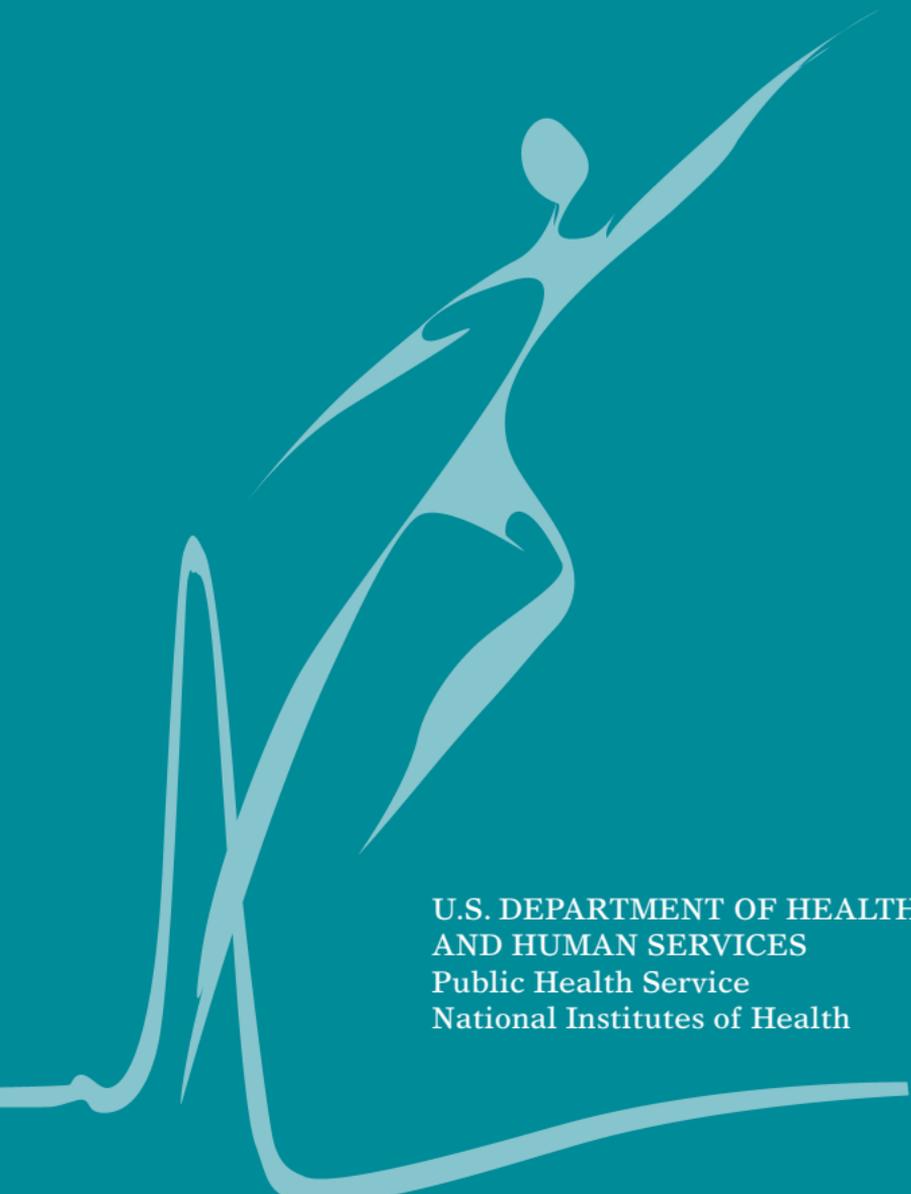


Multiple System Atrophy



U.S. DEPARTMENT OF HEALTH
AND HUMAN SERVICES
Public Health Service
National Institutes of Health



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What is multiple system atrophy?

Multiple system atrophy (MSA) is a progressive neurodegenerative disorder characterized by symptoms of autonomic nervous system failure such as fainting spells and bladder control problems, combined with motor control symptoms such as tremor, rigidity, and loss of muscle coordination. MSA affects both men and women primarily in their 50s. The disease tends to advance rapidly over the course of 9 to 10 years, with progressive loss of motor skills, eventual confinement to bed, and death. There is no remission from the disease. There are currently no cures.

What causes MSA?

The cause or causes of MSA are unknown, but the symptoms reflect the dysfunction and eventual loss of nervous system cells in several different areas in the brain and spinal cord that control the autonomic nervous system and coordinate muscle movements. The hallmark of MSA is the buildup of a protein called alpha-synuclein in oligodendrocytes, a type of glial (support) cell. MSA and Parkinson's disease are called *synucleinopathies* because they share a similar buildup of alpha-synuclein and cause loss of the dopamine-producing cells that control motor movements.

What are the common signs or symptoms?

The initial symptoms of MSA are often difficult to distinguish from the initial symptoms of Parkinson's disease and include:

- fainting or lightheadedness due to orthostatic hypotension, a condition in which blood pressure drops rapidly when rising from a seated or lying down position;
- bladder control problems, such as a sudden urge to urinate, difficulty emptying the bladder completely, or, for men, difficulty achieving an erection;
- clumsiness, loss of balance, and frequent falls;
- slurred speech, a croaky, quivering voice, or difficulty swallowing; and
- decreased spontaneous movement, tremor, or rigid muscles.

Doctors divide MSA into two different types, depending on the most prominent symptoms at the time an individual is evaluated:

- the *parkinsonian type* (MSA-P) has primary characteristics of Parkinson's disease, such as moving slowly, stiff muscles, and tremor, along with problems of balance, coordination, and autonomic nervous system dysfunction; and
- the *cerebellar type* (MSA-C), with primary symptoms featuring difficulty swallowing,

slurred speech or a quavering voice, along with ataxia (problems with balance and coordination).

Some people with MSA may start out with the symptoms of one type and then develop increasingly more severe symptoms of the other type as the disease progresses. Most people with MSA will require an aid for walking, such as a cane or walker, within a few years after symptoms begin.

Additional symptoms of MSA include:

- *contractures* in the hands or feet that prevent the joints from moving freely;
- *Pisa syndrome*, an abnormal posture in which the body appears to be leaning to one side like the Leaning Tower of Pisa;
- *disproportionate antecollis*, in which the neck bends forward and the head drops down;
- *deep, uncontrollable sighing or gasping*;
- *inappropriate laughing or crying*.

How is MSA diagnosed?

Making a positive diagnosis of MSA is difficult, particularly in the early stages, because it so closely resembles Parkinson's disease or one of the many other ataxia disorders.

After taking a clinical history and performing a brief neurological examination, a doctor may order a number of tests to help make the diagnosis. These tests include

autonomic function tests, sphincter electromyography, urine analysis (and other tests to assess bladder function), and magnetic resonance imaging (MRI), which may reveal characteristic patterns of damage in the brain. People with MSA typically don't have any improvement in their symptoms if they are given the anti-Parkinson drug levodopa. However, this lack of response isn't a strong enough indicator, by itself, to make a positive diagnosis of MSA. Unfortunately, the only definitive diagnosis for MSA comes at an autopsy of brain tissue after death; however, doctors often use criteria developed under the auspices of the National Institute of Neurological Disorders and Stroke (NINDS) to help in the diagnosis of "definite," "probable," or "possible" MSA.

How is it treated?

There is no cure for MSA. Currently, there are no treatments to delay the progress of neurodegeneration in the brain. But there are treatments available to help people cope with some of the more disabling symptoms of MSA.

The fainting and lightheadedness from orthostatic hypotension is often treated successfully with simple interventions such as adding extra salt to the diet and avoiding heavy meals and alcohol. Some people with MSA sleep with the head of the bed tilted up or use a compression body stocking. The drugs fludrocortisone (for those on a high

salt diet) and midodrine are sometimes prescribed to increase blood volume and narrow blood vessels. Drinking a glass or two of water before getting out of bed in the morning can also help raise blood pressure.

Bladder control problems are treated according to the nature of the problem. Anticholinergic drugs, such as oxybutynin, may help reduce the sudden urge to urinate. Limiting fluid intake after the evening meal and taking desmopressin at night can reduce episodes of night-time bedwetting.

Impotence and erectile dysfunction are treated in several ways, including injections, suppositories, or implants. The drug sildenafil is effective for erectile dysfunction but may worsen orthostatic hypotension.

Difficulties with swallowing and breathing eventually require that people with MSA use an artificial feeding tube or breathing tube.

Muscle spasms and contractures usually benefit from physical therapy that builds strength and encourages people to remain mobile for as long as possible.

What research is being done?

The NINDS supports research about MSA through grants to major medical institutions across the country. A great deal of research is ongoing to learn why synuclein buildup occurs in MSA and Parkinson's disease, and how to prevent it.

Where can I find more information on MSA and Parkinson's disease?

For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN

P.O. Box 5801
Bethesda, MD 20824
800-352-9424
www.ninds.nih.gov

The following organizations support MSA research and in some cases may be able to provide additional information:

Dysautonomia Foundation

315 West 39th Street
Suite 701
New York, NY 10018
212-279-1066
www.familialdysautonomia.org

Familial Dysautonomia Hope Foundation, Inc.

121 South Estes Drive
Suite 205D
Chapel Hill, NC 27514
919-969-6636
www.fdhope.org

National Dysautonomia Research Foundation

P.O. Box 301

Red Wing, MN 55066-0301

651-267-0525

www.ndrf.org

**Shy-Drager/Multiple System Atrophy Support
Group, Inc.**

P.O. Box 279

Coupland, TX 78615

866-SDS-4999 (737-4999)

www.shy-drager.com



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