Management of Patients With Neurologic Infections, Autoimmune Disorders, and Neuropathies

LEARNING OBJECTIVES

On completion of this chapter, the learner will be able to:

1. Differentiate among the infectious disorders of the nervous system according to causes, manifestations, medical care, and nursing management.

2. Describe the pathophysiology, clinical manifestations, and medical and nursing management of multiple sclerosis, myasthenia gravis, and Guillain-Barré syndrome.

3. Use the nursing process as a framework for care of patients with multiple sclerosis, myasthenia gravis, and Guillain-Barré syndrome.

4. Describe disorders of the cranial nerves, their manifestations, and indicated nursing interventions.

5. Develop a plan of nursing care for the patient with a cranial nerve disorder.
The diverse group of neurologic disorders that make up infectious and autoimmune disorders, and cranial and peripheral neuropathies present unique challenges for nursing care. Infectious processes of the nervous system sometimes cause death or permanent dysfunction. Autoimmune disorders usually have a slow, progressive course, requiring the nurse to manage symptoms and facilitate patients’ and families’ understanding of the disease process. Cranial and peripheral nerve disorders may affect the patient’s comfort, functional independence, and self-esteem.

The nurse who cares for patients with these disorders must have a clear understanding of the pathologic processes and the clinical outcomes. Some of the issues nurses must help patients and families confront include adaptation to the effects of the disease, potential changes in family dynamics, and, possibly, end-of-life issues.

**Infectious Neurologic Disorders**

The infectious disorders of the nervous system include meningitis, brain abscesses, various types of encephalitis, and Creutzfeldt-Jakob and new-variant Creutzfeldt-Jakob disease. The clinical manifestations, assessment, and diagnostic findings as well as the medical and nursing management are related to the specific infectious process.

**MENINGITIS**

Meningitis is an inflammation of the meninges, the protective membranes that surround the brain and spinal cord. Meningitis is classified as aseptic or septic. In aseptic meningitis, bacteria are not the cause of the inflammation; the cause is viral or secondary to lymphoma, leukemia, or brain abscess. Septic meningitis refers to meningitis caused by bacteria, most commonly *Neisseria meningitidis*, although *Haemophilus influenzae* and *Streptococcus pneumoniae* are also causative agents.

Outbreaks of *N. meningitidis* infection are most likely to occur in dense community groups, such as college campuses and military installations. Though infections occur year round, the peak incidence is in the winter and early spring. Factors that increase the risk for developing bacterial meningitis include tobacco use and viral upper respiratory infection because they increase the amount of droplet production. Otitis media and mastoiditis increase the risk of bacterial meningitis because the bacteria can cross the epithelium membrane and enter the subarachnoid space. Persons with immune system deficiencies are also at greater risk for developing bacterial meningitis. Between 1992 and 1996 there was a 28% increase in the number of new cases reported in the 12- to 29-year-old age group (Rosenstein, Perkins, Stephens et al., 2001). This increase focused attention on the need to develop a vaccine for high-risk populations.

**Pathophysiology**

Meningeal infections generally originate in one of two ways: through the bloodstream as a consequence of other infections, or by direct extension, such as might occur after a traumatic injury to the facial bones, or secondary to invasive procedures.

*N. meningitidis* concentrates in the nasopharynx and is transmitted by secretion or aerosol contamination. Bacterial or meningo-coccal meningitis also occurs as an opportunistic infection in patients with acquired immunodeficiency syndrome (AIDS) and as a complication of Lyme disease (Chart 64-1). *S. pneumoniae* is the most frequent causative agent of bacterial meningitis associated with AIDS (Rosenstein, Perkins, Stephens et al., 2001).

Once the causative organism enters the bloodstream, it crosses the blood–brain barrier and causes an inflammatory reaction in the meninges. Independent of the causative agent, inflammation of the subarachnoid space and pia mater occurs. Since there is little room for expansion within the cranial vault, the inflammation may cause increased intracranial pressure. Cerebrospinal fluid (CSF) flows in the subarachnoid space, where inflammatory cellular material from the affected meningeal tissue enters and accumulates in the subarachnoid space, thereby increasing the CSF cell count (Coyle, 1999).

The prognosis for bacterial meningitis depends on the causative organism, the severity of the infection and illness, and the timeliness of treatment. In acute fulminant presentations there may be adrenal damage, circulatory collapse, and widespread hemorrhages (Waterhouse-Friderichsen syndrome). This syndrome is the result of endothelial damage and vascular necrosis caused by the bacteria. Complications include visual impairment, deafness, seizures, paralysis, hydrocephalus, and septic shock.

**Clinical Manifestations**

Headache and fever are frequently the initial symptoms. Fever tends to remain high throughout the course of the illness. The headache is usually severe as a result of meningeal irritation. Meningeal irritation results in a number of other well-recognized signs common to all types of meningitis:

- **Nuchal rigidity (stiff neck)**: an early sign. Any attempts at flexion of the head are difficult because of spasms in the muscles of the neck. Forceful flexion causes severe pain.
- **Positive Kernig’s sign**: When the patient is lying with the thigh flexed on the abdomen, the leg cannot be completely extended (Fig. 64-1).

**Glossary**

- **ataxia**: impaired coordination of movements
- **bulbar paralysis**: immobility of muscles innervated by cranial nerves with their cell bodies in the lower portion of the brain stem
- **diplopia**: double vision, or the awareness of two images of the same object occurring in one or both eyes
- **dyskinesia**: impaired ability to execute voluntary movements
- **dysphagia**: difficulty swallowing, causing the patient to be at risk for aspiration
- **dysphonia**: voice impairment or altered voice production
- **myoclonus**: spasms of a single muscle or group of muscles
- **neuropathy**: general term indicating a disorder of the nervous system
- **paresthesia**: a sensation of numbness or tingling or a “pins and needles” sensation
- **prion**: a particle smaller than a virus that is resistant to standard sterilization procedures
- **spasticity**: muscular hypertonicity with increased resistance to stretch often associated with weakness, increased deep tendon reflexes, and diminished superficial reflexes
- **spongiform**: having the appearance or quality of a sponge
Meningitis can occur as a complication of other diseases and is an opportunistic infection seen with greater frequency in patients with acquired immunodeficiency syndrome (AIDS).

### Meningitis in AIDS Patients
- Aseptic, cryptococcal, and tuberculous forms of meningitis have been reported in patients with AIDS.
- Acute and chronic forms of aseptic meningitis may occur with AIDS; both are accompanied by headache, but signs of meningeal irritation generally occur with the acute form.
- Aseptic meningitis may be accompanied by cranial nerve palsies. The meningitis is thought to be related to direct infection of the central nervous system by human immunodeficiency virus (HIV) because it can be isolated from the cerebrospinal fluid (CSF).
- Cryptococcal meningitis is the most common fungal infection of the central nervous system in patients with AIDS and has a 50% to 60% relapse rate. Patients may experience headache, nausea, vomiting, seizures, confusion, and lethargy. Treatment consists of IV administration of amphotericin B followed by fluconazole. Maintenance therapy with fluconazole may be necessary to prevent relapse.
- Some immunosuppressed patients develop few if any symptoms because of blunted inflammatory responses; others develop atypical features.

### Meningitis in Lyme Disease
- Lyme disease is a multisystem inflammatory process caused by the tick-transmitted spirochete Borrelia burgdorferi.
- Neurologic abnormalities are seen in later stages (stages 2 or 3). Stage 2 occurs either with the characteristic rash or from 1 to 6 months after it has disappeared.
- Neurologic abnormalities include aseptic meningitis, chronic lymphocytic meningitis, and encephalitis.
- Cranial nerve inflammation, including Bell’s palsy and other peripheral neuropathies, is common.
- Stage 3 (the chronic form of the disease) begins years after the initial tick infection and is characterized by arthritis, skin lesions, and neurologic abnormalities.
- Most patients with stage 2 and 3 Lyme disease are treated with intravenous antibiotics, usually ceftriaxone or penicillin G.
- Meningeal and systemic symptoms begin to improve within days, although other symptoms, such as headache, may persist for weeks.

A rash can be a striking feature of *N. meningitidis* infection, occurring in about half of patients with this type of meningitis. Skin lesions develop, ranging from a petechial rash with purpuric lesions to large areas of ecchymosis.

Disorientation and memory impairment are common early in the course of the illness. The changes depend on the severity of the infection as well as the individual response to the physiologic processes. Behavioral manifestations are also common. As the illness progresses, lethargy, unresponsiveness, and coma may develop.

Seizures and increased intracranial pressure (ICP) are also associated with meningitis. Seizures occur secondary to focal areas of cortical irritability. Intracranial pressure increases secondary to accumulation of purulent exudate. The initial signs of increased ICP include decreased level of consciousness and focal motor deficits. If ICP is not controlled, the uncus of the temporal lobe may herniate through the tentorium into the brain stem. Brain stem herniation is a life-threatening event causing cranial nerve dysfunction and depressing the centers of vital functions, such as the medulla (Rowland, 2000). (See Chap. 61 for discussion of the patient with a change in level of consciousness or increased ICP.)

A fulminating infection occurs in about 10% of patients with meningococcal meningitis, with signs of overwhelming septicemia: an abrupt onset of high fever, extensive purpuric lesions (over the face and extremities), shock, and signs of disseminated intravascular coagulopathy (DIC). Death may occur within a few hours of onset of the infection.

### Assessment and Diagnostic Findings

When the clinical presentation points to meningitis, diagnostic testing to identify the causative organism is conducted. Bacterial culture and Gram staining of CSF and blood are key diagnostic tests (Fischbach, 2002). The presence of polysaccharide antigen in CSF further supports the diagnosis of bacterial meningitis (Rosenstein et al., 2001).

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**Figure 64-1** Testing for meningeal irritation. (A) Kernig’s sign. (B) Brudzinski’s sign.

- Positive Brudzinski’s sign: When the patient’s neck is flexed, flexion of the knees and hips is produced; when passive flexion of the lower extremity of one side is made, a similar movement is seen in the opposite extremity (see Fig. 64-1).
- Photophobia: extreme sensitivity to light; this finding is common, although the cause is unclear.

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Prevention

In 1971, the military began vaccinating all new recruits against meningococcal meningitis, resulting in a dramatic decrease in the incidence. Researchers suggested vaccination of college freshmen as surveillance studies indicated that freshmen living in dormitories were at highest risk for developing meningococcal meningitis. At this time vaccination is not required for college freshmen; however, the American Academy of Pediatrics provides information to college freshmen and their parents about the risk of disease and the availability of vaccination (Bruce et al., 2001; Centers for Disease Control and Prevention [CDC], 2000).

In close contact with patients with meningococcal meningitis should be treated with antimicrobial chemoprophylaxis using rifampin (Rifadin), ciprofloxacin hydrochloride (Cipro), or ceftriaxone sodium (Rocephin) (CDC, 2000). Therapy should be started as soon as possible after contact; a delay in the initiation of therapy will limit the effectiveness of the prophylaxis (Rosenstein et al., 2001). Vaccination should also be considered as an adjunct to antibiotic chemoprophylaxis for anyone living with a person who develops meningococcal infection. Vaccination for children and at-risk adults should be encouraged to avoid meningitis caused by *H. influenzae* and *S. pneumoniae*.

Medical Management

Successful outcomes depend on the early administration of an antibiotic that crosses the blood–brain barrier into the subarachnoid space in sufficient concentration to halt the multiplication of bacteria. Penicillin antibiotics (eg, ampicillin, piperacillin) or one of the cephalosporins (eg, ceftriaxone sodium, cefotaxime sodium) may be used. Vancomycin hydrochloride alone or in combination with rifampin may be used if resistant strains of bacteria are identified. High doses of the appropriate antibiotic are administered intravenously.

Dexamethasone has been shown to be beneficial as adjunct therapy in the treatment of acute bacterial meningitis and in pneumococcal meningitis if given 15 to 20 minutes before the first dose of antibiotic and every 6 hours for the next 4 days. Studies indicate that dexamethasone improves the outcome in adults and does not increase the risk of gastrointestinal bleeding (de Gans & van de Beek, 2002).

Dehydration and shock are treated with fluid volume expanders. Seizures, which may occur in the early course of the disease, are controlled with phenytoin (Dilantin). Increased ICP is treated as necessary (see Chap. 61).

Nursing Management

The patient may be critically ill; therefore, so many of the nursing interventions are collaborative with those of the physician, respiratory therapist, and other members of the health care team. The patient’s prognosis may depend on the supportive care provided.

Neurologic status and vital signs are continually assessed. Pulse oximetry and arterial blood gas values are used to quickly identify the need for respiratory support as the increasing ICP compromises the brain stem. Insertion of a cuffed endotracheal tube (or tracheotomy) and mechanical ventilation may be necessary to maintain adequate tissue oxygenation.

Arterial blood pressures are monitored to assess for incipient shock, which precedes cardiac or respiratory failure. Rapid intravenous (IV) fluid replacement may be prescribed, but care is taken to prevent fluid overload. Fever also will increase the workload of the heart and cerebral metabolism. ICP will increase in response to increased cerebral metabolic demands. Therefore, measures are taken to reduce body temperature as quickly as possible.

Other important components of nursing care include:

- Monitoring body weight, serum electrolytes, and urine volume, specific gravity, and osmolality, especially if the syndrome of inappropriate antidiuretic hormone (SIADH) secretion is suspected
- Protecting the patient from injury secondary to seizure activity or altered level of consciousness
- Preventing complications associated with immobility, such as pressure ulcers and pneumonia
- Instituting droplet precautions until 24 hours after the initiation of antibiotic therapy (oral and nasal discharge is considered infectious)

Any sudden, critical illness can be devastating to the family. Because the patient’s condition is often critical and the prognosis guarded, the family needs to be informed about the patient’s condition and permitted to see the patient at intervals, even though the priority is to address the patient’s need for immediate and intensive treatment. An important aspect of the nurse’s role is to support the patient and to assist the family in identifying others who can be supportive to them during the crisis.

BRAIN ABSCESS

Although brain abscess is relatively rare, it is a complication encountered increasingly in patients whose immune systems have been suppressed either through therapy or disease.

Pathophysiology

A brain abscess is a collection of infectious material within the tissue of the brain. It may occur by direct invasion of the brain from intracranial trauma or surgery; by spread of infection from nearby sites, such as the sinuses, ears, and teeth (paranasal sinus infections, otitis media, dental sepsis); or by spread of infection from other organs (lung abscess, infective endocarditis) (Hickey, 2003). To prevent brain abscess, otitis media, mastoiditis, sinusitis, dental infections, and systemic infections should be treated promptly.

Clinical Manifestations

The clinical manifestations of a brain abscess result from alterations in intracranial dynamics (edema, brain shift), infection, or the location of the abscess (Chart 64–2). Headache, usually worse in the morning, is the most prevailing symptom. Vomiting is also common. Focal neurologic signs (weakness of an extremity, decreasing vision, seizures) may occur, depending on the site of the abscess. There may be a change in mental status, as reflected in lethargic, confused, irritable, or disoriented behavior. Fever may or may not be present.

Assessment and Diagnostic Findings

Repeatepd neurologic examinations and continuing assessment of the patient are necessary to determine the location of the abscess. A computed tomography (CT) scan is invaluable in locating the site of the abscess, after the evolution and resolution of supplicative lesions, and in determining the optimal time for surgical inter-
The patient with a brain abscess is extremely ill, and neurologic deficits may remain after treatment, such as hemiparesis, seizures, visual deficits, and cranial nerve palsies. Focal seizures are the most common sequelae, occurring in about 30% of patients (Hickey, 2003). The nurse must assess the family’s ability to express their distress at the patient’s condition, cope with the patient’s illness and deficits, and obtain support.

HERPES SIMPLEX VIRUS ENCEPHALITIS

Encephalitis is an acute inflammatory process of the brain tissue. Herpes simplex virus (HSV) is the most common cause of acute encephalitis in the United States (Levitz, 1998). There are two herpes simplex viruses, HSV-1 and HSV-2. HSV-1 typically affects children and adults.

Pathophysiology

There are two possible modes of HSV-1 infection. In most cases, primary HSV-1 infection of the buccal mucosa occurs, followed by retrograde spread along the trigeminal nerve to the brain. It is also believed that latent virus in brain tissue may reactivate and result in encephalitis (Roos, 1999). HSV-2 most commonly affects neonates and is discussed in pediatric textbooks (Gutierrez & Prober, 1998).

Clinical Manifestations

HSV-1 encephalitis causes inflammation and necrosis in the temporal lobe, frontal lobe, and limbic system. The initial symptoms include fever, headache, confusion, and behavioral abnormalities (Roos, 1999). Focal neurologic symptoms reflect the areas of cerebral inflammation and necrosis and include behavioral change, focal seizures, dysphasia, hemiparesis, and altered level of consciousness. Focal symptoms are present within 7 days of infection and progress for 14 to 21 days.

Assessment and Diagnostic Findings

Neuroimaging studies, electroencephalography (EEG), and CSF examination are used to diagnose HSV encephalitis. MRI is the neuroimaging study of choice in the diagnosis of HSV encephalitis as it can help identify lesions in the temporal lobe.

The EEG demonstrates a specific wave pattern in 66% of cases of biopsy-proven HSV encephalitis. CSF reveals a high opening pressure and low glucose and high protein levels. Viral cultures are almost always negative. Since 1996, the polymerase chain reaction (PCR) technique has been used to diagnose HSV encephalitis (Roos, 1999). PCR will identify the DNA bands of the HSV specifically. The validity of PCR is very high between the third and tenth day of symptom onset.

Medical Management

Ayclovir (Zovirax), an antiviral agent, is the medication of choice in HSV treatment (Karch, 2002). The mode of action is the inhibition of viral DNA replication. It is usually well tolerated by the patient. To prevent relapse, treatment should continue for up to 3 weeks. Slow administration over 1 hour will prevent crystallization of the medication in the urine. The usual dose of acyclovir is decreased if the patient has a history of renal insufficiency (Karch, 2002). In the rare case of acyclovir resistance, foscarnet sodium (Foscavir) is prescribed (Roos, 1999).
Nursing Management

Assessment of neurologic function is key to monitoring the progression of disease. Comfort measures to reduce headache include dimming the lights, limiting noise, and administering analgesic agents. Opioid analgesic medications may mask neurologic symptoms; therefore, they are used cautiously. Focal seizures and altered level of consciousness require care directed at injury prevention and safety. Nursing care addressing patient and family anxiety is ongoing throughout the illness. Monitoring of blood chemistry test results and urinary output will alert the nurse to the presence of renal complications related to acyclovir therapy.

ARTHROPOD-BORNE VIRUS ENCEPHALITIS

Arthropod vectors transmit several types of viruses that cause encephalitis. The primary vector in North America is the mosquito. Arbovirus infection occurs in specific geographic areas during the summer and fall. The four types of arboviral encephalitis that occur in North America are LaCrosse encephalitis, St. Louis encephalitis, Western equine encephalitis, and Eastern equine encephalitis (Roos, 1999).

Pathophysiology

Viral replication occurs at the site of the mosquito bite. If adequate virus is inoculated, a viremia ensues. The virus gains access to the central nervous system (CNS) via the cerebral capillaries. It spreads from neuron to neuron, predominantly affecting the cortical gray matter, the brain stem, and the thalamus. Meningeal exudates compound the clinical presentation by irritating the meninges and increasing ICP (Roos, 1999).

Clinical Manifestations

All arboviral encephalitis begins with a flu-like prodrome, but specific neurologic manifestations depend on the viral type. LaCrosse encephalitis, for example, may present with focal neurologic symptoms and seizures. Mortality is low but residual seizures may occur. A unique clinical feature of St. Louis encephalitis is SIADH with hyponatremia. The mortality rate is 10% to 20%. The clinical manifestations of Eastern equine encephalitis are acute and carry a high mortality rate of 50% to 75% (Roos, 1999). Although the clinical manifestations of Western equine encephalitis are nonspecific, the morbidity rate is high.

Assessment and Diagnostic Findings

Neuroimaging is not useful in diagnosing many types of encephalitis. In Eastern equine encephalitis, however, CT scan and MRI may reveal lesions in the basal ganglia and thalamus (Roos, 1999). The CSF analysis shows a normal glucose level, elevated protein level, and polymorphonuclear leukocytic pleocytosis. St. Louis, Eastern equine, and Western equine encephalitis viruses are rarely isolated in the CSF (Roos, 1999).

The age of the patient is important information in making a specific viral diagnosis. La Crosse virus encephalitis is the most common pediatric arboviral encephalitis. St. Louis encephalitis affects adults over 50 years of age; Eastern equine encephalitis is not age-specific (Roos, 1999). Western equine encephalitis can present as pediatric encephalitis but is less prevalent.

Medical Management

There is no specific medication for arboviral encephalitis. Medical management is aimed at controlling seizures and increased ICP (Roos, 1999).

Nursing Management

If the patient is very ill, hospitalization may be required. The nurse carefully assesses neurologic status and identifies improvement or deterioration in the patient’s condition. Injury prevention is key in light of the potential for falls or seizures. Arboviral encephalitis may result in death or life-long residual health issues. The family will need support and teaching to cope with these outcomes.

Public education addressing the prevention of arboviral encephalitis is a key nursing role. Clothing that provides coverage and insect repellents should be used in high-risk areas. Community mosquito control is advocated.

FUNGAL ENCEPHALITIS

Fungal infections of the CNS occur rarely in healthy people. The presentation of fungal encephalitis is related to geographic area and a compromised immune system (Leedom & Underman, 2000). The common fungi found around the world that can infect the CNS include Cryptococcus neoformans, Histoplasma capsulatum, Aspergillus, and Candida albicans (Davis, 1999). Other fungi are found only in certain regions: Coccidioides immitis, for example, is found in soil in central California, the southwest United States, northern Mexico, and areas of Argentina (Davis, 1999).

Pathophysiology

The fungal spores enter the body via inhalation. They initially infect the lungs, causing vague respiratory symptoms. In some cases, the fungi may enter the bloodstream, causing a fungemia. If the fungemia overcomes the person’s immune system, the fungus may spread to the CNS. The initial presentation is meningitis followed by encephalitis and brain abscesses. In addition to infecting the brain, the fungi may infect the spinal cord, producing an abscess. The abscess will produce symptoms of spinal cord compression (Davis, 1999).

Clinical Manifestations

The common symptoms of fungal encephalitis include fever, malaise, headache, nuchal rigidity, lethargy, and mental status changes (Davis, 1999). C. neoformans is the most common fungus to infect the CNS. Symptoms of increased ICP related to hydrocephalus often occur (Go et al., 2000). Vascular changes are associated with C. immitis and Aspergillus (Leedom & Underman, 2000). Manifestations of vascular change may include arteritis or cerebral infarction.

Assessment and Diagnostic Findings

CNS fungal infections present a diagnostic challenge because their presentations mimic other causes of encephalitis. The symptoms develop over a 2-week period. Fungal infection may also be present in organs such as the lungs or kidney (Davis, 1999). The presence of a compromised immune system and a history of living
in or recently having traveled to a geographic area where specific fungi are found in the soil may suggest fungal encephalitis (Davis, 1999; Leedom & Underman, 2000). Laboratory evaluation of blood shows an elevated white cell count and anemia. In some cases, serologic tests may show fungal antibodies in serum (Davis, 1999). The CSF shows an elevated white cell count and protein levels. *C. neoformans* is readily identified in the CSF fungal culture. The CSF culture is positive for other fungi in 50% of cases (Davis, 1999).

Neuroimaging is used to identify CNS changes related to fungal infection and will demonstrate fungal foci in organs initially invaded by the fungus. Although both MRI and CT scan are used in the workup for fungal encephalitis, MRI is more sensitive. The MRI may indicate lesions in the basal ganglia or thalamus, as well as hemorrhage, vascular complications, ischemia, aneurysm formation, or hydrocephalus (Go et al., 2000).

**Medical Management**

Medical management is directed at the causative fungus and the neurologic consequences of the infection. Seizures are controlled by standard antiseizure medications. Increased ICP is controlled by repeated lumbar punctures or shunting of CSF. In contrast to patients with cryptococcal meningitis, the use of repeated (once or twice daily) lumbar punctures in patients with fungal infections has been an effective strategy to control increased ICP and has been associated with improved survival with fewer neurologic sequelae (Davis, 1999).

Antifungal agents are given for a specific period of time to cure the infection in patients with competent immune systems. Patients with compromised immune systems will receive antifungal therapy until the infection is controlled, after which they will receive a maintenance dose of the medication for an indefinite period of time.

Although the dose and duration of treatment depend on the causative fungus, amphotericin B (Abelcet, AmBisome, Amphocin, Amphotec, Fungizone, and Fungizone IV) is the standard antifungal agent used in treatment (Karch, 2002). Dosing must be high enough to penetrate the blood–brain barrier without causing renal toxicity.

Fluconazole (Diflucan) or flucytosine (5-FC, 5-fluorocytosine, Ancobon) may be administered in conjunction with amphotericin B. Both can be given orally and may be used as maintenance therapy. Potential side effects of fluconazole include nausea, abdominal pain, headache, dizziness, rash, reversible alopecia, and a transient increase in liver enzymes. When flucytosine is prescribed in conjunction with amphotericin B, bone marrow suppression may occur. Therefore, patients receiving these medications in combination should have leukocyte and platelet counts monitored twice a week (Davis, 1999).

**Nursing Management**

If hydrocephalus develops and inflammation progresses, ICP will rise. Nursing assessment aimed at early identification of increased ICP is necessary to ensure early control and management. (See Chap. 61 for management of the patient with increased ICP.) Patient comfort may be optimized by administering nonopioid analgesics, limiting environmental stimuli, and positioning. Administration of amphotericin B may cause fever, chills, and body aches. Giving diphenhydramine (Benadryl) and acetaminophen (Tylenol) approximately 30 minutes before giving amphotericin B may prevent these side effects. Renal toxicity due to amphotericin B is dose-limiting. Monitoring the serum creatinine and blood urea nitrogen levels may alert the nurse to the development of renal insufficiency and the need to address the patient’s renal status.

Providing support will assist the patient and family to cope with the illness. Work-up of the patient for immunodeficiency diseases such as AIDS may put additional stress on the family. The nurse may need to mobilize community support systems for the patient and family.

**CREUTZFELDT-JAKOB AND NEW-VARIANT CREUTZFELDT-JAKOB DISEASE**

Creutzfeldt-Jakob disease (CJD) and new-variant Creutzfeldt-Jakob (nvCJD) disease belong to a group of degenerative, infectious neurologic disorders called transmissible spongiform encephalopathies (TSE). Although CJD and nvCJD have distinct clinical and histologic differences, they have many features in common. Both are rare and have incubation periods ranging from months to decades. In both, the symptoms are progressive, there is no definitive treatment, and the outcome is fatal.

CJD occurs primarily in adults ages 50 to 70. The incidence of disease is 1 per million worldwide (Weihl & Roos, 1999). nvCJD occurs in younger patients and has a prolonged duration of illness compared to CJD. The risk of nvCJD in the United States is thought to be low as cattle are fed primarily with soy-derived feed (see Pathophysiology, below). Only a few rare cases of TSE have occurred in the United States to date (Weihl & Roos, 1999).

**Pathophysiology**

Although still debated, the causative agent appears to be a prion, a proteinaceous, infectious particle smaller than a virus (Davis & Kennedy, 2000). The prion converts a normal cellular protein to an abnormal form, thus destroying neurons and glial cells. The gray matter takes on a spongy appearance (spongiform changes). Lesions, or plaques, also appear in various locations in the CNS (Weihl & Roos, 1999).

In CJD, the method of transmission is frequently unknown; however, direct transmission (by contact with infected animals) of the prion to humans may initiate the degenerative neurologic process. The disease is also heritable, and familial groups account for approximately 15% of cases, clustering in certain parts of the world. Iatrogenic transmission accounts for approximately 5% of cases and is due to contaminated neurosurgical devices and blood transfusions and the use of cadaver-derived growth hormone (growth hormone is now created synthetically) (World Health Organization, 2001).

Based primarily on an outbreak of cases in England in the late 1980s and through the 1990s, it was discovered that in nvCJD, the primary mode of transmission appeared to be the ingestion of CNS tissue of infected cattle. However, in 1998, additional concerns were raised about the safety of the English blood supply. The prion exists in lymphoid tissue and blood in all of the TSEs, but the incidence is higher in nvCJD. In light of the rising incidence of nvCJD, concern arose about the risk of infection through transfusion of blood products. There is no method available to screen blood for infectivity. All blood must be leukocyte-depleted prior to transfusion. In 1998, the use of plasma derived from citizens of the United Kingdom for use in manufacturing blood-derived products was banned in the U.S. (Weihl & Roos, 1999).
Clinical Manifestations

Many patients with CJD have vague prodromal symptoms prior to specific neurologic changes. Symptoms usually include behavioral changes, dementia, mutism, visual changes, cerebellar, pyramidal, and extrapyramidal signs, and myoclonic jerks. The myoclonic jerks may be spontaneous or precipitated by auditory or tactile stimuli. The myoclonus (spasms) may involve a single muscle group, a limb, or the entire body. The symptoms progress until the patient is completely unaware of the environment and immobilized.

Although the same type of agent, a prion, causes nvCJD, there are distinct differences in the clinical manifestations of nvCJD and CJD. In nvCJD, there are more prion-reactive plaques, referred to as florid plaques, surrounding spongiform tissue throughout the cerebrum and cerebellum. The characteristic EEG changes present in CJD are absent in nvCJD. Anxiety, depression, and behavioral changes are the initial symptoms of nvCJD. Cerebellar symptoms occur, with gait changes and ataxia. Myoclonus is present in most patients diagnosed with nvCJD. Memory and cognitive impairments occur late in the course of nvCJD. Mutism occurs in both nvCJD and CJD (Almond, 1998).

Assessment and Diagnostic Findings

Historically, sharp waves and spikes on the EEG were the only features available to support the diagnosis of CJD. Recent detection of a polyclonal antibody (protein 14-3-3) in CSF has enabled the diagnosis of CJD (Poser, Mollenhauder, Krab et al., 1999). In addition to the presence of a polyclonal antibody in CSF, a protein increase is demonstrated along with the presence of enzymes indicative of neuronal loss. CT scan is used to rule out disorders that may mimic the symptoms of CJD. MRI scans are useful, identifying lesions in the basal ganglia in most cases of CJD. Definitive diagnosis is made by brain biopsy or at autopsy.

Medical Management

After the onset of specific neurologic symptoms, progression of disease occurs quickly. There is no effective treatment for CJD or nvCJD. The care of the patient is supportive and palliative. Goals of care include prevention of injury related to immobility and dementia, promotion of patient comfort, and provision of support and education for the family. The duration of disease is 4 to 5 months in CJD and 16 months in nvCJD, with death occurring as a result of respiratory failure or sepsis (Weihl & Roos, 1999).

Nursing Management

As with medical management, the nursing care of patients is primarily supportive and palliative. Psychological and emotional support of patients and families throughout the course of the illness is needed. This care extends to providing for a dignified death and supporting the family through the processes of grief and loss. Hospice care should be used either at home or in an inpatient facility. (See Chap. 17 for an in-depth discussion of end-of-life issues.)

Prevention of disease transmission is an important part of nursing care. Although patient isolation is not necessary, use of standard precautions is important. Institutional protocols are followed for blood and body fluid exposure and decontamination of equipment. Conventional methods of sterilization do not destroy the prion. The CDC guidelines (based on WHO guidelines) outline the stringent sterilization methods that must be used to destroy the prion on surfaces.

Autoimmune Processes

Autoimmune nervous system disorders include multiple sclerosis, myasthenia gravis, and Guillain-Barré syndrome.

MULTIPLE SCLEROSIS

Multiple sclerosis (MS) is an immune-mediated progressive demyelinating disease of the CNS. Demyelination refers to the destruction of myelin, the fatty and protein material that surrounds certain nerve fibers in the brain and spinal cord; it results in impaired transmission of nerve impulses (Fig. 64-2). MS typically presents in young adults ages 20 to 40, and it affects women more frequently than men (Boyden, 2000).

The cause of MS is an area of ongoing research. Autoimmune activity results in demyelination, but the sensitized antigen has not been identified. Multiple factors play a role in the initiation of the immune process. Geographic prevalence is highest in northern Europe, southern Australia, the northern United States, and southern Canada (Noseworthy, Lucchinetti, Rodriguez et al., 2000). It is believed that an environmental exposure at a young age may play a role in the development of MS later in life.

Genetic predisposition is indicated by the presence of a specific cluster (haplotype) of human leukocyte antigens (HLA) on the cell wall. The presence of this haplotype may promote susceptibility to factors, such as viruses, that trigger the autoimmune response activated in MS. A specific virus capable of initiating the...
autoimmune response has not been identified. It is believed that DNA on the virus mimics the amino acid sequence of myelin, resulting in an immune system cross-reaction in the presence of a defective immune system (Boyden, 2000).

**Pathophysiology**

Sensitized T cells typically cross the blood–brain barrier; their function is to check the CNS for antigens and then leave. In MS, the sensitized T cells remain in the CNS and promote the infiltration of other agents that damage the immune system. The immune system attack leads to inflammation that destroys myelin (which normally insulates the axon and speeds the conduction of impulses along the axon) and oligodendroglial cells that produce myelin in the CNS.

Plaques of sclerotic tissue appear on demyelinated axons, further interrupting the transmission of impulses. Demyelination interrupts the flow of nerve impulses and results in a variety of manifestations, depending on which nerves are affected. Demyelinated axons are scattered irregularly throughout the CNS (Fig. 64-3). The areas most frequently affected are the optic nerves, chiasm, and tracts; the cerebrum; the brain stem and cerebellum; and the spinal cord. Eventually the axons themselves begin to degenerate, resulting in permanent and irreversible damage (Bashir & Whitaker, 2002; Halper, 2001).

**Clinical Manifestations**

The course of MS may assume many different patterns (Fig. 64-4). In some patients, the disease follows a benign course, with a normal life span and symptoms so mild that patients do not seek health care and treatment. Eighty percent to 85% of cases of MS begin with a relapsing–remitting course, with complete recovery between clearly defined symptomatic exacerbations (Noseworthy et al., 2000). This form of the disease does not progress between relapses, although the majority of cases with this initial type of course change to a secondary-progressive course after some years (Halper, 2001). Secondary-progressive MS begins as relapsing–remitting disease but changes to a course in which there is not full recovery but rather continued progression between defined relapses. Ten percent to 20% percent of patients have a primary progressive course (Noseworthy et al., 2000; Halper, 2001), in which symptoms progress throughout the disease, with increasing disability. Primary progressive MS is characterized by continuous decline, with the potential development of quadriparesis, cognitive dysfunction, visual loss, and brain stem syndromes.

The signs and symptoms of MS are varied and multiple, reflecting the location of the lesion (plaque) or combination of lesions. The primary symptoms most commonly reported are fatigue, depression, weakness, numbness, difficulty in coordination, loss of balance, and pain. Visual disturbances due to lesions in the optic nerves or their connections may include blurring of vision, *diplopia*, patchy blindness (scotoma), and total blindness.

Fatigue impairs optimal function throughout the course of the disease. Fatigue is exacerbated when febrile illness, environmental temperature, hot showers, and normal circadian rhythms during the afternoon elevate body temperature. Depression may relate to the pathophysiology or may occur as a reaction to the diagnosis. Suicide as the cause of death occurs 7.5 times more frequently among persons diagnosed with MS than among the age-matched general population. If suicide occurs, it is likely to occur within the first 5 years of diagnosis (Walther & Hohlfeld, 1999).
Pain occurs in 66% of patients with MS. Pain may be due to demyelination of pain fibers, mechanical stress on muscles, bones, and joints due to disability, or treatment measures (Maloni, 2000).

**Spasticity** (muscle hypertonicity) of the extremities and loss of the abdominal reflexes are due to involvement of the main motor pathways (pyramidal tracts) of the spinal cord. Disruption of the sensory axons may produce sensory dysfunction (paresthesias, pain). Cognitive and psychosocial problems may reflect frontal or parietal lobe involvement; some degree of cognitive change (eg, memory loss, decreased concentration) occurs in about half of patients, but severe cognitive changes with dementia (progressive organic mental disorder) are rare. Involvement of the cerebellum or basal ganglia can produce ataxia (impaired coordination of movements) and tremor. Loss of the control connections between the cortex and the basal ganglia may occur and cause emotional lability and euphoria. Bladder, bowel, and sexual dysfunctions are common.

Secondary complications of MS include urinary tract infections, constipation, pressure ulcers, contracture deformities, dependent pedal edema, pneumonia, reactive depression, and decreased bone mass. Emotional, social, marital, economic, and vocational problems may also be a consequence of the disease.

Exacerbations and remissions are characteristic of MS. During exacerbations, new symptoms appear and existing ones worsen; during remissions, symptoms decrease or disappear. Relapses may be associated with periods of emotional and physical stress. MRI studies demonstrate that many plaques do not produce serious symptoms; however, the disease may be very active, as demonstrated by MRI. There also is evidence that remyelination actually occurs in some patients.

**Assessment and Diagnostic Findings**

MRI is the primary diagnostic tool for visualizing plaques, documenting disease activity, and evaluating the effect of treatment. Electrophoresis of CSF identifies the presence of oligoclonal banding (several bands of immunoglobulin G bonded together, indicating an immune system abnormality). Evoked potential studies can help define the extent of the disease process and monitor changes. Underlying bladder dysfunction is diagnosed by urodynamic studies. Neuropsychological testing may be indicated to assess cognitive impairment. A sexual history helps to identify changes in sexual function.

**Gerontologic Considerations**

Due to improved treatment and an increase in the average life span for patients with MS, more individuals are living to become elderly. These patients may have chronic health problems for which they may be taking additional medications that could interact with medications prescribed for MS. The absorption, distribution, metabolism, and excretion of medications are altered in the elderly as a result of age-related changes in renal and liver functions. Therefore, the elderly must be monitored closely for adverse and toxic effects of MS medications and for osteoporosis (particularly with frequent corticosteroid use, which may be required for exacerbations). The cost of medications could lead to poor adherence to the prescribed regimen in elderly patients on fixed incomes.

Elderly MS patients have specific physical and psychosocial challenges. Physical challenges include impaired mobility, spasticity, and bladder dysfunction, impaired sleep, and an increased need for assistance with self-care (Klewer et al., 2001). Psychosocial issues include depression and suicidal thoughts (Klewer et al., 2001).

**Medical Management**

No cure exists for MS. An individualized, organized, and rational treatment program is indicated to relieve the patient’s symptoms and provide continuing support, particularly for individuals with cognitive changes (50%), who may need more structure and support. The goals of treatment are to delay the progression of the disease, manage chronic symptoms, and treat acute exacerbations. Many patients with MS have stable disease and require
only intermittent treatment, whereas others experience steady progression of their disease. Symptoms requiring intervention include spasticity, fatigue, bladder dysfunction, and ataxia. Management strategies target the various motor and sensory symptoms and effects of immobility that can occur.

**PHARMACOLOGIC THERAPY**

Three medications, referred to as the “ABC (and R) drugs,” are currently the main pharmacologic therapy for MS. The interferons beta-1a (Avonex) and beta-1b (Betaseron) reduce the frequency of relapse by 30% and decrease the appearance of new lesions on MRI by 80% (Tsels & Lisak, 1999). Glatiramer acetate (Copaxone) also reduces the number of lesions on MRI and the relapse rate. In March 2002, the FDA approved a fourth agent, Rebif, for the treatment of relapsing MS (PRISMS Study Group, 2001).

All of these medications have multiple immune activities. The interferons reduce T-cell proliferation; glatiramer acetate inhibits antigen-specific T-cell activation (Noseworthy et al., 2000). All of the disease-modifying medications, the “ABC (and R) drugs,” require injections. Interferon beta-1b (Betaseron) is administered subcutaneously every other day, interferon beta-1a (Avonex) is given by intramuscular injection once a week, and glatiramer acetate (Copaxone) is administered by subcutaneous injection every day. Rebif is administered subcutaneously three times a week. Seventy-five percent of patients taking one of the interferons experience flu-like symptoms; these symptoms can be controlled with nonsteroidal anti-inflammatory drugs (NSAIDs) and usually resolve after a few months of therapy (Walther & Hohlfeld, 1999). Patients receiving these injectable medications and their families must be knowledgeable about site reactions and other possible side effects (Ross, 2001).

Mitoxantrone (Novantrone), which received FDA approval in 2000 (Rolak, 2001), is an antineoplastic agent used primarily to treat leukemia and lymphoma. It received approval to treat secondary progressive MS due to its immunosuppressive qualities (Rolak, 2001). Patients need to have laboratory tests (complete blood count) performed, and the results must be closely monitored due to the potential for leukopenia and cardiac toxicity. A few patients (2% to 3%) will develop signs and symptoms of cardiomyopathy and heart failure due to cardiac toxicity (Rolak, 2001).

Corticosteroids modulate the immune response and are used to limit the severity and duration of exacerbations. These agents suppress the immune response and decrease inflammatory change. Nerve conduction is restored with variable degrees of symptomatic recovery. Typically the patient receives high-dose IV methylprednisolone followed by an oral prednisone taper. The nurse must carefully monitor the patient for side effects related to corticosteroids such as mood changes and fluid and electrolyte alterations and teach the patient and family about side effects.

Researchers continue to investigate other possible treatments for MS. Many agents that have been investigated have proven to be too toxic for clinical use. Researchers are studying strategies that facilitate the proliferation of anti-inflammatory cytokines. T-cell vaccination and agents that inhibit oxygen radicals and proteases are under study (Noseworthy et al., 2000). Medications are also prescribed for management of specific symptoms. Baclofen (Lioresal), a GABA agonist, is the medication of choice in treating spasms. It can be administered orally or by intrathecal injection. Benzodiazepines (Valium), tizanidine (Zanaflex), and dantrolene (Dantrium) may also be used to treat spasms. Patients with disabling spasms and contractures may require nerve blocks or surgical intervention. Fatigue that interferes with activities of daily living may be treated with amantadine (Symmetrel), pemoline (Cylert), or fluoxetine (Prozac). Ataxia is a chronic problem most resistant to treatment. Medications used to treat ataxia include beta-adrenergic blockers (Inderal), anti-seizure agents (Neurontin), and benzodiazepines (Klonopin).

Various strategies for pain management can be implemented, based on the type of pain that exists. Acute pain may be treated with antidepressants, opiates, or antiseizure medications. Surgical procedures may be required to interrupt the pain pathway. Subacute pain as well as chronic back pain can be effectively treated with NSAIDs. Physical therapy may also benefit the patient by improving posture and strength.

Pain may also be due to osteoporosis (Maloni, 2000). Perimenopausal women with MS are more likely to develop osteoporosis than those without MS. Immobility, corticosteroid therapy, and estrogen loss play a role in the development of osteoporosis in women with MS. Bone mineral density testing is recommended for this high-risk group (Smeltzer, Zimmerman, Capriotti & Fernandes, 2002). Diagnosis and treatment of osteoporosis are discussed at length in Chapter 68.

Management of bladder and bowel control is often among the patient’s most difficult problems and a variety of medications (anticholinergics, alpha-adrenergic blockers, or antispasmodic agents) may be prescribed. Nonpharmacologic strategies also assist in establishing effective bowel and bladder elimination (see Nursing Process section).

Urinary tract infection is often superimposed on the underlying neurologic dysfunction. Ascorbic acid (vitamin C) may be prescribed to acidify the urine, making bacterial growth less likely. Antibiotics are prescribed when appropriate.

**NURSING PROCESS:**

**THE PATIENT WITH MULTIPLE SCLEROSIS**

**Assessment**

Nursing assessment addresses actual and potential problems associated with the disease, including neurologic problems, secondary complications, and the impact of the disease on the patient and family. The patient’s movements and walking are observed to determine if there is danger of falling. Assessment of function is carried out both when the patient is well rested and when fatigued. The patient is assessed for weakness, spasticity, visual impairment, incontinence, and disorders of swallowing and speech. Additional areas of assessment include the following: How has MS affected the patient’s lifestyle? How well is the patient coping? What would the patient like to do better?

**Diagnosis**

**NURSING DIAGNOSES**

Based on the assessment data, the patient’s major nursing diagnoses may include the following:

- Impaired physical mobility related to weakness, muscle paresis, spasticity
- Risk for injury related to sensory and visual impairment
- Impaired urinary and bowel elimination (urgency, frequency, incontinence, constipation) related to nervous system dysfunction
- Impaired speech and swallowing related to cranial nerve involvement
• Disturbed thought processes (loss of memory, dementia, euphoria) related to cerebral dysfunction
• Ineffective individual coping related to uncertainty of course of MS
• Impaired home maintenance management related to physical, psychological, and social limits imposed by MS
• Potential for sexual dysfunction related to spinal cord involvement or psychological reactions to condition

Planning and Goals
The major goals for the patient may include promotion of physical mobility, avoidance of injury, achievement of bladder and bowel continence, promotion of speech and swallowing mechanisms, improvement of cognitive function, development of coping strengths, improved home maintenance management, and adaptation to sexual dysfunction.

Nursing Interventions
An individualized program of physical therapy, rehabilitation, and education is combined with emotional support. The nursing interventions include face-to-face and telephone interactions that address patient education to enable the person with MS to deal with the physiologic, social, and psychological problems that accompany chronic disease (Madonna & Keating, 1999).

PROMOTING PHYSICAL MOBILITY
Relaxation and coordination exercises promote muscle efficiency. Progressive resistive exercises are used to strengthen weak muscles because diminishing muscle strength is often significant in MS.

Exercises
Walking improves the gait, particularly when there is loss of position sense of the legs and feet. If certain muscle groups are irreversibly affected, other muscles can be trained to take over their actions. Instruction in the use of assistive devices may be needed to ensure their correct and safe use.

Minimizing Spasticity and Contractures
Muscle spasticity is common and, in its later stages, is characterized by severe adductor spasm of the hips with flexor spasm of the hips and knees. If this is not relieved, fibrous contractures of these joints with resultant pressure ulcers over the sacrum and hips (due to diminished sensation and the inability to position the patient properly) occur. Warm packs may be beneficial, but hot baths should be avoided because of risk for burn injury secondary to sensory loss and increasing symptoms that may occur with an elevation of the body temperature.

Daily exercises for muscle stretching are prescribed to minimize joint contractures. Special attention is given to the hamstrings, gastrocnemius muscles, hip adductors, biceps, and wrist and finger flexors. Muscle spasticity is common and interferes with normal function. A stretch—hold—relax routine is helpful for relaxing and treating muscle spasticity. Swimming and stationary bicycling are useful, and progressive weight-bearing can relieve spasticity in the legs. The patient should not be hurried in any of these activities because this often increases spasticity.

Activity And Rest
The patient is encouraged to work to a point just short of fatigue. Very strenuous physical exercise is not advisable because it raises the body temperature and may aggravate symptoms. The patient is advised to take frequent short rest periods, preferably lying down. Extreme fatigue may contribute to the exacerbation of symptoms.

Minimizing Effects of Immobility
Because of the decrease in physical activity that often occurs with MS, complications associated with immobility, including pressure ulcers, exhalatory muscle weakness, and accumulation of bronchial secretions, need to be considered and steps taken to prevent them. Measures to prevent such complications include assessment and maintenance of skin integrity and coughing and deep-breathing exercises.

PREVENTING INJURY
If motor dysfunction causes problems of incoordination and clumsiness, or if ataxia is apparent, the patient is at risk for falling. To overcome this disability, the patient is taught to walk with feet wide apart to widen the base of support and to increase walking stability. If there is loss of position sense, the patient is taught to watch the feet while walking. Gait training may require assistive devices (walker, cane, braces, crutches, parallel bars) and instruction about their use by a physical therapist. If the gait remains inefficient, a wheelchair or motorized scooter may be the solution. The occupational therapist is a valuable resource person in suggesting and securing aids to promote independence. If incoordination is a problem and tremor of the upper extremities occurs when voluntary movement is attempted (intention tremor), weighted bracelets or wrist cuffs are helpful. The patient is trained in transfer and activities of daily living.

Because sensory loss may occur in addition to motor loss, pressure ulcers are a continuing threat to skin integrity. Confinement to a wheelchair increases the risk. See Chapter 11 for a discussion of the prevention and treatment of pressure ulcers.

ENHANCING BLADDER AND BOWEL CONTROL
Generally, bladder symptoms fall into the following categories: (1) inability to store urine (hyperreflexic, uninhibited); (2) inability to empty the bladder (hyporeflexic, hypotonic); and (3) a mixture of both types. The patient with urinary frequency, urgency, or incontinence requires special support. The sensation of the need to void must be heeded immediately, so the bedpan or urinal should be readily available. A voiding time schedule is set up (every 1.5 to 2 hours initially, with gradual lengthening of the interval). The patient is instructed to drink a measured amount of fluid every 2 hours and then attempt to void 30 minutes after drinking. Using a timer or wristwatch with an alarm may be helpful for the patient who does not have enough sensation to signal the need to empty the bladder. The nurse encourages the patient to take the prescribed medications to treat bladder spasticity because this allows greater independence. Intermittent self-catheterization has been successful in maintaining bladder control in patients with MS. (See Chap. 11 for a discussion of intermittent self-catheterization.) If the female patient has permanent urinary incontinence, urinary diversion procedures may be considered. The male patient may wear a condom appliance for urine collection.

Bowel problems include constipation, fecal impaction, and incontinence. Adequate fluids, dietary fiber, and a bowel-training program are frequently effective in solving these problems. (See Chap. 11 for a discussion of promoting bowel continence.)

MANAGING SPEECH AND SWALLOWING DIFFICULTIES
When the cranial nerves controlling the mechanisms of speech and swallowing are involved, dysarthrias (defects of articulation) marked by slurring, low volume of speech, and difficulties in
phonation may occur. Swallowing disturbances (dysphagia) may also occur. A speech therapist evaluates speech and swallowing and instructs the patient, family, and health team members about strategies to compensate for speech and swallowing problems. The nurse reinforces this instruction and encourages the patient and family to adhere to the plan. Impaired swallowing increases the patient’s risk for aspiration; therefore, strategies (eg, having suction apparatus available, careful feeding, proper positioning for eating) are needed to reduce that risk (Galvan, 2001).

**IMPROVING SENSORY AND COGNITIVE FUNCTION**

Measures may be taken if visual defects (the cranial nerves affecting vision may be affected by MS) or changes in cognitive status occur.

**Vision**

An eye patch or a covered eyeglass lens may be used to block visual impulses of one eye when the patient has diplopia (double vision). Prism glasses may be helpful for the bedridden patient who is having difficulty reading in the supine position. People unable to read regular-print materials are eligible for the free talking book services of the Library of Congress or may obtain large-type books from local libraries.

**Cognition and Emotional Responses**

Cognitive impairment and emotional lability may occur early in MS in some patients and may impose numerous stresses on the patient and family. Some patients with MS are forgetful and easily distracted and may exhibit emotional lability.

Patients adapt to illness in a variety of ways, which may include denial, depression, withdrawal, and hostility. Emotional support assists patients and their families to adapt to the changes and uncertainties associated with MS and to cope with the disruption in their lives. The patient is assisted to set meaningful and realistic goals to achieve a sense of purpose, to remain as active as possible, and to keep up social interests and activities. Hobbies may help the patient’s morale and provide satisfying interests if the disease progresses to the stage in which formerly enjoyed activities can no longer be pursued.

The family should be made aware of the nature and degree of cognitive impairment. The environment is kept structured, and lists and other memory aids are used to help the patient with cognitive changes to maintain a daily routine. The occupational therapist can be helpful in formulating a structured daily routine.

**Strengthening Coping Mechanisms**

The diagnosis of MS is always distressing to the patient and family. They need to know that no two patients with MS have identical symptoms or courses of illness. Although some patients do experience significant disability early, others have a near-normal life span with minimal disability. Some families, however, face overwhelming frustrations and problems. MS affects people who are often in a productive stage of life and concerned about career and family responsibilities. Family conflict, disintegration, separation, and divorce are not uncommon. Often, very young family members assume the responsibility of caring for a parent with MS. Nursing interventions in this area include alleviating stress and making appropriate referrals for counseling and support to minimize the adverse effects of dealing with chronic illness.

The nurse, mindful of these complex problems, initiates home care and coordinates a network of services, including social services, speech therapy, physical therapy, and homemaker services. To strengthen the patient’s coping skills, as much information as possible is provided. People who live with chronic illness need an updated list of the assistive devices, services, and resources that are available.

Coping through problem solving involves helping the patient define the problem and develop alternatives for its management. Careful planning and maintaining flexibility and a hopeful attitude are useful for psychological and physical adaptation.

**IMPROVING SELF-CARE ABILITIES**

MS can affect every facet of daily living. After certain abilities are lost, they are often impossible to regain. Physical function may vary from day to day. Modifications that allow independence in self-care should be implemented (eg, assistive eating devices, raised toilet seat, bathing aids, telephone modifications, long-handled comb, tongs, modified clothing). Physical and emotional stresses should be avoided as much as possible because these may worsen symptoms and impair performance. Exposure to heat increases fatigue and muscle weakness, so air conditioning in at least one room is recommended. Exposure to extreme cold may increase spasticity.

**PROMOTING SEXUAL FUNCTIONING**

Patients with MS and their partners face problems that interfere with sexual activity, arising not only as a direct consequence of nerve damage but also from psychological reactions to the disease. Easy fatigability, conflicts arising from dependency and depression, emotional lability, loss of self-esteem, and feelings of low self-worth compound the problem. Erectile and ejaculatory disorders in men and orgasmic dysfunction and adductor spasms of the thigh muscles in women can make sexual intercourse difficult or impossible. Bladder and bowel incontinence and urinary tract infections add to the difficulties.

An experienced sexual counselor helps bring into focus the patient’s or partner’s sexual resources and suggests relevant information and supportive therapy (Sipski & Alexander, 1997). Sharing and communicating feelings, planning for sexual activity (to minimize the effects of fatigue), and exploring alternative methods of sexual expression may open up a wide range of sexual enjoyment and experiences.

**PROMOTING HOME AND COMMUNITY-BASED CARE**

**Teaching Patients Self-Care**

As the disease progresses, the patient and family need to learn new strategies to maintain optimal independence. Teaching of new self-care techniques may be initiated in the hospital or clinic setting and reinforced in the home. Teaching about self-care may address the use of assistive devices, self-catheterization, and administration of medications that affect the course of the disease or treat complications. Although the disease-modifying medications (the “ABC and R medications”) may slow the progression of disease and disability in many persons with MS, they are not effective in all patients. Patients who receive these medications will require teaching and support, while those unable to take them or for whom the medications have not been effective need continued support and assistance in coping with this reality. Exercises that enable the patient to continue some form of activity or that maintain or improve swallowing, speech, or respiratory function may be taught to the patient and family (Chart 64-3).

**Continuing Care**

After discharge, the home care nurse often provides teaching and reinforcement of these new techniques in the patient’s home. Nurses in the home setting assess for changes in the patient’s
At the completion of the home care instruction, the patient or caregiver will be able to:

- State how to access the local chapter of the National MS Society and available resources.
- Discuss the clinical course of MS.
- Identify strategies to manage symptoms (pain, cognitive responses, dysphagia, tremors, visual disturbances).
- State how to prevent complications (pressure ulcers, pneumonia, depression).
- Identify coping strategies.
- Identify ways to minimize fatigue.
- Explain how to prevent injury.
- State ways to adapt to sexual dysfunction.
- Discuss ways to control bowel and bladder function.
- Name benefits of exercise and physical activity.
- Identify ways to minimize immobility and spasticity.
- Describe medication regimen and potential adverse effects.
- Demonstrate correct techniques of administering injectable medications, if prescribed.

Evaluation

EXPECTED PATIENT OUTCOMES

Expected patient outcomes may include:

1. Improved physical mobility
   a. Participates in gait-training and rehabilitation program
   b. Establishes a balanced program of rest and exercise
   c. Uses assistive devices correctly and safely

2. Is free of injury
   a. Uses visual cues to compensate for decreased sense of touch or position
   b. Asks for assistance when necessary

3. Attains or maintains control of bladder and bowel patterns
   a. Monitors self for urine retention and employs intermittent self-catheterization technique, if indicated

Nursing Research Profile 64-1

Quality of Life for People With Multiple Sclerosis


**Purpose**

The purpose of this study was to test an explanatory model of variables influencing health promotion and quality of life in persons with multiple sclerosis (MS). This was the second phase of a project designed to investigate the issues surrounding the health-promotion needs of individuals with chronic disabling conditions and the outcomes associated with the performance of health-promoting behaviors.

**Study Sample and Design**

This was a one-time cross-sectional design using mailed questionnaires. Participants were recruited for the study using targeted mailings to individuals with MS on the mailing lists of national MS Society chapters. The study sample included 786 persons with MS (630 women and 156 men) who returned surveys. Instruments used included the Incapacity Status Scale (ISS), Barriers to Health Promoting Activities for Disabled Persons Scale, Personal Resource Questionnaire, Acceptance of Illness Scale, Self-Rated Abilities for Health Practices scale, Health Promoting Lifestyle Profile-II (HPLP-II), and the Quality of Life Index (QLI).

**Findings**

The analysis revealed that the original model was generally well specified, with an adequate fit to the data. The model supported the hypothesis that quality of life is the outcome of a complex interplay between contextual factors (severity of illness), antecedent variables, and health-promoting behaviors. The revised model, with the addition of two paths between personal resources and barriers, had an improved fit of the data compared to the original model. The original and revised models are depicted in the article. Resources, barriers, self-efficacy, and acceptance accounted for 58% of the variance in the frequency of health-promoting behaviors and 66% of the variance in perceived quality of life.

**Nursing Implications**

Further testing of the model was recommended, especially in samples of differing ethnic and cultural backgrounds. Nurses working with MS patients should be aware that quality of life is the outcome of the interaction of many variables. The authors suggest that nurses can improve health-promoting behaviors and quality of life using interventions that enhance social support, decrease barriers, and increase specific self-efficacy for health behaviors.
b. Identifies the signs and symptoms of urinary tract infection
c. Maintains adequate fluid and fiber intake

4. Participates in strategies to improve speech and swallowing
   a. Practices exercises recommended by speech therapist
   b. Maintains adequate nutritional intake without aspiration

5. Compensates for altered thought processes
   a. Uses lists and other aids to compensate for memory losses
   b. Discusses problems with trusted advisor or friend
   c. Substitutes new activities for those that are no longer possible

6. Demonstrates effective coping strategies
   a. Maintains sense of control
   b. Modifies lifestyle to fit goals and limitations
   c. Verbalizes desire to pursue goals and developmental tasks of adulthood
   d. Adheres to plan for home maintenance management
   e. Uses appropriate self-care techniques to maintain independence
   f. Engages in health promotion activities and health screenings as appropriate

8. Adapts to changes in sexual function
   a. Is able to discuss problem with partner and appropriate health professional
   b. Identifies alternate means of sexual expression

**MYASTHENIA GRAVIS**

Myasthenia gravis, an autoimmune disorder affecting the myoneural junction, is characterized by varying degrees of weakness of the voluntary muscles. Women tend to develop the disease at an earlier age (20 to 40 years of age) compared to men (60 to 70 years of age), and women are affected more frequently (Heitmiller, 1999).

**Pathophysiology**

Normally, a chemical impulse precipitates the release of acetylcholine from vesicles on the nerve terminal at the myoneural junction. The acetylcholine attaches to receptor sites on the motor end plate, stimulating muscle contraction. Continuous binding of acetylcholine to the receptor site is required for muscular contraction to be sustained.

In myasthenia gravis, autoantibodies directed at the acetylcholine receptor sites impair transmission of impulses across the myoneural junction. Therefore, fewer receptors are available for stimulation, resulting in involuntary muscle weakness that escalates with continued activity (Fig. 64-5). These antibodies are found in 80% to 90% of the people with myasthenia gravis. Eighty percent of persons with myasthenia gravis have either thymic hyperplasia or a thymic tumor (Roos, 1999), and the thymus gland is believed to be the site of antibody production. In patients who are antibody negative, it is believed that the offending antibody is directed at a portion of the receptor site rather than the whole complex.

**Clinical Manifestations**

The initial manifestation of myasthenia gravis usually involves the ocular muscles. Diplopia (double vision) and ptosis (drooping of the eyelids) are common. However, the majority of patients also experience weakness of the muscles of the face and throat (bulbar symptoms) and generalized weakness. Weakness of the facial muscles will result in a bland facial expression. Laryngeal involvement produces *dysphonia* (voice impairment) and increases the patient’s risk for choking and aspiration. Generalized weakness affects all the extremities and the intercostal muscles, resulting in decreasing vital capacity and respiratory failure. Myasthenia gravis is purely a motor disorder with no effect on sensation or coordination.

**Assessment and Diagnostic Findings**

An anticholinesterase test is used to diagnose myasthenia gravis. Anticholinesterase agents stop the breakdown of acetylcholine, thereby increasing acetylcholine availability. Edrophonium chloride (Tensilon) is injected intravenously, 2 mg at a time to a total of 10 mg. Thirty seconds after injection, facial muscle weakness and ptosis should resolve for about 5 minutes. This immediate improvement in muscle strength after administration of this agent represents a positive test and usually confirms the diagnosis. Atropine 0.4 mg should be available to control the side effects of edrophonium, which include bradycardia, sweating, and cramping (Roos, 1999).

The acetylcholine receptor antibody titer is elevated as indicated previously. Repetitive nerve stimulation tests record the electrical activity in targeted muscles after nerve stimulation. A 15% decrease in successive action potentials is observed in patients with myasthenia gravis (Heitmiller, 1999). The thymus gland, which is a site of acetylcholine receptor antibody production, is enlarged in myasthenia gravis. MRI demonstrates this enlargement in 90% of cases (Wilkins & Bulkley, 1999).

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**Figure 64-5** Myasthenia gravis. (A) Normal ACh receptor site. (B) ACh receptor site in myasthenia gravis.
Medical Management

Management of myasthenia gravis is directed at improving function and reducing and removing circulating antibodies. Therapeutic modalities include administration of anticholinesterase agents and immunosuppressive therapy, plasmapheresis, and thymectomy.

PHARMACOLOGIC THERAPY
Anticholinesterase agents such as pyridostigmine bromide (Mestinon) and neostigmine bromide (Prostigmin) provide symptomatic relief by increasing the relative concentration of available acetylcholine at the neuromuscular junction. Dosage is increased gradually until maximal benefits (improved strength, less fatigue) are obtained. Adverse effects of anticholinesterase therapy include abdominal pain, diarrhea, nausea, and increased oropharyngeal secretions. Pyridostigmine tends to have fewer side effects (Chart 64-4). Improvement with anticholinesterase therapy is not complete or long-lasting (Heitmiller, 1999).

PHARMACOLOGY

The goal of immunosuppressive therapy is to reduce the production of the antibody. Corticosteroids suppress the patient’s immune response, thus decreasing the amount of antibody production. As the corticosteroid dosage is gradually increased, the anticholinesterase dosage is lowered. The patient’s ability to maintain effective respirations and to swallow is monitored throughout. Prednisone, taken on alternate days to lower the incidence of side effects, appears to be successful in suppressing the disease. The patient sometimes shows a marked decrease in muscle strength right after therapy is started, but this is usually only temporary.

Cytotoxic medications have also been used, although the precise mechanism of action in myasthenia is not fully understood. Medications such as azathioprine (Imuran), cyclophosphamide (Cytoxan), and cyclosporine reduce the circulating anti-acetylcholine receptor antibody titers. Side effects are significant; therefore, these agents are reserved for patients who do not respond to other forms of therapy.

A number of medications are contraindicated for patients with myasthenia gravis because they worsen myasthenic symptoms. Risks and benefits should be weighed by the physician and the patient before taking any new medications, including antibiotics, cardiovascular medications, antiseizure and psychotropic medications, morphine, quinine and related agents, beta-blockers, and nonprescription medications. Procaine (Novocain) should be avoided, and the patient’s dentist is so advised.

PLASMAPHERESIS
Plasma exchange (plasmapheresis) is a technique used to treat exacerbations. The patient’s plasma and plasma components are removed through a centrally placed large-bore double-lumen catheter. The blood cells and antibody-containing plasma are separated; then the cells and a plasma substitute are reinfused. Plasma exchange produces a temporary reduction in the titer of circulating antibodies. Plasma exchange improves the symptoms in 75% of patients, although improvement lasts only a few weeks unless plasmapheresis is continued or other forms of treatment such as immunosuppression with corticosteroids are initiated (Bedlack & Sanders, 2000). IV immune globulin (IVIG) has recently been shown to be nearly as effective as plasmapheresis in controlling symptom exacerbation (Qureshi, Choudhry, Akbar et al., 1999). However, neither therapy is a cure as it does not stop the production of the acetylcholine receptor antibodies.

SURGICAL MANAGEMENT
Thymectomy (surgical removal of the thymus gland) can produce antigen-specific immunosuppression and result in clinical improvement. It can decrease or eliminate the need for medication. In one study 92% of post-thymectomy patients had symptomatic improvement, with 50% of them no longer requiring pharmacologic therapy (Wilkins & Bulkley, 1999). The entire gland must be removed for optimal clinical outcomes; therefore, surgeons prefer the transsternal surgical approach. After surgery, the patient is monitored in an intensive care unit, with special attention to respiratory function. After the thymus gland is removed, it may take up to 1 year for the patient to benefit from the procedure due to the long life of circulating T cells (Wilkins & Bulkley, 1999).

Complications: Myasthenic Crisis Versus Cholinergic Crisis

A myasthenic crisis is an exacerbation of the disease process characterized by severe generalized muscle weakness and respiratory and bulbar weakness that may result in respiratory failure. Crisis may result from disease exacerbation or a specific precipitating event. The most common precipitator is infection; others include medication change, surgery, pregnancy, and high environmental temperature (Bella & Chad, 1998).

Symptoms of anticholinergic overmedication (cholinergic crisis) may mimic the symptoms of exacerbation. Differentiation can be achieved with the edrophonium chloride (Tensilon) test. The patient with myasthenic crisis improves immediately following administration of edrophonium, while the patient with
cholinergic crisis may experience no improvement or deteriorate. If myasthenic crisis is diagnosed, neostigmine methylsulfate (PMS-Neostigmine, Prostigmin) is administered intramuscularly or intravenously until the patient is able to swallow oral anticholinesterase medications. Plasmapheresis and IVIG, which reduce the antibody load, also may be used to treat myasthenic crisis. If cholinergic crisis is identified, all anticholinesterase medications are stopped. The patient receives atropine (Atropine sulfa- fite), the antidote for the anticholinesterase medications.

Neuromuscular respiratory failure is the critical complication of crisis. Respiratory muscle and bulbar weakness combine to cause respiratory compromise. Weak respiratory muscles will not support inhalation. An inadequate cough and an impaired gag reflex caused by bulbar weakness result in poor airway clearance. Values on two respiratory function tests, the negative inspiratory force and vital capacity, will be the first clinical signs to deteriorate. Careful monitoring of these values enables the nurse to monitor for impending respiratory failure. Respiratory support and airway protection are key interventions for the nurse caring for the patient in crisis. Endotracheal intubation and mechanical ventilation may be needed (see Chap. 25). Nutritional support may be needed if the patient is intubated for a long period.

Nursing Management

Because myasthenia gravis is a chronic disease and most patients are seen on an outpatient basis, much of the nursing care focuses on patient and family teaching. Educational topics for outpatient self-care include medication management, energy conservation, strategies to help with ocular manifestations, and prevention and management of complications.

Medication management is a crucial component of ongoing care. Understanding the action of the medications and taking them on schedule is emphasized, as are the consequences of delaying medication and the signs and symptoms of myasthenic and cholinergic crisis. The patient can determine the best times for daily dosing by keeping a diary to determine fluctuation of symptoms and to learn when the medication is wearing off. The medication schedule can then be manipulated to maximize strength throughout the day.

The patient is also taught strategies to conserve energy. To do this, the nurse helps the patient identify the best times for rest periods throughout the day. If the patient lives in a two-story home, the nurse can suggest that frequently used items such as hygiene products, cleaning products, and snacks be kept on each floor to minimize travel between floors. The patient is encouraged to apply for a handicapped license plate to minimize walking from parking spaces and to schedule activities to coincide with peak energy and strength levels.

To minimize the risk of aspiration, mealtimes should coincide with the peak effects of anticholinesterase medication. In addition, rest before meals is encouraged to reduce muscle fatigue. The patient is advised to sit upright during meals with the neck slightly flexed to facilitate swallowing. Soft foods in gravy or sauces can be swallowed more easily; if choking occurs frequently, the nurse can suggest pureeing food to a pudding consistency. Suction should be available at home and the patient and family instructed in its use. Gastrostomy feedings may be necessary in some patients to ensure adequate nutrition.

Impaired vision results from ptosis of one or both eyelids, decreased eye movement, or double vision. To prevent corneal damage when the eyelids do not close completely, the patient is instructed to tape the eyes closed for short intervals and regularly instill artificial tears. Patients who wear eyeglasses can have "crutches" attached to help lift the eyelids. Patching one eye can help with double vision.

The patient is reminded of the importance of maintaining health promotion practices and of following health care screening recommendations. Factors that will exacerbate symptoms and potentially cause crisis should be noted and avoided: emotional stress, infections (particularly respiratory infections), vigorous physical activity, some medications, and high environmental temperature. The Myasthenia Gravis Foundation of America provides support groups, services, and educational materials for patients, families, and health care providers.

MANAGING MYASTHENIC AND CHOLINERGIC CRISIS

Respiratory distress and varying degrees of dysphagia (difficulty swallowing), dysarthria (difficulty speaking), eyelid ptosis, diplopia, and prominent muscle weakness are symptoms of myasthenic and cholinergic crisis. The patient is placed in an intensive care unit for constant monitoring because of associated intense and sudden fluctuations in clinical condition.

IV edrophonium chloride (Tensilon) is used to differentiate the type of crisis. It improves the condition of the patient in myasthenic crisis and temporarily worsens that of the patient in cholinergic crisis. If the patient is in true myasthenic crisis, neostigmine methylsulfate is administered intramuscularly or intravenously. If the edrophonium test is inconclusive or there is increasing respiratory weakness, all anticholinesterase medications are stopped, and atropine sulfate is given to reduce excessive secretions.

Providing ventilatory assistance takes precedence in the immediate management of the patient with myasthenic crisis. Ongoing assessment for respiratory failure is essential. The nurse assesses the respiratory rate, depth, and breath sounds and monitors pulmonary function parameters (vital capacity and negative inspiratory force) to detect pulmonary problems before respiratory dysfunction progresses. Blood is drawn for arterial blood gas analysis. Endotracheal intubation and mechanical ventilation may be needed (see Chap. 25).

When there is severe weakness of the abdominal, intercostal, and pharyngeal muscles, the patient cannot cough, take deep breaths, or clear secretions. Chest physical therapy, including postural drainage to mobilize secretions, and suctioning to remove secretions may have to be performed frequently. (Postural drainage should not be performed for 30 minutes after feeding.)

Assessment strategies and supportive measures include the following:

- Arterial blood gases, serum electrolytes, input and output, and daily weight are monitored.
- If the patient cannot swallow, nasogastric tube feedings may be prescribed.
- Sedatives and tranquilizers are avoided because they aggravate hypoxia and hypercapnia and can cause respiratory and cardiac depression.

GUILLAIN-BARRÉ SYNDROME

Guillain-Barré syndrome is an autoimmune attack of the peripheral nerve myelin. The result is acute, rapid segmental demyelination of peripheral nerves and some cranial nerves, producing ascending weakness with dyskinesia (inability to execute voluntary movements), hyporeflexia, and paresthesias (numbness). In 66% of cases, there is a predisposing event, most often a respiratory or gastrointestinal infection, although vaccination, pregnancy,
and surgery have also been identified as antecedent events (Bella & Chad, 1998). Infection with Campylobacter jejuni (a relatively common gastrointestinal bacterial pathogen) precedes Guillain-Barré syndrome in a few cases (Ho & Griffin, 1999; Lindenbaum, Kissel & Mendel, 2001).

The antecedent event usually occurs 2 weeks before symptoms begin. Weakness usually begins in the legs and progresses upward for about 1 month. Maximum weakness varies but usually includes neuromuscular respiratory failure and bulbar weakness. The duration of the symptoms is variable; complete functional recovery may take up to 2 years (Hickey, 2003). Any residual symptoms are permanent and reflect axonal damage from demyelination.

The annual incidence of Guillain-Barré is 0.6 to 1.9 cases per 100,000. Eighty-five percent of patients recover with minimal residual symptoms. Severe residual deficits occur in up to 10% of patients. Residual deficits are most likely in patients with rapid disease progression, those who require mechanical ventilation, or those 60 years of age or older. Death occurs in 3% to 8% of cases, resulting from respiratory failure, autonomic dysfunction, sepsis, or pulmonary emboli (Bella & Chad, 1998).

Pathophysiology

Myelin is a complex substance that covers nerves, providing insulation and speeding the conduction of impulses from the cell body to the dendrites. The cell that produces myelin in the peripheral nervous system is the Schwann cell. In Guillain-Barré the Schwann cell is spared, allowing for remyelination in the recovery phase of the disease.

Guillain-Barré is the result of a cell-mediated immune attack on peripheral nerve myelin proteins (Ho & Griffin, 1999). The best-accepted theory is that an infectious organism contains an amino acid that mimics the peripheral nerve myelin protein. The immune system cannot distinguish between the two proteins and attacks and destroys peripheral nerve myelin. Studies indicate that an exact location within the peripheral nervous system, the ganglioside GM1b, is the most likely target of the immune attack (Yuki, Ang, Koga et al., 2000). With the autoimmune attack there is an influx of macrophages and other immune-mediated agents that attack myelin, cause inflammation and destruction, and leave the axon unable to support nerve conduction.

Clinical Manifestations

Classic Guillain-Barré begins with muscle weakness and diminished reflexes of the lower extremities. Hyporeflexia and weakness progress and may result in quadriplegia. Demyelination of the nerves that innervate the diaphragm and intercostal muscles results in neuromuscular respiratory failure. Twenty-five percent of patients will require mechanical ventilation within 18 days of symptom onset (Bella & Chad, 1998). Sensory symptoms include paresthesias of the hands and feet and pain related to the demyelination of sensory fibers.

Cranial nerve demyelination can result in a variety of clinical manifestations. Optic nerve demyelination may result in blindness. Bulbar muscle weakness related to demyelination of the glossopharyngeal and vagus nerves results in an inability to swallow or clear secretions. Vagus nerve demyelination results in autonomic dysfunction, manifested by instability of the cardiovascular system. The presentation is variable and may include tachycardia, bradycardia, hypertension, or orthostatic hypotension. The symptoms of autonomic dysfunction occur and resolve rapidly. Guillain-Barré does not affect cognitive function or level of consciousness.

While the classic clinical features include areflexia and ascending weakness, variation in presentation occurs. There may be a sensory presentation, with progressive sensory symptoms, an atypical axonal destruction, and the Miller-Fisher variant, which includes paralysis of the ocular muscles, ataxia, and areflexia (Ho & Griffin, 1999).

Assessment and Diagnostic Findings

The patient presents with symmetric weakness, diminished reflexes, and upward progression of motor weakness. A history of a viral illness in the previous few weeks suggests the diagnosis. Changes in vital capacity and negative inspiratory force are assessed to identify impending neuromuscular respiratory failure. Serum laboratory tests are not useful in the diagnosis. However, elevated protein levels are detected in CSF evaluation, without an increase in other cells. Evoked potential studies demonstrate a progressive loss of nerve conduction velocity (Bella & Chad, 1999).

Medical Management

Because of the possibility of rapid progression and neuromuscular respiratory failure, Guillain-Barré is a medical emergency, requiring intensive care unit management. Careful assessment of changes in motor weakness and respiratory function alert the clinician to the physical and respiratory needs of the patient. Respiratory therapy or mechanical ventilation may be necessary to support pulmonary function and adequate oxygenation. Mechanical ventilation may be required for an extended period. The patient is weaned from mechanical ventilation when the respiratory muscles can again support spontaneous respiration and maintain adequate tissue oxygenation.

Other interventions are aimed at preventing the complications of immobility. These may include the use of anticoagulant agents and thigh-high elastic compression stockings or sequential compression boots to prevent thrombosis and pulmonary embolism. Plasmapheresis and IVIG are used to directly affect the peripheral nerve myelin antibody level. Both therapies decrease circulating antibody levels and reduce the amount of time the patient is immobilized and dependent on mechanical ventilation. Studies indicate that IVIG and plasmapheresis are equally effective in treating Guillain-Barré (Bella & Chad, 1999; Winer, 2002).

The cardiovascular risks posed by autonomic dysfunction require continuous ECG monitoring. Tachycardia and hypertension are treated with short-acting medications such as alpha-adrenergic blocking agents. Hypotension is managed by increasing the amount of IV fluid administered. The use of short-acting agents is important because autonomic dysfunction is very labile.

NURSING PROCESS: THE PATIENT WITH GUILLAIN-BARRÉ SYNDROME

Assessment

Ongoing assessment for disease progression is critical. The patient is monitored for life-threatening complications (respiratory failure, cardiac dysrhythmias, DVTs) so that appropriate interventions can be initiated. Because of the threat to the patient in this sudden, potentially life-threatening disease, the nurse must assess the patient’s and family’s ability to cope and their use of appropriate coping strategies.
Diagnosis

NURSING DIAGNOSES
Based on the assessment data, the patient’s major nursing diagnoses may include the following:

- Ineffective breathing pattern and impaired gas exchange related to rapidly progressive weakness and impending respiratory failure
- Impaired physical mobility related to paralysis
- Imbalanced nutrition, less than body requirements, related to inability to swallow
- Impaired verbal communication related to cranial nerve dysfunction
- Fear and anxiety related to loss of control and paralysis

Collaborative Problems/Potential Complications
Based on the assessment data, potential complications that may develop include the following:

- Respiratory failure
- Autonomic dysfunction

Planning and Goals
The major goals for the patient may include improved respiratory function, increased mobility, improved nutritional status, effective communication, decreased fear and anxiety, and absence of complications.

Nursing Interventions

Maintaining Respiratory Function
Respiratory function can be maximized with incentive spirometry and chest physiotherapy. Monitoring for changes in vital capacity and negative inspiratory force are key to early intervention for neuromuscular respiratory failure. Mechanical ventilation is required if the vital capacity falls, making spontaneous breathing impossible and tissue oxygenation inadequate.

Parameters for determining the appropriate time to begin mechanical ventilation include a vital capacity of 12 to 15 mL/kg, downward vital capacity trend over 4 to 6 hours, and an inability to clear secretions (Bella & Chad, 1999). The potential need for mechanical ventilation should be discussed with the patient and family on admission to provide time for psychological preparation and decision-making. Intubation and mechanical ventilation will result in less anxiety if it is initiated on a nonemergent basis to a well-informed patient. The patient may require mechanical ventilation for a long period. Nursing management of the patient requiring mechanical ventilation is discussed in Chapter 25.

Bulbar weakness that impairs the ability to swallow and clear secretions is another factor in the development of respiratory failure in the patient with Guillain-Barré. Suctioning may be needed to maintain a clear airway.

The nurse assesses the blood pressure and heart rate frequently to identify autonomic dysfunction so that interventions can be initiated quickly if needed. Medications are administered or a temporary pacemaker is placed for clinically significant bradycardia (Winer, 2002).

Enhancing Physical Mobility
Nursing interventions to enhance physical mobility and prevent the complications of immobility are key to the function and survival of these patients. The paralyzed extremities are supported in functional positions, and passive range-of-motion exercises are performed at least twice daily.

DVT and pulmonary embolism are threats to the paralyzed patient. Nursing interventions are aimed at preventing DVT. Range-of-motion exercises, altering positioning, anticoagulation, thigh-high elastic compression stockings or sequential compression boots, and adequate hydration will decrease the risk for DVT.

Padding may be placed over bony prominences such as the elbows and heels to reduce the risk for pressure ulcers. The need for consistent position changes every 2 hours cannot be overemphasized. The nurse evaluates laboratory test results that may indicate malnutrition or dehydration, both of which increase the risk for pressure ulcers. Collaboration with the physician and dietitian will result in a plan to meet the patient’s nutritional and hydration needs.

Providing Adequate Nutrition
Paralytic ileus may result from insufficient parasympathetic activity. In this event, the nurse administers IV fluids and parenteral nutrition as prescribed and monitors for the return of bowel sounds. If the patient cannot swallow due to bulbar paralysis (immobility of muscles), a gastrostomy tube may be placed to administer nutrients. The nurse carefully assesses the return of the gag reflex and bowel sounds before resuming oral nutrition.

Improving Communication
Because of paralysis and ventilator management, the patient cannot talk, laugh, or cry and thus has no method for communicating needs or expressing emotion. Establishing some form of communication with picture cards or an eye blink system will provide a means of communication. Collaboration with the speech therapist may be helpful in developing a communication mechanism that is most effective for a specific patient.

Decreasing Fear and Anxiety
The patient and family are faced with a sudden, potentially life-threatening disease, and anxiety and fear are constant themes for them. The impact of disease on the family will depend on the patient’s age and role within the family. Referral to a support group may provide information and support to the patient and family.

The family may feel helpless in caring for the patient. Mechanical ventilation and monitoring devices may frighten and intimidate them. Family members often want to participate in physical care; with instruction and support by the nurse, they should be allowed to do so.

In addition to fear, the patient may experience isolation, loneliness, and lack of control. Nursing interventions that increase the patient’s sense of control include providing information about the condition, emphasizing a positive appraisal of coping resources, and teaching relaxation exercises and distraction techniques. The positive attitude and atmosphere of the multidisciplinary team are important to promote a sense of well-being.

Diversional activities are encouraged to decrease loneliness and isolation. Encouraging visitors, engaging visitors or volunteers to read to the patient, listening to music or books on tape, and watching television are ways to alleviate the patient’s sense of isolation.

Monitoring and Managing Potential Complications
Thorough assessment of respiratory function at regular intervals is essential because respiratory insufficiency and subsequent fail-
ure due to weakness or paralysis of the intercostal muscles and diaphragm may develop quickly. Respiratory failure is the major cause of mortality, which is reported to be as high as 10% to 20%. Vital capacity is monitored frequently and at regular intervals in addition to respiratory rate and the quality of respirations, so that respiratory insufficiency can be anticipated. Decreasing vital capacity associated with weakness of the muscles used in swallowing, which causes difficulty in both coughing and swallowing, indicates impending respiratory failure. Signs and symptoms include breathlessness while speaking, shallow and irregular breathing, use of accessory muscles, tachycardia, and changes in respiratory pattern.

Parameters for determining the onset of respiratory failure are established on admission, allowing intubation and the initiation of mechanical ventilation on a nonemergent basis. This also allows the patient to be prepared for the procedure in a controlled manner, which reduces anxiety and complications.

Other complications include cardiac dysrhythmias, which necessitate ECG monitoring, transient hypertension, orthostatic hypotension, DVT, pulmonary embolism, urinary retention, and other threats to any immobilized and paralyzed patient. These require monitoring and attention to prevent them and prompt treatment if indicated.

PROMOTING HOME AND COMMUNITY-BASED CARE

Teaching Patients Self-Care

Patients with Guillain-Barré syndrome and their families are usually frightened by the sudden onset of life-threatening symptoms and their severity. Therefore, teaching the patient and family about the disorder and its generally favorable prognosis is important (Chart 64-5). During the acute phase of the illness, the patient and family are instructed about strategies they can implement to minimize the effects of immobility and other complications. As function begins to return, family members and other home care providers are instructed about care of the patient and their role in the rehabilitation process. Preparation for discharge is an interdisciplinary effort requiring family or caregiver education by all team members, including the nurse, physician, occupational and physical therapists, speech therapist, and respiratory therapist.

Continuing Care

Most patients with Guillain-Barré syndrome experience complete recovery. Patients who have experienced total or prolonged paralysis require intensive rehabilitation; the extent depends on the patient’s needs. Approaches include a comprehensive inpatient program if deficits are significant, an outpatient program if the patient can travel by car, or a home program of physical and occupational therapy. The recovery phase may be long and will require patience as well as involvement on the part of the patient and family.

During acute care the focus is on obvious needs, issues, and deficits. The nurse needs to remind or instruct patients and family members of the need for continuing health promotion and screening practices following this initial phase of care.

Evaluation

EXPECTED PATIENT OUTCOMES

Expected patient outcomes may include:

1. Maintains effective respirations and airway clearance
   a. Has normal breath sounds on auscultation
   b. Demonstrates gradual improvement in respiratory function

2. Shows increasing mobility
   a. Regains use of extremities
   b. Participates in rehabilitation program
   c. Demonstrates no contractures and minimal muscle atrophy

3. Receives adequate nutrition and hydration
   a. Consumes diet adequate to meet nutritional needs
   b. Swallows without aspiration

4. Demonstrates recovery of speech
   a. Can communicate needs through alternative strategies
   b. Practices exercises recommended by the speech therapist

5. Shows lessening fear and anxiety

6. Absence of complications
   a. Breathes spontaneously
   b. Has vital capacity within normal range
   c. Exhibits normal arterial blood gases and oximetry

Chart 64-5

Home Care Checklist • The Patient With Guillain-Barré Syndrome

At the completion of the home care instruction, the patient or caregiver will be able to:

- Describe the disease process of Guillain-Barré syndrome.
- Manage respiratory needs: tracheostomy care, suctioning.
- Demonstrate proper body mechanics regarding lifting and transfers.
- Practice gait training and strength endurance.
- Perform range-of-motion exercises.
- Perform activities of daily living and manage self-care:
  - Nutrition
  - Bowel and bladder management
  - Skin care
  - Adaptive equipment for bathing, hygiene, grooming, dressing
- Operate and explain function of medical equipment and mobility aids: walkers, wheelchairs, bedside commodes, tub transfer benches, adaptive devices
- Use coping mechanisms and diversional activities appropriately.
- Implement safety measures in the home.
- Know how to contact and use community resources and the Guillain-Barré Syndrome Foundation International.
Cranial Nerve Disorders

Because the brain stem and cranial nerves involve vital motor, sensory, or autonomic functions of the body, these nerves may be affected by conditions arising primarily within these structures or in secondary extension from adjacent disease processes. The cranial nerves (Fig. 64-6) are examined separately and in sequence (see Chap. 60). Some cranial nerve deficits can be detected by observing the patient’s face, eye movements, speech, and swallowing. Electromyography (EMG) is used to investigate motor and sensory dysfunction. MRI is used to obtain images of the cranial nerves and brain stem. An overview of disorders that may affect each of the cranial nerves, including clinical manifestations and nursing interventions, is presented in Table 64-1. The following discussion centers on trigeminal neuralgia, a condition affecting the fifth cranial nerve, and Bell’s palsy, caused by involvement of the seventh cranial nerve. These are the most common disorders of the cranial nerves.

TRIGEMINAL NEURALGIA (TIC DOULOUREUX)

Trigeminal neuralgia is a condition of the fifth cranial nerve characterized by paroxysms of pain in the area innervated by any of the three branches, but it most commonly occurs in the second and third branches of the trigeminal nerve (Maloni, 2000) (Fig. 64-7). The pain ends as abruptly as it starts and is described as a unilateral shooting and stabbing sensation. The unilateral nature of the pain is an important diagnostic characteristic (Preul, 2001). Associated involuntary contraction of the facial muscles can cause sudden closing of the eye or a twitch of the mouth, hence the name tic douloureux (painful twitch). The cause is not certain, but chronic compression or irritation of the trigeminal nerve or degenerative changes in the gasserian ganglion are suggested causes. Vascular pressure from structural abnormalities (loop of an artery) encroaching on the trigeminal nerve, gasserian ganglion, or root entry zone has also been suggested as a cause.

Trigeminal neuralgia is 400 times more common in patients with MS than in the general population. The pain is more often cyclic and affects men with MS at a higher rate than women with MS (Maloni, 2000).

Early attacks, appearing most often in the fifth decade of life, are usually mild and brief. Pain-free intervals may be measured in terms of minutes, hours, days, or longer. With advancing years, the painful episodes tend to become more frequent and agonizing. The patient lives in constant fear of attacks.

Paroxysms can occur with any stimulation of the terminals of the affected nerve branches, such as washing the face, shaving, brushing the teeth, eating, and drinking. A draft of cold air and direct pressure against the nerve trunk may also cause pain. Certain areas are called trigger points because the slightest touch immediately starts a paroxysm or episode. To avoid stimulating these areas, patients with trigeminal neuralgia try not to touch or wash their faces, shave, chew, or do anything else that might cause an attack. These behaviors are a clue to diagnosis.

Medical Management

PHARMACOLOGIC THERAPY

Antiseizure agents, such as carbamazepine (Tegretol), relieve pain in most patients with trigeminal neuralgia by reducing the transmission of impulses at certain nerve terminals. Carbamazepine is taken with meals. Serum levels must be monitored to avoid toxicity in patients who require high doses to control the pain. Side effects include nausea, dizziness, drowsiness, and aplastic anemia. The patient is monitored for bone marrow depression during long-term therapy. Gabapentin (Neurontin) and baclofen (Lioresal)
**Table 64-1 • Disorders of Cranial Nerves**

<table>
<thead>
<tr>
<th>DISORDER</th>
<th>CLINICAL MANIFESTATIONS</th>
<th>NURSING INTERVENTIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Olfactory Nerve—I</strong></td>
<td>Unilateral or bilateral anosmia (temporary or persistent)</td>
<td>Assess sense of smell.</td>
</tr>
<tr>
<td>Head trauma</td>
<td>Diminished taste for food</td>
<td>Assess cerebrospinal fluid rhinorrhea if patient has sustained head trauma.</td>
</tr>
<tr>
<td>Intracranial tumor</td>
<td></td>
<td></td>
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<tr>
<td>Intracranial surgery</td>
<td></td>
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<tr>
<td><strong>Optic Nerve—II</strong></td>
<td>Lesions of optic tract producing homonymous hemianopsia</td>
<td>Assess visual acuity.</td>
</tr>
<tr>
<td>Optic neuritis</td>
<td></td>
<td>Restructure environment to prevent injuries.</td>
</tr>
<tr>
<td>Increased intracranial pressure</td>
<td></td>
<td>Teach patient to accommodate for visual loss.</td>
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<tr>
<td>Pituitary tumor</td>
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<tr>
<td><strong>Oculomotor Nerve—III</strong></td>
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<tr>
<td><strong>Trochlear Nerve—IV</strong></td>
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<tr>
<td><strong>Abducens Nerve—VI</strong></td>
<td></td>
<td></td>
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<tr>
<td>Vascular</td>
<td>Dilatation of pupil with loss of light reflex on one side</td>
<td>Assess extraocular movement and for non-reactive pupil.</td>
</tr>
<tr>
<td>Brain stem ischemia</td>
<td>Impairment of ocular movement</td>
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<tr>
<td>Hemorrhage and infarction</td>
<td>Diplopia</td>
<td></td>
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<tr>
<td>Neoplasm</td>
<td>Gaze palsies</td>
<td></td>
</tr>
<tr>
<td>Trauma</td>
<td>Ptosis of eyelid</td>
<td></td>
</tr>
<tr>
<td><strong>Trigeminal Nerve—V</strong></td>
<td>Pain in face</td>
<td></td>
</tr>
<tr>
<td>Trigeminal neuralgia</td>
<td>Diminished or loss of corneal reflex</td>
<td></td>
</tr>
<tr>
<td>Head trauma</td>
<td>Chewing dysfunction</td>
<td></td>
</tr>
<tr>
<td>Cerebellopontine lesion</td>
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<tr>
<td>Sinus tract tumor and metastatic disease</td>
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<tr>
<td>Compression of trigeminal root by tumor</td>
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<td></td>
</tr>
<tr>
<td><strong>Facial Nerve—VII</strong></td>
<td>Facial dysfunction; weakness and paralysis</td>
<td>Recognize facial paralysis as emergency; refer for treatment as soon as possible.</td>
</tr>
<tr>
<td>Bell’s palsy</td>
<td>Hemifacial spasm</td>
<td>Teach protective care for eyes.</td>
</tr>
<tr>
<td>Facial nerve tumor</td>
<td>Diminished or absent taste</td>
<td>Select easily chewed foods; patient should eat and drink from unaffected side of mouth.</td>
</tr>
<tr>
<td>Intracranial lesion</td>
<td>Pain</td>
<td>Emphasize importance of oral hygiene. Provide emotional support for changed appearance of face.</td>
</tr>
<tr>
<td>Herpes zoster</td>
<td></td>
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</tr>
<tr>
<td><strong>Acoustic Nerve—VIII</strong></td>
<td>Tinnitus</td>
<td>Assess pattern of vertigo.</td>
</tr>
<tr>
<td>Tumors and acoustic neuroma</td>
<td>Vertigo</td>
<td>Provide for safety measures to prevent falls.</td>
</tr>
<tr>
<td>Vascular compression of nerve</td>
<td>Hearing difficulties</td>
<td>Ensure that patient can obtain balance before ambulating.</td>
</tr>
<tr>
<td>Ménière’s syndrome</td>
<td></td>
<td>Caution patient to change positions slowly.</td>
</tr>
<tr>
<td><strong>Glossopharyngeal Nerve—IX</strong></td>
<td>Pain at base of tongue</td>
<td>Assist with ambulation.</td>
</tr>
<tr>
<td>Glossopharyngeal neuralgia from neurovascular compression of cranial nerves IX and X</td>
<td>Difficulty in swallowing</td>
<td>Encourage use of activity of daily living aids.</td>
</tr>
<tr>
<td>Trauma</td>
<td>Loss of gag reflex</td>
<td></td>
</tr>
<tr>
<td>Inflammatory conditions</td>
<td>Palatal, pharyngeal, and laryngeal paralysis</td>
<td></td>
</tr>
<tr>
<td>Tumor</td>
<td></td>
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<tr>
<td>Vertebral artery aneurysms</td>
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<tr>
<td><strong>Vagus Nerve—X</strong></td>
<td>Voice changes (temporary or permanent hoarseness)</td>
<td>Assess for paroxysmal pain in throat, decreased or absent swallowing, gag and cough reflexes.</td>
</tr>
<tr>
<td>Spastic palsy of larynx; bulbar paralysis; high vagal paralysis</td>
<td>Vocal paralysis</td>
<td>Monitor for dysphagia, aspiration, nasal dysarthric speech.</td>
</tr>
<tr>
<td>Guillain-Barré syndrome</td>
<td>Dysphagia</td>
<td>Position patient upright for eating or tube feeding.</td>
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<tr>
<td>Vagal body tumors</td>
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<tr>
<td>Nerve paralysis from malignancy, surgical trauma such as carotid endarterectomy</td>
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</tbody>
</table>

(continued)
Alcohol or phenol injection of the gasserian ganglion and peripheral branches of the trigeminal nerve relieves pain for several months. However, the pain returns with nerve regeneration.

**SURGICAL MANAGEMENT**

When these methods fail to relieve pain, a number of surgical options are available. The choice of procedure depends on the patient’s preference and health status.

**Microvascular Decompression of the Trigeminal Nerve.** An intracranial approach can be used to decompress the trigeminal nerve. The pain may be caused by vascular compression of the entry zone of the trigeminal root by an arterial loop and occasionally by a vein. With the aid of an operating microscope, the artery loop is lifted from the nerve to relieve the pressure, and a small prosthetic device is inserted to prevent recurrence of impingement on the nerve. This procedure relieves facial pain while preserving normal sensation, but it is a major procedure, involving a craniotomy. The postoperative management is the same as for other intracranial surgeries (see Chap. 61).

**Percutaneous Radiofrequency Trigeminal Gangliolysis.** Percutaneous radiofrequency interruption of the gasserian ganglion, in which the small unmyelinated and thinly myelinated fibers that conduct pain are thermally destroyed, is the surgical procedure of choice for trigeminal neuralgia (Tronnier, Rasche, Hamer et al., 2001). Use of stereotactic MRI for identification of the trigeminal nerve followed by gamma knife radiosurgery is being used at some centers with good results (Maesawa et al., 2001).

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### Table 64-1 • Disorders of Cranial Nerves (Continued)

<table>
<thead>
<tr>
<th>DISORDER</th>
<th>CLINICAL MANIFESTATIONS</th>
<th>NURSING INTERVENTIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Spinal Accessory Nerve—XI</strong></td>
<td>Drooping of affected shoulder with limited shoulder movement Weakness or paralysis of head rotation, flexion, extension; shoulder elevation</td>
<td>Support patient undergoing diagnostic tests.</td>
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<tr>
<td>Spinal cord disorder</td>
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<td>Amyotrophic lateral sclerosis</td>
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<tr>
<td>Trauma</td>
<td></td>
<td></td>
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<tr>
<td>Guillain-Barre syndrome</td>
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</tr>
<tr>
<td><strong>Hypoglossal Nerve—XII</strong></td>
<td>Abnormal movements of tongue Weakness or paralysis of tongue muscles Difficulty in talking, chewing, and swallowing</td>
<td>Observe swallowing ability. Observe speech pattern. Be aware of swallowing or vocal difficulties. Prepare for alternate feeding methods (tube feeding) to maintain nutrition.</td>
</tr>
<tr>
<td>Medullary lesions</td>
<td></td>
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<tr>
<td>Amyotrophic lateral sclerosis</td>
<td></td>
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</tr>
<tr>
<td>Polio and motor system disease, which may destroy hypoglossal nuclei</td>
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<td></td>
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<tr>
<td>Multiple sclerosis</td>
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<td></td>
</tr>
<tr>
<td>Trauma</td>
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</tbody>
</table>

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![Figure 64-7](image) Distribution of trigeminal nerve branches.
Under local anesthesia, the needle is introduced through the cheek on the affected side. Under fluoroscopic guidance, the needle electrode is guided through the foramen magnum into the gasserian ganglion. The divisions of the gasserian ganglion (mandibular, maxillary, and ophthalmic) are encountered sequentially. The nerve is stimulated with a small current while the patient is awake. The patient reports when a tingling sensation is felt. When the electrode needle is in the desired position, the patient is anesthetized briefly and a radiofrequency current (heating current to destroy the nerve) is passed in a controlled manner to injure the trigeminal ganglion and rootlets thermally. The patient is then awakened from the anesthesia and examined for sensory deficits. This is repeated until the desired effect is achieved. The procedure takes less than 1 hour and provides permanent pain relief in most patients. Touch and proprioceptive functions are left intact.

In the patient with trigeminal neuralgia and MS who is refractory to medical pain management, the surgical treatment of choice is trigeminal rhizotomy (Maloni, 2000). See Chapter 13 for care of the patient following a rhizotomy.

**Nursing Management**

**PREVENTING PAIN**

Preoperative management of a patient with trigeminal neuralgia occurs mostly on an outpatient basis and includes recognizing factors that may aggravate excruciating facial pain, such as food that is too hot or too cold or jarring the patient’s bed or chair. Even washing the face, combing the hair, or brushing the teeth may produce acute pain. The nurse can assist the patient in preventing or reducing this pain by providing instructions about preventive strategies. Providing cotton pads and room-temperature water for washing the face, instructing the patient to rinse with mouthwash after eating when tooth-brushing causes pain, and performing personal hygiene during pain-free intervals are all effective strategies. The patient is instructed to take food and fluids at room temperature, to chew on the unaffected side, and to ingest soft foods. The nurse recognizes that anxiety, depression, and insomnia often accompany chronic painful conditions and uses appropriate interventions and referrals. (See Chap. 13 for management of patients with chronic pain.)

**PROVIDING POSTOPERATIVE CARE**

Postoperative neurologic assessments are conducted to evaluate the patient for facial motor and sensory deficits in each of the three branches of the trigeminal nerve. If the surgery results in sensory deficits to the affected side of the face, the patient is instructed not to rub the eye, because pain will not be felt if there is injury. The eye is assessed for irritation or redness. Artificial tears may be prescribed to prevent dryness in the affected eye. The patient is cautioned not to chew on the affected side until numbness has diminished. The patient is observed carefully for any difficulty in eating and swallowing foods of different consistency.

**BELL’S PALSY**

Bell’s palsy (facial paralysis) is due to unilateral inflammation of the seventh cranial nerve, which results in weakness or paralysis of the facial muscles on the affected side (Fig. 64-8). The cause is unknown, although possible causes may include vascular ischemia, viral disease (herpes simplex, herpes zoster), autoimmune disease, or a combination of all of these factors. The incidence is 13 to 34 cases per 100,000; it increases with age and among pregnant women in the third trimester (Campbell & Brundage, 2002; Shmorgun, Chan & Ray, 2002).

Bell’s palsy is considered by some to represent a type of pressure paralysis. The inflamed, edematous nerve becomes compressed to the point of damage, or its nutrient vessel is occluded, producing ischemic necrosis of the nerve. There is distortion of the face from paralysis of the facial muscles; increased lacrimation (tearing); and painful sensations in the face, behind the ear, and in the eye. The patient may experience speech difficulties and may be unable to eat on the affected side because of weakness or paralysis of the facial muscles.

**Management**

The objectives of treatment are to maintain the muscle tone of the face and to prevent or minimize denervation. The patient should be reassured that no stroke has occurred and that spontaneous recovery occurs within 3 to 5 weeks in most patients.

Corticosteroid therapy (prednisone) may be prescribed to reduce inflammation and edema; this reduces vascular compression and permits restoration of blood circulation to the nerve. Early administration of corticosteroid therapy appears to diminish the severity of the disease, relieve the pain, and prevent or minimize denervation.
Facial pain is controlled with analgesic agents. Heat may be applied to the involved side of the face to promote comfort and blood flow through the muscles.

Electrical stimulation may be applied to the face to prevent muscle atrophy. Although most patients recover with conservative treatment, surgical exploration of the facial nerve may be indicated in patients who are suspected of having a tumor or for surgical decompression of the facial nerve and for surgical treatment of a paralyzed face.

**PROMOTING HOME AND COMMUNITY-BASED CARE**

**Teaching Patients Self-Care.** While the paralysis lasts, the involved eye must be protected. Frequently, the eye does not close completely and the blink reflex is diminished, so the eye is vulnerable to dust and foreign particles. Corneal irritation and ulceration may occur if the eye is unprotected. Distortion of the lower lid alters the proper drainage of tears. To prevent injury, the eye should be covered with a protective shield at night. The eye patch may abrade the cornea, however, because there is some difficulty in keeping the partially paralyzed eyelids closed. The application of eye ointment at bedtime causes the eyelids to adhere to one another and remain closed during sleep. The patient can be taught to close the paralyzed eyelid manually before going to sleep. Wrap-around sunglasses or goggles may be worn to decrease normal evaporation from the eye.

**Continuing Care.** When the sensitivity of the nerve to touch decreases and the patient can tolerate touching the face, the nurse can suggest massaging the face several times daily, using a gentle upward motion, to maintain muscle tone. Facial exercises, such as wrinkling the forehead, blowing out the cheeks, and whistling, may be performed with the aid of a mirror in an effort to prevent muscle atrophy. Exposure of the face to cold and drafts is avoided.

### Disorders of the Peripheral Nervous System

#### PERIPHERAL NEUROPATHIES

A peripheral neuropathy (disorder of the nervous system) is a disorder affecting the peripheral motor, sensory, or autonomic nerves. Peripheral nerves connect the spinal cord and brain to all other organs. They transmit motor impulses from the brain and relay sensory impulses to the brain. A mononeuropathy affects a single peripheral nerve; multiple mononeuropathy or mononeuritis multiplex indicates the involvement of multiple single peripheral nerves or their branches. Polyneuropathies are characterized by bilateral and symmetric disturbance of function, usually beginning in the feet and hands. (Most nutritional, metabolic, and toxic neuropathies take this form.)

The most common causes of peripheral neuropathy are diabetes, alcoholism, and occlusive vascular disease. These disorders result in hypoxia or atrophy of the peripheral nerve. Many bacterial and metabolic toxins and exogenous poisons also cause peripheral neuropathy. Because of the growing use of chemicals in industry, agriculture, and medicine, the number of substances causing peripheral neuropathies and the incidence of peripheral neuropathies have increased. In developing countries, leprosy is a major cause of severe nerve disease because *Mycobacterium leprae* invade the peripheral nervous system.

The major symptoms of peripheral nerve disorders are loss of sensation, muscle atrophy, weakness, diminished reflexes, pain, and paresthesia (numbness, tingling) of the extremities. The patient frequently describes some part of the extremity as numb. Autonomic features include decreased or absent sweating, orthostatic hypotension, nocturnal diarrhea, tachycardia, impotence, and atrophic skin and nail changes.

Peripheral nerve disorders are diagnosed by history, physical examination, EMG, and somatosensory evoked potentials.

#### MONONEUROPATHY

Mononeuropathy is limited to a single peripheral nerve and its branches. It arises when the trunk of the nerve is compressed or entrapped (as in carpal tunnel syndrome); traumatized, as when bruised by a blow, or overstretched, as in joint dislocation; punctured by a needle used to inject a drug or damaged by the drugs thus injected; or inflamed because an adjacent infectious process extends to the nerve trunk. Mononeuropathy frequently is seen in patients with diabetes.

Pain is seldom a major symptom of mononeuropathy when the condition is due to trauma, but in patients with complicating inflammatory conditions such as arthritis, pain is prominent. Pain is increased by all body movements that tend to stretch, strain, or cause pressure on the injured nerve and by sudden jarring of the body (eg, coughing and sneezing). The skin in the areas supplied by nerves that are injured or diseased may become reddened and glossy; the subcutaneous tissue may become edematous, and the nails and hair in this area become defective. Chemical injuries to a nerve trunk, such as those caused by drugs injected into or near it, are often permanent.

The objective of treatment of mononeuropathy is to remove the cause, if possible, such as by freeing the compressed nerve. Local corticosteroid injections may reduce inflammation and the pressure on the nerve. Aspirin or codeine may be used to relieve pain.

**Critical Thinking Exercises**

1. A 19-year-old college student is suspected of having meningococcal meningitis. Identify two neurologic changes that may reflect increased ICP. What interventions would be included in your plan of care to protect the patient from injury? The patient’s family has many questions about the disease and their risk of contracting meningitis. Develop a teaching plan that would describe meningococcal meningitis and prophylactic therapy for the patient’s family and close contacts.

2. Your patient has been receiving one of the injectable interferon medications for the treatment of MS for about 6 months. She reports that she is becoming very discouraged because the medication does not seem to improve her symptoms. In addition, she reports that she has developed skin lesions at the injection sites. Identify the areas of assessment that are of priority at this time. Describe nursing interventions, including teaching and referral, that you would consider at this time. Provide the rationale for your interventions.

3. Your patient has been admitted to the hospital with a diagnosis of possible Guillain-Barré. Identify the priorities of assessment for this patient and the nursing and medical interventions that you would anticipate.

4. Your patient has myasthenia gravis. Although she reports that her condition has been stable and that she has been able...
to manage without assistance, she reports increasing weakness and fatigue. What nursing assessments and interventions are warranted for her? What discharge plans are indicated if she is to return home to the care of her family? How would your discharge planning change if she lives alone in an apartment on the second floor of a building without an elevator?

5. A 52-year-old man has just been diagnosed with trigeminal neuralgia. Carbamazepine has been prescribed for pain management. Develop a teaching plan that explains the nature of trigeminal neuralgia, the side effects of carbamazepine, and measures he can take to avoid triggering a painful episode. How would you evaluate the outcome of your teaching plan?

REFERENCES AND SELECTED READINGS

Books

Journals
Asterisks indicate nursing research articles.

CNS Infections

Creutzfeldt-Jakob Disease

Guillain-Barré Syndrome

Multiple Sclerosis


**Myasthenia Gravis**


**Trigeminal Neuralgia and Neuropathies**


**RESOURCES AND WEBSITES**


Myasthenia Gravis Foundation of America, 222 S. Riverside Plaza, Suite 1540, Chicago, IL 60606; (800) 541-5454; (312) 258-0522; fax: 312-258-0461; http://www.myasthenia.org.

National Multiple Sclerosis Society, 733 Third Avenue, New York, NY 10017; (800) 344-4867; http://www.nmss.org.