



MODULE 2

# Clinical Presentation

English Version



## CME Module Title Clinical Presentation

### Learning Objectives

After completing this activity, the participant should be better able to:

- Explain what is meant by the term “clinically isolated syndrome”.
- Identify the various types of MS and their natural history and progression.
- Describe the variability of the clinical course of MS and resulting disability.
- Determine the nature and impact of some of the more common symptoms found in people with MS.
- Outline approaches used to identify some of these symptoms.
- Describe the features of an MS relapse.
- Identify key questions to ask a person with MS who you suspect is experiencing a relapse.
- Assist a person with MS to differentiate between fluctuation in symptoms, relapse and potential infection, and to understand trigger factors.
- Outline the differences between adult-onset and early-onset MS.
- Define late-onset MS.
- Describe other less common variations of MS.
- Specify disease progression factors and symptoms that are predictive of long-term outcomes.

### Target Audience

This activity has been developed to meet the educational needs of nurses who have an interest in optimising the management of people with MS.

### Accreditation

This e-learning training curriculum is accredited by the Royal College of Nursing Accreditation for the award of continuing professional development credits.

This continuing education activity has been approved by the International Council of Nurses (ICN) for the award of International Continuing Nursing Education Credits (ICNECs).

### Credit Designation

The Royal College of Nursing and the International Council of Nursing designates this module of the e-learning training curriculum for a maximum of 5 credits. On completion of the course (i.e. all 5 modules) you will be able to download a Virtual College certificate.

Estimated time to complete this module: 5 hours

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### Method of Participation

There are no fees for participating and receiving CME credit for this activity. During the period of TBC, 2013, through TBC, 2015, participants must; (1) read the learning objectives and faculty disclosures, (2) participate in the entire educational activity, consisting of 5 core modules, (3) complete the post-test for each module by recording the best answer to each question, and (4) complete the online evaluation form for each module. Upon successful completion of all 5 post-tests (75% or better) and online evaluation forms, you will be provided with a statement of credit which you can download, save and print.

### Disclaimer

Participants have an implied responsibility to use the newly acquired information to enhance patient outcomes and their own professional development. The information presented in this activity is not meant to serve as a guideline for patient management. Any procedures, medications, or other courses of diagnosis or treatment discussed or suggested in this activity should not be used by nurses without evaluation of their patient's conditions and possible contraindications on dangers in use, review of any applicable manufacturer's product information, and comparison with recommendations of other authorities.

### Grant Statement

This activity is supported by an educational grant from Novartis Pharma AG.

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## MODULE 2: Clinical Presentation



### 1 Module Introduction

#### Intro

The clinical presentation of MS presents a number of diagnostic challenges. There is no single sign or symptom that is specific to MS and, to further complicate matters, there is a broad range of presenting symptoms. In the early stages of the condition, MS signs and symptoms are frequently short-lived and therefore, may not be readily detectable. This module will review the different types of MS and their clinical features, including atypical presentation of the condition. Common symptoms will be discussed and defined, along with the prognosis of people with MS.

## 2 Types of MS and their Clinical Features

### 2.1 Learning Objectives

This section will describe the different types of MS and their distinguishing clinical features. After review of this section, you should be better able to:



- Explain what is meant by the term “clinically isolated syndrome”.
- Identify the various types of MS and their natural history and progression.
- Describe the variability of the clinical course of MS and resulting disability.

### 2.2 Introduction

#### Intro

The clinical course of MS varies considerably between different individuals. This can present problems in the identification of the type of MS as individuals rarely fit neatly into categories; this also has repercussions for the person with MS themselves. For example, even if diagnosed with a certain type of MS, there is no guarantee that any individual's disease will follow the same course as another's with the same type: there may be wide differences in disability and functional difficulties between individuals<sup>1</sup>.

### 2.3 Early Signs of MS / Clinically Isolated Syndrome

The majority (85%) of people who later develop MS start with an episode of neurological disturbance, usually evolving over days or weeks<sup>2</sup>. This is known as a ‘clinically isolated syndrome’ (CIS), or ‘first demyelinating event’. This is an acute, or subacute, neurological episode lasting at least 24 hours, caused by inflammation/demyelination in one or more sites in the CNS. A person with a CIS can have a single neurological sign or symptom – for example, an attack of optic neuritis – caused by a single lesion (monofocal), or more than one sign or symptom – such as an attack of optic neuritis accompanied by weakness on one side caused by lesions in more than one place (multifocal)<sup>3</sup>.



The majority (85%) of people who later develop MS start with a ‘clinically isolated syndrome’ (CIS), or ‘first demyelinating event’.

A review of people with MS found that 21% started with a CIS of optic neuritis, 46% with long tract symptoms and signs (motor or sensory deficits), 10% with a brain-stem syndrome and 23% with multifocal abnormalities<sup>4-6</sup>.

People who experience a clinically isolated syndrome may or may not go on to develop clinically diagnosed MS. It is important to note here that a person with CIS does not meet the diagnostic criteria for MS. However, studies have shown that when CIS is accompanied by MRI-detected brain lesions that are consistent with those seen in MS, then there is a high risk of a second neurologic event. Individuals with no evidence of MRI-detected lesions are at a relatively low risk of developing MS over the same time period<sup>3</sup>.



People who experience a clinically isolated syndrome may or may not go on to develop clinically diagnosed MS. Studies have shown that when CIS is accompanied by MRI-detected brain lesions consistent with MS, there is high risk of a second event.

## 2.4 Radiologically Isolated Syndrome (RIS)

Over the past decade, the increasing use of magnetic resonance imaging (MRI) in the diagnostic work-up of pathological conditions has contributed to the uncovering of asymptomatic brain pathologies<sup>7,8</sup>. The term ‘radiologically isolated syndrome’ (RIS) was recently proposed to describe asymptomatic individuals who possess radiologic abnormalities highly suggestive of MS<sup>9</sup>, i.e. RIS is detected with a brain MRI in which lesions that look like MS are observed, but who have no signs, symptoms or relapses that would indicate CIS.

Several cohorts of patients with RIS have been studied, mainly retrospectively, and a proportion of them do go on to present with clinical symptoms of MS. This has led to the clinical challenge of whether or not to treat patients with MRI lesions suggestive of MS, given the knowledge that MS disease-modifying therapies work best when given early in the disease course<sup>10</sup>.

## 2.5 Types of MS



According to internationally accepted terminology, the clinical courses of MS are categorised as relapsing remitting (RRMS), secondary progressive (SPMS), primary progressive (PPMS) and progressive relapsing (PRMS)<sup>11</sup>. A relapse is defined as an acute episode of neurological symptoms that worsen for some days and then improve or completely subside over time. A period of 30 days should separate the onset of two events for them to be considered as separate attacks<sup>12</sup>.



Clinical courses of MS are categorised as relapsing remitting (RRMS), secondary progressive (SPMS), primary progressive (PPMS) and progressive relapsing (PRMS).

### 2.5.1 Relapsing-remitting MS (RRMS)

The most common presenting course of MS is relapsing-remitting (RRMS). In about 85% of all people with MS, the condition follows a relapsing-remitting pattern at onset, with relapses occurring randomly over many years and with minimal accumulation of disability or change in disease progression<sup>13</sup>.



In about 85% of all people with MS, the condition follows a relapsing-remitting pattern at onset.

RRMS is characterised by acute attacks of neurologic dysfunction that generally evolve over days to weeks, followed by either complete or partial recovery<sup>11</sup> (see Figure 1). Those with partial recovery have what is called ‘stepwise worsening’. The important point to remember regarding RRMS is the lack of evident change in disease course between relapses. That is, whatever level function is at the end of a relapse, it remains at that level until the next attack<sup>14</sup>.

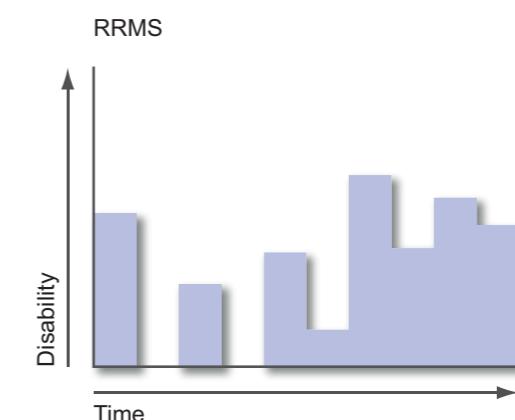


Figure 1. Graphical representation of RRMS

RRMS varies greatly in severity from individual to individual. The annual relapse rate in untreated patients initially averages about 2–2.5<sup>15</sup>, and thereafter it will gradually fall as the years progress. It is considered a poor prognostic sign if the person experiences frequent relapses, especially at the onset of the disease<sup>16</sup>.



The annual relapse rate in untreated patients initially averages about 2–2.5.

## 2.5.2 Secondary-progressive MS (SPMS)

Secondary-progressive MS (SPMS) is defined as progression of clinical disability (with or without relapses and minor fluctuations) after a relapsing-remitting onset<sup>17</sup>. The person with MS does not completely recover from relapses/attacks and disability progresses even between the relapses<sup>18</sup> (see Figure 2). A patient can have superimposed relapses, but the main distinction is a gradual worsening of function between attacks.

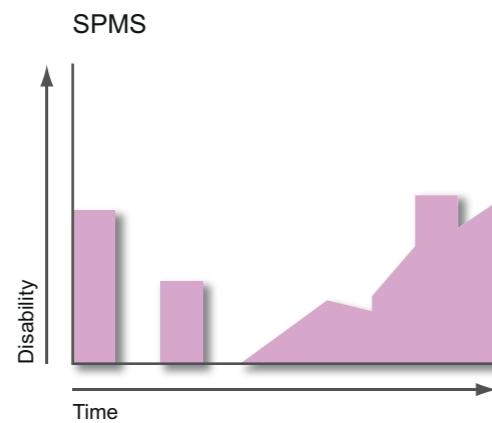


Figure 2. Graphical representation of SPMS

A significant percentage of individuals with RRMS will go on to SPMS. For an individual with RRMS, the risk of developing SPMS is ~2.5% each year, which means that the majority of RRMS ultimately evolves into SPMS within a median time interval of 19 years<sup>19</sup>. In 50–70% of people diagnosed with RRMS, the disease course becomes steadily more progressive over time. This pattern may or may not involve occasional relapses, plateaus, and remissions.



The majority of RRMS ultimately evolves into SPMS within a median time interval of 19 years.

When assessing people with MS, it can be difficult to establish when they are converting from RRMS to SPMS. The Kurtzke Expanded Disability Status Scale (EDSS) can provide the Neurologist with an indicator of whether the patient is entering the secondary-progressive phase of the disease. Patients at EDSS 4.0–5.5 are those most at risk of developing SPMS<sup>20</sup>.

## 2.5.3 Primary-progressive MS (PPMS)

Primary-progressive MS (PPMS) is characterised by a disease course that worsens continuously from onset, with only occasional plateaus or temporary improvements (see Figure 3). In this form of MS, the symptoms develop faster and the disease is progressive from the outset without any discernible relapses or remissions<sup>13,21</sup>.

PPMS is found in approximately 10–15% of the MS population and, as opposed to other forms of MS, there is no increased likelihood for women to have this form of the disease<sup>22</sup>. The unique clinical characteristics of this form make its diagnosis difficult and prolonged<sup>23</sup>.



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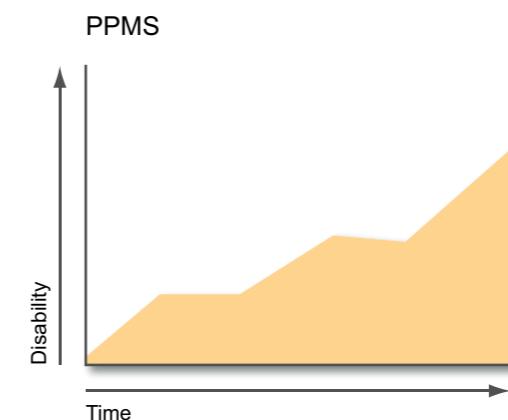


Figure 3. Graphical representation of PPMS

PPMS usually begins later in life (than RRMS) (mean age ~40 years), but in rare cases it can occur at an earlier age. It typically presents with an increasing spastic gait that is already affecting walking ability. The prognosis is poorer than for other forms; the time taking to reach EDSS 6.0 is approximately six years<sup>24</sup>. Definitive diagnostic criteria for PPMS include clinical progression for at least a year<sup>24</sup>. In PPMS the MRI of the brain can look normal as the plaques characteristically form in the spinal cord. Consequently, to diagnose PPMS it is essential for a patient also to have a spinal MRI.

## 2.5.4 Progressive-relapsing MS (PRMS)

PRMS is the least common form of MS, affecting only about 5% of people, and involves continuous disease progression from the outset with superimposed relapses<sup>25</sup> (Figure 4). There is significant recovery immediately following a relapse, but between relapses there is a gradual worsening of symptoms<sup>1</sup>. Identifying this type of MS has important implications for treatment.

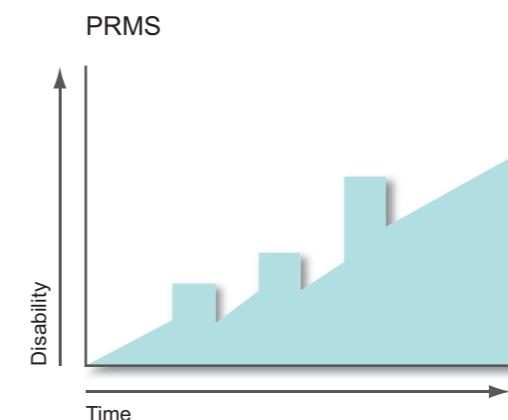


Figure 4. - Graphical representation of PRMS



PRMS is the least common form of MS, affecting only about 5% of people.

## 2.6 Summary



- Multiple sclerosis (MS) usually starts with an acute episode of neurological disturbance.
- There are four types of disease course, defined as relapsing/remitting MS, secondary-progressive MS, primary-progressive MS and progressive-relapsing MS.
- A quarter of all people with MS will do well and manage with no help.



## Reflective learning point

Thinking about radiologically isolated syndrome (RIS), clinically isolated syndrome (CIS) and multiple sclerosis, what do you believe the impact might be on someone who is given these different diagnoses? What is the most important information to communicate to the person in each case?

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Thinking about the main types of MS, outline the key differences between the different types and how you would go about identifying these variations in your day-to-day practice.

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## 3 Typical Signs And Symptoms

### 3.1 Learning Objectives

The diagnosis of MS carries with it an emotional impact that lasts a lifetime and many patients will require emotional support at various points throughout the journey of their condition. This section will discuss some of the more common emotional aspects of MS, along with how MS Nurses can help people cope and adapt to life with the condition. After review of this section, you will be better able to:



- Determine the nature and impact of some of the more common symptoms found in people with MS.
- Outline approaches used to identify some of these symptoms.

### 3.2 Introduction



MS can cause a wide variety of symptoms (*summarised in Figure 5*). At the onset of MS, symptoms can often include visual disturbance. Later, as the disease progresses, weakness, partial loss of movement and spasticity may appear (amongst others).



At the onset of MS, symptoms can often include visual disturbance. Later, as the disease progresses, weakness, partial loss of movement and spasticity may appear (amongst others).

These symptoms can add considerably to the degree of disability and impairment of quality of life (QOL) that patients with MS experience.

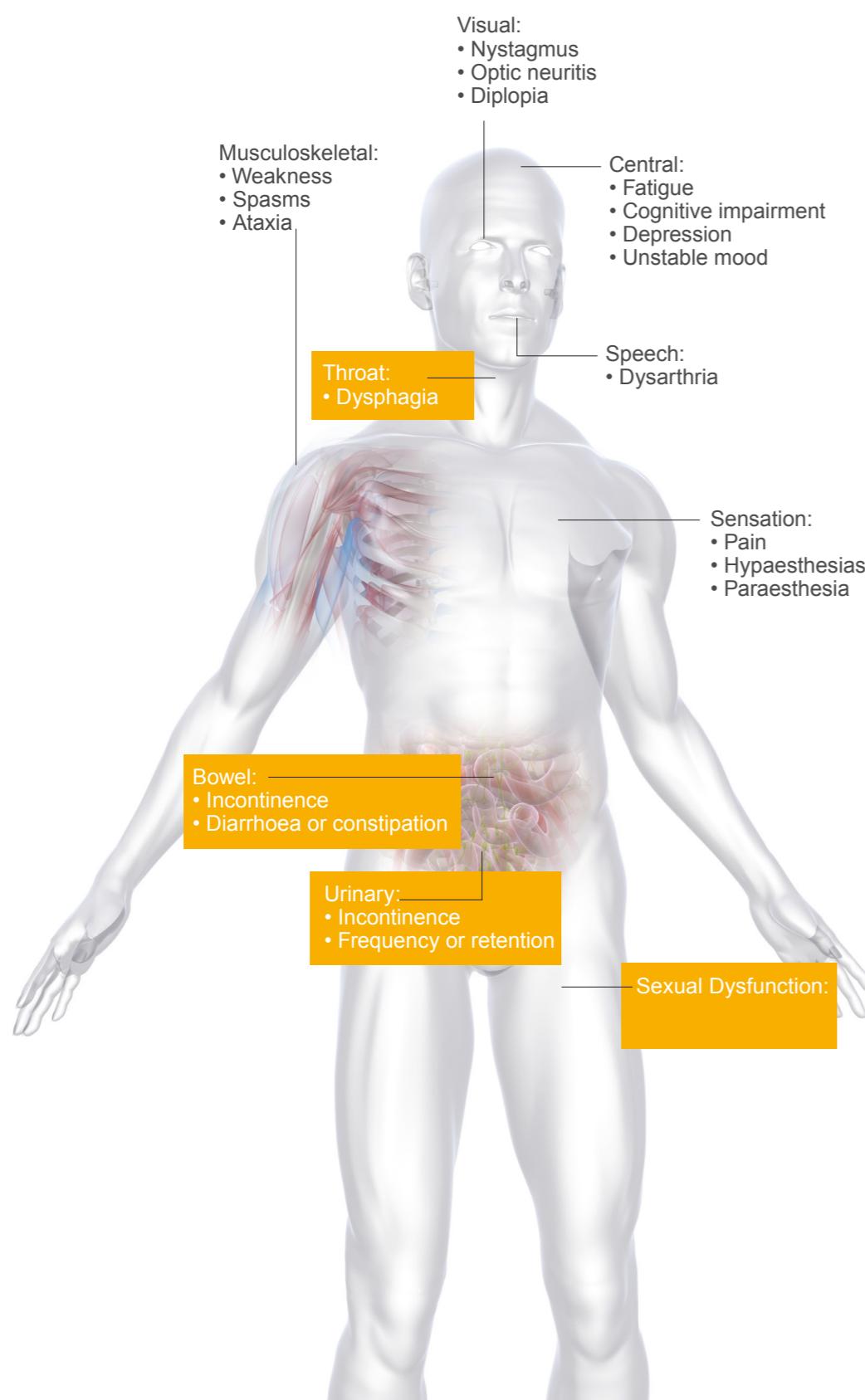


Figure 5. A summary of symptoms of MS

The symptoms of MS are unpredictable and may be mild, moderate or severe. Each person with MS differs in the types and intensity of symptoms they experience, depending on the areas of the CNS that are affected (see *Table 1*). Symptoms can be present in many different combinations and with variable severity. People with MS do not follow a linear trajectory and no two people with MS have exactly the same symptoms. Post-mortem studies have shown that some people have had MS all their lives and symptoms have been so mild they have not been recognised, while other people with MS develop severe disability very quickly following disease onset. However, for most people disability in MS lies somewhere between these two extremes<sup>26</sup>.

Initial Signs and Symptoms	Manifestation
<b>Weakness of the limbs</b>	Loss of strength
<b>Spasticity</b>	Movement-induced, painful muscle spasms
<b>Sexual dysfunction</b>	Decreased libido, impotence in men, diminished vaginal lubrication
<b>Sensory</b>	Paraesthesia (tingling and prickling sensations), hypoesthesia (reduced sensations, numbness), pain (anywhere in the body and can change locations)
<b>Cognitive impairment</b>	Memory loss, impaired attention, difficulty taking in information and with problem solving
<b>Mood disorder</b>	Depression, emotional lability, more rarely euphoria
<b>Visual deficit</b>	Diminished visual acuity, double vision, decreased colour perception; may progress to severe visual loss
<b>Bladder dysfunction</b>	Urinary frequency and urgency, nocturia, uncontrolled bladder emptying, retention, hesitancy
<b>Bowel dysfunction</b>	Constipation, faecal incontinence

Table 1. Indicators of common symptoms in MS

A survey of 2,265 individuals with MS recorded the prevalence rates of common symptoms in MS<sup>27</sup>. Results are displayed in Figure 6.

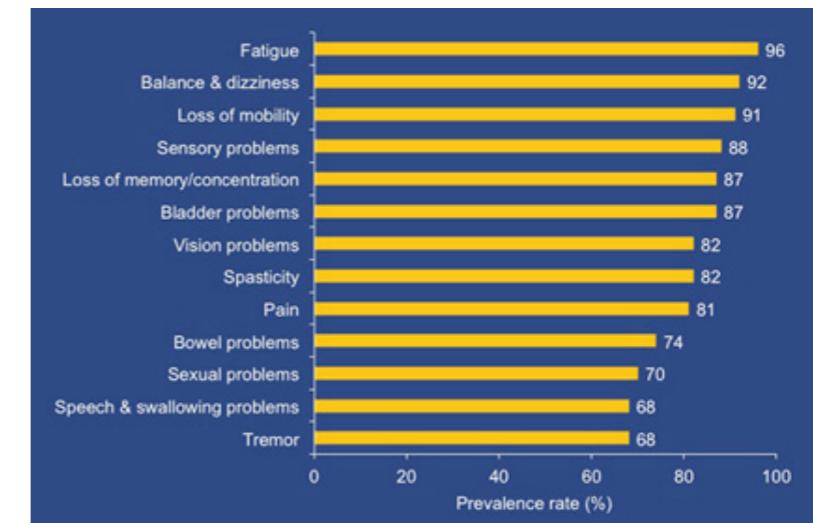


Figure 6. Prevalence of common symptoms in MS<sup>27</sup>

For the MS Nurse, identifying and discussing a person's symptoms can be challenging at times, for several important reasons:

- While some changes are readily apparent — such as walking problems, speech impairments, or tremor — others, including fatigue, bladder and bowel changes, and cognitive and emotional changes, are less visible to the observer.
- While some symptoms are relatively easy for people to discuss — for example, fatigue, double vision, stiffness or pain — others are more embarrassing, such as cognitive symptoms, bladder and bowel dysfunction, sexual dysfunction and even depression.
- While some symptoms are easy for people to associate with a disease that affects the nervous system — including sensory problems, weakness, loss of balance, or visual symptoms — they may neglect to mention other problems that they assume are unrelated to MS (e.g., fatigue, bladder or bowel changes, sexual dysfunction, cognitive changes, or pain).

For all these reasons, it is important that the MS Nurse carries out a complete assessment at every visit, asking about symptoms or changes, even if a person has not mentioned any difficulties. It is equally important to make sure that people living with MS have access to accurate and comprehensive information so that they become experts who can self-manage their condition.



## Nursing tip

Where would you start when assessing for any problems or symptoms that a person with MS may be experiencing – the approach you take, questions you ask etc?

People with MS can present in many different ways. Early in the disease it is not helpful to ask "Do you have any MS symptoms?" because they may not recognise what an MS symptom is. The nurse should start with a review of systems asking specific questions about things such as thinking, memory, vision, strength, walking, bowel and bladder function. Targeted questions such as "Do you have bladder problems such as going too frequently, urgency to void or not feeling like you have fully emptied your bladder?" are the most helpful.



It is important that the MS Nurse carries out a complete assessment at every visit, asking about symptoms or changes, even if a person has not mentioned any difficulties.

## 3.3 Common Symptoms

Some of the more common symptoms of MS will now be defined and described in more detail. Different strategies that an MS Nurse might use to manage these symptoms will be discussed in Module 4 and Module 5.

### 3.3.1 Fatigue

Fatigue is more than tiredness and has been referred to as "pathological exhaustion"<sup>28</sup>. The Centres for Disease Control and Prevention (CDC) defines "pathological fatigue" as fatigue that "is not improved by bed rest and that may be worsened by physical or mental activity".

Fatigue is the most common symptom of MS, occurring in approximately 75–95% of patients in all age groups and with all types of MS<sup>29–31</sup>. Between 50% and 60% of patients with MS report fatigue as their worst problem, regardless of severity of disease or disability, and up to 69% of patients consider it one of the most **disabling features** of MS<sup>32,33</sup>. Fatigue has been cited as one of the two major reasons for unemployment in MS individuals<sup>34</sup>.



Fatigue is the most common symptom of MS, occurring in approximately 75–95% of patients in all age groups and with all types of MS.

Fatigue is a primary determinant of poor QOL in MS<sup>35</sup>, affecting both physical and mental components independent of disability level<sup>32</sup>. Fatigue also has a negative effect on cognitive function<sup>36</sup>. It is often described as an invisible symptom as there is no external evidence or internal damage. This can lead to a lack of understanding and frustration from others, as they may believe that the person is just lazy, often causing social isolation<sup>31</sup> and broken relationships. These views can sometimes be shared by the person with MS as they struggle to deny to themselves how fatigue affects them. It can therefore have a high impact on psychological wellbeing of the person with MS.

#### Disabling features

In a survey of 2,265 people with MS, 94% experienced fatigue, with 87% reporting an impact they rated between moderate and high on their activities of daily living.

Fatigue results from a combination of factors, caused partly by MS itself (known as primary fatigue) and partly by other factors (secondary fatigue) that affect the person with MS more significantly than those without the condition. MS-related fatigue can come on suddenly, even in the morning after a full night's sleep. MS-related fatigue is generally more severe and more likely to interfere with daily responsibilities than normal fatigue. Patient complaints may include tiredness, poor endurance, weakness, difficulty concentrating or mental dullness<sup>37</sup>.

Various other factors can contribute to fatigue in MS, including disturbed or reduced sleep, medication effects, increased muscular effort to accomplish daily tasks, depression, comorbid conditions, infections and relapses<sup>36</sup>.

Fatigue must be distinguished from limb weakness and from depression, though there may be an inter-dependent relationship between depression and fatigue. Fatigue can have a frightening effect, particularly in relation to cognitive difficulties that can be more noticeable when fatigue is being experienced. Similarly, it can be a worrying problem for carers who may fear that psychological problems are developing, or that inactivity is due to laziness<sup>38,39</sup>.

**Heat-sensitive fatigue** is well recognised in MS and it has long been considered a unique dimension of MS fatigue differentiating it from fatigue in other conditions. Changes in temperature can cause some patients with MS to become fatigued. This can be triggered by the weather, hot baths or showers, hot drinks or meals, or feeling feverish as a result of infections. These effects are usually quickly reversed when steps are taken to cool down or when the temperature falls<sup>40</sup>.

### 3.3.2 Sleep Disorders

Sleep disturbances are common in people with MS, with approximately 50% of individuals reporting sleep problems<sup>42</sup>. Treatment of sleep disorders in people with MS is important because poor sleep quality and quantity can affect daily life, causing daytime sleepiness, decreased concentration and memory, worsening depression and the inability to work effectively<sup>43</sup>. Sleep dysfunction can potentially exacerbate other MS symptoms (e.g., mental health problems, fatigue)<sup>44</sup>, and has recently been shown to be an independent predictor of QOL in people with MS<sup>45,46</sup>.

#### Heat-sensitive fatigue

Many people with MS experience a temporary worsening of their symptoms when the weather is very hot or humid, when they run a fever, sunbathe, get overheated from activity, or take hot showers or baths. A definitive symptom of MS 'heat fatigue' is when vision becomes blurred when the person with MS is overheated; this is a phenomenon known as Uhthoff's sign. These temporary changes can result from even a very slight elevation in core body temperature, as little as 0.5°C. An elevated temperature further impairs the ability of a demyelinated nerve to conduct electrical impulses. For many years, the "Hot Bath" test was used to diagnose MS. A person suspected of having MS was immersed in a tub of hot water, and the appearance of neurologic symptoms or their worsening was taken as evidence that the person had MS.

It is important to remember that heat generally produces only temporary worsening of symptoms and does not cause actual tissue damage (demyelination or damage to the axons themselves), however the use of the "Hot Bath" test has been erroneously associated with permanent tissue damage. Heat-related symptoms are generally rapidly reversed when the source of increased temperature is removed<sup>41</sup>. As with so many other MS symptoms, fatigue can be exacerbated by, or conversely cause exaggeration to cognitive/emotional symptoms, spasticity, exercise tolerance and weakness poor nutrition, speech and swallowing problems.



Sleep disturbances are common in people with MS, with approximately 50% of individuals reporting sleep problems.

The most common sleep disorders seen in patients with MS include insomnia, nocturnal movement disorders, sleep-disordered breathing, narcolepsy, restless leg syndrome and rapid eye movement sleep behaviour disorder<sup>47</sup>. Sleep questionnaires were sent to a group of 473 people with MS<sup>48</sup>. Over 46% had moderate-to-severe sleep problems, but most did not use over-the-counter or prescription sleep aids. Factors found to contribute to sleep problems included anxiety, night-time leg cramps, fatigue and nocturia.

Getting a good night's sleep helps to alleviate many common symptoms of MS, including chronic fatigue, mood and memory problems. Sleep quality can be improved by establishing regular habits or good 'sleep hygiene'. Some sleep hygiene recommendations that might be communicated to the person with MS include:

- Stay as physically active as possible during the day, but allow plenty of time to wind down before bedtime.
- Avoid getting overtired by doing too much, as being too tired can make it difficult to get to sleep.
- Ensure some daily exposure to sunlight and avoid bright lights in the evening.
- Avoid unnecessary stress or stimulants (e.g., caffeine, chocolate, alcohol) in the late afternoon and evening.
- Establish a bedtime routine that can include, for example, relaxation, a warm 'milky' drink, taking a warm bath.
- Keep the same nightly ritual every evening throughout the week.
- Only use the bed as a place for sleeping, not for other activities such as watching TV.
- Instead of lying in bed awake, after 15 minutes of not sleeping get out of bed and do something calming or boring, and return to bed after a short period of time.
- Avoid getting too hot; keep the bedroom at a comfortable temperature.

The MS Nurse should ask patients about sleep quality and refer patients for further help with sleep-related issues when indicated. By addressing the causes of poor sleep quality and establishing good daily sleep habits and sleep hygiene, people with MS can greatly improve overall energy levels and QOL.

### 3.3.3 Vision Impairment

Visual disturbance is one of the most commonly reported symptoms in MS, with figures up to 80%<sup>49</sup>, and it is the presenting symptom in 25–50% of cases<sup>50</sup>. Abnormalities of vision are usually due to plaque formation along the optic nerve. There can be loss of visual acuity, double vision or tunnel vision (monocular or binocular, depending upon which nerves have been affected), and photophobia<sup>51</sup>.



Visual disturbance is one of the most commonly reported symptoms in MS, and it is the presenting symptom in 25–50% of cases.

Optic neuritis is the most common ocular disorder in MS. The effects of optic neuritis are visual disturbance and ocular pain. 70% of people with MS will experience optic neuritis during the course of their disease. Like MS itself, it normally affects people aged between 20 and 40 years, affects women more than men, and usually occurs in one eye rather than bilaterally<sup>52</sup>. Not everyone who experiences optic neuritis goes on to develop further symptoms of MS, but a significant proportion do<sup>53</sup>.

Optic neuritis is characterised by dimmed vision with acute pain on eye movement, a consequence of which is blurred, incomplete or jumping vision. It may be accompanied by nystagmus (rapid eye movements). This is usually horizontal but may also be rotary, upbeat and downbeating.

Optic neuritis is usually transient and associated with good recovery. It occurs suddenly, is progressive and usually reaches its peak after about two weeks. Visual recovery takes from 4 to 6 weeks, but colour vision can be severely impaired with other minor defects often persisting. Above all, loss of colour vision is a typical indication of MS; it develops if the optic nerve is inflamed in the region around the central retina section, which is the area responsible for seeing in colour<sup>1</sup>.

### 3.3.4 Tremor

Approximately one third of people with MS have tremor. It is one of the most disabling symptoms of MS, causing the person to become dependent as many daily activities become difficult to perform, e.g., writing, eating, dressing and personal hygiene. People with severe tremor have a high level of disability and a loss of independence in activities of daily living. A survey undertaken as part of the European TREMOR study, found that people with MS who had moderate-to-severe tremor were unable to continue in their employment and had to either give up or greatly modify their leisure activities. Commonly, a person tries to cope with tremor either by **avoidance of movements**, by **compensation strategies** or by adaptation of their external environment.

#### Avoidance of movements

Avoidance of movements: Tremor can be socially isolating. The person with tremor will often avoid movements that make their difficulties obvious. For example, many will refrain from eating or drinking in public.

#### Compensation strategies

Compensation strategies: By using strategies such as pressing the elbow firmly to the side of the trunk, a person may find that their distal tremor is diminished and they are better able to perform movements of the hand even although they will have a shorter arm reach as a result. Those with head tremor (titubation) may attempt to stabilise the head against the shoulder in an attempt to reduce the tremor. Retracting the shoulder girdle and pressing it against the back of the chair, or fixing the elbow in a locked straight position, may give improved distal control.



Approximately one third of people with MS have tremor. People with severe tremor have a high level of disability and a loss of independence in activities of daily living.

Tremor in MS can affect the limbs, trunk, vision and speech. It has been described as the most frustrating MS symptom to treat<sup>54</sup>. Stress and anxiety can exacerbate tremor.

In MS, the two most prevalent tremor forms are postural tremor and intention tremor.

- Postural tremor is present while a position is voluntarily maintained against gravity. This type of tremor is common in people with MS and may include titubation of the head and neck.
- Intention tremor is obvious when a movement is target-directed with an increased amplitude during visually guided movements towards the target. This can be observed during the finger-to-nose test when the person is asked to lift their arm out to the side, then to bend their elbow and touch their nose with their index finger<sup>55</sup>: As the finger approaches the nose the tremor amplitude increases. Intention tremor is related to lesions in the cerebellum and/or connected pathways in the brain stem and the term is often synonymously used with ‘cerebellar tremor’<sup>56</sup>.

Resting tremor, which is observed when a body part is not voluntarily activated and supported against gravity, is unusual in MS<sup>57</sup>.

### 3.3.5 Bladder Dysfunction

Approximately 75% of people with MS experience some type of bladder dysfunction during the course of their disease. The effect of this can be difficult for the person, since bladder control may deteriorate at the same time as mobility worsens, making it increasingly difficult to respond to bladder urgency by hurrying to the toilet.



Approximately 75% of people with MS experience some type of bladder dysfunction during the course of their disease.

There are three types of bladder dysfunction most commonly associated with MS<sup>58</sup>. They are summarised in Table 2.

Nature of Dysfunction	Cause	Common Symptoms
Storage dysfunction	Detrusor hyperreflexia	Urgency, frequency, nocturia, incontinence
Emptying dysfunction	Diminished detrusor contractility Sphincter hypertonia	Urgency, dribbling, frequency, hesitancy, incontinence, infection
Combined storage and emptying dysfunction	Detrusor-sphincter dyssynergy	Urgency, hesitancy, frequency, dribbling, incontinence, infection

Table 2. Types of bladder dysfunction

### 3.3.5.1 Failure to Store (bladder overactivity)

Bladder overactivity is the problem that the person with MS is usually most aware of. It is the most common type of bladder dysfunction in MS, with reported incidences ranging from 26–50%. Symptoms of this dysfunction include urgency, frequency, urge incontinence and nocturia<sup>59</sup>.

It results in a tendency for the bladder to contract unpredictably and sometimes uncontrollably. At its worst, the bladder may seem to have ‘a life of its own’.

After only partial filling the person senses urinary urgency. Incontinence may occur if the contraction pressures are too high and the problem is made worse if mobility is affected and it is difficult to reach the toilet in time.

Another feature of the impaired nerve supply to the bladder muscle is that the normal capacity is diminished, causing urinary frequency. This increases the frequency of emptying from every 3–5 hours (depending on how much is drunk) to hourly or worse.

### 3.3.5.2 Failure to Empty

This type of bladder dysfunction is less common (incidence 19–40%) but it can result in more serious complications than failure to store. Symptoms of failure to empty include frequency, nocturia, retention, overflow incontinence and urinary tract infections.

Although some people with MS are aware that their bladders do not empty properly, others with the same problem are not. For many, needing to void again soon after doing so is an indicator that bladder emptying is poor. Research has shown that if people with MS thought they were not emptying their bladder properly, they were usually correct. However, of those who thought they were **emptying completely**, about half were wrong and were surprised to find how much urine they had been leaving behind<sup>60</sup>.

### 3.3.5.3 Combination Failure to Store and Failure to Empty

This occurs in 24–46% of people with bladder dysfunction in MS. It is often associated with detrusor-sphincter dyssynergia. Symptoms of this type of bladder dysfunction include those associated with both failure to store and failure to empty<sup>62</sup>. If not treated appropriately, this condition can lead to recurrent urinary tract infections, urinary reflux, hydronephrosis and, in extreme cases, renal failure.

### 3.3.6 Bowel Dysfunction

Bowel problems are common among people with MS, although they are generally under reported and neglected<sup>59</sup>. Wiesel and colleagues<sup>63</sup> reported that the prevalence of bowel dysfunction in patients with MS is higher than in the general population (up to 70%)<sup>63</sup>.

Bowel dysfunction does not appear to be associated with the degree of disability; however, it is associated with duration of MS<sup>64</sup>. It is a source of considerable ongoing distress in many people with MS<sup>63</sup>.

In order to control bowel actions, it is necessary to be aware of the need, or '**call to stool**'.

However in MS the messages are often either lost or incomplete, leaving a situation in which it may be difficult to differentiate between the calls.

The two main types of bowel dysfunction in MS are constipation and faecal incontinence, which may coexist:

#### Emptying completely

Incomplete bladder emptying is the result of two things going wrong, both of which are due to spinal cord malfunction<sup>61</sup>:

- **detrusor-sphincter dyssynergia**: the muscle that surrounds the bladder outlet tube (urethral sphincter) does not relax when the bladder muscle contracts, but instead goes into contraction, thus causing an interrupted flow.
- **detrusor hyperreflexia with poorly sustained contractions**: the neural impulses which in health keep the bladder muscle contracting until it is completely empty do not get down the spinal cord. When the bladder does contract, the contractions, although frequent, are poorly sustained.

#### 'Call to stool'

'Call to stool' awareness occurs when the faeces move into the rectum causing the rectum to expand and send messages via the sensory pathways of the need to evacuate. At this point, the finely tuned nerve endings of the rectum are able to differentiate between whether the stool is solid, liquid or wind.

### 3.3.6.1 Constipation

**Constipation** is defined as two or fewer bowel movements per week and/or the use of suppositories, laxatives or enemas more than once per week to promote bowel movements. It is estimated that in the general population of Europe the mean value of the reported constipation rates is 17.1%<sup>65</sup>. Constipation occurs in approximately 36–53% of people with MS.

### 3.3.6.2 Faecal Incontinence

Faecal incontinence is defined as the involuntary passage of stool. In people with MS it occurs in 25% once a week and in 51% less than once a month. Factors contributing to faecal incontinence include constipation that causes rectal distension and overflow, diminished rectal sensation, sphincter dysfunction, certain medications and diet<sup>64</sup>.

### 3.3.7 Sexual Problems

The private and intimate nature of issues relating to sexuality presents a challenge to both people with MS and the healthcare professionals caring for them. Patients who are experiencing sexual problems may be unaware of the extent to which these can be attributed to MS; whilst others may be concerned about the potential impact of MS on their sexuality. It is important, therefore, for nurses working with people diagnosed with MS to be alert to the varying needs of patients for assessment, information and reassurance about sexuality and sexual matters.

The incidence of sexual dysfunction in people with MS varies considerably between different reports. A 2009 review reported that between 50% and 90% of men and 40% and 80% of women are affected<sup>66</sup>. The most frequently reported symptoms in women are anorgasmia or hyporgasmia, decreased vaginal lubrication and reduced libido, and men most frequently experience impotence or erectile dysfunction (ED), ejaculatory and/or orgasmic dysfunction and reduced libido<sup>67,68</sup>.

The prevalence of sexual dysfunction is higher in MS than in other chronic diseases, and almost five times higher than in the general population<sup>69,70</sup>. Such studies tend to focus on physical problems and the total impact of a change in an individual's sexuality is often overlooked.

#### Constipation

Factors that contribute to constipation include<sup>64</sup>:

- neurologic changes.
- lack of sensation in the rectal area.
- weakened abdominal muscles.
- lack of mobility and exercise.
- insufficient fluid intake.
- medications, particularly those used to treat urinary symptoms.
- inadequate bowel routines, particularly the lack of a regular and relaxed time for elimination.



Prevalence studies indicate that approximately 70% of people with MS experience changes in sexual function.

Sexual dysfunction is correlated with the presence of other disabilities, in particular bladder and bowel symptoms, sensory disturbance of the genitalia, weakness of the pelvic floor and spasticity<sup>67</sup>. The associated factors may be recognised as 'risk factors' and alert the nurse to the possibility of sexual dysfunction. In common with other symptoms of MS, those of sexual dysfunction can relapse and remit.

Sexual problems can be described as primary, secondary or tertiary<sup>71</sup>. People with MS may experience dysfunctions at any of these phases.

### 3.3.7.1 Primary Sexual Dysfunction

Primary sexual dysfunction occurs as a result of demyelinating lesions in the spinal cord and brain that directly impair sexual feelings and/or response. Examples include:

- decreased or absent libido.
- altered genital sensations or paresthesias.
- decreased or absent genital sensations.
- decreased frequency or intensity of orgasms.
- erectile dysfunction.
- decreased vaginal lubrication or clitoral engorgement.
- decreased vaginal muscle tone.

### 3.3.7.2 Secondary Sexual Dysfunction

Secondary sexual dysfunction refers to changes in sexual function that result indirectly from other MS symptoms or medical/pharmacological interventions.

The following MS symptoms can interfere with sexual expression:

- fatigue – interferes with interest, activity level and spontaneity.
- spasticity – affects comfort and positioning.
- non-genital sensory paresthesias – reduce comfort and pleasure.
- weakness – interferes with sexual activity.
- pain – reduces sexual activity and pleasure.
- bladder/bowel dysfunction – causes anxiety and discomfort.
- tremor – interferes with sexual activity.
- cognitive impairment – affects attention and psychogenic stimulation.

The following medications can interfere with the sexual response:

- tricyclic antidepressants and selective serotonin reuptake inhibitors (SSRIs) for depression – can inhibit libido and orgasm.
- anticholinergic and antimuscarinic medications – reduce vaginal lubrication.
- antispasticity medications – produce significant fatigue.
- anti-seizure medications used to control pain or tremor – produce significant fatigue.

### 3.3.7.3 Tertiary Sexual Dysfunction

Tertiary sexual dysfunction refers to the psychological, social and cultural issues that interfere with sexual feelings and/or response. Examples include:

- “Disabled people aren’t sexually attractive.”
- “I can’t be both a caregiver and a lover.”
- “If I don’t like myself any more, how can I expect someone else to find me attractive?”
- “This isn’t the same person I married.”
- “He/she doesn’t find me attractive now that I’m a burden.”
- “With everything else that’s going on, sex is the last thing I care about right now.”

It is important to establish in which of these areas a sexual problem is presenting. The range of potential interventions includes (but is not limited to) those which directly enhance sexual activity, the pharmacological management of MS symptoms and counselling directed at self-image or improved communication skills.

It is also important to remember that MS is a condition of young people who may wish to be/remain sexually very active. It is important to consider that the level of physical function or relationship is not relevant to desire for sexual fulfilment.



### Nursing tip

**What might be the right way to start talking about difficult/personal problems, such as bladder/sexual problems, with the person with MS?**

Letting the person with MS know that problems with bowel, bladder and sexual function can be a part of MS is often a reassuring way to start a conversation. It is helpful to discuss the role the nervous system plays in each of the functions discussed and, if possible, provide diagrams for illustration. Letting them know that they are not alone and that, in most cases there are many ways to approach those problems, often relieves anxiety from the start. The nurse as a partner with the patient and family can explore many options to manage MS symptoms. The nurse should also explore other issues related to the symptoms. The female patient with sexual dysfunction may feel it is not worth discussing, yet simple changes such as emptying the bladder before intercourse and alternative positions are often helpful. The patient who has a solid relationship with the MS nurse is in the best position to partner with the nurse to approach the sometimes complex problem of symptom management.

### 3.3.8 Speech Difficulties

At a physical level, MS can affect the production of speech, usually by delays in messages passing through affected nerve pathways to the muscles involved in speech production.

**Symptoms** are variable and often relate to fatigue and stress levels. Some people with MS experience a mild reduction in volume when tired or a slight slurring of speech at the end of the day. Others find their thoughts run ahead of their words<sup>43</sup>.

Speech disturbances have been shown to correlate with severity of demyelination and progressive disease, but not with duration of illness, age or onset of MS<sup>72</sup>. Speech disturbance is uncommon in the initial stage of MS and tends to occur as a later manifestation. Initially the speech disorder is mild and the severity increases progressively with greater degree of neurological involvement; it is more pronounced in individuals in whom the disease process involves a greater number of neurological systems.

It is difficult to determine the number of people who will experience speech and communication difficulties with their MS. Various studies have reported incidences ranging from 44% to 77%<sup>73</sup>. In a large group study of patients with MS, Hartelius and colleagues<sup>74</sup> found that 62% of the group reported speech and voice impairments. The variation in the incidence depends on the severity, duration and stage of disease progression.

#### Symptoms

The main symptoms of speech disturbances in MS are:

##### Imprecise articulation (dysarthria)

Speech can be slow and slurred, with imprecise vowels and consonants, sudden breakdowns or excessive length of sounds. These difficulties are caused by slowness and weakness of tongue movements, and to a lesser extent, by difficulties related with movement of lips and jaw.

##### Voice abnormalities (dysphonia)

There are different types of voice disturbances: harshness, breathiness, hoarseness and a strained-strangled voice. The voice intensity may be low and decrease with fatigue. In some cases, the person can produce only short utterances. These disturbances are caused by abnormalities of breathing and of the laryngeal system. Deficits in breathing may consist of poor breath support and lack of breath control for speech. Deficits in the laryngeal system may be caused by hyper- or hypofunction. Sometimes the voice has an excessive nasal resonance.

##### Abnormal speech melody (dysprosody)

The person with MS may show prosodic disturbances, such as slowness or excessive velocity; poor or excessive pitch variation; and excessive variation of loudness. These disturbances are caused by reduced breathing control, by laryngeal and articulatory dysfunction, or by poor coordination between these components.

Dysarthrias are commonly associated with other symptoms caused by brain-stem lesions, such as head tremor and incoordination of fine motor control. The following speech symptoms have been reported in order of frequency of occurrence:

- impaired loudness control.
- voice harshness.
- defective articulation.
- impaired emphasis.
- impaired pitch control.

Research into the effects of speech therapy on people with MS indicates that therapy can be beneficial. General advice may include reducing background noise before speaking, saying half words on each breath, speaking slowly and facing listeners when speaking. Traditional speech exercises may be beneficial if the problem is very mild (e.g., exercises to assist breathe control for volume).

### 3.3.9 Swallowing Difficulties

Since speech and **swallowing** share the same anatomical structures and some physiological mechanisms, speech disorders can be associated with swallowing disorders.

Swallowing disorders (dysphagia) has been reported in 34% of MS patients, with a significant correlation with pronounced **severity of illness**<sup>75,76</sup>. The disturbances usually involve oral and pharyngeal phases of swallowing, although upper oesophageal sphincter dysfunction has also been detected<sup>75</sup>. This can include difficulty chewing, pocketing food in the cheek, drink dribbling from the mouth, and episodes of coughing/choking when eating or drinking. It is not unusual for people with MS to deny swallowing difficulties, even when family members report concerns.

#### Swallowing

Swallowing has been recognised as one of the most basic biological functions, but the real process is by no means basic. The act of swallowing consists of 3 phases:

- Oral
- Pharyngeal
- Oesophageal

The oro-pharyngeal phases last no longer than 1.5 seconds but involve the coordination of no less than 31 paired muscle groups<sup>73</sup>. Impairment to the neurological control of swallowing results in dysphagia, and may lead to potentially serious effects on respiratory function, nutrition and QOL.

#### Severity of illness

A recent study<sup>77</sup> found that people with MS with dysphagia had a significantly longer disease duration ( $p=0.031$ ) and more neurological impairment in cerebellar functional system ( $p=0.04$ ) when compared with non-dysphagic patients. Dysphagia was significantly more prevalent in people with more neurological disability as measured by EDSS scores ( $p=0.04$ ). These results emphasise the importance of assessment and management of swallowing function in people with MS, particularly in those with a high EDSS score, more severe cerebellar dysfunction and long disease duration.

Self-reports of chewing and swallowing problems generally increase as MS progresses; the incidence of these problems is 65% in the most severely disabled patients<sup>78</sup>. Depending on the location and extent of demyelination, swallowing disorders can relapse and remit along with MS exacerbations.

A speech and language therapist will be able to assess adequacy of swallowing through manual assessment and videofluoroscopy. Following assessment, the therapist will advise on posture, amounts and consistencies of food, and eating environment. If swallowing is considered unsafe and recurrent chest infections or marked weight loss develop, then alternative ways of feeding will be recommended, e.g., nasogastric feeding or percutaneous endoscopic gastrostomy (PEG) feeding.

### 3.3.10 Cognition

Cognitive problems can arise early in the course of MS, although the greater the disease duration and severity, the more likely cognitive problems are to occur. As with physical symptoms, MS may affect some cognitive functions while others are left intact. Cognitive symptoms can worsen during relapse and improve during remission, but more commonly cognitive symptoms develop slowly and gradually.

Approximately 50% of people with MS develop measurable cognitive deficits<sup>79-81</sup>. In 5–10%, cognitive impairment interferes significantly with everyday activities, but it is rarely severe enough to require institutionalised care. Cognitive impairment can occur in patients with little physical disability, and is an independent predictor of subsequent impairment in work and social settings<sup>82</sup>. It can cause difficulties in maintaining employment, performing activities of daily living, and adhering to medication; cognitive impairment interferes with social interactions, strains family relationships and causes significant emotional distress<sup>83</sup>.



Approximately 50% of people with MS develop measurable cognitive deficits.

Cognitive impairment remains relatively mild for most people with MS<sup>84</sup>, but in a small proportion (around 10%) it progresses further to resemble a form of subcortical dementia<sup>85</sup>. Cognitive impairment tends to get slightly worse very gradually, and at an unpredictable pace, over many years. The rate of change depends largely upon the degree of disease activity in the brain.

Some cognitive functions are more likely to be affected than others in people with MS. These include **recent memory**, **attention and concentration**, **information processing**, **executive functions** (such as planning and problem-solving), **visuospatial functions** and verbal fluency<sup>86,87</sup>. General intellect, long-term memory, conversational skill and reading comprehension are most likely to remain intact. General language functions, routine social skills and orientation to person, place and time are rarely significantly affected in MS (even when cognitive impairment becomes severe overall), although some degree of mild word-finding difficulty is common<sup>82</sup>.

There is a wide range of individual variation in the experience of cognitive impairments. Many people with MS experience none. Others may experience impairment in one area only; whilst others may experience a combination of the more common areas of impairment. Even mild cognitive impairment can have a significant impact on a patient's capacity to **function in their daily lives**.

Therefore, evaluation of cognitive functioning should be part of the ongoing neurological and nursing assessment of people with MS<sup>88</sup>.

Motor, sensory and fatigue symptoms may also affect a person's cognitive functioning. Therefore, these symptoms need to be taken into account when evaluating cognitive impairment. Emotional state can also affect a person's cognitive functioning<sup>89</sup>.

**Memory loss** is probably the commonest cognitive problem experienced by patients with MS. Apart from the obvious difficulties presented by forgetfulness, memory loss also has implications in terms of learning new skills.

**Attention and concentration** lapses can also cause problems, especially when attention needs to be divided between tasks.

**Reasoning and judgment**, including new learning, problem solving and behavioural regulation, may also be impaired but, because of the subtle nature of reasoning, this problem is often much less obvious.

**Speed of information processing** can be affected. This is particularly noticeable when people have to deal with information coming to them from different directions.

**Visuo-spatial perception** is also sometimes impaired. Cognitive functions which are less likely to be affected by MS include language, remote knowledge, 'old knowing', previously learned motor skills (e.g., riding a bicycle) and long-term automatic social skills.

#### Function in their daily lives

A significantly slowed speed of information processing has been linked with an increased risk for car accidents, and also with a greater risk of misunderstanding what is been said, which in turn can lead to an increase in interpersonal conflict.

Even mildly reduced problem-solving and organising skills can have a great impact on a patient's ability to successfully juggle the multiple competing demands on their time and energy each day, such as maintaining a job, keeping up with after-school activities of their children, and managing their own personal MS symptom care plans which might require scheduled fatigue- or toilet-breaks.

### 3.3.11 Spasticity

Spasticity affects approximately three quarters of people with MS<sup>90</sup>. In a survey a third of people with MS reported spasticity as either a moderate or the worst symptom they experience on a daily basis<sup>91</sup>. Severity of spasticity was related to duration of MS, severity of disability, number of relapses and worsening symptoms in recent months<sup>91</sup>. Spasticity negatively affects daily activities in up to 44% of those who are affected by it<sup>32</sup>. It can worsen gait problems, physical components of QOL, and bowel or bladder dysfunction<sup>32,92</sup>.



Spasticity affects approximately three quarters of people with MS.

**Spasticity** is one component of the upper motor neurone syndrome that occurs as a result of acquired damage to any part of the CNS, including the spinal cord. It has a range of effects, which can be categorised into positive and negative features; most people will present with a combination of the two<sup>93</sup>.

Spasticity includes active muscle spasms, muscular tightness, stiffness, inelasticity and weakness. Muscle weakness is a frequent first presenting symptom of MS, and spasticity often compounds the weakness. Dizziness, vertigo and numbness can also occur in MS and interfere with mobility<sup>32</sup>. It can decrease range of motion, hinder the initiation or cessation of movement, cause pain, increase fatigue and provoke falls. It is a major source of disability in the lower limbs<sup>90</sup>. People with MS may notice that certain movements are accompanied by increasing rigidity. This often occurs in the morning or after being seated for a length of time; the cramping improves after several minutes. In addition, spontaneous jerks or contractions of the extremities can occur<sup>1</sup>.

#### Spasticity

The control and regulation of normal skeletal muscle activity involves a complex combination of descending motor commands, reflexes and sensory feedback, both from the brain and spinal cord, and from peripheral sensation. During normal movement, influences from the cerebral cortex, basal ganglia, thalamus and cerebellum, travelling via upper motor neurones, adjust, reinforce and regulate the lower motor neurone which connects directly via peripheral nerves to the muscle to form smooth, coordinated muscle activity and maintenance of posture.

In simple terms, spasticity occurs when there is damage to these descending upper motor neuron tracts (e.g., a plaque in MS). This interrupts the regulation of spinal cord and lower motor neurone activity. This can result in enhanced lower motor neurone activity and a consequent increase in muscle activity, in response to peripheral stimuli (e.g., muscle stretch, a urinary tract infection or pressure ulcer)<sup>94</sup>.

#### 3.3.11.1 Consequences of Spasticity

Spasticity does not always cause discomfort or inconvenience for people with MS. The muscle stiffness can compensate for weakness in some cases, and can assist activity that would not otherwise have been possible, as well as assisting with some components of physiotherapy<sup>95</sup>. However, in most cases spasticity does cause problems. The increased stiffness in the muscles consumes a great deal of energy, can hinder coordination and exacerbate other MS symptoms such as fatigue.

Spasticity can affect physical activities such as walking, transferring, picking up objects, washing, dressing and sexual activity. It can also have an emotional impact, on for example, mood, self-image and motivation<sup>96-98</sup>. Safety in sitting and lying can also be compromised due to spasms or persistent poor positioning which can lead to the development of contractures. This can potentially lead to restricted community mobility and social isolation. Therefore, the treatment of spasticity needs to be carefully selected and reviewed over time in order to meet the individual's aims, and to promote and maintain function.

### 3.3.12 Pain

Pain is a common symptom in people with MS. Studies report prevalence ranging from 30% to 90%<sup>99-101</sup>, and it is often one of the presenting symptoms<sup>2</sup>. Pain in MS shows correlations with both anxiety and depression, and can affect all aspects of function and physical and mental QOL domains<sup>32</sup>. Almost one-half of patients with MS and pain report that pain interferes with social activities, work or sleep.



Pain is a common symptom in people with MS. Studies report prevalence ranging from 30% to 90%.

Pain can be difficult to cope with and those who experience it may also be more prone to poorer mental health. Pain can lead to depression, and depression in turn may contribute to the experience of pain. The pain experienced by people with MS can be either primary, a direct result of nerve damage, or secondary, due to disability, for example low back pain from prolonged wheelchair use or poor posture<sup>43</sup>.

A systematic review of pain in MS<sup>100</sup> differentiates between four distinct pain categories:

- continuous central neuropathic pain (e.g., dysesthetic extremity pain).
- intermittent central neuropathic pain (e.g., trigeminal neuralgia, Lhermitte's sign).
- musculoskeletal pain (e.g., lower back pain, muscle spasms or pain relating to tonic spasms).
- mixed neuropathic and non-neuropathic pain (e.g. headache).

An additional source of pain derives from MS-specific drug therapies which, although not serious, may decrease compliance with treatment<sup>102</sup>.

### 3.3.12.1 Neuropathic Pain

Neuropathic pain, or ‘nerve pain’, is usually described as burning, shooting, tingling, stabbing and/or hypersensitivity. People with MS often experience neuropathic pain due to demyelination of the nerves and plaques in the brain and spinal cord. An example of this is trigeminal neuralgia, a severe facial pain, which occurs 300 times more frequently in people with MS than in the general population. In extreme cases, surgery may be performed to alleviate the pain of trigeminal neuralgia, but this may leave the face numb<sup>103</sup>. Lhermitte’s sign is another example of neuropathic pain often triggered by head movement and attributed to demyelination in the cervical area.

### 3.3.12.2 Nociceptive Pain

Musculoskeletal or nociceptive pain is the type of pain experienced when damage occurs to muscles, tendons, ligaments and soft tissue. Muscle spasm and spasticity, common symptoms of MS, can also be a source of nociceptive pain. Many people with MS experience lower back pain, especially if immobility or fatigue means that they are sitting down for much of the time. Sitting places the lower back under more strain than standing and nerves can easily become compressed or pinched. Equally, an alteration of gait may place unusual stresses on the discs between the vertebrae. Such stress can cause damage to the discs and nerves to be trapped, which results in pain in whichever part of the body is served by those nerves. Ligament damage can also occur in MS because of hyperextension of the knee when walking; the subsequent swelling of the knee can cause significant pain<sup>101</sup>.

### 3.3.13 Altered Mobility and Balance

Mobility can be defined as the ability to move freely; it includes moving in bed, getting out of bed, moving into and out of chairs, going up and down stairs and slopes, getting to and from shops and using public transport. It may also include endurance – some people with MS may retain the ability to walk but can do so only over a short distance.

Mobility is impaired in up to 90% of people with MS<sup>32</sup>. Impaired mobility affects functional activity, employment, independence, and physical and mental components of QOL<sup>32</sup>. Several studies have suggested the importance of mobility to productivity and employment. Mobility and hand function are the two largest predictors of leaving the workforce. Of factors affecting QOL in a group of patients with MS, 65% gave mobility the highest priority<sup>32</sup>. In another survey, 70% of people with MS and walking impairment reported that it was the biggest challenge associated with MS<sup>104</sup>. Patient fear about [falling](#) will increase social isolation.

#### Falling

People with MS who are afraid of falling may decrease physical activity, which can reduce strength, endurance, and range of motion<sup>105</sup>. This can in turn increase the risk of falling, which may lead to additional increase in medical costs.



Mobility is impaired in up to 90% of people with MS.

Gait problems can appear any time in the disease course. Fifteen years after diagnosis the probability of requiring assistance for walking is estimated at 40%, and of requiring a wheelchair, 25%<sup>32</sup>.

### 3.3.14 Depression

Major, or clinical, depression is a serious psychiatric disorder that is distinguished from minor depression, discouragement and grief by the persistence, severity and number of symptoms<sup>106</sup>.

A diagnosis of major depression requires an individual to have a sad or irritable mood most of the day and nearly every day for at least two weeks. In addition, there must be four other [symptoms](#) present during this time, which may include:

- loss of interest in the things one is usually interested in and capable of doing.
- feelings of worthlessness or excessive/inappropriate guilt.
- significant increase or decrease in appetite with weight gain or loss.
- change in sleep patterns (waking up 2–3 hours earlier than usual, or increase in number of hours of sleep each day, by several hours or more) recurrent thoughts of death or suicidal feelings and thoughts, fatigue.
- attentional impairment.
- sexual dysfunction.

Major depression is more common in MS patients than in the general population, or even among people with other chronic disabling diseases<sup>107</sup>. Half of all people with MS experience depression at some point in the course of their illness<sup>108</sup>. The prevalence of major depression has been estimated at 26% in those in the 18–45 age range<sup>107</sup>. Depression may be a reaction to effects of MS, a result of lesions in areas of the brain that control emotions, a result of immunologic processes in MS, a side effect of MS medications (e.g., corticosteroids, possibly interferon), or may be a separate comorbidity.

#### Symptoms

Some of the symptoms associated with major depression are also associated with MS. Consequently, it's important to differentiate between social withdrawal associated with depression (due to loss of interest in usual social activities) and the inability to socialise in one's usual manner due to MS symptoms or impairments. If the persistent and severe mood changes that characterise major depression are not present, such symptoms are most likely due to MS.



Half of all people with MS experience depression at some point in the course of their illness.

Depression, which often goes unrecognised and untreated, does not correlate with level of disability in MS<sup>109,110</sup>. It has however been shown to correlate with the degree of neurological impairment; people with MS and depression tend to have a higher lesion load in the medial orbital frontal cortex than non-depressed people with MS<sup>111</sup>. Depression has also been found to be the most important predictor of a decreased QOL in MS patients<sup>112</sup>.

The uncertainty of the future, coupled with the perceived loss of a ‘normal’ life, causes most people with MS to feel depressed at least occasionally. Researchers believe that MS-related depression may be due to a combination of the following:

- psychological reactions to the diagnosis of a chronic illness.
- neuropathology of the disease process.
- anxiety related to the uncertainty of future events.
- grieving over the perceived loss of former self.

Few diseases are as affected by emotional status as MS. Research has shown, for example, that function and performance are much better when people are in good emotional health than when they are depressed or anxious<sup>113</sup>.

The presentation of depression in people with MS frequently differs from that of the general population. In MS, symptoms such as sadness, irritability and anxiety may be the first indicators of an underlying depression. In any person with MS who is depressed, a list of possible contributing factors (such as chronic pain and social isolation) should be drawn up. Interventions should be undertaken to help resolve those contributing factors where possible. Specific antidepressant medication or psychological treatments such as cognitive behavioural therapy should be considered, but only as part of an overall programme of depression management.

### 3.4 Summary



- MS is associated with a number of symptoms, including vision difficulties, speaking and swallowing difficulties, weakness, pain, tremors, bladder, bowel and sexual dysfunction, thinking and memory problems, depression, fatigue and episodic symptoms.
- These symptoms may affect a patient’s functioning and wellbeing.
- Treatment of symptoms may improve quality of life and day-to-day functional ability.
- However, symptomatic treatments do not slow progression of the disease.



### Reflective learning point

Taking into account all of the symptoms discussed, what do you think are the most significant signs to look out for when you meet with your patients?

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How will you go about assessing your patients for any changes they might be experiencing in their symptoms?

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What is the significance of understanding the different symptoms a person with MS might encounter in your role as MS Nurse?

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## 4 Differentiating between Initial Clinical Presentation and a Relapse

### 4.1 Learning Objectives

It is important that a MS Nurse is able to distinguish the features of a relapse. This section will outline the features of a relapse and how the MS Nurse can distinguish a relapse from initial clinical presentation.

After review of this section, you should be better able to:

-  • Describe the features of an MS relapse.
- Identify key questions to ask a person with MS who you suspect is experiencing a relapse.
- Assist a person with MS to differentiate between fluctuation in symptoms, relapse and potential infection, and to understand trigger factors.

### 4.2 What is a Relapse?

A relapse is an episode of neurological symptoms (caused by inflammation or demyelination) that occurs at least 30 days after any previous episode began, lasts at least 24 hours and is not caused by an infection or other cause. A relapse is often described by other names, including an attack, exacerbation, flare-up, acute episode or clinical event<sup>114</sup>.



A relapse is an episode of neurological symptoms (caused by inflammation or demyelination) that occurs at least 30 days after any previous episode began, lasts at least 24 hours and is not caused by an infection or other cause.

During a relapse, new symptoms appear, or old symptoms re-appear, either gradually or suddenly. Symptoms usually come on over a short period of time – hours or days. They often stay for a number of weeks, usually four to six, though this can vary from only a few days to many months. Typical MS symptoms in a relapse include weakness, unsteadiness, bladder disturbance or double vision. Other symptoms of MS, such as fatigue or pain, can be more difficult to categorise as a relapse because they may not have a clear-cut beginning or end.

The frequency of relapses is generally higher early in the disease, but can vary greatly among individuals with MS. Some people will experience several relapses in a year, while others will be relapse free for many years. In one retrospective study in a population of 2,477 patients with relapsing-remitting MS, over three-quarters experienced a five-year relapse-free period<sup>115</sup>. On average, people with MS will experience approximately 0.6 relapses per year, with frequency gradually decreasing during the course of the condition<sup>116</sup>.

Improvement of symptoms for an individual, as well as the degree of recovery, is unpredictable with each relapse. Incomplete recovery has been found to range from 20% to 60% in different studies<sup>117</sup>. Relapses are due to the occurrence of inflammation in some part of the CNS and may be seen on MRI scans as lesions with active inflammation. Complete recovery from a relapse is more typical early in the disease; later in the disease course only partial improvement may be achieved, resulting in accumulation of disabling factors.

### 4.3 The Role of the Nurse

It is important in the first instance for the MS Nurse to ascertain which symptoms have changed and the time period over which the symptoms have deteriorated – typically a relapse will manifest over a few days or a couple of weeks. Symptoms that have deteriorated over a few months or longer are not indicative of a relapse and are more likely to be associated with signs of progression rather than relapse<sup>26</sup>.



It is important in the first instance for the MS Nurse to ascertain which symptoms have changed and the time period over which the symptoms have deteriorated. Symptoms that have deteriorated over a few months or longer are likely to be associated with signs of progression rather than relapse.

It is important to remember that a worsening of symptoms and changes in function may not necessarily be a relapse. Other factors can contribute to such changes (e.g., humidity, increased body temperature, [infection](#), acute or chronic stress or distress).

The worsening of symptoms may “look like” a relapse when in reality it is not. These episodes may be referred to as pseudoexacerbations or pseudorelapses. Once the underlying cause is addressed, pseudorelapses usually remit.

#### Infection

It is not unusual for people to present to the MS Nurse thinking they are experiencing a relapse, only for it to become evident on closer questioning that they have an infection. Any infection can cause a worsening of MS symptoms, although urinary tract infections are often the cause. Infections can be largely asymptomatic while still causing a flare up of MS symptoms, or the symptoms caused by the infection may be confused with those of an MS relapse.

#### 4.3.1 Questions the MS Nurse could ask a person they suspect is experiencing a relapse

- **History of onset of symptoms**

- Ask the person if the symptoms came on sub-acutely/acute.
- When did they start?
- Have they been there for at least 24 hours?
- Ask them how their overall condition is different from how they were 30 days previous.
- What are the symptoms they are experiencing?
- Are these symptoms new or have they had them before?
- Does anything make them worse?

- **How disabling are the symptoms?**

- It is important to establish how these ‘new’ symptoms affect the person on a day-to-day basis; what they can’t do now, that they could do a few days ago.
- How do these symptoms impair them at home or at work?

- **Eliminate a pseudorelapse**

- Prior to confirming a relapse, it is essential to rule out a pseudorelapse, although this is not always straightforward.
- It is essential to ensure patient does not have an infection prior to administration of steroids.
- In some centres, a routine MSU will be taken even if the patient is asymptomatic.
- If there is a recent infection, wait for this to clear before giving steroids.

- **Are the increased symptoms associated with the woman’s menstrual cycle?**

- It is known that the menstrual cycle can affect MS symptoms.
- Some women with MS report they experience increased fatigue and other symptoms approximately 7 days before and 3 days into their period.
- Some women with MS experience an increase in their symptoms associated with menopause.

- **Has the patient commenced any new medication?**

- Some drugs that are prescribed for people with MS have side effects that may mimic a relapse.
- Ask the patient if they have commenced on any new medication recently.

Lifestyle issues may be important in reducing the risk of relapses. A well-balanced diet and regular exercise will promote good health and can help reduce the risk of relapse triggers. Evidence suggests that relapses can be triggered by infections during the three-month period after giving birth and stressful life events<sup>118</sup>. Surgery, general and epidural anaesthesia, and physical trauma are not associated with an increased risk of relapses.

Experiencing a relapse is always a stressful time for people with MS and their families, and can initiate a fresh cycle of grieving and loss<sup>88,119</sup>. For many of those experiencing a relapse, this will mean they could be temporarily unable to work, or struggle to take care of their children. They may not be able to socialise or attend planned events.

People with MS will typically have a lot of unanswerable questions about when their symptoms will resolve, whether they will make a full recovery or if the relapse is the start of a more progressive phase. The social impact of a relapse cannot be ignored and MS Nurses play a particularly valuable role in supporting patients at this time<sup>26</sup>.



The social impact of a relapse cannot be ignored and MS Nurses play a particularly valuable role in supporting patients at this time.

#### 4.11 Summary



- MS is associated with a number of symptoms, including vision difficulties, speaking and swallowing difficulties, weakness, pain, tremors, bladder, bowel and sexual dysfunction, thinking and memory problems, depression, fatigue and episodic symptoms.
- Relapses are common and caused by inflammatory activity within the central nervous system.
- They are classed as fairly acute episodes, defined by an increase in symptoms lasting at least 24–48 hours occurring after a period of stability of at least 30 days.
- The extent of recovery from relapse and the duration of any given relapse are difficult to predict and this understandably causes a great deal of anxiety.
- The role of the MS Nurse in assessing the nature of the problem and providing information and support to patients and their families is key.
- The MS Nurse is also important in ruling out any other possible causes of an exacerbation of symptoms, such as an infection which may require treating in its own right.



## Reflective learning point

What are the key things to watch out for that will help you decide if the patient is having a relapse?

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How would you explain to a patient what happens to them during a relapse?

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## 5 Atypical Presentation

### 5.1 Learning Objectives

Along with the more common types of the disease, MS can also present itself in more uncharacteristic ways, of which it is important for the MS Nurse to be aware. This section will discuss the more atypical presentations of MS, along with their key distinguishing features. After review of this section, you should be better able to:



- Outline the differences between adult-onset and early-onset MS.
- Define late-onset MS.
- Describe other less common variations of MS.

### 5.2 Introduction

#### Intro

MS can be classified according to the stage of the disease course (e.g., early MS) and the age of onset (see *Table 3*). MS is uncommon in children and adolescents, and those who do suffer from it experience a slower disease course than adults.

#### Early MS

Early MS is characterised by the occurrence of one clinical flare plus paraclinical criteria that together allow the diagnosis of MS to be made. It is important to note that at this stage of disease a second flare up has not yet occurred.

#### Late-onset MS (LOMS)

Late-onset MS is defined as the first presentation of clinical symptoms in patients over 50 and its prevalence ranges between 4% and 9.6%. The disease course is often primary progressive and LOMS is associated with a higher and faster rate of progression to irreversible disability than is onset of MS in younger adults.

#### MS in children

MS is uncommon in children and adolescents – around 2-5% of people with MS experience their first symptoms before the age of 16 years. More than 90% of the paediatric MS population have RRMS. Disease course is usually slower than in adults but significant disability may still occur by early adulthood.

*Table 3. MS classified by age of onset*

### 5.3 Paediatric Presentation / Early-Onset MS

Paediatric MS has long been an under-recognised and undertreated MS subgroup. However, over the past ten years awareness has grown of the special diagnostic challenges, the clinical course and the special needs of these patients. While some aspects of the clinical disease in children resemble those of adults, MS in children can also differ dramatically from that in adults in clinical, radiological and laboratory features<sup>120</sup>.

The proportion of patients with MS aged below 16 years is estimated to be 2.7–5%<sup>121</sup>. The frequency is much lower (0.2–0.7%) among young children (aged 10 years and younger)<sup>122,123</sup>. As in adults with MS, **paediatric MS** displays an overall female preponderance. However, the gender ratio varies with age of onset: in children with onset of MS before 6 years of age, the female-to-male ratio is almost equal at 0.8:1. But the female-to-male ratio increases to 1.6:1 for an onset between 6 and 10 years of age, and even further to 2:1 for onset at over ten years of age<sup>124</sup>.

When compared to adults with MS, children with MS have a higher rate of relapse within the first two years of the condition but progress more gradually<sup>127</sup>. Although the clinical course may be favourable initially, patients with EOMS can be more disabled at an earlier age<sup>128</sup>. The risk of secondary progressive MS in children (as in adults) is associated with a higher frequency of relapses and shorter intervals between attacks in the first few years of the disease.

Children often exhibit systemic symptoms, such as malaise, irritability and/or low-grade fever, which are similar to symptoms of encephalomyelitis or metabolic encephalopathy. Children may be under-diagnosed due to the following:

- Acute disseminated encephalomyelitis is more common than EOMS.
- A multitude of CNS diseases mimic MS.
- Diagnosis is often delayed until adulthood.

Being told of the diagnosis of MS is traumatic for the child/young person and their family. The fact that the prognosis is uncertain and the condition is rare contributes to the difficulty the family faces in adjustment. A number of considerations should go into the process of conveying the diagnosis. It is important to emphasise to the family that they are not alone and that online social network groups, support groups and literature specific to the topic of paediatric MS are available.

#### Paediatric MS

There are some additional aspects of the demographic profile of paediatric MS that differ from those in adults with MS. In an outpatient centre in Boston, a higher proportion of African-Americans was found in the paediatric onset category compared to the adult onset MS group (7.4 vs. 4.3%)<sup>123</sup>. Others have noted greater ethnic diversity and ancestry in paediatric MS compared to adult MS<sup>125</sup>. A positive family history of MS is seen in 6–20% of children with the disease<sup>126</sup>.

Challenges exist for both children and parents related to dealing with the unpredictable nature of MS, behavioural changes and the potential for major disability<sup>129</sup>. Shock and dismay are common feelings that parents and caregivers experience when having to face the diagnosis of MS in a child or adolescent<sup>130</sup>. The most common age group to be affected by paediatric MS is adolescence. Sensitivity is essential when dealing with teenagers as they may be particularly vulnerable psychologically. The paucity of research available on the topic and its management can make assisting with psychosocial issues more difficult<sup>129</sup>. As with other chronic medical diseases, a sense of isolation, dependence on long-term treatment and the need for self-management may result in this group being at a greater risk of behavioural problems.



#### Nursing tip

Do you think there are different needs when supporting an adolescent recently diagnosed with MS and their family?

Adolescents recently diagnosed with MS can pose a particular challenge. Reviewing their typical needs in growth and development is of primary importance. They are often feeling the need to be very independent and may not respond well to the restrictions MS may impose as relapses occur. Being told of the diagnosis of MS is traumatic for the young person and their family. The fact that the prognosis is uncertain and the condition is rare contributes to the difficulty the family faces in adjustment. As with other chronic medical diseases, a sense of isolation, dependence on long-term treatment and the need for self-management may result in this group being at a greater risk of behavioural problems. Sensitivity is essential when dealing with teenagers as they may be particularly vulnerable psychologically.



The most common age group to be affected by paediatric MS is adolescence. Sensitivity is essential when dealing with teenagers as they may be particularly vulnerable psychologically.

## 5.4 Late onset

Although there is no general consensus, MS is currently said to be ‘late onset’ (LOMS) if the first symptoms of the disease present after 50 years of age<sup>131</sup>. The diagnosis and clinical management of LOMS is confusing, given that there are several different diseases that can present similar symptoms, including ischaemic cerebrovascular disease, the frequency of which increases with age, and cervical myelopathy of spondyloarthritic origin, considered to be the number one cause of paraesthesia over the age of 50 years<sup>132</sup>.



Although there is no general consensus, MS is currently said to be ‘late onset’ if the first symptoms of the disease present after 50 years of age.

Advanced age is no longer considered to be a diagnostic exclusion criterion for MS. A case has been published of a female patient who was 82 years of age when she presented with her first symptom of MS<sup>133</sup>. In some publications, the term ‘very late onset’ has been used to refer to cases in which the disease presents in the seventh decade of life.

Primary-progressive MS (PPMS) predominates in LOMS, affecting 55–80% of these individuals<sup>131,134</sup>, although disease progression appears similar to those with adult-onset MS<sup>134</sup>. However, it has recently been reported that women with LOMS have a different trajectory in terms of disease progression than women with adult-onset MS<sup>135</sup>. Vigilance is necessary to recognise MS in this population because of its unusual presentation<sup>131</sup>.

## 5.5 MS Variation: ‘Other’ forms of MS

### 5.5.1 Benign MS

There is increasing controversy regarding whether this particular category of MS truly exists. It is thought to occur in 5–10% of MS cases, and features complete recovery from isolated attacks, with little or no accumulation of disability. The attacks may be separated by 10 or more years. Typically, people with benign MS have a Kurtzke EDSS score of less than 3.0. A score of 3.0 indicates moderate disability in one functional system or mild disability in three or four functional systems, though the patient is fully ambulatory<sup>1</sup>. This type of MS often goes undiagnosed for several years and, in many instances, benign MS is diagnosed post-mortem.

### 5.5.2 Malignant MS (Marburg’s variant)

In addition to the MS types already mentioned, there is a variant known as malignant MS. This is a rare and severe form of MS characterised by multiple large lesions scattered throughout the CNS. The demyelination and loss of axons is much more extensive than in all other forms of MS and results in a rapid accumulation of significant disability. It is such an atypical form that diagnosis is often very difficult<sup>136</sup>. However, it will generally progress rapidly without any lasting remission and can result in death within months of onset.

### 5.5.3 Neuromyelitis Optica (NMO / Devic’s Disease)

Neuromyelitis optica (NMO; also known as Devic’s syndrome or Devic’s disease) is an inflammatory disorder with a preference for the optic nerves and spinal cord. Acute transverse myelitis is often the initial manifestation. The principal features are optic neuritis and myelitis, and a tendency to recurrence which led to its classification as a subtype of MS, but it has several unique features. These clinical events also occur commonly in typical MS, however, in NMO they are usually more acute and severe; these characteristics may raise initial diagnostic suspicion of NMO<sup>137</sup>.

Neuromyelitis optica may follow either a monophasic or relapsing course. In monophasic NMO, patients experience either unilateral or bilateral optic neuritis (ON) and a single episode of myelitis, characteristically, but not always, within a very short time of one another, but do not have further attacks. In contrast, patients with a relapsing course continue to have discrete exacerbations of ON and/or myelitis after they meet NMO diagnostic criteria<sup>137</sup>.

## 5.6 Summary



- There are a number of different forms in which MS can present, which although less common, are still important to be aware of.
- MS can present in young children and the older adult, and therefore should be considered as a possible diagnosis in these groups of patients.

## 6. Prognosis (Course of Disease)

### 6.1 Learning Objectives

MS is unpredictable in its overall course, in the type and severity of symptoms experienced by each patient, and in its long-term outcome. This section will discuss the factors that might impact upon the expected course of the disease.

After review of this section, you should be better able to:



- Specify disease progression factors and symptoms that are predictive of long-term outcomes.

### 6.2 Introduction

MS is characterised by considerable variability in prognosis between individuals diagnosed with the condition. Less than 5% of people with MS have very severe disability within the first 5 years after onset, and 10–20% of people remain unaffected without therapy for over 20 years<sup>17</sup>. In the pre-DMT era, the median time from onset to cane requirement, bedbound status and death, was approximately 15, 26 and 41 years, respectively<sup>17</sup>. The **median survival time** is approximately 5–10 years shorter for people with MS than for the age-matched general population<sup>138–140</sup>.



MS is characterised by considerable variability in prognosis between individuals diagnosed with the condition. Less than 5% of people with MS have very severe disability within the first 5 years, and 10–20% of people remain unaffected without therapy for over 20 years.

Although MS is seldom fatal, death can result from secondary complications resulting from immobility, chronic urinary tract infections, and compromised swallowing and breathing.

#### Median survival time

A Danish study, reviewing patients with an onset of MS between 1949 and 1996, found that Danes with MS can expect to live approximately 10 years less than the general age-matched Danish population<sup>138</sup>. In comparison with the general Danish population, those with MS had a higher risk for death from all causes except cancer. Improved survival rates seen over recent decades were due to improved survival in all major disease groups except cancer and cardiovascular diseases for women, and from accidents and suicide in both men and women (for which the death rate remained almost unchanged).

### 6.3 Prognostic Factors

People with new-onset symptoms of demyelinating disease generally want to know whether or not they have MS<sup>141</sup>, and those diagnosed with MS often want to know what to expect in terms of disease progression and future disability<sup>142</sup>. The first question that patients often pose to their healthcare provider is “Will I be in a wheelchair in a few years’ time?” Healthcare providers usually have difficulty answering this question. Up until the early 2000s, MS was generally viewed as a fairly rapidly progressing disease, with 50% of patients reported as needing a cane, crutch or brace to walk 100 m within 15–20 years from disease onset. More recent natural history studies, using comparable survival analysis techniques, report longer times to disability milestones<sup>143</sup>.



#### Nursing tip

What might be your response when someone is worried and anxious about disease progression and is asking questions about potential disability – e.g. will I end up in a wheelchair? Will it shorten my life?

Most patients are anxious when initially diagnosed. They fear the possibility of disability and death. Most say “I don’t want to be a burden to my family”. MS is unpredictable in its overall course, in the type and severity of symptoms experienced by each person, and in its long-term outcome. The uncertainty of prognosis is hard to deal with. Many people ask if there is any way of identifying ‘triggers’ which will cause the condition to worsen but there is very little proof that any particular event or circumstance can be identified. There is some evidence that stressful life events, such as severe emotional stress, can make deterioration more likely but this is controversial.

The nurse may best serve the patient by responding with positive suggestions including maintaining a healthy lifestyle, diet, exercise and not smoking as these are among things known to affect the impact of disability. Starting disease modifying therapy early in the disease course and remaining adherent are also very important as studies have shown a slowing of the progression of disability in those individuals who complied with their therapy. We also have many ways to approach symptoms of MS today and by being proactive we can stall the effects of disability much longer than ever before.



People diagnosed with MS often want to know what to expect in terms of disease progression and future disability.

Although there is no “crystal ball” one can use to predict the future for an individual person, prognostic factors identified in studies of patient cohorts can help clinicians assess the likelihood of a poor prognosis versus a more moderate course (see *Table 4*).

Prognostic Factors	Favourable	Unfavourable
Clinical	A few relapses in the first 5 years	A high number of relapses in the first 5 years
	Good recovery from relapses	Poor recovery after relapses
	Initial presentation of optic neuritis or sensory relapse	Initial presentation of brainstem episode or motor deficits
	Mild relapses not affecting function	Major relapses affecting function requiring steroid treatment
	No change on the EDSS	Positive change on the EDSS
	No change on the MSFC	Positive change on the MSFC
Demographic	Female	Male
	Young age	Older age
MRI	Mild brain and cervical cord involvement	Major brain and cervical cord involvement
	A few T2 lesions representing burden of disease	Multiple T2 lesions
	A few or no gadolinium-enhancing lesions (active lesions)	Multiple gadolinium-enhancing lesions (active lesions)
	A few or no T1 hypointense lesions representing axonal loss	Multiple T1 hypointense lesions (black holes)
	Minimal white matter and gray matter (cortical) involvement	Major white matter and gray matter involvement
	Minimal atrophy	Major atrophy
Spinal Cord Fluid	No oligoclonal bands or normal IgG index and synthetic rate	Oligoclonal bands and elevated IgG index and elevated IgG synthetic rate
Optical Coherence Tomography	Normal retina layer thickness in both eyes	Decreased thickness of the retina layer
	No optic axonal loss	Optic axonal loss

*Table 4.* Prognostic factors in MS<sup>14</sup>

Overall, gender does not appear to be a risk factor with regard to long-term disability in MS when other influential variables are considered, although some natural history studies suggest that male gender might be a negative factor in RRMS and SPMS<sup>144</sup>. Men are more likely than women to have rapid progression, but women have a higher rate of relapse; rates of accumulated disability appear to be comparable between men and women<sup>142</sup>.

Age at onset has some effect on the evolution of disability in MS. Late-onset MS is associated with more rapid progression in some studies, but not all<sup>134</sup>. Mean time to an EDSS score of 6 decreased as age of onset increased in one cohort<sup>145</sup>. Although younger patients have a slower progression, they become disabled at a younger age and, therefore, spend a longer span of their lives disabled<sup>141</sup>.

Characteristics of the initial attack and early disease course have been associated with long-term outcome in MS. In all MS subtypes, a worse prognosis may be more likely in patients whose initial symptoms include motor, cerebellar, brainstem, sphincter, or cognitive involvement, whereas those with sensory or visual symptoms (optic neuritis) are more likely to have a benign course<sup>134</sup>.

The likelihood of disability in RRMS/SPMS and PPMS correlates with the number of neurologic systems involved:

- Complete or nearly complete recovery from an initial attack is a good prognostic indicator; incomplete recovery is associated with poorer prognosis<sup>145,146</sup>.
- A longer interval between the first and second attack is also associated with a more favourable disease course than a shorter interval<sup>145,146</sup>.
- Early accumulation of disability as measured by higher EDSS scores in the first 5 years of disease is a strong predictor of worse prognosis
- Moderate disability (EDSS of 4.0 when not in relapse) within the first year of RRMS is also indicative of greater disability long term.

The uncertainty of prognosis is hard to deal with. Many people ask if there is any way of identifying ‘triggers’ which will cause the condition to worsen but there is very little proof that any particular event or circumstance can be identified. There is some evidence that stressful life events, such as a car accident or severe emotional stress, can make deterioration more likely<sup>118,147,148</sup>. However even this is controversial and there is usually little that can be done to prevent such stresses occurring.



## Nursing tip

How might you continue to support someone who is making a transition to more progressive MS?

People transitioning to a more progressive form of MS are often disheartened and say “There are no drugs for my type of MS”. While currently there are no DMT’s available to truly affect the progressive forms of MS, there are many symptomatic therapies which can be tried to help the person with progressive MS live the best quality of life possible. A comprehensive approach to MS in these cases is essential. Physical and occupational therapy, speech and swallowing therapists, neurocognitive rehabilitation and assistive devices are just a few of the things that can make a significant difference in the lives of a person with progressive MS.

## 6.4 Summary



- A number of prognostic factors have been identified that help predict likelihood of long-term disability.
- None of the factors described have been validated as reliable for predicting the course of an individual patient.
- It is reasonable to expect that the more negative prognostic factors a patient has, the higher the risk of a worse long-term outcome.
- This information may help clinicians identify patients for whom MS is more likely to take a more disabling course.



## Reflective learning point

How will understanding how the MS progresses help me in my day to day role as an MS Nurse?

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## Summary of Module



- Multiple sclerosis (MS) usually starts with an acute episode of neurological disturbance.
- There are four types of disease course, defined as relapsing/remitting MS, secondary-progressive MS, primary-progressive MS and progressive-relapsing MS.
- MS is associated with a number of symptoms, including vision difficulties, speaking and swallowing difficulties, weakness, pain, tremors, bladder, bowel and sexual dysfunction, thinking and memory problems, depression, fatigue and episodic symptoms.
- These symptoms may affect a patient's functioning and wellbeing.
- Treatment of symptoms may improve quality of life and day-to-day functional ability.
- However, symptomatic treatments do not slow progression of the disease.
- Relapses are common and caused by inflammatory activity within the central nervous system.
- They are classed as fairly acute episodes, defined by an increase in symptoms lasting at least 24–48 hours occurring after a period of stability of at least 30 days.
- The extent of recovery from relapse and the duration of any given relapse are difficult to predict and this understandably causes a great deal of anxiety.
- The role of the MS Nurse in assessing the nature of the problem and providing information and support to patients and their families is key.
- The MS Nurse is also important in ruling out any other possible causes of an exacerbation of symptoms, such as an infection which may require treating in its own right.
- There are a number of different forms in which MS can present, which although less common, are still important to be aware of.
- MS can present in young children and the older adult, and therefore should be considered as a possible diagnosis in these groups of patients.
- A number of prognostic factors have been identified that help predict likelihood of long-term disability.

- None of the factors described have been validated as reliable for predicting the course of an individual patient.
- It is reasonable to expect that the more negative prognostic factors a patient has, the higher the risk of a worse long-term outcome.
- This information may help clinicians identify patients for whom MS is more likely to take a more disabling course.

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