

## Stages of Multiple System Atrophy (MSA)

MSA is an adult-onset, progressive neurodegenerative disease that affects movement and autonomic nervous system functions. Issues with movement can be the slow and small movements that are typically seen in parkinsonisms and lack of coordination. The autonomic nervous system is responsible for normal function of involuntary bodily processes, like blood pressure and heart rate, urinary function and sexual function. MSA interrupts the autonomic nervous system, which can lead to problems outlined in the stages below.

As a progressive disease, MSA progresses over time and the distinctions between stages aren't always clear. There are some notable changes that happen throughout the disease, and those are often marked as four main "stages". Knowing what may lie ahead helps to plan and prepare as symptoms and care needs progress over time.

It's important to remember that everyone with MSA experiences the disease differently. Symptoms and their severity vary widely, and someone can be in each stage from anywhere between 1 to 5 years. The symptoms and the impact on daily life can also depend on the specific symptoms of MSA that a person is living with. These differences make it difficult to compare one disease journey to another. For example, some middle or late-stage symptoms may start in the early stage, while others may not experience typical early onset symptoms until later. However, knowing what may lie ahead can inform planning and preparation as symptoms and care needs progress over time.

### Early Stage

The early stage of MSA is when people begin to experience symptoms. Some people in the early stage are bothered enough by their symptoms that they visit their primary care practitioner, a neurologist or a different healthcare professional. Many people in this stage are initially diagnosed with Parkinson's disease or something called cerebellar ataxia due to similar motor symptoms. Some people receive a clinical diagnosis of MSA while in this stage, but not all. Below are symptoms people may experience in the early stage:

#### Motor or movement symptoms people may experience:

- Mild motor symptoms similar to Parkinson's disease may start; including tremors, slowed movements, and muscle stiffness
- Loss of coordination for manual movements and walking
- Balance issues that can lead to falls

#### Speech and swallowing symptoms people may experience:

- Mildly softer and/or slurred speech
- Coughing when drinking liquids or eating certain foods

#### Autonomic symptoms people may experience:

- Difficulties with blood pressure regulation that can cause dizziness and lightheadedness
- Needing to urinate more urgently and more frequently, but rarely leading to incontinence
- Constipation
- Erectile dysfunction

#### Impact on daily living:

- Little to no assistance is needed from other people or devices to perform daily activities, but it can improve safety, convenience or comfort

**Treatment, care or resources people can utilize:**

- Outpatient rehabilitation therapies such as physical, occupational or speech therapy
- Regular exercise
- Possible benefit from oral medications, such as carbidopa-levodopa for parkinsonism symptoms
- Dietary changes or medication to improve blood pressure, urinary function, and constipation
- Specialists as needed, such as cardiology or urology

**Middle Stage**

People often notice that they are moving into the middle stage when they need at least a little assistance to complete daily activities. People who did not get a MSA diagnosis in early stage are likely now diagnosed. The progression and impact of symptoms typically looks something like:

**Motor or movement symptoms people may experience:**

- Worsening tremors, slowed movements, and muscle stiffness
- Worsened coordination in arms and legs that affect manual activities and walking
- Balance issues that can lead to falls

**Speech and swallowing symptoms people may experience:**

- Worsening softness and/or slurring of speech
- Coughing when drinking liquids or eating certain foods

**Autonomic symptoms people may experience:**

- Difficulties with blood pressure regulation that can cause dizziness and lightheadedness
- Needing to urinate more urgently and more frequently, but rarely leading to incontinence
- Constipation
- Erectile dysfunction

**Mood and cognitive symptoms people may experience:**

- Mild cognitive changes, like difficulty organizing information and comprehension
- Depression and anxiety

**Impact on daily living:**

- Some assistance is needed to complete daily activities such as dressing, showering, getting out of chairs or moving outside of the home

**Treatment, care or resources people can utilize:**

- Outpatient rehabilitation therapies with focus on strategies to help with daily activities, safety with ambulation and moving, communication and care partner training
- Exercise from a chair with close monitoring and assistance
- Possible benefit from oral medications, such as carbidopa-levodopa for parkinsonism symptoms
- Dietary changes or medication to improve blood pressure, urinary function, and constipation
- Use of an appropriate walker and evaluation for future use of a wheelchair
- Grab bars or other equipment in the home to make getting around easier and safer
- Social work and palliative care services for additional support and to start discussions around care planning

- Urology and/or pelvic health therapy to manage urinary changes and constipation
- Possible referral for sleep study and to otolaryngologist for noisy breathing at night or during the day (called stridor)
- Occasional or regular professional care services (in-home care or adult day care) may be helpful for added support and assistance for the person with MSA and the family
- Driving evaluation
- Evaluation to capture baseline swallowing function; evaluations will usually be a modified barium study or a fiberoptic endoscopic evaluation of swallowing (FEES)
- Make future healthcare decisions before they need to be made, such as designating a power of attorney and deciding on advanced directives like a do not resuscitate (DNR) order or a living will

### Advanced Stage

At this stage in the disease, individuals are no longer able to complete most or any daily activities on their own. Those with the disease need close supervision and are at higher risk of complications like big falls or infections. People in this stage often have a much more difficult time communicating, which can take a toll on them emotionally. Common symptoms of the advanced stage are:

#### Motor Symptoms people may experience:

- Frequent falls
- Limited walking, especially without assistance
- Abnormalities of posture in some with head drop or spinal deformity.
- Impairment in fine motor movements, like writing or using utensils, that can make some daily activities impossible

#### Speech and swallowing symptoms people may experience:

- Significantly slow, soft and slurred speech to the point of having difficulty being understood
- Increased difficulty with swallowing food and liquids

#### Autonomic symptoms people may experience:

- Difficulties with blood pressure regulation that can cause dizziness and lightheadedness, which can lead to fainting
- Urinary incontinence and possible bowel incontinence

#### Impact on daily living:

- Assistance is required for most or all activities

#### Treatment, care or resources people can utilize:

- Possible continued benefit from oral medications, such as carbidopa-levodopa for parkinsonism symptoms
- Dietary changes or medication to improve blood pressure, urinary function, and constipation
- Permanent catheter for urination
- Working with physical and/or occupational therapy for adaptations to the home environment to support safety and daily functioning, such as toileting devices, lifts and adding foam to corners

- Speech therapy for recommendations on devices and strategies that can help with communication
- At this stage, home health therapy may be more convenient than participating in outpatient physical, occupational and speech therapy
- 24-hour care may be required; if circumstances permit, this can be supplemented with professional caregivers, or moving to an assisted living or skilled nursing facility; respite time is important for the well-being of care partners
- Close monitoring of swallowing function is important due to risk of choking and aspiration and likely need for changes to the consistency of food; it might be helpful to do another swallow evaluation to further understand the current level of swallowing function
- Finalize any healthcare decisions, such as designating a power of attorney and deciding on advanced directives like a do not resuscitate (DNR) order or a living will

### End Stage

The end stage is usually very difficult for care partners and loved ones. This stage may be several months or more but may also occur quickly based on a variety of factors, like if the person has any other chronic medical conditions or if they develop an infection such as a urinary tract infection (UTI). No matter what, the end stage can be a very vulnerable and intense time as the person requires hands-on and near-constant care.

#### Motor symptoms people may experience:

- Inability to walk
- Significant impairment in motor function and movement

#### Speech and swallowing symptoms people may experience:

- Inability to communicate
- Inability to maintain nutrition through eating due to difficulty swallowing

#### Autonomic symptoms people may experience:

- Bladder and bowel incontinence
- Weight loss

#### Impact on daily living:

- Complete support is required for all daily activities

#### Treatment, care or resources people can utilize:

- Hospice services to ease the transition and support quality of life through minimizing physical discomfort for the person with the disease and assisting the family in how to best monitor and provide care.
- A neurologist may still be helpful in finding medications to alleviate symptoms

### How CurePSP Can Help

Medical treatments and supportive therapies should be tailored to your individual needs regardless of stage. We encourage you to talk to your healthcare team and tap into additional CurePSP resources, such as booklets, webinars and support groups, to learn more about MSA and how to best receive the care you need for your unique disease journey. We are here to answer any questions you may have and to help you feel as supported and informed as possible.