Degenerative disorders

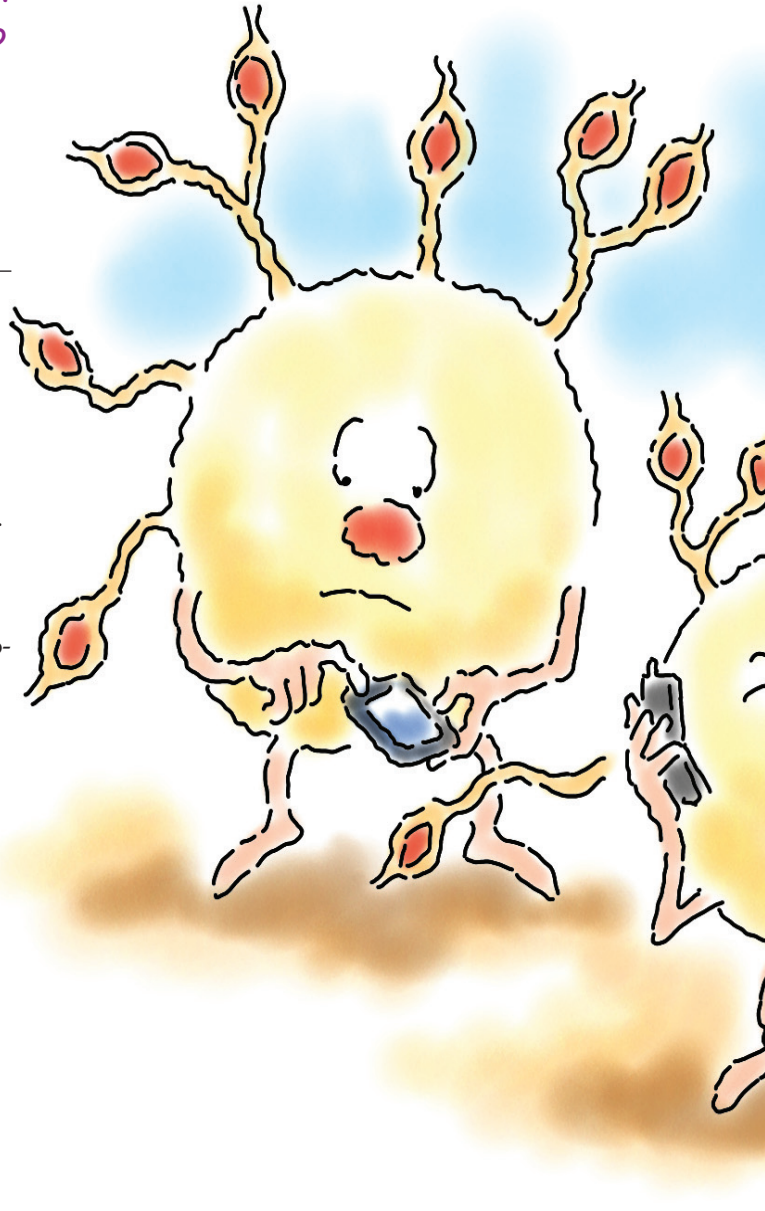
ALS

CE
2.0
ANCC
CONTACT HOURS

Amyotrophic lateral sclerosis and multiple sclerosis are both degenerative diseases that cause significant physical disability. Differential diagnosis is often difficult, so it's important to understand the characteristics of both disorders and their treatment options. We tell you what you need to know.

By Margaret J. McCormick, MS, RN
Clinical Assistant Professor, Nursing • Towson University • Towson, Md.

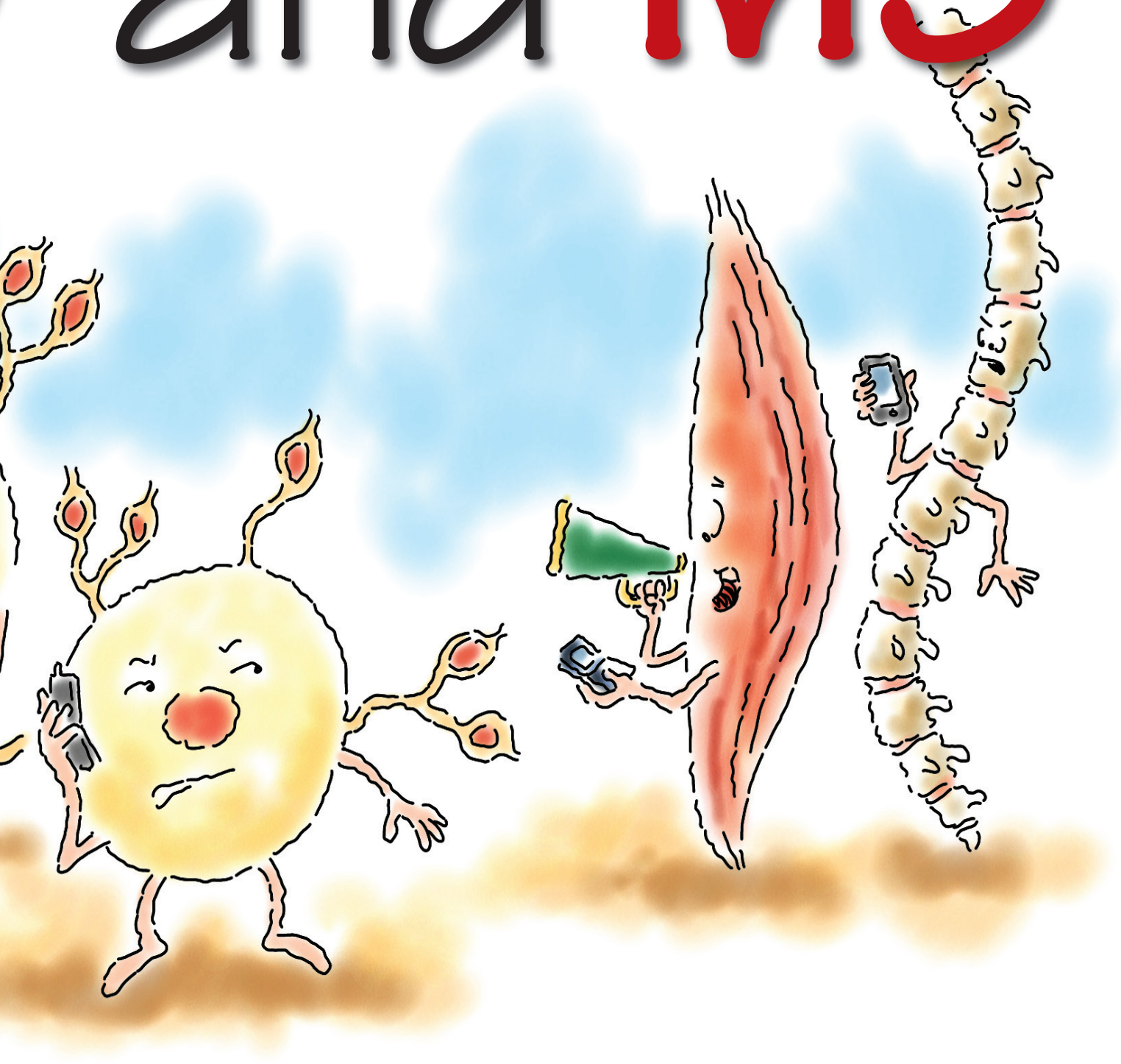
The author has disclosed that she has no significant relationships with or financial interest in any commercial companies that pertain to this educational activity.



Jason, 50, was playing soccer with his son when he tripped several times on the field while passing the ball. Later, his legs were trembling. He thought at first that he was just getting older; however, several months later, he felt his arm twitch slightly and he developed left foot drop. This time, he knew that something was wrong. He went to his healthcare provider, who sent him to a neurologist. He was scheduled for magnetic resonance imaging (MRI) and an electromyogram (EMG). His neurologist said that there could be several possible causes for his symptoms, two of which were amyotrophic lateral sclerosis (ALS) and multiple sclerosis (MS). What's the difference between these two degenerative diseases?

In this article, I'll discuss the characteristics, causes, signs and symptoms, diagnosis, and nursing interventions for both diseases. First, let's review the characteristics of ALS.

and MS



What's ALS?

ALS, better known as Lou Gehrig disease, is a progressive neuromuscular disorder that involves the degeneration of both the upper and lower motor neurons, leading to eventual wasting and atrophy of all voluntary muscles, including the respiratory muscles (see *What happens in ALS*). The term amyotrophic means without muscle nutrition or progressive muscle wasting. This accounts for the lower motor neuron component of the syndrome. Lateral sclerosis is the scarring of the corticospinal tract in the lateral column of the spinal cord and refers to the upper motor neuron part of the syndrome. ALS is a fatal disease, with an average life expectancy after diagnosis of 2 to 5 years. Most patients die of respiratory failure.

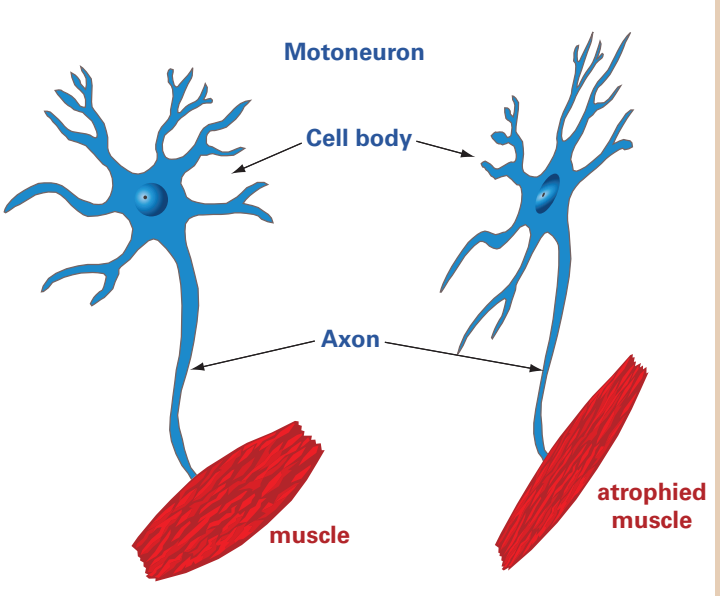
ALS is a diffuse disease, but its onset is focal and asymmetric. There are three different subtypes:

- ***progressive muscular atrophy***—only lower motor neurons are involved
- ***primary lateral sclerosis***—only upper motor neurons are involved
- ***progressive bulbar palsy***—restricted to bulbar muscles (rare). (Most patients who present with initial involvement of bulbar muscles evolve to classic ALS, involving both upper and lower motor neurons.)

Approximately 5,600 people in the United States are diagnosed with ALS each year. The incidence is 2 per 100,000 people. It's estimated that 30,000 Americans have the disease at any given time. The incidence of ALS is higher in men than in women until menopause, with a peak onset of symptoms between ages 40 and 60.

What happens in ALS

- Progressive weakness and wasting of muscles of the arms, trunk, and legs
- Fasciculations and signs of spasticity
- Progressive difficulty swallowing (drooling and regurgitation of liquids through nose), speaking (nasal and unintelligible) and, ultimately, breathing
- Cranial nerve deficits (bulbar symptoms) are present in 20% of cases (prevalence increases with age), along with dysarthria, voice deterioration, and dysphagia



Cause and effect

There's no single cause of ALS. Ten percent of diagnosed patients have a familial form of ALS. The genetic defect is thought to be on chromosome 21 and is involved with the enzyme glutamate. Coded to help destroy free radicals, this enzyme is altered. Glutamate toxicity is now believed to be one of the possible causes or contributors to major neuronal degeneration.

Signs and symptoms

ALS involves a slow, chronic progression of symptoms. The onset of symptoms varies with each patient. One of the first symptoms is usually unexplained weakness in a limb. The patient may notice wrist drop interfering with his work or activities of daily living. He may complain of reduced finger dexterity, cramping, or stiffness. He may develop foot drop that results in a fall or sprain. It's the combination of a weak,

atrophic fasciculating (twitching) muscle with the increased tone and hyperreflexia of the affected limb that are characteristic of ALS.

The upper motor neuron (corticospinal tract) signs are spasticity and hyperactive tendon reflexes, including a positive Babinski reflex (see *Testing Babinski reflex*). The lower motor neuron signs are atrophy, weakness, depressed reflexes, and fasciculations. The cardiac and smooth muscles aren't involved, and there's no loss of anal sphincter tone. Muscle atrophy and weight loss are usually present by the time a patient seeks treatment. Exaggeration of motor expressions and emotional lability may occur when the disease progresses into the corticobulbar projections of the brainstem.

Bulbar symptoms include difficulty swallowing (dysphagia) and/or difficulty speaking (dysarthria). Ocular, sensory, or autonomic dysfunction occurs late in the disease. Nearly all patients develop symptoms of respiratory insufficiency.

Making the diagnosis

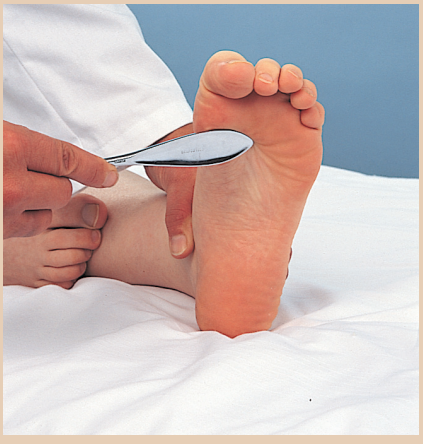
The diagnosis of ALS should be made as early as possible by an experienced neurologist. Recommended mandatory lab tests include:

- erythrocyte sedimentation rate
- C-reactive protein
- hematologic screen
- creatine phosphokinase and lactate dehydrogenase
- thyroid tests
- vitamin B_{12} and folate
- serum protein electrophoresis and immunoelectrophoresis
- electrolytes, including glucose, creatinine, calcium, and phosphorus.

MRI, computed axial tomography scanning, and chest X-rays are used to rule out other conditions, such as MS, brainstem stroke, or tumor. EMG can be used to show fibrillation and fasciculations of the motor units. Muscle biopsy can be used to verify lower motor neuron degeneration and denervation. Nerve conduction studies can be used to assess the amplitude and velocity of nerves.

Testing Babinski reflex

A well-known reflex indicative of CNS disease affecting the corticospinal tract is the Babinski reflex. In a person with an intact CNS, if the lateral aspect of the sole of the foot is stroked, the toes contract and draw together. In a person who has CNS disease of the motor system, the toes fan out and draw back. This is normal in newborns but represents a serious abnormality in adults.



Symptomatic and supportive management

Patients are cared for symptomatically, and supportive management is directed at preventing complications of immobility. Protect the patient from skin breakdown, watch for the development of deep vein thrombosis, and provide pain management. Because ALS is a progressive disease, the patient may be able to transfer from the bed to the chair one day and need medical equipment to help him move the next day.

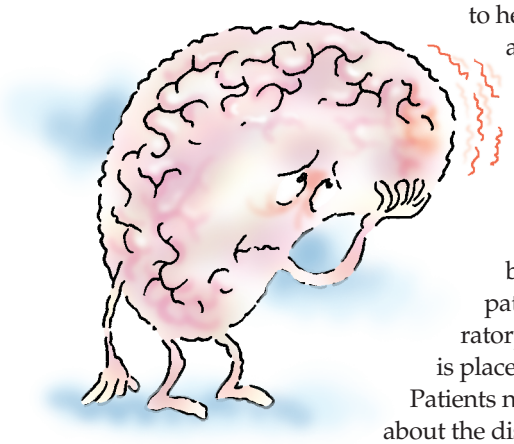
Inpatient and outpatient medications are patient specific and based on symptoms. Clinical guidelines recommend that riluzole 500 mg twice a day be given to block glutamatergic or excitatory neurotransmitters in the central nervous system (CNS). Treatment should be initiated as early as possible after the patient has been diagnosed. Baclofen and tizanidine are given to treat muscle spasms, and cholinergic agents such as pyridostigmine can be used to block acetylcholine release to facilitate impulse transmission across the myoneural junction.

Patients tire easily and complain of spasms and a tight or pulling feeling.

Both ALS and MS are neurodegenerative diseases.

Cramps can be treated with physiotherapy, physical exercise, or hydrotherapy. Drooling and thick mucus can create choking and interfere with eating. Portable mechanical home suction devices can be used to reduce this problem. Also, experimental drug treatments with *Botulinum* toxin injections into the parotid glands are being tried to reduce sialorrhea (excessive saliva).

Give small, frequent high-calorie meals and keep the head of the bed elevated 30 minutes after meals to help prevent possible aspiration. As the disease progresses, nutritional support is provided with the insertion of a gastrostomy tube. This should be done before the patient develops respiratory insufficiency and is placed on a ventilator. Patients need to be educated about the disease and make the decision about choosing or not choosing mechanical ventilation. The primary cause of death for those on ventilatory support is pulmonary infection.



Bronchial secretions can be reduced with the use of mucolytic-like medications, nebulization with saline solution, or an anticholinergic bronchodilator. A mechanical insufflator-exsufflator and nocturnal oximetry are available for home use to help patients with nighttime hypoventilation. Noninvasive positive-pressure ventilation can help improve the quality of life for patients with ALS and allow them to sleep better. The clinical guidelines for ALS recommend that oxygen therapy alone should be avoided because it can exacerbate carbon dioxide retention and mouth dryness.

Patients may also suffer from emotional expression disorder, which causes difficulty controlling emotions such as crying excessively or laughing inappropriately. Guidelines recommend that only troublesome emotional lability should be treated. The life expectancy of a patient with ALS is typically 2 to 5 years after the onset of symptoms, but may be as short as a few months. For this reason, it's essential to provide emotional care for both the patient and his family. A spiritual counselor, mental health worker, or social worker can help the patient and his family with planning end-of-life care.

What's MS?

MS is an immune-mediated progressive neurodegenerative disease of the CNS that typically manifests itself with periods of disease activity followed by intervals of remission. This disease has an inflammatory component, which is associated with a potent cellular and humoral immune response against potential CNS antigens, and a neurodegenerative component, which promotes neuronal loss and causes brain atrophy.

MS is characterized by infiltrates of lymphocytes and macrophages in the parenchyma of the brain. T lymphocytes cross the blood-brain barrier, triggering inflammation and scarring of myelin—the fatty and protein material that surrounds certain nerve fibers in the brain and spinal cord (see *What happens in MS*). Demyelination, or destruction, of these nerve fibers in the white matter of the brain and spinal cord interrupt the conduction of nerve impulses.

There are four different major presentations of the disease:

- ***relapsing-remitting***—patients experience a series of attacks, followed by complete or partial disappearance of symptoms (remission) until another attack occurs (relapse); it may be weeks to decades between relapses (About 65% to 80% of patients begin with this type of MS.)
- ***primary progressive***—a continuous, gradual decline in the patient's physical abilities from the outset rather than relapses. (About 10% to 20% of patients begin with this type of MS.)

• ***secondary progressive***—patients beginning with the relapsing-remitting type of MS enter a phase where relapses are rare and more disability accumulates (About 50% of patients with relapsing-remitting MS will develop this type within 10 years.)
• ***progressive relapsing***—characterized by a steady decline in abilities accompanied by sporadic attacks.

There are cases of MS that are mild and can be recognized only retrospectively after many years. There are also rare cases of an extremely rapid progression of MS symptoms (sometimes fatal) known as malignant or fulminant (Marburg variant) MS.

The prevalence of the disease is 30 to 80 per 100,000 persons. The onset of MS is usually between ages 20 and 50 years old. It's more common in women and those of Northern European descent and is more prevalent in colder latitudes (above the 37th parallel). MS remains a significant cause of neurologic disability in young adults.

Cause and effect

Genetic and environmental factors play important roles in the development of MS. It has been hypothesized that an environmental agent or event, such as viruses, bacteria, chemicals, or lack of sun exposure, acts in concert with a specific genetic predisposition, resulting in immune dysfunction.

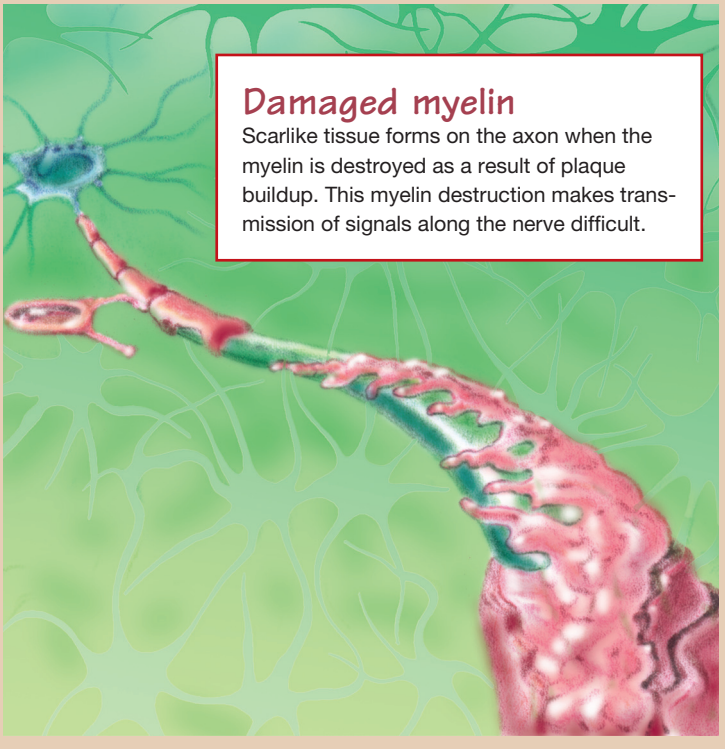
Signs and symptoms

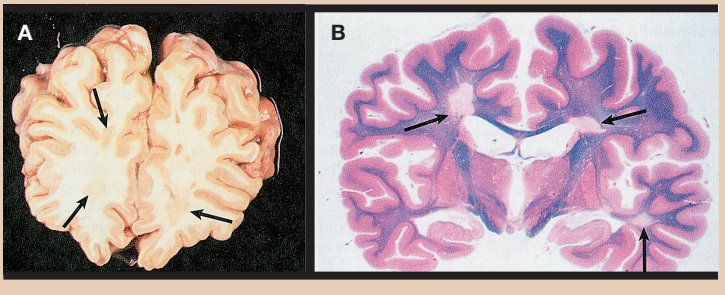
The signs and symptoms of MS may include:
• paresthesia
• ataxia
• vertigo
• weakness and fatigue
• spasticity
• loss of vision or double vision.

Other symptoms include speech and swallowing impairment, bladder and sexual dysfunction, depression, apathy, and loss of memory. Lhermitte sign, a tingling sensation that travels down the trunk or limbs of a person during active or passive neck flexion, is

What happens in MS

• Fatigue and weakness
• Abnormal reflexes (absent or exaggerated)
• Visual disturbances (double vision and nystagmus [involuntary eye movement])
• Motor dysfunction, such as weakness, tremor, and uncoordinated movements
• Sensory disturbances, such as paresthesia, impaired deep sensation, and impaired vibratory and position sense
• Impaired speech (dysarthria)
• Urinary dysfunction, such as hesitancy, frequency, urgency, and retention
• Neurobehavioral syndromes, such as depression, cognitive impairment, and emotional lability





A: In this unfixed brain, the plaques of MS in the white matter (arrows) assume the darker color of the cerebral cortex.
B: A coronal section of the brain from a patient with long-standing MS, which has been stained for myelin, shows discrete areas of demyelination (arrows) with characteristic involvement of the superior angles of the lateral ventricles.

The McDonald criteria for diagnosing MS

Clinical attacks	Objective lesions	Additional data needed
2 or more	2 or more	None. Clinical evidence alone will suffice; additional evidence desirable but must be consistent with MS.
2 or more	1	Dissemination in space by MRI ***or*** 2 or more MRI lesions consistent with MS plus positive CSF ***or*** await further clinical attack implicating other site.
1	2 or more	Dissemination in time by MRI or second clinical attack.
1	1	Dissemination in space by MRI ***or*** 2 or more MRI lesions consistent with MS plus positive CSF ***AND*** dissemination in time by MRI or second clinical attack.
0 (progression from onset)	1 or more	Disease progression for 1 year (retrospective or prospective) ***AND*** 2 out of 3 of the following: • positive brain MRI (9 T2 lesions or 4 or more T2 lesions with positive visual evoked potential test) • positive spinal cord MRI (2 or more focal T2 lesions) • positive CSF.

Source: National Multiple Sclerosis Society. 2005 revised McDonald MS diagnostic criteria. http://www.nationalmssociety.org/download.aspx?id=215.

considered a classic finding in MS. It's due to nonsynaptic impulses traveling in demyelinated axons of the dorsal column in the spinal cord.

Infection, trauma, or pregnancy can precipitate the onset or exacerbation of symptoms. The initial syndrome depends on the portion of the CNS that's involved. Short-lived attacks may be due to minor increases in temperature or serum calcium levels, emotional stress, or when functional demands exceed conduction capacity. Attacks may have motor or sensory symptoms and can begin quickly, lasting only a few seconds or minutes.

Making the diagnosis

When a patient presents with a first episode of neurologic signs or symptoms suggestive of demyelination, a diagnosis of MS is considered. When he presents with a second or subsequent set of neurologic symptoms, he should be referred to an expert for diagnosis according to the McDonald criteria (see *The McDonald criteria for diagnosing MS*). A single neurologic event, such as optic neuritis (reduction or loss of vision in one eye) with MRI lesions compatible with MS, is considered a clinically isolated syndrome. The diagnosis of MS can't be made unless there's another clinical or MRI event.

Other tests used to exclude other possible diagnoses are elevated immunoglobulin G (IgG) found in the cerebrospinal fluid (CSF) and IgG bands found on electrophoresis. Evoked response studies can detect decreased conduction velocity in the visual, auditory, and somatosensory pathways.

Symptomatic and supportive management

When caring for a patient with MS, assess and monitor the patient's level of functioning, existing symptoms, and development

of new symptoms. Encourage verbalization and problem solving. For patients experiencing double vision or blurred vision, an eye patch may be used. Instruct patients to rest their eyes when they're fatigued and to use large-type books when reading. For sensory symptoms such as decreased sensation of temperature, touch, and position sense, patients can use pot holders to protect their hands when cooking and wear gloves in cold weather. Impaired mobility can be helped with the use of assistive devices. Recommendations for home environment safety, such as no scatter rugs and good lighting, are useful to help prevent falls.

Physical therapy can provide gait training, and occupational therapy can help with equipment needs. Adaptive equipment, such as nonspill cups or stabilized plates, can be used to help a patient with intentional tremors of the upper extremities. Be aware of complications of immobility, such as pressure ulcers and genitourinary infections.

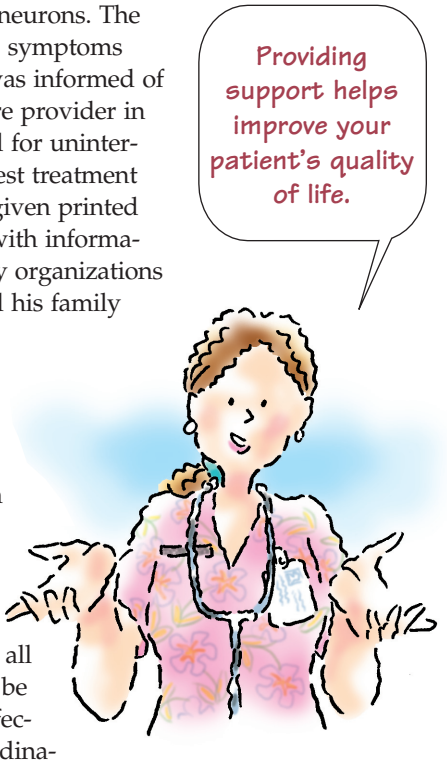
Prune juice and stool softeners can be prescribed to reduce constipation. Increased fluid intake and cranberry juice can help reduce urinary tract infections. If the patient is receiving tube feedings, skin care around the gastrostomy tube is essential. Patients should be kept in high Fowler's position after eating to prevent aspiration.

Medications used to treat MS include corticosteroids to reduce inflammation; antispasmotics, such as baclofen, tizanidine, and clonazepam; and the immune-modulating drugs interferon beta-1a and interferon beta-1b.

Feelings of helplessness are common among patients with MS. It's important that patients receive information about the disease, treatment, and drug therapy. They should have the contact information of their local MS chapter and visiting nurse services.

A difficult diagnosis

Let's check in with our patient. Jason was scheduled for a muscle biopsy to assess whether his muscle damage was present in both the upper and lower neurons. The test results found that Jason's symptoms were caused by ALS. Jason was informed of his diagnosis by his healthcare provider in a private session that allowed for uninterrupted time to review the latest treatment and care options. Jason was given printed materials about the disease, with information on support and advocacy organizations and websites to help him and his family cope with the diagnosis.



Improving quality of life

Providing accurate information can help the patient with ALS or MS feel cared for and supported. Patients and families should be able to take an active part in making informed decisions about all aspects of care. Goals should be attainable and reasonable. Effective communication and coordination between nurses and other members of the healthcare team can help improve the quality of life for patients with ALS and MS. ■

Learn more about it

Andersen PM, Borasio GD, Dengler R, et al. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. an evidence-based review with good practice points. http://www.guidelines.gov/summary/summary.aspx?doc_id=9652&nbr=005168&string=amyotrophic+AND+lateral+AND+sclerosis.

Borazanci AP, Harris MK, Schwendimann RN, et al. Multiple sclerosis: clinical features, pathophysiology, neuroimaging, and future therapies. *Future Neurol.* 2009;4(2):229-246.

Costello, KM. The advanced role of nonphysician practitioners and nurses in managing multiple sclerosis. http://cme.medscape.com/viewarticle/705946.

Clem K, Morgenlander J. Amyotrophic lateral sclerosis. http://emedicine.medscape.com/article/791154.

Dangond F. Multiple sclerosis. http://emedicine.medscape.com/article/1146199.

Houseman G. Symptom management of the patient with amyotrophic lateral sclerosis: a guide for hospice nurses. *J Hospice Palliative Nurs.* 2008;10(4):214-215.

National Collaborating Centre for Chronic Conditions. Multiple sclerosis. National clinical guidelines for diagnosis and management in primary and secondary

care. http://www.guidelines.gov/summary/summary.aspx?doc_id=5063&nbr=003547&string=multiple+AND+sclerosis.

Nettina SM. *Lippincott Manual of Nursing Practice*. 9th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2010: 531,534.

Pathophysiology Made Incredibly Visual! Philadelphia, PA: Lippincott Williams & Wilkins; 2008: 84-85.

Porth CM, Matfin G. *Pathophysiology: Concepts of Altered Health States*. 8th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2009:1284-1287.

Smeltzer SC, Bare BG, Hinkle JL, Cheever KH. *Brunner & Suddarth's Textbook of Medical-Surgical Nursing*. 11th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008:2149,2277-2285,2320-2323.

On the web

These online resources may be helpful to your patients and their families.

ALS

ALS Association: http://www.alsa.org
Mayo Clinic: http://www.mayoclinic.com/health/amyotrophic-lateral-sclerosis/DS00359
MDA: http://www.als-mda.org
National Institute of Neurological Disorders and Stroke: http://www.ninds.nih.gov/disorders/amyotrophiclateralsclerosis/detail_amyotrophiclateralsclerosis.htm
World Federation of Neurology Research Group on Motor Neuron Diseases-ALS: http://www.wfnals.org

MS

Mayo Clinic: http://www.mayoclinic.com/health/multiple-sclerosis/DS00188
Multiple Sclerosis Society of America: http://www.msassociation.org
National Institute of Neurological Disorders and Stroke: http://www.ninds.nih.gov/disorders/multiple_sclerosis/multiple_sclerosis.htm
National Multiple Sclerosis Society: http://www.nationalmssociety.org/index.aspx

For more than 36 additional continuing education articles related to neurologic topics, go to Nursingcenter.com/CE.

CE CONNECTION

Earn CE credit online:
Go to http://www.nursingcenter.com/CE/nmie and receive a certificate *within minutes*.

INSTRUCTIONS

Degenerative disorders: ALS and MS

TEST INSTRUCTIONS

- To take the test online, go to our secure Web site at **http://www.nursingcenter.com/CE/nmie**.
- On the print form, record your answers in the test answer section of the CE enrollment form on page 50. Each question has only one correct answer. You may make copies of these forms.
- Complete the registration information and course evaluation. Mail the completed form and registration fee of $21.95 to: Lippincott Williams & Wilkins, CE Group, 2710 Yorktowne Blvd., Brick, NJ 08723. We will mail your certificate in 4 to 6 weeks. For faster service, include a fax number and we will fax your certificate within 2 business days of receiving your enrollment form.
- You will receive your CE certificate of earned contact hours and an answer key to review your results.There is no minimum passing grade.
- Registration deadline is June 30, 2012.

DISCOUNTS and CUSTOMER SERVICE

- Send two or more tests in any nursing journal published by Lippincott Williams & Wilkins together by mail and deduct $0.95 from the price of each test.
- We also offer CE accounts for hospitals and other health care facilities on nursingcenter.com. Call 1-800-787-8985 for details.

PROVIDER ACCREDITATION

Lippincott Williams & Wilkins, publisher of *Nursing made Incredibly Easy!*, will award 2.0 contact hours for this continuing nursing education activity.

Lippincott Williams & Wilkins is accredited as a provider of continuing nursing education by the American Nurses Credentialing Center's Commission on Accreditation.

This activity is also provider approved by the California Board of Registered Nursing, Provider Number CEP 11749 for 2.0 contact hours. Lippincott Williams & Wilkins is also an approved provider of continuing nursing education by the District of Columbia and Florida #FBN2454.

Your certificate is valid in all states.

The ANCC's accreditation status of Lippincott Williams & Wilkins Department of Continuing Education refers only to its continuing nursing educational activities and does not imply Commission on Accreditation approval or endorsement of any commercial product.