Stages of Corticobasal Degeneration (CBD) and Corticobasal Degeneration Syndrome (CBS)

Corticobasal Degeneration (CBD) is an adult-onset neurodegenerative disease that impacts movement and speech. CBD 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 CBD experiences the disease differently. Symptoms and their severity vary widely, and someone can be in each stage from anywhere between 1 to 4 years. Differences in disease symptoms and rate of progression 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.

Please note: Corticobasal syndrome (CBS) is the clinical diagnosis given during life, while the term CBD is increasingly being reserved for cases confirmed through autopsy. However, many people are still commonly told they have CBD. To maintain consistency with our other resources and what people may have already heard from their doctors, we will continue using the term CBD here.

Early Stage

The early stage of CBD is when people begin to experience symptoms. For many people with CBD, they notice that motor symptoms are only affecting one side of the body. 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 CBD or CBS while in this stage, but not all. Below are symptoms people may experience in the early stage:

Motor Symptoms people may experience:

- Clumsiness
- Difficulty with fine motor tasks, such as buttoning shirts or writing
- A loss of coordination often affecting one limb or side of the body more than the other
- Possible balance loss with falls

Non-motor symptoms people may experience:

- Mild cognitive symptoms
- Difficulties with executive function and language including word-finding
- Loss of initiative

Impact on daily living:

- More time required to complete daily activities
- No assistance from persons or devices is required to perform daily activities, though assistance may improve safety, convenience or comfort

Treatment, care or resources people can utilize:

- Outpatient rehabilitation therapies such as physical, occupational or speech therapy
- Regular exercise

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 CBD or CBS diagnosis in the early stage are likely now diagnosed. The progression and impact of symptoms typically looks something like:

Motor Symptoms people may experience:

- Increasing difficulty with motor tasks in affected limbs
- Muscle tightness and stiffness potentially with pain
- Imbalance and falls, possible involuntary jerking movements

Speech and swallowing symptoms people may experience:

- Mild speech impairment
- Mild difficulty swallowing

Non-motor symptoms people may experience:

- Difficulty organizing information
- Difficulty with comprehension
- Possible impulsivity
- Possible mood disorders

Impact on daily living:

- Some assistance is required to complete one’s daily activities, such as dressing, showering, getting out of low chairs, or ambulating 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
- Possible benefit for oral medications or botulinum toxin (Botox) injections to manage symptoms, such as muscle stiffness
- Evaluation to capture baseline swallowing function (often a modified barium study or sometimes a fiberoptic endoscopic evaluation of swallowing (FEES))
- Evaluation for wheelchair
- Social work and palliative care services for additional support and initiate discussions around care planning
- 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 performing daily activities and communicating, which can take a toll on them emotionally. Common symptoms of the advanced stage are:

Motor Symptoms people may experience:

- Symptoms may start on the other side of the body not originally affected
- Frequent falls
- Limited walking, especially without assistance
- Loss of ability to perform manual tasks
- Muscle tightness and stiffness affecting multiple areas including neck and torso
- Involuntary jerking movements

Speech and swallowing symptoms people may experience:

- Significantly softer and slurred speech, difficulty with articulation
- Increased difficulty with swallowing food and liquids, which can lead to choking

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

Impact on daily living:

- Assistance is required for most or all activities

Treatment, care or resources people can utilize:

- At this stage, home health therapy may be more convenient than participating in outpatient physical, occupational and speech therapy
- 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
- 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
- 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

Non-motor symptoms people may experience:

- Significant weight loss, urinary and bowel incontinence
- Very impaired or non-existent ability to communicate verbally due to loss of voice an 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.