



## Primary Progressive Multiple Sclerosis

### What is primary progressive multiple sclerosis?

Multiple sclerosis (MS) is a disorder of the central nervous system that affects about 400,000 people in the United States. Most people with MS have relapsing symptoms and experience episodes lasting days or weeks of symptoms, such as:

- numbness or tingling
- weakness of a leg or arm
- unsteadiness
- bladder difficulty
- visual symptoms

About one in ten people with MS do not have a relapsing course of the disease. These patients tend to have symptoms that gradually progress without typical relapses. Patients with the gradually worsening disease are considered to have primary progressive multiple sclerosis or PPMS.

### How is primary progressive multiple sclerosis diagnosed?

First steps in diagnosing PPMS may combine a neurological history and exam with results from testing, primarily MRI imaging of the brain and spinal cord, as well as studies such as evoked potentials and spinal fluid testing.

The story of a gradually progressive neurological problem--such as weakness on one side, unsteadiness, or numbness in the legs--is characteristic. The condition must have been present for one or more years to make this diagnosis. The examination should show changes suggesting central nervous system disease. The MRI should show lesions that are characteristic of multiple sclerosis. The spinal fluid often, but not always, shows oligoclonal banding. This is a sign that the immune system is active around the brain and spinal cord. The evoked potentials may show slowing in nerve conduction in the central nervous system. There should be no other diagnosis to cause the symptoms.

### What other diseases can mimic primary progressive multiple sclerosis?

There are other diseases that can look like PPMS and need to be considered. For example, some people inherit a tendency for stiffness and weakness in the legs; a family history may be helpful. Some people with low vitamin B12 levels may develop numbness and stiffness in the legs and will show changes in their MRI scans. Occasionally, Lyme disease may look like MS. Some people with viral infections such as HTLV-1 may have a spinal cord syndrome. Sometimes discs or spinal arthritis may compress the spinal cord, or a tumor may be pushing on the spinal cord. Other diagnoses should be considered when the diagnosis of PPMS is being made.

### **What causes primary progressive multiple sclerosis?**

Just as with multiple sclerosis in general, the cause of PPMS is not known. It is thought that the disease occurs in people who are susceptible to it due to some kind of genetic tendency. It is believed by some to be caused by a trigger, perhaps a virus. However, no specific cause for PPMS is known. PPMS, however, is not infectious.

### **What is the prognosis for primary progressive multiple sclerosis?**

As with multiple sclerosis in general, the prognosis varies in PPMS. Most people start to have symptoms at about age 40 or older, or about 10 years later than is typical of relapsing forms of MS. A recent Canadian study showed that many people with PPMS were able to walk years after a diagnosis, but usually patients tended to get worse over time. This worsening usually affected the ability to walk and was less likely to cause visual symptoms or tremor. Generally, thinking, memory, and intellect are relatively spared with PPMS. People who have PPMS may also have bowel, bladder, and/or sexual problems.

### **Is there any treatment for primary progressive multiple sclerosis?**

At this time, there is no FDA-approved treatment for PPMS. Research studies usually focus on medications for the relapsing forms of MS. There have only been a handful of treatment studies specifically for PPMS; the results so far have not shown a significant treatment effect. The standard FDA-approved medications for MS (interferons, glatiramer acetate, mitoxantrone, natalizumab) have not been proven useful in slowing the progression of PPMS.

In terms of making physical function as good as it can be, people with PPMS will be helped by exercise, stretching, physical, and occupational therapy. Maintaining mobility and fitness are as important in people with

PPMS as with anyone else. In addition, there are medications which may be used to treat symptoms such as bladder and bowel urgency, erectile problems, spasticity, and pain, if such treatments are needed.

Occasionally, intermittent (on and off) intravenous (IV) steroids have been tried in patients with primary progressive multiple sclerosis. Such therapies have provided only limited results in these cases. Also, the chemotherapeutic drug methotrexate has been given in weekly oral doses to patients with PPMS. Results from research trials into new medications for primary progressive MS are still pending.

### **Where can I find additional information on primary progressive multiple sclerosis?**

You may find the following web sites and/or organizations to be useful:

- [www.nmss.org](http://www.nmss.org) (National Multiple Sclerosis Society)
- [www.clevelandclinic.org/mellencenter](http://www.clevelandclinic.org/mellencenter) (Mellen Center)
- [www.MSAA.org](http://www.MSAA.org) (Multiple Sclerosis Association of America)