

## DYSAUTONOMIA INTERNATIONAL

# Summary of Multiple System Atrophy



AWARENESS



ADVOCACY



ADVANCEMENT

Multiple System Atrophy (MSA, also known as Shy-Drager Syndrome) is a rare neurological condition that causes Parkinson's-like symptoms, however MSA patients have more widespread autonomic nerve damage than typical Parkinson's patients. Since MSA can cause widespread neurological damage, it may cause diverse symptoms throughout the body. MSA is one of three conditions of primary autonomic failure, the other two being PAF (Pure Autonomic Failure) and Parkinson's Disease.

Physicians often classify MSA as either MSA-P or MSA-C. MSA-P patients have predominantly Parkinsonian-like symptoms: tremors, muscle rigidity and slowness of voluntary movements. MSA-C patients predominantly show signs of cerebellar dysfunction: gait and limb ataxia (ataxia being a lack of control of muscle movements).<sup>1</sup>

### **Symptoms**

Not every patient has every symptom of MSA. Each patient is unique.

Some of the early symptoms of MSA may include:

- Neurogenic Orthostatic Hypotension (a drop in blood pressure upon standing that can result in lightheadedness, dizziness and even fainting)
- headaches
- abnormally dilated pupils
- loss of bladder control
- dry eyes, dry mouth, dry skin
- impotence

Some of the later stage symptoms may include:

- sleep apnea
- heart arrhythmias
- difficulty speaking
- slow speaking
- stridor (a loud noise during inhalation)
- difficulty eating, swallowing or chewing food
- monotone voice
- low volume of voice

Symptoms that may be seen at varying times during the course of the illness:

- staring with the eyes
- muscle aches and rigidity
- movement difficulties
- cognitive impairment
- loss of sweating in areas of the body
- difficulty digesting foods
- confusion
- depression
- unsteady gait
- difficulty starting any voluntary movement
- loss of fine motor skills
- loss of balance
- difficulty sleeping
- loss of bowel control
- nausea
- vision changes, decreased or blurry vision
- dementia
- unstable or stooped/slumped posture
- difficulty bending arms and legs
- finger-thumb rubbing (pill rolling tremor)

- tremors, which may worsen with excitement, stress or fatigue
- unusual or reduced ability to make facial expressions

### **Who Develops MSA?**

MSA is most often seen in persons over the age of 50, with a slightly higher incidence in men. However, women and younger patients can develop it. Estimates are that approximately 25,000 to 100,000 Americans have MSA at any given time.

### **What Causes MSA?**

It is not known what causes MSA. Researchers are currently investigating this.

### **How is MSA Diagnosed?**

MSA can be a challenge to diagnose. MSA is frequently confused with Parkinson's Disease, Pure Autonomic Failure (PAF), or Progressive Supranuclear Palsy. The top practitioners and researchers in the field have prepared a Consensus Statement on the Diagnosis of Multiple System Atrophy.<sup>1</sup> The work-up may include autonomic testing, EMG testing, blood tests, sleep studies, catecholamine testing, and biopsies. Often, MSA can only be confirmed during an autopsy.

### **Treatment**

There is currently no cure for MSA and no treatments proven to slow the neurological degeneration associated with MSA. However, there are treatments to help manage some of the symptoms, and to help the patient live the fullest life possible. Current therapeutic strategies are primarily based on dopamine replacement and improvement of autonomic failure.<sup>6</sup> Physicians may prescribe medications to help control the patient's blood pressure, gastric motility, sleeping difficulties, tremors, depression, pain and other symptoms. Medications such as Florinef, Midodrine and Sinemet are commonly prescribed.

### **Prognosis**

Unfortunately, MSA usually progresses rapidly over a period of 7 to 10 years, with the mean survival rate of 9.3 years from the time of the first symptom. About 80% of patients are disabled within 5 years of the onset of motor symptoms. It is estimated that only 20% of MSA patients survive beyond 12 years. Patients continue to experience neurological degeneration until they lose motor skills, become confined to bed, and eventually pass away. Many MSA patients succumb to pneumonia and other respiratory infections, choking or cardiac arrest.<sup>2</sup> MSA does not go into remission and there is no cure at this time.<sup>1-6</sup> There are considerable genetic research and clinical investigations taking place to improve quality of life and treatment options for MSA patients at this time.<sup>5</sup>

### **Sources**

1. [Consensus statement on the diagnosis of multiple system atrophy. Journal of the Neurological Sciences.](#) S. Gilman, P.A. Low, N. Quinn, A. Albanese, Y. Ben-Shlomo, C.J. Fowler, H. Kaufmann, T. Klockgether, A.E. Lang, P.L. Lantos, I. Litvan, C.J. Mathias, E. Oliver, D. Robertson, I. Schatz, G.K. Wenning; Volume 163, Issue 1; Pages 94-98; 1 February 1999
2. [National Institutes of Health Autonomic Disorders Consortium](#)
3. J Neurol Sci. 012 Mar 13. [Epub ahead of print]  
[The role of autonomic testing in the differentiation of Parkinson's disease from multiple system atrophy.](#) Kimpinki K, Iodice V, Burton DD, Camilleri M, Mullan BP, Lipp A, Sandroni P, Gehrking TL, Sletten DM, Ahlskog JE, Fealey RD, Singer W, Low PA.
4. Neurol Neurosurg Psychiatry. 2010 Dec;81(12):1327-35. Epub 2010 Jul 26. [Autonomic innervation in multiple system atrophy and pure autonomic failure.](#) Donadio V, Cortelli P, Elam M, DiStasi V, Montagna P, Holmberg B, Giannoccaro MP, Bugiardini E, Avoni P, Baruzzi A, Liguori R.
5. Neurobiol Aging. 2011 Oct;32(10):1924.e5-14. Epub 2011 May 24. [Genetic players in multiple system atrophy: unfolding the nature of the beast.](#) Stemberger S, Scholz SW, Singleton AB, Wenning GK.
6. Ther Adv Neurol Discord. 2010 Jul;3(4):249-63. [Multiple system atrophy: current and future approaches to management.](#) Flabeau O, Meissner WG, Tison F.