

## Stages of Progressive Supranuclear Palsy (PSP)

PSP is an adult-onset neurodegenerative disease that impacts movement, thinking, speech and vision. PSP 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 PSP 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 subtype of PSP 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, it can be helpful to use some of these markers to know roughly where someone is in the disease journey so that the best treatment and care can be decided.

### Early Stage

The early stage of PSP 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 due to similar motor symptoms. Some people receive a clinical diagnosis of PSP while in this stage, but not all. Below are symptoms people may experience in the early stage:

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

- Occasional falls that are often unexplained
- Slowed movements
- Mildly slowed, softer and/or slurred speech
- Occasionally a mild tremor

#### Visual symptoms people may experience:

- Mild double vision

#### Non-motor symptoms people may experience:

- Fatigue
- Mild changes to thinking that may impact ability to easily socialize
- Possible personality or mood changes

#### 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
- Evaluation to capture baseline swallowing function (often a modified barium study or sometimes a fiberoptic endoscopic evaluation of swallowing (FEES))
- Specialists as needed, such as neuro-ophthalmology or urology
- Driving evaluation

## Middle Stage

People often notice that they are moving into the middle stage when they need a bit more assistance completing daily activities. People who did not get a PSP diagnosis in early stage are likely now diagnosed. The progression and impact of symptoms typically looks something like:

### Motor symptoms people may experience:

- Regular falls
- Worsened coordination with walking
- Possible freezing of gait
- More difficulty performing manual tasks, such as using utensils

### Visual symptoms people may experience:

- Difficulty reading and looking down due to eye movement changes
- Dry eyes or blurry vision

### Speech and swallowing symptoms people may experience:

- Worsening slow, soft and slurred speech
- Coughing when drinking liquids or eating certain foods

### Non-motor symptoms people may experience:

- Disinhibited or impulsive behavior
- Difficulty organizing thoughts
- Waking up multiple times at night and daytime sleepiness
- Urinary frequency and urgency
- Constipation

### 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 or botulinum toxin (Botox) injections to manage symptoms, such as muscle stiffness
- Neuro-ophthalmology/optometry for vision concerns
- 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 initiate discussions around care planning
- Urology and/or pelvic health therapy to manage urinary changes and constipation
- Sleep study or sleep specialist to address nighttime sleep quality and daytime sleepiness
- 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 PSP and the family
- 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
- Rigidity in neck, torso and limbs, which can be uncomfortable and make movement difficult

### Visual symptoms people may experience:

- Difficulty focusing on nearby objects because of eye movement issues
- Involuntary eyelid closure and difficulty opening them

### Speech and swallowing symptoms people may experience:

- Significantly slow, soft and slurred speech
- Increased difficulty with swallowing food and liquids

### Non-motor symptoms people may experience:

- Decreased ability to easily participate in conversations because of difficulties with voice and expressing words and organizing thoughts
- Urinary incontinence, including multiple wakings at night to use the bathroom
- Possible bowel incontinence

### Impact on daily living:

- Assistance is required for most or all activities

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

- Possible benefit from oral medications or botulinum toxin (Botox) injections to help with symptoms, such as muscle stiffness
- 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, 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
- 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

## End Stage

The end stage is usually very difficult for care partners and loved ones. This stage may last several months or more but may also happen 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 muscle stiffness

### Non-motor symptoms people may experience:

- Weight loss
- Urinary and bowel incontinence
- Very impaired or non-existent ability to communicate verbally due to loss of voice and inability to organize thoughts

### Impact on daily living:

- Care 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.
- 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.

## 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.