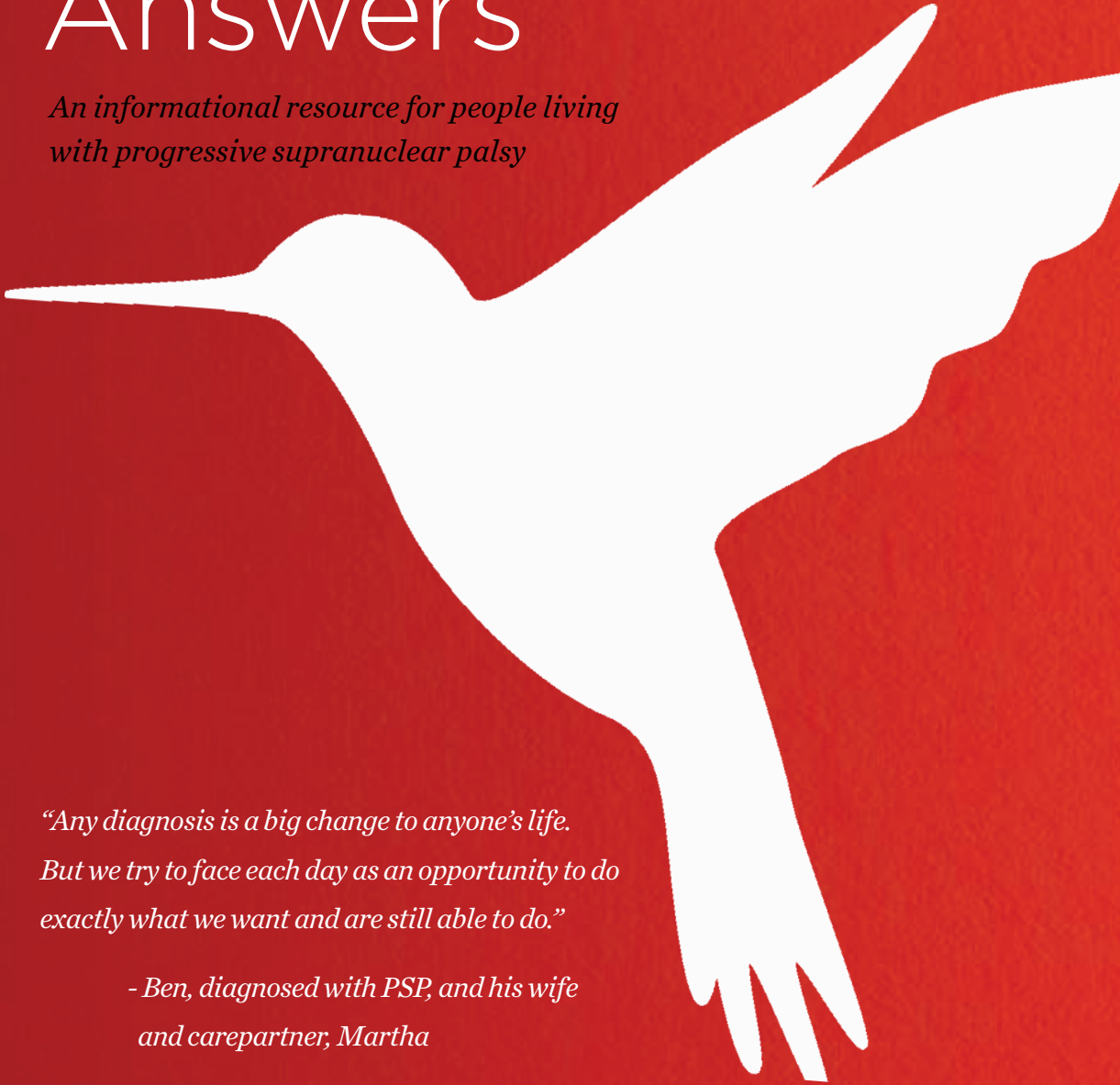


# PSP: Some Answers

*An informational resource for people living  
with progressive supranuclear palsy*



*“Any diagnosis is a big change to anyone’s life.  
But we try to face each day as an opportunity to do  
exactly what we want and are still able to do.”*

*- Ben, diagnosed with PSP, and his wife  
and carepartner, Martha*

# What is progressive supranuclear palsy (PSP)?

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*Progressive supranuclear palsy (PSP) is an adult-onset, neurological disease that impacts movement, thinking, speech, and vision. PSP is commonly referred to as an “atypical parkinsonism,” or sometimes “Parkinson’s-plus” diagnosis, because of its overlap of certain symptoms with Parkinson’s disease. However it often progresses quite differently than Parkinson’s disease, and it shares similar pathology to Alzheimer’s and frontotemporal dementia.*

## How many people are diagnosed with PSP?

PSP is considered rare. It is currently estimated that 10 to 12 people per 100,000 are living with PSP. About 30,000–40,000 people are diagnosed with PSP in the United States. The numbers for PSP are likely underestimates because many people with PSP are misdiagnosed with another condition, such as Parkinson's disease or frontotemporal dementia.

The prevalence for PSP compares to about 1 million with Parkinson's disease and 5 million with Alzheimer's disease. PSP is slightly more common than amyotrophic lateral sclerosis (called ALS, or Lou Gehrig's disease in the U.S. and motor neuron disease elsewhere), although ALS is better known than PSP in that it is easier to diagnose than PSP and often affects much younger people.

We recognize the extra layer of complexities and frustrations that can come along with a rare diagnosis, including healthcare providers who lack familiarity with the disease. One of CurePSP's priority goals is to improve awareness of PSP and related diseases among medical professionals and the general public. Such awareness is critical to accurate diagnosis, which allows patients earlier access to specialized management and care, and stimulates interest among researchers, pharmaceutical companies and funding agencies to support the cause and cure of this unique and puzzling illness.

## What are the common early symptoms of PSP?

PSP can start differently for different people, but there are some commonalities. Balance difficulty, changes to walking, and frequent falls are often the first symptoms for the majority of people with PSP. Other common early symptoms include changes to vision and speech, mild shaking of the hands, difficulty driving a car, and difficulty finding words. Early forgetfulness or personality changes, such as loss of interest in ordinary pleasurable activities, impulsivity, or increased irritability, can also occur and may be misinterpreted as depression or early stage dementia.

## What changes to vision can someone with PSP experience?

Changes to vision can be an early symptom of PSP for many people, while for others it does not appear until a few years later. Abnormalities with eye movements is something that neurologists will assess, which is why they often have you follow their finger and move your eyes in different directions without moving your head.

The most common and characteristic eye movement problem in PSP is an impaired ability to move the eyes up or down. Although Parkinson's disease, other conditions, and normal aging can sometimes cause difficulty moving the eyes up, PSP is nearly unique in also causing problems moving the eyes down. This problem often takes the form of eye movement apraxia, where the patient can move the eyes up or down only after several requests or with a delay after initiating the effort. This symptom can interfere with looking down at a plate while eating or with descending a flight of stairs, among other things.

Another eye movement problem that starts early in the illness is square wave jerks—rapid, involuntary, right-left movements that interfere with the ability to precisely aim the eyes at a target. Large eye movements can also become jerky rather than smooth.

Because aiming the eyes properly is the main difficulty, reading can become challenging, as a person with PSP may find it hard to automatically shift down from line to line. Some people also have difficulty maintaining eye contact during conversation.

Many people with PSP report double vision, which can look like two images of one object or the images may overlap. This can be an early symptom of PSP.

People with PSP can experience abnormal eyelid movement. One such problem is called blepharospasm, which can take the form either of forceful involuntary closing of the eyes for a few seconds or minutes at a time, or of difficulty opening the eyes, even though the lids seem to be relaxed. The person may try to use the muscles of the forehead, or even the fingers, in an effort to open the eyelids (“apraxia of lid opening”). Others may have trouble closing the eyes and blink very little. While about 15 to 25 blinks per minute are normal, people with PSP blink, on average, only about three or four times per minute. This can allow the eyes to become dry. Dry eyes can lead to blurred vision, production of extra tears, or a sensitivity to light called photophobia.

Not everyone with PSP gets all of these visual symptoms. Symptoms vary from person to person and in severity. If you experience changes to your vision, especially if they interfere with your daily functioning and quality of life, you can be assessed by a neuro-ophthalmologist. There may be treatment options and management techniques that can help, depending on the nature and severity of the vision symptoms, such as artificial tears for dry eyes, prism glasses or prism stickers for double vision, and Botox injections or eyelid crutches for forced eyelid closure. Occupational therapists can also provide recommendations for adaptations related to vision that can make daily life easier and the home environment safer.

### **How does PSP impact speech?**

The same general area of the brain that controls eye movement also controls movements of the mouth, tongue, and throat, and these movements also weaken in PSP. In Parkinson’s disease, the primary speech problem is characterized by soft volume. While this can occur in PSP, PSP can also cause the speech to have an irregular, explosive, or rubber band quality (called spastic speech) or a slurred, drunken quality (ataxic speech). These changes to speech and voice can be coupled with difficulty getting out thoughts or finding words, making communication quite challenging for many people with PSP. It can be helpful to work with a speech-language pathologist, especially one who is specialized in Parkinson’s disease and related disorders, to learn exercises and compensatory strategies to help people with PSP continue to communicate as much as possible.

### **Does PSP cause difficulty with swallowing?**

Unfortunately, swallowing problems are common with PSP and are a leading cause of complications. PSP can cause weakness and incoordination of throat muscles, making it more difficult to swallow. This issue increases the risk of food or liquids going into the lungs, which is called aspiration, and can lead to pneumonia.

Changes to swallowing function, which can occur early in PSP, may include coughing on thin liquids or more particulate foods, such as salads or dry cereal. Other signs of swallowing dysfunction can include the sensation of food being caught in the throat, frequent runny nose, watery-sounding voice, occurrences of choking, and recurrent lung infections. Additionally, sometimes people with PSP can eat more impulsively, including large bites or very quickly, which can increase risk with swallowing.

Your doctor may recommend regular examinations of your swallowing function to make sure food and liquid are going down the right way. Additionally, speech-language pathologists can help assess swallowing function as well as provide recommendations for diet and lifestyle modifications and exercises for the throat in order to make swallowing easier and safer.

## **Do people with PSP develop dementia?**

Dementia is a term that refers to the loss of cognitive functioning to the extent that it impacts a person's daily life. There are different types of dementia, and different diseases can cause dementia. In Alzheimer's disease, the most common cause of dementia, the cognitive impairment is primarily in the area of memory. People with PSP can experience changes to cognitive function, especially in more advanced stages of disease. This is usually in the areas of organization and processing of thoughts, planning, multitasking, attention, and word-finding. Sometimes people with PSP are initially misdiagnosed with Alzheimer's disease, or may be diagnosed with frontotemporal dementia, which is a different type of dementia.

Aphasia is a cognitive deficit affecting communication, including finding words, expressing thoughts, and understanding language. Different neurological conditions can cause aphasia. Some people with PSP can experience aphasia. The severity can vary greatly, and for some, it can be an early symptom of PSP.

For many families of someone with PSP, the most challenging aspects of cognitive impairment with PSP are impulsivity, poor judgment, and lack of insight into their cognitive or physical challenges. As examples, someone may jump up from a chair quickly, without waiting for assistance or using a walker, or may eat large portions of food very quickly. Some families have also shared that their loved one with PSP developed a tendency for impulsive and compulsive shopping. A big point of contention for families and healthcare providers alike is when a person with PSP is adamant they can still drive or do other things safely when it is apparent to others that they are unsafe. Impulsivity coupled with imbalance is the primary reason that people with PSP fall.

Apathy can also be a cognitive symptom with PSP, causing people to have less motivation to partake in activities. Cognitive changes in PSP can lead to social withdrawal or irritability, especially when they make communicating thoughts and participating in conversations more difficult.

Even in later stages of PSP, a person can often still respond to a question with accuracy and detail, but because of slowed processing speed, it may take several seconds or minutes and great effort to get out a thought. People with PSP and families can ask people to be patient when communicating. It can also be helpful to work with a speech-language pathologist on strategies to compensate for and adapt to cognitive changes and make communication easier.

## **What happens to someone with PSP over time?**

The name of the disease includes the word "progressive" because, unfortunately, early symptoms get worse and new symptoms develop over time. After five or six years, on average, the imbalance and stiffness worsen to make walking very difficult or impossible. Difficulty with eyesight, speech, and swallowing are additional important features of PSP that eventually occur in most patients. Not everyone experiences all of the symptoms of PSP, and the appearance and progression of these symptoms varies greatly among individuals.

Research has shown that someone with PSP lives, on average, about eight to ten years after the onset of symptoms. Injury from falling is one of the most common complications in PSP. Another is infection, particularly pneumonia and urinary tract infections. Your doctor may recommend regular evaluations of your swallowing function, assessments by a physical therapist and adaptive equipment to improve your safety with ambulation, a consultation with a urologist if challenges with urinary function arise, and other preventative measures.

We recognize this information is scary and overwhelming to learn and to think about. It can be helpful to talk this information through with your medical team and your family, including planning for the future and your wishes for quality of life. CurePSP and the rest of your support system are here to help.

### **Are there different types of PSP?**

PSP is occasionally referred to as Steele-Richardson-Olszewski syndrome after the three physicians who first described the disease in 1963. There are thought to be several different types of PSP.

About half of those with PSP have the Richardson's syndrome type. The most common first symptom of this type of PSP is loss of balance while walking, which can take the form of unexplained falls. People may also experience stiffness or awkwardness when walking, while sometimes the falls are described as attacks of dizziness.

The second most common form of PSP, accounting for about a quarter of cases, is called PSP-parkinsonism. The PSP-parkinsonism type of PSP is more likely to have mild tremor as a symptom, to involve one side of the body more than the other, to have less of a problem with vision and swallowing, and to respond better to drugs for Parkinson's disease.

There are other less common types that are characterized by their predominant clinical symptoms.

Some neurologists may share the diagnosis of a specific PSP type with you, but they likely will not. The different types may involve symptoms that require certain care considerations, but ultimately, PSP, regardless of the type, is treated similarly. Care, support, and management will be tailored to your unique needs.

### **How is PSP diagnosed?**

To diagnose PSP, a neurologist will gather a person's medical history, including neurological symptoms, when they started, and how they impact the person's everyday functioning. They will also perform a physical examination where they will evaluate how a person walks, speaks, and moves their eyes, feet, and hands.

At this time, there is no specific test to diagnose PSP. Neurologists will often refer to magnetic resonance imaging (MRI) to look for changes in the brain. Midbrain atrophy found on a brain MRI can be suggestive of PSP and when viewed from a certain angle, this atrophy can resemble a hummingbird. The "hummingbird sign" on a MRI (sometimes called the "penguin sign") can help to support a clinical diagnosis of PSP. A neurologist may decide to use other tests, such as blood tests, a neuropsychological evaluation, a DaTscan, or positron emission tomography (PET) scan to support the diagnosis of PSP or help rule out other causes of symptoms. Sometimes how someone responds to medications for Parkinson's disease can also help a neurologist make a diagnosis of PSP.

Many people with PSP face a long and confusing diagnosis journey, including a number of tests, specialists, and diagnoses. It is our hope that better awareness of PSP, especially within the medical community, will lead to earlier and more accurate diagnosis.

## How is PSP treated?

At this time, there are no medications to cure PSP or to slow its progression. Some symptoms of PSP can be managed successfully with medications for the same symptoms in other conditions. Drugs for Parkinson's disease that stimulate the brain's dopamine system, particularly carbidopa-levodopa, can be effective in alleviating some of the parkinsonism symptoms in PSP, particularly slowness and stiffness. It has also been found to reduce some of the frequency of falling. Unfortunately, the response to carbidopa-levodopa in PSP is typically not as dramatic or long-lasting as in Parkinson's disease. Sometimes having little to no response to carbidopa-levodopa after a Parkinson's diagnosis can be a sign of an atypical parkinsonism disease such as PSP. Amantadine is another medication often used in Parkinson's disease that may have some benefit in PSP, particularly with freezing of gait.

Botox or Myobloc, two types of botulinum toxin, can be useful for people with PSP who experience blepharospasm (forced closure of the eyelids), excessive drooling, involuntary bending or turning of the neck muscles, jaw clenching, or other types of dystonia. Botox injections should be performed only by an experienced neurologist to minimize risks and side effects.

Other medications can be used to manage other symptoms of PSP, such as sleep, urinary, or mood changes. Your doctor will work with you closely to try different medications, timing, and dosages to maximize the benefits for your symptoms while also trying to minimize side effects.

Deep brain stimulation (DBS) is a brain surgery commonly offered to people with Parkinson's disease. There have been only relatively small case series as examples of people with PSP undergoing DBS, and in most cases it was because they were misdiagnosed as having Parkinson's disease. DBS worsens symptoms for people with PSP, and for this reason, DBS is not recommended for PSP at this time.

Allied health therapies are also important pillars of treatment to address many of the symptoms and challenges faced by people living with PSP. Occupational therapists can assess safety in the home environment and with driving and daily tasks, as well as provide recommendations for strategies and adaptive equipment to support independence and ease of daily activities. As research has shown repeatedly that cardiovascular exercise can slow down progression of motor decline in most neurodegenerative conditions, exercise remains a very important piece of disease management for people with PSP. Physical therapists can teach exercises that are safe to do and help to keep someone active, as well as provide recommendations for appropriate ambulatory equipment. Speech-language pathologists address speech, communication, cognition, and swallowing functions. Clinical social workers and nursing professionals are equally important members of a care team, offering resource connection, help with navigating the healthcare system, and support in decision-making. It is also important to treat anxiety or depression if it is impacting daily functioning and quality of life, which can be done through the use of mood medications and mental health therapy.

## Why is it called “progressive supranuclear palsy”?

“Progressive” is in the name because of how symptoms change and worsen over time. The term “supranuclear” refers to the area of the brain that is affected in PSP (at the base of the brain, in the brainstem), that causes the eye problems in PSP. In general, “palsy” means weakness or paralysis of a part of the body. People with PSP have often difficulty aiming the eyes properly because of weakness or paralysis (palsy) of the muscles that move the eyeballs.



## Why have I heard PSP referred to as an “atypical parkinsonism disorder”?

Many people with PSP may be told, prior to a more definitive PSP diagnosis, that they have “parkinsonism.” They may hear the term “atypical parkinsonism” used as an umbrella term referring to PSP and other related diagnoses. Parkinsonism is a term that describes a constellation of symptoms similar to Parkinson’s disease, including tremor, imbalance, slowness of movement, and stiffness of muscles. Sometimes a provider will say someone has “parkinsonism” in very early stages of symptoms, when it is not yet clear what diagnosis someone has.

“Atypical parkinsonism” is a term frequently used to refer to diseases that look like Parkinson’s disease in many ways, and may even be initially diagnosed as Parkinson’s disease but then have other symptoms not as commonly seen in Parkinson’s disease. These “atypical” symptoms may include earlier and more profound changes to vision, balance, and speech as well as a lack of robust response to the Parkinson’s disease medication carbidopa-levodopa. Other diagnoses that are commonly referred to as atypical parkinsonism diseases include corticobasal degeneration (CBD) and multiple system atrophy (MSA). Because of the similarities of care journeys and needs of PSP, CBD, and MSA, it is not unusual for them to be served by the same healthcare providers, support groups, and organizations, including CurePSP.

## What causes PSP, and what is happening in the brain?

The direct cause of PSP is not fully understood. However, we do know that it has to do with the clumps of tau protein. Tau is a normal protein found in brain cells and is currently thought to help maintain the microtubules, which are stiff rods that function as the brain cells’ internal transportation and skeletal system. In PSP, the tau seems to become abnormally folded, which causes it to stick together and become stuck inside the cell. The areas of the brain that have cells with tau inside of them also exhibit impaired neuron function and neuronal death. The clumps’ technical name is neurofibrillary tangles. Alzheimer’s disease and frontotemporal dementia also involve misfolding of the tau protein.

The symptoms of PSP are caused by a gradual deterioration of brain cells in a few places in the base of the brain. The most important such place, the substantia nigra, is also affected in Parkinson’s disease; damage to it accounts for the symptoms that PSP and Parkinson’s disease have in common. However, several important areas that are affected in PSP are normal in Parkinson’s disease, and vice versa. Additionally, Parkinson’s disease is caused by the misfolding of a different protein, called alpha synuclein, and under the microscope, the appearance of the damaged brain cells in PSP is quite different from those in Parkinson’s disease. Instead, it resembles the degeneration in Alzheimer’s disease and frontotemporal dementia, although the location of the damaged cells is quite different in PSP compared with these other tauopathies.

Genetic differences and exposure to certain chemicals are being explored as possible causes, but ultimately, we are not yet sure why tau misfolds or people get certain brain disorders like PSP. For someone who is personally impacted by PSP, we recognize that not knowing the cause can be extremely frustrating, confusing, and scary. Researchers and doctors are working hard to understand PSP and other related neurodegenerative diagnoses, and we hope their work will lead to more answers and treatment options soon.

## Is PSP genetic?

PSP very rarely runs in families and is not considered a genetic disease. Fewer than one in 20 people with PSP knows of even one other family member with PSP, and detailed neurological



exams of relatives of patients with PSP show no more definite abnormalities than exams of relatives of healthy people.

However, two different variants in the gene on chromosome 17 that encodes the tau protein are more common in PSP than in the rest of the population. One of the variants is called the H1 haplotype. About 95% of people with PSP have this variant on both of their copies of chromosome 17, while this is true for only about 60% of people without PSP. So clearly, the H1 haplotype is (nearly) necessary but far from sufficient to cause the disease. We don't yet know how that version of the tau gene actually affects brain cells. It may increase the amount of tau protein produced or it may change the chemical properties of the protein.

Over the past two decades, a handful of other gene variants not on chromosome 17 have been found to be more common in people with PSP than in people without PSP. These genes help control a variety of critical processes inside of brain cells. More research is needed to identify how these insights can translate into possible PSP prevention and treatment.

### **How can I get involved in research in PSP?**

There is important research being done to look for genetic and environmental contributors to the cause of PSP, as well as research on tests that can lead to more accurate diagnosis and improved care of PSP. Many people with PSP find that participation in research is a meaningful way to help doctors and scientists understand, diagnose, and treat PSP and related diseases. Additionally, participants in clinical trials may benefit from a new treatment that is not generally available and may receive detailed care and attention that is not part of routine medical care.

Because of the rarity of PSP, there can sometimes be the challenge of not having enough eligible people to be included in drug trials. To properly test a drug and understand its impact, patients need to be recruited from a number of sites. CurePSP is working hard to build partnerships with pharmaceutical and biotechnology companies to develop interest and funding for the study of PSP and related diseases. Additionally, CurePSP funds research through various grant programs and supports collaborative research projects through its growing CurePSP Centers of Care network of medical centers across the United States and Canada.

In the United States, clinical trials are listed on a website maintained by the National Institutes of Health, [www.clinicaltrials.gov](http://www.clinicaltrials.gov). You can enter “progressive supranuclear palsy” into the search box. You can also ask your neurologist if they are aware of studies in PSP or consult [curepsp.org](http://curepsp.org) for that information.

Donating your brain to science can be a powerful contribution to the understanding of PSP and other neurodegenerative conditions. Each donated brain is also evaluated by a trained neuropathologist to confirm that the diagnosis of PSP was correct. Setting up brain donation needs to occur early, ideally months or even years prior to someone passing away. Visit [www.psp.org/inedsupport/braindonation](http://www.psp.org/inedsupport/braindonation) to learn about CurePSP's Brain Donation Assistance Program.

### **What can I do to support myself and my family with this diagnosis?**

Building a support team around you is foundational to quality of care and life with PSP. Your support team may consist of your partner, family, friends, support group, religious community, healthcare team, professional care, and others—people who care about you and show up for you.

When living with a chronic and progressive diagnosis, it is important to find the right medical team to support your needs with PSP over time. PSP needs to be managed by a neurologist. This could be a general neurologist, but, if available in your area, you may also choose to work with a neurologist

who has gone through specific training in movement disorders or behavioral/cognitive neurology. Rehabilitation therapists (physical, occupational, and speech therapists) and clinical social workers also play important roles in the care of PSP. You may also benefit from adding other specialists to your team as symptoms and needs arise. These may include a urologist, sleep specialist, neuro-ophthalmologist, and palliative care specialist. Taking care of your emotional health, whether as a person diagnosed with PSP or as a carepartner, is also a priority, and working with a mental health professional to process the experience, foster coping skills, or address other emotional needs can be exceptionally beneficial. When building your care team, it is important you have providers that you have chemistry with and that you trust, and for you to know that you have the right and ability to change your providers if needed.

Completing health care advance directives is an excellent tool for sharing your wishes regarding care with your support team. Health care advance directives address topics such as how aggressive your medical care should be (for example, whether you would want a feeding tube or a machine for breathing, if the need arose) and how you define quality of life. These directives should be completed with your family and your doctor and should be reviewed at least annually in case your wishes change.

Many people living with PSP consider and explore professional care services, such as in-home care, adult day care, or long-term care, depending on their care needs and situation. These services can provide additional layers of support, including companionship or hands-on help for the person with PSP and assistance and respite for the family.

10 Additionally, there can be great value in connecting with other people affected by the same diagnosis as you and your family, through support groups or peer supporters. It can feel validating and uplifting to hear the experiences and insights on how others adjust to life with PSP. You can exchange helpful tips on ways to cope and adapt physically and psychologically with the diagnosis. CurePSP offers a variety of support groups for people affected by PSP. There are also many more for atypical parkinsonism (which can include PSP and other diseases, such as corticobasal degeneration and multiple system atrophy) in the United States and other countries. Visit [www.psp.org/ineedsupport/supportgroups](http://www.psp.org/ineedsupport/supportgroups) for a list of regional support groups as well as virtual, national and international support groups facilitated by or in collaboration with CurePSP. Additionally, many local Parkinson's disease or frontotemporal dementia support groups welcome members with PSP. If you may be interested in starting your own PSP support group, contact CurePSP to learn more and for help in getting started. Additionally, CurePSP offers a peer support network, family conferences, and educational webinars where you can learn about PSP and connect to the community.

We know that a diagnosis of PSP can bring up many emotions, changes, and considerations. No matter how you find support, please remember that you do not have to navigate the PSP journey alone.

**The mission of CurePSP is to raise awareness, build community, improve care, and find a cure for PSP, CBD, and MSA.**

Please contact CurePSP for additional information and resources:

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**Special note regarding the front cover:**

The “hummingbird sign” refers to a unique feature found on brain imaging that is often used to support a clinical diagnosis of PSP. A hummingbird has come to symbolize strength, connection, and hope for many within the PSP community.

C U R E P S P . O R G