Since first thoroughly described by Charcot in 1868, multiple sclerosis (MS) has been viewed as a chronic, frequently debilitating neurologic disease that affects young adults in the prime of their lives. Diagnosis was tedious and difficult. Those affected faced a bleak future. During the latter part of the 20th century, technological advances such as diagnostic imaging (MRI) and neurophysiologic testing (evoked potentials) facilitated the diagnosis of multiple sclerosis and led to more prompt attention and treatment by healthcare providers. This is in direct contrast to care in the early and mid-20th century that was frequently referred to as the “diagnose and adios” era (Labe Scheinberg, personal communication). The treatment of MS has subsequently advanced from a focus on episodic, acute care and rudimentary symptom management, to disease modification and the potential for altering the natural history of what was previously thought to be an unbroken path to severe disability. In addition, a greater understanding and acceptance of the value of psychosocial interventions and rehabilitative services has emerged as a leitmotif of MS care throughout the world.

With its emphasis on acute patient care needs, technological and surgical advances, and primary care interventions, basic and advanced nursing education traditionally has not stressed care of the chronically ill or disabled. The complex nature of MS and its lifelong problems require the skills and direct services of numerous healthcare professionals in addition to neurologists. Nurses who enter this field are often confronted with infor-
Epidemiology, Incidence, and Prevalence

It is estimated that between 250,000 to 350,000 people in the United States are afflicted with MS (Anderson et al., 1992; Vollmer, 1999; Burks & Johnson, 2001). The disease is more common in women than in men by a ratio greater than 2:1, with the difference most marked at younger ages (Kurtzke, 1993; Ebers, 1998).

MS appears to have a higher prevalence in Caucasians than other racial groups. A study done in Hawaii (Alter et al., 1971) showed a higher prevalence in Caucasians than in Asians, native Hawaiians, or blacks living in the same geographic area. Japanese prevalence rates are the lowest of all industrialized countries (Kuroiwa et al., 1983), and the disease is very rare among African blacks (Poser, 1994). The prevalence of MS in those of Scandinavian descent is high, and Bulman and Ebers (1992) showed that the high prevalence of MS in the northern tier of the United States was closely related to the high proportion of people of Scandinavian descent who live there (Ebers, 1998).

It is now believed that MS is the result of an interaction of both genetic and environmental factors (Sadovnick, 1994). It has been theorized that MS susceptibility is under the control of several genes. By inheriting these genes, a person is susceptible to an immunologic stimulus (possibly a virus) that in turn leads to myelin damage and clinical multiple sclerosis (Sadovnick, 1994). The specific genes and how they interact have yet to be identified. For this reason, the interpretation of the data on geographic distribution of MS is difficult (Sadovnick, 1994) and remains a topic of controversy.
There appears to be a markedly uneven geographic distribution of the disease. Kurtzke (1985) identified areas of high, medium, and low risk, according to latitude. In the United States, states south of the 37th parallel of north latitude showed lower death rates than those north of that line, which were well above the national mean. Prevalence studies for groups of Northern Europeans and North Americans who migrated from high-risk areas to low-risk areas (Moffie, 1996; Dean, 1967; Alter et al., 1971) showed that they remained at high risk if they emigrated after the age of fifteen. Those who emigrated prior to age fifteen acquired the low risk of the countries to which they emigrated. Such differences have given rise to a hypothesis that there is a critical age of exposure to unknown causal or triggering factors (possibly a virus), and suggest that there is a long period of latency between exposure and onset of the disease (Garnieri et al., 1993; Ebers, 1998).

This geographic model of distribution has been criticized on the basis that the comparison of prevalence rates was “reported from very different areas, countries, and communities at different times” (Garnieri et al., 1993, p. S17). Garnieri also pointed out that more recent European prevalence studies have contradicted earlier studies. A study in Italy showed that prevalence was actually higher in the southern islands of Sicily and Sardinia (Savettieri, 1983; Rosati, 1990), with similar results from studies done in Yugoslavia (Sepper et al., 1989) and Spain (Martin et al., 1988). According to Garnieri (1993), the “lower MS prevalence ascribed to a lower latitude may in part reflect differences in level, quality, and organization of health services as well as accessibility and case ascertainment, variables that affect the accuracy of the reports and produce bias in prevalence estimates” (p. S17). Studies done in Australia, however, support the correlation with latitude, showing higher prevalence rates in the southern regions, where it is cooler, with no significant differences in ethnic composition that might account for this difference (Hammond et al., 1988).

It has also been shown that the prevalence and pattern of MS can vary over time within a given geographic area. One notable study was done in the Faroe Islands, which experienced a dramatic increase in cases of MS after the arrival of British troops during World War II (Kurtzke & Hyllested, 1979). Kurtzke’s interpretation of this increase was that the British introduced MS to the Faroe Islands, stating that “the only possible explanations are that the British brought either a persistent toxin or a transmissible infection. A toxin cannot explain successive epidemics. Therefore, the cause of MS in the Faroes is a transmissible infection” and that “MS exists in a widespread, transmissible, but neurologically asymptomatic form” (Kurtzke, 1993, p. 412). Kurtzke went on to say that this asymptomatic form, which he calls “primary MS affection (PMSA)” is common in a population in which there is MS, but that it only rarely produces clinical MS symptoms.
It should be pointed out that there is no evidence that MS is a directly transmissible disease since it has not been demonstrated that people who live with, or have frequent contact with, MS patients are at greater risk for the disease; i.e., there is no reason to believe that MS patients are infectious (Granieri, 1993) (Table 1-1).

THE PATHOLOGY AND ETIOLOGY OF MULTIPLE SCLEROSIS

MS is a disease of the central nervous system (CNS) in which the myelin sheath surrounding certain nerve fibers becomes damaged, interrupting the conduction of nerve impulses. The pathologic process begins with the destruction of the myelin, which may slow or interrupt conduction (Allen, 1991). Irregularly shaped macroscopic lesions, which appear to be the result of destruction of the myelin sheath, are scattered throughout the CNS. These lesions, or *plaques*, are found in the white matter and have a predilection for the optic nerves and the white matter of the spinal cord, brainstem, cerebellum, and cerebrum, especially the area surrounding the ventricles (Pallett & O'Brien, 1985). Recently formed lesions show partial or complete degeneration of myelin and perivascular infiltration with lymphocytes and other mononuclear cells, suggestive of an inflammatory process (Pallett & O'Brien, 1985). Axonal damage has been identified in both acute and chronic MS lesions and is believed to be the cause of permanent, irreversible physical and cognitive disability (Trapp et al, 1998).

The etiology of MS is not known although there is believed to be a genetic predisposition in susceptible individuals combined with an unknown environmental trigger (Compson, 1991). The environmental factor is not known but is thought to be viral in origin (Compson, 1991). MS has been shown to occur frequently in specific families, and current theory is that it is multigenic; i.e., results from more than one gene (Compson, 1991; Compson, 2001).

<table>
<thead>
<tr>
<th>TABLE 1-1. Familial Risks for MS</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Up to 2 percent of people with MS have at least one relative with MS</td>
</tr>
</tbody>
</table>

**Empiric recurrence risks (age-adjusted)**

- MS parent-risk for child 4%
- MS person-risk for sibling 4–5%
- MS twin with MS risk for co-twin (fraternal) 3–5%
- MS twin with MS risk for co-twin (identical) 26–26%

*Source: Vollmer, 1999; Sadovnick et al., 1993*
THE COURSE OF THE DISEASE

One of the hallmarks of MS is its unpredictability from person to person and within a given individual over time. Its prognosis is uncertain although there are general prognostic indicators that can suggest whether or not a patient’s disease will follow a specific pattern. In general, women have a better prognosis than men do (Coyle, 1996). Onset at an early age, a monoregional versus a polyregional attack, and complete recovery from an exacerbation portend a favorable prognosis (Coyle, 1996). Brainstem symptoms (such as nystagmus, tremor, ataxia, and dysthria), poor recovery from exacerbations, and frequent attack rate are indicators of a poor prognosis (Coyle, 1996).

The diagnosis of MS is usually made by a neurologist after two or more episodes of unexplained neurologic symptoms have occurred. Recent research studies have indicated that clinically isolated syndromes (CIS)—one exacerbation and positive MRI findings—are the first indication of MS. There is no specific laboratory test for MS, and the diagnosis usually depends on a history that indicates the probability of the disease, a neurologic examination with findings consistent with MS, and positive paraclinical evidence (Sibley, 1990). Most patients fall into the age group of 15–60 years although recent data suggest that as many as 10 percent of MS patients have their first symptoms in their 60s (Sibley, 1990). While the average age of onset is between 20 and 50 years, the disease may start in children. Initial symptoms include numbness, tingling, or weakness of the extremities, visual changes, vertigo, dysthria, ataxia, and urinary frequency and urgency. L’Hermitte’s phenomenon, a transient paresthesia resembling an electrical shock that occurs with forward flexion of the neck, is also common (Sibley, 1990). The diagnostic criteria for MS have evolved from Schumacher et al. (1965), the Poser Committee (1983), and the McDonald consensus (2001) (Tables 1-2, 1-3, and 1-4).

<table>
<thead>
<tr>
<th>Table 1-2. Schumacher Criteria for the Clinical Diagnosis of MS</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Appropriate age (10–50 years)</td>
</tr>
<tr>
<td>• CNS white matter disease</td>
</tr>
<tr>
<td>• Lesions disseminated in time and space</td>
</tr>
<tr>
<td>• Two or more separate lesions</td>
</tr>
<tr>
<td>• Objective abnormalities</td>
</tr>
<tr>
<td>• Consistent time course</td>
</tr>
<tr>
<td>• Attacks lasting more than 24 hours, spaced 1 month apart</td>
</tr>
<tr>
<td>• Slow, stepwise progression for more than 6 months</td>
</tr>
<tr>
<td>• No better explanation</td>
</tr>
<tr>
<td>• Minimum routine laboratory investigation</td>
</tr>
<tr>
<td>• Diagnosis by a physician competent in clinical neurology</td>
</tr>
</tbody>
</table>
TABLE 1-3. Poser Committee Criteria for the Diagnosis of MS

<table>
<thead>
<tr>
<th>Category</th>
<th>Clinical</th>
<th>Paraclinical</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Attacks</td>
<td>Evidence</td>
</tr>
<tr>
<td>Clinical Diagnosis:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Definite</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Probable</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>1</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Laboratory-Supported Diagnosis:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Definite</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Category</th>
<th>Clinical</th>
<th>Paraclinical</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Attacks</td>
<td>Evidence</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Evidence</td>
</tr>
<tr>
<td></td>
<td></td>
<td>CSF</td>
</tr>
</tbody>
</table>

TABLE 1-4. New MS Diagnostic Criteria

<table>
<thead>
<tr>
<th>Clinical Attacks</th>
<th>Objective Lesions</th>
<th>Additional Requirements to Make the Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 or more</td>
<td>2 or more</td>
<td>None; clinical evidence will suffice (additional evidence desirable but must be consistent with MS)</td>
</tr>
<tr>
<td>2 or more</td>
<td>1</td>
<td>Dissemination in space by MRI or positive CSF and 2 or more MRI lesions consistent with MS or further clinical attack involving different site</td>
</tr>
<tr>
<td>1 (monosymptomatic)</td>
<td>1</td>
<td>Dissemination in space by MRI or positive CSF and 2 or more MRI lesions consistent with MS and Dissemination in time by MRI or second clinical attach</td>
</tr>
<tr>
<td>0 Progression</td>
<td>Positive CSF</td>
<td>and</td>
</tr>
<tr>
<td>from outset</td>
<td>1</td>
<td>Dissemination in space by MRI evidence of 9 or more T2 brain lesions or 2 or more cord lesions or 4–8 MRI lesions or positive VEP with less than 4 brain lesions plus 1 cord lesions and Dissemination in TIME by MRI or continued progression for 1 year</td>
</tr>
</tbody>
</table>

MS is diagnosed mainly on a clinical basis due to the difficulty of obtaining appropriate tissue (Fang & Lublin, 1995). The Poser criteria for clinically definite MS require two clinical deficits referable to white matter lesions or a physician’s observation of one deficit with paraclinical evidence of another, either through MRI or evoked potentials. Deficits must be separated in onset by at least 1 month and each must last at least 24 hours (Poser et al., 1983). The somewhat controversial McDonald criteria incorporate increased understanding of MRI parameters and more precisely define the number and location of MRI lesions required for diagnosis. Additionally, no more appropriate diagnoses should exist to account for clinical findings.

Patients can be categorized as having either relapsing–remitting, primary progressive, secondary progressive, progressive–relapsing, benign, or malignant disease (Lublin & Reingold, 1996) (Figure 1-1).

Relapsing–remitting MS is characterized by clearly defined disease relapses with full recovery, and periods between relapses characterized by a lack of disease progression.

Patients with primary progressive MS demonstrate disease progression from the onset with occasional and temporary minor episodes of improvement.

Secondary progressive MS begins with a relapsing–remitting course followed by progression with or without occasional relapses, minor remissions, and plateaus.

Progressive–relapsing MS is progressive disease from the onset, but there are clear acute relapses with or without recovery. The periods between relapses are marked by continued progression.

Benign MS allows patients to remain fully functional in all neurologic systems 15 years after disease onset.

Malignant MS (Marburg’s variant) has a rapid progressive course leading to significant disability or death within a relatively short time after onset (Lublin & Reingold, 1996).

**DOMAINS OF MULTIPLE SCLEROSIS NURSING PRACTICE**

The practice of MS nursing requires that the full range of nursing skills and practice be called into use to serve the MS patient and the family (Maloni, 2001). MS nursing encompasses broad areas of accountability and practice, and includes specific knowledge, skills, and tasks. The domains of MS nursing have been defined as clinical practice, advocacy, education, and research (MSNICB, 2001).
Universal tasks of all nursing care are the establishment of a therapeutic partnership, the performance of a comprehensive assessment, the formulation of a collaborative treatment plan and its implementation, and the assessment of outcomes. Specific requirements in MS nursing care involve full knowledge of the disease and its range of physical, functional, and neurologic implications. Other requirements include skills to teach and empower patients and their families, as well as the ability to advocate for treatments, programs, and services needed by individuals and families affected by MS (Maloni, 2001). The late 20th and early 21st centuries have
seen a change in MS care—and a new recognition of the role of the MS nurse and others who provide nursing services. Nursing care in MS requires creativity, caring, empathy, hope, and a great deal of stamina (Halper, 2001).

**THE WELLNESS MODEL IN MULTIPLE SCLEROSIS NURSING**

The variable pattern of MS—along with the uncertainty and loss of control that the diagnosis brings to the patient and family—impels the nurse to respond with cultural sensitivity and individualized care. Positive approaches to restore control and quality of life tend to reassure anxious patients and their families as they face an unknown future. Nursing care can be divided into direct delivery of services, counseling, education, and support through difficult transitional periods; i.e., following the diagnosis, during an acute exacerbation, during worsening of the disease, or while learning to live with advanced disability. Clark’s wellness model has implications for the nursing process in MS (Clark, 1986) (Table 1-5). In a disease with no cure, the patient and family must assume ongoing responsibility for health care and self-monitoring. In the traditional nursing model, the nurse performs and the patient receives care. The wellness model is a collaboration between the patient and the nurse—a therapeutic partnership whose goal is self-awareness and self-responsibility. Clark defines wellness as a positive striving unique to the individual, in which a person can be ill and still have wellness with a deep appreciation for the joy of living and with a life purpose (Clark, 1986). This wellness focus strengthens and sustains the thera-

| **TABLE 1-5.** Comparison of the Traditional Nursing and Wellness Nursing Processes |
|-------------------------------------------------|-------------------------------------------------|
| **Traditional Nursing Process**                | **Wellness Nursing Process**                    |
| Assess client                                  | Model integrates whole person wellness for the client |
|                                               | Teach client self-assessment procedures          |
| Diagnose                                       | Assess unique learning needs based on client belief systems |
| Set goals                                      | Teach client to set meaningful wellness goals    |
| Develop nursing care plan                      | Develop plan of action with client               |
|                                               | and help client take responsibility for carrying it out |
| Carry out nursing interventions                | Teach client self-care and self-healing measures consistent with client beliefs |
| Evaluate results                               | Teach client to self-evaluate results            |

Source: Clark, 1986.
po
timp
cade
pend
saf
pat
bo
nurs
co
R
reassurance
infection
an
vided
time
assistance
the
acquisition
of
community
ser
require
Patients
of
peutic
partnership
and
can
support
this
relationship
throughout
a
lifetime
of
coping
with
change
and
adapting
to
new
circumstances
(Halper
&
Holland,
1998;
Halper,
2001).

CARE PATTERNS IN MULTIPLE SCLEROSIS—
WHERE NURSES DO NURSING

Acute Care Settings
Patients
with
MS
often
experience
exacerbations
of
their
illness
and
require
acute
interventions
with
corticosteroids,
rehabilitation
services,
and
assistance
with
the
acquisition
of
community
services.
Care
during
this
time
is
usually
limited
to
the
period
of
clinical
worsening
and
may
be
pro-
vided
in
a
hospital
setting,
rehabilitation
facility,
or
at
home.
Treatment
of
an
acute
exacerbation
usually
begins
with
screening
for
an
underlying
infection
(URI,
UTI)
or
an
environmental
trigger
(heat,
humidity).
The
nurse’s
role
at
this
time
is
one
of
direct
patient
care,
education,
counseling,
reassurance,
and
support.

REHABILITATIVE CARE

Rehabilitation
in
MS
can
be
provided
in
an
outpatient
setting,
an
inpa-
tient
unit,
or
at
home.
During
this
period,
nurses
provide
education
and
counseling
to
promote
adaptation
to
a
new
level
of
dunction.
In
addition,
nursing
may
involve
functions
such
as
wound
care;
pulmonary
care;
toilet,
bowel
and
bladder
management;
and
discharge
planning.
Physical,
oc-
cupational,
speech
and
language
therapy
and
vocational
rehabilitation
are
designed
to
help
patients
regain
lost
dunction,
perform
at
a
maximal
and
safe
level,
and
accept
and
adjust
to
periods
of
dependence
and
interde-
pendence
as
the
disease
waxes
and
wanes
(see
Chapter
3).

COMPREHENSIVE CARE

Comprehensive
care
in
MS
is
an
organized
system
of
health
care
designed
to
address
the
medical,
social,
vocational,
emotional,
and
edu-
cational
needs
of
patients
and
their
families
(Halper
&
Burks,
1994).
Comprehensive
care
embraces
a
philosophy
of
empowerment—a
well-
ness
approach
in
which
the
patient
takes
an
active
role
in
planning
and
implementing
health
care
and
self-care
activities
and
acts
as
a
consultant
to
the
team,
which
may
consist
of
physicians,
nurses,
rehabilitation
spe-
cialists, counselors (social workers, psychologists, neuropsychologists), educators, and clergy. Patients must learn to adapt and change in response to alterations in their physical and cognitive functioning. This implies a total commitment by the health care team and the patient to a clearly defined program of “wellness” that looks beyond impairments to each person’s potential (Cobble & Burks, 1985). Comprehensive care centers have proliferated throughout the United States and Canada since the early 1980s. The team approach to MS care has become universally accepted (Halper & Burks, 1994) and has set a standard for care of the chronically ill and disabled that one sees in disease such as HIV/AIDS, cancer, and coronary artery disease. A team leader, a case manager, or a case coordinator (Ignatavicius & Hausman, 1995) may coordinate patient care services provided by the interdisciplinary team. A nurse frequently assumes this role.

HOME CARE

Patients with more advanced disease are frequently cared for in their homes. Many families are faced with day-to-day challenges of caring for loved ones who require total care including personal hygiene, nutrition, transfers, and access to the healthcare system. The nurse in the home care setting is the link to healthcare providers in the community, and is often faced with challenges such as advocacy on behalf of the patient to increase services available in the home and to guarantee a basic level of medical attention (see Chapter 5). Patients who may be severely disabled with no hope of any improvement, may also face medical complications such as pneumonia, skin breakdown, and pulmonary compromise. These patients may request and receive comfort measures in the palliative care model.

LONG-TERM CARE

Long-term care does not necessarily imply nursing home or long-term care facility. It also may mean residing in housing for the disabled, participation in adult day programs, and sustained care within the community. Nurses have a vital role in the long-term care of people with MS: In nursing homes as care providers and supervisors of bedside care; in assisted living programs as monitors of health, function, wellness, and safety; in adult day programs, as part of the team supporting day-to-day activities; and in community centers as educators and advocates for patients and families (see Chapter 5).
MEETING THE CHALLENGES OF MULTIPLE SCLEROSIS NURSING

The challenge of nursing care in MS impels the nurse to answer the following questions when entering into the therapeutic partnership:

- What is the patient’s disease course?
- What treatment(s) have been prescribed?
- How has MS affected the patient’s quality of life?
- What is the patient’s history of adherence?
- How are symptoms interfering with the patient’s functional status?
- How has MS impacted the patient and the family?
- What are the available resources?
- How can I help?

We hope that this book will assist nurses in this wide variety of nursing settings to develop their own philosophy of care, to sustain themselves as MS nurses, and to transfer this model of nursing to other chronic illnesses requiring similar types of interventions.

REFERENCES


Maloni, H. Domains of Multiple Sclerosis Nursing Practice. Presentation, November 2001. Dallas, TX.


