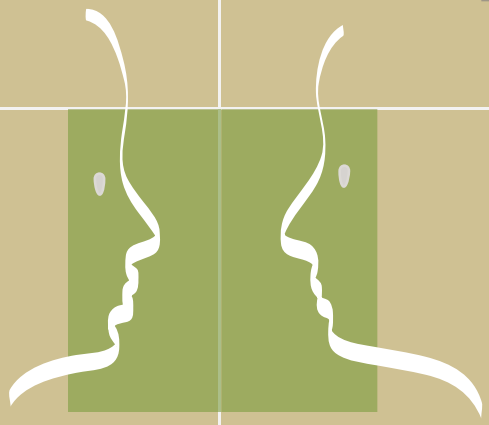
National Multiple Sclerosis Society

Talking with Your MS Patients about Difficult Topics



Talking about Primary-Progressive MS

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Professional Rx Resource Center

Rosalind Kalb, PhD, *Editor*

The National MS Society's Professional Resource Center (PRC) has long been a resource for health professionals providing care to people with multiple sclerosis. The PRC, which houses the most comprehensive library of MS information in the world, provides continuing education opportunities and offers multidisciplinary expertise on MS disease process and management questions, health insurance issues, long-term care options, and the development of MS specialty clinics. The goal of these services is to enhance quality of care and increase access to care for people with multiple sclerosis.

Physicians are invited to consult via email with MS specialist colleagues who serve on our National Clinical Advisory Board
MD_info@nmss.org

Allied health professionals are invited to consult via email with MS specialist colleagues
HealthProf_info@nmss.org

Questions are also welcome at our toll-free number
1-866-MS-TREAT (1-866-678-7328)

Visit our website:
www.nationalMSsociety.org/PRC

To receive periodic research and clinical updates via e-mail, please contact the PRC

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Introduction

This booklet is designed to facilitate conversations with your patients about primary-progressive MS (PPMS). Because there are fewer treatment options for PPMS than for the relapsing courses of the disease, and the prognosis is generally poorer, both you and your patients may find the diagnostic and management issues somewhat difficult to discuss. The information provided here can help pave the way.

Physicians, as well as nurses, rehabilitation specialists and mental health professionals, have a critical role to play in helping people learn how to manage this challenging disease course in ways that promote function, independence, hope and optimal quality of life. The first step is to establish open, comfortable lines of communication.

1. When is it appropriate to talk about the diagnosis of PPMS?

As soon as you have confirmed that the person has multiple sclerosis, and determined that the course appears to be primary-progressive, it's important to share that information with your patient. A clear and open conversation about the diagnosis sets the stage for an effective doctor-patient collaboration to manage the disease.

According to the 2005 Revised McDonald Criteria (Polman et al., 2005), PPMS is defined by at least one year of insidious neurologic progression (i.e., without any acute attacks) *and* two out of three of the following:

- Positive brain MRI (nine T2 lesions *or* four or more T2 lesions with positive visual evoked potentials)
- Positive spinal cord MRI (two or more focal T2 lesions)
- Positive CSF

In order to arrive at this diagnosis, a very careful medical history must be taken to identify any past symptoms or neurologic events that might suggest that the current course is secondary-progressive rather than primary-progressive MS. Engaging both the patient and close family members in this process is essential for obtaining the best possible historical information.

Once the diagnosis of PPMS has been made, it is important to be alert for any acute neurologic events that might signal progressive-relapsing MS (PRMS) rather than PPMS—an important distinction because the treatment options would be somewhat different.

2. What is the most important information to share with my patients about PPMS?

Patients need to know that approximately 10% of people with MS experience a primary-progressive course, which is characterized by steady worsening of neurologic functioning, without any distinct relapses (also called *attacks* or *exacerbations*) or periods of remission. It's important to emphasize that the rate of progression differs significantly from one person to another and each person's rate of progression may vary

over time—but that the progression is continuous, with or without occasional plateaus or apparent improvements along the way. Patients with PPMS need to be reassured that although there are currently no FDA-approved medications for the treatment of PPMS, there is still much that can be done to manage the symptoms; enhance mobility, safety and independence; and promote wellness, emotional well-being and quality of life.

3. How can I explain the differences between PPMS and other forms of MS without causing undue alarm and anxiety?

Although there is a lot of variability among people with PPMS, we know that as a group, they differ in several ways from people with relapsing forms of MS:

- Relapsing MS is defined by inflammatory attacks on myelin in the central nervous system (CNS), during which activated immune cells cause small, localized areas of damage that cause symptoms to abruptly appear or worsen. PPMS involves much less inflammation of the type seen in relapsing MS and more of a gradual progression of symptoms related to degeneration of nerve cells. People with PPMS tend to have fewer brain lesions (also called plaques) than people with relapsing MS, and the lesions tend to contain fewer inflammatory cells. People with PPMS also tend to have more lesions in the spinal cord than in the brain. Together, these differences make PPMS more challenging to diagnose and treat than relapsing forms of MS.
- In the relapsing forms, women are affected 2–3 times as often as men; in PPMS, the sex ratio is 1:1.
- The average age of onset is approximately 10 years later in PPMS than in relapsing MS.
- People with PPMS tend to experience more problems with walking than other people with MS, but less difficulty with other symptoms such as cognitive impairment or visual problems.

Fortunately, there are many strategies available to help people manage their symptoms so that they can remain active and productive.

4. When is the appropriate time to talk with family members?

With your patient's consent, it is important to engage family members as early as possible, particularly since their help will be crucial to the history-taking process. It often happens that a family member remembers a past symptom or event that the patient has forgotten or dismissed as being of no importance. If your patient is initially unwilling to involve family members, you may gradually be able to convince him or her of the value of having family participation and support, and of offering information and support to their loved ones who share the impact of this chronic, unpredictable disease.

5. What is the best format for providing this type of information?

Information about the diagnosis of MS should always be given in person, in a manner that takes into account the patient's intellectual level and emotional state. Without that face-to-face interaction, you won't be able to gauge your patient's reactions or the degree to which he or she has understood what you are saying. Keeping in mind that people who are anxious and upset tend to miss much of what is being said to them, it's a good idea to avoid giving too much information at one time. Scheduling a follow-up appointment within the next few weeks will give the person an opportunity to absorb the news and return with questions and concerns. You might also want to encourage your patient to bring a family member or friend to take notes and act as a second pair of ears. And sending your patient home with print materials about PPMS can also be helpful. Your chapter of the National MS Society (800-344-4867) will be happy to provide you with brochures. Additional information about PPMS is available for your patient on the Society's Web site at www.nationalMSsociety.org/PPMS.

6. What kinds of emotional reactions might I expect from my patients and their family members?

Receiving a diagnosis of MS is frightening enough; the diagnosis of PPMS is even more so. The word "progressive" is easily interpreted as disaster waiting to happen. And the lack of approved medications to treat this disease course causes even more alarm, particularly when physicians don't remind their patients about the available symptom management and rehabilitation strategies. When the message from the physician is heard as "Diagnose and adios"—a phrase coined many years ago by Dr. Labe Scheinberg—patients and family members are likely to feel abandoned, anxious and angry. The best way to support and reassure them is to provide information about the treatment options, along with some assurances that you will work with them to manage the disease and its challenges. This is also an opportune time to refer them to their National MS Society chapter for education and support programs. And, if needed, the Society can provide a referral to a mental health professional with expertise in chronic illness.

7. How can I best answer my patients' questions about the prognosis of PPMS?

With a chronic, unpredictable illness, the message should always be honest but hopeful. Your willingness to be frank and open about potential challenges will lay the groundwork for an effective and trusting working relationship. You can promote hopefulness in your patients in several ways: by emphasizing that every person with MS is different, and that only time will tell whether the progression will be gradual or more rapid; by assuring them that you will work with them to manage the disease and its challenges; and by reminding them that productivity and function are possible even if mobility is affected.

As the disease progresses, patients and family members can easily become overwhelmed by fears of the future. The best strategy is to listen to their concerns and help them connect with the professionals who can best support their efforts to plan and problem-solve effectively.

The National MS Society can provide referrals to employment counselors, home care providers, care managers, financial planners, elder care attorneys, and other community resources—and offer information about long-term care options, as needed.

8. What should I say about treatment of PPMS?

At the present time, there are no medications that have been approved by the U.S. Food and Drug Administration (FDA) for the treatment of PPMS. Because all of the approved disease-modifying therapies work primarily by reducing inflammation in the CNS, they do not work as well in a disease course that is characterized by nerve degeneration rather than inflammation.

Even without an approved disease-modifying therapy, however, there is a lot that people with PPMS and their health care providers can do to manage the disease, including:

- Managing the symptoms
- Enhancing mobility and promoting safety and independence
- Promoting overall health and wellness
- Promoting emotional well-being and quality of life

In talking about these essential treatment strategies, it will be important to inform your patients about the role of other clinicians in their care—nurses, rehabilitation professionals, speech/language pathologists and mental health specialists, among others—and help them access these clinicians in the community. The National MS Society can also help to provide these referrals.

9. What might be the rationale for prescribing one of the disease-modifying therapies off-label for a patient with PPMS?

Any decision about off-label treatment must be made on a case-by-case basis, taking into account your assessment of the patient's condition, his or her wishes, and the willingness of the insurance company to pay for it. Sometimes, for reasons that aren't clear, a person with PPMS does seem to experience a slowing of disease progression with medication, so you may decide that it's worth trying a disease-modifying therapy for a period of time.

When considering off-label treatment, it is helpful to have an open discussion of the associated costs and relative risks and benefits. While the potential risks associated with immunosuppressants or monoclonal antibodies might be too great given the lack of supportive evidence for their use in PPMS, the relatively low risks associated with the injectable medications might be acceptable.

10. What might be the rationale for *not* prescribing one of the disease-modfiying therapies off-label for a patient with PPMS?

It is important to be able to say "no" to a patient's request for a treatment if the possible benefits are minimal, the risks are higher—for example, in an older patient with long-standing PPMS who asks to be put on natalizumab—and the costs are unjustified. Saying "no" can be extremely difficult in the face of a patient's emotional demands—particularly when the person argues that it's his or her choice to assume the risk and costs and not yours. Under these circumstances, it's often helpful to refocus the conversation to emphasize other important aspects of treatment that you plan to pursue. Ultimately, the patient needs to trust your medical judgment or seek out another physician.

11. Why won't most insurance companies pay for a disease-modifying therapy for PPMS?

People who are diagnosed with PPMS are understandably disappointed and frustrated by the unwillingness of insurance companies to cover the costs of medications that other people with MS are taking. While it may not solve the problem of how unfair it feels, it will help them to understand that the insurance industry relies on certain facts—a drug's cost, data from pivotal trials, and FDA recommendations—to decide which medications to cover for which patients. The unfortunate fact that the existing medications do not seem to be effective in PPMS is sufficient reason for insurers to decide not to cover them for this disease course.

12. Why has so little research been done in PPMS compared to the other MS disease courses?

People often ask why so few clinical trials have been done in PPMS compared to the large number in relapsing-remitting MS (RRMS). In answering their questions, it's helpful to say that MS clinicians and researchers share this frustration and are actively looking for potential new therapies to study for PPMS.

Several obstacles have stood in the way:

- The immune-modulating drugs currently used to treat relapsing forms of MS are all primarily anti-inflammatory, and therefore not as effective in PPMS, which involves much less inflammation than the relapsing forms.
- In PPMS, there is a lack of easily identifiable outcomes to measure in clinical trials. In the trials for the approved disease-modifying therapies (the interferon beta medications, glatiramer acetate, natalizumab and mitoxantrone) investigators looked at outcomes such as number of relapses and number of new lesions seen on MRI to determine if people who received the treatment had lower numbers than those who received a placebo (non-active substance). In other words, the investigators looked at things they could easily count over the course of a two or three-year trial. Because people with PPMS don't experience relapses or

the same kind of inflammation in the central nervous system, there are fewer events that can be counted.

- The disease progression that occurs in PPMS can be quite slow—which means that a trial would have to last many years to determine if a treatment slowed or halted that progression.

Researchers are working to identify other ways to measure the changes that occur in PPMS so that they can more easily test potential treatments.

13. When it is appropriate to start rehabilitation interventions?

Rehabilitation, including physical and occupational therapy, is a key component of MS management. In an ideal world, every person living with MS would have ongoing guidance about exercise strategies, energy conservation, symptom managment, assistive technology of all kinds, environmental adaptations, and strategies to deal with any cognitive changes they might experience. However, limitations on insurance coverage will likely make it necessary to save rehabilitation visits for specific time-limited interventions, including: early in the disease course to learn energy-management strategies or develop an at-home exercise program; as the disease progresses to maintain function or learn how to use mobility aids or other forms of assistive technology; as new symptoms develop, such as problems with speech or swallowing, or changes in cognitive abilities.

The key message to patients is that there are a variety of specialists available to help them manage the various kinds of symptoms and changes that can occur over the course of the disease.

14. How can I best assist my patients to deal with disease progression?

Patients tend to be more confident and less anxious when they feel they have a strong relationship with their doctor. You can facilitate that connection by:

- Seeing them on a regular basis (every six months or a year)
- Taking time to respond to their questions and concerns
- Assuring them that you will work with them over the long haul to manage problems that arise
- Focusing proactively on symptom management, including medication, rehabilitation and assistive technology
- Paying close attention to the patient's mood and family issues, and providing referrals to mental health professionals for diagnosis and treatment
- Encouraging the development of an active support network, including connection with the National MS Society and other MS organizations
- Avoiding any message that suggests you have nothing more to offer. Your patient and the patient's family need to know you will be there with them no matter what.

15. What recommendations should I be giving about financial and life planning?

- Have a frank discussion with your patients about the status of their disease and what may happen in the future.
- Refer your patients to their National MS Society chapter for assistance with future planning. The chapter will encourage and support their efforts to:
  - Take a "financial inventory" early in the course of their disease (What are their assets and debts? What about expenses that might now need to be added to their budget, such as medications or physical therapy?).
  - Explore and understand protections that exist for someone who has a pre-existing condition and may want or need to change employment. It is no longer necessary to feel locked into a job to maintain health insurance. Once someone has had group coverage for a year, he or she may have met any requirements relating to pre-existing conditions and be able to switch into another group plan without a penalty or waiting period.
  - Look at options for assistance, e.g., community services, benefits and entitlements that could help address their needs in the future. The best time to be an educated consumer is not when one is in crisis, but when one can take the time to learn about options and gather information for future reference.
  - Refer them to financial planners and attorneys with expertise in chronic illness and disability.
- In your ongoing conversations with patients, encourage them to maintain open lines of communication within their families, thinking together about what would happen if MS imposed changes in the future. Questions to consider include: What if they require assistance with personal care in the future? How available are family members? What other life demands are competing with the needs of the family member with MS? Is the home accessible?

16. Why is it so difficult to find an effective treatment?

The disease-modifying medications we have available today are primarily anti-inflammatory, which means that they do not work as well for a disease course that is more degenerative than inflammatory. The good news is that researchers are working to identify ways to measure the changes that occur in PPMS so that they can more easily test potential treatments. People who are interested in learning about ongoing clinical trials in PPMS can visit the National MS Society Web site at http://www.nationalMSsociety.org/research/ClinicalTrials.

Alongside these efforts to identify effective medications to slow the progression of PPMS, researchers are looking for ways to protect and repair brain tissue that is damaged or lost in MS. The Nervous System Repair and Protection Initiative, funded through the Society's Promise: 2010 Campaign, has brought together four teams of researchers in the U.S. and Europe to study possible mechanisms for preventing damage to brain tissue and restoring function in those individuals who have already experienced significant tissue loss.

Progress is being made with symptom management as well. Sustained-release fampridine—a medication that has been shown in clinical trials to improve walking speed—is moving its way through the new drug development process.

17. What other resources are available?

An important message for people living with PPMS is that they don't need to face it alone. You can help your patients get the support they need by encouraging them to take advantage of available resources:

- **Organizations**
  - **National MS Society** (800-344-4867; www.nationalMSsociety.org)
    - Information Resource Center
    - Publications
      - *Momentum* magazine
      - *Adapting: Financial Planning for a Life with Multiple Sclerosis*
      - *Managing Progressive MS*
      - *A Guide for Caregivers*
      - *At Home with MS: Adapting Your Environment*
      - *So You Have Progressive MS?*
    - On-line programming (Webcasts, podcasts, online classes)
    - Support groups and chat opportunities
    - Financial planning consultations
    - Financial assistance
    - Employment consultations
    - Clinical trial information
  - **Multiple Sclerosis Association of America** (800-532-7667; www.msassociation.org)
    - Reassurance call program
    - Equipment distribution
    - Cooling equipment distribution
    - Barrier-free housing
    - Brochures
    - Lending library
  - **Multiple Sclerosis Foundation** (800-MSFOCUS; www.msfacts.org)
    - Support groups
    - MS Cruise for a Cause
    - Publications
    - Online services
    - Internet helpline
  - **The Heuga Center for Multiple Sclerosis** (800-367-3101; www.heuga.org)

    The Heuga Center is a nonprofit organization that offers MS programs based on the philosophy that people can have a chronic disease and also maintain their health. The programs, which emphasize MS education, exercise, nutrition, stress management and life management skills, help people set personal life goals and give them the strategies, confidence and support to strive for those goals.
- **Books** (from Demos Medical Publishing—800-532-8663; www.demosmedpub.com)
  - Coyle P, Halper J. *Living with Progressive Multiple Sclerosis: Overcoming the Challenges* (2nd ed.), 2008.
  - Holland N, Halper J. *Multiple Sclerosis: A Self-Care Guide to Wellness* (2nd ed.), 2005.
  - Kalb R (ed.) *Multiple Sclerosis: The Questions You Have; The Answers You Need* (4th ed.), 2008.

- Kalb R (ed.) *Multiple Sclerosis: A Guide for Families* (2nd ed.), 2006.
- Karp G. *Life on Wheels: The A to Z Guide to Living Fully with Mobility Issues,* 2008.
- LaRocca N, Kalb R. *Multiple Sclerosis: Understanding the Cognitive Challenges,* 2006.
- Northrop D, Cooper S, Calder K. *Health Insurance Resources: A Guide for People with Chronic Disease and Disability* (2nd ed.), 2007.
- Peterman Schwarz S. *Multiple Sclerosis: 300 Tips for Making Life Easier* (2nd ed.), 2006.
- Rumrill P, Hennessey M, Nissen S. *Employment Issues and Multiple Sclerosis* (2nd ed.), 2008.
- Schapiro R. *Managing the Symptoms of Multiple Sclerosis* (5th ed.), 2007.
- Shadday A. *MS and Your Feelings: Handling the Ups and Downs of Multiple Sclerosis,* 2007.
- Shenkman M. *Estate Planning for People with a Chronic Illness or Disability,* 2008.

Andrew D. Goodman, MD is Professor of Neurology, Chief of the Neuroimmunology Unit, and Director of the Multiple Sclerosis Center at the University of Rochester. Dr. Goodman is a graduate of Rutgers College and the University of Medicine and Dentistry of New Jersey. He did residency training in internal medicine and neurology at Mt. Sinai Medical Center in New York City and completed a research fellowship in the Neuroimmunology Branch at the National Institutes of Health, Bethesda, Maryland. Numerous publications reflect his interests in the immunology and virology of multiple sclerosis as well as clinical and experimental therapeutics research. Dr. Goodman has been principal investigator, site-principal investigator, or on national and international steering committees for numerous clinical trials of new therapies for multiple sclerosis. Dr. Goodman is a past Chair of the Multiple Sclerosis Section of the American Academy of Neurology. He has served the National Multiple Sclerosis Society as Deputy Medical Officer, Co-Chair of the Long-term Care Committee, member of the Client Education Committee, and member of the Executive Committee of the National Clinical Advisory Board.

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