of the strength changes, or they have adjusted to the changes until they have difficulty with a functional activity such as tying shoes or climbing stairs. Physical examination usually demonstrates more widespread weakness and atrophy than reported by the patient. By the time most patients report weakness, they have lost approximately 80% of their motor neurons in the areas of weakness. This demonstrates the plasticity of the nervous system and its drive to adapt to meet functional goals. The weakness spreads over time to include musculature throughout the body. Succeeding symptoms of weakness in other muscles depends on the continued...
loss of motor neurons to the 20% threshold needed for perception of weakness. A typical, but not absolute, pattern of motor progression is early distal involvement followed by proximal limb involvement. In some cases bulbar symptoms herald the onset of ALS, but bulbar symptoms more commonly occur later in the disease. Flexor muscles tend to be weaker than extensor muscles.

Although the atrophy and weakness component of ALS is most obvious, 80% or more of patients show early clinical evidence of pyramidal tract dysfunction (e.g., hyperreflexia in the presence of weakness and atrophy, spasticity, and Babinski and Hoffmann reflexes). Although in some cases the upper motor neuron signs may be absent clinically, Chou has shown on autopsy that significant involvement may be present despite the lack of clinical evidence.

The pattern of ALS onset is highly varied, with several patterns identified by primary area of onset. Lower-extremity onset is slightly more common than upper-extremity onset, which is more common than bulbar onset. Some patients show initial symptoms in distal musculature of upper and lower extremities. A significant diagnostic feature of the pattern of disease is the asymmetry of the weakness and the sparing of some muscle fibers even in highly atrophied muscles. For example, a patient may have weakness of the right intrinsics and shoulder musculature or weakness of the left anterior tibial muscles. Bulbar symptoms are presaged by tongue fasciculations and weakness, facial and palatal weakness, and swallowing difficulties, which result in dysphagia and dysarthria. Pseudobulbar palsy is sometimes present in ALS, manifested by spontaneous laughing or crying unrelated to the situation. Despite the pattern of onset, however, the eventual course of the illness is similar in most patients, with an unremitting spread of weakness to other muscle groups leading to total paralysis of spinal musculature and muscles innervated by the cranial nerves. Death is usually related to respiratory failure.

In a longitudinal study using monthly questionnaires, direct patient interviews, record reviews, physician interviews, and family member interviews, Brooks and colleagues followed 702 patients with ALS. Their findings suggest that spread of neuronal degeneration occurred more quickly to adjacent areas than to noncontiguous areas. The spread to adjacent areas was more rapid at the brain stem, quickly to adjacent areas than to noncontiguous areas. The rate of progression seems to be consistent for each patient but varies considerably among patients. Patients with an initial onset of bulbar weakness (dysarthria, dysphagia) and respiratory weakness (dyspnea) tend to have a more rapid progression to death than patients whose weakness begins in the distal extremities. Death usually follows within 2 to 4 years after diagnosis, with a small number of patients living for 15 to 20 years.

Years of survival after diagnosis may change as drug therapies are developed. In addition, increasing numbers of patients are electing to prolong life with home-based mechanical ventilation as opposed to palliative or comfort care only.

**Medical Prognosis**

In almost all cases ALS progresses relentlessly and leads to death from respiratory failure. The rate of progression seems to be consistent for each patient but varies considerably among patients. Patients with an initial onset of bulbar weakness (dysarthria, dysphagia) and respiratory weakness (dyspnea) tend to have a more rapid progression to death than patients whose weakness begins in the distal extremities. Death usually follows within 2 to 4 years after diagnosis, with a small number of patients living for 15 to 20 years.

**Medical Management**

ALS has no known cure and minimal effective disease-slowing treatments. Mitchell and Borasio have created a table (see Table 2 in their study) that summarizes the results of trials of the many putative ALS-modifying pharmaceuticals. Only riluzole has been approved for treatment of ALS. Riluzole provides very modest improvement over a placebo in both bulbar and limb function, but not in actual strength of muscles. The drug extended lifespan an average of 2 to 3 months. The side effects were minimal in some studies, but fatigue and weakness have been noted in 26% and 18% of patients taking riluzole compared with a placebo.

The popular press has reported on nutritional cures for ALS, including regular use of vitamin E. However, Orrell and colleagues found insufficient evidence to support clinical use of vitamin E supplements in ALS as an additive to riluzole treatment or as adjunctive therapy, although no apparent contraindication was found to taking the supplement. Other nutritional and nonpharmaceutical supplements have had some success in animal models of ALS, but this has not yet been confirmed in humans.

Cannabis has been studied for its effect on spasticity in patients with multiple sclerosis and spinal cord injury. In a study of 131 people with ALS, 13 used cannabis, with reports of reduction in spasticity, pain, and depression. Because of the apparent hopelessness of the diagnosis, many physicians,
## BOX 17-1 AMYOTROPHIC LATERAL SCLEROSIS SEVERITY SCALE: LOWER EXTREMITY, UPPER EXTREMITY, SPEECH, SWALLOWING

### LOWER EXTREMITIES (WALKING)

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>Normal ambulation</td>
<td>Patient denies any weakness or fatigue; examination reveals no abnormality.</td>
</tr>
<tr>
<td>9</td>
<td>Fatigue suspected</td>
<td>Patient experiences sense of weakness or fatigue in lower extremities during exertion.</td>
</tr>
<tr>
<td>8</td>
<td>Difficulty with uneven terrain</td>
<td>Difficulty and fatigue when walking long distances, climbing stairs, and walking over uneven ground (even thick carpet).</td>
</tr>
<tr>
<td>7</td>
<td>Observed changes in gait</td>
<td>Noticeable change in gait; pulls on railings when climbing stairs; may use leg brace.</td>
</tr>
<tr>
<td>6</td>
<td>Walks with mechanical device</td>
<td>Needs or uses cane, walker, or assistant to walk; probably uses wheelchair away from home.</td>
</tr>
<tr>
<td>5</td>
<td>Walks with mechanical device and assistant</td>
<td>Does not attempt to walk without attendant; ambulation limited to less than 50 ft; avoids stairs.</td>
</tr>
<tr>
<td>4</td>
<td>Able to support</td>
<td>At best, can shuffle a few steps with the help of an attendant for transfers.</td>
</tr>
<tr>
<td>3</td>
<td>Purposeful leg movements</td>
<td>Unable to take steps but can position legs to assist attendant in transfers; moves legs purposefully to maintain mobility in bed.</td>
</tr>
<tr>
<td>2</td>
<td>Minimal movement</td>
<td>Minimal movement of one or both legs; cannot reposition legs independently.</td>
</tr>
<tr>
<td>1</td>
<td>Paralysis</td>
<td>Flaccid paralysis; cannot move lower extremities (except, perhaps, to close inspection).</td>
</tr>
</tbody>
</table>

### UPPER EXTREMITIES (DRESSING AND HYGIENE)

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>Normal function</td>
<td>Patient denies any weakness or unusual fatigue of upper extremities; examination demonstrates no abnormality.</td>
</tr>
<tr>
<td>9</td>
<td>Suspected fatigue</td>
<td>Patient experiences sense of fatigue in upper extremities during exertion; cannot sustain work for as long as normal; atrophy not evident on examination.</td>
</tr>
<tr>
<td>8</td>
<td>Slow self-care</td>
<td>Dressing and hygiene performed more slowly than usual.</td>
</tr>
<tr>
<td>7</td>
<td>Effortful self-care performance</td>
<td>Requires significantly more time (usually double or more) and effort to accomplish self-care; weakness is apparent on examination.</td>
</tr>
<tr>
<td>6</td>
<td>Mostly independent</td>
<td>Handles most aspects of dressing and hygiene alone; adapts by resting, modifying (e.g., use of electric razor), or avoiding some tasks; requires assistance for fine motor tasks (e.g., buttons, ties).</td>
</tr>
<tr>
<td>5</td>
<td>Partial independence</td>
<td>Handles some aspects of dressing and hygiene alone; however, routinely requires assistance for many tasks such as applying makeup, combing, and shaving.</td>
</tr>
<tr>
<td>4</td>
<td>Attendant assists patient</td>
<td>Attendant must be present for dressing and hygiene; patient performs the majority of each task with the assistance of the attendant.</td>
</tr>
<tr>
<td>3</td>
<td>Patient assists attendant</td>
<td>The attendant directs the patient for almost all tasks; the patient moves in a purposeful manner to assist the attendant; does not initiate self-care.</td>
</tr>
<tr>
<td>2</td>
<td>Minimal movement</td>
<td>Minimal movement of one or both arms; cannot reposition arms.</td>
</tr>
<tr>
<td>1</td>
<td>Paralysis</td>
<td>Flaccid paralysis; unable to move upper extremities (except, perhaps, to close inspection).</td>
</tr>
</tbody>
</table>

### SPEECH

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>Normal speech</td>
<td>Patient denies any difficulty speaking; examination demonstrates no abnormality.</td>
</tr>
<tr>
<td>9</td>
<td>Nominal speech abnormalities</td>
<td>Only the patient or spouse notices speech has changed; maintains normal rate and volume.</td>
</tr>
<tr>
<td>8</td>
<td>Perceived speech changes</td>
<td>Speech changes are noted by others, especially during fatigue or stress; rate of speech remains essentially normal.</td>
</tr>
<tr>
<td>7</td>
<td>Obvious speech abnormalities</td>
<td>Speech is consistently impaired; rate, articulation, and resonance are affected; remains easily understood.</td>
</tr>
</tbody>
</table>
**Chapter 17**

**Neuromuscular Diseases**

**Box 17-1**

**Amyotrophic Lateral Sclerosis Severity Scale: Lower Extremity, Upper Extremity, Speech, Swallowing—cont’d**

<table>
<thead>
<tr>
<th>Intelligible with Repeating</th>
<th>Rate is much slower, repeats specific words in adverse listening situation; does not limit complexity or length of messages.</th>
</tr>
</thead>
<tbody>
<tr>
<td>6  Repeats message on occasion</td>
<td></td>
</tr>
<tr>
<td>5  Frequent repeating required</td>
<td>Speech is slow and labored; extensive repetition or a “translator” is commonly used; patient probably limits the complexity or length of messages.</td>
</tr>
</tbody>
</table>

**Speech Combined with Nonverbal Communication**

<table>
<thead>
<tr>
<th>4  Speech plus nonverbal communication</th>
<th>Speech is used in response to questions; intelligibility problems need to be resolved by writing or a spokesperson.</th>
</tr>
</thead>
<tbody>
<tr>
<td>3  Limits speech to one-word responses</td>
<td>Vocalizes one-word responses beyond yes and no; otherwise writes or uses a spokesperson; initiates communication nonverbally.</td>
</tr>
</tbody>
</table>

**Loss of Useful Speech**

<table>
<thead>
<tr>
<th>2  Vocalizes for emotional expression</th>
<th>Uses vocal inflection to express emotion, affirmation, and negation.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1  Nonvocal</td>
<td>Vocalization is effortless, limited in duration, and rarely attempted; may vocalize for crying or pain.</td>
</tr>
<tr>
<td>X  Tracheostomy</td>
<td></td>
</tr>
</tbody>
</table>

**Swallowing**

**Normal Eating Habits**

| 10 Normal swallowing                | Patient denies any difficulty chewing or swallowing; examination demonstrates no abnormality. |
| 9  Nominal abnormality              | Only patient notices slight indicators such as food lodging in the recesses of the mouth or sticking in the throat. |

**Early Eating Problems**

| 8  Minor swallowing problems       | Reports some swallowing difficulties; maintains essentially a regular diet; isolated choking episodes. |
| 7  Prolonged times, smaller bite size | Meal time has significantly increased and smaller bite sizes are necessary; must concentrate on swallowing thin liquids. |

**Dietary Consistency Changes**

| 6  Soft diet                        | Diet is limited primarily to soft foods; requires some special meal preparation. |
| 5  Liquefied diet                   | Oral intake adequate; nutrition limited primarily to liquefied diet; adequate thin liquid intake usually a problem; may force self to eat. |

**Needs Tube Feeding**

| 4  Supplemental tube feedings       | Oral intake alone no longer adequate; patient uses or needs a tube to supplement intake; patient continues to take significant (greater than 50%) nutrition orally. |
| 3  Tube feeding with occasional oral nutrition | Primary nutrition and hydration accomplished by tube; receives less than 50% of nutrition orally. |

**No Oral Feeding**

| 2  Secretions managed with aspirator and/or medications | Cannot safely manage any oral intake; secretions managed with aspirator and/or medications; swallows reflexively. |
| 1  Aspiration of secretions          | Secretions cannot be managed noninvasively; rarely swallows. |


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especially those not associated with major medical centers having neuromuscular disease units, do not refer patients with ALS for services, yet few primary care physicians or neurologists have extensive experience in the care of patients and families coping with ALS because of the low incidence of the disease. Yet, referral of patients with ALS to a multidisciplinary clinic typically extends the patient’s lifespan, especially patients with bulbar onset of ALS.25,37

**Muscle Spasms and Pain**

Some patients experience muscle cramps and spasms related to upper motor neuron pathology, and up to 73% of patients complain of pain, typically in the later stages.24 Although most spasms can be relieved with stretching or increased movement, some patients require medications such as quinine or baclofen to relieve symptoms (see Chapter 36 for information on drug therapies). In a review of studies on the treatment of spasticity in ALS, Ashworth and colleagues78 found only one randomized study addressing spasticity: a moderate-endurance exercise regimen decreased spasticity at 3 months after initiation of the program. Stretching and massage may prove helpful for nocturnal muscle cramps.25 Kesiktas and colleagues39 report that in a controlled study of spasticity in patients after spinal cord injury, adding hydrotherapy to a program of medication and exercise decreased severity of spasms and decreased the amount of medication required. A similar response could be hypothesized in patients with ALS. In addition to muscle spasms, patients
report nonspecific aching and muscle soreness, probably related to immobility and trauma to paralyzed muscles during caregiving procedures. However, many patients do not receive adequate pain medication, or the pain is not controlled by the medication taken. A Cochrane review in 2008 found no randomized or quasi-randomized controlled trials of drug therapy for pain in ALS, although several case series reported the use of acetaminophen, nonsteroidal antiinflammatory drugs (NSAIDs), or opioids. Careful administration of medications such as baclofen, tizanidine, dantrolene sodium, and diazepam is useful for some patients with spasticity. Because each has a different action and side effects, the medications may have to be adjusted to find the right dosage and combination. In some patients with severe cramping, botulinum toxin injections might be helpful, but they must be carefully administered to prevent further weakness. Because many patients have compromised respiratory function, the physician must take great care when prescribing pain medication, especially opiates, which are often used when antispasmodics or antiinflammatory pain medications no longer work. Patients should be instructed to keep a daily recording log of the effectiveness of the medication so that the dosage can be adjusted if necessary.

**Dysphagia**

Dysphagia, a difficulty swallowing liquids, foods, or saliva, accounts for considerable misery in the patient with advanced ALS, and it must be dealt with aggressively. Patients with dysphagia have both nutritional and swallowing problems associated with weakness of the lips, tongue, palate, and mastication muscles. As the progressive loss of swallowing develops, patients are also at extreme risk for aspiration. Most patients with dysphagia also have severe problems with management of their saliva (sialorrhea). If a patient has difficulty transporting saliva back to the oropharynx for swallowing, choking and drooling are common. This condition is disconcerting to the affected person, who must constantly wipe the mouth or have someone do it for him or her.

In addition, secretions are often thickened because of dehydration. With pooling of the thickened saliva, the possibility of aspiration is increased. Viscosity of saliva can best be treated by hydration and, in some cases, pharmaceuticals. Drugs, such as decongestants, antidepressant drugs with anticholinergic side effects, and atropine-type drugs, can help control the amount of saliva, provided the patient is well hydrated. In extreme cases, various surgical procedures such as ligation of the salivary gland ducts, severing the parasympathetic supply to the salivary glands, and excision of the salivary glands have been used effectively. Newer treatments to decrease excessive secretions are radiotherapy and botulinum A toxin injections into salivary glands.

Although dietary treatment is not known to be effective in changing the course of the disease, a nutritious diet to meet caloric, fluid, vitamin, and mineral needs must be maintained. Seventy-three percent of patients with ALS have difficulty bringing food to the mouth, making them dependent on others for their dietary needs. Because of the time it takes to be fed, many patients decrease their intake. All patients with dysphagia should be referred for a dietary consultation to determine the choice and progression of solid and liquid foods and supplements. Appel and colleagues describe nutritional plans to maintain nutrition and hydration in patients with motor neuron diseases. Patients with bulbar symptoms and severe dysphagia who are no longer able to consume nutrients orally because of motor control problems and recurrent aspiration may need a percutaneous endoscopic gastrostomy (PEG) for feeding, depending on the patient’s wishes for long-term care. Some evidence exists that the PEG should be performed early in the disease process to prevent severe weight loss and aspiration. Although a PEG does not appreciably lengthen survival time, patients may have less fear of choking or aspiration. Receiving nourishment from a PEG does not prevent the person from taking food orally if desired.

**Dysarthria**

Dysarthria, impairment in speech production, is the result of abnormal function of the muscles and nerves associated with coordinated functions of the tongue and lips, larynx, soft palate, and respiratory system. Speech impairments are the initial symptom in most patients with bulbar involvement. Speech intelligibility is compromised by hypernasality, abnormalities of speed and cadence of speech, and reduced vocal volume. Speech is further compromised by inadequate breath volumes for normal phrasing. A possible option to help patients with severe hypernasality is a palatal lift prosthesis to augment velopharyngeal function. Because little can be done medically to delay the loss of speech control, early referral to a speech therapist is essential. Numerous augmentative and alternative communication systems are now available, the simplest being voice amplification systems or homemade point boards and computer-based head or eye tracking text-to-speech systems that can be modified as the patient status changes. The type of communication system should be chosen with awareness of the patient-caregiver environment.

**Respiratory Management**

Progressive respiratory failure is the primary cause of death in ALS patients. Respiratory failure is related to primary diaphragmatic, intercostal, and accessory respiratory muscle weakness. Respiratory failure should be anticipated and discussed early following the diagnosis of ALS so that patients and their caregivers can express their wishes and develop an advanced directive for care in the terminal phase of the disease.

Physiological tests used to indicate respiratory dysfunction include vital capacity, sniff nasal pressure, and nocturnal oximetry. Clinical signs of increased respiratory dysfunction are dyspnea with exertion or lying supine; hypventilation; weak or ineffective cough; increased use of auxiliary respiratory muscles; tachycardia (also a sign of pulmonary infection with fever and tachypnea); changes in sleep pattern; daytime sleepiness and concentration problems; mood changes; and morning headaches.

In early stages of patient care, physical therapists (PTs) may help manage respiratory dysfunction by providing postural drainage with cough facilitation (suctioning if necessary), especially during acute respiratory illnesses. The patient and care providers should also be taught breathing exercises, chest stretching, and incentive spirometry techniques, as well as postural drainage techniques if the caregivers are prepared to provide such support.
breathing exercises consisting of resisted inspiratory muscle training can facilitate functional respiration, even practicing resisted breathing for 10 minutes three times a day has been shown to result in improved function. An assessment of the home environment is imperative to identify sleeping positions and energy conservation techniques that can be incorporated into the patient’s daily life.

As respiratory symptoms increase, oxygen at 2 L/min or less can be used intermittently at home. When hypventilation with a decline in oxygen saturation becomes common during sleep, resulting in morning confusion and irritability, patients have the option to initiate noninvasive, positive-pressure ventilation (NIV) such as bilevel positive airway pressure (BiPAP). BiPAP, which provides greater inspiratory pressure than expiratory pressure to decrease the effort of breathing, can be administered by either mask or contoured nasal delivery systems. Some evidence indicates that early use of NIV can increase survival time by several months and increase quality of life. When a patient can no longer benefit from NIV, a decision must be made about initiating ventilation by tracheostomy or palliative care. (See also Miller and colleagues for an excellent discussion of practice parameters in the decision-making process related to ventilatory support.) Although in the initial stages of ALS most patients indicate they would not want prolonged respirator dependence at home, patients may change their minds as they adapt to the disease restrictions. A small study of patients who started tracheostomy intermittent positive-pressure ventilation (TIPPV) demonstrated increased long-term survival (2 to 64 months). In another series of 70 patients on long-term TIPPV, 50% of the patients were living after 5 years; however, 11.4% of these patients had entered a ‘locked-in state in which they were unable to communicate in any manner.’ Decisions about long-term respirator use should be made by the patient and involved family members or partners, with input from the interdisciplinary team caring for the patient. Discussions of preferred long-term care options should be revisited as the patient’s condition changes. If a patient decides that home ventilation is a reasonable option, those involved in the decision should visit another physician or therapist who values control and an active lifestyle experiences. Physicians and health care workers who work with the patient and family must be aware of their own feelings and beliefs about prolonging life. For example, a healthy physician or therapist who values control and an active lifestyle may envision a life on a ventilator as intolerable and pass that value on to the patient, who may or may not have the same needs. The patient’s decision, or change in decision, must be respected by the medical team involved in care. In medical centers that use a team approach, patients and families may find support by meeting with counselors or peers with ALS who are making or have made decisions about long-term ventilator care.

Therapeutic Management of Movement Dysfunction Associated with ALS

Perhaps because of the multitude of issues to consider when managing the impairments and limitations associated with ALS, evidence suggests that patients treated by a specialized ALS multidisciplinary team fare better than do those treated by single-source providers, or in general neurology clinics. A Cochrane review of the evidence for multidisciplinary care advantages in this population concluded that the evidence is of low quality, so far, with no controlled trials identified. Whether administered through an ALS-specific team or not, therapeutic management will necessitate examination of the patient’s current status, evaluation of the deficits in relation to patient preferences and needs, and establishment of a plan based on mutually determined and realistic goals. The rate of the patient’s disease progression, the areas and extent of involvement, and the stage of illness must be considered. A patient at the initial stages will have different needs than a patient at later stages who has chosen NIV or tracheostomy ventilation that may extend life span at a markedly reduced mobility level. The goal at all stages is to optimize health and increase the quality of life. With guidance and environmental adaptations, patients with slowly progressing weakness may be able to continue many of their ADLs for an extended number of years. In the final stages of the disease, when the patient is bedridden, programs to increase strength or endurance are not appropriate, and interventions such as stretching may not effectively control contracture development. However, patients may still benefit from positioning and range-of-motion (ROM) exercises to decrease muscle and joint pain related to immobility. The prescription of assistive devices and training of caregivers will also be needed. The efficacy of therapeutic interventions will be related to the timing of interventions, the motivation and persistence of the patient in carrying out the program, and support from family members or caregivers. Objective documentation of outcome measures will help justify the usefulness of therapeutic interventions at all stages of this disease.
Examination

The extent of the therapeutic examination of a patient with ALS will depend on whether the therapist is working as a member of a rehabilitative team or as an independent or clinic-based therapist receiving a referral to evaluate and treat. PTs and OTs working as team members may have a more circumscribed role related to gross motor function and ADLs, with other consultants focusing on bulbar, respiratory, and environmental adjustments. The therapist working in a facility without a neuromuscular disease clinic or in a community or rural environment, however, should be aware of the need to carry out a broad-based assessment. In addition to the standard neuromuscular, musculoskeletal, and functional-level examinations, the therapist should also evaluate the patient’s stated or observed functional problems relative to bulbar and respiratory impairments, environmental blocks to independence, and caregiving demands.

If possible, before the patient’s initial visit, the therapist should contact the patient and request that he or she keep an activity log for several days. If an early contact is not possible, the therapist can assign that task during the initial session. The log should include 15-minute time increments in which the patient or caregiver can record what she or he was doing during a specific period. The log should also indicate whether the patient was experiencing fatigue or pain during the activity and how the patient perceived her or his respiratory status. An example of an activity log and how it is used is shown in Figure 17-2. The sense of fatigue with repetitive muscle activity or functional activity should be specifically tracked by the patient.

Weakness will be the primary deficit, with other problems following depending on the location of strength loss. Muscle weakness and the experience of fatigue may be independent measures of ALS pathology, however. Although weakness may affect balance during gait, patients with ALS have not shown deficits in postural control during quiet stance despite significant paresis or tone changes, possibly because sensation is relatively preserved.

The therapist’s examination will vary depending on the patient’s situation, however, a typical initial assessment may include the following:

- Review of the patient’s medical and activity records, especially time since diagnosis, time course of disease progression to date, current medications, concurrent medical issues, current activities and participation and tolerance for them.
- History should focus on current and recent activities and participation signifying patient’s lifestyle, ADL tasks, hobbies or interests, and work focus; primary complaints, including weakness, fatigue, pain, respiratory status, safety, or speech and swallowing issues; psychosocial support issues (family, caregivers, and agencies); patient’s and family members’ understanding of ALS and the likely progression and prognosis; and patient’s current concerns and goals.
- Screening for multisystem involvement should include checking vital signs at rest, skin integrity, bony abnormalities, sensory integrity, communication ability, and ability to follow multistep commands. More extensive examination of systems showing deficits may be indicated, or the patient may be referred to appropriate health care professionals.
- Baseline testing of muscle strength (manual muscle testing [MMT] or electronic handheld dynamometer testing if standards are clear and can be replicated), ROM, spasticity, and endurance; documentation of any areas of atrophy.
- Assessment of functional activity level (using a standardized test or assessment tool whenever possible) to include, as appropriate: transfers, gait, upper-extremity function, postural control, and assistive devices; suggested tools include the ALS functional rating scale (ALSFRS), the ALS severity scale (ALSSS), timed walk test, or Purdue Pegboard.
- Documentation of pain (type, site, and intensity; use body chart and subjective pain scale); identify what makes pain worse or better.
- Assessment of bulbar and respiratory function. For an in-depth evaluation of bulbar function, the patient should be referred to an ear, nose, and throat clinic or communications disorders clinic unless full evaluation is available in a comprehensive ALS clinic. (See Table 17-1 for bulbar and respiratory evaluation suggestions.)
- Environmental assessment with a focus on energy conservation and safety at current and future functional capabilities.

Brinkmann and colleagues identify standards for assessment of patients with ALS in clinical trials. The review and description of standardized methods for performing recommended tests and measurements is extremely valuable for any therapist assessing and treating patients with ALS.

In evaluating the results of the examination, the therapist should synthesize data to define the following, all of which are necessary for developing goals with the patient.

- Rate of the patient’s disease progression
- Distribution of weakness and spasticity, respiratory factors leading to hypoxemia, and ease of fatigability and bulbar involvement
- Phase of the disease
- Any preexisting impairments and/or activity limitations (see Chapter 8)

Goals of Therapeutic Intervention

Intervention goals and the recommended exercise and activity program designed by PTs or OTs must be based on the patient’s personal goals. Goals are often a difficult area for therapists to discuss with the patient because the disease is progressive despite intervention. Patients, therapists, and physicians commonly assume that because nothing can be done to “cure” the disease, not making additional demands on a patient who is already coping with daily loss is somehow kinder. Some believe that exercise programs may create false hopes that exercise will delay progression. Others believe that exercise will hasten progression. The literature on rehabilitation in neuromuscular disorders, however, suggests that patients with ALS can benefit from carefully designed exercise and activity programs. Active participation in determining goals for therapy can provide the patient and the family with some sense of control over a difficult situation.

The general, broad goals for both patient and therapist are related to maintaining maximal independence in daily living and a positive quality of life for as long as possible.
DAILY ACTIVITY LOG

Instructions: 1) In column I write in what you are doing during the 24 hour period. You may draw a line or an arrow to indicate when the activity occurs for more than one 15 minute time period.

2) In column II indicate whether you are lying down, sitting, standing, or moving actively (walking, etc.) during the activity.

3) In column III on a 10 point scale, indicate how fatigued you feel while performing the activity (No fatigue = 0, extreme fatigue = 10.)

4) In column IV indicate where you feel pain if any and score the intensity on a 10 point scale (No pain = 0, extreme pain = 10.)

Try to fill out your log three or four times a day so you don’t forget what you have been doing. An example is shown below.

<table>
<thead>
<tr>
<th>Time</th>
<th>I</th>
<th>II</th>
<th>III Fatigue level</th>
<th>IV Pain</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>What are you doing?</td>
<td>What position are you in</td>
<td>(lying, sitting, standing, moving)</td>
<td>Location</td>
</tr>
<tr>
<td>5:30 AM</td>
<td>Sleep</td>
<td>Lying</td>
<td>0</td>
<td>neck</td>
</tr>
<tr>
<td>6:00</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30</td>
<td>Breakfast</td>
<td>Sitting</td>
<td>3</td>
<td>neck</td>
</tr>
<tr>
<td>45</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7:00</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>Reading</td>
<td>Sitting</td>
<td>3</td>
<td>neck</td>
</tr>
<tr>
<td>30</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>45</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8:00</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>45</td>
<td>Walk</td>
<td>Standing, Walking</td>
<td>4</td>
<td>neck</td>
</tr>
<tr>
<td>9:00</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30</td>
<td>Nap</td>
<td>Lying</td>
<td>2</td>
<td>neck</td>
</tr>
<tr>
<td>45</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10:00</td>
<td>Reading/TV</td>
<td>Sitting</td>
<td>4</td>
<td>neck</td>
</tr>
<tr>
<td>15</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>45</td>
<td>Walk</td>
<td>Standing, Walking</td>
<td>5</td>
<td>hips</td>
</tr>
<tr>
<td>11:00</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Figure 17-2** Example of a log for monitoring activity level of patients with amyotrophic lateral sclerosis.
### TABLE 17-1  ■ COMMON PHYSICAL FINDINGS IN BULBAR AMYOTROPHIC LATERAL SCLEROSIS

<table>
<thead>
<tr>
<th>ANATOMICAL SITE</th>
<th>INNERVATION</th>
<th>METHOD OF EVALUATION</th>
<th>PROGRESSION OF FINDINGS</th>
<th>PROGRESSION OF SYMPTOMS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>GROUP I</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tongue</td>
<td>XII</td>
<td>Inspect for fasciculations at rest</td>
<td>Fasciculations evident</td>
<td>Dysarthria (disturbance of lingual-alveolar consonants t, d, l, and so on)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Range of motion</td>
<td>Slow, incomplete lateral movements</td>
<td>Inability to clear buccal sulcus of food</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Protrusion</td>
<td>Unable to protrude beyond lips</td>
<td>Marked dysarthria (slow rate and slurring of consonants)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Perform rapid lateral motion</td>
<td>Unable to protrude beyond incisors</td>
<td>Oral transport difficulties</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Atrophy evident</td>
<td>Dietary changes</td>
</tr>
<tr>
<td>Lips</td>
<td>VII</td>
<td>Suck on gloved finger</td>
<td>Lack of suction</td>
<td>Inability to whistle</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Smile or curl lips over teeth</td>
<td>Inability to complete a seal</td>
<td>Inability to use a straw</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hold seal and blow out cheeks</td>
<td>Inability to purse lips</td>
<td>Dysarthria (loss of bilabial consonants p and b)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Atrophy evident</td>
<td>Drooling</td>
</tr>
<tr>
<td><strong>GROUP 2</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Palate</td>
<td>V, X, XI</td>
<td>Visual examination during phonation and stimulation of gag</td>
<td>Unsustained or slow palatal elevation</td>
<td>Dysarthria (hypernasal speech)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Puff out cheeks to check for nasal air leak (hold lips closed if necessary)</td>
<td>Soft palate fails to reach Passavant ridge</td>
<td>Inability to use a straw</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Absence of palatal movement</td>
<td>Nasal air emission during speech</td>
</tr>
<tr>
<td>Muscles of mastication</td>
<td>V</td>
<td>Palpate during bite</td>
<td>Noticeable wasting</td>
<td>Nasopharyngeal reflex on swallowing</td>
</tr>
<tr>
<td>Masseter, temporalis</td>
<td></td>
<td>Visual inspection for wasting</td>
<td>Unable to palpate contraction</td>
<td>Chewing fatigue</td>
</tr>
<tr>
<td>Pterygoids</td>
<td></td>
<td>Move jaw from side to side</td>
<td>No observable lateral jaw movement</td>
<td>Elimination of specific, tough foods from diet</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Dietary changes (soft foods and liquids)</td>
</tr>
<tr>
<td><strong>GROUP 3</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neck and shoulder</td>
<td>XI</td>
<td>Hold arm in coronal plane, hand externally rotated, as patient elevates arm against resistance while the trapezius is palpated</td>
<td>Progressive inability to raise the arm (often asymmetrical weakness)</td>
<td>Inability to comb hair</td>
</tr>
<tr>
<td>Trapezius</td>
<td></td>
<td></td>
<td></td>
<td>Inability to perform facial grooming</td>
</tr>
<tr>
<td>Sternocleidomastoid or mounted head support</td>
<td></td>
<td>Turn the head against resistance applied to opposite side of patient’s chin</td>
<td>Progressive weakness in turning the head against resistance (often asymmetrical)</td>
<td>Inability to lift head when supine</td>
</tr>
<tr>
<td>Vocal cords</td>
<td>X</td>
<td>Mirror or fiberoptic laryngoscopy</td>
<td>Progressive loss of abduction of vocal cords: mild abductor weakness, near-midline paralysis</td>
<td>Inability to support head while sitting; wears neck collar, has weakness</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Paradoxical vocal cord movement</td>
<td>Strangled or strangled voice</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Short of breath (stridor usually not present because of impaired respiratory function)</td>
</tr>
</tbody>
</table>
TABLE 17-1 ■ COMMON PHYSICAL FINDINGS IN BULBAR AMYOTROPHIC LATERAL SCLEROSIS—cont’d

<table>
<thead>
<tr>
<th>ANATOMICAL SITE</th>
<th>INNERVATION</th>
<th>METHOD OF EVALUATION</th>
<th>PROGRESSION OF FINDINGS</th>
<th>PROGRESSION OF SYMPTOMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>GROUP 4</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Extraocular muscles</td>
<td>III, IV, VI</td>
<td>Assessment of extraocular movements</td>
<td>Limitation of extraocular movement</td>
<td>Limitation of gaze</td>
</tr>
<tr>
<td>Respiratory group</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diaphragm</td>
<td>C3-5</td>
<td>Pulmonary function test or handheld respirometer for vital capacity</td>
<td>Diminishing vital capacity: 1.5-2.0 L</td>
<td>Shortness of breath during exertion if patient has remained active</td>
</tr>
<tr>
<td>Intercostal</td>
<td>C7-L3</td>
<td>Cough</td>
<td>1.0-1.5 L</td>
<td>Weak cough</td>
</tr>
<tr>
<td>Accessory muscles of respiration</td>
<td>VII, XI, XII, C5-8</td>
<td>Sustain a vowel Blow against a tissue</td>
<td>0.5-1.0 L</td>
<td>Change in speech phrasing (5-10 syllables per breath)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Speech produced in syllable- by-syllable fashion (if vocal)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Shortness of breath on swallowing</td>
</tr>
</tbody>
</table>


More specific therapeutic goals are (1) maintenance of mobility and independent functioning, to include safe mobility for patient and caregiver; (2) maintenance of maximal muscle strength and endurance within limits imposed by ALS; (3) prevention and minimization of secondary consequences of the disease, such as contractures, thrombophlebitis, decubitus ulcers, and respiratory infections; (4) management of energy conservation techniques and respiratory comfort; (5) determination of adaptive equipment needs to include mobility, self-help and feeding devices, augmentative communication units, and hygiene equipment that supports both patient and caregiver; and (6) eliminating or preventing pain.

**Therapeutic Considerations**

To prevent more rapid functional loss than expected from the natural history of the disease, both the patient and therapist must delicately balance the level of activity between the extremes of inadequate exercise and excessive exercise. Exercise has been recommended for the general public for its many benefits. Inadequate exercise may result in loss of strength and endurance from disuse, as well as secondary problems such as loss of ROM, muscle cramping, and pain. Excessive exercise may result in excessive fatigue and consequent inability to perform ADLs during recovery periods. Overuse injury with excessive strengthening exercise may also lead to unnecessary pain and loss of strength. The next two sections review the evidence for the optimum amount of activity or exercise.

**Disuse Atrophy.** Because ALS is a disease of older adults, patients may not have maintained their aerobic fitness or muscle strength before the onset of their neuromuscular problem. Newly diagnosed patients also commonly report that they had markedly decreased their activity level in the months before diagnosis because of a sense of fatigue or increasing clumsiness from increasing weakness. If the patient had led a sedentary lifestyle before diagnosis, the additional decrease in activity level after the onset of ALS can lead quickly to marked cardiovascular deconditioning and disuse weakness. The disuse weakness lowers muscle force production and reduces muscle endurance.

**Exercise or Overwork Damage.** Anecdotal evidence that muscle activity or overwork exercise can lead to a loss of muscle strength has been reported since the poliomyelitis epidemic of the 1940s and 1950s. During that epidemic, physicians and therapists noted that patients with poor- and fair-grade muscles who exercised repeatedly or with heavy resistance after reinervation often lost the ability to contract the muscle at all (see Chapter 35). Controlled testing of this observation suggests that overwork damage occurs in mostly denervated muscles, not in all muscles. Reitsma noted that vigorous exercise damaged muscles in rats if less than one third of motor units were functional. If more than one third of the motor units remained, exercise led to hypertrophy. An additional mechanism of potential overwork damage is inhibition of the collateral sprouting of intact axons to innervate “orphaned” muscle fibers when other axons degenerate. Yuen and Olney provided evidence that collateral sprouting of intact axons can partially reinnervate orphaned muscle fibers in ALS. In a rat model, highly intensive activity reduced the ability of adjacent axons to sprout after fewer than 20% of intact motor units remained. In contrast, vigorous exercise in a mouse model had no adverse effect on the course of ALS. Lui and Byl reviewed the literature reporting exercise effects in animal models of ALS and calculated an effective size of 1.39 (where numbers over 0.8 are considered large) in favor of exercise. The few negative effects they noted were associated with either very-high–intensity exercise or a slow rate of exercise (slower than usual activity for animals when unrestricted in activity). In addition to generic overwork, evidence exists that repeated maximal eccentric contractions may specifically damage even normal muscle fibers, resulting in muscle weakness of several weeks’ duration. Although
normal muscle eventually adapts to repeated eccentric exercise, whether the reparative effect is possible in patients with neuromuscular diseases is uncertain. Aboussouan reviews some of the specific mechanisms of exercise intolerance in neuromuscular diseases, including mitochondrial dysfunction, abnormal muscle metabolism, impaired muscle activation, and central activation failure.

Many researchers have expressed concern about the possible relation between high-resistance exercise and muscle fiber degeneration in humans with motor neuron disease. Because of the concerns about damage from stressing substantially denervated muscles, Sinaki and Mulder published recommendations in 1978 that patients with ALS not engage in any vigorous exercise and focus instead on exercise associated with walking and daily activities. On the other hand, McCrate and Kaspar review the possible mechanisms by which exercise protects nerves from more rapid degeneration. Evidence regarding the positive benefits of exercise in ALS has been accumulating, with fewer adverse effects than some expected.

Sanjak and colleagues reported that muscle damage does not necessarily result from resistance exercise testing or training, although fatigue occurs more easily during both anaerobic and aerobic exercise. Milner-Brown and Miller found that mild progressive resistance exercise was helpful in neuromuscular disorders if the patient had muscle strength in the good (4/5) to normal (5/5) range. They determined that patients should begin their exercise program early because strength training of muscles with less than 10% of normal function was generally not effective. Aitkens and colleagues noted strength gains of 4% to 20% without deleterious effects after a 12-week program of moderate-resistance (30% of maximum isometric force) exercises in patients with slowly progressive neuromuscular diseases. Kilmer and colleagues, in the same population, found no additional advantage to high-resistance training (12 weeks of exercise using the maximum isometric force the individual was able to lift 12 times) and noted evidence of overwork in some subjects. In a case report of a patient with ALS, strengthening 6 days a week for 10 weeks with proprioceptive neuromuscular facilitation (PNF) patterns using maximal resistance applied manually or with tubing resulted in strengthening of 14 muscle groups out of 18 with no adverse effects. Aksu and colleagues compared a supervised versus home exercise protocol in 26 ambulatory ALS patients. They noted that supervised breathing exercises, stretching, manually applied resistance exercise with PNF, and functional mobility training 3 days a week for 8 weeks resulted in small gains in function in the first 4 weeks and a slower decline over the subsequent 10 months compared with home-based breathing, stretching, and active ROM exercises. The groups were not randomly allocated but were not significantly different in the measured variables at baseline. In a randomized controlled trial, Drory and colleagues assigned 25 patients with ALS to a group continuing their normal daily activities or a group participating in a moderate daily program of exercise individualized for each patient. The primary exercise focus was to have muscles of the trunk and limbs work against “modest” loads while undergoing significant shortening (not lengthening or eccentric contractions). The exercises were completed twice daily for 15 minutes at home with phone contact by the treating therapist every 14 days. Data were evaluated for 3 and 6 months after initial assessment. All patients showed continued disease progression; however, in all cases, at the 6-month assessment patients who exercised showed positive effects in maintenance of muscle strength, less fatigue, less spasticity, less pain, and higher functional ratings. In another randomized controlled trial, moderate load and moderate-intensity resistance exercises prescribed individually to patients with ALS in the early stages resulted in significantly less decline in function, small improvements in strength, and no reported adverse effects, compared with patients who performed stretching exercises alone. A Cochrane review designated the quality of the Drory and colleagues (2001) study as “fair” and the Dal Bello-Haas and colleagues study as “adequate.” Table 17-2 summarizes some of the studies of strength training in neuromuscular diseases.

Fewer researchers have considered endurance in neuromuscular disorders. Sanjak and colleagues noted that exercise energy requirements during bicycle ergometry testing were greater than expected, possibly because of motor inefficiency caused by weakness. Work capacity and maximal oxygen consumption were decreased, but heart rate, respiratory responses, and blood pressure were within normal limits. Wright and colleagues found small positive physiological effects from an aerobic walking program in patients with slowly progressive neuromuscular disorders. Pinto and colleagues provided eight ALS patients with NIV during exercise to compensate for respiratory insufficiency. Patients walked on a treadmill for 10 to 15 minutes to the point of subjective fatigue, leg pain, heart rate above 75% of resting value, or desaturation of oxygen not correctable with NIV. In comparison to a nonexercising control group, the exercising group had a significant reduction in the rate of decline of respiratory function test results, strength, and function over the 1-year training period.

Endurance training for longer than 10 to 15 minutes in patients with ALS may be restricted by central fatigue, the decreased ability to recruit all motor units or develop high discharge rates, and not merely respiratory function. Sharma and colleagues explored the mechanism of fatigue in ALS. Both maximum voluntary contraction and tetanic force decreased in patients with ALS compared with controls following a 25-minute low-intensity intermittent exercise, but with similar recovery. Fatigue may thus be a consequence of chronic denervation resulting in secondary muscle changes such as altered muscle metabolism and impaired calcium kinetics along with the loss of motor unit activation.

In addition to strength and endurance gains from exercise, ongoing, gentle exercise programs may also help decrease persistent pain and muscle stiffness that often accompany weakened, overtaxed muscle groups. A case study of a patient with ALS undergoing a focused exercise program revealed a positive psychological effect on the patient’s coping strategies. Besides exercise programs, some preliminary evidence exists to suggest that creatine supplementation may increase isometric power in patients with ALS over the short term. Modafinil has been noted to have potential in helping with severe fatigue in ALS. Many studies focus on the impact of exercise on muscle strength; however, knowledge of impairments does not necessarily correlate directly with functional status.
### TABLE 17-2 ■ SUMMARY OF STRENGTH TRAINING STUDIES IN NEUROMUSCULAR DISEASES

<table>
<thead>
<tr>
<th>AUTHOR</th>
<th>STUDY POPULATION AND SAMPLE SIZE</th>
<th>DURATION OF TRAINING</th>
<th>TRAINING MODALITY</th>
<th>TRAINING PROTOCOL</th>
<th>RESPONSE(S)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vignos and Watkins, 1966⁴⁹¹</td>
<td>Various neuromuscular diseases (NMDs) (24)</td>
<td>12 months</td>
<td>Weight training (multiple muscle groups)</td>
<td>Unspecified, but based on 10-repetition maximum (RM)</td>
<td>Strength increased; percentage increase correlated with initial strength</td>
</tr>
<tr>
<td>Milner-Brown and Miller, 1988⁸⁸⁸</td>
<td>Various NMDs (12)</td>
<td>&gt;12 months (variable)</td>
<td>Weight training (elbow flexion and knee extension)</td>
<td>Initially one set of 10 reps based on 15 RM performed on alternate days; gradually increased to a maximum of five sets 4 days/week; protocol individualized</td>
<td>Strength increased significantly when the initial degree of strength loss was not severe (&lt;10%)</td>
</tr>
<tr>
<td>McCartney et al, 1988²⁹⁷</td>
<td>Various NMDs (12)</td>
<td>9 weeks</td>
<td>Weight training (arm curl and leg press)</td>
<td>3 days/week; initially two sets of 10-12 reps at 40% of 1 RM; gradually progressed to three sets of 10-12 reps (one set at 50%, 60%, and 70% of 1 RM); contralateral arm control</td>
<td>Strength and muscular endurance increased; considerable intersubject variability</td>
</tr>
<tr>
<td>Aitkens et al, 1993⁶⁹⁰</td>
<td>Slowly progressive NMD (27) and able-bodied controls (14)</td>
<td>12 weeks</td>
<td>Weight training (elbow flexion, knee extension, grip one side only)</td>
<td>3 days/week; resistance at 30% of 1 RM; work increased commensurate with ability</td>
<td>Significant improvement in most isokinetic strength measures (not grip) in both groups; cross-training effect</td>
</tr>
<tr>
<td>Kilmer et al, 1994⁴⁹⁰</td>
<td>Slowly progressive NMD (10) and able-bodied controls (6)</td>
<td>12 weeks</td>
<td>Weight training (elbow flexion, knee extension, one side only)</td>
<td>3-4 days/week; high-resistance exercise (resistance based on 12 RM; progressed from one to five sets of 10 reps)</td>
<td>Results mixed; increase in leg strength but decrease in arm strength in NMD</td>
</tr>
<tr>
<td>Lindeman et al, 1995²²⁵</td>
<td>MD (33) and HMSN (29); nonexercise control group</td>
<td>24 weeks</td>
<td>Weight training (knee extension and flexion, hip extension and flexion)</td>
<td>3 days/week; initially three sets of 25 reps at 60% of 1 RM; progressed to three sets of 10 reps at 80% of 1 RM</td>
<td>In MD group, no change in strength In HMSN group, increased strength of knee extensors; no adverse effects</td>
</tr>
<tr>
<td>Drory et al, 2001⁸⁹¹</td>
<td>ALS (25): randomly assigned to treatment or control groups</td>
<td>24 weeks</td>
<td>Moderate load, trunk and limbs, concentric contractions</td>
<td>Twice daily, 15 min per session</td>
<td>Treatment group: maintenance of strength, less fatigue, less spasticity, less pain, higher function Increased ROM, strength in treatment group; function sustained better in treatment group</td>
</tr>
<tr>
<td>Aksu et al, 2002⁵⁰⁵</td>
<td>ALS (26): convenience assignment to treatment and control groups</td>
<td>8 weeks</td>
<td>Breathing exercises, PNF, stretching, vs stretching and ROM and breathing exercises</td>
<td>3 days/week supervised vs home</td>
<td>Treatment group had increase in leg muscle strength; no change either group in 2-min walk test</td>
</tr>
<tr>
<td>Dawes et al, 2006⁶⁸⁶</td>
<td>Various NMDs: 11 randomly allocated to control group, nine to treatment group</td>
<td>8 weeks</td>
<td>Walking and strengthening exercises</td>
<td>Walking for 20 min at light to moderate intensity alternating days with progressive resistance and repetitions in strength</td>
<td></td>
</tr>
</tbody>
</table>

Continued
Although some research has shown improvements in muscle force production with strengthening and endurance training, associated functional improvements were evident in some studies, but not others. Jette and colleagues calculated the percentage of predicted normal maximal isometric force (%PMF) relative to four walking levels in patients with ALS: unable to walk, walking within the home only, walking in the community with assistance, and independent walking in the community. Although they found great variation in muscle force production between and within the different levels of walking for each patient, they demonstrated that relatively small changes in force production were associated with losses of functional levels. For example, on average, when an independent ambulator began to need assistance in the community, the lower-extremity strength dropped to less than 54%PMF. When the patient became an in-home ambulator only, the average strength dropped to approximately 37%PMF, and it was approximately 19%PMF when the patient was no longer able to walk. Jette and colleagues acknowledge that many factors need to be considered when interpreting their work; however, their study relates functional skills to isometric muscle force production in a concrete way. Factors such as spasticity, age at onset of ALS, prior levels of fitness and activity, and psychological factors, including past responses to extremely challenging situations and satisfaction with social support, must also be considered.

Based on the evidence and current practice, exercise prescription in the early stages of ALS should address the following:

1. To improve compliance, include both a formal exercise program and enjoyable physical activities.
2. Include activities with opportunities for social development and personal accomplishment.
3. Strengthening programs should emphasize concentric rather than eccentric muscle contractions; use moderate resistance rather than high resistance; and focus on muscles that have at least antigravity strength.
4. Endurance programs should be monitored for signs of fatigue, more so when continuous activity lasts longer than about 15 minutes. Activity programs should include rest periods.
5. Patients should ensure that they have adequate oxygenation, aeration, and carbohydrate loads as well as adequate fluids before exercising.
6. Muscle strength must be monitored to assess for possible overwork weakness; in unsupervised programs, patients must be instructed about signs and symptoms that indicate overwork, including feeling weaker within 30 minutes after exercise, having excessive soreness 24 to 48 hours after exercise, and experiencing severe muscle cramping, heaviness in the extremities, or prolonged shortness of breath; and therapists should check with an independently exercising patient regularly to assess whether any deterioration in strength may be from progression of the disease or overwork weakness.

If a patient shows evidence of significant, persistent weakness after institution of an exercise program or persistent morning fatigue after exercise on the previous day, the therapist must carefully redesign the patient’s exercise program and activity level and increase the frequency of monitoring the patient’s program. The program must be adjusted as the disease progresses. Figure 17-3 is a diagram showing the appropriate exercise “window” for use in working with a patient with a neuromuscular disorder.

**Therapeutic Interventions**

Maintenance of strength and endurance requires daily activity and repetitive muscle contractions. In normal persons, absence of muscle contraction can result in decreases of 3% to 5% in muscle strength per day. If the patient’s exercise level requires less than 20% of the maximal voluntary contraction of the muscles, a decrease in strength will occur; yet overwork must be avoided. Sinaki has described three phases and six substages of ALS with recommended exercise levels (Box 17-2). Although therapists should not assume that all patients will fit precisely within the stages as described, the stages do provide sugges-
### BOX 17-2  ■ EXERCISE AND REHABILITATION PROGRAMS FOR PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS ACCORDING TO STAGE OF DISEASE

<table>
<thead>
<tr>
<th>PHASE I (INDEPENDENT)</th>
<th>PHASE II (PARTIALLY INDEPENDENT)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Stage 1</strong></td>
<td><strong>Stage 4</strong></td>
</tr>
<tr>
<td><strong>Patient Characteristics</strong></td>
<td><strong>Patient Characteristics</strong></td>
</tr>
<tr>
<td>Mild weakness</td>
<td>Hanging-arm syndrome with shoulder pain and sometimes edema in the hand</td>
</tr>
<tr>
<td>Clumsiness</td>
<td>Wheelchair dependent</td>
</tr>
<tr>
<td>Ambulatory</td>
<td>Severe lower-extremity weakness (with or without spasticity)</td>
</tr>
<tr>
<td>Independent in activities of daily living (ADLs)</td>
<td>Able to perform ADLs but fatigues easily</td>
</tr>
<tr>
<td><strong>Treatment</strong></td>
<td><strong>Treatment</strong></td>
</tr>
<tr>
<td>Continue normal activities or increase activities if sedentary to prevent disuse atrophy</td>
<td>Heat, massage as indicated to control spasm</td>
</tr>
<tr>
<td>Begin program of range-of-motion (ROM) exercises (stretching, yoga, tai chi)</td>
<td>Preventive antiedema measures</td>
</tr>
<tr>
<td>Add strengthening program of gentle resistance exercises to all musculature with caution not to cause overwork fatigue</td>
<td>Active assisted passive ROM exercises to the weakly supported joints; caution to support, rotate shoulder during abduction and joint accessory motions</td>
</tr>
<tr>
<td>Provide psychological support as needed</td>
<td>Encourage isometric contractions of all musculature to tolerance</td>
</tr>
<tr>
<td><strong>Stage 2</strong></td>
<td><strong>Stage 5</strong></td>
</tr>
<tr>
<td><strong>Patient Characteristics</strong></td>
<td><strong>Patient Characteristics</strong></td>
</tr>
<tr>
<td>Moderate, selective weakness</td>
<td>Severe lower-extremity weakness</td>
</tr>
<tr>
<td>Slightly decreased independence in ADLs, such as:</td>
<td>Moderate to severe upper-extremity weakness</td>
</tr>
<tr>
<td>■ difficulty climbing stairs</td>
<td>Wheelchair dependent</td>
</tr>
<tr>
<td>■ difficulty raising arms</td>
<td>Increasingly dependent in ADLs</td>
</tr>
<tr>
<td>■ difficulty buttoning clothing</td>
<td>Possible skin breakdown as a result of poor mobility</td>
</tr>
<tr>
<td>Ambulatory</td>
<td><strong>Treatment</strong></td>
</tr>
<tr>
<td>Continue stretching to avoid contractures</td>
<td>Encourage family to learn proper transfer, positioning principles, and turning techniques</td>
</tr>
<tr>
<td>Continue cautious strengthening of muscles with manual muscle testing (MMT) grades above F+ (3+); monitor for overwork fatigue</td>
<td>Encourage modifications at home to aid patient’s mobility and independence</td>
</tr>
<tr>
<td>Consider orthotic support (e.g., ankle-foot, wrist, thumb splints)</td>
<td>Electric hospital bed with antipressure mattress</td>
</tr>
<tr>
<td>Use adaptive equipment to facilitate ADLs</td>
<td>If patient elects home mechanical ventilation (HMV), adapt chair to hold respirator unit</td>
</tr>
<tr>
<td><strong>Stage 3</strong></td>
<td><strong>Stage 6</strong></td>
</tr>
<tr>
<td><strong>Patient Characteristics</strong></td>
<td><strong>Patient Characteristics</strong></td>
</tr>
<tr>
<td>Severe selective weakness in ankles, wrists, and hands</td>
<td>Bedridden</td>
</tr>
<tr>
<td>Moderately decreased independence in ADLs</td>
<td>Completely dependent in ADLs</td>
</tr>
<tr>
<td>Easily fatigability with long-distance ambulation</td>
<td><strong>Treatment</strong></td>
</tr>
<tr>
<td>Ambulatory</td>
<td>For dysphagia: soft diet, long spoons, tube feeding, percutaneous gastrostomy</td>
</tr>
<tr>
<td>Slightly increased respiratory effort</td>
<td>To decrease flow of accumulated saliva: medication, suction, surgery</td>
</tr>
<tr>
<td><strong>Treatment</strong></td>
<td>For dysarthria: palatal lifts, electronic speech amplification, eye-pointing electronics</td>
</tr>
<tr>
<td>Continue stage 2 program as tolerated; use caution not to fatigue to point of decreasing patient’s ADL independence</td>
<td>For breathing difficulty: clear airway, tracheostomy, respirator if patient elects HMV</td>
</tr>
<tr>
<td>Keep patient physically independent as long as possible through pleasurable activities such as walking</td>
<td>Medications to decrease impact of dyspnea</td>
</tr>
<tr>
<td>Encourage deep breathing exercises, chest stretching, postural drainage if needed</td>
<td><strong>Special Considerations</strong></td>
</tr>
<tr>
<td>Prescribe wheelchair, standard or motorized, with modifications to allow eventual reclining back with head rest, elevating legs</td>
<td><strong>Special Considerations</strong></td>
</tr>
</tbody>
</table>

tions for interventions on the basis of degree of impairment, functional limitations, and level of disability. In the following section, staging patterns are used as the framework for therapy interventions. Staging information is particularly helpful to therapists who do not have the opportunity to work with large numbers of patients with ALS.

Most patients need specific guidance about what type of activities and exercises they should do. Although many physicians may suggest to patients that they increase their activity level, their suggestions are seldom specific. Examples of exercise advice that patients have recalled are “Try to move around as much as possible,” “Walk some more,” and “Be active, but don’t overdo it.” Because changing their typical exercise pattern is difficult for most patients, even when they know doing so is important, referral for a physical therapy consultation can be helpful.

**Phase I (Independent): Stages 1 to 3.** A program to increase activity must be specifically designed, with input from the patient about willingness to participate and knowledge of the patient’s environmental situations and social support systems. In the early stages of the disease, patients should be encouraged to continue as many prediagnosis activities as tolerated. For example, a golfer should continue to golf for as long as possible. Walking the course should be encouraged if it is not too fatiguing. When walking or balance becomes difficult on uneven terrain, the golfer can use a golf cart, decrease the number of holes played, move to a par 3 course, or hit balls at a driving range. If upper-extremity weakness is a major problem that interferes with swinging the club for distance shots, the player can continue playing the greens or on putting courses. Some golfers may need adaptations to club handles with nonskid material such as Dycem (Dycem Non-Slip products, www.dycem.com) or Scoot-Gard (Vantage Industries Product) to prevent the club from rotating on impact.

Patients with newly diagnosed ALS who had a sedentary lifestyle before diagnosis should be encouraged to increase their activity level. This may include activities that require muscular effort within or around the home, such as sharing household and gardening tasks or beginning a walking program around the neighborhood. After diagnosis, some patients begin searching for in-home exercise devices such as bicycles and rowing machines. As with healthy persons who start an exercise program after the purchase of exercise equipment, patients with ALS are not likely to use the equipment consistently if they did not before a diagnosis. The search for a “perfect” exercise machine may reflect the patient’s desperation to do something tangible. Without taking away the patient’s motivation to exercise, therapists can encourage participation in exercise programs that do not require expensive equipment, such as walking or working out to specific exercise routines. A clever therapist can make a video for each patient that includes stretching and gentle exercise programs that elicit muscle contractions from all functional muscle groups (by using inexpensive elastic bands or small weights) with follow-up breathing, “warm down,” and relaxation exercises. Patients could follow a program of six maximal isometric contractions held for 6 seconds and isotonic elastic band exercises at submaximal levels to maintain and improve muscle strength. Patients should exercise for short periods several times a day rather than attempting to exercise all muscle groups in one session.

For most patients in the early stages of ALS, pleasurable, natural activities such as swimming, bowling (can gradually decrease weight of ball if shoulder strength is a problem), walking, bicycling (three-wheeler may be needed or in-home stationary bicycle, either of which must be evaluated for easy mounting and dismounting), or tai chi should be recommended. Some patients prefer to exercise alone, whereas others will gain confidence and companionship by joining a group activity. Listening to the patient’s desires related to group activities is important. The dropout rate is high among those who have been pressured to participate. Some spouses or family members are supportive of the patient’s activity needs and will join the patient in his or her regimen. If possible, the spouse and family members should be engaged in the treatment planning process.

The therapist must observe the patient completing her or his entire recommended activity program. The patient’s response to the program must be monitored because fatigue from exercise sessions can interfere with the ability to carry out other normal daily activities. If the patient becomes too exhausted at the end of a session, he or she may learn to fear exercise and may become depressed about the decreased activity status. This depression may lead to decreased activity and further deconditioning (see Chapter 6).

**Phase II (Partially Independent): Stages 4 and 5.** During phase II, the goal of physical and occupational therapy intervention should be to help the patient adapt to limitations imposed by weakness and spasticity, an increasingly compromised cardiorespiratory status, and possible pain from stress related to weakness or muscle imbalance. This transition stage is often frightening for patients because the decrease in function and independence becomes clear; therapists should accentuate what the person can do and how accommodations can be made to help maintain independence. After a full physical assessment of the patient’s motor status similar to the initial evaluation, the patient, family members, and therapists (including PT, OT, and speech therapists) should discuss treatment options and adaptive devices that can help the patient remain as independent as possible.

During late phase I and through phase II, many patients show significant weakness of both upper- and lower-extremity musculature, but each patient has his or her own pattern and rate of progression of weakness and onset of spasticity, bulbar, and respiratory symptoms. A typical patient at this time may have marked weakness of the intrinsic muscles, shoulder muscle weakness (in some cases “hanging arm” syndrome) with shoulder pain, and generalized lower-extremity weakness (in some cases more severe distally). Patients may be able to walk within the home environment, but many patients have precarious balance and fall easily because of muscle weakness. At this stage, most patients report fatigue with minimal work and have to rest frequently when carrying out ADLs. ROM can deteriorate quickly in this phase of the disease, requiring daily stretching to end range for the calf, quadriceps, hip adductors, trunk lateral flexors, and long finger flexors.

Moderate exercise can have a modest effect in reducing spasticity.

Patients at this point, even if ambulatory, should consider using a wheelchair outside the home to conserve energy. Factors to consider in choosing a wheelchair include extent of insurance coverage or financial assistance
programs for purchase of wheelchair (some policies or programs may provide only one type of wheelchair or only one wheelchair, either motorized or manual); transportability of motorized chair from home to community and work (few motorized wheelchair brands fold for stowing in car trunk, and few families can afford to purchase a van that will allow the patient to drive or be driven while in a motor chair); reclining potential of chair back and headrest (preferably electric) to allow the patient to shift weight and rest while in the chair during later stages of the disease; removable arm rests for ease of transfer; potential for headrest attachment or extension; potential mounting area for portable respirator equipment if needed; and ease with which caregiver can help patient with chair mobility transfers. Chairs should have lumbar support and appropriate cushioning to prevent pressure ulcers.

At this stage, patients with more advanced bulbar symptoms begin to experience dysarthria and may need guidance in dealing with communication issues. Murphy indicated four major reasons for communication: to identify needs or request help, share information, respond politely in social situations, and maintain social closeness. The primary focus of communication for the study participants was to maintain social closeness. Although few patients had any instruction in ways to deal with communication problems, most patients and caregivers created ways to make themselves understood, such as giving cues about the topic and context, creating a “shorthand” language, and checking with the dysarthric speaker to ensure that the listener understands the patient correctly. A number of patients in the study who had significant dysarthria commented that attempting to communicate socially was extremely tiring. Therapists who are guiding patients with energy conservation techniques should be aware of the exhaustion that can be associated with communication. A number of strategies recommended by the American Speech-Language-Hearing Association can be used by the person with ALS to deal with the effects of dysarthria, including the following:

- Reduce background noise in the room.
- Face the person while talking.
- Use short, simple phrases rather than long, complicated ones.
- Take the time to say what needs to be said; do not allow people to rush conversation.
- Make extra use of body language, such as gestures and facial expressions, and use writing to supplement speech, if possible.
- Do not worry about saying things correctly; if the basic message being conveyed is understood, then that is enough.

Also in this stage, some patients and families may need support to identify adapted feeding systems (special utensils, adapted plates, adjustable tables) and hygiene equipment if transfers within the family bathroom are problematic.

Because Mr. Turner in Case Study 17-1 was cared for in a neuromuscular disease clinic, he benefited from input from multiple specialists working as a team to help him maintain his independence. Unfortunately, many patients do not have the benefit of such a coordinated treatment environment. Therefore, when necessary, the therapist must be in a position to provide input on adaptive and safety devices and bulbar issues if other specialist input is not available. Therapists working in smaller communities and rural areas most likely need to be chameleon-like to play many therapeutic roles when working with the patient with ALS.

CASE STUDY 17-1 MR. TURNER

Mr. Turner is a 45-year-old man diagnosed 2 years ago with ALS. He lives at home with his wife, who works full time, and two teenage children. Mr. Turner is a computer programmer for an engineering firm in the area. Since his diagnosis, Mr. Turner has been able to continue his full-time work schedule, although he states that he is no longer able to touch type and can type with the index fingers only. He has noticed that his shoulders and neck hurt (4 out of 10 on a numerical pain rating scale) after an hour at the computer. In the last 2 weeks he has found it fatiguing to walk to the cafeteria for lunch (approximately 100 meters), and he fears that he will be knocked down when walking in crowds. He dropped his tray last week, which was embarrassing, so he decided to eat in his office even though he misses the socialization and opportunity to discuss work issues with his colleagues.

Mr. Turner has been able to continue most of his nonwork activities, although he is no longer able to operate his sailboat independently and is having trouble maintaining his balance when golfing. Also when golfing, he now uses a cart and plays only nine holes. He states that his wife and children are supportive and that they have made some changes in the home environment to accommodate his increasing weakness. He also revealed, however, that his children seem frustrated with him because he is so much slower than he was before the illness.

On assessment, Mr. Turner showed marked wasting of hand intrinsics. He was unable to abduct or flex either shoulder past 90 degrees. His right shoulder showed considerable atrophy, especially of the deltoid and supraspinatus muscles. All other upper-extremity movements were weakened but in the G (4–) range. His neck posture was forward: neck extension is F+ (3+), neck flexion is G− (4–). Scapular winging was noted bilaterally. No spasticity or loss of passive ROM was evident in the upper extremities. Lower-extremity musculature showed generalized weakness at the F (3) to F+ (3+) range, with left musculature weaker than right, marked wasting of the foot intrinsics, and a cavus foot position bilaterally. Spasticity of the hip adductors and hamstrings was noted (Modified Ashworth Scale grade 2), but no passive ROM loss was detected in the lower extremities. Most obvious during gait was inadequate dorsiflexion for heel strike and no propulsion during heel-off. He showed a bilateral corrected gluteus medius pattern on weight bearing. He needed to pause to lock each knee during weight bearing and at times he pushed his knee into extension with his hand. He had great difficulty ascending and descending the four steps to enter his home. There were no stairs to negotiate at work.

Until this appointment, Mr. Turner had not been willing to discuss the use of adaptive equipment or a wheelchair. During

Continued
prior clinic visits his decisions were supported and he was told that when he was ready, therapists would work with him and his family to help with equipment decisions.

Mr. Turner also showed some early bulbar signs. He noted that he sometimes had to catch drool when working intensely, and that his pillow was moist in the morning. Food sometimes got stuck in his cheek area and he could not move it out with his tongue. Swallowing was still adequate for eating all foods; however, he had had a few coughing episodes when drinking coffee and wine. He showed increased use of accessory musculature when breathing but had no reports of respiratory distress. His cough was adequate to clear secretions.

With input from the therapist, Mr. Turner and his wife identified the following general goals:

1. Increase mobility while conserving energy
2. Control fatigue and pain of upper extremities and neck during computer work
3. Maintain maximal muscle strength and ROM (patient reported that he felt stiff)
4. Identify safety issues within the home and work environment and adjust household and work environment to prepare for the time when Mr. Turner could not ascend and descend stairs safely.

A treatment plan was discussed to achieve the following:

1. Increase mobility. Because of his increased walking difficulties, Mr. Turner decided to use a front-wheeled walker with a seat attachment at home. Because of his hand grip weakness, he felt most stable using attached forearm troughs. For his worksite, he selected a motorized wheelchair so that he could maintain his independence at work. Although he found that he could push an ultralight manual chair, his upper-extremity strength was clearly decreasing. Mr. Turner decided that he preferred the motorized chair to an electric scooter because of the financial cost of switching devices when the scooter no longer provided adequate postural support.

   Because Mr. Turner’s insurance and Medicare would not fund an additional manual chair and because the family had no way to transport the electric wheelchair, the ALS Society loaned the family a manual wheelchair for home use. Although not ideal, it was functional. Mr. Turner’s son made some inexpensive adjustments to adapt the chair for a headrest, and his daughter and grandchildren repainted the chair to his specifications.

   Because Mr. Turner wanted to keep as active as possible and use his walker within the home, he was fitted with bilateral ankle-foot orthoses (AFOs) with a flexible ankle joint and preibial shell to facilitate knee extension. Straps were simple overlap style because Mr. Turner had poor thumb and grasp control.

2. Decrease fatigue and pain of upper extremities. Mr. Turner was taught some simple ROM exercises of the neck and arms to perform every half hour while working at the computer. In a simulated work environment the therapist noted that Mr. Turner had a forward head position when working at a computer similar to his workstation. The height of the computer was adjusted to decrease his neck strain, and the desk height was adjusted to allow his wheelchair to fit under the desk so that his arms could rest fully on the surface.

   He felt immediate relief with the adaptations. He was also fitted for a soft neck collar to wear when he felt he needed more neck support. (As his condition worsened, he learned to rest his head on the headrest of his chair and recline slightly for a few minutes every 15 minutes.)

3. Maintain maximal muscle strength and ROM. Mr. Turner was taught as many self-ranging maneuvers as possible, which he was encouraged to do in small segments frequently throughout the day. For example, his series of motions included neck rotations, side bends, and flexion and extension within strength limits; upper-extremity motions with the exception of shoulder flexion and abduction past 90 degrees; hip flexion, abduction, and rotations; full knee extension; and all ankle motions. When using the walker, Mr. Turner was encouraged to extend each hip fully and to stretch his heel cords. Mrs. Turner and their adult children were taught to administer full ROM exercises, including trunk rotations, with special attention to ranging of the shoulder to prevent impingement. Simple massage techniques were also taught to all family members who felt comfortable with the task.

   Mr. Turner had been active before the onset of ALS and he liked to exercise. He rented a portable pedaling unit to attach to a chair at home. He pedaled two to four times a day, with no additional resistance, to the point at which he felt fatigue (usually 3 to 5 minutes at this stage). He carefully monitored his soreness and fatigue level after exercise and increased and decreased his pedaling depending on how he felt immediately and several days after exercise. Mr. Turner felt invigorated by this exercise, which he usually did while watching television. He was also taught a series of simple elastic band exercises, with tensile strength adjusted according to his ability to contract his muscles without fatigue. Mr. Turner was also shown a series of isometric exercises for all muscle groups to do throughout the workday. Because he had some foot and ankle edema, he was encouraged to wear lightweight pressure stockings while sitting. Mr. Turner also had access to a swimming pool, and he was encouraged to carry out walking and upper-extremity exercises as long as another adult was with him in the water at all times.

4. Assess environment of home and work. Occupational therapy input was requested to help with ADL aids such as reachers, utensil adaptors to facilitate grip, rubber pen grippers, key adaptors to permit turning, and thumb abduction splints to assist in pincer grasp. Mr. Turner’s OT made several visits to his worksite and home to identify adaptations of the environment for safety and independence. His wheelchair was eventually adapted with universal joint arm troughs to decrease his effort during self-feeding and basic upper-body hygiene. Ramps were recommended for home entry, and nonpermanent safety rails were placed in the bathroom. Mr. Turner was able to assist with transfer to a shower chair, and the shower head was replaced with a handheld unit.

   A speech pathology consultation was also requested. Using information from the PT’s manual muscle testing, the speech pathologist carried out a thorough bulbar evaluation and provided information about swallowing.
techniques. The speech therapist focused on ways to
decrease drooling and ways to cope with food pocketing
(tongue mobility was impaired) by using techniques
such as hand pressure on the cheek to push food back
to the center of the mouth. The therapist also instructed
Mr. Turner and his wife how to prepare foods with textures
that were easily swallowed and manipulated. Mr. Turner
had lost 5 pounds during the last 6 months, so he was
also referred to the dietician for information about how
to maintain nutritious calorie intake.

PROGRESSION OF THE DISEASE
Within 3 to 4 months after initial examination, Mr. Turner was
no longer able to continue working despite workplace adapta-
tions. At home, he became more dependent. Mr. Turner had great
difficulty adjusting to his physical dependence. Because of
his slow onset of dysphagia and his augmented communication
system, he was able to continue control over his expressive,
cognitive, and emotional life for another few months. Initially
Mr. Turner angrily resisted his wife’s attempts to help him with
eating and dressing tasks. This began to alienate her and the
children until a family meeting was held with their medical
social worker and PTs and OTs. All family members had the
opportunity to express their frustrations. A major irritation to
the children was what they perceived to be their constant waiting
for their father to complete a task. Mrs. Turner was most irritated
when Mr. Turner yelled at her when she attempted to help even
though he frequently expressed anger about his clumsiness.
Mr. Turner sadly admitted that he was having increasing diffi-
culty with his ADLs and was sometimes too tired after dressing
to participate in family activities. At the end of the meeting, the
family had worked out a compromise plan. Mr. Turner would
continue to do as much as possible for himself. He would specif-
ically ask for help from Mrs. Turner when he wanted it so she
did not get caught in his anger about needing help. He preferred
that the children not have to take any role in his care at this point
but realized that he might need their help later. Visiting nurse
support was requested twice a week to help with bathing, and
the OT was requested to make another home visit to help with
toileting needs. Mr. Turner felt comfortable with his wife and
children carrying out ROM exercises. A therapy home visit was
arranged to review the exercise and positioning program as well
as respiratory exercises and postural drainage techniques.

As Mr. Turner became totally dependent, he needed 24-hour
care. Professional nurses were provided through his insurance
contract 14 hours a day from 6:30 AM to 8:30 PM. Family mem-
bers provided care until midnight. Initially Mr. Turner was able
to activate a bell at night to call for help. His wife and children
followed a schedule to turn him every 3 hours throughout the
night. When Mr. Turner became respirator dependent and was
no longer able to call for help, it became clear that the nighttime
responsibilities were taking a heavy toll on his wife, who
worked full time, and the children, who were in high school and
college. Fortunately the family was able to pay for a nurse as-
stant to remain at Mr. Turner’s bedside throughout the night,
although the family members all felt that they had no privacy.
Although the family was committed to having Mr. Turner re-
main at home until his death, all agreed that they needed respite.
Thus several week-long hospitalizations were made to give the
family a break in the constant care needs.

Although Mr. Turner had elected HMV, he also had signed
a durable power of attorney for health care, indicating that he
did not want treatment for infections and that palliative care for
comfort should direct his treatment. He had a strong lust for
life, but he had come to accept his impending death. He did not
have strong religious views, but he had talked with all his care-
givers and therapists about his concerns related to death. He
freely expressed his fear of “nonbeing.” Because his caregivers
and therapists were willing to talk about his and their own feel-
ings, Mr. Turner came to believe that he would live on in the
minds, hearts, and behaviors of those he had known. This idea
seemed to give him great comfort. He particularly liked to talk
to others about special times they had had together and how
their interactions had affected each other. To help Mr. Turner
process his death, his family, friends, and medical team put
together an album of pictures and statements about their time
gether. Mr. Turner frequently liked to have his wife read
through the book with him. His family continued to carry out
his ROM exercises and massage because Mr. Turner had indi-
cated that the treatments provided him physical comfort and
the spiritual closeness he needed with his family. His primary
treatment during the last few days consisted of morphine to
decrease his respiratory discomfort. After 5 to 6 months of
being totally dependent for all care and respiratory function,
Mr. Turner died at home in his sleep after a respiratory illness.

Phase III (Dependent): Stage 6. PTs and OTs are
usually less involved in the care of the patient in phase III,
and nursing personnel become more active. During this
phase, therapists make home visits to support caregivers
and respond to questions about pain control, bed mobility,
positioning to prevent pressure ulcers, ROM, and equip-
ment adaptations. Therapists should be sure to teach
all caregivers some basic body mechanics to use during lift-
ing or positioning devices if spasticity or paralysis leads
to caregiving difficulties (e.g., excessive adductor tone and
collar tubes interfering with hygiene and bowel care) or
tissue damage and pain. If nursing care providers do not
give advice on pressure relief beds or mattresses of air or
foam, therapists should be prepared to do so. Unfortu-
nately, many insurance providers and Medicare may not
fund special mattresses, and they can be costly. Therapists

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may also need to review postural drainage techniques with caregivers.

Of greatest importance in phase III, and sometimes in earlier stages, is the patient’s ability to communicate. In the earliest manifestation of dysarthria, therapists train patients to slow the speech rate and cadence, exaggerate lip and tongue movements, and manage phrasing through breath control.83 Although spouses and caregivers can often interpret their partner’s or patient’s severely dysarthric speech (see earlier discussion of phase II), most patients who use NIV or invasive ventilation for a prolonged period need to find nonverbal methods to communicate. If severe bulbar impairments precede extremity paralysis, paper and pencil, alphabet and word boards, and adapted computer keyboards can be used with minimal upper-extremity or finger strength for pointing. The American Speech-Language-Hearing Association provides suggestions for developing communication boards with the specific language most appropriate for the patient’s situation.112 For example, the board may be designed with commonly needed sentences, words used in the person’s daily life, and the alphabet. As the person’s ability to finger point decreases, the language board can be redesigned. When no extremity movement is possible, subtle neck movements or pressures, eye gaze, eye blink, upper facial movements, and electroencephalographic activity can be harnessed to operate communication devices.114,115 Learning to use electroencephalographic interfaces, however, takes months of intense training and may not provide a reasonable system for communication for most patients with ALS.116

Some patients with hypernasality benefit from using an orthodontic palatal appliance. Patients with a tracheostomy may benefit from use of a Passy-Muir (Irvine, Calif) speaking valve tracheostomy tube. These devices require recommendation by communication specialists. As speech quality deteriorates and sound projection wanes, the spouse or caregiver can use an electronic speech amplifier to magnify the patient’s speech. Speech pathologists and therapists have information on commercially available amplifying devices that are often used by persons with hearing problems but can be used by hearing people to amplify the speech of a person with severe weakness of phonation.

When selecting a communication device, therapists must work closely with the patient and family members to ensure that the system is compatible with patient skills and communication needs and preferences. Expensive systems commonly lie unused because of simple factors such as lack of proximity to the patient, interference of the unit with personal care, increased caregiver workload to manage the unit, and slowness of communication processing. The best systems are tailored to the precise needs of the patient; however, many patients do not have the financial or insurance support to purchase the device, and many patients in the end stages of ALS do not have the time to wait for systems designed for their specific needs. Therefore commercially manufactured systems may be most appropriate. (See Cook and Hussey114 for a comprehensive list of communication devices and control interfaces.)

Some patients and caregivers learn to communicate effectively with simple eye gaze, eye blinking, and clicking techniques with Morse code or self-developed codes. At minimum, patients with no ability to communicate or move and their caregivers must have some system to communicate emergency needs; for example, looking to the right means “help” and looking to the left means “pain.” Therapists should help patients develop alternative modes of communication before intelligible speech becomes impossible. (See also Cobble117 for information on language impairments.) In addition to communication systems, environmental control systems can be programmed to turn on and off television, lights, and other electronic units with the same type of switching units used for communication (e.g., eye blink, infrared beam, head movement pressure). Unfortunately, these devices are often expensive and may not be available to all patients. (See Cook and Hussey114 for a comprehensive review of environmental control systems.) Financial support is often not extended for high-tech equipment by third-party payers because of the patient’s limited life expectancy. The ability to communicate and call for help, however, is of paramount importance with completely dependent patients.

By phase III most patients have significant problems eating and maintaining nutrition, although these problems may manifest in earlier stages. Patients often report choking or coughing after swallowing liquids or problems moving food around in the mouth or to the back of the throat for swallowing. These problems are best handled medically and can be assessed with videofluoroscopy or videendoscopy. The aggressiveness of treatment intervention depends on the patient’s preference and whether she or he still wants to attempt any oral feeding (e.g., syringe feeding, oral gastric tubes) or wishes to have a PEG or another alternative to oral feedings implemented. Therapists, however, can help patients and caregivers develop strategies that improve eating and nutrition, such as adjusting eating position, changing head and neck alignments, adding thickeners to liquids, and adjusting portion sizes and texture of foods.7

Psychosocial Issues

Giving the bad news of a terminal diagnosis is difficult for even the most experienced clinician. In dealing with the diagnosis of ALS, most physicians now believe that the diagnosis, prognosis, and possible patterns of progression should be shared with the patient and family or partners and caregiving friends. Only by knowing the truth can patients and families deal openly with one another and make plans for the future.118 McCluskey and colleagues119 suggest that those giving the medical or therapeutic diagnosis should attend to good practice parameters when giving bad news, such as creating the appropriate setting, identifying patient and caregiver needs, asking what patients and caregivers want to know, providing knowledge, exploring feelings of the patients and caregivers, and formulating a strategy for dealing with the situation. Patients and family members seldom remember what they are told when first given a terminal diagnosis. They do, however, remember how the information was given. Therefore information should be given honestly but with a sense of hope. All information need not be given at the time of diagnosis. Rather, the patient and family can be exposed to more in-depth information over a number of sessions when they have the opportunity to ask questions that occur during the assimilation process. Therapists, especially those working in isolation from a comprehensive clinic, should also follow these guidelines by
providing information, helping the patient and family identify goals, and establishing a plan for intervention. Patients should know that the goals will have to be adjusted and plans reset as the disease process continues. If patients and families know that they can contact the therapist for support and advice, many of the negative aspects of the illness can be confronted in a positive manner. Preferably, an appointment for a follow-up visit will be set so patients and family members feel that contact with the care provider is expected.

Information about transitions related to nutrition, communication, and respiratory functions should be delivered to patients and families in time to make thoughtful decisions rather than just before a time of crisis, such as after a choking episode or during a respiratory arrest. Care should also be taken to respect the cultural and spiritual views of the patient and family. Preferably, patients and family members will prepare an advance medical directive that should be reviewed with the physician at least every 6 months.

Therapists treating patients who do not have access to a multidisciplinary ALS clinic should remember that they are often the person who works most closely with the patient, and they should plan on spending enough time with the family to respond to concerns and help with problem solving. Patients will progress through the diagnostic process with different responses and at different rates on a continuum from taking a cognitive approach by asking many questions and reviewing the most current research to the extreme of marked denial and disinterest in participating in any medical or therapeutic recommendations.

Purtilo and Haddad identified four major fears of the patient who has a terminal condition: fear of isolation, fear of pain, fear of dependence, and fear of death itself. Patients with progressive diseases often see their social contacts decrease. Mr. Turner in Case Study 17-1 was concerned when he was no longer able to join his colleagues in the company cafeteria. After he received his motorized wheelchair he was able to continue his social contacts until his bulbar symptoms progressed to a point that he chose not to eat in public. When Mr. Turner lost the ability to speak and had to use his computerized speech system, he noticed that fewer colleagues stopped by his office to talk because of the slowness of the communication process. Although he understood the problem, Mr. Turner mourned the loss of friendship and his loss of standing as a competent computer expert. Because of his need for social contact, Mr. Turner continued to work until he could no longer tolerate the sitting position. His fear of isolation increased when he became homebound.

Although colleagues came for visits regularly at first, as Mr. Turner progressed to a near locked-in state only a few close friends came by for brief visits. Mr. Turner’s greatest fear was being separated from his family and abandoned to hospital care with inconsistent staffing patterns. Fortunately, in his community, Mrs. Turner was able to set up visitations from several church members, clerics, and hospice volunteers.

Fear of uncontrolled pain is common among people with terminal diseases. Patients need assurance that their pain will be controlled. Fortunately, today pain medications can be administered in many forms, dosages, and frequencies that can be tailored to the patient’s specific needs. In a study of the final month of life with ALS, caregivers reported that a major emphasis of care was to eliminate as much pain and discomfort as possible, even if it shortened the patient’s life. Keeping a pain log of intensity, type, location, and time of pain may provide the physician with information necessary to best prescribe dosages. Many patients with ALS do experience significant pain from musculoskeletal sources, persistent spasms, or spasticity and pressure sores. Most of these problems can be handled with appropriate pain medications, muscle relaxants, careful positioning, frequent ROM exercises, and tissue massage. Undertreated and uncontrolled pain is associated with a patient’s seeking information on assisted suicide. Some patients who expressed interest in assisted suicide options did not follow up because of religious beliefs and concerns about possible loss of life insurance coverage for surviving family members.

A major concern of patients with ALS is the dependence necessary for ADLs associated with late phase II and phase III of the disease. Because the process is gradual, most patients have the opportunity to make adjustments. The dependency issues and resulting privacy issues are more uncomfortable for some patients than for others, especially for the person who has always valued self-control and independence. Some patients are concerned about their increasing dependence because of the consequences of increasing burden of care on spouses or other caregivers. That concern for others sometimes causes patients to choose hospital, nursing home, or in-patient hospice care over home care during the terminal stage of the disease. Not all patients with terminal illness react the same way during the dying process. Throughout the process, patients and family members may cycle back and forth through a range of different emotional and coping reactions: depression, anger, hostility, bargaining, and acceptance and adaptation (order is not implied). How the patient coped with life’s difficulties before the illness and her or his prior relationship patterns often direct how the patient will deal with the terminal illness. In one study, patients adjusted most successfully to the changes in their functional status if they did not look back to the past and compare their losses to their future.

Health care providers and family members often have great difficulty coping with a patient who is depressed; they may make repeated efforts to “talk the person out of” the depression. Medical professions must be able to distinguish between depression that can be destructive and the mourning or grieving that is a necessary and vital response to dealing with loss. In both states the person may feel a level of withdrawal, sadness, apathy, loss of interest in activities, and cognitive distortions. In a depressive state, however, the patient experiences an accompanying loss of self-esteem. A person in mourning rarely experiences that loss of self-esteem essential to a diagnosis of depression. The grieving person’s feelings are congruent with the degree of loss experienced. A person who grieves for what is lost but who has adapted to the prognosis may make plans for the impending death. Such behaviors are positive coping strategies. However, depressive symptoms related to hopelessness, uncontrolled suffering, and perceived burden on caregivers are more related to a choice for treatment discontinuance of feeding or ventilatory support.

The issue of depression is complicated by the pseudobulbar effect of emotional lability (inappropriate laughing and crying), which is manifested by approximately 50% of patients with ALS. This emotional lability is not under complete control of the patient and is often misunderstood by family members and caregivers. Although current treatment is antidepres-
sant medications, underlying clinical depression may or may not be present that would respond to higher doses of antidepressant medication and counseling.120

Yet, pressuring a patient who appears depressed to see a mental health clinician can lead to loss of trust if the patient is not comfortable talking about feelings or confiding in a counselor. Therefore, OTs and PTs and other persons involved in the direct care of a dying patient may find that their patients feel safer talking with nonprofessional counselors or psychotherapists about the burden of their care on family members or their own impending death. Rehabilitation personnel should, therefore, be aware of local options for in-home support services, palliative care, and end-of-life options and services and be prepared to listen to the patient’s concerns if the patient expresses the need for emotional support.

**Caregiver Issues**

Often in the concern for the patient’s needs, health care professionals pay little attention to the effect a person’s degenerative illness has on other members of the family. ALS significantly affects the person’s extended family because the patient gradually becomes increasingly dependent on family members, partners, or caregiving friends for physical care, social arrangements, cognitive stimulation, and emotional support. For some families, the spouse may have to take on additional work, return to work, or, in the case of some older women, join the workforce for the first time to deal with the financial stresses that occur when chronic illness invades the family unit. Family members must absorb the former family duties of the dependent person. For example, a spouse or child may have to handle all the cooking, cleaning, or other household chores or work to help support the family. Once the patient becomes dependent, the caregiver may need to reduce or discontinue employment to take care of the patient. All family members may have to become involved in the physical care of the increasingly dependent person with ALS.

Children of patients with ALS also have to deal with major changes in their lifestyle. Although they may love their parent who is sick, at some level most are frustrated with factors such as the need to provide physical care to parents. This is a difficult problem for children who have not had a positive relationship with that parent. Children living in the home of a parent who is dying of ALS also express frustration about the lack of privacy in their home when nursing personnel and attendants are present, interruptions in family and personal life plans, embarrassment because of the parent’s appearance and dependency, lack of attention from the caregiving and working parent, and fear of financial crises (e.g., possible loss of home, no financial support for college).

The entire family is affected by the sick person’s increasing dependency and impending death. In a small study of 11 family caregivers, many caregivers felt frustrated and resentful because their lives were consumed with the caregiving responsibilities. Most caregivers had adjusted to some degree after 2 to 4 years. Caregivers who adjusted most successfully learned to take time for themselves without guilt and to tap their social support systems for help.126,128 Similarly, 40 caregivers of young adults with severe disabilities reported being overwhelmed by the physical requirements of daily care and felt a severe loss of spontaneity in their lives.129 They also reported a sense of isolation from everyday social interactions. Although they highly valued their social support systems, they expressed frustration that few people offered instrumental or direct service support, such as respite care or help with medical appointments, housekeeping, or shopping. Despite the stresses of caregiving, the caregivers felt positive about their roles in helping the dependent adult by finding meaning in their acts of caregiving.129 Fortunately, most families manage to cope with the process—the major contributing factor being the coping ability of families before the illness. To be really effective, the therapist working with the patient with ALS must be prepared to help families and caregivers find appropriate ways of coping with the emotional, social, and physical stress of caregiving. For example, therapists should present, without pressing, adaptive equipment options to patients when they first start to show impairment in functional ability. If shown how the equipment will help them maintain independence, most patients are receptive to its use. Even when presented in a positive way, however, a wheelchair or adaptive devices may be resisted long after the adaptations would facilitate mobility and ADLs. Therapists must be attentive to patients’ feelings and fears at this time because use of a wheelchair heralds to many patients the beginning of the end.

Other factors that affect the family of a patient with ALS include medical insurance and differing levels of long-term care coverage. Some families are fortunate to have excellent coverage that provides extensive home nursing support, whereas other families are unable to cope with the financial stresses and must accept public assistance during the final stages of the disease. As opposed to Germany and Japan, which provide long-term nursing care insurance, in the United States financial stress on patients with ALS can reach more than $150,000 per year for ventilation support at home.63 Financial burden significantly impacts patient and caregiver decisions. (See Case Study 17-1 and end-of-life issues resources at www.nlm.nih.gov/medlineplus/endoflifeissues.html#cat1.)

**GUILLAIN-BARRE Syndrome**

**Pathology and Medical Diagnosis**

In the past 15 years a broad spectrum of inflammatory demyelinating polyradiculoneuropathies has been identified. GBS, or acute inflammatory demyelinating immune-mediated polynoeuropathy, is the most common form of the disease. GBS affects nerve roots and peripheral nerves, leading to motor neuropathy and flaccid paralysis with possible sensory and ANS effects.130 Purely motor forms and mixed motor and sensory forms of GBS have been identified.131 Unlike ALS, GBS usually has a good prognosis, with most patients returning to their prior functional status by 1 year after onset.

The incidence of GBS is approximately one to four cases per 100,000 persons. A variant form is acute motor axonal neuropathy, which, like GBS, has a good prognosis. Less common forms are acute motor and sensory axonal neuropathy, which has a less positive prognosis (and which some consider to be a distinct type of peripheral neuropathy); Miller-Fisher syndrome, with primarily cranial nerve symptoms, ataxia, and areflexia;132 and chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), which causes progressive relapsing and remitting numbness and weakness.133 Epidemiological studies show that males are affected by GBS twice as often as are females.134
Approximately 27% of patients with GBS have no identified preceding illness; however, more than two thirds had symptoms of an infectious disease 2 weeks before the onset of GBS symptoms. Although no consistent predisposing factors are known, evidence exists to support connections with Campylobacter jejuni, Mycoplasma pneumoniae, cytomegalovirus, and Epstein-Barr virus. In GBS the spinal roots and peripheral nerves are infiltrated with macrophages and T lymphocytes. Macrophages then attack and strip the myelin sheaths. In milder cases of GBS the axons are left intact and the nerves are remyelinated, typically in a matter of weeks. However, in some cases, the axons also degenerate, with recovery dependent on axonal regeneration from intact elements, which takes months and may be incomplete. In acute axonal motor neuropathy, macrophages invade the axon directly, leaving the myelin intact. Some evidence exists in a substantial number of patients with GBS that axonal loss is related to long-lasting or permanent muscle weakness.

Because of damage to the myelin sheath, saltatory propagation of the action potential is disturbed, resulting in slowed conduction velocity, dyssynchrony of conduction, disturbed conduction of higher frequency impulses, or complete conduction block. Partial conduction block is most often seen in the early stages of GBS, and the conduction block increases as the patient reaches a plateau. The most common conduction block findings are in the peroneal nerve, followed by the tibial nerve. Proximal conduction block is evident more often than distal conduction block. In axonal neuropathy, conduction block is more severe, and the number of functional motor units is decreased (Figure 17-4). The diagnostic criteria for GBS are detailed in Box 17-3.

**Clinical Presentation**

GBS in both children and adults is characterized by a rapidly evolving, relatively symmetrical ascending weakness or flaccid paralysis. Motor impairment may vary from mild weakness of distal lower-extremity musculature to total paralysis of the peripheral, axial, facial, and extraocular musculature. Severe fatigue is present in 38% to 86% of patients with GBS, depending on the cutoff point used to define severity and the age of the sample, with a positive correlation between severe fatigue and age. Tendon reflexes are usually diminished or absent. Twenty percent to 38% of patients may require assisted ventilation because of paralysis or weakness of the intercostal and diaphragm musculature. Impaired respiratory muscle strength may lead to an inability to cough or handle secretions and to decreased vital capacity, tidal volume, and oxygen saturation. Secondary complications such as infections or organ system failure lead to death in approximately 5% of patients with GBS. Approximately 35% to 50% of patients develop some cranial nerve involvement, primarily facial muscle weakness, although patients may also develop oropharyngeal and ocu-lomotor involvement.

ANS symptoms are noted in approximately 50% of patients. Low cardiac output, cardiac dysrhythmias, and marked fluctuations in blood pressure may compromise management of respiratory function and can lead to sudden death. Other typical ANS symptoms may result in peripheral pooling of blood, poor venous return, ileus, and urinary retention.

Sensory symptoms such as distal hyperesthesias, paresthesias (tingling, burning), numbness, and decreased vibratory or position sense are common. The sensory disturbances often have a stocking-and-glove pattern rather than the dermatomal distribution of loss. Although the sensory problems are seldom disabling, they can be disconcerting and upsetting to patients, especially during the acute stage.

Pain was identified as a significant presenting symptom reported in the original articles describing GBS. When
pain was prominent, patients spontaneously revealed its presence during a medical history. Therefore therapists who may be working with patients with an onset of low back pain not associated with known injury or stress and reports of paresthesias (pins and needles) and vibratory or decreased tendon reflexes should evaluate or monitor for possible GBS.\(^{149,150}\)

The most common description of presenting pain was of muscle aching typically associated with vigorous or excessive exercise. Pain was usually symmetrical and reported most frequently in the large-bulk muscles such as the gluteals, quadriceps, and hamstrings and less often in the lower leg and upper-extremity muscles. Some pain reported during late stages of the illness was described as “stiffness.” Pain was consistently more disturbing at night.\(^{150}\) As the disease progresses, some patients experience severe burning or hypersensitivity to touch or even air movement, which can interfere with nursing care and limit therapy interventions.

The types of pain reported include paresthesias, dysesthesias, axial and radicular pain, joint pain, and myalgias.\(^{151}\) Dysautonomia (orthostatic hypotension, blood pressure instability, cardiac arrhythmias and sometimes bowel and bladder dysfunction) is relatively common in patients with GBS requiring ventilatory support; in one prospective study of 297 patients, cardiac arrest associated with dysautonomia was the leading cause of death.\(^{137}\) In patients with paraplegia or quadriplegia, approximately one fourth had problems with urinary retention caused by detrusor areflexia or overactivity, overactive urethral sphincter, and disturbed bladder sensation.\(^{134}\) The possibility of deep vein thrombosis (DVT) and pulmonary embolus must also be monitored and prophylactic treatment used.\(^{152}\)

### Medical Prognosis

Although some patients have a fulminating course of progression with maximal paralysis within 1 to 2 days of onset, 50% of patients reach the nadir (the point of greatest severity) of the disease within 1 week, 70% by 2 weeks, and 80% by 3 weeks.\(^{145}\) In some cases the process of increasing weakness continues for 1 to 2 months. Onset of recovery is varied, with most patients showing gradual recovery of muscle strength 2 to 4 weeks after progression has stopped or the condition has plateaued. Although 50% of the patients may show minor neurological deficits (e.g., diminished or absent tendon reflexes) and 15% may show persistent residual deficits in function, approximately 80% become ambulatory within 6 months of onset of symptoms. The most common long-term deficits are weakness of the anterior tibial muscle and, less often, weakness of the foot and hand intrinsic muscles, quadriceps, and gluteal musculature. Three percent to 5% of patients die of secondary cardiac, respiratory, or other systemic organ failure.\(^ {134,151}\) Fatigue or poor endurance was also noted as a long-term consequence of GBS, possibly attributable to deconditioning and peripheral fatigue related to muscle fatigue during the healing process.\(^ {141,153}\) Vasjar and colleagues\(^ {154}\) also report that fatigue and poor exercise tolerance were common persisting symptoms in children who appeared to have fully recovered from acute GBS.

Although often not the focus of most studies on the long-term impact of GBS, sensory deficits (impaired response to pinprick, light touch, and vibration and proprioception in combination with other sensory losses) are an ongoing problem for patients 3 to 6 years after recovery from acute GBS. In a study of 122 subjects, 38% showed sensory deficits in the upper extremities\(^ {155}\) and 66% had ongoing sensory deficits of the lower extremities.\(^ {151}\) The muscle aches and cramps experienced by some of these patients appeared to be related to sensory rather than persistent motor dysfunctions as usually thought.

Overall, factors associated with a poor prognosis are severity of muscle weakness (especially quadriplegia), the need for respiratory support, cranial nerve involvement associated with loss of eye movement and swallowing, rapid rate of progression from onset, length of time to nadir, older age at onset, history of gastrointestinal illness, and recent cytomegalovirus infections.\(^ {134,142}\) In a prospective study of 297 patients with GBS in Italy, disease severity was not associated with time to clinical recovery, but it did predict ultimate outcome, along with shorter length of time to nadir, older age at onset, evidence of axon damage, and recent gastroenteritis.\(^ {137}\)

### Medical Management

Medical treatment depends on the rate and degree of ascending paralysis. Because most patients return to their prior functional status, excellent supportive care during the acute stage is imperative. Respiratory compromise should be expected, and all patients, including those with limited paralysis and sensory dysfunction, must be closely monitored for the rapid onset of pulmonary and cardiac decompensation or cardiac arrhythmias, paroxysmal or orthostatic hypotension, urinary retention, and paralytic ileus caused by dysautonomia.\(^ {152}\) Because of the possibility of sudden respiratory failure, patients with evidence of GBS must be hospitalized so that immediate cardiorespiratory support can be given if functional vital capacity (FVC) falls below 20 mL/kg or
oxygen saturation falls below 75%.\(^{144}\) Patients who progress to respiratory paralysis must be treated in an intensive care environment where adequate respiratory function can be maintained, secondary infections can be prevented or limited, and metabolic functions can be carefully monitored. The patient should be intubated if the FVC falls below 12 mL/kg or if the patient is increasingly dyspneic even if FVC is above the cutoff level.\(^{145,156}\) Twenty-five percent of patients who experience respiratory failure will develop pneumonia.\(^{151}\) Even if daytime respiration seems adequate, night-time respiratory insufficiency (sleep-disordered breathing) should be ruled out if patients have persistent sleepiness or fatigue.\(^{141}\)

Patients with GBS in the intensive care unit (ICU) on ventilation and with varying levels of paralysis and sensory dysfunction feel trapped and out of control because they cannot express their needs. These patients can usually hear well and most can see what is happening around them. They benefit from being oriented to time, having the personnel explain all procedures, and having some means of obtaining help. Therapists can work with the ICU staff to provide the patient with alternative forms of communication, such as eye blink, clicking, and communication boards designed for their needs. Having some form of communication and knowing that they will not be left alone will help prevent traumatic stress reactions.\(^{152}\)

In addition to the intensive monitoring of progression and supportive care required for patients with GBS, two specific immunotherapy-based treatments—plasma exchange (removal of plasma from withdrawn blood with retransfusion of the formed elements back into the blood) and intravenous immunoglobulin (IVIg) (taking blood from a vein, separating plasma, and returning the blood cells with a plasma substitute)—have been under investigation for their ability to decrease the duration of respirator dependence and the time to onset of improvement. Systematic reviews of these interventions as of 2010 have found that plasma exchange decreases recovery time and is most beneficial if begun within the first week of diagnosis and can be beneficial up to 30 days after diagnosis.\(^{157}\) Plasma exchange is also cost-effective as used in patients with mild, moderate, or severe courses of GBS.\(^{158}\) IVIg is somewhat safer and easier to administer than plasma exchange; IVIg speeds recovery by the same amount of time as plasma exchange and is more effective than supportive care only. Adding IVIg to plasma exchange did not improve time to recovery any more than either treatment alone.\(^{159}\) High-quality evidence is available to support IVIg use in adults with GBS; the quality of evidence is slightly less high to support its use in children with GBS.\(^{160}\)

Although corticosteroids have been used to decrease the inflammatory process in GBS since the 1960s, a review of clinical studies of corticosteroid effectiveness showed that corticosteroid treatment alone does not hasten recovery from GBS.\(^{161}\) Hughes and colleagues have developed practice parameters associated with these findings.\(^{162}\)

**Therapeutic Management of Movement Dysfunction Associated with Guillain-Barré Syndrome**

Therapeutic management of the movement deficits associated with GBS includes supportive management during the acute phase, prevention of long-term medical comorbidities during the acute through early recovery stages, and rehabilitation throughout recovery.\(^{163}\) With the assumption that the patient will have significant return of function within months, therapists must help maintain the integrity of functioning systems, address pain, teach compensatory strategies, and appropriately promote increasing activity after the plateau. The immediate needs of the patient will change as the patient moves through the acute stage, the plateau at the nadir, and the recovery stage of GBS before and after muscles attain antigravity strength. Transitioning between the changes in immediate therapeutic goals necessitates careful examination of the current status, progression of disease, and needs of the patient.

**Examination**

A comprehensive examination of the patient’s movement and function includes factors shown in Box 17-4. The extent of the examination in any one session depends on the patient’s condition and ability to participate. History taking should include the course of the disease, along with any recent illness, preexisting neuromotor or other medical conditions, current concerns, and the patient’s immediate goals. Screening tests can help determine whether sensory and autonomic systems are involved along with motor systems. Checking vital signs at rest and immediately after activity, assessing skin integrity especially in immobile patients, screening cranial nerve performance, and noting communication ability are all important components. Additional testing of sensation (and documentation on a body chart, for example) or autonomic systems may be required if the screening tests indicate.

In GBS, assessment of muscle strength and ROM as specifically as possible is important so the patient’s course of progression or improvement can be tracked, possible patterns leading to contractures can be predicted and prevented, and the appropriate level of exercise can be implemented. MMT, dynamometry, or isokinetic testing could be useful in various stages; goniometry is typically used for ROM testing. Full MMT and joint ROM may require several sessions in the initial stages, and a few specific muscles and joints may be selected (e.g., sternocleidomastoids, deltoids, triceps, flexor carpi ulnaris, lumbricals, iliopsoas, gluteus medius, anterior tibialis, flexor hallucis longus; shoulders, fingers, ankles) to test for changes weekly.

Several factors may interfere with complete assessment in the initial stage. Patients who report considerable pain during handling or active movement may not tolerate or may be unwilling or unable to cooperate with testing. The therapist should track the patient’s level of pain, for example, on a numerical rating of pain scale, to help distinguish between weakness and loss of ROM related to pathological condition, immobility, or pain.

Fatigue and respiratory difficulties may also preclude complete strength assessment in a single session. Fatigue may result from deconditioning, increased effort required to perform similarly with weakened muscles, and inability to recruit sufficient motor units to maintain contractions.\(^{164}\) Fatigue can be documented in relation to amount of activity tolerated (with specific symptoms noted before rest is required) or with a questionnaire such as the Fatigue Severity Scale (FSS), Fatigue Impact Scale (FIS), or the Visual Analogue Scale for Fatigue (VAS-F).\(^{143}\) Functional tests may include standardized scales of independence in ADLs or balance, tests of manual dexterity, and temporal measures.
of gait. Chehebar and colleagues\textsuperscript{163} review some of the pros and cons of standardized tests such as the Barthel Index, modified Hughes scale of GBS disability, and the Functional Independence Measure. Health-related quality-of-life measures used in related populations include the Nottingham Independence Measure. Health-related quality-of-life measures used in related populations include the Nottingham Independence Measure. Health-related quality-of-life measures used in related populations include the Nottingham Independence Measure. Health-related quality-of-life measures used in related populations include the Nottingham Independence Measure. Health-related quality-of-life measures used in related populations include the Nottingham Independence Measure. Health-related quality-of-life measures used in related populations include the Nottingham Independence Measure.

Changes in the patient's condition should be monitored with serial MMT, ROM assessments, sensory testing, and functional status examinations. See Karni and colleagues\textsuperscript{166} for suggestions on serial functional assessments. Before the patient is discharged from the hospital or rehabilitation unit, therapists should complete an assessment of the patient's home environment so that appropriate safety and adaptive equipment can be in place in time for the patient's return home.

**Respiratory and Dysphagia Examination**

Therapists are usually involved early in the care of patients with GBS. For patients with respiratory or bulbar paralysis, the therapist's initial contact may be in the ICU. Although most hospitals have fully equipped ICUs, a therapist working in a rural or smaller community hospital may be the first person to note a patient's changing respiratory status during an evaluation and treatment session for muscle weakness or back pain. Therefore the therapist must be prepared to advise nursing and medical staff about the need to test oxygen saturation levels and FVC. Therapist attention to respiratory complications is particularly important in the managed care environment, which discourages hospitalization if presenting symptoms are not life endangering.\textsuperscript{145} A simple estimate of FVC can be done at bedside. If after taking a large breath the patient can count out loud only to 10, the forced vital capacity is approximately 1 L and intubation should be considered. Complete information on the PT's evaluation of patients in acute respiratory failure is provided by Irwin and Tecklin.\textsuperscript{167}

Patients who have been intubated or who have cranial nerve involvement with oral motor weakness commonly have a high incidence of aspiration. Patients with severe oral-motor problems and dysphagia should be evaluated thoroughly and treated by a therapist skilled in oral-motor dysfunction and feeding. This may be a speech therapist, OT, or PT depending on the facility. Patients with a feeding tube (PEG) should receive their feedings in a relatively upright position and should remain in that position for 30 to 60 minutes after feeding to decrease the chance of aspiration. According to Logemann,\textsuperscript{168} approximately 40% of patients receiving bedside swallowing assessments have undetected aspiration. Therefore the bedside evaluation should be considered only a preliminary step in the diagnostic process. In addition to careful assessment of oral-motor control, some clinicians recommend cervical auscultation to listen to swallowing sounds, particularly during the acute phase of the illness.

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**BOX 17-4 □ FACTORS TO CONSIDER IN THE EXAMINATION OF PATIENTS WITH GUILLAIN-BARRÉ SYNDROME**

<table>
<thead>
<tr>
<th>HISTORY</th>
<th>MOTOR FUNCTION</th>
<th>SENSORY SYSTEM</th>
<th>AUTONOMIC SYSTEM</th>
<th>PSYCHOSOCIAL SYSTEMS</th>
<th>ELECTRODIAGNOSTIC TESTING</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patterns and sequence of symptom onset</td>
<td>Visual inspection to identify symmetry of muscle bulk and function</td>
<td>Identify pattern of sensory loss or changes (use body chart)</td>
<td>Blood pressure resting and immediately after activity (prone, sitting, standing, if possible)</td>
<td>Identify patient and family concerns in acute circumstances and concerns about long-term issues that may affect patient and family. Assessment need not be extensive if referral can be made for social service evaluation of patient and family financial concerns, day-to-day living problems (e.g., transportation, child care), support systems, and coping strategies.</td>
<td>Nerve conduction velocity. (Physician will order these studies to be performed by a clinician skilled in the procedures. This may be a physical therapist, physician, or technician depending on facility.)</td>
</tr>
<tr>
<td>Recent illness or injury, prior episodes of sensorimotor problems</td>
<td>Myotatic reflexes, rule out tonic reflexes</td>
<td>Identify specific type of sensory change (e.g., paresthesias, anesthesia, hypesthesias) (use body chart)</td>
<td>Heart rate resting and immediately after activity, dysrhythmias</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Manual muscle testing, carefully identifying pattern of weakness (testing should be as muscle specific as possible rather than assessing muscle groups only; use form for serial recording)</td>
<td></td>
<td>Body temperature stability</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Presence of muscle fasciculations</td>
<td></td>
<td>Bowel and bladder control</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>Cranial nerves</td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Range of motion (use form for serial recording)</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td></td>
<td>Equilibrium reactions sitting and standing (if testable)</td>
<td></td>
<td></td>
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<td></td>
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<tr>
<td></td>
<td>Current functional status (activities of daily living, including bowel and bladder function, ambulation)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Endurance and experienced fatigue</td>
<td></td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

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With evidence of swallowing difficulties and possible aspiration, the patient should be referred for comprehensive testing with videofluoroscopy. Swallowing can also be assessed by techniques such as fiberoptic endoscopy, ultrasound, electroglottography to determine laryngeal movement, and scintigraphy, which involves scanning a radioactive bolus during swallowing.\(^\text{169}\) (Refer to section on medical management of ALS for suggestions for dealing with dysphagia.)

### Intervention Goals

General goals for the care of the patient with GBS, to be specified with reference to the patient’s preferences, include the following:

- Facilitate resolution of respiratory problems and dysphagia
- Minimize pain
- Prevent contractures, decubitus ulcers, and injury to weakened or denervated muscles
- Introduce a graduated program of active exercise while monitoring overuse and fatigue
- Resume psychosocial roles and improve quality of life

### Therapeutic Interventions

In a Cochrane review of exercise in people with peripheral neuropathies, no randomized or quasi-randomized controlled trials were identified for patients with GBS as of September 2009.\(^\text{135}\) However, some treatment programs used for patients with other neuromotor dysfunctions can be adapted for use with patients with GBS.

### Respiratory and Cranial Nerve Dysfunction

Depending on the facility, PTs may be involved in the respiratory care of patients with GBS. PTs may conduct chest percussion, breathing exercises, resistive inspiratory training, or strict protocols to prevent overfatigue of respiratory muscles while weaning patients from mechanical ventilation.\(^\text{170}\) Goals of treatment are related to increasing ventilation or oxygenation, decreasing oxygen consumption, controlling secretions, and improving exercise tolerance. See Irwin and Tecklin\(^\text{167}\) for coverage of treatment programs and techniques appropriate for the GBS patient with acute or residual respiratory dysfunction.

When patients are placed on mechanical ventilators, communication can be difficult and frustrating.\(^\text{171}\) The rehabilitation team can help develop and execute alternative means of communication.

In the more severe cases of GBS, cranial nerve involvement can lead to multiple complications such as dysphagia and vocal cord paralysis. In many facilities, speech pathologists or OTs are responsible for establishing a dysphagia treatment program. Therapists responsible for treatment of patients with dysphagia and swallowing problems should refer to Logemann’s classic text on the evaluation and treatment of swallowing disorders.\(^\text{168}\) Therapeutic goals are the prevention of choking and aspiration and the stimulation of effective swallowing and eating. The act of chewing and swallowing is complex and requires coordinated reflexive and conscious action. Intervention is focused on positioning (upright with head tilted slightly forward),\(^\text{171}\) head control, and oral-motor coordination (e.g., sucking an ice cube, stimulating the gag response, facilitating swallowing with quick pressure on the neck and thyroid notch timed with intent to swallow). A conscious swallowing technique is introduced with thick liquids and progressed to thinner liquids after the patient’s oral-motor coordination response is enough to control movement of fluids. Once the patient has good lip closure, fluids should be introduced one sip at a time from a straw cut to a short length to minimize effort. Semisoft, moist foods are gradually introduced (pasta, mashed potatoes, squash, gelatin). Any crumbly or stringy foods (coffee cakes, cookies, snack chips, celery, cheeses) should be avoided, and the patient should not attempt to talk or be interrupted during eating until choking does not occur and swallowing is comfortable and consistent.\(^\text{172}\) Feeding training should occur during frequent, short sessions to prevent fatigue. Therapists should be prepared to use the Heimlich maneuver if choking occurs or have a suction machine available at bedside.

### Pain

If pain seems to be a major factor limiting the patient’s passive or active motion, the treatment team should determine the best approach to alleviate pain. According to one study, patients with GBS did not seem to show a consistent response to any specific pain medication, although six of the 13 patients seemed to have a positive response to codeine, oxycodone plus acetaminophen (Percocet), and oxycodone plus aspirin (Percodan).\(^\text{147}\) Some patients may find relief with medications used to treat neurogenic pain, such as the tricyclic antidepressants, carbamazepine, or gabapentin (anticonvulsants).\(^\text{151}\) For patients who do not respond to conventional analgesics or tricyclic antidepressants, a short course of high-dose corticosteroids can lead to pain relief.\(^\text{144}\)

Some patients with neuropathy have noted decreased pain after using transcutaneous electrical nerve stimulation (TENS).\(^\text{173,174}\) Although no study has examined the effect of TENS specifically on pain associated with GBS, it might be a treatment option to help with desensitization in patients whose pain is not controlled with passive movement or pain medications.

Another option is capsaicin, the active ingredient in chili peppers, which when applied topically interacts with the sensory neurons to relieve pain from peripheral neuropathies.\(^\text{151}\) Therapists, wearing gloves, apply a topical anesthetic until the area is numb. The capsaicin is then applied topically. The capsaicin remains on the skin until the patient starts to feel the heat, at which point it is promptly removed. Because the nerves are overstimulated by the burning sensation, the sensory gateway is unable to report pain for an extended period.\(^\text{175}\)

Some patients who experience extreme sensitivity to light touch, such as from movement of sheets, air flow, and intermittent touch contact, benefit from a “cradle” that holds sheets away from the body. Some find relief if the limbs are wrapped snugly with elastic bandages, which provide continuous low pressure while warding off light and intermittent stimuli. Alternatively, the patient’s pain response can be desensitized through methodical stimulation with frequent, consistent stimuli to the affected area for short durations to allow acclimatization.\(^\text{170}\)

### Contractures, Decubitus Ulcers, and Injury to Weakened or Denervated Muscles

#### Positioning

In the acute stage of GBS, rehabilitation will focus on positioning and passive ROM to prevent contractures and decubitus ulcers.\(^\text{176}\) Preventing pressure sores...
should consider how best to maintain the physiological position of the hands and feet. Research has shown that mild continuous stretch maintained for at least 20 minutes is more beneficial than stronger, brief stretching exercises. Thus the use of splints for prolonged positioning is superior to the use of short bursts of intermittent, manually applied passive stretching for maintaining functional range. Although some facilities still use a footboard to control passive ankle plantarflexion, most therapists now use moldable plastic splints that can be worn when the patient is in any position. Because ankle-foot splints often prevent visual inspection of the heel position, care must be taken to ensure that the heel is firmly down in the orthosis and that the strapping pattern is adequate to secure the foot. The strap system must be simple enough to be positioned properly by all staff and family members caring for the patient. The ankle-foot splint should extend slightly beyond the end of the toes to prevent toe flexion and skin breakdown from the toes rubbing on sheeting. Care should be taken not to compress the peroneal nerve with the splint as it crosses the fibula, a particularly vulnerable area after the loss of muscle mass in the lower legs from the GBS. Wrist and hand splints may be prefabricated, resting-style splints, or molded to meet the patient’s specific needs. Because spasticity is not a problem in the patient with GBS, a simple cone or rolled cloth may be adequate to maintain good wrist, thumb, and finger alignment for short-term immobility.

Range of Motion. To be effective, the ROM program must start within the first couple of days of hospitalization and include both accessory and physiological motions to increase circulation; provide lubrication of the joints; and maintain extensibility of capsular, muscle, and tendon tissue. Passive ROM exercises to the ends of normal range for all extremity joints, fingers and toes, neck, and trunk should be performed twice daily—more frequently if the patient has no active movement. Patients can be instructed to perform the ROM exercises themselves if they can move actively without pain or fatigue; during the acute stage of declining strength, they should be observed during ROM activities to ensure adequacy of the range and any changes in quality of movement. If the patient cannot complete movement through full range independently, a therapist or well-instructed and monitored caregiver can assist the patient in moving to the end of range. This may not be easy if the patient has pain with motion. Knowing whether to “push through the pain” or stay within the limits of pain is often a great dilemma for the therapist. The therapist needs to find a balance between working for full joint range and reacting to the patient’s reports of pain. If the ends of ranges start to become stiff, stretch should be slow and sustained at the end point for 10 to 30 seconds.

Denervated or weakened muscles can be injured easily; therefore the therapist is responsible for ensuring that joint structures are not damaged and that ROM activities are done with appropriate support of the limb to prevent sudden overstretching. Instruction to caregivers regarding passive ROM activities must include details such as externally rotating the shoulder during abduction to prevent impingement and ensuring that the subtalar joint is in the neutral position during dorsiflexion to avoid overstretching of the midfoot. In hospitals where the patient is treated by a changing therapy or nursing staff or by family members, a positioning schedule with diagrams, a splinting plan, and ROM recommendations should be presented in poster format at the patient’s bedside to facilitate consistent treatment.

ROM can usually be maintained with standard positioning and ROM programs. Nevertheless, some patients, especially those who have reported severe extremity and axial pain early during the disease process and those who have been quadriplegic and respirator dependent for prolonged periods, may develop significant joint contractures despite preventive interventions. As with patients with spinal cord or severe head injuries, heterotopic ossification has been reported in patients with GBS. Meythaler and colleagues note that early mobilization was related to therapeutic decreases in serum calcium levels and suggest that aggressive ROM (but not hard or abrupt movements that may injure the muscle) may impede the effects of heterotopic bone overgrowth, which can have a severe impact on ROM. Once heterotopic ossification has been identified, treatment includes modification of ROM exercise to use only active and passive motion within the pain-free arc.

Soryal and colleagues reported on three patients with GBS who had marked residual contractures that limited function after strength improved. None of the patients had radiological signs of erosive arthropathy or inflammatory joint disease. Soryal hypothesized a number of possible mechanisms for the limitations in ROM: (1) therapists and nurses may have been reluctant to take patients who reported marked pain during passive movement through the full ROM; (2) the contractures may have been a result of pain or damage caused by inappropriate excessive passive movement of hypotonic and sensory-impaired joints and muscles (often caused by poor movement of the patient in bed or by poorly trained staff or family members moving limbs); (3) the paralysis may have resulted in lymphatic stasis with accumulation of fluid in tissue spaces and nutritional disturbances; and (4) vasomotor disturbances resulting from autonomic neuropathy may have led to adhesions and fibrosis. Although the authors found few reports describing contractures as a significant residual problem, they suggested that ROM programs must be defined precisely as to frequency and duration, particularly for patients reporting early joint pain.

Some patients will prefer to position their limbs so muscle and tendons are in the shortened range in an attempt to decrease muscle pain. This may lead to capsular contractures. The therapist should be aware of changes in “end feel” over time when testing ROM of each joint to determine if capsular and ligamentous structures are also becoming more restricted as the muscle and tendon tissue shortens. Patients
who have intact sensation of pain and temperature may respond positively to the use of heat (up to approximately 45° C or 113° F) before stretching to decrease muscle pain and facilitate tissue elongation before stretching. Several basic studies of rat tail tendon and the relation between load and heat have shown that attenuating permanent length increases in collagenous tissue is possible with a combination of heat and stretch.181-184 (Caution: Heat should not be used on a patient with a sensory deficit that inhibits ability to distinguish differences in temperature.)

On the basis of evidence that continuous passive motion (CPM) is effective in maintaining joint range in both rabbits and human beings,185 Mays186 described a case study of a patient with GBS (quadriplegia with 7 days of mechanical ventilation) who had persistent pain and stiffness of the upper extremities and fingers approximately 3 months after the onset of GBS. CPM of the hands and fingers was added to a program of occupational therapy that included ROM, splinting, and ADLs. The author reported an increase in the rate of recovery of finger range and a decrease in pain after use of CPM. Numerous other studies have reported the value of CPM in maintaining or increasing ROM after hip and knee surgery. It may be a useful adjunct to traditional therapy for patients with GBS, especially those who continue to develop contractures with standard, intermittent ROM programs. Patients with severe paraparesis or dysesthesias may not be able to tolerate CPM equipment.

Massage also may play a positive role in maintaining muscle tissue mobility and tissue nutrition while limiting the amount of intramuscular fibrosis development. The use of massage in patients with GBS has not been reported; however, it makes intuitive sense that it may be a useful adjunct to ROM exercises in patients who do not have marked hypersensitivity to touch, significant muscle pain, or a history of DVT. Patients with or without a history of DVT who are immobile for long periods or who have concomitant cardiac illnesses may have marked swelling of the distal limbs. After medical clearance, edema-specific massage and limb-elevation techniques may be useful if tolerated by the patient. Early active ROM exercises creating “muscle pumping” contractions in muscles with at least fair strength can help prevent uncomfortable edema.

**Progressive Program of Active Exercise while Monitoring for Overuse and Fatigue**

Although most patients with GBS recover from the paralysis, the course and rate of recovery may vary significantly among patients. The decline of strength may take 2 days to 4 weeks, with a plateau of a few days to a few weeks after the nadir. Strength returns over the course of weeks to months, depending on whether the disease process affected only myelin or the axons themselves. Strength usually returns in a descending pattern—opposite to the pattern noted during onset of the disease. No evidence exists to indicate that active exercise can change the rate of progression of the disease or regrowth of myelin or axons, although it may improve function through increased strength and aerobic capacity once muscles are reinnervated. The major goal of therapeutic management throughout the course of GBS must be to maintain the patient’s musculoskeletal system in an optimal ready state, prevent overwork, enhance circulation and cardiorespiratory endurance within the limits of active movement, and pace the recovery process to obtain maximal function as reinnervation occurs.

In the acute stage of GBS, active exercise is limited to whatever the patient can move without pain or excessive fatigue. Slings or adaptive devices may help support the weight of a limb to continue active movement in a gravity-eliminated plane for those muscles that have lost antigravity strength. As the disease reaches its nadir, activity remains limited. Once weakness stops progressing, passive maintenance of ROM may be the only activity possible for immobile patients. As strength begins to return after the plateau, therapists must prescribe limited amounts of low-resistance activities, with strict avoidance of antigravity strain on the muscles until strength reaches the 3/5 (Fair) range of MMT. Active exercise can be added very slowly, with frequent rest periods and monitoring to avoid fatigue.175,185 Activity should be halted at the first point of fatigue or muscle ache; abnormal sensations (tingling, paresthesias) that persist for prolonged periods after exercise may also indicate that the exercise or activity level was excessive. Any progression of resistance or repetitions of strengthening exercises should be monitored for 3 to 7 days for increase in weakness, muscle spasms, or soreness before exercises are progressed further.186 If additional weakness or soreness ensues, the additional activity must be eliminated for several days, with reinitiation at a lower level of resistance or number of repetitions and more gradual increase. Work simplification and energy conservation strategies may be useful to improve function in the recovery stage of GBS.170 As strength increases, additional resistance may be applied to those muscles showing good recovery while avoiding strain on muscles that have not yet reached the same level, frequently the most distal musculature. Even when strength has returned throughout, rehabilitation and exercise may need to continue to address fatigue that may persist at each of the International Classification of Functioning, Disability and Health (ICF) levels: body function and structure, activity, and participation.141 For an example of treatment progression during the acute stage from week 1 through week 12, see Figure 17-3. After the initial stages of upright activity after any period of bed rest, therapists must progress patients with GBS very carefully because 19% to 50% of this population show orthostatic hypotension along with dysautonomia.139,140 A program to improve tolerance to upright position can be started in the ICU if the patient is on a circle electric or Nelson standing bed. If a standing bed is not available, a sitting program can be initiated as soon as it is tolerated. A progressive standing program can be instituted when the patient’s respiratory system and ANS are no longer unstable and the patient can be moved to a tilt table. Caution should be taken to stabilize the patient fully to maintain alignment and to limit activity in muscles having strength below the fair range. When beginning training, some patients benefit from using an abdominal binder or foot-to-thigh compression stockings if tolerated. Because of the relation between poor hydration and hypotension, therapists must ensure the patient is well hydrated before beginning upright or standing tolerance programs.139

As was discussed in the section on therapeutic considerations for patients with ALS, a muscle that has significant denervation is more likely to respond to exercise with overwork fatigue (see Figure 17-3 for the therapeutic window for exercise). Studying the effect of exercise on rat muscle after nerve injury, Herbison and colleagues187 identified a loss of contractile proteins during initial reinnervation. After reinnervation the same amount of exercise resulted in muscle
Rehabilitation Management of Clients with Neurological System Pathology

Hypertrophy. Bensman reported on eight patients who had stabilized after acute polyradiculoneuritis (among them patients with GBS). All eight patients had a temporary loss of function after strenuous physical exercise. Three patients apparently had significant decreases in strength. All patients were then placed on a program of passive ROM exercises, and an increase in muscle strength was noted. Recurring episodes of a temporary loss of function appeared to be related to strenuous exercise and fatigue. The current position for patients with GBS, then, is that excessive exercise during early reinnervation when only a few functioning motor units are present can lead to further damage rather than to the expected exercise-induced hypertrophy of muscle.

During the initial stages of exercise, the repetitions per exercise period should be low and the frequency of short periods of exercise should be high. As reinnervation occurs and motor units become responsive, the early process of muscle reeducation exercise used by the therapist may be similar to that used after polio. To encourage active contraction of the muscle the therapist should carefully demonstrate to the patient the expected movement. The therapist then passively moves the patient’s limb while the patient observes. After gaining a clear picture of what movement is expected, the patient is encouraged to control muscles. Facilitatory techniques such as skin stroking, brushing, vibration, icing, and tapping may be used in conjunction with the muscle reeducation process if the sensory and pain status of the patient permits. The patient is taught to reassess his or her movements and make corrective responses. As the patient gains strength, the movements are translated into functional activities.

### TABLE 17-3 MEDICAL STATUS OF PATIENTS WITH GUILLAIN-BARRÉ SYNDROME AND POSSIBLE TREATMENT OUTLINE

<table>
<thead>
<tr>
<th>MEDICAL STATUS</th>
<th>TREATMENT*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tracheostomy</td>
<td>Week 1:</td>
</tr>
<tr>
<td>Respirator dependent</td>
<td>Postural drainage every 3 hours around the clock</td>
</tr>
<tr>
<td>Complete cranial nerve paralysis</td>
<td>Passive ROM exercises to all joints</td>
</tr>
<tr>
<td>Quadriplegia</td>
<td>Splinting (molded plastic) of hands and feet to maintain functional position</td>
</tr>
<tr>
<td></td>
<td>Positioning, splinting, and ROM program schedule posted at bedside</td>
</tr>
<tr>
<td></td>
<td>Weeks 2-5:</td>
</tr>
<tr>
<td></td>
<td>Postural drainage decreased to two times each shift (every 8 hours)</td>
</tr>
<tr>
<td></td>
<td>Passive ROM exercises, physiological and accessory motions, gentle stretching of intercostal musculature, trunk rotations</td>
</tr>
<tr>
<td></td>
<td>Continue splinting and positioning program</td>
</tr>
<tr>
<td></td>
<td>Family education: family members taught gentle physiological ROM techniques, with attention to correct shoulder patterns and simple massage techniques</td>
</tr>
<tr>
<td></td>
<td>Weeks 6-7:</td>
</tr>
<tr>
<td></td>
<td>Postural drainage two times each shift (every 8 hours)</td>
</tr>
<tr>
<td></td>
<td>Continue ROM program, splinting, and positioning</td>
</tr>
<tr>
<td></td>
<td>Begin to build tolerance of upright sitting with good trunk alignment</td>
</tr>
<tr>
<td></td>
<td>Begin facilitation of active facial and tongue muscle activity in patterns necessary for swallowing, eating, and speaking; speech pathology, occupational therapy consultation for dysphagia training</td>
</tr>
<tr>
<td></td>
<td>Family members active in care, helping with ROM, splinting, and positioning schedule as they choose</td>
</tr>
<tr>
<td>Respirotor set on intermittent mandatory ventilation</td>
<td></td>
</tr>
<tr>
<td>Weaning to respirator at night by end of week 7</td>
<td></td>
</tr>
<tr>
<td>No active muscle contractions except eye opening and lip movements</td>
<td></td>
</tr>
<tr>
<td>Dysphagia</td>
<td>Weeks 8-12:</td>
</tr>
<tr>
<td></td>
<td>Postural drainage one time each shift</td>
</tr>
<tr>
<td></td>
<td>Chest stretching, breathing exercises</td>
</tr>
<tr>
<td></td>
<td>Dysphagia program in collaboration with speech consultant</td>
</tr>
<tr>
<td></td>
<td>Muscle reeducation program with electromyographic biofeedback progressing to gravity-eliminated exercises using suspension slings attached to bed</td>
</tr>
<tr>
<td></td>
<td>Tilt-table standing program to increase tolerance to upright (wearing positioning splints if necessary)</td>
</tr>
<tr>
<td></td>
<td>Collaborate with occupational therapist for treatment in wheelchair with suspension slings to facilitate active arm motion in gravity-limited position</td>
</tr>
<tr>
<td></td>
<td>Exercise, rest, positioning schedule posted</td>
</tr>
<tr>
<td></td>
<td>Family, patient educated about stimulating activity level to prevent fatigue, overuse of reinnervating muscles</td>
</tr>
</tbody>
</table>

ROM, Range of motion.
*Treatment depends on rate of recovery.
with impaired judgment often need a strict schedule of rest and activity. Patients and staff also need to be reminded that prolonged sitting in bed or in a wheelchair, even when supported, may tax the axial musculature. A program of gradual sitting should be instituted, with the final goal being independent, unsupported sitting with functional equilibrium reactions. In busy hospitals a schedule of sitting and activity should be posted in clear view at the patient’s bedside.

As reinnervation progresses and strength and exercise tolerance increases, the therapist may choose to use facilitative exercise techniques such as neurodevelopmental sequencing\textsuperscript{189} or PNF\textsuperscript{190,191} to recruit maximal desired contraction of specific muscle groups. Although PNF techniques are excellent for eliciting maximal contraction, care must be taken not to overwork the weaker components of the movement pattern. A positive aspect of PNF techniques is that they can be tied in with functional patterns such as rolling, which is necessary for bed mobility, transitions to quadruped, kneeling, sitting, standing, and gait.

Because patients with GBS are transferred from acute care facilities to rehabilitation, skilled nursing, or home environments more quickly than in the past, therapists must be careful to document any serial negative changes or plateaus in motor, sensory, or respiratory impairments or functional status that may herald a relapse.\textsuperscript{192} Although 65% to 75% or more of patients with GBS show a return to clinically normal motor function, 2% to 5% of patients have a recurrence of symptoms similar in onset and pattern to the original illness.\textsuperscript{193} Recurrence of symptoms should trigger immediate cessation of activity and possibly medical reassessment in case of respiratory insufficiency.

Anecdotal and empirical evidence shows that patients with GBS can continue to show deficits during strenuous exercises that require maximal endurance. Four soldiers who were considered clinically recovered from GBS (normal motor power with or without reappearance of reflexes and the absence of sensory impairment) were unable to pass the Army Physical Fitness Test (APFT), which is designed to measure a minimal acceptable age-related level of physical fitness for military duty (maximal effort to challenge respiratory and muscular endurance, strength, and flexibility). Before onset of GBS, the four patients had all exceeded the APFT standards. None was able to pass the APFT as long as 4 years after the illness, indicating that the persistent deficit interfered with their ability to continue their military careers.\textsuperscript{195} The possibility of long-term endurance deficits should be considered when patients appear to have reached full recovery but report difficulty when returning to work or activities that require sustained maximal effort.\textsuperscript{194,195}

So far, no pharmaceutical agents have been helpful in alleviating fatigue in this population. In a study of the use of amantadine to relieve severe fatigue in 74 patients with GBS randomly allocated to treatment or placebo groups, the groups showed no difference in any of the primary or secondary measures recorded.\textsuperscript{196} Determining the effectiveness of interventions to affect fatigue may be complicated by differences in measures of experienced fatigue (subjectively reported) versus physiological fatigue (central or peripheral reduction in voluntary muscle force production) and the weak relationship between these in many neuromuscular disorders.\textsuperscript{197}

Cardiovascular fitness may also be compromised after recovery from GBS. This may be caused by altered muscle function, but it is also related to deconditioning from an imposed sedentary lifestyle.\textsuperscript{154} Several studies have reported the effect of endurance exercise training after GBS. In one case study a 23-year-old woman with a chronic-relapsing form of GBS with onset at age 15 years was placed on a walking and cycling program at 45% or less of her predicted maximal heart rate reserve. The low-intensity exercise program was selected to prevent possible fatigue-related relapse. After the program, the subject had improved her walking time 37%, walking distance approximately 88%, and cycle ride time more than 100%. Although no standardized or formalized recording of functional level was recorded before and after the exercise program, the patient reported that her energy level for ADLs was a “little higher” and that stair walking was easier.\textsuperscript{198} In another single-subject study of a 54-year-old man 3 years after onset of GBS with residual weakness, the authors demonstrated similar improvements in cardiopulmonary and work capacities as well as leg strength after a 16-week course of a thrice-weekly aerobic exercise program. The subject also reported expanded ADL capabilities. The authors suggested that their training regimen may disrupt the cycle of inactivity after recovery from GBS that leads to disuse atrophy and further deconditioning in patients with mild residual weakness.\textsuperscript{198} Fehlings and colleagues\textsuperscript{199} tested muscle strength and endurance in a group of children at least 2 years after acute onset of GBS. Although the children appeared essentially recovered, endurance of the arm muscles was lower than that of the lower extremities. They hypothesize that the typical walking, running, and cycling activities that the children participated in were sufficient to improve strength and endurance of lower-extremity muscles, and they recommended that children be encouraged to participate in activities such as swimming to improve upper-extremity endurance. Controlled tetherball and volleyball activities are also appropriate. Tuckey and Greenwood\textsuperscript{200} reported positive results of treatment with partial body-weight support (PBWS) treadmill exercise for a patient with severe GBS. Garssen and colleagues\textsuperscript{201} reported a 20% reduction in fatigue levels, along with improved physical condition and strength, after a 12-week intensive bicycling exercise program for patients several years after the onset of GBS.

Improvements in strength and endurance after GBS may continue for months to years. A prospective study following 6 patients for 18 months after onset of GBS recorded continuing improvement of muscle strength on average throughout the assessment period, and yet the average strength of major muscle groups had not yet reached that of healthy controls.\textsuperscript{202} Although the traditional thought has been that little clinical improvement occurs after 2 to 3 years, Bernsen and colleagues\textsuperscript{203} found that 21% of the patients in a study of 150 patients after recovery from acute GBS reported improvement after 2.5 to 6.5 years, although the authors thought the perception of improvement was related to improved sensory function. Of future research and clinical interest are the long-term consequences of GBS and how the normal aging process will affect patients who have some mild residual effects—for example, whether some patients will develop increasing weakness over time similar to persons with postpolio syndrome.\textsuperscript{139}

For those patients who experience significant losses in proprioception after GBS, sensory reintegration activities and high repetitions of task practice may help to redevelop motor engrams that are based on the altered sensory perception.\textsuperscript{139}

Patients with GBS have a significantly reduced health-related quality of life compared with control subjects at
approximately 1 year after onset, associated with decreased functional scores and changes in work status. Although physical training may be expected to improve functional scores and work capabilities, Bussmann and colleagues found little correlation between physical fitness and other domains. They hypothesized that training has psychological components, such as positive effect on mood and self-confidence, that influence quality of life in addition to physical changes.

Adaptive Equipment and Orthoses

Judicious use of orthotic devices and adaptive equipment should be considered an integral part of the rehabilitation process. The purpose of the orthotic and adaptive devices is twofold: (1) to protect weakened structures from overstretch and overuse and (2) to facilitate ADLs within the limits of the patient’s current ability. Orthotic devices and adaptive equipment should be introduced and discontinued on the basis of serial evaluations of strength, ROM, and functional needs. For example, a hospitalized patient who has poor (2/5) middle deltoid strength may practice upper-extremity activities such as eating while using suspension slings. A thumb position splint may be used temporarily to aid thumb control in grasping tasks.

Most patients will need a wheelchair for several months until strength and endurance improve. As strength returns, patients recovering from severe paralysis may need to change from use of a wheelchair with a high, reclining back with a head rest to use of a lightweight, easily maneuverable chair. A quandary for the therapist is to predict how long a wheelchair will be necessary and whether it should be rented or purchased as the patient progresses through different stages of recovery. While moving from wheelchair mobility to independent ambulation, patients will usually progress from parallel bars to a walker with a seat to allow frequent resting, and then to crutches or a cane. Because wheelchairs, walkers, crutches, and canes, especially custom appliances, are expensive and not always covered by insurance, the therapist should carefully consider the cost to the patient during the recovery process.

Although most patients with GBS are able to walk within 8 months of onset, many show a prolonged residual weakness of calf and, most commonly, anterior compartment musculature, requiring the use of an AFO. The decision whether to use a prefabricated orthosis or custom appliance is not always simple. Several temporary orthotic measures can be considered. For example, if the patient shows good gastrocnemius-soleus strength with mild weakness of the dorsiflexors, a simple elastic strap attached to the shoelaces and a calf band may be sufficient to prevent overuse of the anterior compartment muscles. An old-fashioned, relatively inexpensive spring wire brace, which can be attached to the patient’s shoes to facilitate dorsiflexion, is a good choice for patients who report sensory hypersensitivity when wearing a plastic orthosis.

Most therapy units today have access to varied sizes of plastic, fixed-ankle AFOs that can be used until a decision is made to have the patient fitted with custom AFOs. A newer system of prefabricated AFOs with adjustable ankle motion cams has been developed that allows the therapist to limit plantar flexion and dorsiflexion to the specific needs of the patient. For patients with reasonable control of plantar flexion and dorsiflexion but with lateral instability because of peroneal weakness, a simple ankle stirrup device such as the AirCast Air-Stirrup Ankle Brace (AirCast, Summit, NJ) can be used temporarily to provide lateral ankle stability. Although few patients with GBS need knee-ankle-foot orthoses (KAFOs) on a long-term basis, inexpensive air splints or adjustable long-leg metal splints to control knee position are sometimes helpful when working on standing weight bearing and during initial gait training. See Chapter 34 for additional information on orthotics.

Psychosocial Issues

Although most patients with GBS have a good recovery over a period of 2 or more years, the acute stage of the disease can be frightening, especially to patients who progress to complete paralysis and respiratory failure. Nancy, in Case Study 17-2, reported that she was terrified during the time she was totally paralyzed (including eyelid movement) and on a respirator. She said that nurses, doctors, and hospital staff seemed to assume she could not hear because she was unable to respond in any manner. In her words,

“They acted like I was already dead, and I thought I would be from the way they were talking. The thing I hated the most was when the night nurses from the registry would come in and ask how to make the ventilator work! I felt panicked. Can you imagine having your life depend on a machine and knowing that the person who was supposed to make it work had no idea what to do if a tube came unconnected? They were always worried about my blood pressure. Who wouldn’t have high blood pressure in that situation! The thing I liked about my therapists was that they told me what they were going to do even when I couldn’t respond. They didn’t just start doing things or pulling on me like other people did.”
next 3 days she developed flaccid quadriplegia and within 5 days she had complete cranial nerve involvement. She was weaned from the respirator after 29 days after several episodes of pneumonia. After extubation, she had swallowing and speech problems that resolved by discharge at 3 months after onset. During the acute stage, she was catheterized because of urinary retention and was treated for a bowel obstruction. Sensation was normal for perception of temperature changes and deep pressure. Proprioception was diminished at the ankle, knee, and fingers. Paresthesias and hypesthesias, aggravated by light touch, were present in a glovelike pattern in both hands and a stocking pattern in both feet.

Nancy’s physical therapy treatment began in the ICU. Formal strength testing was inappropriate; passive ROM was full but felt stiff at ends of ranges in the wrist, fingers, and ankles. The goals were to assist in respiratory care, prevent joint contractures, and prevent stasis ulcers during the period of immobility. Although her postural drainage treatment was performed by using respiratory therapy techniques in conjunction with aerosol medication by intermittent positive-pressure ventilation (IPPV), PTs began a course of chest stretching techniques in coordination with a fastidiousROM program performed twice a day by a therapist and on the evening and night shifts by a nurse. A pressure relief mattress was ordered for her bed. To prevent contracture development, an OT fabricated bilateral wrist and finger splints; a PT molded ankle splints to maintain 90 degrees of dorsiflexion with neutral eversion-inversion. A positioning and ROM schedule in poster form with pictures of positions and ROM patterns was posted at Nancy’s bedside.

Because Nancy reported severe hypersensitivity to light touch or to any passive movement of her limbs, a cradle was placed on the bed to prevent sheets from touching her and to prevent air flow changes from irritating her skin. She was fitted for above-knee light pressure stockings, which seemed to decrease her sensitivity to light touch.

Progression of the GBS process seemed to plateau at approximately 15 days after onset with a gradual return of respiratory function complicated by infections. Weaning from the respirator was difficult, and the PT played a major role in instructing Nancy, the staff, and her family in appropriate breathing exercises to be performed every 1 or 2 hours. Because her parents wanted to be involved with her care, they were taught ROM techniques with special attention to correct shoulder ROM techniques. The PTs continued to follow Nancy twice a day to ensure that accessory motions were completed with correct ROM techniques. The PTs continued to follow Nancy twice a day to ensure that accessory motions were completed with correct ROM techniques.

Follow-up of Nancy’s outpatient therapy showed that she continued to make gradual recovery over the next 1.5 years. She returned to school 3 months after rehabilitation discharge using a wheelchair. She graduated to a walker, then to forearm crutches, and finally to independent ambulation. She refused to be seen using a walker at school, so she continued to use the wheelchair at school until she was independent on crutches.

As part of her positioning program, Nancy was placed in a supported semisitting position while on the respirator. As muscle control returned, a muscle reeducation program was initiated that focused initially on the head and trunk and then on the upper and lower extremities. Exercise periods were limited to 15 minutes twice a day. She would have benefited from more frequent short sessions; however, this was not possible. Her parents were shown how to guide her active exercise program cautiously so that she was able to exercise more frequently at low repetitions. When each muscle group reached an MMT grade of Fair (3) or greater, Nancy was allowed to use the muscles in functional activities with specified limitations in activity duration. When she was able to tolerate upright sitting and had some bed mobility, Nancy was transferred to a Nelson bed in which she could begin a gradual standing weight-bearing program.

A speech therapist worked with Nancy in the ICU to help her relearn safe swallowing patterns and to reintroduce her to different-textured foods. A dietician had been working with Nancy throughout her hospitalization to ensure adequate nutrition while intubated, and she worked closely with the speech therapist to progress Nancy’s diet as she became able to handle liquids and solids.

After being weaned from the respirator and transferred to the general floor, Nancy was brought to the physical therapy department for treatment, which was frequently done in conjunction with occupational therapy. As strength increased, she began a program of resisted exercise. Trunk and upper- and lower-extremity PNF patterns were used as the primary exercise technique; however, great caution was used to avoid overworking weak muscle groups evoked during use of the PNF pattern. A full mat program with rolling and coming to sitting was also instituted. OTs focused on graduated use of Nancy’s upper extremities, first using overhead slings attached to a wheelchair and later using a lap board to support her weakened shoulder musculature while practicing hand activities.

After 2 months of hospitalization, Nancy was discharged home to return for daily outpatient rehabilitation. Because Nancy appeared to be regaining strength well, she was provided with an ultralight rental wheelchair through her insurance for use until a final determination was made for long-term need. Nancy was also fitted with prefabricated adjustable AFOs, which were designed to provide adequate ankle support. After 4 to 6 months a determination would be made about expected recovery of her persistently weakened dorsiflexors. If Nancy appeared to need AFOs for a prolonged period, a set of specifically molded AFOs would be ordered. At discharge, both the PT and OT made a home visit with the hospital social worker and parents to determine what home adaptations and support services would be necessary.

Follow-up of Nancy’s outpatient therapy showed that she continued to make gradual recovery over the next 1.5 years. She returned to school 3 months after rehabilitation discharge using a wheelchair. She graduated to a walker, then to forearm crutches, and finally to independent ambulation. She refused to be seen using a walker at school, so she continued to use the wheelchair at school until she was independent on crutches.

She returned to school 3 months after rehabilitation discharge using a wheelchair. She graduated to a walker, then to forearm crutches, and finally to independent ambulation. She refused to be seen using a walker at school, so she continued to use the wheelchair at school until she was independent on crutches. She continued to wear bilateral AFOs but was weaned from full-time use approximately 14 months after discharge. During the weaning process, Nancy wore her AFOs at school while hiking and for any walking distance over four city blocks or if she heard her feet begin to slap from fatigued dorsiflexors. By 14 months, Nancy showed no evidence of overuse weakness after her regular activities, although she had difficulty with endurance activities in her physical education classes. When hiking, she carried her AFOs to use when she expected a long downhill trek to prevent overwork from eccentric muscle activity. By age 19 years—3 years postonset—Nancy had returned fully to her normal activity level.
Skirrow and colleagues remind clinicians that the “intensive care patient is plunged into a world of machines that flash and beep; of tubes and wires that seem to spring from almost every orifice; and of mind-numbing sedative and analgesic medications.” Needless to say, evidence is increasing that patients treated in acute trauma rooms or ICUs can have posttraumatic stress disorder (PTSD). Particularly vulnerable are patients who have had previous traumatic experiences. PTSD places patients at marked risk for increased startle responses, extreme vigilance or anticipation of painful events, sleep disorders, terrifying dreams, and dissociative flashbacks after leaving the ICU; sometimes these symptoms are left untreated for years after the experience. Patients discharged from prolonged ICU experiences, especially those who had respiratory failure, have an increased incidence of anxiety, depression, and panic disorders years after discharge.

In a nursing study of patient experiences in the ICU, researchers found that patients often felt anxious, apprehensive, and fearful. The patients expected ICU nurses to be experienced and technically adept, but those who felt most secure despite the traumatic ICU experiences felt that the nurses were vigilant to their needs and offered personalized care. A point clearly made by Nancy in the case study. Although one might expect ICU staff to be carefully tuned in to patient needs, the highly technical nature of modern ICUs may attract personnel less focused on individual patient care, or it may prevent caring staff from attending to the little kindnesses that are so comforting to critically ill patients. Baxter suggests that caregivers in the ICU try to orient patients to what is being done, to approach the patients within their field of vision, and to minimize unexpected noises and sudden touching.

Although most patients recover well from GBS, 3 to 6 years after onset of GBS 38% of patients in a Dutch study had to make a job change to accommodate their physical limitations. After 6 years following onset of GBS 38% of patients in a Dutch study had to make a job change to accommodate their physical limitations. After 6 years following onset of GBS 38% of patients in a Dutch study had to make a job change to accommodate their physical limitations. After 6 years following onset of GBS 38% of patients in a Dutch study had to make a job change to accommodate their physical limitations.

In summary, the rehabilitation program for a person with GBS must be graded carefully according to the stage of illness. In the acute care environment when respiratory deficits are present, the initial emphasis is directed toward support of maximal respiratory status through postural drainage, chest stretching, and breathing exercises. Because of prolonged bed rest and immobility related to weakness, accessory and physiological ROM must be maintained with around-the-clock efforts. Splinting or positioning devices are recommended to maintain functional positions during prolonged periods of immobility. A gradual program to increase upright tolerance is begun when respiratory and autonomic functions have stabilized. Therapists must keep in mind the potential to damage denervated muscles with aggressive strengthening programs when developing a rehabilitation plan and a home-based conditioning program. Perhaps as a result of cautious exercise programs, cardiovascular conditioning appears to lag significantly behind strengthening; so endurance training should specifically follow the return of strength. Adaptive equipment and orthoses should be used as needed to protect weakened muscles, facilitate normal movement, and prevent fatigue during the reinnervation process. Although a rehabilitation program has been found to make a measurable difference in patient long-term recovery, many patients are being discharged without follow-up care. Therefore therapists should be assertive in ensuring that their patients with GBS have ongoing contact with rehabilitation specialists who can guide the recovery process (see Case Study 17-2).

**DUCHENNE MUSCULAR DYSTROPHY**

Pathology and Medical Diagnosis

*Muscular dystrophy* refers to forms of hereditary myopathy characterized by progressive muscle weakness associated with deterioration, destruction, and regeneration of muscle fibers. During the process, muscle fibers are gradually replaced with fibrous and fatty tissue. Each of the inherited forms of myopathy (e.g., Becker dystrophy, myotonic dystrophy, limb-girdle dystrophy, and facioscapulohumeral dystrophy) has its own unique genetic and phenotypic characteristics. (For a comprehensive review of the forms of muscular dystrophy and myopathy, see Dubowitz.)

Because Duchenne (pseudohypertrophic) muscular dystrophy (DMD) is one of the most commonly known forms of muscular dystrophy, it is used as a model for discussion of treatment implications for therapists. DMD is a disease of progressive muscle weakness leading to total paralysis and early death in the late teens or young adulthood. It has an incidence of 13 to 33 cases per 100,000 live births and a new mutation rate of approximately 1 in 10,000 (i.e., one third or more of cases occur in families without a history of DMD). The abnormal gene for DMD has been detected on the X chromosome at band Xp21.2, which encodes for dystrophin. A 427-kD cytoskeleton protein in the membrane. Because it has an X-linked recessive pattern, the disease affects males almost exclusively. However, in nearly one third of DMD cases, DNA analysis is normal and diagnosis must be confirmed by protein analysis or immunohistology tests. In almost 100% of patients with DMD there is a complete absence of dystrophin from muscle tissue. This loss of dystrophin results in a weakened cell membrane that is easily damaged in muscle contraction. However, loss of dystrophin alone is not considered the sole explanation of the severity and lethality of muscular dystrophy.

Laboratory studies show serum creatine kinase (CK) elevated more than 100 times normal in early stages of the disease. These CK levels decrease over time with loss of muscle mass. Elevated CK level is evident at birth long before symptoms are evident. Muscle biopsy specimens show degeneration with gradual loss of fiber, variation in fiber size, and a proliferation of connective and adipose tissue. Histochemical studies indicate loss of subdivision into fiber types, with a tendency toward type I fiber predominance. Electromyographic studies show patterns of low-amplitude, short-duration, polyphasic motor unit action potentials.

Although the absence of dystrophin is usually discussed relative to skeletal muscle, dystrophin is also evident on the membrane surfaces of the cardiac Purkinje fibers and is thought to contribute to the cardiac conduction problems seen in DMD. Cardiac involvement is present in more than 60% of boys with DMD across all ages; however, the common electrocardiogram and electrocardiographic abnormalities are reflected early in clinical complications in 30% of...
boys until late stages of the disease, when more than 95% of boys have significant cardiomyopathy. Because of the increased life span secondary to in-home ventilation for respiratory failure, nearly 20% to 30% of deaths can be attributed to cardiac disease.\textsuperscript{216}

The average IQ of boys with DMD is approximately 85, with one third of the boys testing below 75, as reflected in delayed developmental milestones. A specific deficit in verbal intelligence and verbal memory that leads to significant impairment in later cognitive development has been identified.\textsuperscript{217,218}

**Clinical Presentation**

Although histological studies have indicated that DMD may be identified in the fetus as early as the first trimester, symptoms are seldom noted until the child is 2 to 5 years of age. When recalling the child’s early development, parents often state that the affected child was more placid and less physically active than expected.\textsuperscript{219} The earliest obvious manifestations of DMD, however, may be the delay of early developmental milestones, particularly crawling and walking. In many cases the onset is gradual. Parents or teachers may first identify a problem because the boy is noted to have difficulty keeping up with peers during normal play activities and to be somewhat clumsy, with frequent falling when attempting to run, jump, climb structures, or negotiate uneven terrain. By age 5 years, symmetrical muscle weakness can usually be clearly identified by MMT. Deep tendon reflexes may be absent by 8 to 10 years or earlier. Sensation is normal.\textsuperscript{220}

The typical progression of weakness is symmetrical from proximal to distal, with marked weakness of the pelvic and shoulder girdle musculature preceding weakness of the trunk and more distal extremity muscles. Bowel and bladder function is usually spared. Progression of weakness is slow but persistent. Weakness of trunk and lower-extremity musculature typically leads to changes in gait at 3 to 6 years of age. Muscle mass continues to decline, with increasing weakness of the trunk, anterior neck, and upper-extremity musculature affecting functional activities. A typical child will continue walking until about age 12 or 13 years, at which time the process of transition to a wheelchair becomes imperative. A rapid decrease in strength may occur after prolonged periods of immobilization caused by illness, injury, or surgery.\textsuperscript{221}

**Progression of Lower-Extremity Weakness**

Before age 5 years, hypertrophy of the calf muscles is frequently noted. Pseudohypertrophy is evident as the muscle tissue is replaced by fat and fibrous tissue. Even in the early stages of the disease, few boys with DMD walk with a normal gait pattern. Because of early pelvic girdle muscle weakness, most young boys retain a developmentally immature, wide-based gait pattern. An early distinctive feature of DMD is the Gowers maneuver, in which the child gets up from the floor by using his arms to crawl up his own legs (Figure 17-5).\textsuperscript{219}

Muscle imbalance occurs in typical patterns as a result of weakness and contractures. As the posterior hip muscles weaken, the child must arch his back when standing and retract his shoulder girdle to maintain the center of gravity behind the hip joint. This creates a pattern of lumbar lordosis with protrusion of the abdomen. As the quadriceps weaken, the child must maintain his knees in hyperextension to place the axis of rotation posterior to the line of gravity. At this point, mild equinus contractures caused by a muscle imbalance between the plantar and dorsiflexors may help the child maintain knee control because the gastrocnemius-soleus group provides a torque that opposes knee flexion. If plantar flexion contractures become severe, however, the child will not be able to maintain standing balance because his base of support is too small and his ankle adaptive strategies are nonfunctional.

Once the child stops weight bearing, development of severe equinovarus deformities is common. Figure 17-6 shows a pattern of progression of muscle imbalance affecting the trunk and lower extremities in stance. Note the increasing lordosis and plantar flexion as the boys attempt to maintain their center of gravity posterior to the hip joint and anterior to the knee joint.

**Progression of Gait Pattern Changes**

The typical changes in gait pattern over time are identified in Figure 17-7; however, age alone is not an adequate index of predicted gait pattern. Many factors influence how long a child will be able to ambulate. Contributing factors are rate of progression of weakness; severity of contractures (hip flexion, external rotation, abduction, knee flexion, and plantar flexion— inversion contractures occur as disease progresses); influence of body weight; degree of respiratory compromise; type of treatment interventions such as bracing, surgery, and exercise; extent of family support; and the child’s personal motivation to ambulate. When the child can no longer ambulate functionally, a wheelchair must be ordered to fit the specific needs of that child within his home and community environment. (For an extensive analysis of changes in gait pattern see Sutherland and colleagues.\textsuperscript{222})

**Progression of Upper-Extremity Weakness**

The upper-extremity pattern of weakness is similar to that in the lower extremities, with proximal musculature being affected before distal musculature. Functional changes related to weakness of upper-extremity musculature, however, usually lag behind those in the lower extremities by 2 to 3 years. The early weakness of the scapular stabilization muscles interferes with controlled movement of the arms and hands during reaching. The child gradually loses biceps and brachioradialis function, followed by continued deterioration of triceps and more distal musculature. The marked instability of scapular musculature is clearly evident when the child tries to elevate his trunk with his arms (e.g., when attempting to use crutches) or when he is lifted from under the shoulders.\textsuperscript{220,222} A classic test of scapular stability is the test for the Meryon sign, in which the child slips from the examiner’s grip as the child is being lifted from under the arms (Figure 17-8). Typical progression of upper-extremity weakness is shown by use of the reaching test (Figure 17-9).

By the time the child reaches stage 3 of the reaching test, he needs considerable help with eating, hair care, and oral hygiene. Because of major trunk involvement and marked lower-extremity weakness, the child will also be dependent for most ADLs, such as hygiene, dressing, and transferring. Weakness of the respiratory muscles (diaphragm, chest wall, and abdominal musculature) is usually evident by the tenth or twelfth year, although the diaphragm remains functional.
Figure 17-5 ■ Child demonstrating Gowers maneuver necessary to achieve upright posture because of pelvic and trunk weakness caused by Duchenne muscular dystrophy.

Figure 17-6 ■ Pattern of progression of muscle imbalance affecting trunk and lower extremities in Duchenne muscular dystrophy.

Figure 17-7 ■ Early through late stages of ambulation in Duchenne muscular dystrophy demonstrating changes in alignment at loading response, midstance, and terminal stance phases of gait. (From Hsu JD, Furumasa J: Gait and posture changes in the Duchenne muscular dystrophy child. Clin Orthop Relat Res 288:122–125, 1993.)
longer than do the intercostal and accessory muscles. A progressive, sometimes severe scoliosis may contribute to respiratory compromise. Pure respiratory failure, restrictive lung disease, or respiratory failure caused by infection is the usual cause of death, most commonly at age 18 to 25 years. Typical functional stages in DMD are identified in Box 17-5. See Emery and Muntoni for a comprehensive review of the clinical process of DMD.

Medical Intervention

Treatment of Primary Pathology

DMD has no cure. Some clinicians suggest that until an effective treatment can be found, the best way to decrease the number of children with DMD is through genetic counseling. Serum CK is elevated in the female carriers, and genetic molecular probes of possible carriers are now available to identify deletions within the Xp21 region (the short arm of the X chromosome) at a 95% accuracy level. Of course, some families may have belief systems that do not allow consideration of pregnancy termination to prevent having a boy with possible DMD. Those views must be respected. Prenatal diagnosis of DMD for women without a family history of the disease is not yet practical.

Despite much effort, an effective pharmaceutical agent has not been identified to treat DMD. In a Cochrane review, Manzur and colleagues concluded that glucocorticoid corticosteroid therapy improves muscle strength in the short term of 6 months to 2 years; however, adverse effects such as weight gain, excessive hair growth, osteoporosis, and behavioral problems were noted. Researchers have also attempted to implant the normal precursor muscle cells or myoblasts directly into dystrophic mice and, in several cases, into children with DMD to precipitate the proliferation of normal donor muscle cells into the host muscles of dystrophic subjects, but results have not led to significant improvement. Animal studies using helper-dependent adenoviral vectors for dystrophin gene transfer to muscles in dystrophic mice show promise for patients with DMD.

Although no cure for DMD is on the horizon despite the positive research on gene transfer, the functional status of the patient, quality of life, and life expectancy can be influenced with thoughtful, functionally based treatment and supportive care. Figure 17-10 provides an overall scheme for the management of DMD.

Figure 17-8: Meryon sign shows lack of scapular stability as the child slips from the examiner’s grip when lifted from under the arms.

Figure 17-9: Method of evaluating the working hand as demonstrated by the reaching test.
Assistance in respiration progresses in steps. In the first step, a self-inflating manual ventilation bag may be sufficient. Step two is associated with manual and mechanically assisted cough techniques. Steps three and four consist of the institution of nocturnal and daytime ventilation, respectively. Step five consists of tracheostomy, if the patient and family prefer.

Sleep-disordered breathing and hypoventilation are common in the later stages of DMD, and the onset is often subtle. Early symptoms include repeated nighttime awakenings, early morning headache, and daytime sleepiness. Inexpensive oximetry can be used in the home to identify nighttime oxygen desaturation if polysomnography with continuous carbon dioxide monitoring is not available.

Because sleep hypoxia is common in the later stages of DMD, IPPV or noninvasive ventilation by nasal mask or mouthpiece is recommended to control oxygen desaturation at night. Eventually most boys with DMD enter a stage of constant hypoventilation throughout the day and night, and a decision needs to be made about the use of 24-hour ventilation support. Daytime ventilation should be considered when waking $P_{CO_2}$ exceeds 50 mm Hg or hemoglobin saturation is lower than 92% while awake. Motorized wheelchairs can be adapted to handle ventilator systems so that the boys can remain active and mobile.

Once a patient with DMD requires daytime and nighttime ventilation and has severe bulbar muscle weakness, a decision must be made to elect ventilation by tracheostomy.
Management of Duchenne muscular dystrophy (DMD). Coordination of clinical care is a crucial component of the management of DMD. This care is best provided in a multidisciplinary care setting in which an individual and family can access expertise for the required multisystem management of DMD in a collaborative effort. A coordinated clinical care role can be provided by a wide range of health care professionals depending on local services, including (but not limited to) neurologists or pediatric neurologists, rehabilitation specialists, neurogeneticists, pediatricians, and primary care physicians. It is crucial that the person responsible for the coordination of clinical care be aware of the available assessments, tools, and interventions to proactively manage all potential issues involving DMD. ABG, Arterial blood gas; ACE, angiotensin-converting enzyme; DMD, Duchenne muscular dystrophy; ECG, electrocardiogram; GI, gastrointestinal; MEP, maximum expiratory pressure; MIP, maximum inspiratory pressure; PCF, peak cough flow; ROM, range of motion. (From Bushby K, Finkel R, Birnkrant DJ, et al: Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. Lancet Neurol 9:77–93, 2010.)

or palliative care. Ventilation by tracheostomy allows higher ventilation pressures and a better patient-ventilator interface. However, use of a tracheostomy requires careful stoma hygiene to prevent infections and mucus plugs and requires 24-hour caregiver vigilance. Although many patients and families adapt well to tracheostomy use, the ability to speak audibly may be affected. Consideration must be given to use of a speaking valve system. Several cases of pneumothorax have been reported with long-term IPPV. Also, as increasing numbers of patients use long-term tracheostomy-based ventilation, the potential for tracheal erosion or tracheobronchomalacia, which must be monitored to prevent hemorrhaging, is increasing. As with patients with ALS, many significant treatment and ethical decisions must be made by the patient, family, and health care providers when submitting to prolonged HMV. Patient autonomy and family input after adequate patient education about prolongation of life by tracheostomy ventilation must be respected.

Cardiomyopathy is present in 59% of children with DMD by 10 years of age, but the cardiac problems seldom become symptomatic until the end stages of DMD because the child’s decreased activity level does not stress the weakened heart muscle. In later stages of the disease, however, cor pulmonale with right-sided heart failure may occur. Medical treatment of any cardiac symptoms generally follows the conventional interventions. Some boys with severe scoliosis that creates cardiac compression may require correction by spinal fixation. Retrospective data suggest that children treated before ventricular dysfunction with corticosteroids have a lower incidence of cardiac involvement.
**Nutritional Concerns**

Excessive weight gain that impairs functional ability is a frequent and difficult problem for children with DMD and their families. The typical active child needs approximately 2400 calories daily to maintain weight and grow; however, the child with DMD who is more sedentary or who is wheelchair dependent may need 1200 or fewer calories to maintain weight. Because of decreased esophageal and intestinal motility, exacerbated by weak or absent abdominal muscle strength, a healthy low-fat diet should be encouraged with adequate bulk foods, stool softeners, and fluids to facilitate bowel function and motility. Problems with obesity are often related to the family’s typical pattern of eating and nurturing. The child and family members may “feed” their anxiety or depression about the disease. In many cases, family members and friends feel that the child’s only pleasure may be eating. Although this may seem true, caring for a totally dependent obese teenager or young adult can become problematic for both the child and the caregivers. Before obesity becomes an issue, the child and his family should be referred for comprehensive nutritional advice from a specialist experienced in dealing with childhood obesity. Suggestions for adapting eating behavior and food choices will not be followed if they are too restrictive or unreasonable for the child’s social situation.

Although obesity is a common problem for children with DMD (greater than 54%), malnutrition is also common. Malnutrition usually occurs in the late stages of the disease as a result of dysphagia. Special care must be taken to provide adequate nutrition after spinal surgery. One review showed that postsurgical weight loss was related to the inability to self-feed; therefore the investigators suggest that before surgery a feeding evaluation should be done and an appropriate plan should be put in place to prevent postsurgical malnutrition.

As the disease progresses, some children develop problems swallowing, and then weight loss and malnutrition can become an issue. To decrease the possibility of aspiration, careful attention must be paid to food textures and chewing and swallowing functions (see page 537 for information on dealing with bulbar symptoms). Depending on the patient’s and family’s decisions about prolongation of life, some patients now elect to have a permanent PEG placed once self-feeding and swallowing become a problem rather than a pleasure. Even if the patient can still swallow and enjoys eating in the late stages of DMD, the patient may not be able to physically take in adequate calories; the PEG allows the delivery of needed calories and fluids beyond what the patient can take orally (see page 563 for information on dysphagia and eating issues). Consensus is that body weight and body mass index should be reviewed regularly and family education on nutrition should be an ongoing process. Evaluation of swallowing should be assessed by taking a history of choking episodes and observing the child eat different foods and fluids. Videofluoroscopy should be used to determine if aspiration is a problem, and appropriate adjustments in feeding should be instituted under the supervision of the appropriate therapist. (See also Bushby and colleagues.)

**Treatment of Scoliosis**

Scoliosis is a frequent complication of DMD, with a reported incidence of nearly 90%. Consequences of severe scoliosis are increased respiratory problems in boys with respiratory compromise, chronic pain related to musculoskeletal problems, sitting tolerance difficulties, and caregiving issues. Figure 17-11 presents an example of a boy with moderate scoliosis that affects sitting posture. Note the pelvic asymmetry that would seriously affect sitting alignment.

Scoliosis tends to occur in two basic patterns: the early-onset form (seen in approximately 23%), which becomes evident before the child begins to use a wheelchair, and the late-onset form, which develops, on average, 4 years after wheelchair dependency. In the early-onset form the curve usually becomes severe and progressive, leading to pulmonary compromise and structural-based pain. In the late-onset form the course is usually mild. Unfortunately, attempts to control sitting posture through the use of a spinal orthosis and wheelchair seating inserts (inserts that place the child in lumbar lordosis to lock facets, thereby preventing rotation and lateral collapse, or, more commonly, lumbar and thoracic lateral supports) have been disappointing. Bach states that thoracolumbar bracing is never indicated to slow scoliosis development in DMD and it cannot substitute for surgical correction; however, spinal bracing may improve comfort and postural stability in some patients who are not eligible for surgical correction because of severe respiratory or cardiac involvement.

Efforts have been made to delay the time of onset of scoliosis with steroid treatment protocols. Evidence supports the hypothesis that onset of scoliosis can be delayed; however, a longer follow-up period would be required to determine if scoliosis can be prevented.

Cervellati and colleagues reported on a study of 20 boys treated from 1985 to 1995 and concluded that early surgery significantly reduces the risk factors associated with severe spinal deformities. The period after spinal surgery requires careful coordination of medical, respiratory, and physical therapy services. Depending on the hospital culture, PTs may be responsible for the pulmonary drainage and breathing exercise programs as well as typical passive and active programs while the child is in ICU and postsurgical care environments. Preferably, therapists should introduce postural drainage and breathing techniques as well as exercise expectations to the child before surgery to gain better cooperation after surgery.

Figure 17-11  Moderate scoliosis affecting sitting stability.
Treatment of Other Musculoskeletal Dysfunctions

The primary effect of progressive weakness in DMD generally results in secondary effects such as decreases in muscle extensibility, joint contractures, and bone demineralization. Strength loss diminishes the ability to move actively through full range, shift out of static positions, balance muscle forces around a joint, and avoid fibrotic changes in muscle tissue. Loss of ROM from muscle shortening and joint stiffening will occur if not aggressively prevented. Once present, contractures can severely complicate function. Long bone fractures in children with DMD are a serious problem that can have a significant long-term impact on ambulation. In a study of 378 patients, 21% had incurred fractures, primarily from falling. Leg fractures predominate in independent ambulators and wheelchair users, whereas upper-extremity fractures more often occurred in boys using KAFOs. Twenty percent of those who had fractures lost the ability to ambulate.

In standard treatment protocols for children with DMD who have impairing loss of ability to walk independently, bilateral KAFOs are used in conjunction with surgical release of contractures. At the point of surgery, a pattern of contractures has magnified the effect of weakness from the loss of approximately 60% of muscle mass. Surgery is typically followed by an aggressive therapy program. Bach and McKeon studied 13 boys with DMD who had surgery to release lower-extremity contractures. Seven boys were ambulating independently before surgery (early surgery group), and six boys were preparing to use or had begun to use a wheelchair before surgery (late surgery group). Depending on the contraction pattern, the boys underwent surgical procedures that typically included subcutaneous release of the Achilles tendons and hamstring muscles and fasciotomy of the iliobibial bands. Four patients had rerouting of the posterior tibialis to the dorsal surface of the second or third cuneiform to balance the foot and prevent the often severe varus position of the foot. Boys in the late surgical group required more extensive inpatient rehabilitation, whereas boys in the early surgical group were treated as outpatients after a short hospitalization. Physical therapy was started on the second postoperative day. The program consisted of general conditioning exercises of the trunk and extremities (e.g., rolling, trunk stabilization, neck and head control), stretching exercises, and intensive weight bearing in standing while wearing bilateral long-leg casts or below-knee casts, depending on the surgery. One child participated in a pool therapy program. Bach and McKeon suggest that early surgery for contractures followed by intensive physical therapy can prolong brace-free ambulation. The number of falls experienced by the boys decreased markedly after the surgery and rehabilitation period. Boys in the early intervention groups benefited from the surgical interventions more than the boys in the later intervention groups. All patients and their families in the early surgery group thought that the procedures were helpful. Boys in the late surgery group, however, stated either that they would not have had the surgery if they had a chance to decide again or that they had no opinion. Roposch and colleagues reviewed the records of 91 boys with the typical equinovarus deformity in DMD and strongly recommended surgical intervention, including a posterior tibialis transfer, over conservative, nonsurgical treatment to maintain foot position and lengthen time of ambulation.

Manzur and colleagues carried out a randomized, controlled trial of 20 boys with DMD (ages 4 to 6 years) to study the effect of early release of contractures versus conservative (stretching) programs. The boys were followed for 12 months or more. Surgery corrected the contractures and improved the speed of gait and transfers over conservative treatment as measured at 12 months, but a 2-year follow-up of six of the boys who had surgery revealed a recurrence of ankle contractures. In addition, some of the boys in the operated group showed more rapid deterioration. The authors did not recommend routine early surgery to relieve contractures.

Therapeutic Management of Movement Dysfunction Associated with Duchenne Muscular Dystrophy

Like ALS, DMD has a relentless and incurable progression toward total dependence and eventual early death. The differences are the population (children rather than adults) and time course, with DMD taking 15 to 25 years rather than the 3 to 5 years typical of adults with ALS. In ALS and GBS, strength and endurance remain the primary impairments of DMD, with secondary problems such as contractures and respiratory problems following from immobility. Unlike in the other neuromuscular disorders, the endurance problems in DMD are related to peripheral fatigue, fatigue stemming from the muscles themselves rather than from the lack of ability to recruit additional motor units. As in ALS and GBS, therapeutic management in DMD will involve evolution of the intensity and frequency of exercise to correspond to changes in the strength and endurance of the patient. In all three disorders, the general therapeutic goals are to maximize function, manage discomfort, and promote optimal quality of life. The differences among the disorders mean that the actual form of the exercises and interventions in DMD may require adaptation to suit a child or adolescent. Ideally, a team of specialists should be involved in the long-term care of a child with DMD and his family. The therapist’s primary role is twofold: to perform serial examinations of the child’s movement capabilities and to adjust the child’s intervention program as the disease progresses. Even with relentlessly progressive diseases, rehabilitation programs can have potential psychological benefits, such as more positive coping strategies, while physical activity continues to decline.

Examination

A typical therapy examination should include a history, systems review, and tests and measures to assess muscle strength, endurance, and ROM impairments along with levels of activity and participation. In some facilities the therapist also collects data on the child’s pulmonary status. History taking should include the course of the disease, any recent illnesses or losses of function, coexisting neuromotor or other medical conditions, current concerns, and the goals of the patient and family. Screening tests can help rule out sensory deficits, identify cardiac and respiratory issues, and determine skin integrity, especially in im mobile patients. Checking vital signs at rest and immediately after activity, noting communication ability, and assessing ability to follow multistep commands are all important components. When screening tests indicate a deficit, follow-up should occur with additional testing or referral to
the appropriate professional. The tests and measures appropriate for assessing the movement dysfunction of patients with DMD include measures of strength, ROM, function, activity, and quality of life. Palmieri and colleagues review many of the measures reported in the literature for use in this population.

**Manual Muscle Testing.** MMT is used extensively for measuring muscle strength of children with DMD and is relatively reliable if consecutive examinations are made by the same rater. Interrater reliability of scores in the gravity-eliminated position have been shown to be highest in this population. DMD shows a linear pattern of decreased muscle strength (loss of about 0.25 MMT unit per year from ages 6 to 13, and 0.06 MMT unit per year from age 13 on) without marked increases in the rate of deterioration in strength over time. Thus, marked, precipitous changes in muscle strength noted in a few months with initiation of bracing or wheelchair use for example, or immobilization after fracture, generally reflect disuse atrophy rather than disease progression. Such transitory weakness may respond to increased activity and exercise. The history and medical records can help differentiate weakness stemming from various sources and thus determine the potential for strengthening. Cable and strain-gauge tensiometers, handheld myometers and dynamometers, and isokinetic dynamometers may also be useful for a more discriminating documentation of muscle strength.

**Range of Motion.** ROM is assessed with goniometry in most cases of DMD. As with MMT, serial ROM evaluations should be completed by the same therapist because intrarater reliability is higher than interrater reliability in this population. The two-joint muscles are most prone to de- 

**Functional Status.** The child’s functional status continues to be relatively stable for some time even when MMT indicates that the child is losing strength. Because the weakness is gradual, many children develop remarkably adaptive adjustments in movement patterns to remain functional even with marked strength loss. Lue and colleagues developed the Muscular Dystrophy Functional Rating Scale (MDFRS) to standardize assessment of the functional impact of muscular dystrophy, including people with DMD—more than half of those tested. The MDFRS consists of 33 items covering mobility, basic ADLs, arm function, and impairment (including contractures, strength of the trunk and neck, scoliosis, and respiratory issues). The developers reported test-retest and interrater reliabilities of 0.98 to 0.99 and good evidence of validity. Brooke and colleagues and Vignos and colleagues have described previous functional scales for use in DMD; the MDFRS compares favorably with each of these, with some advantages for determining the child’s status and for predicting appropriate care, perhaps because it is longer. As part of any functional assessment in DMD, adaptive behaviors should be noted. For example, a child may not be able to lift his arm overhead, but he may use his fingers (strength often remains intact even after respiratory support is necessary) to “craw” his chest to reach his head or he may lean forward to approximate his chest to his hand or use his other arm or a lever system to assist with activities.

For ambulatory patients, gait velocity can help predict how long the patient has before transitioning to a wheelchair. In a longitudinal study of 51 boys with DMD, 100% of those who took 9 seconds or more to walk 30 feet were wheelchair bound within 2 years. McDonald and colleagues also recommend the use of the 6-Minute Walk test as a standardized and functional measure of endurance for this population. Slight modifications may be necessary to keep younger children on-task for this test. Observational gait analysis can help to identify adaptive behaviors and use of compensatory strategies during locomotion.

**Respiratory Function.** The PT’s role in evaluating respiratory status in children with DMD will vary depending on the facility and area of the country in which the therapist works. For more in-depth information regarding evaluating pulmonary status, refer to Chapter 30 or see Irwin and Tecklen. At a minimum the therapist should evaluate bulbar function, cough effectiveness, and FVC (a simple spirometer available in most clinics is adequate). For more sophisticated testing, the child should be seen by a pulmonary function specialist. In addition, the therapist may monitor activity levels via armbands or pedometers or may assess metabolic equivalents or caloric consumption to design the optimal activity program for children with DMD and obesity. One method of testing a child’s energy cost during ambulation is the energy expenditure index, which divides walking heart rate (WHR) minus resting heart rate (RHR) by walking speed (distance [D] divided by time [T]): EEI = [WHR – RHR]/[D/T]). Determinations of energy expenditure while walking may factor into the decision to transition to a wheelchair, at least for longer distances.

In late stages the therapist may need to assess the child’s bulbar function to prevent swallowing and aspiration problems caused by tongue and oral-facial muscle weakness.

**Therapeutic Goals.** The basic goals for a therapeutic program are straightforward: (1) to prevent contractures that can lead to further disability and pain, (2) to maintain maximal strength and endurance and prevent disuse atrophy, (3) to facilitate maximal functional abilities by using appropriate adaptive equipment, (4) to maintain maximal respiratory muscle strength and movement of secretions, and (5) to
foster realistic child and family expectations within the context of the environment. These are broad-based goals; the therapist will need to write more specific, time-oriented goals for a particular episode of care.

**Therapeutic Interventions**

Younger children with disabilities are usually eligible for school-based therapy services. However, therapists increasingly act primarily in the role of consultant rather than direct service provider, especially for older children. Much of the child’s exercise program must be carried out at home by parents or caregivers. When both parents work outside the home or when the child lives in a single-parent home with a working parent, compliance with home programs can be problematic. As many exercise activities as possible should be encouraged within the child’s school day so that parents can focus on parenting, nurturing, general caregiving, and simple positioning and bedtime exercises. Under the supervision of a consulting therapist, the child’s therapy often can be provided in some form at the child’s school if on-site therapists, personal attendants, or adaptive physical education teachers are available.

**Respiratory and Dysphagia Care.** In the school therapy environment, where most children with DMD are monitored, the therapist should be prepared to provide the child and family with methods to improve breathing efficiency. In the early stages of the disease, the child and family can be taught simple breathing exercises stressing diaphragmatic breathing, full chest expansion, air shifts, and rib cage stretching. Most children enjoy playing with handheld incentive spirometer units and blowing bowling games (e.g., bubbles, pinwheels). Respiratory exercise in different studies has resulted in improvement in respiratory endurance, ventilatory muscle endurance but not respiratory muscle strength, and both respiratory muscle strength and endurance. In the last study, two thirds of the 27 subjects had DMD, with percent predicted vital capacities of 27% to 96% that had decreased over the 6 months immediately preceding the exercise protocol. The exercise protocol, monitored via a visual feedback system, consisted of twice-daily sessions of 10 cycles of resisted inspiratory breaths at 70% to 80% of the patient’s maximum inspiratory pressure, plus 10 maximal static inspiratory efforts that reached at least 90% of the maximally generated inspiratory pressure. The intervention lasted for 2 years, with increases noted in the first 10 months and a plateau maintained through the end of the training period. Winkler and colleagues noted the first 10 months and a plateau maintained through the end of the training period. Winkler and colleagues noted the first 10 months and a plateau maintained through the end of the training period.

Although inspiratory exercises tend to be the focus of interventions, expiratory inefficiency may play a major role in the inability to clear secretions. Once the child begins to have difficulty clearing secretions, the family should be taught manual or mechanically assisted postural drainage techniques as long as the patient has an adequate cough. Patients who need support with coughing can be taught “air stacking” techniques (taking a series of breaths without exhaling between breaths) to increase intrathoracic pressure needed to cough effectively. Some patients respond well to manual coughing assistance. Increasingly patients and caregivers are being taught to use a mechanical insufflator-exsufflator (positive pressure followed by negative pressure) to stimulate coughing. These techniques should be reviewed and used aggressively whenever the child is bed bound for more than 1 or 2 days and before and after all surgical procedures. Physical therapy interventions, such as postural drainage and breathing exercises, are invaluable in preventing early death from respiratory failure. The Muscular Dystrophy Association continually updates its information on breathing and respiratory care.

In end stages of DMD when the child is dependent, dealing with oral-motor problems that may interfere with eating and swallowing is imperative. Techniques such as positioning, increased sensory input (texture, temperature), and volume changes in foods may improve the child’s swallowing and allow the child to continue taking food orally. The interventions are similar to those described for ALS. The Muscular Dystrophy Association also publishes informational manuals dealing with dysphagia problems (see www.mdausa.org).

**Prevention of Contractures.** Diligent ROM exercises for the whole body will require cooperative efforts of the rehabilitation team and the patient and family. Stretching may progress as weakness dictates, from active to active-assisted to passive to prolonged elongation phases using positioning, splinting, orthoses, and standing devices. During the ambulatory phase of the disease, focus should be on the hips, knees, and ankles. Later, focus will shift to the shoulders and the elbow, wrist, and finger flexors. At the first sign of loss of end ROM, the therapist should adjust the child’s program to include specific stretches.

Evidence provides a protocol for stretching in people with normal muscles to increase ROM: stretches performed 2 to 3 days a week, once per day, held for 10 to 30 seconds for three or four repetitions over a 6-week time frame. Unfortunately, no such evidence exists for the best stretching protocols in DMD to maintain ROM. Palmieri and colleagues recommend that stretching be performed a minimum of 4 to 6 days per week for any joint or muscle group.

The stretch should be slow to avoid muscle reflex contractions, and sustained at the end point for 10 to 30 seconds. To increase muscle extensibility, dry or wet heating, electromagnetic stimulation, or a warm bath may help; for best effect, follow a bath by drying with prewarmed towels to avoid shivering and muscle stiffening.

In a 2010 Cochrane review of the best methods for increasing ankle ROM in patients with neuromuscular disease, only two studies of DMD were noted, with interventions of early surgery or prednisone use. Surgery eliminated the contractures, but in most cases the contractures had recurred by the 2-year follow-up. Prednisone...
had no significant effect on ROM in comparison to a placebo or when comparing two different doses.\textsuperscript{284} Hyde and colleagues\textsuperscript{285} noted an annual delay of 23\% in the development of contractures at the Achilles in boys with DMD randomly allocated to a group receiving both stretching and night splints compared with boys who had stretching alone. Brooke and colleagues\textsuperscript{220} reported similar findings, and Scott and colleagues\textsuperscript{290} noted that boys who had both AFOs and stretching were able to continue walking longer than boys who did not. This evidence indicates that multiple simultaneous strategies may be most beneficial in preventing shortening and maximizing function. Patient and family preferences must be considered for any plan to be effective, however. Some young patients do not tolerate night splinting well. In such cases, AFOs to control plantarflexion contractures may be preferred over long-leg orthotics that prevent knee flexion contractures or align hips (using an additional bar between legs to control rotation).

Early in the course of the disease process, both parents and the child must be educated about the expected changes in muscle balance and how they can play an active role in preventing or limiting the impact of contractures caused by muscle imbalance. Because contractures at the hip, knee, and ankle interfere with the mechanical alignment necessary to stand erect and walk, each day the child should be encouraged to move his own limbs to end ranges through normal play activities to slow development of contractures related to sedentary positioning. Some research supports the view that the combination of positioning, stretching, and splinting should begin before contractures exist. For example, the child can be encouraged to watch television or play video games while lying prone with legs aligned out of the common "frog leg" (hip abduction and external rotation) pattern. Once a child has significant hip flexor or iliobibial band contractures, stretching techniques must be specific because simple prone positioning can force the lumbar spine into excessive lordosis. Although difficult to accomplish in some mainstreamed school environments, positioning the child in a standing frame during several class periods helps provide prolonged stretch to hip, knee, and ankle musculature. Later in the course of the disease, resting hand splints are appropriate to control shortening of the long finger flexors.\textsuperscript{221}

Although development of contractures of the hip, knee, and ankle from muscle imbalances has been thought the cause of early loss of ambulation instead of weakness,\textsuperscript{287} others believe that weakness causes the loss of ambulation instead.\textsuperscript{280} Some authors note that loss of ambulation can occur from either case.\textsuperscript{250} Limiting contracture development facilitates mobility and handling throughout the course of the disease, however, and the best approach to contractures is to prevent them.\textsuperscript{290}

**Exercise and the Maintenance of Maximal Functional Level.** Because DMD affects muscles throughout childhood and adolescence, when strength and endurance are generally developing, effectiveness of strengthening and aerobic exercise has been difficult to assess.\textsuperscript{267} Training programs may maximize muscle and cardiorespiratory function, but they have also led to reports of weakness after physical exercise.\textsuperscript{288} The debate over the value of exercise in DMD and the relative lack of controlled trials have limited the ability of clinicians to provide evidence-based therapy. No definitive protocols can be provided at this time. In general, however, both strengthening and aerobic exercises should be considered, the frequency and intensity of which should be appropriately prescribed based on the disease course and the patient’s abilities and goals.

Strengthening exercises have had mixed support in the past.\textsuperscript{289} de Lateur and Giaconi\textsuperscript{290} noted small gains in strength of the exercised compared with the unexercised quadriceps muscle of four boys with DMD during and for 18 months after a 6-month exercise program of submaximal isokinetic muscle contractions, 30 repetitions, 4 to 5 days per week. No postexercise weakness or increases in deterioration were noted in the exercised muscles. Vignos and Watkins\textsuperscript{291} instituted a home program of maximal resistance exercises for 1 year; the 14 patients with DMD in the exercised group improved in strength for the first 4 months and then reached a plateau, compared with declines in strength of the control group. Scott and colleagues\textsuperscript{292} noted diminished strength after a strengthening home-exercise program for 18 boys, although with no control group, possible reductions in disease progression could not be confirmed.

Evidence for the effectiveness of strengthening exercises in other muscle disorders is insufficient\textsuperscript{293} and cannot thus be generalized to DMD. Elder\textsuperscript{294} reviewed animal studies suggesting that dystrophic mice trained on a treadmill showed increased damage to muscle tissue, whereas forced swimming in dystrophic mice had no adverse effect. In a case review of three generations of patients with facioscapulohumeral muscular dystrophy (seven cases and one suspected case), Johnson and Braddom\textsuperscript{295} noted asymmetrical weakness of the upper extremities. They related the weakness to patterns of overuse (dominant side or side used most often in work activities). On the basis of their information and additional evidence that muscle-derived enzymes (CK and myoglobin concentrations in blood) were markedly elevated in patients with DMD after prolonged exercise,\textsuperscript{296} repetitive exercise may be contraindicated.\textsuperscript{297} In contrast, Cup and colleagues\textsuperscript{298} reported that in their review of 33 studies of exercise therapy for neuromuscular diseases, they found absent or negligible adverse effects; one study reported that “3 of 20 patients decreased their training for 1 or 2 sessions due to delayed-onset soreness.”

Given the evidence to date, Hasson\textsuperscript{299} concluded that exercise consisting of brief periods of low- or high-intensity activity can improve strength for patients with minimal to moderate weakness. The increased recruitment of motor units from training effects also may improve muscle coordination and reduce disuse atrophy. However, exercise programs have minimal effect on strength of muscles already severely weakened.

In addition to active and resistive exercise programs, Scott and colleagues\textsuperscript{290} completed a small study of the effect of intermittent, long-term, low-frequency electrical stimulation on dystrophic anterior tibialis muscles. They demonstrated a significant increase in mean voluntary contraction force and suggested that electrical stimulation can have a beneficial effect if used with children whose muscles are not already markedly weakened. Zupan\textsuperscript{290} supports this finding, but children under treatment were unable to maintain strength beyond 4 to 5 months.
Evidence for the effect of aerobic training in DMD is sparse. Hasson in a review of exercise studies of patients with muscular dystrophy, reports that oxygen consumption improved with endurance training, although whether repetitive endurance training at moderate or high intensity (70% of VO₂max) causes muscle damage is unknown. Muscle biopsies in DMD have revealed reduced or missing nitric oxide synthase, necessary for sufficient nitric oxide levels. Nitric oxide normally limits vasoconstriction in muscles during and after exercise and also provides cytoprotection and antiinflammation in muscle tissue. Muscle fatigue in DMD may thus be exacerbated by ischemic exercise. However, aerobic training in other muscular diseases has shown indications of positive effect on aerobic capacity as well as measures of activities and participation, so generalization to DMD has a possible rationale. In addition, strengthening exercises in combination with aerobic exercises in other muscle disorders have been shown to have a likely positive effect.

Overall, the data from animal and human studies suggest that submaximal exercise is not harmful and it may be helpful in maintaining maximal function if the patient does not exercise into marked fatigue. Because muscle endurance and peak power are diminished in addition to muscle strength, a focus on program design related to functional exercises individualized to each child’s functional requirements is recommended.

Ideally, the child’s exercise can be incorporated into measurable activities adapted for children with movement and weakness-related balance problems. Many ambulatory children enjoy ball activities, walking-based simple obstacle courses, parachute games, table tennis, cycling (preferably tandem), and especially swimming. Swimming is an excellent exercise for children with DMD because they often are quite buoyant because of their increased fat/muscle ratio. Many children can continue to float or swim independently on their backs even when nonambulatory (if supervised) and able to move only distal musculature. The Muscular Dystrophy Association has an excellent guide to water-based exercises: “No Sweat Exercise: Aquatics.”

A safe indicator of extent and intensity of exercise is that the patient should recover from exercise fatigue after a night’s rest. When designing an active play program, therapists should review the types of muscle contractions that the activity requires, considering that possible muscle damage occurs when muscles are active and functioning in an eccentric manner. Concerns about damage from eccentric muscle contractions were supported in animal studies in which dystrophic muscles were found to be more susceptible to stretch-induced muscle damage.

Figure 17-12 shows responses of normal and impaired muscle to exercise. (See Eagle’s report on exercise in neuromuscular diseases.) In a summary of findings on effects of physical exercise on conditioning in muscular dystrophy, Ansved found that the scientific basis for clear recommendations on exercise prescription is poor, but evidence does show the importance of maintaining an active lifestyle with limitations on high-resistance and eccentric training activities.

**Maintenance of Ambulation.** As DMD progresses, the child’s posture (a result of both weakness and contracture) and gait pattern abnormalities become extreme and he must work harder to maintain balance while walking. Most children gradually discontinue walking about a year after they lose their ability to deal with stairs or when daily ambulation time decreases to less than 30 minutes per day. Toward the end of the child’s independent walking stage, he has a marked anterior pelvic tilt with lordosis and a protuberant abdomen. His shoulders are retracted and he may hold his hands behind his hips or elevated in a mid-guard position to stabilize his hips. He has a severe waddling gait with a shortened stride, and he must carefully lock his knees at each step. He falls frequently, which may result in fractures of the lower or upper extremities.

If the child and his family have followed an aggressive ROM, positioning, and activity program, the child’s walking time may be extended by months. In most cases, however, the contractures from muscle imbalance continue relentlessly and the child begins to need support when walking. When contractures at the hip, knee, and ankle show evidence of interfering with the child’s ability to stabilize each joint during stance, most children are referred for surgery to restore functional joint motion. Figure 17-13 shows the typical walking pattern of a boy with DMD who is being considered for release of contractures and bracing.

Bracing either before or after surgery may be indicated to assist with positioning and stabilizing joints for function. Ideally, bilateral KAFOS should be measured and fitted in final form before surgery to release contractures so the child can begin upright weight bearing in the KAFO the day after surgery. KAFOS are commonly fabricated of molded plastic thigh units (ischial weight-bearing quadrilateral socket) with metal joints at the knee (drop locks) and ankle (or a flexible plastic ankle component) (Figure 17-14). If the orthoses are not immediately available, the child can begin the standing program in long-leg casts. Casting must be kept to a minimum because of the risk of disuse atrophy in immobilized muscles. (See Grossman and colleagues for a review
of the effect of immobilization on normal muscle and appropriate therapy interventions.)

In the hospital, standing in bilateral KAFOs can be initiated on a tilt table. Most children are fearful after surgery and report significant pain when their legs are moved or if they are placed upright. For therapy to be successful during this early standing stage and during passive ROM exercises, the child must have adequate pain medication. If the child is not properly medicated in the first few days after surgery, the therapist may have to deal with difficult, resistant behaviors of the child that persist long after the pain should have subsided. Pain protocols must be discussed before the child’s surgical procedures. The child should be medicated at least 30 minutes before the therapist’s visit.

Gait training is usually begun within 48 hours after surgery. Initial work focuses on helping the child regain his sense of standing balance because his old patterns of equinus, lordosis, and shoulder retraction may no longer be adaptive. The child should be allowed to find his own best center of balance, and he should be allowed to use compensatory gait deviations necessary to allow the best mobility and stability. Depending on the child’s upper-extremity strength and control, he may progress from parallel bars for balance assist, to pushing a wheelchair or weighted walker, to balance assist from a therapist with a safety strap to prevent falls. Some children who seem to need a walker for balance transition do best if they use a walker with forearm rests and vertical hand grips, which seem to help them stabilize their arms more effectively than a standard walker. Fortunately, most children do learn to walk independently without support again after surgery, although they are unable to negotiate steps or inclines or rise from the floor independently. Hyde and colleagues report that 24 of 30 boys treated with KAFOs were able to achieve functional ambulation again. Vignos and colleagues report in a review of long-term treatment of DMD that a combination of operative procedures, orthotics, stretching, and a program of standing and walking resulted in extended walking until a mean age of 13.6 years and standing for 2 years after that. With the early use of surgery and bracing procedures to maintain ambulation, the expected deterioration in muscle strength and function as a result of becoming sedentary in a wheelchair is deferred.

Because most children with DMD are discharged home within a few days of surgery, PTs must provide options for continuing standing within the home. Standing frames are often available through the child’s school district or therapy unit. If they are not, the therapist can help the family build a simple standing frame for home. This frame often can be made from a piece of plywood, or a gluteal strap system can be attached to a table at home. If possible the child should be positioned just forward of the line of gravity to encourage back extension with facet stability and to allow the child better head control in the presence of weak anterior neck muscles. Use of swivel walkers has been recommended by some therapists and physicians because the child does not need upper-extremity control for support. Although the concept of hands-free walking seems logical, boys with DMD had more difficulty using the walkers compared with children with paraplegia because of the more delicate postural adjustments needed by children with dystrophy and their greater sensitivity to the motion restriction of the swivel
walker. In addition, older children with DMD are seldom willing to wear externally visible bracing outside the home or school system. Some therapists have reported success with the ORLAU variable center of gravity swivel walker (Mopac Ltd., Eau Claire, Wisconsin)\(^1\), however, support for its use is not widespread.

Bakker and colleagues\(^2\) reviewed the literature on the effectiveness of treatment with surgery and KAFOs. They found that the scientific strength of the studies was poor. Although the treatment approach seemed to prolong the walking time, whether it extended functional walking was not clear. The children who benefited most were highly motivated and had slower rates of deterioration.

**Transition to Wheelchair.** Although surgical and orthotic interventions may prolong ambulation within the home and classroom past the predicted time for cessation of independent walking (8 to 12 years), most children begin to use a wheelchair for community mobility and long distances before this time. When children begin to spend more time in their chair, the rate of development of contractures, disuse weakness, and obesity increases.\(^2\) Because of this more rapid deterioration in the child's functional skills, professionals and parents often discourage the child from using a chair for mobility. Children, however, tend to welcome use of the chair because they have more energy for their social interactions and learning tasks.\(^3\)

Selection of the appropriate wheelchair is often difficult for the patient and family because of the multiple decisions that must be made. Few children with DMD can propel a manual wheelchair for more than a few years because of their increasing upper-extremity weakness. In addition, their propulsion speed in their manual chair is seldom adequate to keep up with their peers. Eventually, the child will need a motorized chair. Although this provides tremendous freedom for the child, a motorized chair presents problems to many families because transporting the chair requires a van and lift unit, which is seldom funded by insurance. Ideally, the child should have both a manual and a motorized chair; however, in today's health policy climate, parents or advocates often must engage in protracted efforts to obtain adaptive equipment for the patient.

An important consideration when purchasing a wheelchair is the trunk support system. Traditionally, boys with DMD are thought to develop a gravity collapse of the spine related to their functional sitting posture. To control the collapsing spine, spinal orthoses and seat inserts to lock the spine in extension (to prevent lateral bending and rotation) are frequently recommended. Unfortunately, the effectiveness of positioning devices to control the development of scoliosis has been disappointing.\(^4\) The therapist therefore should work with the child, the family, and the orthopedist to determine the best system to maintain optimal spinal alignment and trunk stability as the child weakens. In addition, as the child becomes more physically dependent, the chair may need to be fitted with a pressure-relief molded seat and trunk cushions, elevating leg rests, and a reclining back with a head rest.\(^5\) The Tilt-in-Space chair (LABAC Systems, Denver) is a good example of a chair that can be motorized to allow mobility as well as maximal adjustment of seat position by using mouth control systems. It can also be adapted for a respirator attachment. The decision about the type of power chair necessary in the later stages of disease progression takes considerable thought. Therapists, the patient, and the parents or caregivers must review environmental constraints, access issues, social goals, and work and recreational needs.

Because of the problems associated with increased wheelchair use, the therapist must work closely with the family and any school-based personnel to design a realistic plan to prevent rapid deterioration in strength and independent function. If possible, the child's standing program in KAFOs should be continued at school and at home as long as possible, with a goal of 3 to 5 hours of standing per day. With mainstreaming, however, continuing a standing program at school is sometimes difficult because attendants and equipment are not available, the child may need to move from room to room for different classes, and the child may not like being singled out for special treatment. It is helpful to caregivers if the child continues to wear his KAFOs when using the chair until he is totally dependent for transfers and can no longer be pivoted from the chair to another surface.

If the child uses a motorized wheelchair, directional control systems must be adapted to each child's needs. Most young people with advanced DMD do well for years with a standard joystick hand control system; however, because of extended survival times relative to the long-term use of mechanical ventilation, many patients must have their control systems adjusted frequently to minimize the need for muscle control, such as pinch strength. The need for ventilation support while using the wheelchair does not seem to interfere with the ability to drive.\(^6\) (See Cooper\(^7\) for a comprehensive manual on wheelchair selection. This information is equally valuable for patients with ALS and GBS.)

When the child can no longer tolerate the sitting position, some children have continued to attend school on a gurney. Once the person with DMD is no longer able to attend school or work, the home environment will need to be adapted for maximal self-direction despite significant physical dependence. Both low- and high-tech environmental control systems are more readily available today than they were 10 years ago. Television control units, voice-activated telephones, switch-activated bed controls, and page turners are among the low-tech systems. Sip-and-puff, blink-operated, and voice-activated control units can be adapted to operate most electronic devices. OTs and PTs can provide invaluable support to the person with DMD and the caregivers by making several home visits to suggest modifications and adaptive devices and systems. (See Cook and Hussey\(^8\) for detailed information on assistive technology systems. Also see an excellent website for home automation, environmental control, and electronic aids for daily living [EADLs]: www.makoa.org/ecu.htm.)

**Psychosocial Issues**

Psychosocial issues related to DMD are family issues. At the time of the child’s diagnosis, the parents are often emotionally devastated and cycle back and forth through many phases of denial, anger, sadness, and active coping, especially if they feel guilt that they “caused” their child's disease. This process tends to recur when the child does not meet expected normal physical and social milestones or when he reaches predicted stages of deterioration, such as the transition to a wheelchair. Because children with DMD have concomitant developmental and cognitive
delays or issues, educational and social interactions can be compromised in addition to the physical changes. Because DMD is a multisystem multiprocess disease, early in the child’s life the family should be guided to encourage the child’s independence and to discourage overprotection. Therapists can play an important role in helping the child and family identify realistic goals for independence. In addition, therapists can be instrumental in extending independence and a sense of self-direction by anticipating patient needs for adaptive equipment and identifying appropriate assistive devices and environmental control systems that empower the person with DMD and provide relief for caregivers from the constant attention required by a completely dependent person. Key to family support is access to a multidisciplinary clinic with specialists in neurology, pulmonology, orthopedics, rehabilitation services, psychology, social work, and dietetics. Only through comprehensive clinics do families of children and adults with DMD receive the level of education and support necessary to deal with the changing levels of function and demands on family systems. Psychosocial support should be made available to the child and family during predictable times of crisis. Major times of crisis occur around the age of 5 years when the child begins to realize his differences, at age 8 to 12 years when the child loses the ability to walk independently, during the adolescent years when social interactions become restricted, and around the time of high school graduation when the child and family must face vocational limitations and almost certain death within the next decade. Transition times are often accompanied by depression, withdrawal, and anxiety in the child and family members because parents had a marked preoccupation with their sons and a diminished expression of enjoyment. Predictably, the integrity, strength, and intragenerational and intergenerational function and coping styles of the child’s family contribute a great deal to the way the family responds to the child’s progressive deterioration. Extended periods of anxiety and depression should be treated vigorously with cognitive interventions, support groups, respite care, and, when appropriate, short-term anxiolytics and antidepressants. Repeated opportunities to discuss end-of-life care must be given to both the child and parents. Professionals, however, tend to underestimate the quality of life for patients with end-stage DMD; therefore patients and family members must be educated about long-term options for ventilatory support or palliative care well ahead of any respiratory emergency that might occur to ensure that the patient's desires are respected.

Because of the extended life opportunities for DMD patients who may now live into their 20s, home care requirements, the impact of in-home care on family members, and the financial impact must be fully reviewed and support systems put in place before caregiving stress becomes overwhelming. Positive family functioning while caring for a dependent child or adult with DMD is correlated with caregiver health and hardiness and requires multiple levels of family support from family, friends, and professionals. Increasingly, young men with DMD are attending college even though they may require 24-hour assistance with ADLs and monitoring of ventilation equipment. To date, parents are providing most of the care to their children with DMD by attending colleges or living in dorms or apartments with their child. With life extended with ventilation, parents and the young person with DMD should begin early to plan for a future with maximal decision making by the young adult with DMD. This mindset of a “future” requires considerable problem solving by all people involved in the care of the young adult. Parents of children with DMD should involve their child early in life to make appropriate decisions about care, learn about medical needs and practices, and deal with finances necessary to run a home or hire an attendant. These issues related to independence (even though physically dependent) and caregivers are now being discussed by patients with DMD and their caregivers.

Parents and the child should be given the opportunity to discuss the impending death in an accepting environment with persons who are experienced in dealing with degenerative diseases. Because the child and family have long anticipated the child’s death and have made transitions through many levels of grieving, the process of separation and mourning may have occurred before the child’s death. Each child and family member should therefore be helped to deal with the process according to his or her own pace and in response to individual needs. The child’s death is sometimes considered a welcome relief. This feeling of relief, however, is often accompanied by survivor guilt and a tremendous sense of loss of life focus for the family members whose lives have been so intertwined with that of the child’s. Ideally, arrangements should be made for the family to meet with the professionals with whom they feel most comfortable several weeks after the child’s death and again several months later so that the family (and caregivers) can deal with their thoughts and feelings (Case Study 17-3).

CASE STUDY 17-3  ■ JEREMY

Jeremy was 3 years old when he was diagnosed with DMD. He lived at home with his mother and a 5-year-old sister. There was no known family history of DMD, although family lore suggested that a cousin died quite young from pneumonia and a “wasting disease.” Jeremy was referred for a medical evaluation when a playground supervisor at his preschool noted that he was clumsy when running and that he had difficulty on the playground climbing equipment and the slide. He also had difficulty rising from the ground and needed to hold on to a railing when stepping up a stair.

During a medical history, Jeremy’s mother said that she had noticed that he was “slow to develop” but was not worried because she thought he was just a “late bloomer.” A muscle biopsy was positive for a diagnosis of DMD. A physical therapy evaluation 3 months after diagnosis showed ROM to be within normal limits for all joints. Muscle weakness was evident on
SUMMARY

In this chapter, discussion of three different diseases reveals the varied effects of neuromuscular pathology on a person’s day-to-day function. ALS is an adult-onset degenerative disease of the upper and lower motor neurons; GBS is an inflammatory process affecting the PNS of children and adults; and DMD is an inherited degenerative disease manifesting in childhood that affects muscle tissue. In all three conditions the therapist must design a therapy program that will provide the patient with the impetus to become or remain as active as possible without causing possible muscle damage from excessive exercise demands or overwork.

Therapists must be aware of their own feelings and reactions to patients with severe neuromuscular diseases. Working with patients with GBS is usually a positive experience because most patients attain full recovery despite their often severe disability during the acute illness and long recovery period. Working with patients with degenerative terminal diseases, however, draws deeply on the therapist’s emotional and spiritual strength. A typical response of health care professionals is to view these patients’ conditions as hopeless and to assume that the patients must also perceive their existence as hopeless, depressing, and without value. Research does suggest an increased incidence of depression...
and demoralization in patients with degenerative, terminal diseases compared with nonaffected populations. Other research, however, has indicated that many patients perceive their own life satisfaction much more positively than professionals would believe. Therapists must tap into patients’ positive energy to design treatment programs that respect patients’ goals and life plans within the context of their environment.

Limited evidence exists to document the effectiveness of rehabilitation for patients with progressive neurological diseases. Determining the most appropriate exercise and therapeutic intervention programs therefore requires diligent examination of the dysfunctions and needs of the individual patient and assessment of the effects of interventions appropriately adapted from use in other populations.

Because few medical-clinical facilities see a large enough sample of patients with any of these three diagnoses, therapists must align with their professional organizations to institute nationwide, multisite research studies to provide clear evidence of effectiveness of therapy in these populations.

References
To enhance this text and add value for the reader, all references are included on the companion Evolve site that accompanies this textbook. This online service will, when available, provide a link for the reader to a Medline abstract for the article cited. There are 326 cited references and other general references for this chapter, with the majority of those articles being evidence-based citations.
CHAPTER 19  Multiple Sclerosis

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KEY TERMS
autoimmune disease
axonal damage
benign
demyelination
disease-modifying agents
exacerbation
immune system
lesion
neuroprotection
plaques
primary progressive
progressive relapsing
relapse
relapsing remitting
remission
secondary progressive

OBJECTIVES
After reading this chapter the student or therapist will be able to:
1. Describe the pathological processes, prevalence, and clinical presentation of people with multiple sclerosis.
2. Compare and contrast the types of multiple sclerosis and the common disease progression in each.
3. Discuss the medical management of the disease and the disease symptoms.
4. Describe how the International Classification of Functioning, Disability and Health provides a common language for describing the impact of disease on people with multiple sclerosis and how it provides a framework for rehabilitation management.
5. Describe the outcome measures that can be used to examine people with multiple sclerosis that cover body system problems (impairments), functional skill and activity limitations, and participation restrictions.
6. Develop a rehabilitation plan of care using evidence-based interventions to maximize patient function and quality of life.

OVERVIEW OF MULTIPLE SCLEROSIS
Pathophysiology
Multiple sclerosis (MS) is a chronic, inflammatory disease of the brain, optic nerve, and spinal cord mediated by the immune system. It is characterized by lesions of disseminated focal demyelination accompanied by variable axon damage and destruction and reactive gliosis. Initially, MS was thought to be a disease of the white matter (WM); however, recent investigations have shown that the gray matter (GM) is significantly involved. Lesions found in the GM typically contain demyelination and loss of neurons without the immune system infiltrates and inflammation characteristic of lesions in the WM. Tissue damage has been found outside the focal lesions throughout the GM that is associated with brain and spinal cord atrophy. These areas of demyelination and axonal damage interfere with normal conduction of neural signals, leading to a disruption of function.

Early in the course of the disease, focal inflammatory WM lesions are composed of immune system components that produce demyelination, axonal injury, and loss of oligodendrocytes. Astroglia activated by the damaged neurons produces gliotic scarring (visualized as sclerosis in postmortem brain tissue) called plaques. Active disease is followed by periods of remission in which acute inflammation is reduced. Axonal remyelination occurs but is highly variable and is related to recovery of function during periods of remission. The degree of axonal loss is associated with the severity of the inflammation; however, axons are spared in the majority of WM lesions. Treatment in the initial stages of the disease is aimed at reducing inflammation and immune system infiltration with disease-modifying agents (DMAs).

Later in the course of the disease, inflammation becomes uncommon while demyelination and axonal loss continue, suggesting replacement by a neurodegenerative disease process. Disease progression becomes more constant with a lack of exacerbation. The motor, sensory, and cognitive disability that accumulates in the advanced stages of the disease appears to be associated with the cortical GM pathology. Owing to the lack of inflammation, DMAs have not been shown to be beneficial in the later stages of the disease.

Incidence and Prevalence
MS is the primary cause of nontraumatic disability in young and middle-aged adults and the most common inflammatory condition of the central nervous system (CNS). It is reported that approximately 350,000 to 400,000 people in the United States and over 2.5 million people worldwide have the disease. People are most commonly diagnosed at age 20 to 50 years, with an average age of 32. However, MS can be diagnosed in people of any age. Approximately 5% of all patients with MS are diagnosed before their sixteenth birthday.

MS is found in people who reside above the northern or below the southern 40° latitude with greater frequency than those who live closer to the equator (Figure 19-1). Given the
increased sun exposure of people living closer to the equator, lack of vitamin D is being investigated as a potential factor contributing to disease development. Many researchers believe that exposure to an infectious agent may trigger the disease process: Epstein–Barr virus is currently considered a likely candidate.

Women are affected two to four times more frequently than men. Even so, men are more likely to have a more aggressive disease progression and a worse prognosis. Caucasians with Northern European ancestry have the greatest incidence of MS, whereas people of Asian, African, or Hispanic ethnicity are at lower risk. African Americans have a lower incidence, but become disabled earlier than Caucasians, suggesting that tissue destruction occurs earlier and more rapidly. Inuits, Yakutes, Hutterites, Hungarian Romani, Norwegian Lapps, Australian Aborigines, and New Zealand Maoris do not appear to develop MS. Being diagnosed with MS may be related to age, gender, genetics, geography, or ethnic background. An identical twin with MS means that the other twin will have a 25% chance of diagnosis, suggesting something beyond genetics. Having a first-degree relative with MS will increase the risk of disease from 1/750 to 1/40.

**Types of Multiple Sclerosis and Clinical Characteristics**

At least four types of MS have been identified (Figure 19-2). Although the course of the disease is highly variable even within a subtype of MS, there are characteristics common to each.

The initial neurological episode or attack is typically identified as clinical isolated syndrome (CIS). Symptoms must last for at least 24 hours and can be monofocal or multifocal. If there are lesions present on magnetic resonance imaging (MRI), there is a high risk of developing MS. In one group of people with CIS followed for 20 years, 63% were diagnosed with definite MS.

Relapsing remitting MS (RRMS) represents about 85% of people with MS, characterized by exacerbations (attacks, flairs, relapses) that can last days to months and are typically followed by periods of improved function. During remissions, function can return to prerelapse levels, but most frequently it does not recover fully. Attacks normally occur with a frequency of one or two per year. Approximately 90% of people with RRMS transition to SPMS after 20 years or around 40 years of age.

In secondary progressive MS (SPMS), relapses decrease in frequency over time and convert to a slow steady progression of increasing disability or disease severity. Relapses may occur early in SPMS but gradually lessen over time. People with RRMS eventually convert to SPMS 10 to 20 years after diagnosis.

It is thought that the clinical disability associated with SPMS results from the neurodegeneration that occurs as a result of tissue injury that accumulates from early in the disease process. In addition to less inflammation, there is a greater amount of brain atrophy in people with SPMS compared with RRMS. Figure 19-3 shows the natural history of RRMS and SPMS, comparing the change in brain volume with increasing clinical disability and disease burden.

Primary progressive MS (PPMS) is less common, affecting only 10% to 15% of people with MS. From disease onset, progression results in a gradual worsening of symptoms without relapses. People tend to be older when diagnosed (late 30s or early 40s), have fewer abnormalities on brain MRI, and respond less favorably to standard MS therapies. Progressive myelopathy is commonly associated with PPMS.

Progressive relapsing MS (PRMS) is the least common form (5%). This form of MS typically begins with a progressive course with clear relapses or exacerbations.

Benign MS is identified when symptoms occur once and never recur. This happens in roughly 25% of cases. Recently, Sayao and colleagues reported that 52% of people with benign MS had not developed MS 20 years later. However, the remainder of people went on to develop MS, with at least 21% requiring the use of a cane.
Multiple Sclerosis

The authors could not identify any criteria associated with either developing MS or continuing to have the benign form.

The risk of a more rapid disease progression is correlated with older age at diagnosis; male sex; initial symptoms involving the motor, sphincter, or cerebellar systems; multifocal disease at onset; shorter time between first and second attacks; and frequent attacks in the first 5 years postdiagnosis.\textsuperscript{7,14}

Clinical Manifestations

MS can affect the optic nerve and any tissue within the brain or spinal cord, so almost any neurological symptom can result. Individual assessments are required to identify the problems present. Even so, the following list constitutes the most common problems encountered by people with MS.

Fatigue

Of people with MS, 65% to 97% report fatigue during the course of the disease; as many as 40% of people with MS state that fatigue is their most disabling symptom.\textsuperscript{13} There are two types of fatigue in people with MS: primary and secondary. Primary fatigue, often called lassitude, is caused by the effects of the demyelination and axonal destruction and its effect on nerve conduction. Restorative rehabilitation has little effect on primary fatigue from neurodegeneration. Secondary fatigue results from problems such as deconditioning, infections, sleep disturbances, poor nutrition, medication side effects, other medical conditions (such as thyroid disease), and heat intolerance. Clinicians should be extremely careful to separate the types of the fatigue in order to determine the most appropriate interventions.

Sensory Impairments

Sensory impairments are among the most common symptoms associated with MS and can affect the visual, somatosensory, and vestibular systems.\textsuperscript{16} The most common problem of the visual system is optic neuritis, which can produce blurry or double vision and/or painful eye movements and nystagmus. Somatosensory or proprioception disturbances can include dyesthesias (tingling, buzzing, or vibrations) or anesthesias (complete loss of sensation in part of the body). People may experience paresthesia or anesthesia in half of the body—upper or lower or side to side—or below a certain spinal cord level. Dyesthesias may be limited to small body areas such as a patch of skin on the head or a single upper or lower extremity. Vestibular system involvement occurs in 20% of people with MS at some time during their disease course\textsuperscript{17} and may manifest as dizziness and/or vertigo. People with MS can have pain associated with the damage to neural tissue (neuropathic pain). This may manifest as neuralgias with burning, itching, or electric shocklike sensations. Lhermitte sign is an electric shock–like shooting sensation that can run into the upper extremities or down the back in response to flexion of the neck.

Motor Systems Impairments

Deficits in the motor system include weakness, spasticity, ataxia, and tremor. Paresis or muscular weakness is frequently seen in people with MS and is associated with several causes. Like fatigue, weakness can be caused by damage to the myelin and axons of motor and premotor neurons in the CNS that can manifest in many different patterns including monoparesis, paraparesis, hemiparesis, or quadriparesis. However, additional causes of muscle weakness can also be associated with disuse deconditioning and may also result in muscle atrophy. When muscle weakness or loss of motor control is seen in the muscles of speech, it results in dysarthria. Paralysis or total loss of muscle strength occurs with less frequency but can be devastating for patients. Several patterns of paralysis (or “-plegia”) occur in people with MS including paraplegia, hemiplegia, and quadriplegia.
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A broad clinical definition of spasticity is a velocity-sensitive resistance to muscle stretch or a muscle spasm during movement. Some people report heaviness in the limbs, difficulty moving a joint, jumping of the extremities, or involuntary painful movements. Muscle spasms or cramping are frequently experienced by people with MS. Eighty-four percent of people with MS report spasticity, with 34% indicating that their spasticity is moderate to severe. Female sex or longer disease duration are both associated with higher prevalence of spasticity. Spasticity has been highly correlated with patient-reported disability and poorer quality of life (QOL). Spasticity may change according to position and may result from increased effort during activity or from the presence of a noxious stimulus such as an infection, skin lesions, fractures, renal stones, distention of bladder or colon, or other physiological stressors such as certain medications (DMAs or serotonin reuptake inhibitors) or psychological distress. Environmental factors such as tight clothing, hunger, or elevated body or air temperature may also lead to increased spasticity. Spasticity can cause muscle contractions, skin breakdown, pain, and sleep disturbances, which often lead to secondary activity limitations and participation restrictions that limit performance of activities of daily living (ADLs) and mobility.

Ataxia occurs in up to 80% of people with MS at some point in their disease progression. This motor deficit can occur from disturbances in the vestibular system or cerebellum or a loss of proprioception. Ataxia or a lack of coordination can manifest as difficulty with walking or difficulty with movements of the extremities such as overshooting or undershooting targets (dysmetria) or an inability to produce rapid alternating movements (dysdiadochokinesia). Occasionally, patients experience sustained body positioning (dystonia) of the extremities or head and neck. In different research studies, tremor is reported by 25% to 58% of people with MS, with the majority of people experiencing mild to moderate dysfunction. Action tremor, both postural and intention, are found in people with MS, pointing to the cerebellum as a likely source (see Chapter 21). Tremors affect the head, neck, vocal cords, limbs, and torso, with the upper extremities having the greatest occurrence.

MS affects many of the systems required for postural control and balance, including sensory input (visual, somatosensory, and vestibular), central processing, and motor output. Therefore it is not surprising that over 50% of people with MS report falling one or more times in the previous 6 months.

**Bowel and Bladder Dysfunction**

The incidence of bowel problems (35% to 68%) and bladder problems (52% to 97%) make them common in people with MS, as reported by two research studies. Symptoms include urinary urgency, nocturia, or retention of urine or feces. Incontinence of either system can also occur. Neurogenic detrusor muscle overactivity is the most common urological impairment in people with MS; 20% have detrusor muscle underactivity, and only 10% report no symptoms.

**Sexual Dysfunction**

Sexual dysfunction affects 40% to 85% of women with MS and 50% to 90% of men. It can manifest as erectile dysfunction, impotence, inability to achieve orgasm, and, in men, retrograde ejaculation.

**Cognitive Impairments**

Cognitive dysfunction occurs in roughly 40% to 70% of people with MS, with 70% demonstrating mild to moderate impairment. Although cognitive problems can occur at anytime, abilities affected early in the course of the disease are verbal fluency and verbal memory. Other cognitive dysfunctions common in people with MS include impairments in memory, processing speed, executive functioning, attention, and visuospatial learning. There is a fair correlation between cognitive decline and ability to work and unemployment because of the impairments in short- and long-term
Depression
Depression is two to three times more common in people with chronic health conditions than in the general population and has a greater incidence than other neurological conditions. From 26% to 50% of people with MS have been reported to experience depression during the course of the disease. Several factors contribute to the high incidence of depression in people with MS. The fact that MS is a chronic, progressive, and unpredictable disease that affects people in their early to middle adult years, is often invisible, and limits participation in many life roles often leads to a perceived reduction in QOL. Suicide is of great concern for people with depression, and rates are significantly higher in people with MS than in the general population.

Depression is associated with a lower QOL significantly higher in people with MS than in the general population. From 26% to 50% of people with MS have been reported to experience depression during the course of the disease. Suicide is of great concern for people with depression, and rates are significantly higher in people with MS than in the general population. Depression is associated with a lower QOL and other symptoms of MS including fatigue, disability, pain, and cognitive impairment.

Heat Intolerance
Uthloff phenomenon is a temporary worsening of MS-related problems associated with an increase in core body temperature. Such increases can occur with physical exertion such as exercise or with a change in the environment such as hot baths or showers, hot weather, and hot air temperature.

MEDICAL MANAGEMENT
Diagnosis
Historically, people with MS would wait for a diagnosis for a year or more. Although there are no definitive tests that diagnose MS, the addition of MRI has accelerated diagnosis. In 2001 the International Panel on the Diagnosis of Multiple Sclerosis updated criteria to include MRI, visual evoked potentials, and cerebrospinal fluid (CSF) analysis. The 2005 Revised McDonald Criteria for MS diagnosis were designed to make the diagnostic process even more efficient and easier.

The Poser criteria require the presence of two separate episodes over time, plus evidence of two or more lesions in separate brain or spinal cord regions identified by radiological imaging studies. Even with the improved technological measures used to facilitate diagnosis, an accurate clinical history is critical. Often patients will recall episodes of transient symptoms that did not last long enough to require attention by a primary care provider.

In addition to the clinical history, MRI studies have improved diagnosis of MS. Although T2-weighted MRI images show MS lesions as hyperintense and identify new or active lesions, MRI has been shown to overestimate clinical relapses. Conventional MRI with T1 weighting identifies lesions as hypointense (black holes) and is able to identify brain atrophy. T1 imaging demonstrates a stronger correlation with clinical status and disease severity than the lesion load found with T2 weighting. Gadolinium-enhanced T1-weighted MRI images show active MS lesions as hyperintense (white).

Two additional medical tests can be used to aid in the diagnosis of MS and differentiate it from other diseases and conditions. The first is the analysis of CSF. This requires a lumbar puncture in which CSF is gathered and analyzed to identify oligoclonal bands representing the presence of immune system proteins indicating that the body is attacking itself. The majority of people with MS have oligoclonal bands; however, because people with other diseases or conditions also have oligoclonal bands, the test is not specific for MS. The lack of oligoclonal bands at diagnosis has been related to a slower progression of the disease and increased time to reach markers of disability such as walking with an assistive device or confinement to a wheelchair.

Evoked potentials record the nervous system’s response to stimulation of a specific sensory pathway (visual, auditory, vestibular, or general somatosensory). Demyelination and axonal degeneration cause a slowing of signal transmission along neurons and therefore will increase the response time to an externally applied sensory stimulus. Damage to the optic system is a common first symptom in MS, and therefore visual evoked potentials are often most helpful in diagnosis.

Disease severity and progression are monitored by ongoing medical checkups, MRI imaging, and the use of several outcome measures. The Kurtzke disease severity scale was developed to allow primary care providers a way to measure clinical disability and chart disease progression. It has been replaced by the Expanded Disability Status Scale (EDSS) (Table 19-1). The EDSS is a 10-point ordinal scale completed by a physician or physician extender, with 0 indicating no disability and 10 indicating death caused by MS. Using a cane relates to an EDSS score of 6.0. The National MS Society (NMSS) Task Force on Clinical Outcomes Assessment also recommends the Multiple Sclerosis Functional Composite (MSFC) as a measure of disease severity and progression. This set of outcome measures is used to

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**Figure 19-4**  T2-weighted magnetic resonance imaging (MRI) scan of plaques associated with multiple sclerosis. Plaques are indicated by arrows. (From Frey H, Lahtinen A, Heinonen T, Dastidar P: Clinical application of MRI image processing in neurology. Int J Bioelectromagnet 1(1), 1999.)
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chart change in physical and cognitive function and will be discussed later in this chapter. It includes three tests that measure upper-extremity function (Nine-Hole Peg Test [NHPT]), lower-extremity function and mobility (25-Foot Timed Walk [25FTW]), and cognitive function (Paced Auditory Serial Addition Test [PASAT]).

Medical management of MS has two major goals: long-term management of the disease and exacerbations and symptomatic management. Early after diagnosis with CIS, it is recommended that people take DMAs. Recent evidence suggests that as the disease progresses it becomes less inflammatory and more neurodegenerative. Therefore medications aimed at reducing inflammation will be less effective as the disease progresses. Fox\(^2\) suggests that early treatment is needed to compensate for the later stages of the disease when inflammation is less prevalent.

**Medications**

**Disease-Modifying Agents**

DMAs are aimed at reducing immune system dysfunction, thereby reducing damage to neural tissue and long-term disability for people with RRMS. There are several different medications that act on various components of the immune system with the intention of modifying the course of the disease (Table 19-2). In general, these drugs are approved for use with RRMS and are used off-label for other forms of MS and have been shown to reduce the number of attacks experienced. The majority of the drugs require injections; however, in 2010 the U.S. Food and Drug Administration (FDA) approved the first oral DMA, fingolimod. Measurement of therapeutic effectiveness includes relapse rate, progression of disability (EDSS), and quantitative evidence of lesions on MRI. All DMAs have side effects (see Table 19-2), but rarely are they serious. These medications are costly, and some people do not respond well or tolerate the side effects. It is common that people will try more than one type before finding the DMA they tolerate the best.

**Antiinflammatory Medications**

High-dose corticosteroids (such as prednisone or methylprednisolone) are used to reduce inflammatory response during exacerbations for people with RRMS. Although no medications have demonstrated effectiveness in people with

<table>
<thead>
<tr>
<th>SCORE</th>
<th>FUNCTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.0</td>
<td>Normal neurological examination findings</td>
</tr>
<tr>
<td>2.0</td>
<td>Minimal disability</td>
</tr>
<tr>
<td>3.0</td>
<td>Moderate disability</td>
</tr>
<tr>
<td>4.0</td>
<td>Ambulates 12 hours without aid</td>
</tr>
<tr>
<td>5.0</td>
<td>Disability impairs activity (walks 1500 feet without assistance)</td>
</tr>
<tr>
<td>6.0</td>
<td>Intermittent or unilateral constant assistance</td>
</tr>
<tr>
<td>6.5</td>
<td>Bilateral support required (walker, crutches, two canes)</td>
</tr>
<tr>
<td>7.0</td>
<td>Unable to walk 15 feet without assistance</td>
</tr>
<tr>
<td>8.0</td>
<td>Basically constrained to bed</td>
</tr>
<tr>
<td>9.0</td>
<td>Bedridden</td>
</tr>
<tr>
<td>10.0</td>
<td>Death from multiple sclerosis</td>
</tr>
</tbody>
</table>

## Table 19-2: Disease-Modifying Agents: Indications and Side Effects

<table>
<thead>
<tr>
<th>FDA-Approved Disease-Modifying Agents</th>
<th>Indication</th>
<th>Common Side Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>IFN beta-1a (Avonex)</td>
<td>CIS</td>
<td>Flulike symptoms</td>
</tr>
<tr>
<td>IFN beta-1a (Rebi)</td>
<td>RRMS</td>
<td>Injection-site reactions</td>
</tr>
<tr>
<td>IFN beta-1b (Betaseron)</td>
<td>SPMS</td>
<td>Depression</td>
</tr>
<tr>
<td>IFN beta-1b (Extavia)</td>
<td>CIS</td>
<td>Elevated liver enzymes</td>
</tr>
<tr>
<td>Glatiramer (Copaxone)</td>
<td>RRMS</td>
<td>Injection-site reactions</td>
</tr>
<tr>
<td>Natalizumab (Tysabri)</td>
<td>RRMS</td>
<td>Progressive multifocal leukoencephalopathy</td>
</tr>
<tr>
<td>Mitoxantrone (Novantrone)</td>
<td>RRMS</td>
<td>Cardiotoxicity</td>
</tr>
<tr>
<td>Intravenous infusion</td>
<td>SPMS</td>
<td>Treatment-related leukemia</td>
</tr>
<tr>
<td></td>
<td>PRMS</td>
<td>Infection risk</td>
</tr>
<tr>
<td>Fingolimod (Gilenya)</td>
<td>RRMS</td>
<td>Alopecia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Amenorrhea</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Increased liver enzymes</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Headache</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Diarrhea</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Back pain</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cough</td>
</tr>
</tbody>
</table>

CIS, Clinical isolated syndrome; FDA, U.S. Food and Drug Administration; IFN, interferon; RRMS, relapsing remitting multiple sclerosis; SPMS, secondary progressive multiple sclerosis.
PPMS, anecdotal evidence suggests that intermittent pulses of intravenous methylprednisolone can help slow progression of clinical disability in some patients. A host of additional medications are used to manage the symptoms associated with MS. Each will be discussed as part of symptom management. Also refer to Chapter 36 for additional information.

**Symptom Management**

**Fatigue**

The fatigue experienced by people with MS is generally divided into primary and secondary causes. Fatigue from primary causes results from the disease itself or to heat intolerance and is defined by the term *MS lassitude.* Heat intolerance may result in a temporary worsening of symptoms. It is sometimes referred to as *pseudoexacerbation* and occurs when core body temperature rises with exposure to raised ambient temperature or metabolic activity such as exercise. However, in addition to MS lassitude, other causes can include side effects of medications used in the treatment of MS, deconditioning from reduced activity levels, poor nutrition, infections or other medical conditions, depression, or sleep disturbances. Several medications combined with rehabilitation strategies have been recommended for management of fatigue. Amantadine (Symmetrel) and modafinil (Provigil) are frequently prescribed.

**Spasticity**

Spasticity can interfere with physical function and hygiene. However, spasticity can also add support to weakened limbs, allowing more effective mobility. The goal of medical management of spasticity is to maintain full range of motion (ROM) of muscle and soft tissue structures to allow maximal physical function and proper hygiene. Haselkorn and colleagues describe the clinical practice guidelines for managing spasticity in people with MS written by the Multiple Sclerosis Council. A complete assessment of the spasticity and how it affects the individual’s life is required. Typically, successful management includes both pharmaceuticals and rehabilitation.

When spasticity is the result of CNS impairments, medical management often includes the use of oral pharmacotherapy including baclofen (Lioresal) or tizanidine (Zanaflex). Adjuvant therapies include diazepam (Valium) or clonazepam (Klonopin), dantrolene (Dantrium), gabapentin (Neurontin) or levetiracetam (Keppra), clonidine (Catapres), or muscle relaxants. Each of these drugs can have negative side effects that interfere with movement and therefore rehabilitation.

Management of focal spasticity may include local anesthetics such as lidocaine, bupivacaine, etidocaine, all of which are short acting with side effects of CNS and cardiovascular toxicity and hypersensitivity. Neurolysis treatment with phenol or alcohol is longer acting; however, these agents can have the side effects of pain, swelling, fibrosis, and dysesthesias. Focal spasticity affecting functional muscle groups can also be effectively treated with neuromuscular blocking agents including alcohol, phenol, or botulinum toxin. Botulinum toxin type A (Botox) has been shown to improve spasticity as measured by the Ashworth Scale and the hygiene score, but no changes were noted in spasm frequency score. Blocks last 1 to 3 months with relatively few side effects. Similarly, botulinum toxin type B was shown to reduce hip adductor spasticity. Clinical practice guidelines recommend that neuromuscular blocks be performed by appropriate specialists in conjunction with a rehabilitation program.

Refractory spasticity is defined as unsuccessful treatment with oral medications and/or rehabilitation. In this situation two other options exist: surgery or placement of an intrathecal baclofen pump (ITB). Surgical procedures include tendon lengthening or tendon transfer and are performed to maintain adequate hygiene or prevent or correct contractures and therefore preserve function. Intrathecal pumps, inserted into the spinal cord, allow adjustable drug delivery. Baclofen, the drug of choice for the intrathecal pump, can be given in higher doses; use of the pump avoids the side effects often encountered when the drug is taken orally. Relapses are more commonly reported in people on oral medications than those using ITB. People using ITB also report higher levels of satisfaction, less spasticity, and fewer painful spasms compared with those on oral medications.

**Pain**

Both nociceptive and neuropathic pain can be present in people with MS. Therefore it is important to discern the type of pain in order for the most appropriate treatment to be rendered. Nociceptive pain can often be treated with analgesics (acetaminophen, nonsteroidal antiinflammatory drugs [NSAIDs], or opioids) and is more amenable to physical therapy (discussed later under rehabilitation management). Neuropathic pain generally requires pharmacological intervention, although an interdisciplinary team approach may be valuable. First-line medications for neuropathic pain that occurs in the spinal cord are calcium channel blockers (gabapentinoids) or N-methyl-D-aspartate (NMDA) antagonists (ketamine). When pain is present in the head, the primary treatment is opioid drugs such as antidepressants (tricyclics) or anticonvulsants (gabapentin or pregabalin).

In the case of trigeminal neuralgia, the first choice is often carbamazepine. Refer to Chapter 32 on pain management for additional information.

**Mobility**

Physical rehabilitation is the primary intervention used to manage mobility dysfunctions. However, one medication has recently been FDA approved to improve gait. In clinical studies dalfampridine (Amphra) demonstrated the ability to improve walking speed in people with MS. However, changes in the quality of gait or movement were not measured.

**Tremor**

Tremor management using medications such as isoniazid, carbamazepine, ondansetron, or cannabis extract has been minimally effective. Surgical interventions including stereotaxic thalamotomy and deep brain stimulation have been studied, but the evidence to support the effects on functional status and disability is lacking. The effectiveness of other options including physical therapy, tremor-reducing orthoses, and extremity cooling have yet to be proven beneficial in clinical trials.
**Rehabilitation Management**

**Overview**

Chronic neurodegenerative conditions, such as MS, result in a loss of physical and cognitive function from the destruction of neurons and from a lack of activation of the affected systems. People with MS experience physical and cognitive impairments potentially leading to inactivity and resultant deconditioning (Figure 19.5). This often becomes a cycle that is difficult to break. One question that frames the rehabilitation strategy chosen is whether the focus should be compensation for or restoration of lost function. Compensation includes interventions such as wheelchairs or walkers to assist with mobility or braces for absent or inadequate muscle power. Restoration is aimed at increasing the capacity of the system—for example, maximizing cardiovascular endurance by increasing maximal oxygen uptake or restoring full ROM. Therefore, prescribing programs, activities, and exercises that provide an adequate stimulus to produce adaptation is critical to restore function or improve motor and cognitive performance. Although each patient case is unique, the most likely answer is that both strategies will be employed. The challenge for rehabilitation professionals is to sort out how much of a patient’s dysfunction arises from neurodegeneration, which necessitates compensation, and how much occurs from inactivity and system deconditioning, in which case system capacity can be restored to some extent. Rehabilitation professionals must choose therapeutic interventions based on whether compensation or restoration is the goal.

Rehabilitation for people with MS occurs in every setting: inpatient hospitals, outpatient clinics, skilled nursing facilities, home care settings, and the community. With the current climate of decreasing access to and reducing coverage for rehabilitation, therapists must be able to make evidence-based arguments to primary care providers and insurers, as well as patients, to support effective therapeutic interventions that will achieve the goals of optimal physical and cognitive functioning, safety, and QOL.

For rehabilitation professionals managing people with MS, the International Classification of Functioning, Disability and Health (ICF) model (refer to Chapter 1) provides an excellent framework for assessment and management regardless of the setting in which the patient or client is encountered. Although guided by the opening interview and chart review, the initial assessment must include how the individual with MS is functioning in home, at work, and in recreation environments and which impairments of bodily structure or function might be contributing to the identified activity limitations and participation restrictions. Rehabilitation professionals must consider how personal and environmental factors may impede or facilitate achievement of rehabilitation goals. Personal factors in people with MS may include whether the patient is heat intolerant, experiences MS-related fatigue, or has the confidence or motivation to perform certain tasks. Environmental factors that may be of particular importance for the patient with MS may be living in a hot climate or having access to cooling equipment such as air conditioning or cooling garments. It is critical to understand how the disease affects the lives of both individual patients and their caregivers. Outcome measures designed to test impairments, activity, and participation, along with assessments of environmental and personal factors, will help health care professionals understand the deficits of their patients and determine the best place to focus rehabilitation efforts and monitor the patient’s response to intervention.

Because of the myriad CNS lesions and variable clinical presentations in people with MS, there is no one approach that is the gold standard for rehabilitation management. Whatever the approach, evidence is growing that rehabilitation is beneficial. Intensive inpatient therapy programs provide long-term improvement in a number of functional skills, participation, and QOL but may not change underlying impairments. Prospective studies have shown that intensive inpatient rehabilitation improves disability and QOL and that these benefits can be long lasting.

High-intensity programs in the outpatient clinic or home environment offer evidence of short-term symptomatic changes that have translated into improved participation and QOL.
Assessment
The initial interview must include a quick screen or questioning about the body systems and areas that are commonly impaired in people with MS and the problems commonly encountered: motor strength, coordination, spasticity, sensory disruption (vestibular, visual, and somatosensory), bladder control, depression, and cognition. If impairments are present, there is a strong likelihood of negative impact on the patient’s ability to perform ADLs or participate in activities related to work, home, and leisure. All patients with MS must be asked if they have fallen in the last 6 months because of the high rate of falling in people with MS. Results of the interview and chart review will help develop hypotheses about which potential impairments might be contributing to the patient’s or client’s physical or cognitive dysfunctions. Therefore the examination needs to be designed to observe the problematic tasks and test the hypotheses developed.

During the assessment, examiners must determine if the problems identified by the patient (or those found by the assessor) fit within their scope of practice or whether the patient requires a referral to an appropriate health care professional. A good example is identifying people with depression using a quick two-question screen. According to Mohr and colleagues, these questions are 98.5% sensitive for identifying major depressive disorder. The two questions are (1) “During the past 2 weeks, have you often been bothered by feeling down, depressed, or hopeless?” and (2) “During the past two weeks, have you often been bothered by having little interest or pleasure in doing things?” An answer of yes to either question should trigger a referral to the patient’s primary care provider for follow-up.

The physical examination might then start with testing the patient’s ability to perform functional activities that the patient or his or her caregivers have identified as problematic. This might include performance of transfers, gait, ADLs, or cognitive tasks as well as the specific activities that the person states are compromised in his or her work, home, and recreational life. There are several measures of QOL that also cover participation issues relevant to people with MS.

Comprehensive lists of standardized tests and measures for impairment, activity limitations, and participation restrictions or QOL are provided in Chapter 8. The next section of this chapter will primarily focus on the tests and measures found to be valid and reliable in the examination of individuals with MS.

Assessing Body System Problems Contributing to Activity Limitations
In general, standardized methods of examining muscle strength and endurance, somatosensation, vision, coordination, cardiovascular status and endurance, posture, muscle tone, reflexes, ROM, pain, and cognition are useful in examining patients with MS. As with many neurological conditions, abnormal posturing or pain may necessitate using nonstandardized test positions or methods that must be noted in the patient documentation. If a patient is unable to attain the normal test position while performing a muscle strength test, the assessed strength is noted along with the position in which the muscle or muscle group was tested.

Spasticity. Spasticity can be measured using resistance to passive ROM and Ashworth and Modified Ashworth Scales. However, because these scales measure spasticity at rest, they may not reflect the degree to which spasticity may be interfering with function. Careful observation of the patient’s movements may also inform the clinician about how spasticity is affecting the patient’s ability to move.

Ataxia and Incoordination. Few standardized tests have been developed to specifically measure ataxia. One recent test is the Scale for the Assessment and Rating of Ataxia (SARA). Although this test has yet to be validated in people with MS, it has good reliability and validity in patients with cerebellar dysfunction, a common problem in people with MS.

Tests of nonequilibrium coordination are designed to measure the presence of dysmetria or dysdiadochokinesia, both of which occur in patients with MS. However, these tests (including finger to nose, heel to shin) are somewhat subjective and are therefore difficult to use to demonstrate improvement after an intervention. However, using a stopwatch during these tests can be an important tool to record objective data. Count the number of repetitions of a given activity performed in a set amount of time (e.g., how many alternating forearm supinations and pronations can be performed in 30 seconds), or record the time it takes to complete a set number of repetitions of a given activity (e.g., how long it takes to complete five alternating supination-pronation movements). Refer to Chapter 21 for additional assessment tools.

Vestibular Dysfunction. The vestibular system is affected by MS both centrally (lesions in the vestibular nuclei or cerebellum) and at the entry site of cranial nerve VIII. However, benign paroxysmal positional vertigo (BPPV) can also occur. The techniques used to assess and treat the effects of vestibular disorder in an individual with MS are the same as those discussed in Chapter 22. When vestibular symptoms are present, Williams and colleagues suggest evaluation using computerized platform posturography (CPP) in people with MS with minimal to mild disability. It is important to keep in mind that the patient with MS will often have additional problems that might require modification of the vestibular intervention—for example, heat intolerance or additional visual or somatosensory deficits.

Fatigue. Identifying if and when fatigue occurs in individuals with MS is important to assessment and the structuring of intervention. Questions should address the type of fatigue, whether mental or physical; when during the day it occurs; whether it is related to physical or mental exertion; and what the person with MS does, if anything, to relieve it. In addition, fatigue-related self-report scales can help the rehabilitation professional gain an understanding of the perceived impact that fatigue may be having on a patient with MS. Two of the commonly used scales are the Modified Fatigue Impact Scale and the Fatigue Severity Scale. These measures may also aid the therapist in determining if the intervention had any impact on the patient’s perceived level of fatigue.

Cognition. The PASAT, recommended by an expert panel of the National MS Society, is a test for cognitive impairments in people with MS. A more recent test, the Audio Recorded Cognitive Screen (ARCS), appears to be a more comprehensive cognitive assessment developed for people with dementia but the psychometric properties have not yet been determined in people with MS. However, Lechner-Scott and co-workers found that compared with
the PASAT, the ARCS was similar in detecting impairments of cognition and more sensitive at identifying problems with memory or executive impairments.

**Assessing Activity Performance and Participation**

Outcome measures assess the ability of an individual to perform an activity or task as well as assess the perception of the person to use those tasks to fulfill life roles. Following are activity and participation measures commonly used in people with MS. An individual’s perceived ability to participate may also be included in some QOL outcome measures that are included in the following sections.

**Balance.** Balance is foundational to upright movement and is produced by a complex interaction among sensory inputs, central processing, and motor responses. It can be discussed under both body structure and function or activity. In either case balance dysfunction has been identified in people with MS with minimal as well as more advanced disability. Cameron and Lord report the three most common problems with balance to be delayed response to postural perturbations, increased body sway while standing quietly, and an inability to move outside the base of support.

Whereas some balance tests focus on stationary or static tasks that allow observation of body sway in standing, including single-leg stance test, Romberg test with eyes open or eyes closed, tandem stance, and CPP, others add movement and challenge dynamic balance (Functional Reach Test, Tinetti Performance-Oriented Mobility Assessment [POMA], and Berg Balance Scale [BBS]). Other tests challenge anticipatory balance (reactions to perturbations related to self-generated movement) or reactive balance (perturbation tests, CPP). Frzovic and co-workers found that single-leg stance, tandem stance, response to external perturbations, and the Functional Reach Test were able to distinguish people with MS from healthy controls.

Several authors have studied measures of balance in people with MS. Cattaneo and colleagues determined that four tests measuring balance during standing and gait and self-perception of balance had good intrarater and interrater reliability. The two tests measuring balance during standing and movement were the BBS and the Dynamic Gait Index (DGI).

CPP provides an objective assessment of sensory contributions to balance dysfunction in people with MS. In particular, the Sensory Organization Test is useful in identifying the relative sensory contributions (visual, vestibular, and proprioceptive) to stationary balance and response to perturbation. Understanding the sensory conditions under which the patient loses balance and falls assists the therapist in providing exercises that will challenge these conditions in a safe and controlled manner. For example, the patient who relies heavily on visual input to maintain balance (conditions with eyes closed in the Sensory Organization Test) would be provided exercises and activities that challenge the vestibular and proprioceptive systems, such as standing on foam while the eyes are closed.

Developed by Horak and colleagues, the Balance Evaluation Systems Test (BESTest) is an instrument examining complex balance disorders that includes the six domains that underlie orientation and postural stability: biomechanical constraints, stability limits and verticality, transitions and anticipatory postural reactions, reactive postural responses, sensory orientation, and stability in gait. Both interrater reliability in people with parkinsonism and content validity are good, but testing in other populations has not yet been completed. There is an abbreviated version of the BESTest, the mini-BESTest, that covers four of the six systems, focusing on dynamic balance. These promising tests may offer the clinician a better way of identifying which components of orientation and postural control are dysfunctional, which may allow more targeted interventions.

The Activities-specific Balance Scale (ABC) is a questionnaire that rates people’s self-perception of how confident they are to perform activities that challenge their balance. The Dizziness Handicap Inventory (DHI) assesses three domains of disability related to dizziness: physical, emotional, and functional. The sum score or each subscale score can be reported. Higher scores mean greater levels of handicap and disability. Cattaneo and colleagues found that both the ABC and DHI tools discriminated between fallers and nonfallers and were therefore good predictors of fall status in people with MS. Refer to Chapter 22 for additional information on balance.

**Gait.** Gait can be measured in myriad ways depending on the goal of the assessment. Speed, distance, and quality may all be important to the patient and therapist. Observational gait analysis is the gold standard for clinical measurement of gait quality. Although motion-analysis laboratories are able to provide detailed kinetic and kinematic assessment of joint angles and gait cycle, it is costly and typically not available in most clinical settings. Instrumented mats such as the GaitRit can provide clinicians with temporal and spatial gait parameters such as step length, step width, cadence, and single-leg support and double-leg support times. Although this is less costly than motion analysis, it may still be out of reach for many clinics. Gait speed and velocity can also be measured by having the patient walk a given distance while being timed. These walks can occur at a self-selected pace or as fast as the person can walk safely. Several short-distance timed tests exist, the 25FTW and the timed 10-meter gait test, both of which have shown to have good reliability and sensitivity to change. The 6-minute walk test (6MWT) measures walking endurance and is recommended by the NMSS Task Force on Clinical Outcome Measures as a measure of walking ability that is sensitive to change. Gijbels and co-workers report that the 6MWT was better at predicting habitual walking in people with mild to moderate MS than the 25FTW. However, the 25FTW may be more sensitive to change when compared with the EDSS. The 6MWT distance was reduced in people with MS compared with healthy controls and was inversely related to disability.

Two additional performance-based tests, the DGI and the Timed Up-and-Go Test (TUG), combine walking with other functional tasks. The DGI measures the ability of an individual to walk while adding various challenges such as slowing down or speeding up, head turning, stepping over or around obstacles, and stair climbing. It was developed to assess gait dysfunction associated with peripheral vestibular disease. McConvey and Bennett found the DGI to be a reliable and valid tool for use in people with MS. The TUG
test combines walking with transfers and turning. It is frequently used in both clinical and research settings and has been shown to be reliable in measuring function in people with MS.93

The Multiple Sclerosis Walking Scale—12 (MSWS-12) is a 12-item patient-rated questionnaire that measures the perception of the impact of MS on walking ability. This scale has good reliability and validity and may be very useful to document patient perceived change in walking ability before and after intervention.100,101

**Upper-Extremity Tests of Function.** Movement impairments of the upper extremities can result in decreased ability to perform ADLs and other functional activities. Standardized tests such as the Box and Block Test (BBT)102 or the NHPT103 provide objective data about unilateral manual dexterity or the ability to manipulate objects. Both tests are inexpensive but do require some equipment and a stopwatch. The NHPT is part of the MSFC and therefore has been used extensively in evaluating people with MS.

**Composite Tests.** An expert panel of the NMSS recommended the use of the MSFC,54,104 including the 25FTW, the NHPT, and the PASAT. The MSFC has been tested against lesion load as measured via MRI, EDSS scores, and QOL measures, showing that it has good validity and reliability and is sensitive to change.104-106 Each component scale of the MSFC can also be used independently to monitor physical and cognitive function as written previously.

**Assessing Quality of Life**

QOL measures are patient-report tools that evaluate the value a person places on his or her abilities and limitations and how these affect the individual’s social, emotional, and physical well-being. Many of these tools include questions that address an individual’s perception of how well he or she is able to fulfill life roles and how the disease affects this participation. In a meta-analysis of exercise training on QOL in people with MS, Motl and Gosney,107 found that disease-specific measures of QOL detected larger changes than generic QOL measures. Several measures have been commonly used to evaluate people with MS: the Multiple Sclerosis Quality of Life–54 (MSQLI-54)113 and the Multiple Sclerosis Quality of Life Inventory (MSQOL).114 The multidimensional MSQOL-54 was based on the Health Status Questionnaire (SF-36), with 18 additional items specific to MS covering fatigue, and cognitive and sexual functioning. There are 12 subscales that cover physical function, role limitations—physical, role limitations—emotional, pain, emotional well-being, energy, health perceptions, social function, cognitive function, health distress, overall QOL and function, and change in health. The measure takes about 15 minutes to complete and requires 15 to 20 minutes to score. Reliability is good to excellent in people with MS.113

The MSQOLI was developed by the Consortium of Multiple Sclerosis Centers Health Research Subcommittee in 1997. It is composed of 10 components covering issues important in MS. It includes the Health Status Questionnaire, Modified Fatigue Impact Scale, MOS Pain Effects Scale, Sexual Satisfaction Survey, Bladder Control Scale, Bowel Control Scale, Impact of Visual Impairment Scale, Perceived Deficits Questionnaire, Mental Health Inventory, and MOS Modified Social Support Survey. It takes about 45 minutes to administer the complete set of questionnaires and does not provide a sum score for all tests. There is good test-retest reliability for the MSQOLI even in people with MS and cognitive dysfunction.109 A shortened version of the tool exists, but the psychometric properties have not been thoroughly tested.

**Disease Severity Measures**

Disease severity is a measure of disablement. Interventions that change function (e.g., improve walking distances or decrease reliance on assistive devices to move) can reduce disability. There is also compelling evidence that exercise may actually modify disease progression in people with MS. Therefore disease progression may be used to assess the impact of an intervention on the patient’s perceived level of disability. Although the EDSS11 is the gold standard for assessing disease severity, it requires a trained primary care provider to administer. Disease Steps112 and Guy’s Neurological Disability Scale (GNDS)108 are two additional disability scales that have demonstrated good correlation with the EDSS. Whereas Disease Steps must be administered by a professional, GNDS can be given to patients to complete on their own.110

**Interventions**

The goals of rehabilitation for persons with MS are to maximize and maintain function and prevent complications so that they can participate fully in all aspects of their lives. The variable presentation that people with MS can manifest requires rehabilitation professionals to be flexible and creative. The plan of care developed to manage a patient must be linked to the impairments, activity limitations and participation restrictions identified during the assessment. Research provides evidence for the most effective interventions and must be coupled with the desires and needs of the individual with MS. The rehabilitation program must be negotiated with the patient/client in consultation with caregivers when available or appropriate.

The National Clinical Advisory Board of the National MS Society recommends that rehabilitation occur whenever there is a sudden or gradual decline in function or an increase in impairment that has a negative impact on an individual’s safety, independence, mobility, or QOL. In addition, it is recommended that rehabilitation be a part of a comprehensive health care plan at all stages of the disease.115

Regardless of the type of intervention chosen, evidence is growing that increased activity, whether cognitive or physical, may have a neuroprotective effect on the brains of people with neurological insults. In fact, Golzari and colleagues116 demonstrated that an 8-week, 24-session, combined exercise program improved muscle strength and balance and reduced disability in people with MS. In this study, levels of proinflammatory immune system mediators were measured before and after the intervention. The authors demonstrated that this dosage of exercise reduced markers of inflammation in the blood. This is one of the first studies in people with MS showing that inflammation and therefore the disease process may be altered by the application of an exercise intervention, suggesting a role for rehabilitation in neuroprotection and not simply symptom management. This also implies that rehabilitation, specifically
exercise, should occur early in the course of the disease and not only after clinical disability has occurred. However, the exact dosage, intensity, or type of exercise required to produce activity-dependent neuroplasticity is not yet known. At least one study in an animal model of MS, experimental allergic encephalomyelitis, has shown the beneficial effects of exercise.\(^\text{117}\)

In prescribing a rehabilitation program for persons with MS, each individual’s level of fitness and physical and cognitive resources including memory, judgment, strength, endurance, spasticity, balance, and coordination must be taken into consideration. In addition, therapists must investigate the person’s level of fatigue and heat sensitivity. If present, these factors will require modification of the rehabilitation program, including where the activity is performed, in what environment, and the time of day in relation to fatigue level and the other tasks the individual must perform. In other words, to be successful, the rehabilitation program must fit into the framework of the person’s life.

Rehabilitation can occur in a variety of locations: inpatient, outpatient, home, and the community. Figure 19-6 shows a physical therapy–led community-based exercise program for people with MS in which group activities addressing strength, balance, and endurance are modified for each individual. In addition, a number of health providers can be members of the rehabilitation team, including nurses, occupational therapists, physical therapists, speech-language pathologists, psychologists, neuropsychologists, and physicians.

**Exercise**

Historically, exercise was thought to worsen disability and bring on exacerbations. Medical advice warned patients that overexertion could hasten relapse and progression. There now exists clear evidence that this is not the case. Regular, appropriate exercise has been shown to increase strength, aerobic capacity, overall function, and QOL. In 1996 Petajan and colleagues published a seminal study in which a 60% \(V\hat{o}_2\) max aerobic ergometer exercise program was well tolerated in people with MS and did not provoke remission.\(^\text{118}\) After 10 weeks, participants had improvements in \(V\hat{o}_2\) max, work capacity, isometric strength, and blood lipids and reduced depression, anger, and fatigue. In a 2009 systematic review of the literature,\(^\text{119}\) exercise was shown to be an effective intervention for people with MS to improve muscle strength, endurance, mobility-related actions, and to a lesser extent mood compared with control conditions. This evidence did not suggest the superiority of one particular type of exercise program over others. It is very important to note that adverse effects were rarely seen in any of the exercise studies, and when they did occur they did not last for longer than 24 hours, indicating that exercise is safe for people with MS.

In a review of the exercise literature, White and Dressendorfer\(^\text{120}\) recommend that endurance exercise programs for people with MS with mild to moderate disability use the following guideline: perform regularly, two or three sessions per week, at an intensity of 65% to 75% heart rate maximum, and last 20 to 30 minutes per session. Resistance exercise should include 15 to 18 repetitions for one to three sets initially with a goal of increasing to three to four sets. Training should last at least 12 weeks.\(^\text{121}\) Owing to heat intolerance, exercise should incorporate intermittent rest periods that allow heat to dissipate.\(^\text{120}\) Heesen and colleagues\(^\text{122}\) developed a guideline for exercise prescription for people with MS for all levels of disability (Table 19-3).

Prescribed early in the course of the disease when mild to moderate disability is present, exercise can be used to restore function by reducing physical or cognitive decline from disuse or deconditioning. As clinical disability accrues in the later stages of the disease, exercise may then be used to compensate for missing function or prevent secondary complications—for example, stretching hip adductor muscles with decreased range of motion to allow adequate personal hygiene to occur.

**Evidence-Based Interventions for Specific Problems**

**Fatigue**

Fatigue is one of the most frequent and disabling symptoms associated with MS and is best managed with a multidisciplinary team composed of physicians, physical therapists, occupational therapists, and nurses. As described earlier, the causes of fatigue can be divided into two basic categories:
primary and secondary. Primary fatigue related to demyelination and neurodegeneration may have fewer options for treatment. Secondary fatigue caused by deconditioning, comorbidities, depression, poor nutrition, heat intolerance, sleep disturbance, and medications may be more easily managed. Several strategies for fatigue management have been reported and show promise; however, few research studies have demonstrated effectiveness in randomized controlled trials or in comparisons among approaches. Interventions for fatigue management include cooling devices, energy conservation education training, exercise, and a multifaceted class aimed at teaching people with MS how to manage their fatigue.

One study found that the cooling suit was shown to improve all dimensions of fatigue on the Fatigue Impairment Scale (physical, cognitive, and psychosocial) in a small multiple-case study. Although recommended in the clinical practice guidelines on fatigue and MS by expert opinion and anecdotal reports of people with MS, little additional evidence exists to support cooling as a therapeutic intervention. Two additional studies have shown that cooling garments can reduce symptoms of fatigue and improve ambulatory ability.

Exercise shows promise as an intervention that can improve fatigue for people with MS that may improve muscle weakness caused by disuse and deconditioning. However, no one type of exercise, resistance or aerobic, or program has been proven most effective. One program included a 5-day-per-week, 30-minute bicycle aerobic training program for 4 weeks that improved fitness and showed a tendency for reduced fatigue. This study had an age, sex, and activity level control group. Di Fabio showed that a prolonged outpatient rehabilitation program in patients with progressive MS led to a decrease in MS-related symptoms, including fatigue. However, there was no control group. A randomized study comparing bicycle training with yoga found that fatigue improved in both groups, with neither group shown to be better than the other.

Energy conservation is defined by the fatigue and MS guidelines of the Multiple Sclerosis Council for Clinical Practice Guidelines as energy effectiveness and includes an analysis of individuals’ home, work, and leisure activities and the environments in which they occur in order to develop activity modifications designed to reduce fatigue. This can include a variety of strategies such as reducing energy expenditure through activity and modification, workspace organization and improving efficiency of movements; balancing work and rest periods; delegating tasks; evaluating standards and prioritizing activities; and using assistive technologies that conserve energy usage.

In a randomized controlled trial, a 6-week community-based energy conservation class using the strategies listed previously was compared with a wait-list control group. Immediate post-course improvements in fatigue were noted and were present after a 1-year follow-up period.

The multidimensional fatigue management class “Fatigue: Take Control” was developed based on the recommendations of the Fatigue Management Guidelines of the NMSS from 1998. The content of the 6-week class includes many of the aspects of fatigue management education and training that were described previously. The pilot study found that participants had less fatigue compared with a wait-list control group. These classes are often offered by local chapters of the NMSS.

Patients may need to be prescribed assistive devices for ADLs. People with MS who have spasticity have a greater cost of walking. Using wheeled mobility for longer-distance outings (to the shopping mall, an extended event, on vacation) can conserve energy and extend the time a person can participate in activities of importance to him or her. However, therapists should be aware that using assistive devices such as walkers or crutches actually increases energy expenditure for elderly people, and therefore the need for improved support must be balanced with the increased energy burden an assistive device might add.

**Spasticity**

Several rehabilitation strategies to manage spasticity are available, including ROM, stretching, light pressure or stroking, cold therapy, electrical stimulation, and education. Although none of these interventions is supported by

<table>
<thead>
<tr>
<th>LEVEL OF DISABILITY</th>
<th>EDSS LEVEL</th>
<th>TRAINING PROGRAM</th>
</tr>
</thead>
<tbody>
<tr>
<td>None: no fatigue or thermosensitivity</td>
<td>0</td>
<td>Full exertion, aerobic and resistance exercise, no extreme sports</td>
</tr>
<tr>
<td>Minimal: limited fatigue and heat sensitivity; minor balance or gait problems</td>
<td>1-2</td>
<td>Monitored exercise program including strengthening and endurance using a variety of exercise types, precooling if heat-sensitive, avoid overtraining</td>
</tr>
<tr>
<td>Moderate: limited gait; may have spasticity, weakness, ataxia, balance problems</td>
<td>3-5</td>
<td>Deficit-driven exercise protocols including strengthening and endurance training using methods tolerated, walking, cycle ergometry, precooling if needed</td>
</tr>
<tr>
<td>Severe: cannot participate in all daily activities; short-distance, aided walking only</td>
<td>6-7</td>
<td>Movement preservation, stretching, targeted strengthening needed for task-specific training</td>
</tr>
<tr>
<td>Bedridden</td>
<td>8-9</td>
<td>Primarily passive movements to maintain motion, breathing exercises</td>
</tr>
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EDSS, Expanded Disease Severity Scale.
strong research evidence, many are used routinely in clinical practice (ROM, stretching). Other approaches (cold therapy, light pressure or stroking) are recommended for use in conjunction with stretching or ROM programs. Regardless of the technique employed, educating individuals and caregivers about the importance of adhering to a spasticity management program is essential. The Multiple Sclerosis Council for Clinical Practice Guidelines recommends, based on expert opinion, stretching a muscle with spasticity for 60 seconds or longer or using a prolonged stretch, lasting hours, with braces or splints.

Cold can be applied in a number of ways: baths, towels, or cooling garments. There are multiple quasi-experimental research studies that suggest an improvement in spasticity for a brief period after cooling; however, the number of subjects and study methods make these results equivocal. Nilsagård and co-workers found subjective reports of improved spasticity after a single session of cooling, although no statistically significant differences in spasticity measures were found.

**Balance and Postural Control**

Balance is foundational to the ability to stay upright and perform dynamic movements. It is a frequent problem in people with MS and results in a person limiting his or her participation in home, work, and leisure activities. Abnormalities of balance along with cane use and poor performance on tests of balance and ambulation can increase the risk of falling. Other fall risk factors that have been identified include fear of falling, male sex, poor concentration or forgetfulness, and urinary incontinence. Rehabilitation programs must be based on a thorough understanding of the impairments and personal and environmental factors that may be contributing to the balance dysfunction. Cattaneo and co-workers compared the effects of three balance interventions on falling and other measures of balance. Three rehabilitation groups were included: one in which motor and sensory strategies were targeted, the second focusing on motor strategies alone, and the third group not receiving balance-specific training. The greatest reduction in falls and improvement on the BBS were associated with group one, and the least with group three. Hayes compared 12 weeks of standard physical therapy with high-intensity resistance exercise (60% to 80% maximal contraction) added to standard therapy and found that standard therapy produced better balance outcomes. In addition, strength and the ability to ascend and descend stairs were all better in the standard therapy group. Importantly, people with MS tolerated the high intensity resistance exercise without problems. One pilot study found that a 12-week, biweekly aerobic exercise program did not improve balance as measured by the Functional Reach Test but did result in an improvement in walking distance. For additional intervention strategies on balance, refer to Chapter 22.

**Mobility**

People with MS rate gait as one of the most important bodily functions; gait is often adversely affected in people with MS. Gait disturbances have been observed in people with MS even before disability is measured on the EDSS scores. Lesions in the brain and spinal cord produce a wide variety of potential impairments that can adversely affect gait. In a review article by Kelleher and colleagues, imbalance, fatigue, spasticity, incoordination, muscle weakness, and sensory system impairments were all reported to negatively affect ambulation ability. Therefore addressing each of these impairments has the potential to improve gait. A recent literature review of therapeutic interventions for mobility problems suggests that a variety of different methods can be used to improve ambulation. Snoek and Motl performed a meta-analysis of exercise studies aimed at improving walking mobility in people with MS and found that greater effects were associated with supervised exercise training, programs of less than 3 months’ duration, in mixed samples of people with RRMS and progressive MS.

Task-specific gait training has been evaluated in people with MS. A randomized controlled trial compared two different treatment groups—facilitation and task-specific training—that each received 15 to 19 1-hour treatment sessions over 5 to 7 weeks and found that both improved 10-m gait speed, stride length, and balance; however, there was no control group. Treadmill training has been investigated in several small, pilot or case studies with promising results of improved QOL, energy expenditure, and gait parameters.

Several exercise studies have an association with improved gait. A combined resistance and aerobic home program lasting 23 weeks improved gait speed for short and longer distances in exercise compared with a control group. Rampello and co-workers compared a neurorehabilitation program with an aerobic training program of similar duration (three times per week for 8 weeks). The authors found that aerobic training improved walking distances and speeds and measures of aerobic capacity over the neurorehabilitation group. Both groups had QOL improvements in emotional well-being and health distress; the neurorehabilitation group demonstrated improved mental health.

An additional technique that shows promise for improving mobility in people with MS is an evaluation and intervention approach that uses small amounts of weight placed on the torso in response to identified balance dysfunction. Balance-Based Torso-Weighting (BBTW) is an intervention that uses directional loss of balance in both static and dynamic assessment to determine where small amounts of weight (generally less than 1% to 1.5% of body weight) are placed in a treatment orthotic called BalanceWear. The BalanceWear orthotic can be worn during the performance of activities in therapy or daily for home, work, or leisure activities. A recent randomized controlled trial in people with MS who reported gait abnormalities showed that when wearing the weighted BalanceWear orthotic participants increased their gait speed compared with no weight controls, and improved TUG scores compared with a standard weighted control.

When people with MS do not respond to therapeutic interventions to restore function, mobility assistive devices such as canes, crutches, walkers, wheelchairs, and scooters are used to enhance mobility through compensation. Mobility-assisted technology (MAT) can improve function in people with moderate to severe impairments of ambulation and may reduce activity limitations and participation restrictions by reducing fatigue and enhancing energy conservation to allow greater involvement in work, family, social,
vocational, and leisure activities. Other MAT technologies include functional electrical stimulation (FES), neuroprostheses, and orthotics. FES is applied to specific muscles or muscle groups to activate weak muscles. Some of these stimulators can be built into a neuroprosthesis that can be set up for use during exercising or walking. Orthotics such as the ankle-foot orthosis (AFO) or hip flexion assist orthosis (HFAO) can compensate for muscle weakness in the lower extremity, improve foot and knee positioning, and reduce energy expenditure. Therapists often work cooperatively with orthotists to ensure proper fit. Use of wheeled mobility devices such as a manual wheelchair, power wheelchair, or scooter requires a formal evaluation by an occupational or physical therapist with justification that it is required for mobility at home at least on a part-time basis. Therapists must take a long-term view of the projected needs of the patient when prescribing wheeled mobility, as most insurance companies will replace this equipment only every 5 years.

**Pain and Dysesthesias**

The occurrence of pain in people with MS is often underestimated. Pain can be acute, as in optic neuritis or Lhermitte syndrome, or chronic, as in dysesthesias in the limbs or joints due to mechanical pain related to abnormal positions or repeated movements that cause abnormal wear and tear on the musculoskeletal system. Occupational and physical therapists can address poor body mechanics and weakness and poor movement patterns with retraining, and soft collars may help reduce Lhermitte syndrome. However, little evidence supports these interventions. Transcutaneous electrical nerve stimulation has been suggested anecdotally by Kassirer as beneficial for reducing pain. Cognitive-behavioral therapy has been researched for managing chronic pain, but little evidence exists for using it in people with MS.

**Bladder Dysfunction**

Urinary incontinence and retention are common and often embarrassing problems for people with MS. Patients may be advised to avoid bladder irritants including caffeine, alcohol, concentrated urine, and infection. Physical therapists may work with patients to assess the factors contributing to bladder dysfunction by retraining hyperactive or weak pelvic floor muscles using biofeedback techniques and exercise. Nurses may need to teach patients with urinary retention intermittent catheterization. Refer to Chapter 29 for additional information on pelvic floor dysfunction and its treatment.

**Cognition**

Strategies for managing cognitive impairments include compensation techniques such as memory notebooks, diaries, calendars, and computer-assisted programs for memory, attention, or other executive functions. Neuropsychologists, speech-language pathologists, and occupational therapists can all direct cognitive rehabilitation programs. Strategies for coping with cognitive impairments are often shared with the other members of the health care team for reinforcement with patients. There is growing evidence to support psychological interventions for people with mild to severe MS-related cognitive deficits, aimed at alleviating depressive symptoms and helping people cope with and adjust to their impairments. However, the evidence is not yet convincing for specific programs addressing attention and executive functioning. O’Brien was able to recommend the use of a modified story technique to address learning and memory deficits in people with MS. In a systematic review Maitra found that cognitive behavioral therapy programs performed by occupational therapists were positively correlated with improvement in Functional Independence Measure (FIM) scores. Refer to Chapter 27 for additional information regarding interventions with individuals with cognitive problems.

**Dysphagia and Dysarthria**

Dysphagia or difficulty with chewing and swallowing becomes more prevalent in people with MS as the disease progresses. Therapists facilitate proper swallowing with exercises that will improve posture to prevent aspiration and strengthen muscles of mastication. Other interventions may include diet modifications and education for the patient and his or her family or caregivers. Dieticians may be consulted to facilitate proper food choices.

Dysarthria from the disruption of muscular control in the central and peripheral speech mechanisms leads to abnormalities of speed, range, timing, strength, sound, and accuracy of speech movements. Speech-language pathologists determine therapy programs that take into consideration the stage of the disease and speech quality. Typical programs may include exaggerating articulation, increasing voice volume, and increasing strength of oral musculature. Exercise programs designed to increase respiratory muscle strength have not been successful in improving voice quality or production.

**SUMMARY**

This chapter has focused on the pathophysiology, clinical presentation, medical management, and rehabilitation of people with MS. Understanding the type of MS, clinical disability, and stage of the disease will help therapists determine the best assessment and intervention strategies for management of the rehabilitation program. Using the ICF framework will facilitate the assessment of the impairments, activity limitations and participation restrictions affecting patients and clients. In addition, including the environmental and personal factors present will help tailor the program to the patient’s needs. Using QOL measures developed for people with MS should help the therapist understand the entire range of problems that patients may have.

Many websites are available to assist therapists and their patients with MS to understand the disease and find resources to help them manage the disease. The National MS Society (www.nationalmssociety.org) and the Multiple Sclerosis Foundation (www.msfocus.org) are both excellent resources.

**References**

To enhance this text and add value for the reader, all references are included on the companion Evolve site that accompanies this textbook. This online service will, when available, provide a link for the reader to a Medline abstract for the article cited. There are 160 cited references and other general references for this chapter, with the majority of those articles being evidence-based citations.
CASE STUDY 19-1

INITIAL INTERVIEW
Mrs. P. is a 54-year-old woman with a 28-year history of RRMS. She was first diagnosed after her third daughter was born and remembers having a lot of trouble walking. Mrs. P. is concerned about her trunk weakness, back pain, and difficulty with walking; she often stumbles, especially when she does not use her single-point cane. She reports having fallen twice in the past year when she lost her balance and was unable to catch herself. One fall was at home, and one in the backyard. Therefore she has been using the cane more, especially on days when she feels off balance. Mrs. P. is limited to 10 minutes of walking and standing secondary to trunk fatigue and difficulty balancing. She is overweight and reports some bladder incontinence and heat sensitivity. Mrs. P. is a homemaker with 4 children; the youngest is 11. Leisure activities include playing the piano, singing, doing the Wii balance exercise for 20 min/day, and doing 10 minutes of treadmill walking at 3.0 mph after using a cooling vest. After her treadmill walking, she feels fatigue for 3 to 4 hours. Recently she has noted having more difficulty with singing and at times feels out of breath. Mrs. P.’s goals are improved posture, better breath control, no back pain, the ability to walk without stumbling or using a cane, and the ability to keep up with her children and her busy life.

ASSESSMENT
Mrs. P.’s Disease Steps classification is 3 (she uses a cane intermittently and is able to walk for 100 feet without it), and her EDSS score is 6.0. Vital signs are within normal limits (WNL) at rest and for exercise. This patient is cognitively intact and reliable in her response to questions. Her active and passive ROM is WNL throughout her extremities, trunk, and neck. She has selective motor control with normal tone. Manual muscle tests of bilateral upper extremity (UE) were normal, with the lower extremity (LE) 4/5 except for right hip flexion 3+/5, hip extension-abduction and plantarflexion 3/5. Abdominals 2/5, back extensors 3/5 (able to lift trunk against gravity through full range with difficulty and unable to take resistance). Sensation to light touch (LT), pain, and proprioception are intact throughout except for bilateral (B) feet, noted to have diminished sensation to LT. In sitting her posture is extremely slumped (from 30 to 45 degrees when fatigued) with notable thoracic kyphosis. She requires standby assist from supine to prone secondary to trunk weakness and instability. She requires use of B UEIs in weight bearing to move from sitting to standing. During observational gait analysis, she demonstrates an asymmetrical step length with the left longer than the right and a right heel strike that is notably loud or audible. Her TUG score is 8 seconds using B UEIs to stand up. Tinetti balance (POMA) = 14/16 and gait = 6/12 for a total score of 20/24 (19 to 24 risk for falls). Single-limb stance on the right = 4 seconds and left = 6 seconds, tandem stance = 4 seconds. Perturbation tests reveal loss of stability with an anterior nudge (posterior loss of balance [LOB]), posterior nudge (anterior LOB), and lateral and upper and lower trunk (LOB to opposite side). Rotational resistance tests to the right upper and lower trunk result in a stepping response, and the patient is unable to maintain stability, resulting in a stepping response. Results of rotational resistance tests to left upper and lower trunk are normal.

PLAN OF CARE AND GOALS
Mrs. P. had weakness in B LEIs, balance problems, and an unstable gait with an increased risk of falling, interfering with her functional mobility and QOL. The physical therapy plan included balance and gait training, improved posture and time standing, and increased endurance and cardiovascular fitness. Goals included a decreased fall risk with an improved Tinetti score of 25/28, improved B LE and trunk strength (4/5 in all muscle groups), improved endurance to stand and walk to 30 to 45 minutes, decreased back pain to 0 to 1/10 on most days, and improved endurance and cardiovascular fitness to 45 minutes to 1 hour in 12 to 24 weeks.

INTERVENTION
BBTW placement of 1.5 pounds of weight to the torso to address the perturbation and rotational asymmetries (posterior right upper and lower trunk and anterior near navel) was effective in reducing her kyphotic posture and impaired reactive balance control. The rigid component of the BBTW resolved back pain immediately; she felt better breath control and trunk support, and her Tinetti balance 15/16 and gait 12/12 = 27/28 significantly improved. TUG score remained the same, but she no longer needed her UE to stand up. Her posture also improved to 50% less kyphosis. During gait while weighted, a softer (inaudible) right heel strike was noted, and her step length was even. Mrs. P. expressed that she felt much steadier and more balanced with the BBTW BalanceWear vest and was thrilled to have no back pain.

Mrs. P. was seen in a managed care setting and was able to make significant progress with her physical therapy program. She was seen in physical therapy (1×/week × 1 month, 2×/week × 2 months, 1×/month × 3 months) to improve her posture, strength, balance, and fitness. Breath-control exercises using her diaphragm were implemented to improve singing, monitoring progress with an inspiratory spirometer. Because this patient understood the principles of exercise, she was advised to perform the specific exercise until she experienced a decrease in the quality of movement or the muscles fatigued. A general stretching program was initiated, including specific stretches to improve her posture in sitting and standing. For strengthening exercise she started with one set of 8 to 15 reps; resistance was increased by 2% to -5% when 15 reps were efficient for major muscle groups including hip flexion-extension-abduction, heel raises for plantarflexion, rowing (shoulder retraction) with yellow Thera-Band in standing, curl-ups in hook-lying, and quadruped weight shifts, which were performed 2× to 3×/week. To address her deconditioning, an interval treadmill-training program was implemented on alternating days. Recommendation was to maintain the intensity of 3.0 mph at 65% to 75% of HRmax and do intervals of four blocks of 3 minutes with 1- to 2-minute rest breaks in between and to continue to use the cooling vest before exercise. She continued with the Wii balance program, increasing to 30 minutes on the days she was not doing the treadmill. On those days she also performed specific balance exercises in the corner, beginning with her eyes open (single-leg stance; tandem stance). Mrs. P. was advised to use the BBTW rigid vest for 2 hours during functional activities and walking and to perform her exercises with it every other day. If she became less steady, she was advised to wear the vest an additional 1 to 2 hours per day.