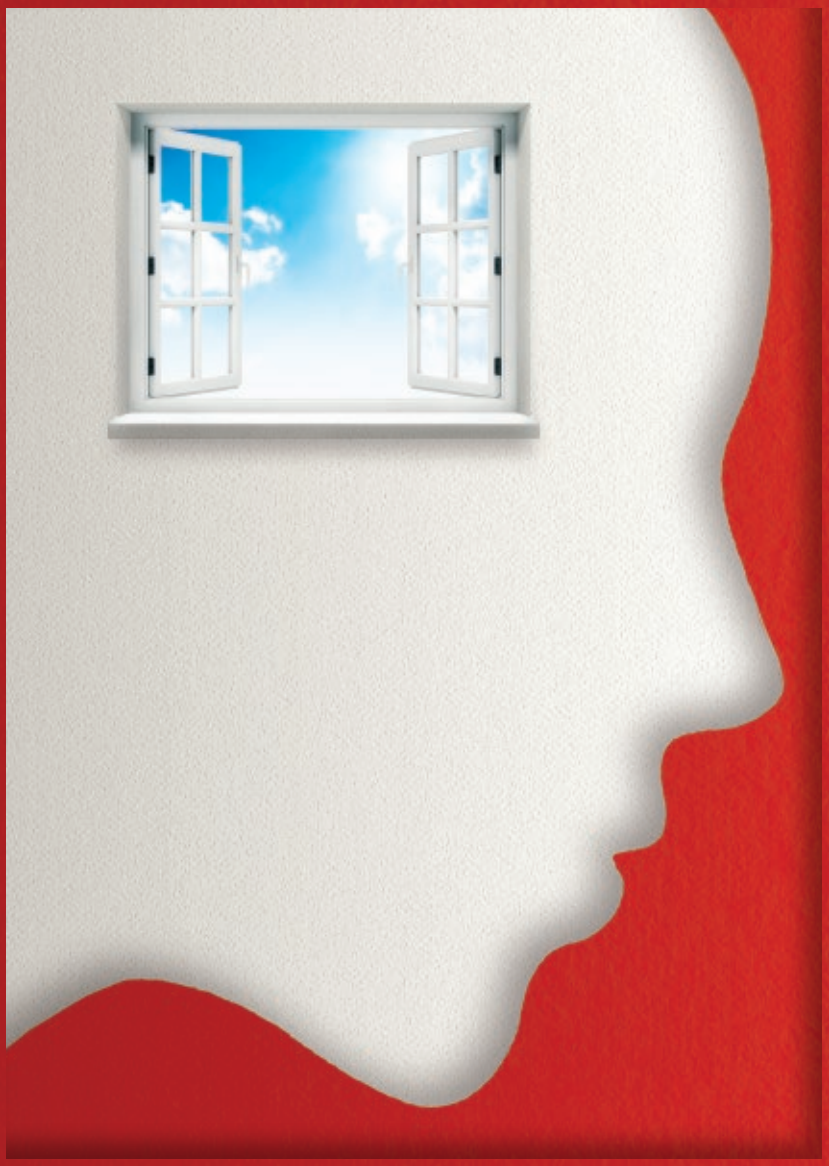
curePSP®

UNLOCKING THE SECRETS OF BRAIN DISEASE®

# Guidebook



**A RESOURCE FOR PEOPLE LIVING WITH PRIME OF LIFE NEURODEGENERATIVE DISEASE**

*Information you need to know*
*written by healthcare professionals*

Progressive supranuclear palsy (PSP)
Corticobasal degeneration (CBD)
Multiple system atrophy (MSA)

2020 EDITION


**EDITED BY**

LAWRENCE I. GOLBE, MD

DIANE BRESLOW, MSW, LCSW

# DEDICATION

*This guidebook is dedicated to the many volunteers who tirelessly contribute untold hours, diverse talents, knowledge, and great compassion toward making a difference in the lives of others.*

*Whether professionals in the medical and allied healthcare field, business professionals, or those whose knowledge from firsthand experience uniquely qualifies them to serve, all are experts in giving and compassion.*

*We at CurePSP are grateful for every generous act volunteers contribute, to the mission and to the well-being of those diagnosed, their caregivers, and the friends and families on this journey. Without them, we would not have come this far.*

*With them, we will go far.*

curePSP®

UNLOCKING THE SECRETS OF BRAIN DISEASE®

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# A Message from CurePSP's Director of Clinical Affairs

*Lawrence I. Golbe, MD*

Anyone embarking on the journey with PSP or a related disease as a patient, family member, or caregiver knows that finding detailed information on the disease is not easy. Few doctors know much about PSP or have time to share what they do know. Medical literature is too technical; articles provided by organizations devoted to Parkinson's disease are too superficial; and much of what's on the Internet is just plain wrong. One of the founding purposes of CurePSP in 1990 was to correct that state of affairs. This book is one big way of doing that.

Research into PSP has made important progress in the past few years. Drug companies have started clinical trials of treatments to slow the ongoing progressive course, and more are entering that arena each year using a variety of approaches to the problem. Genetic studies have revealed a few DNA variants, each with a contribution to the cause of the disease that's too subtle to elevate the risk to family members but important enough to point to new drug targets. New techniques have become available for rapidly testing drugs in the lab using stem cells. A new set of diagnostic criteria for PSP and for its several newly identified variants has been published and validated. The same set of criteria can identify the early, uncertain stage of PSP. Progress is being made in finding a diagnostic test of blood or spinal fluid, and a new type of PET brain scan may, in the next year or two, prove able to identify PSP before it even causes symptoms.

CurePSP has been at the forefront. We have formed collaborations with multiple organizations large and small to bring their resources to bear on "our" disease. We continue to provide grant money to established, top researchers and to students doing faculty-supervised summer projects on PSP. We convene an annual international scientific conference at which researchers share their findings and ideas. We have formed a network of clinical centers at medical schools with verifiable expertise in PSP, our Centers of Care initiative.

Perhaps one measure of our success has been documented in a paper that two colleagues and I published in the journal Movement Disorders Clinical Practice in early 2018. We wanted to know if people with PSP were receiving that diagnosis with less delay than was the case in past decades. Sure enough, in the patient population in New Jersey, the average delay from initial symptom to a diagnosis of PSP, or even a suspicion of it on the written record, declined from 44 months in the 1990s to 29 months in the 2010s. This dramatic improvement in the awareness of PSP by physicians may in large part be the work of CurePSP, which, since its founding 1990, has educated physicians and laypersons in recognizing early features of PSP.

Please let us know how we can continue to improve this guide in future editions. We hope it is helpful, readable, authoritative, and above all, hopeful.

**Lawrence I. Golbe, MD**
*Director of Clinical Affairs, CurePSP*

## A Message from the Editors

*Lawrence I. Golbe, MD, and Diane Breslow, MSW, LCSW*

We are pleased to bring you the updated 2020 Edition of CurePSP's Guidebook. In the foundation's commitment to providing up-to-date information, we have revised and expanded upon the previous edition. We also hope the reorganized Table of Contents and the merging of some of the articles make the book easier to use and understand.

The Guidebook is devoted to helping patients and families learn more about PSP and related diseases that we call "prime of life" neurodegeneration. These diseases tend to strike (though not always) during middle age, when people often have careers, family responsibilities, and active lives. We realize that everyone affected by these diseases is at a different place in the journey. You and your loved one may not experience everything this book discusses. In addition, resources may vary depending upon where you live. Equally important, these diseases can carry with them profound emotions and changes that vary dramatically from one patient-family unit to another. Therefore, we want you to use the book as an ongoing resource for yourself and your family. A good approach might be to read material relevant to you at the time, rather than attempting to tackle the entire manual at once.



Whatever your needs might be, and whenever they occur, we want this book to be beneficial to you. Also, please contact CurePSP if you have questions or wish to request any other of the foundation's informational and educational materials. We want you to know that you are not alone. CurePSP—our staff, support group leaders, and volunteers—and all the families who have been through these diseases are your community of support.

**Website:** curepsp.org
**Email:** info@curepsp.org
**Phone:** 800-457-4777 and 347-294-3873 (CURE)

Lawrence I. Golbe, MD

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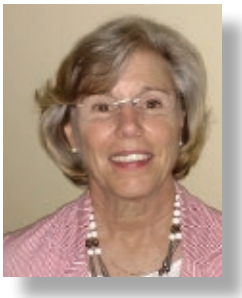
## About the Editors

*Lawrence I. Golbe, MD,* is Emeritus Professor of Neurology at Rutgers Robert Wood Johnson Medical School in New Brunswick, NJ. He has worked closely with CurePSP since 1992, currently as Director of Clinical Affairs, a member of its Board of Directors, and chair of its Medical Advisory and Scientific Advisory Boards. He graduated from Brown University and New York University School of Medicine and completed residency training at NYU/Bellevue. He was the first to measure the prevalence of PSP and to assess its epidemiological risk factors. He devised the PSP Rating Scale, which, since its publication in 2007, has become the standard clinical measure and treatment outcome marker for PSP worldwide. He has a referral practice for patients with PSP and CBD and conducts multiple ongoing research studies in PSP. He is the author of the 2019 book, *A Clinician's Guide to Progressive Supranuclear Palsy*.



*Diane Breslow, MSW, LCSW,* is a Licensed Clinical Social Worker in Illinois. She has worked with CurePSP since the early 2000s, when she served on the organization's multidisciplinary advisory board and participated in the Midwest Family Conference. She has been an active member of CurePSP's Patient Carepartner Advocacy Committee (PCAC) since 2015. In 2018, she became the organization's Social Work Clinical and Education Consultant. In that role, she develops and oversees the Committee's initiatives, consults to patients and families, and writes educational materials. Diane graduated from the University of Missouri in Columbia and received her master's degree, with a specialty in family therapy, from the University of Maryland School of Social Work. Throughout her career, Diane has applied her expertise in family dynamics to helping improve the quality of life for individuals and families affected by movement disorders.



## A Message from the Chair of CurePSP's Patient and Carepartner Advocacy Committee

*Ileen McFarland*

Those of us who have walked the walk, either as a patient or carepartner associated with one of the rare brain diseases, know all too well the daily challenges these diseases present. Patients are stunned at their diagnosis and do not know where to turn, and carepartners feel powerless and unqualified in their efforts to comfort and provide care for their loved ones. We here at CurePSP are sensitive to these circumstances. Over the years, we have developed programs and support networks to assist you. We have compiled an array of written materials that offer concrete resources and education about the entire course of disease and disease features: the physical, mental, emotional, and family impact of disease. You can request information simply by calling our office.

We have some 70 in-person support groups in the U.S. and Canada. We also offer eight online support groups. We can expect both of these numbers to grow. Patients and carepartners come together to learn about and discuss issues and to ease their burden. Well-trained facilitators, some of whom have had personal experience with one of the diseases, lead these support groups. Periodically, the support group leaders schedule professional speakers, such as neurologists, physical therapists, occupational therapists, speech therapists, experts in legal matters, social workers, and hospice representatives, just to name a few. There are also online webinars that address specific subject matter related to the disease, such as managing carepartner stress; physical, occupational, and speech therapies; "how to" presentations; and others. At all levels—written, online, and through our programming—we continue to expand our educational reach.

We offer Family Conferences at various locations nationwide twice a year. These conferences are for patients and carepartners, and include support group sessions the day before the conference starts.

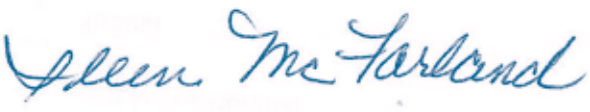
Additionally, we have approximately 100 Peer Supporters nationwide and others in Australia, Canada, India, Kuwait, and Argentina. These volunteers play a very important role by making themselves available for patient, family, and carepartner phone calls. Most of our Peer Supporters have lost a loved one or a friend to one of the brain diseases and are familiar with the journey. They can provide useful information that will assist the patient and carepartners, or just listen with understanding to your story.

The above, and more, information can be accessed through our website, *www.curepsp.org*.
Our committee and the entire CurePSP team continually work to strengthen and increase advocacy, education, and support programs, with the aim of providing tools to ease the burden of patients and their families.

Educating our physicians and healthcare professionals is also crucial to our mission. The more they know, the sooner they can provide an accurate and timely diagnosis and treatment recommendation. For healthcare professionals, we provide educational materials in print and video, and on the web. We also present specialized curriculum for general neurologists, physical therapists, occupational therapists, and speech and language therapists across the U.S. We have established working relationships with professional organizations that provide us the opportunity to present at their conferences. In their local areas, our many volunteers and support group participants also play an important role in educating the neurologists and healthcare professionals.

**We are here for you... you are not alone.** As research continues to search for a cure, we are sensitive to your daily challenges and needs. We can and want to provide you with resources and support during your journey with neurodegenerative disease. Get in touch with us and let us know your concerns. We can't make the disease go away, but we can help you persevere. Contact *info@curepsp.org* or 347-294-2873 (CURE).





**Ileen McFarland**

*Chair, CurePSP Patient and Carepartner Advocacy Committee*

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PART ONE

# The Diseases

**Lawrence I. Golbe, MD**

*Professor of Neurology,*
*Rutgers Robert Wood Johnson Medical School*
*Director of Clinical Affairs and Scientific Advisory Board Chairman, CurePSP*

## *What is progressive supranuclear palsy (PSP)?*

Of the approximately 15,000-20,000 people in the United States with progressive supranuclear palsy (PSP), few, if any, had ever heard of the disease before their diagnosis. In fact, most patients with PSP report that their family doctors knew nothing about it until a neurologist made the diagnosis. As of now, three of every four people with a diagnosis of PSP could have been diagnosed earlier, if their doctor had suspected it and performed the appropriate examination. However, it is appearing in medical journals more and more often, which will help doctors become familiar with PSP. This chapter should help patients and their families do the same.

## *Why has no one heard of PSP?*

PSP is rare: No one even realized it existed until 1963, when several patients were first described at a national neurology research convention and the disease was given its name. In retrospect, at least 12 cases of PSP had appeared in the medical literature between 1909 and 1962, but because of its resemblance to Parkinson's, it wasn't recognized as a distinct disease. The brain under the microscope is almost identical to that of "post-encephalitic parkinsonism," a common condition in the early 20th century but now nearly extinct, which also made for erroneous diagnoses during that era.



Although PSP is slightly more common than the well-known amyotrophic lateral sclerosis (called ALS, or Lou Gehrig's disease in the U.S. and motor neuron disease elsewhere), ALS is easier to diagnose than PSP and often affects much younger people. Each year an average of 1.1 people per 100,000 are newly diagnosed with PSP; five or six people per 100,000 are living with the disease. These figures are nearly identical wherever they have been carefully measured, which is in only three countries—the U.K., the U.S., and Japan.

## *What are the common types of PSP and their early symptoms?*

PSP is occasionally referred to as Steele-Richardson-Olszewski syndrome, after the three physicians who first described the disease in 1963. The most common form is Richardson's syndrome, after Dr. J. C. Richardson, who only recently retired from a career in neurology research. About half of everyone with PSP has the Richardson's syndrome type. The most common first symptom, which occurs, on average, when a person is in her or his 60s, is loss of balance while walking. This may take the form of unexplained falls or of a stiffness and awkwardness in a person's gait that can resemble Parkinson's disease. Sometimes the falls are described by patients as attacks of dizziness. This often prompts the doctor to suspect an inner ear problem or hardening of the arteries supplying the brain.

The second most common form of PSP is called PSP-parkinsonism. Its early stages more closely resemble those of Parkinson's disease, with less emphasis on balance problems and behavior changes and more on tremor. These typically have a better early response to antiparkinson drugs than is typical for PSP. PSP-parkinsonism comprises about a quarter of all PSP.

## *What are the most common early symptoms of PSP?*

Balance difficulty, usually with falls, is the first symptom in a majority of people. Other common early symptoms can be misinterpreted as depression or even as senility. These include forgetfulness and personality changes, such as loss of interest in ordinary pleasurable activities or increased irritability. Less common early symptoms are trouble with eyesight, slurred speech, mild shaking of the hands, and difficulty driving a car. Freezing of gait can be a first and only symptom for several years and difficulty finding words, or aphasia, can be a first and most prominent issue.

## *Are there other, rare types of PSP?*

Another type, called corticobasal syndrome, affects limbs on one side of the body far earlier and more severely than on the other side. It is a rare type that was described only in 2018, mostly in Japan, and starts with ataxia, which is a specific type of coordination problem arising from the cerebellum and resembling drunkenness. Most people with these minority phenotypes eventually do develop the more classic signs and symptoms of PSP.

## *What happens next?*

The name of the disease includes the word "progressive" because, unfortunately, the 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. Trouble with eyesight eventually develops in almost all cases and can sometimes be as disabling as the movement difficulty. Difficulty with speech and swallowing are additional important features of PSP that eventually occur in most patients.



## *What does the name "supranuclear palsy" mean?*

In general, palsy is a weakness or paralysis of a part of the body. The term "supranuclear" refers to the nature of the eye problem in PSP. Although some patients with PSP describe their symptom as blurring, the actual problem is an inability to aim the eyes properly because of weakness or paralysis (palsy) of the muscles that move the eyeballs. These muscles are controlled by nerve cells residing in clusters or nuclei near the base of the brain, in the brainstem. Most other brain problems that affect the eye movements originate in those nuclei, but in PSP the problem originates in parts of the brain that control those eye-movement nuclei. These "higher" control areas are what the prefix *supra* in *supranuclear* refers to.

## *Is the visual problem the most important part of PSP?*

In most cases, the visual problem is at least as important as the walking difficulty, though it does not typically appear until three to five years after. Because aiming the eyes properly is the main difficulty, reading often becomes challenging. The patient finds it hard to automatically shift down from line to line. This is very different from just needing reading glasses. An eye doctor unfamiliar with PSP may be baffled by the patient's complaint of being unable to read a newspaper despite normal ability to read the individual letters on an eye chart. Some patients have their mild cataracts extracted in a vain effort to relieve such a visual problem.

Another common visual problem is an inability to maintain eye contact during conversation. This can give the mistaken impression that the patient is senile, hostile, or uninterested. The same eye movement problem can create the symptom of tunnel vision and interfere with driving a car.

The most common and characteristic eye movement problem in PSP is an impaired ability to move the eyes up or down. This can interfere with eating or with descending a flight of stairs, among other things. This problem is not usually as vexing for the patient and family as the inability to maintain eye contact or to coordinate eye movements while reading, but is much easier for the doctor to detect. Reduced vertical eye movement is usually the first clue that the diagnosis is PSP. Although other conditions, particularly Parkinson's disease 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. In most people with PSP, the difficulty in downward eye movement starts out as a slowing of that movement. This can also interfere with vision, but can be very difficult for a physician to detect. 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.

Yet another eye problem in PSP can be abnormal eyelid movement—either too much or too little—called blepharospasm. A few patients experience forceful involuntary closing of the eyes for a few seconds or minutes at a time, while others have difficulty opening the eyes, even though the lids seem to be relaxed.

They may try to use the muscles of the forehead, or even the fingers, in an effort to open the eyelids (called apraxia of lid opening). About 20% of patients with PSP eventually develop one of these problems. Others, on the contrary, have trouble closing the eyes, and therefore, these people blink very little. While about 15–25 blinks per minute is normal, people with PSP blink, on average, only about 3–4 times per minute.

This reduced blinking can allow the eyes to become irritated and react by producing extra tears, which in itself can become annoying.



### *What sort of speech problems occur?*

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. Speech becomes slurred in most patients after three or four years, on average, although it is the first symptom in a few patients. In Parkinson's disease, the speech problem is characterized by soft volume and rapid succession of words. In PSP, however, the speech may have an irregular, explosive, or rubber-band quality (called spastic speech) or a drunken quality (ataxic speech) or may have the same softening of speech as Parkinson's disease. Most commonly, there is a combination of at least two of these three features in the speech of patients with PSP.

The speech difficulty of PSP, in combination with the forgetfulness, slow mental responses, personality change, apathy, and poor eye contact during conversation can create an erroneous impression of senility or dementia. True dementia of a sort does occur in many people with PSP, however, and is discussed below.

### *What about the swallowing problems?*

This is the source of the most important and dangerous long-term complications of PSP. Swallowing tough foods or thin liquids can become difficult because of throat muscle weakness or incoordination. This tends to occur later than the walking, visual, and speech problems, but can become very troublesome if the patient tends to choke on food or if food goes into the breathing passages. Usually, problems managing thin liquids precedes difficulty with solid food because the swallowing muscles have difficulty creating a watertight seal that separates the path to the stomach from the path to the lungs. This is true with many neurological diseases. For non-neurologic conditions, such as stricture of the esophagus, the difficulties start with solid foods. Repeated, minor, often unnoticed episodes of small amounts of food and drink dripping into the lungs can cause pneumonia. Often, it is not apparent to the physician or family that the PSP patient's pneumonia is in fact the result of subtle aspiration. But aspiration pneumonia is the most common cause of death in PSP.

The risk of aspiration is aggravated by the tendency to overload the mouth or to take big gulps of beverages because of a loss of inhibition or a reckless impulsiveness that can be partly involuntary.

## *Does PSP lead to dementia as in Alzheimer's disease?*

Most patients do eventually develop some degree of mental impairment during the course of the disease. Some, however, are mislabeled as having Alzheimer's disease. This is not unlike the situation in Parkinson's disease.

Dementia in PSP, if it does occur, does not feature the memory problem that is so apparent in Alzheimer's disease. Rather, the dementia of PSP is characterized by slowed thought, difficulty resisting impulses, and difficulty synthesizing several different ideas into a new idea or plan. These mental functions are performed mostly by the front part of the brain (the frontal lobes). In Alzheimer's, on the other hand, the problem is mostly in the part of the brain just above the ears (the temporal lobes), where memory functions are concentrated.

Alzheimer's disease also includes either difficulty with language (such as trouble recalling correct names of common objects) or difficulty finding one's way around a previously familiar environment. Fortunately, these symptoms almost never occur in PSP. Nevertheless, these problems can interfere to a major degree with the ability to function independently, and the patient's irritability in some cases can make it difficult for caregivers to help.

Slowing of thought can cause major problems for people with PSP by making it difficult to partake in conversation. A question may be answered with great accuracy and detail, but with a delay of several seconds to minutes. Probably the most important aspect of PSP dementia is apathy. People with PSP seem to lose interest in their surroundings, creating the impression of lost thinking ability and interfering with family interactions.



## *How is PSP different from Parkinson's disease?*

Both PSP and Parkinson's disease cause parkinsonism (with a small p)—a combination of stiffness, slowness, and clumsiness. This is why PSP may be difficult to distinguish from Parkinson's disease early on. However, shaking (tremor), while prominent in about 70% of people with Parkinson's disease, occurs in only about 10% of people with PSP. A more common type of tremor occurring in PSP is irregular, mild, and present only when the hands are in use, not at rest as in Parkinson's disease.

Patients with PSP usually stand up straight or occasionally even tilt the head backwards and tend to fall backwards, while those with Parkinson's usually are bent forwards. The problems with vision, speech, and swallowing are much more common and severe in PSP than in Parkinson's. Parkinson's causes more difficulty using the hands and more stiffness in the limbs than does PSP. Finally, the medications that are so effective for Parkinson's disease offer much less benefit in PSP. The PSP-parkinsonism variant of PSP is more likely than typical PSP to have a tremor, 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.

Most drugs for Parkinson's disease enhance, replace, or mimic a brain chemical called dopamine. Parkinson's responds better to such drugs than does PSP because dopamine deficiency is by far Parkinson's most important abnormality. In PSP, deficiencies of several other brain chemicals are at least as severe as the dopamine deficiency, and no good way exists to replace those. Also, in PSP, there is damage to the brain cells that receive the dopamine-encoded messages, while these remain intact in Parkinson's.

### *What about treatment with medication?*

Several prescription medications can help patients with PSP, in some cases. Levodopa and carbidopa are the almost universally prescribed generic form of the brand name Sinemet. Levodopa is the component that helps the disease symptoms; carbidopa simply helps prevent the nausea that levodopa can cause. When levodopa came along in the late 1960s, it was a revolutionary treatment for Parkinson's, but it is of only modest benefit in PSP. It can help the slowness, stiffness, and balance problems of PSP to a degree, but usually not the mental, speech, visual, or swallowing difficulties. About 50% of those with PSP-parkinsonism respond to levodopa/carbidopa, while the figure is only 14% for Richardson's syndrome. The drug typically loses its benefit after two or three years, but a few patients with PSP continue to respond.

Some patients with PSP require large dosages to see an improvement—up to 1,200 milligrams of levodopa (with carbidopa) per day—so the dosage should generally be raised to at least that level under the close supervision of a physician, unless a benefit or intolerable side effects occur sooner. The most common side effects of this drug in PSP are confusion, hallucinations, and dizziness. These typically disappear after the drug is stopped. The most common side effect in patients with Parkinson's disease, involuntary writhing movements (chorea or dyskinesias), occur very rarely in PSP, even at high dosages.

Patients with PSP should generally receive the standard Sinemet or generic carbidopa/levodopa preparation rather than the controlled-release (Sinemet CR or generic carbidopa/levodopa ER) form. The CR or ER form is absorbed from the intestine into the blood slowly and can be useful for people with Parkinson's disease who respond well to carbidopa/levodopa but need to prolong the number of hours of benefit from each dose. In PSP, however, such response fluctuations almost never occur. Because the CR or ER is sometimes absorbed very little or erratically, a poor response in a patient with PSP might be incorrectly blamed on the fact that the disease is usually unresponsive to the drug. Such a patient might actually respond to the standard form, which reaches the brain in a more predictable way.



For people with PSP who cannot swallow pills safely, a solution is to crush a regular levodopa-carbidopa tablet into a food or beverage that is easily swallowed. The drug dissolves best in acidic beverages such as juices or sodas.

Another formulation of levodopa-carbidopa combines those two drugs with a third drug, entacapone, in the same tablet called Stalevo. The entacapone slows the rate at which dopamine is broken down. It is useful for patients with Parkinson's but rarely, if ever, in PSP.

There are three dopamine receptor agonists drugs on the market for Parkinson's—Mirapex (pramipexole), Requip (ropinirole), and Neupro (rotigotine, which comes only as a skin patch). For PSP, these rarely give any benefit beyond that provided by carbidopa/levodopa and may cause hallucinations and confusion, excessive involuntary movements, dizziness, and nausea.

Antidepressants have also had some modest success in PSP, sometimes relieving the depression that can be part of the disease. There are many antidepressants and none has been shown to be superior to any others. The older, tricyclic antidepressants seem to be no less effective in PSP than the newer reuptake blocker antidepressants.

Amantadine has been used for Parkinson's since the 1960s. It can be effective against the PSP gait disorder even if Sinemet is not, possibly because it affects more than just the dopamine system. Its benefit generally lasts only a few months, however. Its principal potential side effects are dry mouth, constipation, confusion, swelling of the ankles, and a pink skin discoloration in a lacy pattern called livedo reticularis. In people with PSP, the dosage should be kept low, generally no more than 200 mg per day, because of the potential for confusion or agitation.

Drugs for dementia, including Aricept (donepezil), Reminyl (galantamine), and Exelon (rivastigmine), enhance the activity of the brain chemical acetylcholine and are modestly useful against the dementia of Alzheimer's disease. They can also be somewhat useful in Parkinson's disease and other forms of dementia. None of them has been found helpful in PSP, but rivastigmine is probably worth trying. A fourth anti-Alzheimer drug, Namenda (memantine), acts on a different brain chemical, glutamate. It works no better for PSP than the others and can cause confusion and agitation in patients with PSP.

Although the jury is still out, one possible success story is the dietary supplement Coenzyme Q-10 (CoQ10), which is available without a prescription. It helps the body's cells produce energy from sugar and oxygen and is a normal constituent of the mitochondria, the tiny compartments in our cells where that chemical process occurs. One small study gave modestly favorable results. Another gave similar benefit but was too small to be sure that its outcome was not just the result of chance. The required dosage of the standard formulation of CoQ10 is probably at least 1,200 mg per day and perhaps as high as 2,400 mg. The newer liposomal form probably gives the same benefit at 300 mg per day. As a nonprescription item, CoQ10 is not covered by drug insurance and costs $50 to $100 month. Therefore, people with PSP should carefully consider the meager evidence to date for the benefit of CoQ10 before taking that long-term financial plunge. Their neurologists should do a careful exam upon starting the drug and repeat it two months later to determine whether the treatment is working, and if not, discontinue it.



Botox or Myobloc, two types of botulinum toxin, are a different sort of drug that can be useful for people whose PSP is complicated by blepharospasm. A very dilute solution of the toxin, which is produced by certain bacteria that can contaminate food, can be carefully injected by a neurologist into the eyelid muscles as a temporary remedy for abnormal involuntary eyelid closure. Botox can also be used for involuntary turning or bending of the head that occurs in PSP, but injecting it into the neck muscles can sometimes cause slight weakness of the nearby swallowing muscles. In PSP, where swallowing is already impaired in many patients, caution should be used when considering use of Botox in neck muscles.

## *What about other experimental drugs?*

In the past 20 years, many drugs have been tested in patients with PSP. Some of these are intended to actually slow the long-term brain cell loss, thereby slowing the progression of the disease. Unfortunately, none has helped. The most promising were riluzole, which modestly helps amyotrophic lateral sclerosis (ALS); davunetide, which is a fragment of a protein that helps maintain brain cells' internal skeletons; and tideglusib, which prevents the abnormal attachment of phosphate to the tau protein, preventing it from clumping up in the brain cells.

The last drug, tideglusib, did show a glimmer of promise: MRI scans showed less loss of brain mass in the patients on tideglusib than in those on placebo. This may justify further study of the drug, but it is doubtful that any drug company will want to pursue it after it failed to slow progression of the actual symptoms of the disease.

Two other approaches to slowing the progression of PSP have entered clinical trials. One gives antibodies designed to attack the abnormal tau protein as if it were an invading virus or bacterium. There are two such trials, sponsored by the companies Bristol-Myers Squibb and AbbVie. Another uses a version of the cancer drug Taxol to help maintain the brain cells' inner skeletons, which break down in PSP.

Two current, small, preliminary trials use existing drugs that are marketed for other diseases. One study administers Salsalate, a nonsteroidal anti-inflammatory drug, which may prevent the tau protein from aggregating via a mechanism unrelated to its anti-inflammatory effect. Another tests the drug methylene blue or LMTX in Alzheimer's disease, hoping it will also be useful in PSP.

Two drugs that reduce the tendency of tau protein to aggregate by altering the ability of phosphate molecules to attach to it will enter trials in the next year or two. The two drug companies involved are Asceneuron and Merck. Fortunately for PSP sufferers, drug companies have started to act on the realization that a prevention or disease-slowing treatment for PSP, where the market is tiny, could also work in Alzheimer's disease, where the market is huge.

### *Is tube feeding advisable for advanced patients?*

For extreme cases of poor swallowing where choking is a definite risk, the placement of a tube through the abdomen into the stomach (called gastrostomy, percutaneous endoscopic gastrostomy, or PEG) may be advised. PEG feeding may allow patients to regain lost weight, avoid hunger, and receive the nourishment they need to fight off other potential complications of PSP. A patient receiving the necessary nutrients and fluids is much happier and stronger overall and will probably find general movement, speech, and thinking easier.



PEG placement may be considered when any of the following occur: a first episode of aspiration pneumonia, small amounts of aspiration with each swallow, significant weight loss from insufficient feeding, or when the prolonged time required for a meal disrupts the operation of the household.

The PEG tube can be inserted with the patient awake but sedated, often as an outpatient procedure. The tube is clamped shut and hidden under the clothes when not in use. The feeding can be managed easily at home by pureeing the family's regular food in a blender and injecting it into the tube with what looks like a basting syringe. The skin site where the tube enters requires only a little care that can be provided easily by a family member or even by the patient in some cases. If the need for tube feeding abates (as through a new medication, for example), normal oral feeding can be resumed, and the tube can be kept as a backup or else removed.

One potential downside of tube feedings for some patients is a loss of feeling "whole" or part of humanity. The issue of how a feeding tube will affect a patient's quality of life must be considered carefully by the family, physician, and if possible, the patient, and sometimes ethical or spiritual advisors. Some patients in the advanced stages of PSP may feel that their quality of life is so poor that prolonging that life by having a PEG installed is not what they want. It's important to note that some nursing homes will advise PEG placement because it reduces the personnel time needed to feed the patients and because third-party payers often will pay an additional fee for tube feeding but not for the time-consuming task of hand feeding a patient by mouth.

## *Do any of the new brain operations for Parkinson's work for PSP?*

Not so far, unfortunately. The operations for Parkinson's disease fall into two categories. One is based on the theory that the basal ganglia (the group of nuclei that control movement) are overactive. At present, the most common operation to dampen down this overactivity is subthalamic nucleus stimulation. The previous approach, pallidotomy, is rarely performed now. In PSP, however, the output area of the basal ganglia is damaged, so its activity is already dampened. The operation would only make things worse.

There are, however, trials now in progress in people with PSP to test stimulation of the area of the brain that serves balance—the pedunculopontine nucleus (PPN). The PPN is in the brainstem, which is an area tightly packed with critical circuitry. The procedure seems to be acceptably safe and does seem to help the balance problem in some patients with Parkinson's disease, but the overall improvement in patients is still undetermined.

In patients with Parkinson's, there is also an operation that attempts to replace the lost dopamine-producing brain cells. This operation is unlikely to work for PSP because, while in Parkinson's, most of the movement problem is caused by loss of the main dopamine-producing nucleus (the substantia nigra), the movement problems in PSP are caused by the loss of many additional nuclei. Many of those other nuclei receive their input from the substantia nigra, so replacing only the first link in the chain would not help much. In addition, at this point, it would cause too much trauma to the brain to replace cells in all of the nuclei involved in PSP.

An exciting experiment in progress in Milan, Italy, but one that has only a slim chance of success, is the injection of stem cells directly into the arteries feeding the brain. This project is still in the very early phases designed to test its safety. If it succeeds, then a large project will test for benefit. The intent is not to replace lost cells, as the injections land in random spots, but to stimulate the brain to produce its own growth factors to repair the damage of PSP.



## *What about other non-drug treatment?*

Formal physical therapy is worth a trial in PSP, especially with the goal of teaching the patient to use gait-assistive devices, such as a walker.

Certain exercises done in the home on a regular schedule can keep the joints limber. Exercise also has a clear psychological benefit that improves a sense of well-being for anyone with a chronic illness. For specific exercises, consult one of the books for patients with Parkinson's disease or the pamphlets distributed by the national Parkinson organizations.

The special balance problems in PSP dictate caution in performing any exercises while standing. Many useful exercises can be performed seated in a chair or lying on a mat. Using a stationary bicycle is usually feasible as long as there is help in mounting and dismounting safely. The best strategy is to have an evaluation and treatment plan from a physical therapist or physiatrist (a physician specializing in rehabilitation of chronic conditions).

Probably the most important part of dealing with PSP is for the patient's family to understand that the problems with visual inattention and personality changes are part of the illness. The patient is not lacking willpower nor "faking." Furthermore, many of the problems in PSP are intermittent and can be aggravated by the patient's mental or emotional state. For example, walking, writing, and eating may be poor one hour and better the next. The family should understand that these fluctuations are not under the patient's control and that nagging and shouting usually make matters worse. A wise policy is to be prepared to take advantage of the good periods to have an outing, a relaxing shower, or some other activity that would be more difficult at another time.

Walking aids are often important for patients with PSP. If a walker is required, it should be weighted in front with sandbags over the lower rung because of the tendency for PSP patients to fall backwards. A better but more expensive solution is a large, heavy walker resembling a small shopping cart with three or four fat, soft rubber wheels and a hand brake. The tendency to fall backwards can also be countered by the use of built-up heels. Leg braces are not helpful because the problem in PSP is coordination and balance rather than actual muscle weakness.

Shoes with smooth soles are often better than rubber-soled athletic shoes. In many people with PSP, the gait disorder includes some element of "freezing," a phenomenon that makes it difficult to lift a foot from the ground to initiate gait. People with such symptoms can fall if they move their body forward before the foot moves. In these cases, smooth soles could make it easier to slide the first foot forward.

Handrails installed in the home, especially in the bathroom, may also be helpful. The difficulty looking down dictates that low objects, such as throw rugs and low coffee tables, be removed from the patient's living space.



To remedy the difficulty of looking down, bifocals or special prism glasses are sometimes prescribed for people with PSP. These are worth trying but can be of limited value because there is not only a problem moving the eyes in PSP but also a problem directing the person's attention (the "mind's eye") to objects located below the eyes.

### *What is the cause of PSP?*

The symptoms of PSP are caused by a gradual deterioration of brain cells in a few tiny but important places in the base of the brain. The most important such place, the substantia nigra (pronounced sub-STAN-cha NYE-gra), is also affected in Parkinson's disease. Damage to it accounts for the symptoms that PSP and Parkinson's have in common. However, several important areas that are affected in PSP are normal in Parkinson's, and vice versa. And, under the microscope, the appearance of the damaged brain cells in PSP is quite different from those in Parkinson's and instead resembles the degeneration in Alzheimer's disease. In addition, the location of the damaged cells is quite different in PSP and Alzheimer's. Also, PSP lacks amyloid plaques, which are deposits of waxy protein that are a hallmark of Alzheimer's.

### *But what causes the brain cells to degenerate in the first place?*

No one knows yet, but we have some clues. In the brain cells that are degenerating in PSP, there is an abnormal accumulation of the normal protein tau. These clumps of tau, once they reach a size that can be seen through a microscope, are called neurofibrillary tangles. The normal function of tau is to help support the microtubules, which have two important jobs: one is to form the internal "skeleton" of the brain cells, and the other is to serve as a "monorail" system, transporting nutrients around the cell. We don't know whether the problem is that the tau is defective from the time of its manufacture or if it is damaged later, or even if it remains normal but is produced in excess. If it is damaged, the nature of that damage could be the excessive attachment of phosphate (see the section on PSP preventive drugs). Or, the excessive phosphates

on the tau molecules could simply be how the brain minimizes the effects of tau protein that is misbehaving for some other reason.

Regardless of its origin, the excessive phosphate on the tau protein molecules causes the tau to misfold. Ordinarily, tau protein that's not attached to microtubules floats around without a fixed shape in the cell's fluid, like a strand of overcooked spaghetti in boiling water. But when it misfolds, it assumes a more rigid structure, like a strand of dried-out spaghetti. Like spaghetti, it's sticky and it forms clumps with other misfolded tau molecules.

In the past few years, evidence has been discovered that a single strand of misfolded tau can cause normal, freely floating copies of tau protein to misfold in the same way. This is called a templating process. The newly misfolded copies then cause other copies to misfold, and so on, in a chain reaction. Then the misfolded tau molecules start to clump. The clumps are toxic to the cell, which eventually dies. Even before the cell dies, it releases misfolded, clumped tau protein into the fluid surrounding the cells. That tau is then taken up by neighboring healthy cells that undergo the same damaging chain reaction of tau misfolding, templating, and clumping. In this way, the process of brain cell malfunction and death spreads slowly through the brain. A similar process is thought to occur in most of the neurodegenerative diseases but with different kinds of proteins in different diseases. This insight raises the tantalizing possibility that a drug that prevents that templating process could prevent all neurodegenerative diseases.

Since the 1980s, it's been known that one type of protein in the cell, called prion protein (pronounced PREE-on), does in fact misfold, template itself, and form toxic clumps to cause certain neurodegenerative diseases, such as mad cow disease and Creutzfeldt-Jakob disease. Those diseases progress very rapidly and can be transmitted from one individual to another via exposure to diseased tissue. But misfolded prion protein behave very differently from misfolded tau protein, and PSP, Parkinson's, Alzheimer's, and the other neurodegenerative diseases progress far more slowly and are not transmittable between people. They should not be lumped with the true prion diseases.



## *Is PSP genetic?*

PSP only very rarely runs in families. Fewer than one in 20 people with PSP knows of even one other family member with PSP. 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 60–77% of the rest of us. So clearly, the H1 haplotype is (nearly) necessary but far from sufficient to cause the disease.

We're still not quite sure how the H1 haplotype increases PSP risk. One possibility is that it simply increases the amount of tau produced, which causes that protein to stick together, even if it's not misfolded. Another possibility, discovered only recently, is that it causes too many methyl groups to stick to the tau gene, which alters its function. A methyl group is simply a carbon atom with three hydrogens that can attach to large molecules, including DNA. Methylation is a normal way for the cell to regulate the function of DNA, thereby affecting the function of genes without actually changing the content of the genetic code like ordinary mutations do. This is exciting because certain molecules have the potential to be developed into drugs that could alter DNA methylation.

Insights into the other PSP-related variant in the tau gene were published in 2011 by a group of scientists working in the Charles D. Peebler Jr. Genetics Program, sponsored by CurePSP. The approximate location of this defect in the tau gene is known, but its relationship to the brain cell loss of PSP is not yet understood. This has been the subject of intense research since 2010. CurePSP's Peebler Genetics Program has also discovered several other genetic variants that are more common in people with PSP than in those without PSP. One, called EIF2AK3, makes an enzyme that helps control the brain's system for disposing of misfolded proteins. This could explain why the clumps of tau protein form. Another gene newly implicated in PSP, called STX6, helps direct the movement of packets of important chemicals within brain cells. Transporting such packets is one of the functions of the microtubules, the internal skeleton or monorail system that the tau protein maintains. A third, MOBP, is the blueprint for the manufacture of a protein in brain cells' myelin sheath, which serves as a layer of electrical insulation on the wires connecting brain cells together. Still other PSP-associated genes help control the body's immune system, although their exact relation to PSP remains unknown.

In 2016, CurePSP, in conjunction with the Tau Consortium, formed the PSP Genetics Consortium, an international team of investigators. Unlike the 2010 study, which was only able to find the approximate location of genes, this effort will work out the genetic code of all genes that make proteins in a group of people with PSP and in a similar group without PSP. This will not only discover additional genes but will also show exactly what those "typos" in the genetic code are.

The next step will be to figure out how those errors damage the normal function of brain cells. Those insights, in turn, will provide new ideas for drug targets. In other words, once scientists work out which steps in the brain cells' normal function become disordered as a result of these PSP-related gene variations, they will know where new drugs could be directed to prevent the process from proceeding further. Coupled with a test to detect PSP in its earliest stages, before it actually causes any disability, such a drug treatment would amount to a PSP prevention. This multistep process is the basis of CurePSP's Research Roadmap to a cure.



### *Could PSP be caused by toxins?*

There is evidence that chemicals in the environment or diet may contribute to the cause of PSP. Surveys of PSP patients have shown, on average, lesser educational attainment in people with PSP. This suggests that part of the cause of PSP may be certain occupational factors exposing people to different chemicals than are encountered by people with more sedentary or office-bound occupations. Another possibility is that people with less education tend to live in areas closer to industrial sites, some of which may generate toxins.

One important clue to a possible dietary factor in the cause of PSP comes from the island of Guadeloupe in the Caribbean. People there are far more likely to develop PSP and other atypical parkinsonisms than people elsewhere. A questionnaire survey on Guadeloupe revealed that people there with PSP-like illnesses were more likely than others to have consumed two native fruits called sweetsop and soursop. These fruits have since been shown to harbor toxins that, when given to laboratory rats, cause damage to the brain very similar to human PSP. We don't yet know what foods in the Western diet, if any, may contain similar toxins. Research on that question is underway.

Another intriguing geographical cluster of PSP exists in a group of suburban towns in northern France. The area was the site of metal-related industry that contaminated the soil in that area over much of the 20th century. Investigations are now underway to narrow the long list of toxins that could explain this cluster and then assess the role of such toxins in PSP elsewhere.

## *How can I help research?*

CurePSP welcomes donations to its research grants program. Since its inception in 1997, the program has provided over $10 million to institutions and senior researchers with excellent track records of productivity and to junior people with original ideas and first-rate training. CurePSP favors projects with the potential to produce preliminary findings that would support an application to a government agency for a much larger grant in the future. It does not restrict its grants to any country or continent.

The various national organizations that sponsor research in Parkinson's disease sometimes sponsor deserving PSP research. Their support of research in Parkinson's disease adds to our knowledge of PSP.

Another way to help research and yourself is to participate in studies of PSP if requested by a researcher. This may take the form of answering questionnaires, having medical examinations or tests, or taking experimental medication. There are so few people with PSP in any one geographical area that each person can make a very important contribution. Joining the mailing list at CurePSP will allow PSP researchers to contact you regarding participating in new research studies.

## *Should I join a support group?*

There can be great value in joining a group of other people with the same problem. You can exchange helpful tips on ways to cope physically and psychologically with the limitations of the illness and can learn more about the problem and its treatment from guest speakers. Many large medical centers have a Parkinson support group that welcomes members with PSP. While there are far fewer people with PSP than Parkinson's in one geographical area, several dozen successful PSP support groups have been organized in the U.S., usually in more densely populated areas. All it takes is one organizer with some time and energy. See the CurePSP Support Groups chapter for information.



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## *What is corticobasal degeneration (CBD)?*

As its name states, corticobasal degeneration is a loss of brain cells emphasizing the cerebral cortex and the basal ganglia. The cortex is the outer layer of the cerebrum, the gray matter where most thinking, speech, and sensory perception occur. The basal ganglia are also gray matter, but they're deeper in the cerebrum and they coordinate movement. Basal refers to the base of the brain; ganglia are collections of brain cells in one location devoted to a single purpose.

## *What are its main signs and symptoms?*

CBD is considered one of the "parkinsonian" disorders, or "parkinsonisms." There are about a dozen such diseases, all of which produce some degree of slowness, muscle stiffness, balance problems, and sometimes tremor. Most people with CBD have these things, but in addition they have unusual difficulty performing complex limb movements, such as cutting food, buttoning, or typing. There is often a tendency to hold part or all of a limb in a fixed posture called dystonia. There can also be very rapid, irregular, small movements of muscles called myoclonus.



The unusual feature about CBD is that it is almost always very asymmetric, with one side affected much earlier and worse than the other. In half of people with CBD, it's the left side, and in half, it's the right. The side affected has no relationship to the person's handedness.

An important part of CBD that is often difficult for patients and families to understand is apraxia. This is the loss of the ability to perform a complex movement that required some practice to learn in the first place. Manual tasks or gestures become clumsy, and walking can become frozen for several seconds at a time. Common examples of apraxia are losses of the ability to use eating utensils or to manage buttons. An unusual but dramatic type of apraxia is alien limb phenomenon, where one hand seems to belong to someone else and can perform actions that oppose the person's intentions. Another unusual type of apraxia is arm levitation, where one arm tends to move upward involuntarily.

Contributing to the movement problem of CBD is a sensory problem. It's not a simple loss of sense of touch but an inability to interpret spatial complexity involving touch. This can take the form of an inability to recognize common objects by feel alone or the inability to know the position of a finger or a limb in space.

## *How does CBD start?*

The first symptom is usually apraxia in one hand, but sometimes it starts as Parkinson's disease does, with general slowing and stiffness, perhaps with a mild tremor and slurred speech. Over the next few months, the symptoms worsen, and the dystonia may start. Sometimes CBD starts with aphasia, which is difficulty producing or understanding language.

## *What happens later?*

The apraxia and dystonia usually spread to the other arm within two or three years and can affect the legs and feet as well, which can cause unsteady walking. There can be slurring of speech and difficulty swallowing liquids, with coughing and even irritation of the lungs from fluids that drip down. Some people develop difficulty multitasking or organizing their thoughts. Some individuals can lose some behavioral inhibitions.

## *How common is CBD?*

CBD is very rare: about five people per million, which comes to about 1,600 people in the United States, most of whom have not yet been accurately diagnosed. For Parkinson's, the figure is 700,000, and for Alzheimer's disease, 5.8 million.

## *How does CBD differ from the more common brain degeneration disorders?*

Unlike Parkinson's, CBD usually has little tremor. Unlike PSP, CBD has only mild problems with balance and eye movements. However, CBD does have features that these other diseases usually lack, such as apraxia, dystonia, and myoclonus. Unlike all of these other diseases, CBD is highly asymmetric in its limb involvement, meaning that either the right or left limbs start to show problems well before the opposite side and remains the worse side throughout. Parkinson's disease is also asymmetric but not to the same degree.

In CBD, some degree of cognitive loss is present in half of individuals at the outset and in 70% eventually. If it does occur, it does not feature the memory problem that is so apparent in Alzheimer's disease. Rather, the dementia of CBD is often characterized by trouble in planning, resisting impulses, and synthesizing information into a new idea. These mental functions are performed mostly by the front part of the brain (the "frontal lobes"). In Alzheimer's, on the other hand, the problem is mostly in the part of the brain just above the ears (the "temporal lobes"), where memory functions are concentrated. CBD may in some cases also feature difficulty in recalling or understanding words ("aphasia"), loss of visual-spatial skills, loss of ability to order a complex manual task ("apraxia"), disinhibited social behavior, and general loss of interest in one's problems or in one's environment ("apathy").



## *What happens in the brain?*

In people with CBD, some groups of brain cells break down and die off. Central to that process is the abnormal folding and clumping of a protein called tau, which is a normal component of the brain. Its job is to help maintain the microtubules, which are stiff rods that function as the brain cells' internal transportation and skeletal system. It appears that the cells' death is caused by the tau protein clumps themselves and not by the reduced availability of tau for its normal function. The clumps' technical name is neurofibrillary tangles.

We know several reasons why tau might misfold, but we don't know which one(s) are relevant to CBD. Tau forms similar aggregates in about a dozen other diseases, including Alzheimer's disease, PSP, chronic traumatic encephalopathy (CTE), and about half of all cases of frontotemporal dementia. The specific disease depends on the chemical characteristics of the tau aggregate and which brain cells are involved. We don't yet understand what determines these.

### *What are the various subtypes of CBD?*

The microscopic brain abnormalities of CBD can affect different parts of the brain, producing different sets of outward symptoms in different people. The first variant to be described in the medical literature is called corticobasal syndrome and is the most common. It affects about half of all people with CBD and is the archetype of CBD described above.

About 25% of people with CBD have an outward appearance that resembles the most common form of PSP, which features problems with balance, eye movement, speech, and swallowing. Another 15% have frontotemporal dementia, with inappropriately uninhibited behavior and difficulty organizing thoughts. Then there are two rare forms, each accounting for about 5% of the total. One has a dementia similar to that of Alzheimer's disease, with important problems with memory or spatial orientation. The other is a form of aphasia, a problem with language, in this case difficulty finding words and obeying rules of grammar.

All forms of CBD at some point include, in almost all patients, a degree of "motor parkinsonism," meaning stiffness, slowness, soft speech, and reduction in facial expression, sometimes with balance difficulty and tremor..

### *Is CBD genetic?*

CBD almost never runs in families. However, a variant in the gene on chromosome 17 that encodes the tau protein is a little more common in CBD than in the rest of the population. Called the "H1 haplotype," it occurs in 92% of people with CBD and in 60-77% of the rest of the population. So the H1 haplotype is (nearly) necessary but far from sufficient to cause the disease.



We're still not quite sure how the H1 haplotype increases CBD risk. One possibility is that it simply increases the amount of tau produced, which causes that protein to stick together, even if it's not misfolded. Another possibility, discovered only this year, is that it causes too many "methyl groups" to stick to the tau gene, which alters its function. A methyl group is simply a carbon atom with three hydrogens. It can be attached to large molecules including DNA. Methylation is a normal way for the cell to regulate the function of DNA, thereby affecting the function of genes without actually changing the content of the genetic code like ordinary mutations do. This is exciting because certain molecules have the potential to be developed into drugs that could alter DNA methylation.

A detailed analysis of the genetics of CBD was published in 2015 by an international group of researchers. The project was partly sponsored by CurePSP. It found five genetic variants to be associated with CBD in addition to the H1 haplotype in the gene for tau. One of these genes, called MOBP, is also associated with PSP. It is the blueprint for the manufacture of a protein in brain cells' myelin sheath, which serves as a layer of electrical insulation on the wires connecting brain cells together. These findings await confirmation by other research groups using other methods.

If these gene variants are confirmed, the next step will be to figure out how those errors damage the normal function of brain cells. Those insights, in turn, will provide new ideas for drug targets. In other words, once scientists work out which steps in the brain cells' normal function become disordered as a result of these CBD-related gene variations, they will know where new drugs could be directed to prevent the process from proceeding further. Coupled with a test to detect CBD in its earliest stages, before it actually causes any disability, such a drug treatment would amount to a CBD prevention.

## *How is CBD treated?*

Unfortunately, CBD almost never responds to levodopa, the drug that is the mainstay of Parkinson's disease treatment. However, because there are rare exceptions, and because the diagnosis of CBD may be wrong, it's usually worth a try in people with apparent CBD and who have important muscle rigidity or slowing of movement. A typical approach is to start the carbidopa-levodopa 25/100 size at one tablet once a day on a full stomach and to increase each week by one tablet per day until reaching three per day (taken as one tablet three times per day). The dosage can be increased at weekly intervals to six tablets per day, then nine, then 12. If that doesn't help, then the drug should be quickly tapered and discontinued. The most common side effects of carbidopa-levodopa in people with CBD are nausea and sleepiness, each occurring in about 10% of patients.

There is no evidence that other antiparkinson drugs—such as the dopamine agonists (pramipexole, ropinirole, rotigotine patch)—help, and, in fact, they can have more side effects than levodopa. An exception may be amantadine, an old antiparkinson drug with a complex chemical mechanism that can sometimes help the gait freezing of CBD. The dosage of that drug should not exceed 200 mg per day because of its possible side effects of confusion, constipation, and urinary retention.

The dystonic muscle spasms that sometimes occur in CBD may respond to muscle relaxant drugs, such as cyclobenzaprine, baclofen, and tizanidine. A medication for seizures called levetiracetam can also help and may be better tolerated than the traditional muscle relaxants. The most common side effect of these drugs is sleepiness. The myoclonus of CBD, in the unlikely event that it is troublesome, may respond to clonazepam or levetiracetam.

A different sort of drug that can be useful for people whose CBD is complicated by dystonia is botulinum toxin. This substance is produced by certain bacteria that can contaminate food. A dilute solution can be injected into overactive muscles. The effect takes one to two weeks to start and lasts two to three months before having to be repeated.

Unfortunately, deep brain stimulation surgery, which can be so useful in Parkinson's, does not help CBD.



## *Is physical therapy useful?*

Formal physical therapy is worth a trial in CBD, especially with the goal of teaching the patient to use gait assistive devices, such as a walker. Certain exercises done in the home by oneself on a regular schedule can keep the joints limber. For anyone with CBD, or any chronic illness, exercise also has a clear psychological benefit: improved sense of well-being.

For people with gait or balance problems, many useful exercises can be performed seated in a chair or lying on a mat. Using a stationary bicycle is usually feasible as long as the person has help in mounting and dismounting safely. The best strategy is to obtain a neurologist's order for an evaluation and a treatment plan from a physical therapist or physiatrist (a physician specializing in rehabilitation of chronic conditions). The same advice applies to the utility of occupational therapy in improving the apraxia and dystonia of the hand(s) that is so common in CBD.

### *Are there experimental treatments for CBD?*

Not so far, but there are experimental trials for PSP, and the two diseases are very similar at the molecular level. As far as we can tell at this point, anything that helps PSP could also help CBD. So it's a good idea to keep an eye on new developments in PSP. One problem in mounting drug trials in CBD is that there are so few patients in any one place that many sites would have to be recruited and coordinated (and paid) to have a proper trial.

Another problem is that about half of the patients who have the outward corticobasal syndrome actually have corticobasal degeneration, so any benefit of a drug could be diluted by the non-CBD patients in the trial, creating a false-negative result. Furthermore, any positive result in such a trial could be explained by an effect in some of the non-CBD patients, a messy result that would not satisfy most researchers, drug companies, or the FDA.

### *How long do people with CBD live?*

Most people with CBD encounter life-threatening complications between five and ten years after the first symptoms appear. However, some live much longer. Survival is enhanced by dedicated caregiver support, professional nursing care as needed in the more advanced stages, maintaining general health, and, perhaps most important, by an optimistic and hopeful attitude of the patient and family.



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## *What is multiple system atrophy (MSA)?*

Multiple system atrophy is a disease of the brain and spinal cord. It is often classified as one of the "parkinsonian" conditions because it often resembles Parkinson's disease, at least for the first few years. The resemblance is in the general slowness, stiffness, and balance loss. But MSA usually has at least two other important categories of symptoms to some degree. One is impairment of the cerebellum, producing a coarse tremor, drunken-appearing walk, and slurred speech. The other is impairment of the autonomic nervous system, which maintains such things as blood pressure, sleep, bowel action, and bladder emptying. The result can be fainting, insomnia, constipation, and urinary urgency or incontinence. Many other less common symptoms can occur, too.

## *How common is MSA?*

MSA is rare, with about 13,000 sufferers in the United States, most of whom have not yet received a correct diagnosis. This compares with about 20,000 with progressive supranuclear palsy (PSP), which is a similar condition; about 700,000 with Parkinson's disease; and 5.8 million with Alzheimer's disease. New cases of MSA arise in about six people per million per year. This means that in the U.S., about five people are newly diagnosed with MSA each day.



## *Are there different types of MSA?*

There are three types that grade into one another. MSA emphasizing slowness and stiffness was once called striatonigral degeneration. MSA emphasizing autonomic problems was called Shy-Drager syndrome. Cases emphasizing cerebellar problems were called sporadic olivopontocerebellar atrophy. These terms were discarded in 1989, when all three were found to be variants of the one disease, which then received its current name. What all three variations have in common is a type of protein that forms clumps in the same types of cells. The differences among the three types of MSA are dictated by which parts of the brain or spinal cord are most involved.

## *What is the cause of MSA?*

The ultimate cause (called the etiology of the disease) is not known. MSA almost never occurs twice in a family, and there are no confirmed clusters related to occupation, industry, diet, ethnicity, or geography. A variant in a gene called alpha-synuclein (SNCA) occurs more often in people with MSA than in the rest of the population, but this accounts for only a small fraction of the overall cause of the disease. Various chemical toxins have also been implicated, but these results have not been confirmed in multiple studies.

## *What's going wrong in the brain and spinal cord cells?*

The direct cause of the loss of brain and spinal cord cells (the pathogenesis of the disease) in people with MSA is not fully understood. However, it has to do with the clumps of alpha-synuclein protein mentioned above. Alpha-synuclein is a normal protein that is necessary for brain cells to signal to one another. The current favored theory is that too much alpha-synuclein is made. Once it reaches a certain concentration in the cell, it starts to stick together. The resulting blobs, when they are still too small to be seen with a microscope, are toxic.

The larger clumps seen under the microscope, called glial cytoplasmic inclusions, may actually be the brain's attempt to solidify the tiny, toxic clumps floating around inside the cell into a hard, relatively harmless form.

The same protein, alpha-synuclein, accumulates in the brain cells in Parkinson's disease, but in somewhat different parts of the brain. Also, in MSA, the initial accumulation and cell loss appear to be in the glia, the electrically inactive supporting cells of the brain. In Parkinson's disease, on the other hand, the problem starts in the neurons, the electrically active cells.

## *At what age does MSA start?*

The average age at which the symptoms of MSA begin is only 53. This is younger than the averages of 59 for Parkinson's and 63 for PSP.

## *What happens to someone with MSA?*

All three kinds of MSA feature important balance problems that can eventually require assistance in walking or even a wheelchair. All three types can also display difficulty in the aspect of thinking called executive function. This is what allows us to organize information by categories, understand abstractions and instructions, create and follow a plan, and inhibit inappropriate actions. These abilities often decline to some extent in people with MSA.

Other features occur in all three types of MSA. Perhaps the most important is difficulty swallowing, which obliges one to modify the diet to exclude thin liquids or tough solids. All three types of MSA can also have difficulties in moving the eyes, which can interfere with reading, and in speaking, which may require speech therapy and communication devices.



People with MSA of the parkinsonian type tend to get more muscle stiffness and general slowness. Those with the cerebellar type develop a coarse tremor when moving the limbs and difficulty aiming their limb movements or walking in a straight line, much like someone who is drunk. Those people with MSA that emphasizes the autonomic problem have a variety of symptoms, including urinary urgency and incontinence, constipation, erectile disturbance in men, lightheadedness or even fainting upon standing, noisy or irregular breathing, sleep disruption, and difficulties with temperature regulation. Most people with MSA have some combination of the three types rather than a pure form.

Some other problems with movement can occur in MSA. The hands can gradually assume abnormal, fixed postures called dystonia. This can be prevented to some degree by stretching exercises and can be treated in some cases by botulinum toxin (Botox) injections into the hands or forearms. Some people with MSA hold their heads bent forward to an extreme degree, a condition called antecollis. This too may respond to Botox injected into the neck muscles, although care must be taken not to exacerbate the swallowing problems via leakage of the Botox to the immediately adjacent muscles. Another issue in some people is sudden, rapid jerks of a limb or of the trunk. These issues can be annoying but are almost never large enough to interfere with normal movement. This is called myoclonus and can be treated with medication.

Finally, a tremor, which is not nearly as prominent as in most people with Parkinson's, can occur in MSA. This tremor generally happens when the limb is in use rather than at rest (as in Parkinson's). The MSA tremor may respond to medication but usually is too mild to require treatment.

Some people with MSA develop obstructive sleep apnea. This is where the upper airway tends to close during sleep, causing insomnia and sometimes insufficient blood oxygen levels. It can be treated by wearing a mask during sleep that connects to a kind of air pump that keeps the airway open, called continuous positive airway pressure (CPAP). This is usually best managed by a specialist in sleep medicine or pulmonary medicine rather than a neurologist.

### *Is MSA a fatal disease?*

On average, someone with MSA lives about seven or eight years after the onset of symptoms. This short survival is not a direct effect of the disease but of complications of the difficulties in swallowing safely and moving around. The most common of these complications are pneumonia, urinary tract infections, and blood clots in the legs that lodge in the lungs. Other potentially dangerous complications of MSA are low blood pressure, slow heart rate, sleep apnea, and injuries from falls.

### *How is MSA treated?*

We have no treatment or prevention for the underlying brain disorder, but some of the individual symptoms can be managed successfully with medications that raise blood pressure, enhance sleep, inhibit bladder emptying, or stimulate the bowel. Drugs for Parkinson's that stimulate the brain's dopamine system sometimes work for a few years for the general slowness and stiffness in MSA.

### *What happens eventually?*

Unfortunately, the progression of MSA is almost as rapid as that of PSP, with the average time from initial symptom to requiring a wheelchair of five years. Death occurs on average seven to eight years after the initial symptoms, usually from complications of the immobile state, such as pneumonia or other infections. Keep in mind that this is only an average duration of survival. Many people with MSA survive longer.

### *What research is being done?*



A critical defect in the brain cells in MSA is accumulation of the normal protein alpha-synuclein. The same protein accumulates in Parkinson's disease but in a different set of brain cells. In 2018 alone, 416 research papers on MSA were published in scientific journals, up from 227 papers in 2010. As scientists understand more about the various brain degenerative disorders, such as Alzheimer's, Parkinson's, PSP, Lou Gehrig's disease, and MSA, many commonalities among them are being revealed. That means that any breakthrough in one could benefit the others. It is entirely realistic to expect that after researchers find a prevention or a way of halting the progression of one of these diseases, the others will benefit similarly.

### *How can I help research in MSA?*

One way is to be alert for trials of new medication or new diagnostic tests. These are listed on a website maintained by the National Institutes of Health, *www.clinicaltrials.gov*. You simply enter "Multiple System Atrophy" into the search box. Participants in clinical trials may not only benefit from a new treatment that is not generally available, but they also often receive detailed care and attention that is not routine, even at excellent medical centers. Study participants also get the satisfaction of helping in the fight against their illness. Other trials may look for new genetic or environmental contributors to the cause of MSA. Discovering these could point to potential preventative measures or treatments.

PART TWO

# Managing Symptoms

## Managing Cognitive and Behavioral Changes

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The cognitive and behavioral changes of PSP can present a major challenge to the caregiver. Cognition concerns functions such as speech, memory, organization of thought, solving problems, and initiating new ideas. Behavior, on the other hand, concerns mood, appropriateness of behavior, and sociability.

The single most important piece of advice for those whose PSP is causing cognitive problems is to stay physically, mentally, and socially active.

It is important to establish a daily routine and stick to it, using a written or digital daily planner. The medications that may modestly help the memory problems of Alzheimer's do not, unfortunately, help much in PSP and can cause gastrointestinal and bladder overactivity, with cramps, nausea, diarrhea, urinary urgency, or even incontinence in some cases.



What appears to be depression in PSP is usually apathy, a very common feature of PSP that does not respond to antidepressants. It may respond to amantadine or to amphetamine-like medications, but this should be discussed carefully with the neurologist or psychiatrist, as those drugs can have significant side effects.

Caregivers must come to terms with the fact that the patient will not return to their previous normal cognitive or behavioral state. They must choose their battles carefully and avoid arguing with the patient, instead redirecting them to a more appropriate activity or location. It's okay to tell white lies as long as it's for the patient's safety and comfort. Support groups can be very helpful, both for the caregiver and the patient.

Respite services to keep the caregiver from burning out are very useful and may be available through local community agencies. A social worker can advise on respite care.

The cognitive and behavioral problems of PSP are likely to worsen temporarily during a period of unrelated stress. For example, pain arising from a bedsore, arthritis, sciatica, tight clothing, or an undetected infection can cause confusion, agitation, or exaggeration of any other existing cognitive/behavioral deficit. Common examples are bladder infections, pneumonia, infected bedsores, and even the common cold. Any physical illness, even if not painful, can do the same thing. Ordinary fatigue and lack of sleep are other common culprits, as is stress arising from excessive social expectations or pressure from the family to perform what the patient can no longer perform.

Another common cause of worsening cognitive or behavioral symptoms is inappropriate medication, such as sedatives, tranquilizers, sleeping pills, pain pills, antiparkinson medications, antidepressants, antihistamines, and antipsychotics (medication for hallucinations or delusions). Of course, use of alcohol or other recreational drugs can do the same.

People with PSP commonly have disinhibited behavior. It most commonly takes the form of attempting to perform physical tasks that are dangerous for them, such as getting up to walk unassisted, reaching for things beyond their grasp, overloading the mouth, speaking too loudly, constantly groaning, or repeating words or phases of speech unnecessarily. Rarely, disinhibited behavior can be sexual in nature, consisting of inappropriate remarks, touching oneself or others, or removal of one's clothes.

For both the sexual and non-sexual disinhibited behaviors, the caregiver should be careful not to react emotionally or to shout at the patient. Firmly but calmly correct the misbehavior, explain the problem, and redirect the patient's attention and activity. Sometimes antidepressant medication can help with this sort of behavior. Inappropriate sexual behavior may merely reflect a need for physical closeness that can be satisfied by a normal hug or backrub. Overloading the mouth can be avoided by cutting the food into small pieces or, if necessary, by feeding the patient. Disinhibited attempts to walk should not be controlled by seat belts or other strapping devices but by placement of a table in front of the seated patient or, even better, by an attentive aide. When the aide is not available, the patient can be put into a hospital bed with bedrails; however, creating a constant bedbound state is not an appropriate treatment for behavioral issues.

## Dry Eye Syndrome and Visual Issues in PSP

### Onur Melen, MD



#### *Definition of Dry Eye Syndrome*

Many people in the general population develop dry eye syndrome (also referred to as keratitis sicca). Left untreated, it can cause damage and roughness to the surface of the eyeball. If dry eyes are a problem, it is always best to consult an eye professional for advice and treatment. Dry eye syndrome is a common condition thought to affect approximately 60 million Americans. In some instances, the condition is characterized by dry, irritated eyes, caused by a lack of lubricating tears, which stems from an imbalance between tear production and tear volume drainage via the nasolacrimal ducts (NLD).

Dry eye syndrome can also result from excessively watery eyes due to tears lacking the proper balance of mucus, water, and oil to coat the eyes properly. The tear film is made up of a mucus layer against the eye, a middle aqueous (water) layer, and an outer lipid (oily) layer. All three components are critical to a normal tear film. If any of the three layers of the tear film are deficient, the eye may suffer symptoms of dry eye.

There are two types of tears: lubricating and reflex. Lubricating tears protect the eyes against the elements. They are produced in a steady flow throughout the day and spread across the eye by the blink reflex. Reflex tears flood the eye suddenly whenever the eye is irritated. They can flow to such a degree that tears roll down the cheek. This excessive watering may occur when the eyes are irritated from smoke, smog, or other irritants like onions, or when the eyes lack proper protection from lubricating tears.

#### *Symptoms of Dry Eye Syndrome*

Symptoms of dry eyes may include burning, stinging, redness of the eyes, and tearing. The tearing seems paradoxical at first but is explained by the fact that an underlying dry eye may become irritated, perhaps sending a signal for increased tear production to flush out the eye. This response is physiologically equivalent to the presence of a foreign body such as a hair or a grain of sand in the eye.

### *Diagnosis and Treatment*

An ophthalmologist who has the patient's history can usually diagnose dry eye syndrome, though the exam confirms the diagnosis. The doctor's exam may show a reduced tear volume and rapid tear break-up time (the time for dry spots to occur on the cornea). A drop of fluorescein dye in the tear film allows the tear film to be better visualized. Some ophthalmologists will test tear production using specially prepared tear-strips (paper strips designed to absorb tears, which appear as a discolored area on the strip).

The severity of dry eye syndrome generally dictates the course of treatment. In most cases, the patient is advised to use artificial tear drops or eye ointment on a regular basis, perhaps four times a day. If the condition is treated with artificial tears chronically, many ophthalmologists will recommend non-preserved artificial tears. Although most of these products can be obtained without prescription, it is highly advisable to consult an eye doctor for advice. Excessive or prolonged use of artificial tears can disrupt the natural production of tears, leading to further aggravation of the condition instead of providing desired relief. A humidifier in the home, especially next to the bed at night, has been found to be particularly helpful for many patients. Because of the hardness of tap water in most areas, however, distilled water is usually required. Hard water will create an airborne mineral dust, which may make the humidifier less effective for its intended purpose

### *Visual Disturbances of PSP*



PSP does not lead to blindness. Any patient with PSP who begins to lose eyesight should see an eye specialist before attributing the loss of vision to PSP. The disorder does affect the control centers in the brain that generate eye movements. Thus, patients progressively lose the ability to move their eyes. During the early stages of the disorder, the eyes still move fully but slowly, particularly in upward and downward directions. Most patients do not experience any eye symptoms at this stage. As time goes on, the range of eye movements decline. Eventually total paralysis of eye movements ensues. In those with PSP, upward and downward gaze is more impaired than side-to-side gaze.

Inability to look down creates several problems. First, reading becomes difficult, and at times impossible, because patients can't look down and use their bifocals. Sometimes people with PSP cannot locate food on their plates. Family members may report that patients unknowingly leave food on their plates. The inability to look down impairs the ability to walk down stairs or to step off curbs, thereby increasing the risk of falls.

Inabilities to look up and sideways are less problematic. While reading, some patients find it hard to shift their gazes to the beginning of the next line after reaching the end of the previous line. Impaired horizontal gaze makes it difficult to scan the surrounding environment, giving patients the impression that their peripheral vision is constricted or that they have tunnel vision. This may lead to a sense of insecurity while walking and driving, activities that patients can still do early in the disease course. When patients express complaints about looking up or sideways, ophthalmologists may prescribe a new pair of glasses or even cataract removal, no matter how mild the cataract might be. Neither of these measures helps. Consulting with a neuro-ophthalmologist or neuro-optometrist can be very helpful, as they are more likely to be knowledgeable about how neurodegenerative diseases affect the eyes.

Another frequent cause of reading difficulty for those with PSP is convergence insufficiency. Besides not being able to look down, the patient cannot aim the eyes in the direction of the nose to focus at near objects and reading material. Because of the combination of failure to converge their eyes and to look down, many patients give up reading.

### *Eyelid Problems in PSP*

Patients with PSP blink less frequently, giving the impression that the patient is staring. Because the blink reflex wipes out tears, the reduced rate of blink can lead to an accumulation of tears. This may lead to transient blurring of vision.

Sometimes patients experience repeated spasms of the eyelids leading to forceful closure of the eyes. This is called blepharospasm. A milder version of this is apraxia of eyelid opening. The patient has preserved the ability to blink, but, from time to time, they cannot open their eyelids after blinking. Often the person is forced to use their fingers to manually lift the eyelids.

### *Treatments*

There are several ways of managing the eye symptoms. Unfortunately, there is no satisfactory treatment of paralysis of eye movements. Eye exercises are futile. Patients who cannot look down and read through their bifocals should hold the reading material higher to eye level so that when they read, the eyes are positioned straight ahead rather than down. Use of a ruler may help in finding the beginning of the next paragraph. Special prism glasses best help patients who have convergence insufficiency. Those who can't look side to side are advised to train themselves to turn their heads while walking and driving.

Blepharospasm and apraxia of lid opening are treated with botulinum toxin injections into the area around the eyelids. The results are good but last only for three to four months. Repeated injections are necessary to keep the condition under control.

Taping the eyelids gently to the forehead is another simple method to keep lids open. "Eyelid crutches" attached to glasses can also help keep the lids open. Finally, in the worst-case scenario, surgery may be necessary to remedy eyelid spasms. The surgeon removes part of the muscle responsible for eyelid closure. In the hands of an experienced plastic surgeon, results are often good, rendering an acceptable aesthetic appearance.

As discussed earlier, dry eye is easily treated by liberal use of lubrication eye drops that are available over the counter.



## Speech, Communication, and Swallowing

### Laura Purcell Verdun, MA, CCC/SLP

### *Overview of the Role of the Speech-Language Pathologist (SLP)*

As with any progressive neurological condition, early intervention is the key to maintaining or increasing communication effectiveness and swallowing function. At the time of diagnosis, or as soon as a patient or carepartner notices changes in speech or swallowing, is the time to seek a referral for a certified and licensed speech-language pathologist (SLP). SLPs are healthcare professionals trained to evaluate and treat individuals with speech, voice, language, and swallowing problems. It is much easier to learn effective strategies and techniques to keep the speech and swallowing mechanisms highly functional than it is to rebuild what is lost. Additionally, strategies can be implemented to compensate for impairment. It is never too late to seek an SLP and get assistance to promote speech and swallowing ability.

## Swallowing

### *Adapting to Swallowing Problems*

Progressive supranuclear palsy (PSP), corticobasal degeneration (CBD), and multiple system atrophy (MSA) are progressive neurodegenerative diseases dominated by motor symptoms. Changes in swallowing and communication are hallmarks of PSP, CBD, and MSA. These changes often occur early in the disease progression, which may help to distinguish them from Parkinson's disease. The intent of this section is to serve as a guide, providing information and strategies to enhance swallowing and communication abilities specific to PSP, CBD, and MSA. Management of swallowing and speech problems requires changing intervention strategies as the disease progresses. Consultations with your physician and speech pathologist are recommended to tailor a program to your specific needs.

PSP, CBD, and MSA are neurodegenerative disorders that develop swallowing difficulties. The primary causes of death are infection and pulmonary complications in the setting of immobility, feeding dependence, and swallowing difficulties. It is important that aggressive efforts are pursued to evaluate and manage these symptoms in an attempt to minimize complications such as malnutrition, dehydration, and aspiration pneumonia. A plan should be developed that will be useful through all stages, including late stages of the disease.

### *Why should I be concerned about swallowing?*



Self-feeding and swallowing problems are very common in PSP, CBD, and MSA. Dysphagia is the medical term for disordered swallowing. Dysphagia can lead to aspiration, meaning food, liquids, saliva, or stomach contents go down the wrong way into the trachea and toward the lungs instead of toward the stomach. This can lead to the development of aspiration pneumonia, an infection that develops in the lungs. Management of swallowing difficulties should promote swallowing safely and easily, along with maintaining adequate intake of fluids and calories.

There are many reasons for self-feeding and swallowing difficulties in PSP, CBD, and MSA. In PSP, there is impairment of self-feeding, including bilateral supranuclear gaze palsy resulting in difficulty looking down. This limits the ability to see the plate during meals. Behavioral changes such as disinhibition can cause rapid drinking or mouth-stuffing. Stiffness can also occur, which interferes with self-feeding. The control and timing of the swallowing mechanism may be affected by an extended head and neck posture, which alters the proportional relations of the mouth and throat. The swallow mechanism may also be affected by cognitive changes or by lack of awareness of deficits. Cough ability may also be impaired across diagnoses.

Slow and incomplete chewing as well as swallowing apraxia are observed in CBD. Apraxia is the inability to perform purposeful movements, so that foods are held in the mouth and not swallowed. Apraxia can also affect the hands and interfere with the ability to manipulate the plate and utensils.

In MSA, there may be difficulty sitting upright at mealtimes and a tendency toward holding foods or liquids in the mouth.

### *What is normal swallowing?*

To manage swallowing difficulties, it is helpful to understand some of the basics of the normal swallowing mechanism. Swallowing is a complex sequence of events that occurs in three phases: the oral, pharyngeal, and esophageal phases. The oral phase involves the placement and manipulation of food or liquid in the mouth and moving it from the front to the back of the mouth. The pharyngeal phase is more automatic and

controls movement of the food and liquid from the back of the mouth through the throat, protecting the airway directed to the esophagus. The esophageal phase involves moving the food or liquid through the esophagus (the food tube) to the stomach. There should be no contamination of the airway or the back of the nose, and nothing should be left behind in the mouth or throat. The phases of swallowing are under voluntary and involuntary control. Therefore, certain aspects of the swallowing mechanism can be modified more than others.

## *How do I know if there is a problem with swallowing?*

There are several warning signs of potential problems with swallowing, including:

- Drooling and difficulty managing secretions
- Food collecting in the mouth
- Apparent increased effort with swallowing
- A "wet" voice quality
- Abnormal posture
- New onset trouble swallowing medications
- Increased length of mealtime
- Low-grade fever
- Chest congestion
- Trouble talking
- Coughing and choking with a red face
- Coughing more during mealtimes than at other times of the day
- Unintentional weight loss
- Need for the Heimlich maneuver
- Need for diet modifications
- Pneumonia



Some patients with PSP may not recognize that they have an eating or swallowing problem, whereas people with CBD and MSA may be more aware. Coughing is a normal response. Silent aspiration occurs when the individual does not cough when food, liquids, or saliva go down the wrong way toward the lungs. If any of these signs occur, you should notify your physician. A referral should be made to a speech pathologist who specializes in swallowing problems, preferably one who is familiar with PSP, CBD, MSA, or related movement disorders.

## *What is aspiration pneumonia?*

Aspiration pneumonia is an infection that forms in the lungs following the aspiration of food, liquid, saliva, or stomach contents. This can happen when a person develops problems with swallowing. Clearly, aspiration in and of itself can place an individual at risk for developing aspiration pneumonia. However, studies have shown that there are other contributing factors that can increase the risk for developing aspiration pneumonia. These include poor oral hygiene, immobility, and dependence on others for feeding assistance. Additionally, the presence of a feeding tube does not eliminate the potential for aspiration pneumonia. In fact, it may potentially increase the risk. The development of aspiration pneumonia is a complicated process with multiple factors.

### *How is aspiration pneumonia treated?*

Treatment for aspiration pneumonia may include:

- Chest physical therapy: Therapists work to encourage deep breathing and coughing through positioning and percussion of the chest and back. The purpose of this therapy is to help loosen the secretions.
- Suctioning: Nurses use a small tube attached to a suction machine, placed in the nose or mouth, and briefly advanced into the lungs. The purpose of suctioning is to remove the secretions in the lungs caused by the pneumonia.
- Antibiotics: Medications used to treat the infection.
- Oxygen: Occasionally, while the infection resolves, people need oxygen administered through a face mask or nasal cannula.

### *What is a swallowing study?*

The swallowing evaluation usually consists of a clinical examination and a swallowing study. The clinical examination includes a discussion with the patient and caregiver, identifying concerns, experiences, and observations related to the potential swallowing problem. It is often beneficial to maintain a diary of the swallowing difficulties observed, including the type of foods or liquids and a description of the surrounding circumstances. This will make the swallowing evaluation more thorough and the recommendations more specific to the needs of the patient. The clinical visit also includes an initial assessment of swallowing skills, including observations of eating and drinking. At this time, suggestions to promote safer and easier swallowing may be provided. These suggestions are based on an understanding of normal swallowing physiology and the changes observed in PSP, CBD, and MSA. The clinical examination is also beneficial to determine the need for an imaging study.



The clinical examination may be followed by an imaging study, either a videofluoroscopic swallowing study (VFSS) or a fiberoptic endoscopic evaluation of swallowing (FEES). A VFSS, also called a modified barium swallowing study (MBS), is a video-recorded X-ray examination of the swallowing mechanism during which foods, liquids, and pills containing some form of barium are presented. Varied consistencies and volumes are presented because some things are easier to swallow than others. A fiberoptic endoscopic evaluation of swallowing (FEES) may be performed in a clinic, but this study is typically of less benefit, as it does not visualize the mouth, only the throat, during the swallow. It can also be limited by compromised posture and movement. A swallowing study is used for multiple purposes: to document current swallowing ability, to rule out non-neurogenic causes of dysphagia, to identify strategies that may enhance the ease and safety of swallowing, to assist in patient education, and perhaps to guide timing of feeding tube placement. The swallowing study should replicate the feeding environment. The decision to proceed with this examination is made on a case-by-case basis. Swallowing status should be frequently monitored by repeating the study as the disease progresses.

### *Will swallowing therapy help?*

Currently, there are few reports in the literature as to the role of direct swallowing exercises in any neurodegenerative diseases, much less in PSP, CBD, and MSA. It is difficult to define the benefits of such efforts given the rapidly progressive nature of these disorders. Emphasis should be placed on patient and caregiver education, diet modifications, and feeding and swallowing strategies. The goals are to avoid nutritional deficiency and dehydration, to reduce the risk of choking and aspiration pneumonia, and to continue oral intake as long as possible for enjoyment and independence.

## *What can be done to improve the mealtime success?*

- Sit upright during meals. For MSA in particular, where antecollis is common, support the patient to sit upright against the chair back.
- Eliminate mealtime distractions such as television or conversation.
- Concentrate on maintaining a slow, steady rate of self-feeding or feeding assistance (PSP and CBD). Make sure food or liquid is swallowed before the next bite or sip.
- Keep the plate of food in the line of vision by placing it on a book or other stable object that will provide sufficient elevation (PSP).
- Use a pair of reading glasses instead of bifocals, which are often ineffective in the setting of impaired downward gaze (PSP).
- Experiment with different plates and utensils. Plates with a rim can be useful for keeping food on the plate. Try using a spoon instead of a fork, or a fork with a modified grip, to make it easier to hold.
- Experiment with different cups. Use of straws is not usually advised because the liquid accelerates into the throat, making it more difficult to control. Flexi-cups aid in drinking without tilting the head back.
- Consider a consultation with an occupational or physical therapist to optimize positioning and self-feeding.

## *Examples of eating and drinking adaptive devices and utensils*

- Flexi-Cut Cup • *www.alimed.com/flexi-cut-cups.html*
- Independence Flo Tumbler • *www.kcup.com/flo.htm*
- Wedge Cup • *www.wedgecup.net*
- Provale Cup • *www.alimed.com/provale-cup.html*
- Maroon Spoons • *www.alimed.com/maroon-spoons.html*
- Scooper Plate with Non-Skid Base • *https://www.southpaw.com/non-skid-scooper-bowls-and-plates.html*
- Skidtrol Non-Skid Bowl • *www.maddak.com/skidtrol-nonskid-bowl-p-27963.html*
- Bruce Medical Supply • *www.brucemedical.com*.

## *What are some strategies to make it easier and safer to swallow?*

- Make sure the mouth and throat are clear of excessive secretions/saliva prior to eating or drinking.
- Maintain the head in a slightly chin-down position while eating, drinking, and taking medications, specifically avoiding extending the head backwards.
- Avoid taking too large a bite, drinking too rapidly, or taking more than one bite at a time.
- Alternate food and liquid swallows to assist with clearance of dry foods or more textured foods.
- Experiment with different food consistencies. Avoid highly textured foods (red meat, raw vegetables, crusty breads, etc.) or dry particulate foods (rice, pretzels, potato chips, crackers, cookies, etc.). Consider changing to soft, moist foods such as casseroles, pasta, canned fruits; or cooked vegetables, fish, and chicken, complemented with gravies, sauces, and condiments.
- Avoid items of mixed consistencies, such as cold cereals, chicken noodle or vegetable soup, and fruit cocktail. These items should be blended into one consistency.
- Thin liquids may be difficult to swallow because they move faster through the mouth and throat. Restrict the amount per swallow or thicken liquids. Information on commercially available thickeners is provided later in this document. Potato or banana flakes, fruit purees, tofu, tapioca, and oatmeal can also be used.

- Carbonated beverages may be easier to swallow than still, clear liquids and may result in less residue in the throat after the swallow.
- Try cold liquids versus room-temperature liquids.
- It may be easier to take medications with a spoonful of a puree, such as pudding or applesauce, instead of with liquids. Do not use gelatin. Do not crush medications unless approved by a physician. Take one medication at a time.
- Consider smaller, more frequent meals.
- Use the less-affected side for self-feeding (CBD).
- Consider that some foods melt at body temperature, such as ice cream and gelatin, and may increase the aspiration risk.
- Consider using yogurt for smoothies to maintain consistency.

*Any other suggestions?*

- It is important to maintain good oral hygiene to minimize the risk of aspiration pneumonia and to enhance ease and enjoyment of oral intake.
- Minimize use of dairy products, which may make secretions thicker, thereby interfering with swallowing.
- A portable suction machine may be of value to assist with clearance of secretions, particularly at mealtimes. Both electric and manual oral suction pumps are available. Your physician can provide a prescription.
- Drinking more water, or sometimes carbonated beverages, may help break up secretions. Your physician may have suggestions for over-the-counter or prescription medications to thin or reduce secretions. Generally, try to avoid decongestants, as these dry up secretions, making them thick and ropey and more of a challenge to manage.
- It is important that the caregiver be educated in the use of the Heimlich maneuver. Ask your physician, nurse, or therapist for instructions.
- Monitor for unintentional weight loss.
- Evaluate prescribed medications because some of them may cause the swallowing problem (or make it worse) or cause dry mouth.
- It is important to be alert to the signs of aspiration pneumonia, including increased chest congestion, chronic low-grade fever, increased cough (particularly with mealtimes), and change in sputum. Does the affected individual cough more at mealtimes than at other times of the day?
- The use of nutritional supplements, such as Ensure, Carnation Instant Breakfast Drink, Boost, Sustacal, Benecalorie, etc., may be beneficial to increase caloric intake quickly and easily. Consider the "plus" version of these products for more concentrated calories and nutrition.
- Supervision during mealtimes is always a good idea.
- Consult with a dietitian to ensure that your meal plans contain all the nutrients that you need.

## Thickeners

Thickening liquids are sometimes used to slow the rate that liquids move through the mouth and throat, improving airway protection. These thickeners are selected because they are readily available or can be obtained through most local pharmacies within 24 hours. These products do not require a prescription. However, you should consult your physician and speech pathologist as to whether these are appropriate. There has been some concern that thickeners, when aspirated, are not well tolerated by the lungs. In addition, some experts believe that thickeners may further reduce liquid intake, as regular thin liquids are generally everyone's preference.

In addition to the thickeners, some of these companies also carry pre-thickened liquids and other foods designed for safer and easier swallowing. SimplyThick is a good choice if you are diabetic.

- Thick-it & Thick-it 2 • *www.thickit.com*
- ThickenUp • *www.nestlehealthscience.us/brands/Resource/home*
- SimplyThick • *www.simplythick.com*
- Thick & Easy • *www.hormelhealthlabs.com/brand/thick-easy/*

## Cookbook Suggestions

Below are some readily available cookbooks with emphasis on swallowing difficulties:

- Achilles E & Levine T. *The Dysphagia Cookbook.* 2003. Cumberland House Publishing.
- Best J. *Down Easy: A Cookbook for Those with Swallowing Difficulties.* 2012. JudyBestCookx.
- Larson J. *Preparing Puree Meals: Comprehensive Guide and Puree Recipe Book for People with Swallowing Difficulty*. 2017. CreateSpace Independent Publishing Platform.
- Mercer SL & Grachuk A. *Safe Swallowing with Dysphagia: A Puree Cookbook for Dysphagia-Related Lifestyles.* 2015. CreateSpace Independent Publishing Platform.
- Weihofen D, Robbins J, Sullivan P. *Easy to Swallow, Easy to Chew Cookbook: Over 150 Tasty and Nutritious Recipes for People Who Have Difficulty Swallowing.* 2002. Houghton Mifflin Harcourt.
- Wilson JR & Piper MA. *I Can't Chew Cookbook: Delicious Soft Diet Recipes for People with Chewing, Swallowing, and Dry-Mouth Disorders.* 2003. Hunter House, Inc.
- Wolff D. *Essential Puree – The A to Z Guidebook with 67 Pureed Recipes for the Dysphagia Diet.* 2016. Gourmet Puree LLC.
- Woodruff S & Gilbert-Henderson L. *Soft Foods for Easier Eating Cookbook: Recipes for People Who Have Chewing and Swallowing Difficulties.* 2007. Square One Publishers.



### *Alternative Nutrition: What do I need to know?*

All or most of nutrition and hydration can be provided in liquid form via a feeding gastrostomy tube. A feeding tube may be indicated if there is evidence of recurrent aspiration pneumonia, greatly increased duration of mealtimes, progressive weight loss, or dehydration—despite efforts to optimize feeding, trouble swallowing coexisting with depressed alertness, or clinical evidence of frequent aspiration or significant silent aspiration. Little is known of the role, timing, and benefits of tube feeding in PSP, CBD, and MSA, particularly in people with advanced disease. Since aging and nutritional deficiencies may severely compromise a person's potential to confront PSP, CBD, and MSA, it is unclear whether early and aggressive nutritional therapy may delay its progression. Feeding tube placement may be indicated if the swallowing impairment far outweighs other aspects of the disease. Placement of a feeding tube does not eliminate the potential for aspiration pneumonia, as gastric contents and saliva can still be aspirated. Additionally, for enjoyment purposes, people with a feeding tube may continue to eat and/or drink.

### *What is a feeding tube?*

A feeding tube is a soft plastic tube that is inserted into the gastrointestinal (GI) tract to provide an alternate route for nutrition, liquids, and medications.

Feeding tubes may be inserted through the nasal passageway for short-term use (nasogastric/NG tube). But for those who require long-term use, the tube is placed directly into the stomach through the abdominal wall. Although usually placed in the stomach, the tube can alternatively be inserted further down in the digestive tract—in the jejunum. The type of tube and location of placement is determined by the physician and other medical considerations. The tube can either be placed surgically with a gastrostomy tube (G-tube)

or jejunostomy tube (J-tube), or endoscopically with a percutaneous endoscopic gastrostomy (PEG) or percutaneous endoscopic jejunostomy (PEJ).

PEGs are the most commonly used type of feeding tube for people with neurological disorders. They are not painful and are not easily visible when wearing normal clothes. When not in use, they are capped and can simply be taped or bound to the belly to prevent moving around under clothing.

### *What do I need to consider as I make a decision about a feeding tube?*

A common decision faced while living with PSP, CBD, or MSA is whether to have a feeding tube placed. This discussion should take place sooner rather than later and be revisited as needed. Prior to determining that a feeding tube is the right course of action, the facts need to be reviewed. Discussions should be initiated prior to a health crisis. The person with PSP, CBD, or MSA and their family should agree in advance with the doctor about what is hoped to be accomplished. Decisions must revolve around the assessment of burdens and benefits. This requires value judgments and consideration of quality of life.

## Speech and Communication Difficulties

### *Adapting to Communication Difficulties*

Problems with communication tend to be present and progressive in all individuals with PSP, CBD, and MSA. Dysarthria is the term used for speech disorders that result when the muscles cannot move with the correct range of movement, strength, speed, force, or coordination, causing slurred imprecise speech, or slowed or rapid speech. Dysphonia is the term used for disorders of voice that affect the sound that comes from the voice box (larynx). Common examples of dysphonia are hoarseness, low volume, strangulated speech, whispery speech, and vocal strain. To address the changes in communication, some comparisons with Parkinson's disease may provide insight.



### *Tips to Enhance Communication*

Most people with communication difficulties prefer to attempt verbal communication for as long as possible, even when their speech becomes hard to understand. The following strategies are designed for the listener and to enhance the communication environment:

- Eliminate distractions and reduce background noise (TV, radio, newspaper, large groups of people, an open door, etc.)
- The listener should face the speaker and be an active listener.
- Keep questions and comments brief.
- Ask one question at a time, allowing time for a response.
- Allow enough time for the person to convey their message.
- Stick with one topic at a time.
- Ask for the topic of the message so that you can use context cues to help with interpretation.
- Ask targeted "yes/no" format questions.
- Pay attention to gestures and facial expressions.
- Ask for clarification when you do not understand, or repeat what you think was said in the form of a question, such as, "Did you say...?"
- Keep to familiar topics.
- Give the person choices to ease decision-making, such as, "Do you want coffee or tea?" rather than, "What do you want to drink?"
- Be patient.

### *Strategies for the Speaker*

- It is often useful to consult with a speech pathologist who will recommend specific exercises and compensations to address limitations in speech and voice.
- Keep sentences short.
- Repeat the entire sentence when necessary, versus an isolated word, providing a context for the listener.
- Take a deep breath before speaking and breathe often.
- Exaggerate and be deliberate with all speech sounds.
- Use gestures.
- Speak one sentence at a time without immediate repetition, remembering the potential for involuntary repetition of speech in PSP (called palilalia).
- Speak loudly and slowly.

Reports of successful speech intervention for individuals with PSP, CBD, and MSA are limited and infrequent. Strategies beneficial to the voice and speech impairments of Parkinson's may have variable benefit. Reasons for limited success include the presence of cognitive impairment, the relatively rapid progression of the disease, delay in an accurate diagnosis, and delay in referral to therapy services.

A speech pathologist can be helpful at all stages of the disease, preferably with early involvement and intervention that emphasize good speaking habits before the underlying problems become so severe that new learning is difficult. It is critical to discuss the limitations and severity of the communication problem, as well as the communication needs. Therapy efforts should focus on increasing loudness, articulatory precision, minimizing repetitions in the setting of palilalia, and enhancing awareness and participation in communication efforts. A therapy program called the Lee Silverman Voice Treatment® (LSVT® • *www.lsvtglobal.com*) is frequently applied to the communication changes in Parkinson's. The emphasis of this program is loud voice productions with intensive training and practice in an attempt to optimize voicing effort and performance. This is appropriate to the Parkinson's communication impairment because the primary problems are low volume, rapid speech, and imprecise articulation. LSVT may not have the same impact in PSP, CBD, or MSA, given that the speech problems are more pervasive and that apraxia is a problematic component in CBD. However, it is reasonable to pursue this strategy, or others, for a trial period of time. A strategy called "communication circles" may be helpful. This is when family members and friends are recruited to try to reinforce target speech behaviors.



### *What is assistive or alternative communication?*

When speech abilities are no longer meeting communication needs, or when speaking is effortful and tiring, communication may be more efficient using alternate means. Handwriting is the most accessible and portable means of alternative communication, such as using a dry erase board. However, writing may be less effective when greater motor impairment is present. Alphabet boards can be used to identify the first letter of each word as it is spoken, or to spell the entire message. Communication books with words or pictures can be used to facilitate conversation. Text-to-speech options with tablets and smartphones are of good quality and easy to access using apps. Text can often be scripted and saved to facilitate conversation and expression of daily needs or appointments with medical providers. Insurance presently will not pay for tablets or apps. Medical-grade augmentative communication systems with more varied means of access are available as well.

Below are some of the commonly used communication apps:

- MyVoice • https://apps.apple.com/us/app/myvoice-tap-or-type-to-talk/id1111359925
- UrVoice • https://apps.apple.com/us/app/urvoice-aac-text-to-speech-with-type-and-talk/id850150951
- Text to Speech – Durbridge • https://apps.apple.com/us/app/text-to-speech/id712104788
- Sono Flex – Tobii Dynavox • https://apps.apple.com/us/app/sono-flex/id463697022
- GoTalk NOW • https://apps.apple.com/us/app/gotalk-now/id454176457
- TouchChat HD • https://apps.apple.com/us/app/touchchat-hd-aac/id398860728
- Proloquo2go • https://apps.apple.com/us/app/proloquo2go/id308368164

These apps also may be found on the Android app store for those who do not use an Apple device.

### *How can I find someone to recommend a communication device?*

The speech pathologist evaluates and trains patients to use communication systems. Ask if they have experience working with augmentative/assistive communication devices and if they can determine which device best meets the needs and abilities of a particular patient.

A team of professionals, including a speech pathologist and perhaps an occupational therapist, a physical therapist, and a rehabilitation therapist usually conducts evaluations. The evaluation includes an assessment of speech, language, visual, and physical-motor abilities. An evaluation will allow patients and families to make informed choices from firsthand experience with a variety of devices. These rehabilitation teams usually work in centers that specialize in assistive technology—in rehabilitation hospitals, university clinics, and nonprofit organizations. These centers often have a variety of different equipment for patients to try. Some sites even allow for a loaner period to further confirm appropriateness. Insurance and private organizations often cover the cost of such devices, called speech-generating devices (SGD), with appropriate supportive documentation.



Resources for augmentative communication can be obtained from the following organizations:

- American Speech-Language-Hearing Association (ASHA) • www.asha.org
- International Society for Augmentative & Alternative Communication (ISAAC) • www.isaac-online.org
- RESNA Technical Assistance Project • www.resna.org
- United States Society of Alternative & Augmentative Communication (USSAAC) • www.ussaac.org

### *What is a voice amplifier, and will that help me?*

While the voice usually becomes abnormally loud in PSP patients, the opposite occurs in some PSP patients and in those with MSA and CBD. A voice amplifier increases the loudness of speech, may minimize the strain and fatigue associated with speaking, and improve the success of communication. A voice amplifier tends to be most beneficial when the voice impairment surpasses the speech impairment, meaning that the voice is weak but the speech is relatively well articulated. When selecting an amplifier, consider the quality of the amplifier, cost effectiveness, portability, and a location to secure the microphone. Headset microphones are ideal for positioning and ease of use. Amplifiers are available for telephone handsets. There are many personal voice amplifier devices available, including these two:

- Chattervox • www.chattervox.com
- Spokeman • www.luminaud.com/spokeman_amp

### *Communicating in an emergency*

There should be a mechanism in place within the home to communicate with one another, such as a walkie-talkie or a bell. It may be wise to consider putting a medical alert system in place in case there is a need for outside medical attention.

# Maintaining Good Oral Hygiene

**Nancy Brittingham, BS, RDH**

With so many PSP symptoms so difficult to treat, this article addresses something that we all CAN DO to manage an important activity of daily living: maintaining good oral hygiene. When this task becomes difficult for the patient, it is time for the caregiver to step in and assist. As a caregiver, our list of tasks is ever-increasing and can seem overwhelming. We have so much to do, and oftentimes, oral hygiene care becomes a low priority and a routine that is easily forgotten. Yet, maintaining good oral hygiene should be a priority because doing so will help the patient maintain oral comfort, prevent bad breath, reduce gum disease and tooth decay, and reduce pneumonia or lung infection caused by oral bacteria.

As PSP and CBD progress, swallowing problems develop as a result of reduced coordination of throat muscles. One result is difficulty forming the watertight seal that separates the pathway to the lungs (the trachea) from the pathway to the stomach (the esophagus). Food "going down the wrong pipe" is called pulmonary aspiration. Frequent episodes of small amounts of liquid and food dripping into the lungs can cause lung inflammation, or aspiration pneumonia. Aspiration pneumonia is a bacterial infection that can be fatal. Good oral hygiene may not prevent aspirating during eating and drinking, but it will minimize additional bacterial growth in the mouth that can be aspirated during coughing or choking spells. Dental care is just as important to one's health and daily routine as is taking medications and getting physical exercise.



## *Tips for Your Dental Visit*

- Make an appointment with your dentist as soon as possible after diagnosis. Take a list of ANY medical conditions and allergies you have and all the medications you're taking, including nonprescription drugs, inhalers, nutritional supplements, and skin patches.
- Educate the dental staff about your disease. Bring printed information. It is important for the dentist and hygienist to understand your symptoms to be able to work most effectively with you.
- Bring all insurance, billing, and legal information. The dentist may need legal consent before treatment begins.
- Although most dental insurance plans cover two cleanings a year, it would be a good investment for those with special needs, such as people with PSP or CBD, to consider three or four per year.
- Be on time for your appointment.
- Ask your dentist about prescribing a fluoride dental paste or rinse to help prevent decay.
- Request a fluoride treatment.
- Ask your dentist to make sure partials or dentures are fitting correctly and that adjustments are made in a timely manner.
- Make sure your X-rays are up to date.
- Make sure the dental office is wheelchair accessible.

## *How to Brush Teeth*

Make sure your teeth are brushed at least twice a day, with the last brushing being after the last meal or snack in the evening.

### *Some ways to make the task easier for someone with difficulty in gripping a toothbrush handle:*

- An electric/power toothbrush will help.
- Another way to build up the handle of the toothbrush is to wrap a washcloth around the handle and secure it with rubber bands. Wrapping rubber bands around the handle before wrapping the wash cloth will reduce slippage of the washcloth.
- Commercially available thick handles into which the toothbrush can be slid.
- Toothbrushes are available with thick handles ergonomically shaped to fit the hand, with the head of the toothbrush angled in a way to allow better contact with the teeth when there is limited elbow and shoulder mobility.

### *Other tips*

- After eating, rinse the mouth with water and use a wet washcloth to sweep through the folds of the cheek. These techniques help to remove food that may be tucked away inside the mouth.
- Use fluoridated toothpaste and also consider using over-the-counter fluoride rinses or a non-alcohol mouthwash.
- If teeth are sensitive, try desensitizing toothpaste.
- If you wear dentures, remove them and use a soft-bristled toothbrush to scrub gums.
- If you wear a partial, remove it so that all areas of the teeth and gums can be cleaned.



### *How to brush and take care of another person's teeth*

- The bathroom does not have to be the only place that you help someone brush their teeth. It may be easier for the person to sit at the kitchen table. Make sure you have good light so you can see inside the person's mouth.
- First, wash your hands, and then put on disposable gloves.
- Sit or stand in a position where you can see all the teeth.
- Have your toothbrush, bowl or basin, rinsing cup, floss, and towel within reach.
- Make sure your manual or electric toothbrush has a small head with SOFT bristles. You may need to use a child's toothbrush.
- Use a pea-sized amount of fluoride toothpaste. If toothpaste bothers someone with swallowing problems, brush with just water instead.
- Replace the toothbrush every three months. Replace it earlier if bristles are worn, and always after a contagious oral or respiratory tract illness. This is something so many dental patients forget to do.
- Be sure to use an apron, towel, or bib draped over a wide area beneath the patient's chin to catch drops of saliva.
- If standing in front of the patient is too difficult, try working from the side and cushioning the patient's head with your other arm, lightly holding their head next to your body. You may find it easier to stand behind the person. See what works best for both of you.
- Make sure the person is comfortable and is seated as upright as possible. Place the toothbrush on the gum line. Brush one or two teeth at a time in small, gentle circles. In order to see well, you can lift the lip or cheeks with another toothbrush or a finger on your other hand. Set up a pattern so that you clean all the surfaces of the teeth—outside, inside, and chewing surfaces.
- Gently brush the inside of the cheeks and the top of the tongue. Help the person rinse with plain water. If they have difficulty, sweep the mouth with gauze.

### *Tools that may help*

- Toothpaste dispenser (pump or hands free)
- Reusable single oral hygiene instrument for patients with difficulty swallowing and expectorating. This is a toothbrush with a hollow handle attached to a suction device to remove fluids during brushing that would otherwise have to be expectorated. 800-325-9044 • *www.trademarkmedical.com/personal-oral.html*
- Mouth swabs, such as Toothettes, Sage Foam Oral Swab, McKessin Oral Swabs with Dentifrice, or Monkcare Disposables Oral Swab Sticks
- Suction bulb
- Portable oral suction device (may assist with the clearance of secretions)
- Water irrigating device or water flosser, such as Waterpik Cordless Profloss, Panasonic Dental Flosser, Sonicare Air Flosser, or WaterPik Water Flosser
- Tongue scraper, such as Gum Dual Action Tongue Cleaner and Scraper or Brosite Tongue Cleaner Scrapers
- Dental Wipes, such as EZGO Deep Cleaning Teeth Wipes, Hisight Fresh Breath Deep Cleaning Teeth Finger Brush, or Lurrose Disposable Finger Toothbrush

### *Flossing*

It is difficult enough to floss one's own teeth. Flossing another person's teeth is even more difficult and can be frustrating. Flossing is a tough job that takes practice. Use whatever type of floss is easiest for you: waxed, unwaxed, plain, or flavored. Wrap 18 inches around your middle fingers. Then, use your thumbs to snap the floss in between the teeth. Try moving the floss up and down, or horizontally back and forth. There are commercially available aids to help you with flossing.



### *Water irrigating*

Water irrigating devices use water under pressure to flush out debris and plaque between and around teeth. This is an excellent tool if the patient can use the device without aid.

### *Denture and partial denture care*

Dentures and partials are very delicate and may break if dropped even a few inches. When handling dentures, always stand over a folded towel or a basin of water. Like natural teeth, dentures must be brushed daily to remove food deposits and plaque. Brushing dentures keeps one's mouth healthy and prevents permanent stains. Use a brush designed for cleaning dentures or a toothbrush with soft bristles. Do not use hard-bristled brushes because they can damage dentures. Some denture wearers use hand soap or mild dishwashing liquid. Do not use other powdered household cleansers, which may be too abrasive. Avoid bleach because it can whiten the pink portion of the denture. When cleaning the denture or partial, the first step is to rinse away loose food particles. Moisten the brush and apply denture cleanser. Brush every surface, scrubbing gently to avoid damage. Next, rinse the denture with water or an antiseptic rinse. Do not allow the denture to dry out because it can lose its shape. Dentures should be taken out at night and placed in a denture cleanser soaking solution or in water.

If not fitting correctly, dentures can cause pain and ulceration of the supportive gum tissue. Be attentive to these complaints. The patient will need to see the dentist for denture adjustments.

### *Tube feeding and dental care*

Oral health care is absolutely necessary for people receiving tube feedings. The same oral health protocol applies to reduce bacteria in the mouth.

### *Additional Tips*

- Avoid smoking.
- Use sugar-free candies.
- Minimize overly salty foods.
- Avoid alcohol, caffeine, and citrus-containing drinks.
- Add club soda or sparkling water to cut through thickened secretions.
- For dry mouth, use sugarless cough drops or over-the-counter dry mouth products.

### *Drooling*

Drooling is a common problem for patients with PSP. Incomplete swallowing causes an accumulation of saliva in the mouth. The salivary pool overflows from the corners of the mouth. This may cause sores and cracking in these areas of skin. Check with your dentist to see if you may need a prescription cream to treat these areas.

### *Conclusion*

Good oral hygiene is very important. Take time out to take care of your well-being. Being able to prevent problems today is well worth it tomorrow. With ingenuity, patience, care, and sometimes humor, oral hygiene will become more routine and less difficult. Your efforts will result in healthier teeth and gums and will minimize risk of aspiration pneumonia.



## How Physical Therapy Can Help

**Heather Cianci, PT, MS, GCS**

*"Physical therapists are movement experts who optimize quality of life through prescribed exercise, hands-on care, and patient education." (apta.org)*

Physical therapy (PT) plays an important role in helping individuals with PSP, CBD, or MSA and their carepartners to cope with mobility changes that occur as the disease progresses. Commonly, individuals are referred to physical therapy because of changes in their walking, balance, and activities of daily living. Ideally, physical therapy should begin immediately following diagnosis, not later when loss of mobility and falls are already happening.

Establishing a relationship with a therapist early on in the disease course can help with the management of physical changes that occur as the disease progresses. Individuals with PSP, CBD, and MSA benefit not only from continual exercise and physical activity but also from the social engagement that PT and exercise offer.

### *What type of tests do physical therapists perform?*

- Muscle strength and range of motion (flexibility)
- Transfers—in and out of chairs, beds, cars, and from the floor
- Gait (walking) and balance
- Coordination
- Posture
- Safety in the home
- Confidence and fear of falling

### *What are the goals of physical therapy in the treatment of PSP, CBD, MSA, and other neurodegenerative diseases?*

- To maintain safe, functional mobility for as long as possible
- To prevent or limit falls and injuries
- To educate and train carepartners to safely assist with functional mobility and home exercise programs
- To prevent or reduce pain, if any
- To make recommendations for home safety modifications

### *What types of treatments do physical therapists provide?*

Physical therapists provide many types of treatments. For example, they:

- Design exercise programs to help patients maintain or improve strength, flexibility, coordination, and balance. Even with disease progression, exercise can help maximize a person's function.
- Treat joint or muscle pain that interferes with activities of daily living. When left untreated, pain associated with muscular tightness and injuries from falls can lead to less mobility and less independence.
- Provide training in correct or new ways to walk to minimize injuries from falls.
- Train in correct or new ways for getting in and out of a car, in and out of a chair at a table, and many other types of movement.
- Teach carepartners proper body mechanics and techniques for assisting with mobility. When individuals with PSP, CBD, and MSA can no longer safely move on their own, carepartners often have to assist with mobility. Learning proper mobility assistance techniques helps keep carepartners free from injuries such as back strains.
- Make referrals to movement and exercise programs in the community. Exercising with a group not only promotes mobility and fitness but also helps with maintaining social interaction and limiting isolation.
- Make recommendations and provide training for walking devices, wheelchairs, and other equipment that can make mobility easier and safer.
- Perform home safety assessments and teach new ways of performing older tasks, such as holding a handrail on the wall when opening and closing a door.

*Examples of Devices*

- Rolling walkers (rollators) for walking
- Transfer aids
- Gait belts
- Bed rails
- Chair and bed risers to make surfaces higher
- Motorized lift chairs
- Specialized wheelchairs

*With Any of the Diseases*

- Maintain a wide and staggered stance while assisting someone with activities of daily living.
- For freezing of gait, do not fight the freeze in an attempt to break free from it.

*At the First Sign of Shuffling or Freezing, Remember the Four S's:*

- Stop
- Stand tall and take deep breaths
- Shift weight side to side
- Step—take a large step



## Physical Therapy Strategies and Recommendations for PSP

- Begin physical therapy sooner rather than later. Early training in the correct way to stand can be helpful, but many individuals will eventually need someone with them at all times for safety.
- Because of problems with eye movements, particularly the downward gaze, individuals with PSP often trip or fall on uneven floor surfaces or objects on the floor. Training in new ways to scan the environment, performing eye movement exercises, and having the correct amount of support and a clutter-free space are crucial.
- The "rocket response" (moving to stand too quickly and unsafely because of impulsivity) can be problematic in individuals with PSP. Simply reminding the individual with PSP to not get up without help may not be enough. A bed or chair alarm to alert others of the patient moving about can be helpful in these situations.
- Individuals with PSP tend to fall frequently, and oftentimes backwards. Weighting the front of rollators and heel wedges in shoes may be helpful in these cases. Ultimately, most individuals will need someone with them at all times when walking, both for verbal cues and for physical assistance.
- Generally, rollators with front swivel wheels work nicely to help improve walking safety. It is very important to have a PT evaluation to determine which device is best. With disease progression, different gait and mobility devices will be needed at different times.
- It is recommended that you try various techniques with the assistance of a therapist to determine which technique works best.

### *Chair Transfer Technique for Patients*

- Slide to the front of the chair.
- Keep feet wide apart and under the knees.
- Lean weight forward and then rise up by pushing on arm rests with your hands (Nose over Toes).

### *Chair Transfer Technique for Carepartners*

- Assist patient with sliding to the front of the chair and properly positioning feet.
- Stand to the side of the individual.
- Place one arm under the individual's arm and your other arm on their back.
- Never stand in front of the individual and pull them forward by the hands.
- Keep your feet wide apart and knees gently bent.
- Gently guide the individual forward and up.

### *Recommended Features of Chairs*

- Avoid low, soft chairs and couches. Choose firm, supportive surfaces.
- Avoid chairs without arm rests.
- Avoid chairs with wheels.

### *Bed Transfer*

- It is recommended that you try various techniques with the assistance of a therapist to find out which technique works best.
- Never pull an individual by the neck or arms; rather, guide partner by placing your hands on their shoulders, trunk, or hips. Take your time, and break the task down into small steps; do not try to do it all at once.
- Generally, helping the individual to roll on their side and then sit up works nicely. Having a handrail to hold improves the safety and ease.

### *Bed and Bedroom Tips*

- Avoid flannel sheets and heavy, bulky blankets. Try lighter, smoother bedding and clothing.
- Make sure the bed is not too high.
- Equip the bed with handrails.
- Try a hospital bed with elevating head and feet options.
- Use nightlights.
- Keep a clear path from the bed to the bathroom, or use a urinal or commode.

### *For Walking*

- Gait training with a focus on preventing small steps, having adequate foot clearance and heel strike, and not moving too quickly.
- Safe turning techniques, such as making U-turns in open spaces, avoiding pivot turns, and avoiding crossing one foot over the other.

### *For Antecollis (A Neck That Has Tilted Forward)*

- Stretching and positioning/bracing
- A tilt wheelchair or recliner
- Botox is not often used because of the possibility of causing further speech and swallow problems

### *For Dolls' Eye Movement (Eyes Moving in Opposite Direction of Head Turn)*

- Stop before turning. Turn head in direction of turn. Work to bring the gaze in direction of turn. Perform turn.
- Early on in the disease, tilting the head down may help with bringing the gaze down. Eventually, this will not be the case.

### *To Prevent Backward Falls*

- Avoid bending or reaching low and standing up quickly.
- To open doors, cabinets, refrigerator, etc., stand sideways in a wide, staggered stance with one hand on a steady surface.



## Physical Therapy Strategies and Recommendations for Corticobasal Degeneration (CBD)

### *Recommendations That Patient and Carepartner Consult with Physician Regarding*

- Dystonia: botulinum toxin, stretching, positioning/bracing
- Aerobic, therapeutic, and balance exercises along with fall prevention training
- Use and exercise of affected limb
- Stretching, positioning, and bracing to prevent contractures
- Gait training based on needs
- Appropriate assistive devices for walking (may not be able to use because of "alien limb," apraxia, or contractures; may need physical assistance or wheelchair instead)
- Safe turning techniques:
  - Make U-turns in open spaces.
  - Avoid pivot turns.
  - Avoid crossing one foot over the other.

## Physical Therapy Strategies and Recommendations for Multiple System Atrophy (MSA)

### *For Bradykinesia*

- Aerobic and flexibility exercises
- Large movements

### *For Orthostatic Hypotension*

- Increase salt in diet (with physician's approval).
- Rise slowly and rest after position changes.
- Wear abdominal compression binder (not pressure stockings).
- Elevate head of bed.
- Eat smaller, more frequent meals.
- Avoid increased time in motionless positions.
- Avoid warmer temperatures.
- Avoid movements that could provoke dizziness.

### *For Parkinsonian-Type Gait*

- Focus on large steps and heel stride.
- Use appropriate assistive devices for walking.
- Practice safe-turning techniques: U-turns in open spaces, avoiding pivot turns, and crossing one foot over the other.
- Make appropriate home modifications.
- Use appropriate adaptive equipment/devices.

### *For Cerebellar-Type Gait*

- Heavier or weighted devices may be helpful

### *Practice Safe Turning Techniques*

- U-turns in open spaces
- Avoiding pivot turns
- Avoid crossing one foot over the other

### Beyond Physical Therapy

*General Exercises*

- Performing regular exercise helps with strength, flexibility, posture, and mobility.
- Exercise also promotes cardiovascular fitness and a general sense of well-being and helps with stress reduction.
- Exercise takes many forms. Playing with the grandchildren, dancing, walking, chair aerobics, yoga, and even gardening (sitting or standing) all qualify as exercise. Aquatic (water) exercise is often recommended because water increases buoyancy and weightlessness. This helps people with mobility problems move with more ease and less fear of falling. The resistance water provides is also good for muscle strengthening. In addition, exercising in warm water can help muscles feel relaxed and less painful. For those who are comfortable in the water, aquatic therapy is a worthwhile form of exercise. Contact your local YMCA, community center, or health club to see what programs they offer.
- The trick to staying with any type of exercise or exercise program is to find one that is enjoyable for you.
- For a more detailed and individualized exercise approach, talk with your physical therapist. Happy exercising!



## Adapting to Functional Changes: How an Occupational Therapist Can Help

**Tony Gentry, PhD, OTR/L, FAOTA**

Though there is no cure for PSP at this time, there are a wide range of adaptive strategies, environmental modifications, and assistive technologies to support continued enjoyment of meaningful activities even as physical abilities change. Occupational therapists (OTs) are rehabilitation professionals who specialize in helping people with disability to manage everyday life. Working with an OT can teach adaptive techniques for performing ordinary tasks in new ways. Restructuring activities can conserve energy. Patients and families can make the home safer and more functional. Lastly, OTs will help patients find and use assistive technologies that will allow them to remain meaningfully engaged in life.

OTs undergo a rigorous education with graduate degrees and national certification to assist the people in managing physical, functional, perceptual, and cognitive changes related to disability. They are trained to operate from a person-centered perspective focused on patients' and families' personal goals rather than on "cookbook" prescriptions. In this way, they build a supportive social and environmental context that allows the patient and family to continue doing the things they care about, despite PSP. The OT may address functional vision, physical skills, cognition, home safety and access, work, leisure, and community participation. They will work collaboratively with the patient and family to build the best mix of supports to help you. Though every person with PSP has a unique situation and requires an individualized approach, what follows is a look at some common problems in PSP and how an OT may be able to help.

## *Functional Vision*

Vision is our most far-reaching sensory ability, involved in nearly everything we do. The visual changes associated with PSP can prove disabling. The visual challenges that people with PSP may face include difficulties in downgaze, blurred or double vision, reduced blink rate (leading to painful, dry eyes), photosensitivity (especially outdoors in bright sunlight), interrupted smooth tracking and steady gaze, and a reduced ability of the eyes to converge while watching objects move closer.

OTs collaborate with other vision professionals to support improved functional vision. Those who have begun to experience vision difficulties will want to meet with each of the vision specialists, since they address different visual challenges. Physicians known as neuro-ophthalmologists specialize in identifying and treating eye problems caused by neurological processes. Neuro-optometrists, also called behavioral optometrists, can provide glasses with adaptive lenses for combatting double vision, photosensitivity, or visual tracking difficulties. These professionals work closely with OTs to address neurologically-based vision problems.

The OT's role in vision care may include recommendations for home modification, task adaptation, and assistive devices intended to help you optimize visual performance during everyday tasks. For instance, it is important to make sure that the lighting at home is bright but without glare, especially in the areas used for reading, cooking, grooming, or working. Clearing clutter from floors, cabinets, and work surfaces allows overtaxed eyes to find things more easily and helps minimize pathway obstacles.

Wherever possible, one should improve visual contrasts in the home. For example, wall switch faceplates and doorknobs may be replaced with contrasting colors to make better visual targets. Florid wallpaper can play havoc with already-challenged eyesight. Flat colors and clearly marked surfaces work better. An OT can visit the home to recommend changes tailor-made for the individual.



For those with downgaze difficulty, a downward-tilted mirror at the bathroom sink or over the kitchen stove may help in seeing items that fall below the visual range. You can purchase mirrored prism glasses (see Resources below) that work in the same way as downward tilted mirrors, allowing one to read as always—by holding a book in the lap. Bookstands and tablet holders can be adjusted to keep reading material at eye level. Magnifying glasses, as well as increased print size and contrast on a tablet or computer screen, can help compensate for blurred vision. Tablets and computers also have the capability to read aloud any onscreen printed material. The OT can help you incorporate these tools and strategies.

Visual difficulties can make community outings a chore. Gray-tinted wrap-around glasses are a good choice for reducing photosensitivity in bright daylight. While walking, use of a cane can help identify surface changes and obstacles on the street. Allowing extra time for any activity in the community is advisable, as is seeking quieter outings at less busy times, so as not to overtax the visual abilities. The OT can teach a systematic head-bobbing technique to allow scanning the visual environment during walking.

## *Fall Safety and Mobility*

If there have been falls or a feeling of weakness in the legs, a talk with an OT can provide ways to make everyday tasks safe. The OT may recommend a physical therapist for ambulatory challenges as well. A tub bench or shower seat may be very useful here. These sturdy, waterproof furniture items are available at medical equipment houses and pharmacies. They allow one to get in and out of the tub safely, sitting down first, then swinging the legs in. Taking a shower sitting down conserves energy while reducing the risk of falling. Adding a shower mat, a grab rail, and a shower hose may help as well. For those who prefer baths to showers, some newer hydraulically-driven bath chairs raise and lower one safely into and out of the tub. The OT can recommend exactly the right combination for the individual's needs and bathroom size.

Getting on and off the toilet (or any chair in the house) can be a cumbersome task and a fall risk. Medicare and other insurance plans cover three-in-one commodes that fit over the toilet. These items also serve as a bedside commode and even as a shower chair. They have armrests and can be raised or lowered to fit one's height. One version comes with a spring-loaded seat that slowly lifts the person to their feet. The same principle works on spring-based or motorized sit-to-stand recliner chairs available from medical supply houses.

Many people with PSP have trouble getting in and out of cars. A good strategy is to back up to the car seat, sit down, then bring the legs into the car one at a time. When getting out, the strategy is to bring the legs out first, then to lean forward to come to a stand. If there is difficulty pivoting on the car seat, a Frisbee-shaped pivot-disk can help. Or a plastic garbage bag on the seat can make sliding or pivoting easier when getting in or out.

### *Mobile and Smart Technologies*

Over the past decade, computers, mobile phones, wrist-worn health trackers, and smart home technologies have become increasingly useful and affordable. For people with visual, physical, or cognitive challenges, these technologies can be real life-changers. Personal computers and tablets incorporate a range of accessibility settings to support people with visual, hearing, dexterity, and cognitive issues. Cell phones come in a wide variety of styles, some with extra-large keys, photo identification screens, or one-touch dialing. Smartphones can be customized with a personalized suite of apps that can track exercise, provide medication reminders (or to let the cat out), read books aloud, serve as a map, or operate entirely by voice command. Wrist-worn devices can supplement a smartphone. For example, some of these devices can detect falls and even notify a caregiver or emergency service of a fall. There are so many tools and apps available these days that finding the ones that will work best for you can be daunting. An OT can help in trying out and choosing the right devices and software accommodations for each person's needs.



Inexpensive smart home tools can automate the home to provide lighting when needed, conserve energy, and reduce the risk of falls. Motion-activated or remote-controlled lights, curtains, fans, and doors can all be linked to a smartphone or can be operated by a voice assistant, such as the Amazon Echo, Google Home, or Apple HomePod. Passive occupant monitors driven by the same interfaces can help offsite caregivers keep an eye on their loved ones without being intrusive. Timed pill dispensers, robotic vacuum cleaners, automatic plant-watering and pet-feeding devices, and dozens of other smart appliances can be incorporated into the home. Choosing the right suite of tools, installing them correctly, and learning to use them can be challenging. The OT can help in exploring these assistive technologies. Though insurance plans often help defray the cost of an OT consult, the tools themselves are rarely covered by insurance. Many states in the U.S., however, have provisions for low-interest loans to pay for them, through assistive technology loan programs.

PSP is an insidious and relentless disease, but there are many ways to face its challenges safely and effectively. Some of the available options have been discussed here. Remember, too, that the OT is a specialist in supporting adaptations for everyday function. As for any other medical professional, however, the patient, family, or referring doctor should ask if the OT is familiar with PSP, how many years of practice in neurological care they have had, and for examples of approaches they have used to help people with PSP. In this way, you can be sure to get the care needed from clinicians with PSP experience.

*Resources*

- American Occupational Therapy Association • www.aota.org • This informative website includes neuro-rehabilitation links and informative articles.
- Centers for Independent Living (CILs): These locally-managed, state-funded centers, run by individuals with disability, are great sources for help with home modification, community mobility, and assistive technology. There's probably one near you. • www.acl.gov/programs/aging-and-disability-networks/centers-independent-living
- Independent Living Aids: This online medical supply store also carries assistive devices for vision difficulties, mobility needs, and daily living. • www.independentliving.com
- Maxi Aids: This online medical supply store specializes in visual assistive devices. • www.maxiaids.com
- Neuro-Optometric Rehabilitation Association: This educational site has information on treating diplopia, dry eye, visual tracking difficulties, etc., with links to neuro-optometrists in your area. • www.noravisionrehab.org
- Performance Health: This online medical supply store has tools for falls prevention, mobility, daily living, and more. • www.performancehealth.com

## PSP and Constipation

### Lawrence I. Golbe, MD



As we age and our metabolism slows, so does the process of elimination. Constipation is a common problem for the elderly, all the more so for people diagnosed with chronic neurological disorders such as progressive supranuclear palsy (PSP), multiple system atrophy (MSA), corticobasal degeneration (CBD), and Parkinson's disease.

*What is constipation?*

Constipation is defined by both frequency and quality of bowel movements. No "normal" bowel movement frequency applies to everyone. If the stool is so hard that its passage is difficult, or if long intervals between bowel movements produce abdominal discomfort, then constipation is present and should be treated. Other symptoms of constipation may include bloating, lethargy, and general abdominal discomfort.

**Scientific Basis**

In parkinsonian disorders, clusters of cells in the spinal cord that control the action of the intestines degenerate. In Parkinson's disease, the nerve cells in the intestines themselves undergo the same sort of degeneration as those in the brain and spinal cord. In such disorders, reduction in bowel movement frequency, or even constipation, may precede—by many years—the limb movement problems. There is even a respectable theory that the cause of Parkinson's disease is a toxin, perhaps ingested, perhaps produced by normal intestinal bacteria, that is absorbed into the tissues of intestines, where it starts to cause constipation and only later reaches the brain.

In normal intestinal functioning, food is primarily liquefied by strong digestive juices in the stomach so that it can travel smoothly through the small intestine, where nutrients are absorbed. Next, rhythmic contractions called peristalsis propel remaining material into the large intestine, where excess fluid is reabsorbed. Parkinsonian disorders can impair the peristalsis of the large intestine, allowing excessive time for its contents to lose their fluid, producing stool that is dry and hard.

### *Fluid is Foremost*

Liquids, like water and juice, add fluid to the colon and soften the stool bulk. Drinking 48–64 ounces (2 quarts) of clear fluids per day is the single most important tool for managing chronic constipation. Beverages containing alcohol and caffeine are actually dehydrating and should be used sparingly. Milk products can also worsen constipation and should not be counted in the daily total of necessary fluids.

If you have problems choking on thin liquids, try using thickening products, which are readily available at your pharmacy. In addition, fluid intake can be enhanced using treats like Jell-O, frozen fruit bars, or fruit smoothies.

### *Fiber is Fundamental*

Low fiber intake aggravates constipation by decreasing the necessary bulk and texture required for stools to form and pass properly. Most Americans eat about 8–10 grams of fiber per day, whereas about three times that amount is optimal to prevent constipation. Dietary fiber is composed of the non-digestible components of grains, fruits, and vegetables.



Try increasing small servings of whole grain pasta or rice, beans, peas, and deeply colored vegetables throughout the day. If swallowing or choking problems occur, these healthful foods can be pureed without altering taste. Red meat and dairy products can actually make constipation worse, so portions of these should be minimized.

### *Role of Exercise*

Inactivity is the enemy of regular elimination. Constipation often worsens after a period of marked immobility, such as a hospitalization or accident that reduces mobility. Regular walking aids regular bowel function. If walking is not possible because of balance or other problems, exercises such as pumping the legs, crawling, or waist-bending can help. Also, twice-daily abdominal massage from the rib cage to the pubic bone can act as a mechanical cue to the bowel.

### *Early Treatment*

If you are getting sufficient fluids and fiber but need additional help to restore regular bowel function, several remedies may be useful. It is important to check with your personal healthcare provider before starting these interventions.

- Weak tea (green or black) or hot water—6 ounces with the juice of ½ lemon on waking each morning helps stimulate bowel action.
- Prune juice cocktail: Mix ½ cup applesauce, ½ cup prune juice, and 2 tablespoons miller's bran. Store in a covered container in the refrigerator. Take one tablespoon per day initially, and gradually increase if needed.
- Over-the-counter stool softeners containing docusate sodium can be very helpful. Choose a product that does not combine a stimulant laxative with the stool softener.
- Fiber products containing methyl cellulose or psyllium are less desirable and should be used only if there is ample fluid intake throughout the day. Otherwise, these products can further dry and harden the stool.

### *Advanced Treatment*

Laxatives are drugs that stimulate the muscular action of the large intestine, as opposed to merely making the stool softer or bulkier, which is what the early treatment measures do. Laxatives tend to lose their effect with repeated use, so they should not be used daily. Many types of enemas contain laxative drugs. The use of any laxatives, even those sold over the counter, should be discussed with one's physician or another healthcare professional.

### *Complications*

Constipation can become more than just bothersome. Untreated, constipation leads to straining to pass stool, causing hemorrhoids and anal fissures (small tears in the skin). Constipation over an extended period can cause stool to compact so tightly in the rectum that the normal pushing action of the rectum cannot expel the mass. This is known as a fecal impaction and can quickly become a medical emergency. Seek medical help immediately if home management for constipation is no longer effective.

## PSP and the Bladder

**Lawrence I. Golbe, MD**

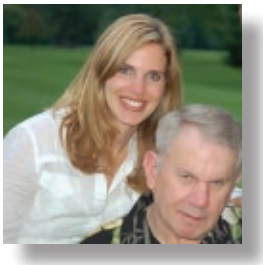
Many brain disorders, including all of the parkinsonian disorders, can affect the urinary bladder. The normal nerve circuitry linking the bladder to the spinal cord perceives the stretching of the bladder by a large quantity of urine. These circuits normally set in motion a reflex, not unlike the knee jerk reflex, that stimulates the muscle in the bladder wall. This squeezes the bladder, causing it to empty. It is the job of the normal brain to inhibit this function. When we voluntarily initiate the act of emptying the bladder, we temporarily suspend this inhibitory brain function. Patients with PSP have difficulty inhibiting reflex bladder emptying. As the bladder slowly fills with urine from the kidneys, this produces sudden urinary urges (urgency) and, in many cases, incontinence, a condition sometimes called irritable bladder. In PSP, degeneration of a small cluster of nerve cells near the lower end of the spinal cord (the nucleus of Onuf) contributes to the problem.

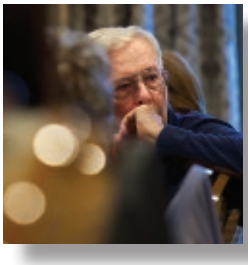
Urinary urgency and incontinence can be treated in a variety of ways. First, it is wise to check for a bladder infection. The inflammation caused by infection can irritate the bladder lining, fooling its sensory nerves into perceiving that they are being stretched. This stimulates the reflex contraction of the bladder wall muscle. Treating the infection with antibiotics and fluids can correct this problem. There are many possible causes of bladder infections, including enlargement of the prostate in men, which can be treated by medication or surgery, and dropped bladder in women, which can be treated with surgery. The overall disability of PSP can cause a bladder infection by making trips to the bathroom difficult and infrequent, allowing urine to accumulate excessively in the bladder, which permits bacteria in the urethra (the tube from the bladder to the outside) to spread into the bladder and grow. Many people with PSP drink little fluids, either because they cannot swallow easily or because they cannot easily walk to get a drink. This reduces the frequency of urination and the natural cleansing of the urethra by regular flows of urine.

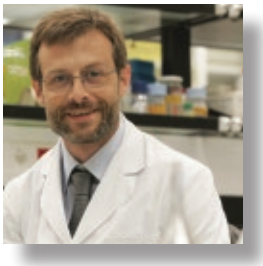
If bladder infection is not the cause of urinary urgency and incontinence, the symptom often responds to drugs that stimulate the muscle at the neck of the bladder. Even when the bladder wall muscle contracts abnormally, the ability of the bladder to empty, and the sensation that it needs to do so, can be inhibited in this way. Two of the more popular drugs are oxybutynin (Ditropan) and tolterodine (Detrol). Both drugs can cause dry mouth and constipation. The latter is often already a problem in PSP and can be treated by fluids, fiber, stool softeners, and, in more severe cases, laxatives. These drugs must not be overused, as they can actually worsen the urinary problem by making it so difficult to empty the bladder that it overfills, weakening the bladder wall muscle.

Some drugs used to treat the movement disorder of PSP, such as amantadine and amitriptyline, can share this side effect of worsening the urinary difficulty. Incontinence during the night is especially common because of the reduced availability of a caregiver to aid the trip to the bathroom and because the recumbent position increases blood flow to the kidneys, producing more urine per hour. Nighttime incontinence can be reduced by avoiding drinking fluids after supper. However, this must not be accomplished at the cost of reducing the total daily fluid intake and causing dehydration. If urinary incontinence in PSP fails to respond to these measures, it may be necessary to use an absorbent pad or adult diaper. The advice of a visiting nurse is often useful in choosing between this method and a urinary catheter. The nurse can also provide instruction in avoiding skin irritation. For men, a condom (Texas) catheter is usually preferable to an indwelling (Foley) catheter because the latter allows bacteria to ascend the urethra, giving the bacteria a surface in the bladder on which to grow and cause infection.





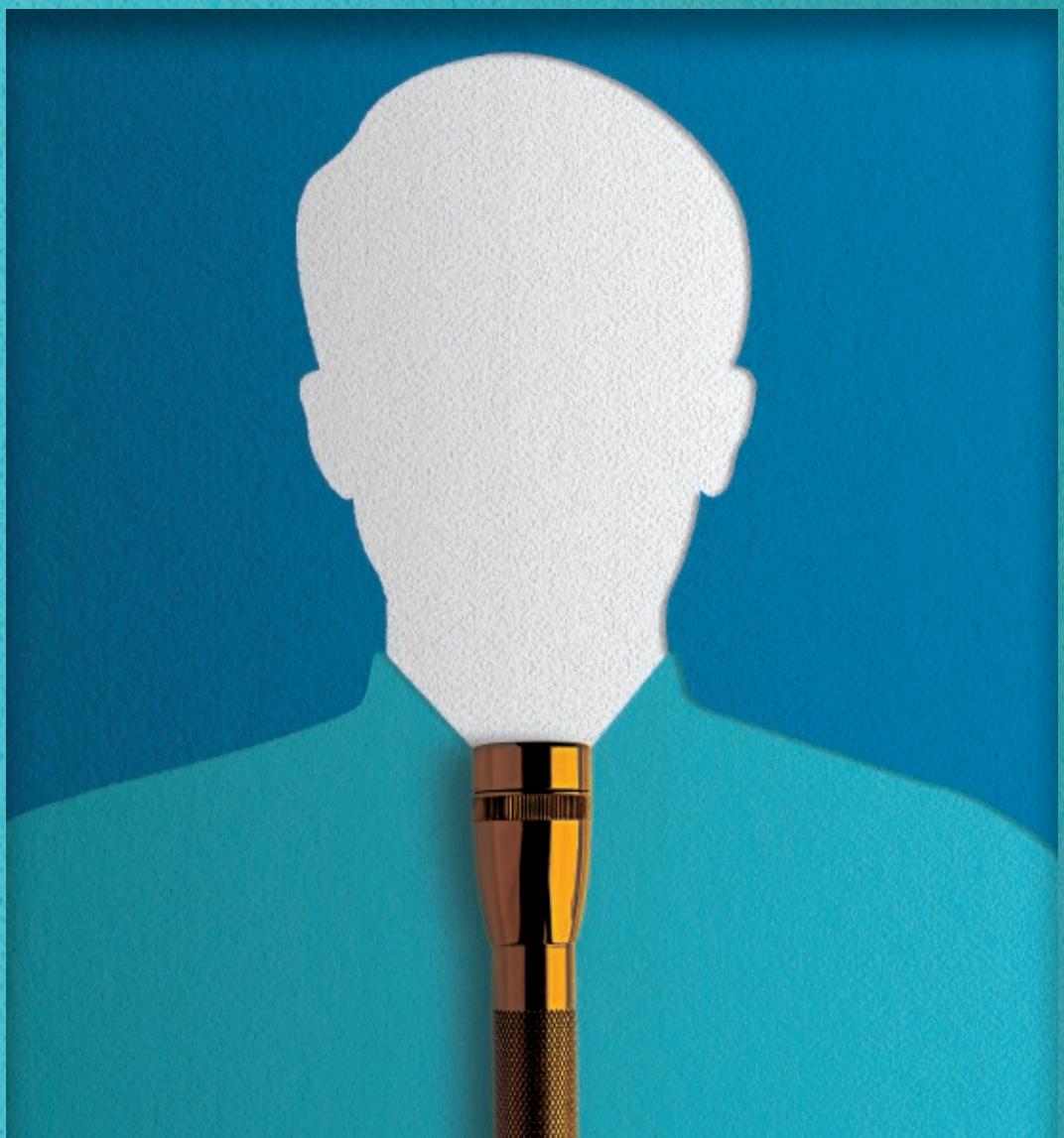




PART THREE

# Managing Care

## and Getting Professional Support



# Building Your Personal Advisory Team

**Diane Breslow, MSW, LCSW**

## *Who doesn't want to feel in control?*

When facing uncertain and unfamiliar territory—illness, trauma, and transitions—most people fare better if they receive information (not too much but not too little either) and if they can develop a plan (albeit a flexible one). I once read a quote from a family member of a loved one with a neurodegenerative disorder: "The worst day of this disease was the day of diagnosis. The best day was the day that we, as a family, understood that we could find ways to handle it. What we needed was a sense of control—and some power."

That sense of control can come from many different realms, people, and perspectives, both professionally and personally. First we will look at the why and the how of building your core healthcare team. Then we will explore other sources of support that can help to fortify and sustain you along your journey.

## *The Rationale for Team Care*

It is an understatement to say that PSP, CBD, and MSA are intricate, complicated diseases. Movement disorders involve so much more than disorders of movement. Besides affecting mobility, these diseases have an impact on the autonomic nervous system—activities of daily living, sleep, mood, emotional well-being, communication, cognition, and social and family relationships. The diseases are chronic, meaning that they continue over a long period of time. In addition, they are progressive, which means that symptoms increase over time. Given this complexity and impact, we can easily understand that a single healthcare discipline cannot address all of an individual's and family's concerns. Rather, these diseases must be treated holistically. Clearly, medical diagnosis and treatment are the foundation upon which you will build your team. After you have established a relationship with a movement disorders neurologist, many other healthcare disciplines, types of interventions, support, and support programs will play a significant part in treatment and management.

## *The Goal of Team Care*

The overarching goal of team care is the delivery over time of the right blend of clinical care, information, education, emotional support, and programs designed to improve quality of life. Ideally, this kind of interconnected or holistic care is provided in a coordinated, seamless manner—across settings, professional disciplines, and time. In many instances, team care requires an individual (carepartner, family member, or care manager) who is designated—and dedicated—as the point person, communicator, and keeper of appointment schedules, team members' contact information, patient information, etc. Last but not least, you want a care team that views patients and families as essential partners in their own care. In other words, you want to participate in the development of a continuous care plan and in ongoing decision-making.

## *The Professional Healthcare Team*

Hopefully this Guidebook gives you insight into the roles of multidisciplinary professionals who will make up your team. You will not be involved with each of these specialists all at the same time. However, at some point in the course of the disease, you will likely encounter them. Therefore, in this book, experts in many of the relevant professions have contributed their knowledge. Those professionals on your team may include:

- Movement disorders neurologist (specialist) and their clinical nurse
- Primary care provider (PCP)
- Physical therapist (PT)
- Speech and language pathologist (SLP)
- Nutritionist/Dietician
- Clinical social worker (MSW)
- Occupational therapist (OT)
- Pharmacist
- Psychiatrist and/or neuropsychologist
- Neuro-ophthalmologist

## *Becoming a Partner in Your Care*

There are steps you and your loved one can take to feel more comfortable as a partner and collaborator on the care team.

- Learn about each team member's role.
- Find out how to access their professional services.
- Acquire their contact information.
- Become educated about the disease and its management.
- Ask questions.
- Encourage family or friends to accompany you to medical and therapy appointments and your support group.
- Establish communication with your team.
- Obtain the name of your particular point person on the team: the person who will either answer your questions or triage them to the team member who can best respond.



## *Your Neurologist Visit*

To make the most of your time with the movement disorders specialist, come to the appointment prepared:

- Bring a list of all current medications, including dosages and times.
- Inform the neurologist of health and family changes, or hospitalizations, since the last visit.
- Write down—and bring—questions and health concerns.
- Ask for explanations of medical terms, medication changes, and recommendations.
- Ask about referrals—such as rehabilitation therapist, social worker, and classes—even if, or especially if, the doctor does not raise the subject.
- Take notes.
- Provide your neurologist with the contact information of your primary care physician.
- Before leaving the appointment, make sure you understand the plan: medication schedule and changes, potential side effects, referrals, and date of return visit.

### *Expanding Your Support Network Beyond Healthcare*

Begin to consider that your team is made up of everyone who has, or could potentially have, a part in helping make your lives as patient or caregiver better, fuller, or easier. This support system may include family members, friends, neighbors, an exercise teacher or trainer (for patient or carepartner), volunteers, clergy, a paid caregiver, and respite care. Your growing network may also consist of various kinds of groups, such as music, dance, or exercise classes; a support group; service or advocacy organizations; and religious affiliations.

As a carepartner, do not be afraid to ask for help. Many people want to help but may not know what you need or even how to approach the subject. Determine which obligations—whether in caregiving or in day-to-day life—you could comfortably relinquish. Then, make a plan to do just that. Write down the names of people who might assist you, and for which activity or task. It may take a while, or even be an ongoing endeavor, for you to coordinate this type of plan. But it will be well worth it.

### *Conclusion*

Remember that your loved one's care and your own self-care are continuous and ongoing. Over time, you will integrate new professionals, friends, and volunteers into your lives. Some people may assume a lesser role; others, a greater role. Your support network is fluid yet a constant in your life.



## The Carepartner Plan: A Guide to Assessing and Prioritizing Your Needs

**Diane Breslow, MSW, LCSW**

It is likely that you, as a family caregiver, provide full-time or close-to-full-time help for your loved one. More than likely, you are involved in assisting your loved one with activities of daily living and medical tasks as well as maintaining a household; shopping and preparing meals; organizing records, documents, and appointments; transporting your loved one to health and rehabilitation; keeping up with social and family relationships; and many other tasks. You are likely providing this care in your home. At the same time, you may be working, raising children or grandchildren, or dealing with your own health or personal issues. Caregivers have an enormous, and often underappreciated, job.

Although the caregiver journey is unique for each individual caregiver, it carries common stresses, concerns, fears, and rewards. Research from the National Alliance for Caregiving shows that the top four caregiver concerns are:

1. Keeping your loved one safe
2. Managing your own stress
3. Finding activities that will engage the person with the illness
4. Carving out time for one's self

Research also reveals that, when caregivers are asked what they want, the majority respond that they want information about coping with being a caregiver. This information takes several forms: knowledge about the disease, comfort with the caregiving role, and dealing with stress.

**Caregiver burden** is defined as: "The strain or load borne by a person who cares for an elderly, chronically ill, or disabled family member or other person... The point where the experience is no longer a viable or healthy option for either the caregiver or the person receiving the care." (Kasuya, RT, Polgar-Bailey P, Takeuchi R. Caregiver burden and burnout: A guide for primary care physicians. *Postgraduate Medicine* 1000; 108(7)119-123).

We also now know for a fact what Dr. Nicholas Christakis from Harvard wrote in the *New England Journal of Medicine*, Feb. 16, 2006: "Certain diseases take an especially heavy toll on the health of the spouse-caregiver. It is the **disablingness** of a condition, not necessarily a terminal nature of the condition, that contributes to caregiver stress." The Carepartner Plan is designed to help you, the carepartner, to understand and manage your own particular caregiving situation so that the role can continue to be—or return to being—a healthy and viable option for you.

**The Carepartner Plan** is based on the premise that caregivers' needs comprise seven broad categories whose order of importance will vary as circumstances and disease symptoms change. Therefore, you should revisit and re-evaluate the seven areas to determine your own particular needs at any given time. The seven categories include:



## *Disease Education*

- Understanding the symptoms and how the diagnosis was made
- Treatments and symptom management
- The impact of the disease on the patient's and family's everyday lives
- Resources and sources of information
- What does it mean to be a carepartner?

## *Time Management*

- Make a daily and weekly list of things to do. Make it manageable and realistic.
- Prioritize—do the most important or difficult things first. Check off what's done.
- Do several errands at one time, rather than going on multiple, time-consuming outings.
- Take a small task with you if you are going someplace where you may have to wait.
- Delegate what can be delegated.
- Forget unnecessary tasks.
- Take a break when pressure gets too great, or as a reward.
- Don't do so much in one area that you cannot be effective in another.
- Break large tasks into smaller, more doable parts.
- Establish and try to maintain routines.
- Recognize that you will have to expend some amount of time and energy on the unexpected and on things beyond your control.

### *Self-Care, Health, and Respite*

- Build in quality time for yourself—1 to 1½ hours a day—and protect it.
- Keep up with your own needs, hobbies, and regular activities. Hold on to your own sense of self.
- Exercise. It leads to better sleep, decreased tension and depression, and increased energy.
- Eat a balanced, nutritious diet. Drink water.
- Recognize when you are stressed.
- Get enough rest.
- Take time to relax.
- Maintain a sense of humor.
- Get regular checkups and keep your own medical appointments. If you provide a lot of physical, hands-on assistance to your loved one, take particular care of your back. Ask a physical therapist for tips on lifting and see if there are assistive products that can help you or the person with the illness.
- Think about your future beyond the caregiving role. What goals do you hope to achieve, and how can you achieve them? Can your care recipient help you achieve them?
- Know your limits, set limits, and bring in outside help (family or paid worker) so that you can take a break.

### *Emotional Support and a Support Team*



- You alone can do it, but you cannot do it alone. Caregiver or carepartner is an identity or role that you grow into.
- Share the care: Emotional support can be derived from obtaining physical and concrete help, as well as emotional help.
- Develop your coping skills, ways to express and release difficult emotions. Give yourself permission to feel sad, frustrated, or irritated—but also to enjoy. Try not to be critical of yourself in moments of anger. Give yourself credit, not guilt. Grieve. Laugh. Love. Hope. Forgive your mistakes. Focus on the present, the needs and rewards of the day. Use positive self-talk, such as "I am doing a good job." Know that it's okay to cry. Write about your experiences and feelings: Research shows that writing helps.
- Develop your emotional and spiritual support network, including your healthcare team (physician, nurse, social worker, rehab professionals), your family, friends, neighbors, support group and individual support group members, clergy, volunteers, and online support forums. Seek comfort from your faith, faith community, and spiritual practices. Find meaning, insight, understanding, and your own inner strengths. Adjust your expectations: Life and you are not perfect. Accept changes as they occur. Get help if necessary. Remember, it is a strength, not a weakness, to ask for help, including emotional help or counseling. Each person experiences depression in a unique way. It is important to take seriously any symptoms you experience that could signal depression. You should not feel embarrassed or ashamed. There are several core symptoms you should be aware of:
    - Sleeplessness
    - Loss of appetite
    - Difficulty concentrating
    - Feelings of hopelessness and worthlessness
    - Feeling slowed down or restless inside
    - No interest in once pleasurable activities
    - Thoughts of death or suicide

If you think you may be depressed, talk to a doctor or mental health professional about your symptoms. Find a supportive professional that you trust and with whom you feel comfortable. In most cases, depression is effectively treated with antidepressant medications, psychotherapy, or a combination of both, plus activities such as regular exercise, spirituality, supportive social interactions, and meditation.

### *Your Relationship with the Person with the Diagnosis*

- Maintain open communication.
- When conversing, remove or turn off loud and distracting noises.
- Express love and appreciation, and also concerns and feelings of frustration. Don't let negative feelings transform into interfering resentments.
- Share special time together apart from caregiving tasks.

### *Medical, Financial, and Care Decisions*

- Define and clarify issues, whether around family participation in caregiving, advance directives, or long-term care options.
- Devise steps for carrying out plans.
- When making decisions about giving hands-on care, ask if your decision promotes your loved one's independence: Do not confuse caring with doing.

### *Community Resources*

- Physical and practical assistance and products
- Caregiving and disease-specific associations
- Educational materials and seminars
- Legal, such as Power of Attorney for Health Care
- Financial, such as disability assistance or medication assistance
- Professionals, such as specialist physicians, nurses, therapists, social workers, and clergy



### *Conclusion*

Once carepartners have identified their concerns, they can then more easily move on to a step-by-step plan.

In summary:

- Using the seven headings described above, rank your needs and concerns in priority order.
- Address your most pressing needs first. View the other categories with less urgency and plan to revisit them at a later date.
- Think about and jot down action steps that you can take.
- Discuss your ideas with people on your healthcare team and with those family and friends who are closest to you.
- Devise a step-by-step plan.
- Implement the steps—with help from others.

## Understanding Advance Healthcare Planning

**Diane Breslow, MSW, LCSW**

CurePSP is committed to providing its constituents—patients, families, caregivers, and healthcare providers—with up-to-date educational information about topics relevant to living with neurodegenerative disease.

It is of utmost importance that patients understand the disease and its potential treatments, decide their wishes for future care—including end-of-life care—and discuss these wishes with their family.

The earlier in disease progression these topics are addressed, the better for everyone—patient, family, and healthcare providers. The benefits of Advance Planning are many-fold—for example: a comfort or freeing-up feeling in having decided one's plans for possible disease complications; diminishing of fears through the course of the disease; and helping the family and the team stay on the same page in their ability to honor and carry out the patient's wishes.

This article will help you recognize possible late-stage treatment options for neurodegenerative disease, as well as understand the various kinds of planning documents.

### *Education about Advance Directives: Making the Case*

A 1997 study in the *Annals of Internal Medicine* (Hoffmann et al.) found that most people do not communicate in advance to their families and healthcare providers their wishes about treatment. Research published in the July 2017 issue of *Health Affairs* demonstrates that statement is still true. In fact, only one-third of adults in the United States have advance directives (ADs). Furthermore, people living with chronic illnesses were only slightly more likely than healthy individuals to express their wishes in written documents.

Research studies consistently report that receiving education about advance directives significantly results in increased completion of such documents. (*JAMA 1994, University of Massachusetts Amherst College of Nursing 2013, General Medicine 2014, Journal of Pain and Symptom Management 2017*, among others).

It is up to each individual to decide if and how they want to think about their medical destiny. Education is key to ensuring that medical-legal issues are discussed, understood, and acted upon.

### *Myths and Facts about Advance Directives*

*Myth: Federal law requires that every person have advance directives.*

**Fact: The Federal Patient Self-Determination Act of 1991 obligates healthcare institutions and professionals to ask patients if they have advance directives and to provide information and education about advance directives.**

*Myth: Most seriously ill patients have discussed cardiopulmonary resuscitation (CPR) with their doctor.*

**Fact: Most patients have not discussed CPR with their physician. Yet, the vast majority want to have this discussion.**

*Myth: An attorney is required to complete one's advance directives.*

**Fact: Legal forms are required, but the services of a lawyer are not. The conversation is best started with one's physician and closest family members or trusted person. An attorney can be helpful in drafting personalized documents.**

*Myth: Some states in the U.S. do not recognize advance directives.*

**Fact: Advance directives are recognized in all 50 states. However, states differ regarding requirements such as number of witness or necessity of a notary. Some states maintain registries that allow quick access by agents and healthcare providers.**

*Myth: Advance directives should be kept in a safe deposit box with other important papers.*

**Fact: A copy of your ADs should be readily available at home. Patients should also give a copy to their physicians and proxy (agent). It is a good idea to make extra copies in the event you need to be admitted to the hospital.**

*Myth: With an advance directive in place, the person gives up control.*

**Fact: Drawing up an advance directive gives you the opportunity to decide, state, and put in writing the end-of-life care you want, thereby helping your agent represent your choices when the time comes.**

## *The Basics*

So, what are advance healthcare directives? They are documents designed to ensure that a person's wishes—to accept or refuse treatments—are met through written directives. Healthcare institutions (hospitals, home health, nursing homes, etc.) are legally obligated to: 1) ask patients if they have advance directives, and 2) provide information and education about advance directives. It is up to each one of us to decide, while we are well and capable, how we will want unexpected medical events to be handled. By exercising our freedom to decide for ourselves our fate or the course of actions, we greatly decrease the chances of an arbitrary exercise of outside authority in determining our futures. Our aim in having ADs is to give us maximum control of our lives and our futures, and to provide peace of mind to our loved ones.



Making healthcare decisions involves trying to understand a lot of information that may be confusing, daunting, and scary. There are multiple issues to consider and a variety of documents to obtain, draw up, sign, and periodically review. The definitions and explanations below present an overview of the documents and will help you decide which ones are pertinent for you. The Resources section below will guide you to more detailed information and next steps.

## *Values History*

A Values History Form is designed to help individuals reflect upon their own personal value system before and while making healthcare decisions. Approaching advance directives from a values perspective enables people to make healthcare choices in line with their values. The intention of a Values History is not only to help an individual understand their own wishes regarding advance care planning, but also to convey one's ethics, views, and principles to family members. In that spirit, values questions include topics such as your overarching attitude or philosophy toward life and health, your goals, what makes your life feel worthwhile, what you fear, what you enjoy, your current health and health challenges, your views about independence-dependence, your role in significant relationships, your spiritual or religious beliefs and how those affect your feelings about illness, your attitudes and relationships with your healthcare providers, and more.

Examples of different Values forms include:

- Values History from the University of New Mexico Center of Health Law and Ethics
- *Ethical Wills: A Modern Jewish Treasury* edited by Jack Riemer and Nathaniel Stampfer (Schocken Books)
- *Ethical Wills & How to Prepare Them* edited by Jack Riemer and Nathaniel Stampfer
- *Ethical Wills: Putting Your Values on Paper* by Barry K. Baines, MD

## *Resuscitation Orders*

In the event that one's heart ceases to beat or is in a life-threatening abnormal rhythm, an individual has the right to choose what, if any, type of resuscitative intervention they would accept.

### Do Not Resuscitate (DNR), also called Do Not Attempt Resuscitation (DNAR) and Allow Natural Death (AND)

Do Not Resuscitate orders are doctor's orders instructing medical personnel, nursing, and hospital staff not to try to return your heart to a normal rhythm by any means: cardiopulmonary resuscitation (CPR), electric shocks (defibrillation), or medications. On the other hand, a doctor may specify in writing the specific means by which you would or would not want to be revived. In addition, Do Not Resuscitate orders mean that you refuse life-support measures.

These orders require the patient's consent and signature, two witnesses, a physician's written order and signature, and inclusion in the patient's medical record. If a hospitalized patient does not want CPR under any circumstances, the physician is obligated to write the DNR order, thereby permitting nursing staff and code teams to withhold CPR. In out-of-hospital situations, such as at one's home or in a dedicated hospice facility, many states require a specific out-of-hospital form to be completed and signed by the patient or patient's proxy and the physician.

### Do Not Intubate (DNI)

Do Not Intubate orders instruct medical personnel of a nursing facility or hospital that the patient does not want to be put on a ventilator, or breathing machine, to prolong their life.



## *Living Will*

A Living Will allows one to put into writing their wishes about future medical treatment should one have a terminal condition (incurable, irreversible, and where death is imminent) and be unable to communicate. The Living Will outlines one's desire to withhold "heroic measures." It usually addresses resuscitation and life support; however, it may cover more preferences and interventions. For example, Living Wills can specify directions about particular death-delaying procedures one wants or does not want, such as artificial life support, transfusions, and dialysis. The key points to remember about Living Wills are: 1) They refer to a terminal condition; 2) They do not designate a patient's spokesperson or proxy.

### *Durable Power of Attorney for Health Care (DPOA-HC)*

The Durable Power of Attorney for Health Care is a signed legal document that designates and authorizes an agent to carry out a person's healthcare and life-support wishes at any time in the future that one loses the ability to speak or make one's own decisions. Every state in the United States has passed a state law recognizing Durable Power of Attorney for Health Care. However, certain regulations may differ from state to state, such as whether a notary is required.

In addition to naming a healthcare agent, the person who is establishing a DPOA-HC also identifies alternate or successor agents who will serve—one at a time, and in listed order—if the original agent can no longer function in the role. A DPOA-HC can include instructions regarding life-support measures, artificial nutrition (tube feeding and what type) and hydration (intravenous fluids), mechanical ventilation (tracheotomy), comfort care, if or when to end life-sustaining measures, CPR, anatomical organ gifts, and disposal of remains. It is crucial that anyone who is completing a DPOA-HC discusses their wishes with their chosen agent and successor(s). If those people indicate that they would not be comfortable expressing your personal wishes, then you need to choose another agent, or successor, as the case may be. If the time comes that your designated agent is unsure of your wishes in a specific situation, that agent is required to speak in your "best interest."

Remember to periodically review, update (if necessary), re-date, and initial the original document.

DPOA-HCs can be ended any time by informing your physician and agent of your decision and destroying the document, or by drafting, signing, and dating a new one.



### Practitioner Orders for Life-Sustaining Treatment (POLST) or Medical Orders for Life-Sustaining Treatment (MOLST)

POLSTs are signed practitioner's orders. They act as standing medical orders and apply to all healthcare personnel (EMTs, emergency room personnel, etc.) and in all settings (homes, assisted living facilities, nursing homes, etc.). The type of practitioners who can sign the POLST varies by state, and may include a physician, advance practice registered nurse, nurse practitioner, and physician assistant. POLSTs provide guidance about one's care near the end of life, allowing healthcare personnel to act immediately—and in line with your wishes—in an emergency. POLSTs spell out what specific care should be administered or withheld *at the present moment in time* for a specific patient, as directed by a physician. The POLST Paradigm is in effect in most states of the United States.

Because these are signed medical orders, they are portable in states where POLST programs have been legislatively determined. This means that all healthcare facilities and emergency service providers in the region have agreed to recognize and abide by the orders, regardless of where the document was originally signed. POLST is particularly relevant to patients expected to transition from one healthcare setting to another.

The form is completed after a careful discussion between the doctor and all parties involved. The form contains three sections: cardiopulmonary resuscitation, medical interventions, and artificially administered nutrition. The patient can choose no nutrition by tube, defined trial period of tube nutrition, or long-term artificial nutrition by tube.

For more information, please see the POLST website: *www.polst.org*.

### *Mental Health Treatment Preference*

This type of directive asks you to state whether you would allow electroconvulsive treatment (ECT) or psychotropic medicine if you have a mental illness and are unable, at the time, to make these decisions for yourself. In addition, you can express whether you will accept admission to a mental health facility for up to a certain number of days (for example, 17 days). You can name someone to speak for you. These orders are written and have an expiration (such as, three years from signature date). They require witnesses. Mental health treatment directives can be canceled in writing as long as you are not receiving mental health treatment at the time of cancellation.

It is important to note that not all states have a mental health treatment advance directive, and requirements may vary by state.

## *Guardianship*

A guardian is a legally responsible individual who is court-appointed—on a limited-time or full-time basis—when an individual is not able to make their own decisions and there are no advance directives and designated healthcare proxy, or no next of kin (commonly referred to as surrogates). Other instances that might necessitate a court-appointed guardian include families in which multiple first-degree relatives cannot agree—even with mediation, or where a next of kin is clearly acting in their own self-interest and not that of the patient. Fortunately, formal guardianship is rarely required.



## *Points Pertinent to PSP and Related Diseases*

Patients and family members often put off what they view as the morbid discussion of whether artificial nutrition and hydration (ANH, or tube feeding) should be used if swallowing problems or advanced dementia keep the patient from eating and drinking normally. Without the discussion, however, a crisis situation can develop. If the person with PSP is unable to make their wishes known, then it is left to the Durable Power of Attorney for Health Care to decide whether or not to place a feeding tube.

If the person with PSP is competent and able to make care decisions, then the question of whether to accept ANH can be made based on individual circumstances and beliefs. But what if the patient is no longer able to make personal decisions and has not previously completed a Durable Power of Attorney for Health Care? The person acting as the health decision surrogate (spouse, partner, adult child, sibling, close friend) may struggle with the ANH decision and not have much time to think about it. The following information may be of help.

If ANH is being suggested because the patient is at risk for aspirating or has had a bout of aspiration pneumonia, it is important to know that the feeding tube will not prevent bacteria-laden saliva and nasal secretions from getting into the lungs and causing infection. In other words, patients could still die of pneumonia.

Placing a feeding tube in a severely demented person can result in the patient trying to pull the tube out and needing to be physically restrained. This can worsen any agitation the person might have and may lead to the use of sedative medications. In the Cochrane review done in London, doctors searched for evidence that tube feeding improved the quality of life for people with advanced dementia—but could not find any. They, in fact, found some evidence that tube feeding increased mortality and morbidity, and reduced quality of life.

Decision makers also worry that by not choosing ANH, they are dooming the patient to a long and painful death. Reports from conscious dying patients indicate that thirst and appetite decrease naturally at the end of life. Conscious elderly patients slip quickly into a coma that is free of pain; observation of unconscious patients indicates that their dying process is quite peaceful. If the body is shutting down in preparation for death, artificial hydration—the process of giving intravenous fluids—can actually cause distress. The body is unable to rid itself of excess fluids at this time, and they can build up in the lungs, making it hard to breath.

In an article that appeared in the *Journal of General Internal Medicine*, the authors point out that we seem to have lost sight of the difference between a person who dies because they stop eating and drinking, and one who stops eating and drinking because of the natural dying process. Hopefully in the future, we will have clinical studies that define more clearly when the use of ANH in advanced PSP adds quality as well as quantity to a patient's life and when ANH inhibits the natural process of dying.

## *Summary*

With this basic information in hand, we hope you will discuss and draw up your wishes in conversation with your family, your physician, and an experienced social worker or nurse. Doing so will provide you with control over decisions about your care even if you are unable to communicate in the future and will provide your family or agent with valuable information about your care wishes.

## *Resources*

- American Bar Association • 800-285-2221 • *www.americanbar.org*
- Compassion and Choices • 800-247-7421 • *www.compassionandchoices.org*
- Family Caregiver Alliance • 800-445-8106 • *info@caregiver.org* • *www.caregiver.org*
- National Academy of Elder Law Attorneys • 520-881-4005 • *www.naela.org*
- National Hospice and Palliative Care Organization Caring Info • 800-658-8898 • *www.caringinfo.org*
- National Institute on Aging Advance Care Planning • *www.nia.nih.gov*
- National POLST Paradigm • 202-780-8352 • *info@polst.org* • *www.polst.org*
- State medical societies in the U.S. by state • *www.emedevents.com* • *www.thedoctors.com*
- Values History from University of New Mexico Institute for Ethics • 505-272-4566 • *hsc-ethics@salud.unm.edu* • *https://hscethics.unm.edu/directives/values.html*

## Compassionate Allowances

Source: Social Security Administration website

Compassionate Allowances (CAL) are a way to quickly identify diseases and other medical conditions that, by definition, meet Social Security's standards for disability benefits. PSP, CBD, MSA, and other brain disorders are on the list of conditions allowable for Compassionate Allowances. The CAL program helps reduce waiting time for a disability determination for individuals.

The Compassionate Allowances program identifies claims where the applicant's disease or condition clearly meets Social Security's statutory standard for disability. By incorporating cutting-edge technology, the agency can easily identify potential Compassionate Allowances. The Social Security Administration (SSA) uses the same rules to evaluate CAL conditions when evaluating both Social Security Disability Insurance (SSDI) or Supplemental Security Income (SSI) programs.

SSA receives information from the public, comments from the Social Security and Disability Determination Service communities, counsel from medical and scientific experts, research with the National Institutes of Health (NIH), and information from past public outreach hearings regarding potential CAL conditions. For more information on how to submit a potential CAL condition to the Social Security Administration, go to their website at *www.ssa.gov/compassionateallowances* or call 800-772-1213.



## The Social Worker's Role

### Diane Breslow, MSW, LCSW

Living with a chronic, progressive illness like PSP, MSA, or CBD can pose many different and changing challenges over a long period of time. Each of the diseases is "family disease." While one person may have the diagnosis, the whole family experiences the effects. Social workers play a key role in helping people with these diseases and their families to deal with those challenges and to navigate the healthcare system, community services, and the many feelings and situations encountered.

Because symptoms and abilities change over the years, many families find that what worked or helped at one point may not be the best option at another time. Whether you are seeking advice in managing the impact of disease progression or need help finding resources (such as a support group or new housing possibilities), talking with a social worker can be an important step in helping explore new options and creative ways of living well.

#### *Why Social Work?*

Social workers offer an important skill set and mindset. Social workers subscribe to the biopsychosocial model of assessment and care. That is, they are trained in the theory that individuals and families are a composite of complex physical (biological), emotional (psychological), family, and social/cultural aspects, all of which combine to play a role in behavior and relationships. Because the problems of neurodegenerative disease impact people in so many ways, disease management requires a comprehensive team approach: one that includes social workers and other allied healthcare professionals (physical therapist, speech

therapist, occupational therapist) who can intervene on all levels. Some social workers, as well as rehabilitation professionals, specialize in these diseases and their unique challenges.

Ideally, social workers can provide the best care in an ongoing relationship with clients in which trust can be built over time. Regular visits afford the social worker the opportunity to understand the client's perspective, strengths, and unique challenges—and to work with clients to develop a mutually agreed upon treatment plan. Sometimes clients meet with a social worker only once—for a specific issue, for information about a service, or at a point of transition or crisis. Some patients and their families also talk with social workers at periodic intervals, as specific needs emerge during the course of the disease.

Social workers aim to understand each person and family's unique situation and goals. Therefore, social work intervention is not a one-size-fits-all approach. Every case is viewed and treated individually. What one couple or family finds helpful may be very different for another couple. For example, one couple wants help to remain in their own home, while another couple wants to understand assisted living options and locating an appropriate residential community. Furthermore, it is fairly common for one person in a couple to see a situation differently from the other, or for adult children to hold different views from one another. A woman with PSP may believe she can still travel the world, while the husband/carepartner feels quite reluctant about this. Or one adult child views Dad as "not changed at all," while the other adult child treats Dad as helpless. These kinds of different perspectives can create new conflict or cause old conflicts to re-emerge. Social workers are trained, experienced, and successful at dealing with these kinds of issues.

### *Reasons to See a Clinical Social Worker*

- To better understand the disease and to discuss reactions to receiving the diagnosis
- To talk about when and how to share the diagnosis with family members and friends, or at the workplace
- To find new ways to cope with feelings such as sadness, depression, anger, fear, or frustration
- To develop new attitudes and habits of flexibility, persistence, and adaptation, with a focus not on cure but rather on living the best life possible and creating new meaning
- To plan for the future, including advance directives, insurance issues, home care, and housing options
- To express and deal with losses—physical, emotional, and social
- To adapt to changes in family roles and dynamics, such as taking on the role of carepartner
- To find ways to build or maintain good communication with a carepartner and other family members
- To deal with different kinds of reactions and myths or misinformation of family, friends, and the general public
- To learn about and access resources: community services, programs, classes, and support groups

### *Resources a Social Worker Might Suggest*

- Associations like CurePSP
- Home safety evaluation
- Workplace accommodation
- Driver safety evaluation
- Social Security Disability
- Legal assistance
- Exercise classes and videos

- Transportation
- Support groups
- Caregiver support
- In-home help
- Respite
- Palliative or hospice evaluation

## *The Social Worker's Clinical Roles*

- Individual and family counseling, both short- and long-term
- Facilitating support groups for diagnosed individuals as well as caregivers and loved ones
- Providing resources and disease education to patients, families, and other healthcare workers involved in the patient's care
- Linking patients and families with appropriate community services
- Care management or case coordination with patient's other healthcare providers
- Peer-to-peer (one-to-one) pairings of patients with other patients, or caregivers with other caregivers
- Telephone consultation to patients and families
- Advocate for multidisciplinary interventions, including physical therapy, speech and swallow therapy, and occupational therapy



## *Facts about Social Work*

- Social work degrees are awarded at the bachelor's, master's, and doctoral levels.
  Most licensed social workers have a Master's in Social Work (MSW), a state requirement.
- All 50 states in the U.S. license social workers. However, many people without licenses call themselves social workers. The best way to get clarity is to ask the professional you are seeing if they are a licensed social worker. "Licensed social worker" is a regulated title; "social worker" is not.
- LMSW, LCSW, LGSW, and LSW are typical initials you might see after a licensed social worker's name. The meaning of these titles varies among states.
- The National Association of Social Workers (NASW) is the main professional social work governing body.
- In many settings, licensed clinical social work services are reimbursable.
- Some healthcare settings where you can find social workers:
  - Medical institutions and rehabilitation centers
  - Adult day programs
  - Home health agencies
  - Assisted living and skilled nursing facilities
  - Veteran's Administration (VA) centers
  - Palliative and hospice care
  - Private practice

# Understanding Levels of Home- and Community-Based Services and Residential Care

**Diane Breslow, MSW, LCSW**

CurePSP is committed to providing its constituents—patients, families, caregivers, and healthcare providers—with up-to-date educational information about topics relevant to living with PSP, CBD, MSA, and other neurodegenerative diseases.

Among the ever-increasing population of older and disabled adults and their caregivers and family members, confusion and misunderstanding abound with regards to the broad spectrum of available choices for home- and community-based services, residential facilities, long-term care, and rehabilitation. Part of the reason for this confusion is that, throughout the 50 states, a lack of consistency exists in terminology, regulations, licensing, on-site services, costs (up-front and add-ons), amenities, and more.

This article will help you understand the spectrum of services and care options for adults with physical impairments or chronic disease. You will become aware of questions to ask when searching for a type of care or care facility.

Each of the services and care levels discussed are potentially appropriate for people with PSP and related diseases. That said, it is most important and acceptable for family caregivers to inquire as to a staff's experience with prime of life neurodegenerative disorders like PSP. Family members can and should make providers aware of the symptoms and needs of these patients, and offer to ascertain educational materials, speakers, and in-service training for staffs.



## *Adult Day Services (also called Older Adult Day Programs, or Adult Day Care Centers)*

Adult day services are community-based, daytime group programs for cognitively or functionally limited adults who require supervision, socialization, structure, and some personal assistance. The programs are provided in a safe and secure setting, sometimes in stand-alone buildings and sometimes within hospital campuses or continuing care retirement communities (CCRCs).

Adult day centers feature exercise, socialization, and recreational activities.

*Other possible components are:*

- Transportation
- Health-related services, such as medication reminders, incontinence care, nurse oversight
- Meal and snack
- Social work services, such as counseling, information, referrals, and care coordination

Features and fees vary from program to program, and state to state. It is important to ask specific questions about what is and is not included in any one center.

Note: In addition to the benefits to participants, adult day programs afford family caregivers much-needed respite from the demands of caregiving full-time for someone who needs constant supervision.

### *Medical/Rehabilitation Services: Physical, Occupational, and Speech Therapies*

Even with a neurodegenerative disorder such as PSP, rehabilitation therapies can offer helpful safety instructions and can help re-stabilize an individual's functioning. The skilled rehabilitation therapies of physical therapy, occupational therapy, and speech therapy are provided in multiple different settings:

**Inpatient rehabilitation** takes place in either a dedicated rehabilitation center or institute, or in a rehabilitation unit of long-term care facilities or hospitals. To qualify for admission to an inpatient rehab center, one must meet specific criteria related to one's ability to participate in and benefit from daily, intensive, multiple therapy sessions.

**Day rehabilitation treatment centers** offer community-based treatment in an outpatient setting or clinic. Some day rehab programs are housed within a larger rehabilitation institute. The patient must meet specific criteria related to ability to undertake several hours of skilled therapy throughout the day.

**Outpatient centers** provide therapy in a community clinic, hospital, or agency. Therefore, as with day rehabilitation, the person must be able to leave their home for therapy. Unlike day rehabilitation, outpatient therapy consists of a one-hour session at a time of physical, speech, or occupational therapy.

**Home health care:** If a patient is deemed to be home-bound, then skilled care or rehabilitation therapies along with visits from a registered nurse, can take place in one's home. During the time that a case is open for home rehab, the patient is also eligible to receive a bath aide. However, once the course of rehabilitation ends, so too does the bath service.

Medicare or insurance covers skilled rehabilitation therapies, subject to initial evaluation and periodic re-evaluations.



### *Non-Medical or Personal In-Home Care*

Non-medical or personal home care refers to personal care with activities of daily living, such as bathing, grooming, and dressing. In-home care providers are also called companions, personal aides, home health aides, homemakers, or personal caregivers. These kinds of home-care providers work for themselves (privately) or are employees of an agency that takes responsibility for setting fees, making caregiving assignments, insuring and bonding the caregivers, and training them.

Home health agencies offer caregiving services in any of three ways:

1. By task, such as bathing assistance
2. By blocks of time or shifts, such as 4 hours or 8 hours
3. Live-in

For the most part, personal care is a private expense. Medicare or health insurance does not cover personal care. However, it may be covered by one's long-term care insurance policy. In addition, some states, through their local Area Agencies on Aging, offer subsidized, and usually "capped," personal care services for individuals with limited financial means.

### *Independent Living: Senior Apartments, Active Senior Communities, and Retirement Homes*

Independent living is a broad term that encompasses senior apartments, active senior communities, and retirement homes. These types of buildings are age-restricted (for example, 55 and over, or 65 and over) and are not licensed to provide personal care or nursing services. Basic services include a certain number of meals (per day, week, or month, depending on the particular facility), housekeeping or laundry (this could

mean laundry or only linens, or also personal clothing), social activities, well-being checks, and transportation to doctors and outings. For an additional cost, personal care assistance can be purchased, either on-site or from an outside home care company (see above). Some independent living buildings are federally or state subsidized; however, most are private-pay. Families need to ask detailed, specific questions about what features and services are and are not included in the cost of any one independent living facility. Costs and inclusions can vary greatly.

### *Assisted Living*

Assisted living refers to an entire building, a specified unit of a building, or a part of a continuing care retirement community (CCRC) that is licensed to provide 24-hour-a-day personal care for activities of daily living, in addition to offering the basic services of independent living (see above): meals, housekeeping, and social programs. However, the assisted living license—and its definition of which care services or how many—can differ from state-to-state. *Assisted living is the fastest-growing type of residential care with the most discrepancy in definition.* Again, it is crucial for prospective residents to ask very specific questions as to the type of care—and caregivers—offered in a particular building. In general, assisted living caregivers are trained, certified aides who assist with daily tasks—for example: bathing, dressing, escort to meals, medication setup and dispensing, and routine checks on residents. The role of a registered nurse (RN) may vary from facility to facility. At the least, the RN should be the one who sets up and stores a person's medications. The aide may be the person who delivers the medicines to the resident. Even the definition of the term "delivers" regarding medication may vary. "Deliver" could mean: to drop off the medication in the person's room; to pour it into the hand of the resident, who then takes the medicine; or to administer the medicine to the person. Assisted living facilities may also supply transportation to and from medical appointments, errands, and group outings. Some facilities may offer rehabilitation therapies, hospice care, and specialized care for different disorders (such as dementia). If the facility offers rehab or hospice, the same regulations for physician referral, evaluation, and insurance or Medicare coverage apply as when someone is living in their own home. In addition, residents are free to choose their own rehabilitation provider, even if the facility houses its own or another company on-site.



### *Supportive Living*

Supportive living refers to state-funded facilities—in some but not all states—for seniors who need assistance with daily living, as in assisted living (above). Individuals need to check with their own state's Area Agency on Aging or a social worker familiar with community resources.

### *Skilled Nursing Facilities (SNFs) or Nursing Homes*

Skilled nursing facilities are institutions that are licensed and regulated by state and federal governments to provide room and board, daily personal care, supervision, medical care and oversight, and 24-hour-a-day care from registered nurses (RNs) and certified nursing assistants (CNAs). Physicians (MDs) serve as medical directors of skilled nursing facilities. Many, if not most, nursing homes are certified for Medicare and/or Medicaid coverage for a certain number of beds (residents) per facility. Medicare certification allows for short-term rehabilitation stays only. Medicare and supplemental insurance policies usually cover 100 days of nursing home care that results from a hospitalization and meets the criteria for rehabilitation therapies. For those people who are not on Medicaid or the short-term allowance of Medicare, skilled care is a private expense and/or may be covered by a long-term care insurance policy.

Skilled nursing facilities are composed of various types of care:

- **Custodial or Basic Care:** Assistance with personal care (activities of daily living such as bathing, dressing, or toileting), ambulation, safety, supervision, and medication dispensing
- **Skilled Care:** Treatments or procedures, such as wound care; intravenous medications or feedings; or managing of machinery, such as respirator or ventilator
- **Sub-Acute Care:** Short-term rehabilitation stay (discussed above) to provide rehabilitation from an acute illness, injury, or exacerbation of a disease process
- **Respite Care:** Short-term or trial stays in the facility; or "vacation stays" while the family caregiver takes a break. Respite is a private-pay service. Additionally, some states and private or non-profit foundations may offer respite funds.

### *Continuing Care Retirement Communities (CCRCs)*

Continuing care retirement communities (CCRCs) are residential campuses comprised of a number of buildings, offering a range of care options, all on the same grounds. Within such a community there might be independent living, assisted living, and skilled nursing. However, not all CCRCs provide all levels of care. Most CCRCs require an upfront investment of some amount as well as monthly payments. Specifics differ from community to community, so it is important to ask questions and do comparison shopping.

### *Special Care Units (SCUs) and Memory Care Units (MCUs)*

Some states license, and thereby recognize the designation of, special units that exist within facilities that are already licensed to give care. Examples of specialty licenses are memory care assisted living and memory care skilled nursing. If facilities advertise as memory care and are located in states that assign such licenses, then the facility must have that license.



### *Patient Medical Information*

No matter where your loved ones live or attend programs such as those described here, they should always keep with them, and in a specified location in their living space, written medical information in the event of an emergency. This document is often referred to as a "vial of life." It should contain the following information:

- Date updated
- Name, address, phone number
- Medications, dosages, frequency or times
- Drug allergies
- Medical conditions
- Surgeries (including year)
- Blood type
- Allergies
- Inoculations
- Physicians (primary care, movement disorders neurologist, and any other key doctors) and their phone numbers
- Durable Power of Attorney for Health Care
- Emergency contacts

Because information can change, you should regularly review and update the document. In case of emergency, carepartners should keep a copy with them and place another copy in the home—in a visible spot, such as taped to the refrigerator.

## *Resources*

- AARP • *www.aarp.org*
- Administration for Community Living • *www.acl.gov*
- Aging Life Care Association • *www.aginglifecare.org*
- Eldercare Locator • 800-677-1116 • *www.eldercare.acl.gov*
- Medicare • 800-MEDICARE • *www.medicare.gov*
- Medicare Nursing Home Compare • *www.medicare.gov/nursinghomecompare/*
- Medicare & You Official Government Handbook • *www.medicare.gov/medicare-and-you*, or obtain your print copy by calling 800-MEDICARE
- National Center for Assisted Living (NCAL) of the American Health Care Association • 202-842-4444 • *www.ahcancal.org*
- National Institute on Aging • *www.nia.nih.gov;* Long-Term Care • *www.nia.nih.gov/health/caregiving/long-term-care*
- Social Security Administration • 800-772-1213 • *www.ssa.gov*
- Veteran's Administration Helpline • 800-827-1000 • *www.va.gov*
- Vial of Life: This nonprofit provides materials for allowing first responders access to relevant medical data in your home • *www.vialoflife.com*

## *Caregiver Associations*

- Family Caregiver Alliance • *www.caregiver.org*
- National Alliance for Caregiving • 301-718-8444 • *www.caregiving.org*
- National Family Caregivers Association • 800-896-3650 • *www.nfcacares.org*

## CurePSP Support Groups

**Wendy M. Resnick, RN, MS, CS**

### *What is a CurePSP support group?*

A CurePSP support group is a voluntary gathering of people who share common experiences, situations, or problems related to living with a prime of life disease such as PSP, CBD, or MSA. Group members offer each other emotional and practical support and reduce the sense of isolation that is associated with rare brain diseases. The purpose of the support group is to help people with PSP and other prime of life diseases to develop new and existing methods of coping with the problems associated with these diseases.

### *Who attends CurePSP support group meetings?*

A very diverse group of people attends these groups. Brain diseases are not specific to any race, gender, socioeconomic group, or religious background. As is true of most support groups, there is a common bond: people whose lives are significantly affected by PSP and related brain disorders. Because of the mature discussions that take place during the meetings, the appropriate age for people attending the meetings is 18 years and older.

The following types of individuals can be involved in attending support group meetings. Many groups involve both the person with the disease and family members or significant others, since sharing mutual concerns and perspectives can often be especially helpful. However, some group meetings are just for family members or patients separately, creating a welcome haven for individuals to share their personal point of view as to how they experience dealing with their loved ones.

Professionals may be invited to attend support groups—for example, nurses, social workers, physicians, or anyone who serves as a paid helper who provides assistance to people with these diseases and their families.

### *What happens at meetings?*

The meetings provide the opportunity to share information and mutual support. Meetings might consist of group sharing without a specific agenda, a planned program with literature provided, or an open discussion surrounding a topic of interest. Whatever the format for a particular meeting, there is always an opportunity for personal sharing.

## *What do people learn?*

**Information about PSP and related neurodegenerative brain diseases:**

- What are typical symptoms?
- What constitutes a thorough evaluation and diagnosis?
- How do these diseases vary from person to person?
- What can be expected as the disease progresses?
- What treatment options and clinical trials exist?

**Information about available resources in the community:**

- Where do I go for help?
- What disability benefits are available and how do I apply?
- What options are available for respite care?
- What if I have to admit my loved one to a nursing home?

**Information about identifying methods of coping:**

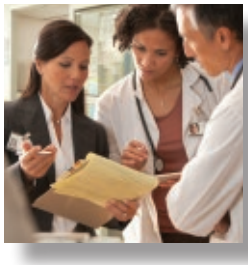
- What if there is no help at times?
- What if there is family conflict, or lack of understanding on the part of well-meaning relatives and friends?
- How do family members deal with the possibility of needing to place a loved one into a nursing home?

Feelings of anger, fear, guilt, frustration, and grief interfere with decision-making. Talking about feelings with others helps to sort things out. Through sharing together, people help each other appreciate the range of responses to situations, their growing knowledge of the disease, and their ability to make use of practical coping strategies.



## *How do I find a CurePSP support group?*

Please go to our website *www.psp.org/ineedsupport/supportgroups/* where you will find listings. If you don't have access to the Internet, please call Joanna Teters at 347-294-2871.



# When to Stop Driving: The Hard Conversation

**Lissa Robins Kapust, MSW, LICSW**

## *PSP and Driving Safety*

Driving is a complex skill that demands high-level cognitive and physical abilities. Many view driving as a marker of independence. However, PSP will likely jeopardize safe driving at some point in the illness. Those with PSP and their family members will need to monitor the course of the illness and the impact of symptoms on driving safety. It is important to recognize that no two patients with PSP will progress in the same way: each situation is different. But because driving can impact personal and public safety, it is an issue that bears careful monitoring and in most people with PSP, is impaired early in the course of the illness, typically after only two or three years, often sooner. One way to monitor driving is for family members to ride as passengers with the person who has PSP. It can be a red flag when family members no longer allow the person with PSP to drive with others, especially with children.

PSP is an illness that affects motor and cognitive domains. Motor symptoms such as stiffness and slowness may limit the speed and accuracy of manipulating and switching between the gas pedal and the brake. For some people, eye movement problems lead to an inability to scan the environment actively and accurately. This may be further exacerbated by neck stiffness, which limits the ability to turn the head for compensatory scanning. The ability to judge distances is impaired early on in PSP.



As important as the motor skills necessary for safe driving are cognitive and emotional factors. PSP can decrease speed of processing information, which is important in all aspects of driving and decision-making. This is especially true when unexpected situations arise and a decision needs to be made quickly. Poor sleep, a common issue in PSP, can contribute to problems maintaining attention. Judgment errors add to the risk of safe driving. Other contributing behavioral changes—such as impulsivity, depression, and anxiety—also negatively impact driving safety.

**Our society values independence and the ability to drive.** But when a person becomes a danger to themselves or others, it is time to consider giving up this privilege. Different physical and cognitive conditions may impair driving. It is advisable to ask physicians about conditions or medications that may affect driving ability. It is important for families to observe driving behavior over time.

Some of the warning signs of driving problems include:

- Hitting curbs
- Dents or scrapes on the car or garage
- Getting lost
- Diminished insight and judgment
- Impulsive behavior
- Driving too slowly for the conditions
- Incorrect signaling
- Making turns too widely or sharply (inability to judge a turn's radius)
- Changing lanes without looking
- Making wrong judgments, causing other drivers to constantly honk or take evasive action

- Stopping at green lights instead of red lights
- Not understanding signage
- Getting confused at freeway entrances and merges
- Driving the wrong way on a one-way street

There is no right way to resolve the driving difficulty issue. The responses of individual family members may vary. Those involved with the care of the driver need to remain focused on the self-respect of their loved one and the safety of others on the roadways. Understandably, one doesn't want to prematurely end driving, but waiting too long can jeopardize safety and result in serious injury or death.

## *What to Do*

1. Begin discussions and planning early; anticipate with the patient that there may be a time when they can no longer drive. When possible, involve the driver in these conversations. Open and early communication can help avoid a crisis later. Many carepartners permit their loved ones to drive longer than they know they should, causing carepartner anxiety and fear of putting others at risk.

2. When it is time to give up the keys, explain everything to your loved one and do not become critical of them. "Medicalizing" the problem can help. For example, let your loved one know that PSP results in symptoms that interfere with safe driving. You are there to support the person and tell them you understand their feelings.

3. Never leave the keys out where they can be found.

4. Assure your loved one that they can depend on you to meet their transportation needs.



## *Determining Driving Safety*

It is sometimes hard to know the degree to which illness impacts safe driving. If the impaired driver resists driving cessation, talk to the neurologist or family doctor. Based on the office visit and the patient's history of crashes, the physician may believe that they can strongly recommend the patient stop driving. However, sometimes it is hard to know from the office visit whether the patient is still safe to drive, since driving is an overlearned skill (like riding a bike). Each state differs in its procedures, but you or the neurologist may choose to contact your Division of Motor Vehicles and supply the DMV with a Driver Medical Evaluation form. In some states, the driver will then be notified of the need for a formal driving assessment. Another good option is to check to see if driving safety can be assessed through a nearby rehabilitation hospital or another hospital offering this service. Your physician can order such an evaluation. These formal assessments are valuable since the emotionally-charged decision about driving is turned over to skilled professionals who can render an objective opinion. These assessments, often done by an occupational therapist, measure key cognitive functions linked to driving safety, as well as visual and physical functions related to driving. The most comprehensive programs also include an on-road test, most often conducted in the car of a certified driving instructor. If patients do well with this assessment, they and the family can be reassured. Often, follow-up testing is necessary. The guidelines of the American Academy of Neurology suggest six-month follow-up for anyone with a progressive neurological condition.

For information on physical and cognitive impairments and driving, contact your local DMV and ask for the Regional Driver Safety Office. Explain the situation and ask about resources.

*Conclusion*

Having the discussion about driving safety is difficult. Driving cessation can be viewed as a sign that one is sick, with the need for major lifestyle changes. Sometimes the person has insight into driving difficulties and can participate in helping make the decision about when to stop driving. But often it is more challenging. Driving alternatives—and in some cases, a move—can help, but, as a non-driver, the spontaneity of going out for an ice cream on a hot July evening may not be possible. Hopefully, with thoughtful discussion, the person with PSP will adjust and the need for more authoritarian measures will not be needed (for example: reporting to the DMV, or removing or disabling the car).

Despite resistance to ending driving, some individuals are actually relieved to stop driving. They may have been in some very anxiety-provoking situations on the road, and they may not have shared this information with family.

In any case, work with the person with PSP to define themselves as much more than a driver—rather, as a person with many strengths. Professional counselors can provide additional help for those experiencing significant mood changes as a result of driving cessation. The family can highlight for the affected individual other times in their life when they have adjusted to change with strength and resilience. Hopefully the individual can draw on these past strengths and family support to manage the transition to no longer driving.



## Understanding Palliative and Hospice Care

### Diane Breslow, MSW, LCSW

CurePSP is committed to providing its constituents—patients, families, caregivers, and healthcare providers—with up-to-date educational information about topics relevant to living with PSP, CBD, and MSA.

The terms *palliative care, hospice care, end-of-life care, comfort care,* and *supportive care* are widely misunderstood and often used interchangeably. They also may conjure fear, stigma, and myth. One of the most common misconceptions is that palliative care and hospice care mean "giving up."

This article will help you understand the care options defined as palliative care and hospice care. In addition, you will find a list of available resources to assist you in making informed decisions about enlisting palliative or hospice services.

**Comfort care** is the umbrella term for patient care that is focused on symptom control, pain relief, and quality of life. Comfort care includes supportive care to family members by explaining the care plan, responding to questions and concerns, and providing emotional comfort.

## *Myths and Facts*

*Myth: Palliative care and hospice care are the same.*
**Fact: Palliative care and hospice care do not mean the same thing. This section, and the various agencies listed in the Resources section below, can help you understand the differences between palliative and hospice care.**

*Myth: Palliative care starts at the time of "terminal" diagnosis.*
**Fact: Palliative care can start upon diagnosis of a serious, chronic illness like PSP, CBD, or MSA, or at any time throughout a patient's disease progression.**

*Myth: Medicare covers hospice but not palliative care.*
**Fact: Medicare pays all hospice costs. Depending on your Medicare benefits and medical treatments, Medicare can also pay for palliative care. *It is important that you or your healthcare provider check with your insurance carrier regarding your benefits.***

*Myth: Palliative care does not permit medical, rehabilitation, or curative treatment.*
**Fact: Palliative care allows for symptom relief and also for tests, curative treatments, and therapies for disability. Hospice permits the patient to receive symptom relief (such as blood pressure or pain medications) but does not authorize the patient to receive curative or life-prolonging treatments. In some cases, patients enrolled in hospice may still receive a treatment typically thought of as life-prolonging (such as chemotherapy) when the purpose of that treatment is to bring comfort, not to extend life. *Make sure that you, and your physician, understand these distinctions as they apply to your particular case.***

*Myth: Palliative care occurs at home, while hospice care is provided in a facility.*
**Fact: Both palliative and hospice care can be provided wherever a person resides—in their own home, an assisted living facility, or a nursing home. In addition, some hospitals have palliative care teams, while some hospices offer the option of stand-alone hospice facilities. *Talk with your doctor or social worker about the best choice for you.***

*Myth: Hospice provides full-time care at the end of life.*
**Fact: Family members, paid caregivers, or the staff of a facility (assisted living or nursing home) give daily care to patients who are on hospice. The hospice team recommends and sets up a schedule for its staff—the hospice nurse, social worker, chaplain, volunteers, and others—to see the patient. If a person has chosen to die at home, meaning that family and friends are providing the hands-on daily care, hospice team members teach the lay people how to care for the dying person. Someone from the hospice team is always available by phone.**

## *Similarities Between Palliative Care and Hospice Care*

**BOTH are specialized care for people with serious illnesses.**

**BOTH require a physician's order.** The order is not for hospice per se; rather, it is for a hospice evaluation. A member of the hospice team—usually a nurse, sometimes the hospice physician—meets with the patient and family to explain hospice and to determine if the patient meets the admission criteria. If the patient and family find out that the patient is eligible, they can then decide whether or not they will take that route. In other words, choosing palliative or hospice care is a step-by-step, joint decision-making process involving hospice, the patient and family, and someone from the patient's healthcare team—physician, nurse, social worker.

**BOTH palliative care and hospice care are focused on relief from symptoms, including pain and stress.** Even on hospice, when a patient is not being treated curatively, they receive medical care that is helpful and contributes to comfort, such as blood pressure medication.

**BOTH aim to improve quality of life and dignity for the patient and the family.**

**BOTH are made up of teams that include a physician, nurse, social worker, nutritionist, and volunteer visitors, and may also include music, art, and massage therapists.**

**BOTH include the service of helping patients and families do advance care planning.**

**BOTH palliative care and hospice care recognize dying as part of the normal process of living. Neither palliative nor hospice care hastens or delays death.**

**BOTH result in decreased hospitalizations and decreased use of unhelpful services, and in the patient and family's higher perceived quality of care and quality of dying** (often seen as "not dying in the hospital"). (See research studies by Mitchell et. al., 2007; Teno et. al., 2011).

An Internet perusal of hospice care research identifies numerous references supporting the premise that palliative care and hospice care do not hasten death:

1. *"...hospice enrollment is not significantly associated with shorter survival; for certain terminally ill patients, hospice is associated with longer survival times."*
   *(Journal of Pain and Symptom Management, March 2007)*
2. *"...patients with terminal cancer that disenrolled from hospice care had significantly higher rates of hospitalizations...than patients who remained under the care of hospice."*
   *(Journal of Clinical Oncology, October 1, 2010)*
3. *Researchers found that patients receiving palliative care reported a higher quality of life through the final course of their illness.*



## *Differences Between Palliative Care and Hospice Care*

**When the care begins:** Palliative care can begin at any point in a person's disease process and can be provided concurrently with curative treatments. A referral for palliative care is not dependent upon prognosis or life expectancy. In fact, palliative care can help patients understand treatment options over time and throughout their disease.

Hospice care is for people in the final phase of life-limiting illness when life-saving treatment no longer works or is wanted, and the physician believes that the person has six months or less to live if the illness were to run its natural course. The patient who agrees to hospice understands that their illness is not responsive to treatments that intend to slow or cure it. Provided that a proper re-evaluation is conducted and appropriate hospice eligibility criteria are met, hospice services can be renewed beyond the initial six months. Likewise, a person could choose to discontinue hospice care and re-enroll at a later date.

**Who provides the care:** Palliative care is provided by a healthcare team of doctors, nurses, and other specialists who work with the patient's current treatment team.

Hospice care is provided by a designated hospice multidisciplinary healthcare team of practitioners: nurses, doctors, social workers, spiritual counselors, massage therapists, creative arts therapists, and trained volunteers. In some cases, if the patient has had a palliative care team, those practitioners continue to provide services but in the new role of hospice care. A hospice team member is available by phone 24 hours a day, 7 days a week.

Patients receiving palliative or hospice care can continue to see their primary care physician and other specialists, though insurance coverage may vary; check with your insurance carrier for limitations.

### *Families' Experiences Associated with High Satisfaction Regarding Hospice Services*

(National Hospice and Palliative Care Organization, 2005—based upon 116,974 surveys from 819 hospices; National Institute on Aging, July 2016)

- Family members concurred that the hospice team regularly informed them about their loved one's condition.
- The family felt that the hospice team provided family members with the right amount of emotional support.
- Family members felt that the hospice team provided them with accurate information about the patient's medical treatment.
- Family members could identify one nurse as the team leader in charge of their loved one's care and care plan.

### *The Take-Away Message*

Although similar, palliative care and hospice care are two distinct care options available to people living with chronic illness or disease. In particular, hospices vary widely as to the services, professional staff, and frequency of visits they offer. A referring practitioner cannot (and should not) promise or assure which services a patient and family will receive from a hospice organization. Families need to consult with hospice agencies on an individual basis to ask questions specific to one's own situation. We at CurePSP hope that you will discuss your needs and concerns with your family, your physician(s), and an experienced social worker or nurse, and then interview at least two different palliative care and/or hospice agencies to determine which one is the best fit for you.



### *Resources*

- American Academy of Hospice and Palliative Medicine • info@aahpm.org • www.palliativedoctors.org
- Center to Advance Palliative Care • 212-201-2670 • capc@mssm.edu • www.getpalliativecare.org
- Centers for Medicare & Medicaid Services • 800-633-4227 • www.medicare.gov • www.medicare.gov/coverage/hospice-care
- Death with Dignity • 503-228-4415 • www.deathwithdignity.org
- Education in Palliative and End-of-Life Care (EPEC) • 312-503-3732 • info@epec.net • www.epec.net
- Eldercare Locator • 800-677-1116 • www.eldercare.acl.gov
- Family Caregiver Alliance—National Center on Caregiving • 800-445-8106 • www.caregiver.org
- Five Wishes • www.fivewishes.org
- Grandfolk • www.grandfolk.com
- Hospice and Palliative Nurses Association • 412-787-9301 • hpna@hpna.org • www.advancingexpertcare.org
- Hospice Association of America—National Association for Home Care and Hospice • 202-546-4759 • hospice.nahc.org
- Hospice Foundation of America • 800-854-3402 • info@hospicefoundation.org • www.hospicefoundation.org
- National Hospice and Palliative Care Organization • 800-658-8898 • caringinfo@nhpco.org • www.caringinfo.org • www.nhpco.org
- National Institute on Aging Information Center • 800-222-2225 • niaic@nia.nih.gov • www.nia.nih.gov
- U.S. Department of Veterans Affairs • www.va.gov/GERIATRICS/Guide/LongTermCare/Hospice_Care.asp

## Caring from a Distance

### Susan C. Imke, FNP, GNP-C

Relatives who live far from loved ones coping with chronic neurological illness are often at a loss as to how to be helpful in ways that are both meaningful and practical. It can be tempting for family members at a distance to offer advice that may not be welcome or easily implemented.

Part of my work is facilitating family conferences for the purpose of making elder care decisions for people with PSP and related neurodegenerative disorders. Sometimes, the son or daughter who lives furthest away is the one with the strongest opinion regarding what Mom or Dad needs. This can result in friction with weary or exhausted front-line caregivers.

While relatives not struggling with issues of daily care do have a valuable perspective to share, this objective wisdom must be expressed with great tact and without criticism for family members who live close by and carry the greater responsibility.

All family members need to avoid making rash promises to the person with chronic health problems. Rather than saying, "Of course we'd never put you in a nursing home," express your empathy for a parent's preference to live at home, and your willingness to explore alternatives as care needs change.

So, what *can* long-distance relatives do to be helpful, short of moving back home? Perhaps some of the following suggestions from caregivers in support groups around the U.S. will meet needs for your family:

- Agree on a designated day and time to call each week. Do not assume that "no news is good news." Many older parents keep their own counsel even during tough times, not wanting to bother children who are busy with their own lives. Make the call faithfully. A short call more frequently may be better than an extended telephone visit. You may need to do most of the talking. Share your own family's activities since you last spoke.
- Try not to over-quiz the elders about health issues; this may put them on the defensive. If you are the son, speak for yourself. Your parents may dearly love their daughter-in-law but are likely to feel grateful to hear from you personally.
- Inquire tactfully whether financial help is needed. Many parents won't ask for help of a monetary nature, even if living on a fixed income and the rising cost of medication can mean eliminating basic necessities.
- If a regular subsidy is unacceptable, offer something specific.
- Picking up the tab for pads and diapers is useful for the person with incontinence. Paying for a month's supply of medication that is not covered by insurance can be a great relief.
- Send a creative caregiver package once a month. This might be a flower arrangement, restaurant gift certificate, or a spa basket filled with elegant bath goodies. Even a simple card with an encouraging hand-written message can mean a lot in the midst of a trying day!
- Budget funds for regular visits to see your family. Consider getting your own ride from the airport. The primary caregiver should not have to take care of you, too! If crowded quarters put a strain on everyone, reserve a nearby hotel room. You are not there to be entertained but to provide a listening ear and a helping hand.

- Educate yourself about the medications your loved one is using, and if helpful, report problems and changes to their physicians. Inquire about the health of the "well" parent. Is your mom's mammogram overdue? Does your dad neglect his own medical checkup because he's preoccupied with taking care of his wife? Ask your parents to schedule an appointment with the neurologist for a time when you can accompany them. The doctor will benefit from hearing your observations, and you will learn much about your parents' communication style and be able to reinforce the medical instructions afterwards.
- Once or twice a year, offer and follow through with a 3- to 5-day respite stay for the primary caregiver. Arrive a day or two prior to the caregiver's leave of absence so that you can learn necessary skills to help your care recipient. If filling in personally is not possible, offer to pay for a respite stay in an extended care facility. Your family may be saving for a rainy day when it's pouring outside!
- If your parents are able to travel, invite them to visit you. For some older couples who are living with chronic neurological disease, it is easier to travel and be a guest in your home than to host family visits at their home. An added benefit when parents are visiting you is that relatives back home get a welcome break from their usual caregiving responsibilities.
- When the time comes, speak up as the out-of-towner with less caregiving burden, to affirm the difficult decision to place your loved one in a residential care facility. It is a mistake to allow worries about cash flow, sibling rivalry, or dwindling inheritances get in the way of doing what is best for the key players whose lives are most affected.



## Respite Care

### Diane Breslow, MSW, LCSW

*What is respite?*

Caregiving is a tough job with long hours. To maintain your health and sanity and provide the best care for your loved one, respite is essential. Respite refers to short-term, temporary care provided to people with disabilities so that their families can take a break from the daily routine of caregiving. Respite services may sometimes involve overnight care for an extended period of time. Respite is an insurance policy against overtaxing yourself so that you can continue to provide quality care for the person with PSP, CBD, or MSA. Respite care enables families to take vacations or even just a few hours of time off. Respite is often referred to as "a gift of time."

*What kinds of services are provided, and where?*

Services are provided in many ways depending on the provider, the needs of the family, and available funds. The two primary categories of respite include in-home and out-of-home services. The following are typical services provided in each category.

### In-Home Respite

**Informal, Personal Arrangements:** This refers to a situation in which a family caregiver prepares and trains a couple of friends or other family members to fill in. Every family caregiver should have a back-up plan, and more than one option, in case someone is not available.

**Home-Based Services:** A trained, licensed, insured, and bonded employee of a private or state agency comes to the home. Ideally, service is available 24 hours a day, 365 days a year. Even if you do not employ regular home care, you can use a home help agency for the purpose of respite.

**Companion Services:** These services are provided by individuals who are trained in caring for adults and children with disabilities. This type of service is sometimes provided through a non-profit or service organization. In many cities across the country, Area Agencies offer respite care to the at-home spousal or family caregiver.

**Trainer Services:** A caregiver may be selected by the family or may be a family member. This individual receives training from a respite program. These caregivers may be paid or unpaid.

### Out-of-Home Respite

**Family Care Homes:** Respite is provided in the provider's home. The home and the individual providing care should be licensed under state regulations.

**Respite Adult Day Care:** As discussed in the section "Understanding Levels of Home- and Community-Based Services and Residential Care," community-based adult day programs provide all-day, structured programming to adults with physical and cognitive impairments. While enhancing patients' socialization and keeping patients safe, these adult day centers also offer family caregivers a respite from the demands of full-time caregiving for someone who needs constant supervision.

**Residential Facilities:** Most long-term residential facilities—independent living, assisted living, and nursing homes—offer short-term, overnight respite stays.



## *How do family caregivers benefit from respite services?*

**More than providing direct relief, the benefits of respite care can also include:**

- Relaxation: Families can relax, gain peace of mind, and renew their humor and energy.
- Enjoyment: Families can enjoy favorite pastimes and pursue new activities.
- Stability: Respite can improve the family's ability to cope with daily responsibilities and maintain stability during crisis.
- Preservation: Respite helps preserve the family unit and lessens the pressures that might lead to institutionalization, divorce, neglect, or abuse.
- Involvement: Families can become involved in community activities and be less isolated.
- Time off: Couples or families can take a needed vacation or merely spend time together and time alone.
- Enrichment: Respite makes it possible for family members to establish individual identities and enrich their own personal growth and development.

**Things to Consider When Seeking Respite Services in Your Community**

- What kinds of services do I need? For example, long-term, short-term, or both, and why?
- Do I prefer services in my home or at an outside setting?
- Do I want to use an individual or an agency?
- Does the agency provide the type of services I need?
- What is the cost of services? How is payment arranged?
- What are the training and experience of the care providers? Will they need—and accept—additional disease-specific, hands-on instruction and education to meet the patient's needs?
- How, and by whom, are the care providers supervised?
- Does the program maintain up-to-date information about medical and other needs? Is there a written care plan?
- What procedures does the program have for emergencies?
- Can family members meet and interview the people who care for the patient?
- How far ahead of time do services need to be arranged?
- Are families limited to a certain number of hours of service?

**If Service is Provided Out-of-Home**

- What is a typical daily schedule?
- How are the meals handled?
- Is the program able to meet the individual's specific needs for physical assistance, medication, food preparation, and the like?
- Does the program offer transportation?
- What is the client-staff ratio?

Many family caregivers have a difficult time reaching out for help and relief from their caregiving responsibilities. Sometimes, a respite trial can be a good way to experience what it is like to let someone else care for your loved one. You can gain knowledge and clarity by thinking through and discussing your questions with others. Hopefully, your experience with respite meets the dual goals of helping you—the carepartner—to feel revived and refreshed while at the same time taking care of your loved one's needs.



PART FOUR

# Being A Carepartner

# Keeping Families Strong

**Ileen McFarland**
**Diane Breslow, MSW, LCSW**

PSP, CBD, and MSA are very challenging diseases that have significant impact on patients as well as their families. It is important to respect and accept that each person in the family may have different reactions and coping styles. Nonetheless, family members will benefit from exploring how each of you can grow stronger while traversing this difficult journey.

## *Family Roles*

You may notice that, once a diagnosis is given, family members assume different positions or roles in the family system. While not an exhaustive or rigid list, the following role descriptions may help you gain a better understanding of your family dynamics and feel less alone.

**The Leader:** Often, one family member steps up to become the Leader. They take control of the situation. They obtain a wealth of disease information. They initiate actions such as making medical appointments. They appoint themselves in charge of communication with other family members. And they are the one to offer support. Being actively engaged is one way that the family "leader" deals with emotions.

However, the negative or flip side to this leadership role is that the Leader may become overburdened and burned out. Wanting to appear strong, they may have difficulty expressing emotions or asking for help, lest other family members view them as slacking in this role. Sometimes, the Leader can grow to resent those family members who are less involved than they are. Even if they do not verbalize these feelings, they will likely express them in actions.



**The Supporter:** Supporters are more than willing to help, but they lay out clear boundaries. They are most comfortable taking on specific tasks that have a defined beginning and end—for example, accompanying Dad to a doctor's appointment. The Supporter may be reluctant to provide hands-on help and may seem distant from other family members. The Supporter may openly express feelings of anger or resentment, which can be particularly hard for Leaders, who view themselves as taking on the lion's share of responsibility.

The other side of the Supporter's role can be a sense of feeling excluded and uninformed about the patient's disease and treatment. Knowing that their involvement is "only" task-related, the Supporter may feel that they do not have the right to ask for more information. Because they have set limits on what they will and will not do, they may also feel less valued, or de-value themselves. If they get a sense that other family members resent their boundaries, they may feel guilty or angry.

**The Bystander:** The Bystander pulls away during the course of the disease. They may reduce overall communication with the family and may avoid direct interaction with the person who is ill. They are very unlikely to take on caregiving tasks. Their lack of involvement often makes other family members feel taken advantage of, disappointed, frustrated, or angry. The bystander may have very understandable reasons for their distance.

Nonetheless, they are missing out on the rewards of caregiving: feeling helpful, feeling closer to the ill person or others in the family, or finding new meaning or purpose in life. Bystanders may feel helpless or ashamed about their role. They may become depressed, which, because of their self-imposed isolation, other family members may not recognize.

**The Only One:** In certain cases, only one person in the family is responsible for the care of the patient. Yet, at some point, every caregiver will need assistance with the patient's day-to-day care, not to mention back-up plans for emergency situations. The enormity of being the Only One clearly dictates the need for additional support. Over time, the Only One may become socially isolated and may compromise their own physical and mental health in favor of the patient's well-being. It is important for other family members, friends, clergy, and community members to reach out to the Only One in an effort to help them to develop a support network.

## *Characteristics of Strong Families*

Many factors contribute to and characterize family strength. The first is a **feeling of safety and trust** within a family. Strong families exhibit a respect for, and an encouragement of, sharing thoughts and feelings with each other, even in the face of disagreement, conflict, and intense emotions. Greater closeness and strength ensue when family members behave respectfully, listen openly to one another, and try to understand different points of view.

**Family history and tradition** can also foster family strength and a sense of togetherness. If families can maintain their customs and observances, even during illness, they have a better chance of staying close, keeping their family traditions alive, and building more memories.

Another characteristic of a strong family is the **willingness of individuals to help one another and to share responsibilities.** Most likely, each family member, whether near or far, has a special ability or talent that they can offer. For example, one individual may be more adept with financial records and bookkeeping. Another family member may enjoy shopping or running errands. Another may accompany the patient to doctor or therapy appointments. There are plenty of tasks to be distributed and shared.



**Resilience** is another component of strong and healthy families. "Resilience" comes from the Latin root meaning "leap back from" or rebound. Resilience is the capacity not only to withstand hardship but also to spring forward from it, often with a newfound sense of courage, determination, and inner resources. Resilient people can adapt to change, find new and creative solutions to complex problems, and accept what has become the "new normal." As the diseases of PSP, CBD, and MSA change and worsen over time, the "new normal" also changes. Families are called upon to be flexible, cooperative, tolerant, accommodating, and willing to compromise. No one can be this way all of the time. Yet, even while feeling conflicted or overwhelmed, strong families strive for this ideal of resilience and flexibility.

**Open boundaries:** Strong families let others in. People are not afraid or ashamed to ask for help, whether physical or emotional, in a healthy family. Open-boundary families welcome outside support and viewpoints.

## *Practical Pointers for Building a Strong(er) Family*

Although the person with the diagnosis is usually the family's central concern, family members also need to focus on themselves and how they can best work together. To that end, here are some pointers.

**Realize that there are stages of adapting to living with chronic illness.** Be patient with yourselves and the process. The stages are:

1. Disbelief, or rejection of the diagnosis
2. Recognition: The reality is hitting. Now what?
3. Reorganization: Bringing others into the circle of people who know and may be able to provide support
4. Resolution: The disease is real. What does it, or what will it, require? How will we build our network of support and find resilience and hope?

**Share positive emotions.** Openly express love, affection, and appreciation of one another.

**Accept each other.** Recognize the humanness in one another; respect each person's efforts to cope under such difficult circumstances.

**Define roles.** As a family, make decisions as to who will take on a particular task. Recognize that roles will need to change over time.

**Communicate.** Set up routines to stay in touch; attempt to resolve conflicts directly and quickly.

**Ask for help.** Develop solutions to new and ongoing problems by asking for help, by being open to others' ideas, or by joining a support group.

**Build in a sense of family togetherness.** Take time out to decompress and simply enjoy being with one another. Include your loved one who has the disease in your relaxing activity, be it music, family charades, or a meal.

**Maintain traditions.** Continue your family's meaningful activities and celebrations. These provide continuity and are a reminder of your family's history and love. However, also recognize that certain holidays can be highly stressful and anxiety-producing. Give yourself permission to miss a celebration if you feel that's best for you or your loved one.

### *Conclusion*

It is essential to acknowledge that everyone is human and that each person is dealing with multiple stressors all at the same time. Express your concerns and fears. Ask each other for support. Reach out to family members who are distant and try to express some of your own feelings and fears. This can open the door to a genuine exchange and a new understanding. Most importantly, stay close to your loved one who has the disease. Remember who they are, what you love about them, and that each moment you share is precious.



## Talking to Children and Grandchildren

**Elaine S. Book, MSW, RSW**
**Diane Breslow, MSW, LCSW**

A diagnosis of PSP, MSA, or CBD for an individual is actually a diagnosis for the family. It changes life for everyone in the family, including children of all ages. Supporting the children in the family will require preparation; awareness of age, stage, and personality of the children; and your own acceptance of, or coming to terms with, the condition.

### *Possible Effects on Children*

Just as the symptoms of PSP, MSA, and CBD vary and change over time for each person, so do the reactions, emotions, and needs of each individual child within the family. Their adjustment to hearing about the diagnosis and to the progression of the condition may be related to specific symptoms, the nature of the relationship with the affected individual, the family structure, the family's social supports, and other influences such as culture, religion, and outside influences.

Common feelings some children may experience include:

- Shame or self-consciousness about the way the condition presents (for example, a parent in a wheelchair)
- Anxiety about the stress in the family as a result of the diagnosis and symptoms of the condition
- Sadness, fear, anger, and grief regarding the changes and loss of experiences with the person who has PSP, MSA, or CBD
- Loneliness when one parent spends increasingly more time providing care to the person with the condition
- Worries about their risk of developing the condition, about the families' finances (especially if income has changed and care costs have increased), and about the impact of the condition on their own future aspirations and decisions
- Feeling overwhelmed by caregiving responsibilities
- Frustrations with their own activities being restricted (such as not wanting to bring friends home, or feeling bad about going out while one parent is home potentially burdened by symptoms and the other parent may be burdened by caregiving responsibility)

Remember that young children cannot always identify their emotions and instead may express them through behaviors (for example, increased clinginess, fears, inattention at school, attention-seeking behaviors).

Living with a challenging illness in the family can also have positive effects on children. The experience can strengthen family relationships, help improve coping skills, and instill a sense of pride and confidence in one's ability to handle life's trials. In addition, children who are exposed to chronic illness develop an earlier, and strong, sense of empathy and compassion, which sometimes leads to life-changing educational and career choices. Just as a caregiver and affected spouse may experience personal growth as a result of the illness, children too may grow and develop in unanticipated ways.



Be mindful that as children grow up, there may be difficult times or behaviors that are not related to the disease in the family but are more likely a result of the child's or teen's social and emotional developmental stage. Children who are managing daily stressors in their own individual lives may have a harder time coping with the illness in the family.

### *What, when, and how to talk with your children or grandchildren about these diseases*

Many parents are understandably concerned about what, when, and how to tell the children about the diagnosis. Whatever the age of the children, think about:

- Sharing the diagnosis and the process of having received the diagnosis
- Providing information about PSP, MSA, or CBD that is appropriate to the child's age and developmental stage
- Using language that is easy to understand and that focuses on symptoms and behaviors that the child sees and can relate to
- Communicating your best understanding of disease progression, in response to the child's questions and in amounts of information that the child may be able to digest at one time
- Practicing what you are going to say before talking with the children
- Planning for a time to talk that will be free of interruptions, especially for the initial conversation about the diagnosis

Below are examples of easy-to-understand language that address some typical concerns that children may have:

- PSP/MSA/CBD is a progressive (changing) neurological (brain) illness that can affect a person's physical movements and other ways the body works. Sometimes these are changes you can see (poor balance that leads to falls, voice changes), and sometimes they are changes you can't see (forgetfulness, sad mood) but that still affect your parent/grandparent.
- PSP/MSA/CBD is not contagious. You cannot get it from touching someone or even being around them. It is rare for PSP/MSA/CBD to be genetic (inherited or passed down in your family).
- There are different treatments to help with the symptoms (physical and mental signs), including medications (pills). There are also physical therapists, occupational therapists, and speech therapists to help with some of the symptoms.

## *When to talk with children/grandchildren*

In general, secrecy can be more harmful than sharing the truth about health issues, as children tend to be quite perceptive. In addition, children are usually more afraid of the unknown than they are of information that is presented at their level. Children are inclined to say that they want more information rather than less, and earlier on in the condition rather than later. They also want to know what to expect as the disease progresses. Be sensitive to the timing of your discussions. For example, bring up the topic when everyone is least burdened, not at the end of long day when you/they are tired, or before a test or major sporting event, or when rushing out the door.

## *How to talk with children/grandchildren*



Your attitude and the way the diagnosis is presented can affect how children cope from the start. You want to be honest, hopeful, and mindful of tone of voice and choice of words. Encourage children to let you know what is on their minds and to ask questions. Communicate honestly, openly, and frequently about how this disease impacts the family. Discuss worries, find solutions together, and highlight what is working well. Consider asking, "How are things different for you now that Mom has MSA?" or "Is there something that I can do that would be helpful?" By acknowledging a child's feelings and fears and by providing the responses and reassurances children need, you will help replace their fear with knowledge. You want to provide an environment that is comfortable for children to ask questions. Answer the specific question the child has asked. And it is ok if you do not know all the answers or if you are unsure how to respond to difficult questions. Let the children know that their question is a good one and that you will get back to them with a good answer once you have thought about it.

Also, consider talking about research, possible new treatments, and specialists who are working hard to improve treatments and quality of life for people with PSP, MSA, and CBD.

## *Ways to support children and maintain family wellness*

Children may want to talk about the condition, but not with you. Ensure that they have someone to talk to (a friend, parent of a friend, teacher, coach, relative, member of clergy, or healthcare professional). Connect with other adults in their life who may offer support to children as needed. These adults may also be able to keep an eye out for changes in children that may be related to disease progression or the family's coping. Peer support is another option. Your neurologist, healthcare team, or organization may be able to match your child with another child in a similar situation. If a child displays significant behavior changes that begin to interfere with their normal day, seek a referral to a social worker or other mental health professional.

Journaling is a strategy that provides a safe space to express thoughts and feelings that children may not want to share with anyone else. This can include writing, drawing, poetry, random words, or images. Purchase a journal book and some pens, and suggest some prompts (for example: Something I found difficult today, Something I am worried about, Something that made me smile, etc.).

Children should be encouraged to continue leading their lives and remaining active and involved with their interests, hobbies, and friends. Check that your children have support systems at school, in the family, and in the community. Arrange for younger children to have play dates. Ask extended family to spend special time with your child. As children grow into teens and young adults, they may need these resources and support to encourage and validate making decisions that are right for them and their future.

Rather than isolating children from the person with the condition, find ways to allow the children to assist and to be involved. Be creative in helping the child to continue the connection with the person with PSP, MSA, or CBD. Children might want to hold packages for the person, push a wheelchair, sign greeting cards, be responsible for some household chores, play board games, tell jokes, watch a TV series, read a book, or help make a favorite recipe. Being involved in these ways will provide children with the opportunity for personal growth and for coming to terms with the illness in a healthy, productive way. Empower children by encouraging them to be involved in a fundraiser or awareness event. Being personally involved with chronic illness expands one's horizons and opens one's heart to being non-judgmental about others' limitations or differences.

## *Conclusion*

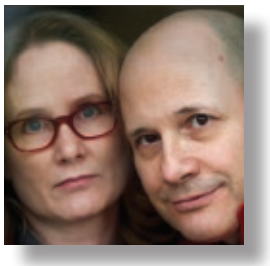
In general, children need to feel secure knowing that you will be able to manage whatever comes, that you will get the help that is needed, that you love them, and that you will all be ok—maybe different, but ok.

Children, like adults, need permission and time to grieve the changes and ongoing losses in their lives. Understandably, they may have to work through some sadness before adjusting to a new reality. Focus on what still exists—love, time together, and shared interests.

## *Resources*

- Pittsburgh Healthcare Report: This site has some good general information on prime of life neurodegeneration. • *www.pittsburghhealthcarereport.com/helping-kids-cope-when-a-parent-is-sick/*
- Marjorie E. Korff PACT Program: This site details how to support children at different ages. See the left sidebar of this page for age-specific tips. • *www.mghpact.org/for-parents/parenting-principles/developmental-perspective/overview*
- *How to Help Children Through a Parent's Serious Illness* by Kathleen McCue
- *Raising an Emotionally Healthy Child When a Parent is Sick* by Paula Rauch and Anna Muriel
- *The Etiquette of Illness* by Susan Halpern, with a chapter "Talking to Children about Illness"





# Facing Loss and the End of the Caregiver Role

## Diane Breslow, MSW, LCSW

Throughout the course of one's life, every loss helps prepare us for subsequent losses. Because of the progressive nature of PSP, CBD, MSA, and other neurodegenerative diseases, caregivers and others close to the patient have already experienced and grieved the step-by-step loss of the person they knew and loved. Yet, the death of the person you were caring for brings with it an additional loss and subsequent life transition: the end of your caregiver role and a period of mourning for that role. It is the beginning of a new and uncharted path in your own life.

The purpose of this article is to help you with the grieving process and to map a new course for yourself. Grief is not necessarily predictable nor the same for everyone. Every person moves through the series of steps at their own pace. Therefore, the suggestions contained here are meant to be general guidelines.

### *Grief*

The grieving process is unique to each individual, and its so-called stages are not sequential or linear. Nevertheless, each person is likely to pass through similar phases of grief and to experience common issues and emotions. In 1969, Elisabeth Kubler Ross was the first of many subsequent theorists to posit a framework for the stages of grief.

Her theory is comprised of five stages:

- Denial
- Anger
- Bargaining
- Depression
- Acceptance

A more recent theory, The Grief Cycle, posits the emotions of grief in a dynamic circle, rather than in stages (see the book by Shelly O'Brian listed with the Resources at the end of this chapter), as follows:

- Shock: Numbness, disbelief, inability to think straight
- Protest: Acknowledging the deep sorrow; consequently feeling anger, guilt, sadness, fear of what's next, yearning. Feelings, such as anger, at the person who died or at those who tried to help them are normal. Guilt is also normal: for example, guilt about the death of your loved one, guilt that you have survived, or guilt that you feel a sense of relief that your caregiving duties are over.
- Feeling of disorganization: Depression, loneliness, anxiety, confusion (perhaps because you feel relief that the suffering has ended not only for the one you cared for but for yourself as well), loss of identity, vulnerability
- Reorganization and redefining of self: A gradual return to "normal" functioning with new habits and routines but still feeling "not quite right"

Give yourself permission to experience and feel these emotions, and their fluctuations. Expect "anniversary" grief reactions on the dates of memorable occasions and holidays. With that in mind, mentally allow yourself to grieve for at least one year of seasons, holidays, and anniversary events without your loved one. Even consider planning for anniversary reactions. For example, think about what would feel right for you on a particular occasion. Let your family and friends know this, and ask them to honor your wishes. Do as much, or as little, as you can handle and allow yourself to be flexible and change plans if you feel you need to. Anniversary times are stressful enough as is, and grief can intensify that feeling of stress.

*Moving on—making a new life after caregiving*

Inside the circle of grief is the resolution, which the theory calls Recovery. Recovery means learning to live apart from and in spite of the loss. What can you do to help yourself arrive at this place of Recovery, or "Now what?"

**Take Care of Business**

- In view of the multiplicity of your losses—the loss of a dear loved one, the loss of your role and perhaps identity, the loss of much that filled your days and your mind—please, allow yourself time before plunging into the tasks of sorting through belongings, settling finances, passing on personal items, and managing family business matters.
- Delay making major life changes, such as selling a home, moving, or leaving a job.

**Take Care of Yourself**

- The basics: Nourishment, hydration, rest, exercise, relaxation breathing or meditation, and making and keeping doctor and dentist appointments.
- Exercise: Regular exercise contributes to improved circulation, flexibility, sleep, and sense of well-being.
- Breathing: Breathe in through your nose while counting to four, then exhale through your mouth to a count of four. Repeat 5 to 10 times.
- Accept offers of companionship and help from others.
- Rediscover what you enjoy.



**Interact with People**

- Perhaps you will want to or need to reconnect with people that you may not have seen frequently during your caregiving days and years.
- Reach out to trusted friends who will listen to you and support you in your grieving process and in building a new life.

**Develop Daily and Weekly Routines**

- Take time to figure out how you might want to spend your newfound time.
- Take small steps toward the goal of establishing new routines, activities, and interests.
- Structure at least a few days a week around a particular activity, class, job, or volunteer responsibility that gives you purpose and meaning. Perhaps schedule an activity where others rely on your presence.
- Volunteering: Many organizations offer volunteer opportunities on weekly schedules and also as one-time events. Consider how much time you might want to give. Decide whether you want to utilize your professional expertise or try a new area of interest. Think about location: Is it important to you to volunteer in a particular neighborhood, such as close to home or work? If you choose, you can even find volunteer jobs that you can do from your own home, such as recording audio books or making phone calls.

**Continue Your Own Healing**

- Talk about your loss. Face your thoughts and feelings about it.
- If you belonged to a caregiver support group, consider returning to the group at least once for reminiscence, goodbyes, and closure—or even for longer, if you are so inclined.
- Think about whether you would gain solace through volunteer work for CurePSP or another organization related to the disease or particular issues you were dealing with.
- Resume or start a creative endeavor, such as painting, music, crafts, or writing. Any of these activities enable self-expression and the possibility of seeing things from a new perspective.
- Join a bereavement or grief support group, either in person or online. If, in the past, you have been a member of support groups, you know firsthand some of the benefits: commonality among participants, feeling less alone, receiving understanding and feeling understood, the reward of reaching out to others, socialization and relationships, information, and resources.
- In addition to planned online groups, there is an app called Goodgrief. It asks you a few questions about your loss, offers additional filters, and then connects you on a one-to-one basis to people in similar situations.
- Turn to your faith or faith group for guidance.
- Keep alive the good memories of your loved one. Talk about them. Reminisce about the positive moments you and your loved one shared. Retain precious mementos. Enjoy special photographs with your friends and family members. Include children or grandchildren in conversations and, in general, in the processes of grieving and healing. Create a memory book. Throughout, bear in mind that, although death ends a life, it does not end a relationship or your feelings about the person.
- Reflect on, and embrace, the personal rewards of having been a caregiver: Did you learn patience? Acceptance? Taking one day at a time? Did you discover inner strengths and resources you didn't know you had? Do you have a sense of pride and contentment that you were there for your loved one?
- Be aware that grief may re-appear even after you thought it might be over.
- Reach out for professional help from a psychotherapist or bereavement counselor, particularly if grief persists for many months, interferes with your day-to-day life, causes you to lose (or not regain) interest and pleasure in life, or makes you preoccupied with thoughts of death or a desire to die.

## *The take-away message*

We hope these ideas and suggestions provide you with directions, options, and affirmation as you grieve the loss of your loved one and map a new course for your life. Grief and moving on are processes that take time, are not necessarily linear, and are different for every individual. For many people, grief continues—in varying degrees and occasions—for a long time. Give yourself the time, the space, and the course that is right for you.

## *Resources*

- AARP • 888-687-2277 • *www.aarp.org*
- Hospice Foundation of America • 800-854-3402 • *info@hospicefoundation.org* • *www.hospicefoundation.org*
- National Hospice and Palliative Care Organization • *www.caringinfo.org/i4a/pages/index.cfm?pageid=3367*
- National Library of Medicine • *https://medlineplus.gov/caregivers.html*
- *Peaks and Valleys – Integrative Approaches for Recovering from Loss* by Shelly L. O'Brian, LCSW, 2014.
- *Today's Caregiver* Online Magazine, After Caregiving topics • *www.caregiver.com/topics/caregiving*

## The 7 Deadly Emotions of Caregiving

### Paula Spencer Scott

Nobody would ever choose a smiley face as the perfect symbolic emoticon for a caregiver. Caregiving for an ailing loved one is just too stressful, often triggering damaging emotions that can not only undermine your good work but harm your health, as well. Here's how to cope:

**Caregiver emotion trap #1: Guilt**
*Guilt is virtually unavoidable as you try to "do it all."*

*What causes guilt:* Guilt stems from doing or saying what you believe is the wrong thing, not doing what you perceive to be enough, or otherwise not behaving in the "right" way, whether or not your perceptions are accurate. Caregivers often burden themselves with a long list of self-imposed "oughts," "shoulds," and "musts." A few examples: I must avoid putting Mom in a nursing home. I ought to visit every day. I shouldn't lose my temper with someone who has dementia.

*Risks of guilt:* Caregiver guilt is an especially corrosive emotion because you're beating yourself up over faults that are imagined, unavoidable—or simply human. That's counterproductive at a time when you need to be your own best advocate.

*What you can do:* Lower your standards from ideal to real; aim for a B+ in the many aspects of your life rather than an across-the-board A+. When guilt nags, ask yourself what's triggering it: A rigid "ought"? An unrealistic belief about your abilities? Above all, recognize that guilt is virtually unavoidable. Because your intentions are good but your time, resources, and skills are limited, you're just plain going to feel guilty sometimes—so try to get comfortable with that gap between perfection and reality instead of beating yourself up over it.

**Caregiver emotion trap #2: Resentment**
*This emotion is still so taboo that many caregivers are loathe to admit it.*

*What causes resentment:* Caregivers often feel put-upon and upset because of imagined slights by others, including siblings and adult children who don't do enough to help. Caregiver resentment is especially felt toward the person being cared for, when the caregiver's life feels hijacked by responsibility and out of her or his own control.

*Risks of resentment:* Without enough support or noncaregiving outlets, feelings of being ignored, abandoned, or criticized can fester into anger and depression.

*What you can do:* Simply naming this tricky emotion to a trusted confidante can bring some release. Try venting to a journal or anonymous blog. Know that resentment is a very natural and common response to long-term caregiving, especially if your work life, marriage, health, or outside activities are compromised as a result. Know, too, that you can feel this complicated emotion yet still be a good person and a good caregiver.

*See also: Chapters on Managing Symptoms and Managing Care and Getting Professional Support.*

### Caregiver emotion trap #3: Anger

*Some people outwardly show their anger more than others, but almost no one is never angry.*

*What causes anger:* We get mad for reasons both direct (a balky loved one, an unfair criticism, one too many mishaps in a day) and indirect (lack of sleep, frustration over lack of control, pent-up disappointment).

*Risks of anger:* Chronic anger and hostility have been linked to high blood pressure, heart attack and heart disease, digestive-tract disorders, and headaches. Anger that builds up unexpressed can lead to depression or anxiety, while anger that explodes outward can jeopardize relationships and even harm others. Managing caregiver anger not only helps your well-being but makes you less likely to take out your fury on your loved one.

*What you can do:* Rather than trying to avoid anger, learn to express it in healthy ways. Simple deep-breathing exercises can channel mounting anger into a calmer state, for example. Talk yourself down with soothing chants: It's okay. Let it go. Ask yourself if there's a constructive solution to situations that make you angry: Is a compromise possible? Would being more assertive (which is different from anger) help you feel a sense of control? Laughing at absurdities and idiotic behavior can provide a healthier biological release than snapping.

### Caregiver emotion trap #4: Worry

*A little goes a long way, but sometimes we can't turn off the fretting.*

*What causes worry:* Good intentions, love, and wanting the best for your loved ones are the wellsprings of worry. Focusing intensely on the what-ifs provides a perverse kind of comfort to the brain: If we're worrying, we're engaged. Of course, that ultimately triggers more worry and upset because it's engagement without accomplishing anything.

*Risks of worry:* Being concerned is harmless. Over worry and obsessing, however, can disrupt sleep, cause headaches and stomach aches, and lead to mindless eating or undereating.

*What you can do:* If you notice worrying thoughts interfering with getting through the day or sleeping at night, force a break to the cycle. Try setting a timer and resolving to focus on something else when the five minutes is up. Then flip negative thoughts to their productive side: How can you help? Can you call someone? Are there possible solutions? And don't be shy about seeking out a trained counselor to help you express and redirect obsessive ruminations more constructively.

### Caregiver emotion trap #5: Loneliness

*Your world can shrink almost before you realize what's happened.*

*What causes loneliness:* Friends may back away out of uncertainty or a belief they aren't wanted. Intense time demands lead you to drop out of outside activities. If you're dealing with dementia, the loss of your loved one's former level of companionship is another keenly felt social loss adding to isolation.

*Risks of loneliness:* Your very brain is altered: People with large, rich social networks have different brain structures, new research finds. Loneliness seems to curb willpower and the ability to persevere, and it can lead to overeating, smoking, and overuse of alcohol. Lonely people also have more cortisol, the stress hormone. And social isolation is a risk factor for dementia.

*What you can do:* Expand your social circles, real and virtual. Arrange respite help, so you can add at least one outside activity, such as one you've dropped. Take the initiative to reach out to old friends and invite them over if you can't get out easily. Consider joining a support group related to caregiving or your loved one's illness. In online support groups, you can find kinship with those who know just what you're going through.

**Caregiver emotion trap #6: Grief**

*Don't think this one applies yet? Think again.*

*What causes grief:* Although most people link grief with death, anticipatory grief is a similar emotion especially felt by caregivers who are coping with a loved one's long-term chronic illness.

*Risks of grief:* "Long goodbyes" can trigger guilt as well as sadness if one mistakenly believes that it's inappropriate to grieve someone still alive. Mourning the loss of a beloved companion is also a risk factor for depression.

*What you can do:* Know that your feelings are normal and as painful as "real" (postmortem) grief. Allow yourself to feel sadness and express it to your loved one as well as to supportive others; pasting on a happy face belies the truth and can be frustrating to the person who knows he or she is ill or dying. Make time for yourself so that you're living a life outside of caregiving that will support you both now and later.

**Caregiver emotion trap #7: Defensiveness**

*Protecting yourself is good—to a point.*



*What causes defensiveness:* When you're doing so much, it's only natural to bristle at suggestions that there might be different or better approaches. Especially if you're feeling stressed, insecure, or unsure, hearing comments or criticisms by others, or reading information that's contrary to your views, can inspire a knee-jerk response of self-protection: "I'm right; that's wrong!"

*Risks of defensiveness:* While nobody knows your loved one and your situation as well as you do, being overly defensive can make you closed-minded. You risk losing out on real help. You may be so close to the situation that you can't see the forest for the trees. For example, a social worker or friend may have a perspective that points to what really might be a better way.

*What you can do:* Try not to take everything you hear personally. Instead of immediately getting cross or discarding others' input, vow to pause long enough to consider it. Remember the big picture. Is there merit in a new idea, or not? What you're hearing as a criticism of you might be a well-intentioned attempt to help your loved one. You may decide things are fine as is, and that's great. But if you start from a point of calm and confidence, the focus becomes (as it should be) your loved one, not you.

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Paula Spencer Scott is the author of *Surviving Alzheimer's: Practical Tips and Soul-Saving Wisdom for Caregivers*.

# Make Meaning to Stay Positive


## Janet M. Edmunson, MEd


Taken from her book, *Finding Meaning with Charles*, and used with permission.

It wasn't until about the third year of Charles's disease that the Serenity Prayer had its greatest impact on me. But I have always loved it, and it means the most to me when I am going through tough times: "Grant me the serenity to accept the things I cannot change, courage to change the things I can, and wisdom to know the difference."

I have really latched onto the concept of "accepting the things I cannot change." Though we tried to fight it, Charles's disease was going to take away his abilities, and eventually, his life. Our choice was to accept that or deny it.

I don't know for sure what Charles's choice was. He was determined not to let the disease change his life and goals. He tried hard to keep contributing as best he could. That was how Charles tackled everything in life. He denied the obstacle and set out to conquer it. He wouldn't succumb. He fought all the way to the end. Is that acceptance or denial?

I, however, consciously chose to accept it and make the most of it.

My greatest learning through this experience came when Charles and I attended the Mind/Body Medical Institute program which, at the time, was held at Beth Israel Deaconess Hospital in Boston. The instructor, Peg, talked about acceptance, explaining that the way to accept the things that we have no control over, such as an illness, is to make meaning out of it. Wow! Make meaning out of it! Her explanation suddenly allowed me to consciously look at what Charles and I were going through and identify where it helped us grow and where it allowed us to have an impact on others that we wouldn't have had without the adversity of his disease.

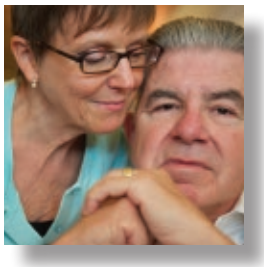
Taking a proactive approach to making meaning out of our situation helped me to positively focus on the opportunities and not plunge into depression. I was determined to help Charles reach whatever potential his life could give. And I was amazed to see how Charles became even more influential—even after he could no longer talk. As a caregiver, I found it important to focus on this greater purpose. My goal for caregiving went beyond making sure Charles was safe and physically cared for. I wanted to ensure that he still lived life to the fullest—to whatever degree the disease would allow.

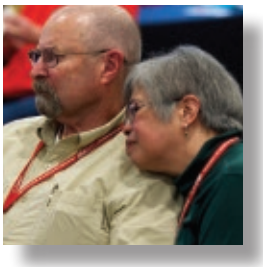
Accepting Charles's disease and making meaning out of it didn't mean that we didn't feel pain. Coping with this type of degeneration was difficult physically and emotionally for Charles, the person with the disease, as well as for me, the caregiver. We faced many trials—some successfully, others not. But we both became better people through experiencing his disease.

M. Scott Peck starts his book *The Road Less Traveled* with the sentence "Life is difficult." He goes on to explain that once we accept this, we can begin to make the most of life. Charles and I had discussed this concept a number of times when we faced problems at work or with other people. The misfortune of his disease forced us to face our greatest life difficulty, truly testing our ability to accept adversity and then move on.

I don't know of anyone who expressed this thought better than Viktor Frankl in his book *Man's Search for Meaning*. Frankl survived the atrocities and indignities of a concentration camp in World War II. He realized there that to renew our inner strength, we need to have a future goal. He quoted Nietzsche's words, "He who has a why to live for can bear with almost any how." I found that "finding meaning" is a way to define the why. The act of looking for and finding meaning in Charles's disease focused and empowered me.







PART FIVE

# Making Plans

## Estate Planning

*You can read or download CurePSP's Estate Planning Guide and Organizer at http://plannedgiving.curepsp.org or contact Joanna Teters at teters@curepsp.org or call 347-294-2871 to have one mailed to you.*

### *There are several benefits to making an estate plan:*

**Peace of mind for you —** An estate plan is designed to provide for you during life should the unforeseen (your incapacity) happen and for your family when the foreseen (your passing) does occur.

**Peace of mind for your family** — An estate plan and documents will help guide your family if they need to make difficult decisions about your care and provide the authority they might need to do so, and to know what to do when you are gone. Consider your plan a final gift to your family and other loved ones at the very time they need it the most.

**Distribution that you want —** Without an estate plan of some type, the laws of your state determine what happens to your property. This is called intestate succession (property inheritance when there is no will). Very likely the distributions it dictates will NOT be the ones you would have chosen. And no state distribution law provides for gifts to friends or charities, or makes provisions for your pets. Make sure what you've earned and accumulated in your lifetime goes to help those you love and causes you care about.



**Provide for your family –** An estate plan is especially important if you have minor children, as it will name a guardian to care for your children. In many instances, it will establish a trust to help ensure their financial well-being.

**Financially wise –** A good estate plan will help streamline the distribution process, minimize administrative costs, and possibly reduce taxes that might otherwise be owed. That means you leave the most you can to the people you love and the causes you care about.

### *Related to your final wishes*

- **Will.** A valid will is generally typed, dated, and signed by you as well as two legally competent witnesses. States differ as to whether a handwritten will, with or without witnesses, is valid.
- **Revocable Living Trust.** This can be used instead of a will as the main document disposing of your property. You might hear it referred to as a "living trust" or "RLT." The trust is created while you are living, most often people serve as their own trustee, and the power to change and even revoke it can be retained. The living trust becomes irrevocable upon your death. A living trust requires that you actually transfer your property into it for it to be effective.

*There are pros and cons with each approach, and an estate planning attorney can advise you as to which is best for your situation.*

*Note: Even if you decide upon a revocable living trust, you should still have what is called a "pour-over" will. It catches any property that was, intentionally or inadvertently, left out of the trust during your life and is not transferred in another way. While this property will still need to go through probate, it will eventually be distributed according to your trust instructions instead of being distributed under state law provisions.*

- **Beneficiary designations.** These are the forms you fill out when you do things like open a bank or stock brokerage account, establish an IRA or other type of retirement plan, or purchase a commercial annuity or life insurance policy. They specify who will receive whatever remains upon your passing (or the death benefit in the case of life insurance).
- **Form of ownership.** Jointly owned property that is "jointly owned with right of survivorship" passes directly to the surviving joint owner regardless of what the will or living trust might provide. This is most often seen with real estate but can involve other types of property as well. If you live in a community property state, your half of the community property will pass automatically to your spouse. These latter two means of passing property can have a profound impact on how your overall estate is distributed and should be considered as part of any coordinated plan.

## *Provide for physical and mental incapacity*

- **Power of Attorney (POA) for financial matters.** This document grants to someone you trust the ability to act on your behalf for a variety of potential transactions and responsibilities. When the POA becomes effective, the extent of the authority granted can be tailored to your particular desires.
- **Power of Attorney for healthcare decisions.** This document appoints someone to make decisions for you regarding medical treatment if you are not able to do so. It allows you to specify who is in charge of making critical treatment decisions and, perhaps more important, who does not have that authority.
- **Health Care Directive.** Sometimes referred to as an "advance directive" or "living will" (not to be confused with a living trust), this specifies the type of end-of-life treatment you want to receive. It is a directive to the physicians treating you and for the person holding your Health Care Power of Attorney.
- **Physician's Order for Life Sustaining Treatment (POLST).** This allows for your doctor, working with you, to document for the benefit of healthcare providers your wishes regarding resuscitation and other life-sustaining procedures.



## *Steps to having an estate plan*

*Depending on your situation, creating an estate plan doesn't have to be overly difficult or expensive. Here are some practical steps to get you started:*

1. Take inventory of what you own. List all of your assets and their approximate value. Include pertinent information about that asset.
2. Make a list of tangible personal property such as jewelry, dishes, books, furniture—items other than real estate and investments—and who is to receive each item upon your passing. You may want to maintain this as a separate list rather than designating this in your will, for maximum flexibility.
3. Think about your goals for your estate plan, for example, whom you want to benefit, how you want to treat each of your children, any special needs that you want to provide for, what happens if you and your spouse both pass away close in time, and if there are charities or organizations you want to remember. Your attorney will most likely ask you about goals you didn't consider, but at least you'll have a head start on those that are most top-of-mind.
4. Consider whom you would like to name as your agents, e.g., the executor of your will, the trustee of your trust, and the person to hold your power(s) of attorney, and gather pertinent information about them.

5. Go see an attorney, preferably one who specializes in estate planning. If you don't have one or know of one to call, check with family, friends, or co-workers for recommendations.
6. Follow through on whatever actions are decided upon in the meeting with your attorney. Rely on the advice of your attorney and other professional advisors as you make your decisions.
7. Share your plans with others. Key documents are of little or no value if no one knows what they say or where to find them when they are needed. This is especially true for the person(s) you have designated to serve as your personal administrator/executor under your will or the trustee of your living trust. It's also important to give loved ones at least a general sense of what to expect, so that there won't be surprises later on.

## *Frequently asked questions*

**Do I need to have an estate plan?**
Yes. Regardless of the size of your estate, you still want what you have to go to those you love and care for and that your wishes are carried out. But a good estate plan does far more than that. It cares for you as well as your things. It grants a Power of Attorney for financial and health matters should you become incapacitated and states your wishes regarding final medical care. Your estate documents become a last expression of what you have valued in your life, expressed through a personal statement and by what you leave to whom. By being thoughtful and organized about your affairs, you will have left a final, loving gift to your family and friends.



**Do I need to see an attorney?**
Yes. Estate planning is a very complex area of the law and shouldn't be left to a one-size-fits-all arrangement. This is especially true when you have a combined family. What is best for your sister and brother-in-law is not necessarily best for you! While there is a cost involved in preparing your plan, it is modest compared to the value of having appropriate arrangements for your family, minimizing probate fees and costs, and possibly saving state and federal estate taxes.

**How often should I update my plan?**
It is a good idea to update your plan every seven to 10 years. Some people have an annual checkup with their attorney. Certainly whenever there is a significant event in your life such as the birth of a child or grandchild, sale of a business, retirement, or death of a spouse or other loved one, you should review your plan for necessary changes.

**What if I have a plan but want to change one thing?**
If your plan is fairly current, it is easy to make a change or two, such as adding a charitable beneficiary. Your attorney can prepare an amendment to your will (called a "codicil") or to your living trust. Many times this can be done quickly and for a nominal cost.

The information provided here is offered solely as general education information and is not intended to be a substitute for professional estate planning or legal advice. Because the laws of each state vary and your own circumstances are unique, you should seek the advice of your own attorney, tax advisor, and/or financial planner before deciding on a course of action and in creating your estate plan.

# CurePSP Brain Tissue Donation Program


## Lawrence I. Golbe, MD

*Supported by the*
*Eloise H. Troxel Memorial Brain Bank at the Mayo Clinic*
*Jacksonville, Florida*


CurePSP asks you to consider brain tissue donation when the clinical diagnosis is made. A brain bank at the Mayo Clinic in Jacksonville, Florida, one of the leading such facilities in the world, is supported in part by CurePSP, which provided the initial impetus for the brain bank's founding in 1998.

The final autopsy report from the brain bank provides the authoritative diagnosis as well as information about other brain changes that may exist, including those that may have contributed to a clinical misdiagnosis. The Brain Bank provides tissue samples to reputable scientists worldwide for investigation into the genetics and other aspects of the causes of neurodegeneration. Of course, the donor's name or other identifying information is removed before the tissue is shared outside of the Mayo Clinic.

After a few weeks, the next-of-kin will receive a comprehensive autopsy report that often offers closure after many years of suffering and caring for a loved one. The Brain Bank does not charge the family for its services. Neither does it require payment from researchers to whom it supplies brain tissue.



## *Questions and Answers*

**Q. Why make your decision in advance?**

**A. For several reasons:**

- Your loved one can die suddenly, which is extremely stressful. A decision may have been made to donate the brain; however, without making prior arrangements, it is possible that the donation will not occur. The 24- to 48-hour window to perform the brain donation after death cannot always be met on short notice, especially on weekends and holidays.
- The patient, family members, and other loved ones should be involved in the decision. It may take time for everyone to come to an agreement. The Brain Bank coordinator is available to answer any questions and assist in making the arrangements.
- It can be difficult to locate a pathologist to perform the tissue collection. It is most important to have someone lined up in advance to make sure this procedure is accomplished within 24–48 hours after death.
- It is helpful to talk with family and friends, as well as the funeral home, about the planned arrangements. Planning in advance and the support of others will help during the time of grief. In addition to the brain donation, securing your loved one's medical records in advance is a significant help to the pathologist conducting the examination. Medical records show the disease progression. Records can be correlated with autopsy results.

**Q. Who may authorize a brain donation?**

**A. Legally, the patient and/or next-of-kin are the people who sign the Autopsy & Research Consent Form (available in our brochure).**

If the patient's spouse is deceased, the oldest child is considered next-of- kin. In some states, the patient or next-of-kin may sign the consent form prior to death. In other states, consent is not legally binding unless signed after death.

**Q. Will there be a need for any other tissue or organ donation?**

**A. No. In most instances where these diseases are suspected, only brain tissue will need to be examined for diagnosis.**

**Q. Where is the brain donation performed?**

**A. If death occurs in a hospital, and if the attending physician has ordered the procedure, the tissue collection will likely be performed in that hospital.**

If death takes place in a nursing home, hospice, or at home, then the body will have to be transported to the funeral home, crematorium, hospital, or medical examiner's office for the collection to take place. In that case, the funeral home may charge for transport.



**Q. Can there be an open casket?**

**A. Yes.** Collecting brain tissue for diagnosis and research leaves no disfigurement to the body. You must inform the pathologist or diener (pathologist's assistant) that there will be an open casket.

**Q. Will it be visibly noticeable that the brain has been removed?**

**A. Only on close inspection would anyone discover that a brain tissue collection has been performed.**

**Q. How do I find a pathologist who will perform the brain donation?**

**A. Please contact the Mayo Clinic Brain Bank coordinator,** Rachel LaPaille-Harwood, at *lapaille-harwood.rachel@mayo.edu* or 904-953-2439. The Brain Bank has a list of pathologists in the U.S. and Canada to help you locate a professional in your area. You can also ask a funeral director or the patient's neurologist.

**Q. Are there other ways to definitely confirm a diagnosis of neurodegenerative disease?**

**A. While clinical diagnosis has been greatly advanced, there is no way to confirm a diagnosis for most of these diseases other than by examining brain tissue.**

**Q. How long does it take for the autopsy report to be released?**

**A. Please allow up to 90 days for receiving the results of the autopsy.** The report will be sent to the person who is listed as next-of-kin.

**Q. I don't have any known brain disease; can I still donate my brain?**

**A. Yes, the Mayo Clinic Brain Bank actively searches for healthy brains.** These serve as valuable control brains in research studies. It is very important to compare pathological changes in diseased brains with healthy brains. This helps scientists to better understand disease processes and to develop novel therapeutic strategies. Please contact the Mayo Clinic Brain Bank for more information on healthy brain donation. Please note that CurePSP cannot provide financial assistance for the donation of healthy brains. If you have any questions, you may contact Dr. Lawrence Golbe at CurePSP, at *golbe@curepsp.org* or 347-294-2872.

After your decision to donate has been made, important paperwork is required to perform a legal tissue collection. Please follow the guidelines below and complete all four forms. You can download the forms from *www.psp.org/ineedsupport/braindonation*. Or you may contact Joanna Teters at *teters@curepsp.org* or 347-294-2871, and we will send you a printed copy of the forms.

**Please follow these seven steps:**

1. Contact the Brain Bank coordinator, Rachel LaPaille-Harwood, who will help you understand the next steps and procedures. Phone: 904-953-2439, Monday-Friday, 7:00 a.m. to 3:30 p.m. (U.S. Eastern Time). Email: *lapaille-harwood.rachel@mayo.edu*
2. Enroll in the Brain Bank by completing the Brain Bank Questionnaire (in the above-mentioned brochure) and mailing, faxing, or emailing it to the Brain Bank ahead of time. It is important that this information be on record once the brain arrives at the Mayo Clinic.
3. Please send copies of the Medical Release Form to all of the following physicians and neurologists:
   a. Those who are listed on the Autopsy & Research Consent Form
   b. Those who have treated the patient for a neurodegenerative disease
   c. Those whose clinical records could assist the researchers at the Brain Bank
4. The Medical Release Form authorizes physicians who diagnosed and/or treated the patient's neurological disease to send copies of their clinical notes to the Mayo Clinic. Only the patient or the next-of-kin can authorize the release of these records, which are important to the Mayo Clinic's researchers.
5. Complete the Autopsy Information Form. Make sure to have it placed in the patient's chart or medical files. The pathologist will sign and send the form, along with the brain tissue, to the Mayo Clinic Brain Bank.
6. Complete the Autopsy & Research Consent Form, signed by the patient and/or next-of-kin. Make sure to have it placed in the patient's chart or medical files. This is the actual consent for donation of a postmortem brain. This form can be signed only by the following individuals, and in this order of preference: patient, spouse, oldest adult child, parent, adult sibling, guardian, or power-of-attorney. For tissue collection to occur, the Autopsy & Research Consent Form, with original signature(s), and the Autopsy Information Form must accompany the deceased. Without a fully signed Autopsy & Research Consent Form, no brain donation is possible.

7. At the time of death, all family members and healthcare professionals need to know of the patient's wish to donate their brain. Please make sure that you have the pathologist's contact details available, so that they can be contacted immediately.

The Brain Bank coordinator can answer any questions about the donation process, assist in getting copies of the patient's medical records for use in ongoing research projects, and help locate a pathologist in your area to collect the tissue.

If the patient dies at home, in a nursing home, or with hospice, the funeral home or crematorium will be involved in arrangements for the tissue donation. At times, the procedure can be performed at the funeral home or crematorium. In other cases, it may be necessary to transport the body to a hospital or medical examiner's office for the procedure. If the patient dies in the hospital, be sure that the physician has placed an order in the patient's chart to have the tissue collected and sent to the Mayo Clinic Brain Bank. We emphasize that the most difficult step in this process is to identify someone nearby who is willing and able to remove, prepare, and ship the brain quickly after death. This is the most important reason to make arrangements in advance.

### *Cost of Brain Tissue Donation*

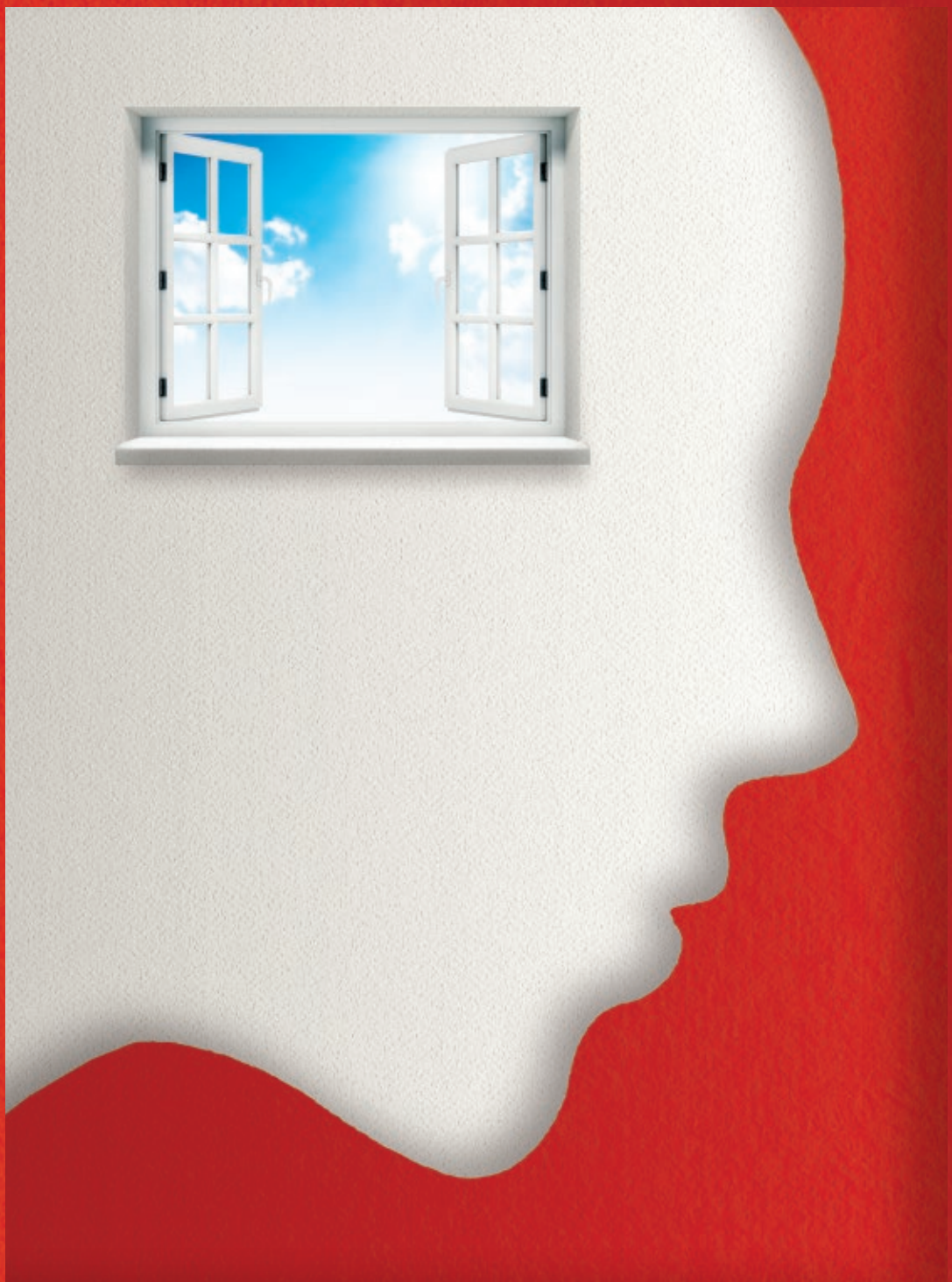
Neither CurePSP nor the Mayo Clinic imposes a charge for any part of the brain donation process. However, charges are typically imposed by the technician removing, preparing, and shipping the brain, and the funeral home that transports the deceased and provides a facility for brain removal. This cost typically ranges from $500 to $2,000 and is the responsibility of the family.

At CurePSP, we recognize that the expenses of the brain donation can be prohibitive for some families. A generous donor has contributed funds to create the CurePSP Brain Tissue Donation Fund. This has allowed CurePSP to provide financial assistance of up to $750 to families who wish to donate brain tissue but may have financial constraints. If your family needs assistance with the tissue collection costs, please contact Joanna Teters at 347-294-2871 or *teters@curepsp.org*. We emphasize that this program has limited resources and is intended only for families whose financial situation would not otherwise allow brain donation to occur.



PART SIX

# Appendix



## Glossary of Terms

**Activities of Daily Living (ADL):** Functions that are typically performed as part of a person's daily routine, such as dressing, bathing, eating, toileting, leisure activities, socialization, transferring, walking, and other functions of daily living. They are descriptions of physical functions that are useful tools when planning helping services for older people.

**Acute:** Referring to symptoms of abrupt onset, often of marked severity or intensity.

**Adult Day Care Centers:** Adult day healthcare centers are a program of services provided under health leadership in an ambulatory care setting for adults who do not require 24-hour institutional care. They offer supervised social and educational activities, including exercise, special events, nutrition, music, art, guest speakers, and family counseling.

**Akinetic:** Referring to absence of or poverty of voluntary movement; loss of the ability to move all or part of the body.

**Alzheimer's Disease:** A progressive degenerative disease of the brain of unknown cause. Alzheimer's disease is characterized by widespread loss of nerve cells, particularly in the outer region of the brain (cerebral cortex), with distinctive neurodegenerative changes (including "plaques" and "neurofibrillary tangles") and reduced activity of certain neurotransmitters of the brain. The disease is the most common cause of dementia or progressive deterioration of thought processing and acquired intellectual abilities. Associated symptoms include initial forgetfulness with increasingly severe memory impairment; disorientation and confusion; loss of the ability to recognize familiar people or objects through sensory stimuli (agnosia); and speech disturbances. The disorder may also be characterized by restlessness and agitation; an increasingly impaired ability to conduct purposeful movements; personality disintegration; and symptoms of psychosis, such as the perception of sights, sounds, or other sensations in the absence of external stimuli (hallucinations) and false beliefs of persecution despite evidence to the contrary (paranoid delusions).



**Ambulant (Ambulatory):** Able to walk; may be used to describe patients who do not require a wheelchair or are not confined to bed.

**Ambulation:** The act of walking.

**Antibodies:** Specialized proteins that function as an essential part of the immune system. Antibodies are produced by certain white blood cells (B cells) in response to the presence of specific, usually foreign proteins (i.e., antigens), helping the body to neutralize and destroy the invading microorganism, foreign tissue cell, or other antigen in question.

**Antioxidants:** Agents that inhibit or neutralize potentially harmful compounds known as free radicals. Free radicals are produced during metabolic activity. High levels of free radicals may eventually lead to impaired functioning and destruction of neurons and other cells in the body. Certain antioxidants are thought to neutralize free radicals before cellular damage occurs.

**Apraxia:** Loss of the ability to sequence, coordinate, and execute certain purposeful movements and gestures in the absence of motor weakness, paralysis, or sensory impairments. Apraxia is thought to result from damage to the cerebral cortex, such as due to stroke, brain tumors, head injury, infection, or neurodegenerative disease. It may also occur as a result of impaired development of the cortex as in certain neurodevelopmental disorders, including Rett syndrome. Apraxia may affect almost any voluntary movements, including those required for proper eye gaze, walking, speaking, or writing.

**Area Agencies on Aging (AAA):** The national network of senior service providers funded by Title III of the Older Americans Act (OAA) in 1965. They are administered through state and local government by the Administration on Aging. The Area Agency on Aging serves the needs of people 60 years and older.

**Assisted Living:** Facilities that are licensed to provide 24-hour assistance for those with the inability to perform some or all of the Activities of Daily Living (ADLs).

**Assistive Technology (AT):** Assistive technology helps individuals with disabilities perform activities that may otherwise be difficult or impossible. Examples of AT are wheelchairs, specialized computer keyboards, computers, and software that can read print out loud, communication devices that talk for individuals, and magnifying devices for those with low vision.

**Atrophy:** Wasting away or loss of a cell, tissue, or organ due to disease, malnutrition, insufficient blood supply, or other causes, such as loss of skeletal muscle mass due to peripheral nerve damage.

**Basal Ganglia:** Set of brain structures including the striatum and the substantia nigra, deep in the cerebral hemispheres. The basal ganglia are primarily involved in motor control, as well as other roles such as motor learning, executive functions and behaviors, and emotions.

**Bilateral:** Having or affecting two sides.

**Blink Rate:** The number of times per minute that the eyelid automatically closes—normally 10 to 30 times.

**Botulinum Toxin (BTX):** Any of a group of toxins, designated as A through G, produced by *Clostridium botulinum* bacteria. Localized injection of minute amounts of commercially prepared BTX may help to relax an overactive muscle by blocking the release of acetylcholine, a neurotransmitter responsible for the activation of muscle contractions. It was originally introduced in the 1970s for the treatment of misalignment of the eyes (strabismus) and involuntary contraction of eyelid muscles (blepharospasm) associated with dystonia or facial nerve disorders. There are four major forms of botulinum toxin in use for the treatment of medical conditions: onabotulinum toxin A (Botox), incotobulinum toxin A (Xeomin), abobotulinum toxin A (Dysport), and rimabotulinum toxin B (Myobloc).



**Bradykinesia:** Slowness of movement.

**Bradyphrenia:** Slowness of thought as seen in many brain diseases.

**Brainstem:** The region of the brain consisting of the medulla oblongata, pons, and midbrain. This area of the brain forms a connection between other brain regions and the spinal cord. Most of the 12 pairs of cranial nerves from the brain arise from the brainstem to regulate breathing, swallowing, digestion, heartbeat, blood pressure, pupil size, as well as the other basic movement of the eyes, face, palates, and neck. In addition, sensory functions, such as reflexes, vision, hearing, and taste are regulated. The brainstem also coordinates balance functions. Degeneration of one of its areas called the pedunculopontine nucleus is important in the balance loss of PSP.

**Carbidopa:** A drug that, when combined with levodopa, slows the peripheral breakdown of the levodopa, thereby allowing more of the levodopa to enter the brain.

**Case Management:** The primary goal of case management is to maintain the highest functioning, safety, and independence of each person by linking her or him to the needed services and resources available. A case manager will perform a comprehensive assessment in the home to determine a person's physical health, mental health, and safety needs, including limits and strengths. Based on this assessment, the case manager prepares a care plan.

**Case Managers:** Responsible for assessing, planning, implementing, coordinating, monitoring, and evaluating actions and services that patients and families need. Case management may need to be licensed or certified by states or other organizations depending on the funding agency's standards.

**Central Nervous System (CNS):** The brain and spinal cord. The CNS, which receives sensory impulses from and sends motor impulses to the peripheral nervous system (i.e., nerves outside the CNS). It controls thought, guides movement, registers sensations, and controls the activity of all parts of the body.

**Cerebellum:** A large structure at the lower back part of the brain responsible for the coordination of movement and balance, motor learning, and some cognitive function.

**Cerebral Cortex:** Outer layer of the brain; the wrinkly surface that you see when looking at a whole brain. The cerebral cortex is responsible for higher thought processes including speech, memory, attention, perception, consciousness, perception, and decision making. The cerebral cortex is divided into two hemispheres that are connected through the corpus callosum. Four different lobes on each side can be identified: the frontal, parietal, temporal, and occipital.

**Cerebrospinal Fluid (CSF):** The fluid that flows through and protects the brain. One hundred to 150ml of CSF flow in the four cavities (ventricles) of the brain, the spinal cord's central canal, and the space (known as the subarachnoid space) between the middle and inner layers of the membrane (meninges) enclosing the brain and spinal cord. Laboratory analysis of CSF, usually obtained via lumbar puncture, may help to diagnose central nervous system infections, certain tumors, or neurologic disorders. During lumbar puncture, CSF is removed from the spinal canal via a hollow needle inserted between certain bones of the spinal column within the lower back (i.e., usually the third and fourth lumbar vertebrae).



**Certified Nursing Assistant (CNA):** Provide bedside patient care under the direction of licensed vocational nurses and registered nurses. The CNA usually assists older adults in Activities of Daily Living. They may be employed by state nursing facilities.

**Chorea:** Jerky, irregular, relatively rapid involuntary movement that primarily involves muscles of the face or extremities. Choreic movements are relatively simple and discrete or highly complex in nature. Although involuntary and purposeless, these movements are sometimes incorporated into deliberate movement patterns.

**Chromosomes:** Thread-like structures within the nuclei of cells composed of DNA (deoxyribonucleic acid) that carries genetic information ("the genes") involved in directing cellular activities, thus controlling functioning and determining the expression of inherited traits.

**Cognition:** Mental actions and the process of acquiring knowledge such as perception, memory, awareness, reasoning, judgment, intellect, and imagination.

**Complementary and Alternative Medicine:** Complementary and alternative medicine, as defined by National Institutes of Health, is a group of diverse medical and healthcare systems, practices, and products that are not currently considered to be part of conventional medicine. It is important to check when using alternative medicine that these therapies were thoroughly tested in evidence-based (and not anecdotal) experiments and that they are FDA-approved.

**Computerized Tomography (CT) Imaging or Scanning:** An advanced diagnostic scanning technique during which cross-sectional images of tissues and organs are produced by passing X-rays through the body at various angles.

**Contractures:** Fixed resistance to passive stretching of certain muscles due to shortening or wasting (atrophy) of muscle fibers or the development of scar tissue (fibrosis) over joints.

**Corticobasal Degeneration (CBD):** CBD is considered one of the "parkinsonian" disorders, or "parkinsonisms." There are about a dozen such diseases, all of which produce some degree of slowness, muscle stiffness, balance problems, and sometimes tremor. Most people with CBD have these things, but in addition they have unusual difficulty performing complex limb movements such as cutting food, buttoning, or typing. There is often a tendency to hold part or all of a limb in a fixed posture called dystonia. There can also be very rapid, irregular, small movements of muscles called myoclonus. The unusual feature about CBD is that it is almost always very asymmetric, with one side affected much earlier and worse than the other. In half of people with CBD, it's the left, and in half, it's the right. The side affected has no relationship to the person's handedness.

As its name states, corticobasal degeneration is a loss of brain cells emphasizing the cerebral cortex and the basal ganglia. The cortex is the outer layer of the cerebrum, the gray matter where most thinking, speech, and sensory perception occur. The basal ganglia are also gray matter, but they're deeper in the cerebrum and coordinate movement. Basal refers to the base of the brain and the ganglia are collections of brain cells in one location devoted to a single purpose.

**Custodial Care:** Usually refers to care rendered in a skilled nursing facility. Custodial care does not require a licensed medical provider, such as a nurse or therapist.

**Degenerative:** Marked by or pertaining to deterioration; particularly, deterioration of the function or structure of tissue or changes from a higher to a lower or less functionally active form.

**Delirium:** A sudden and typically temporary state of confusion, agitation, lethargy, or alteration of sensory perception caused by a medical, as opposed to a psychiatric, condition. A few common causes of delirium are intoxications by alcohol or other chemicals, alcohol or drug withdrawal, concussions, brain infections, dehydration, insufficient oxygen supply to the brain, or a recent epileptic seizure. Delirium will usually resolve if the cause can be corrected.



**Delusions:** Persistent, aberrant beliefs.

**Dementia:** A neurological condition characterized by a progressive decline in intellectual functioning, resulting in impaired judgment, memory, and abstract thinking; disorientation; and personality disintegration. Dementia may result because of various underlying conditions, including certain neurodegenerative diseases, such as Alzheimer's disease or Huntington's disease; brain injury or tumors; inflammation of the brain (encephalitis); successive strokes; or a condition known as normal-pressure hydrocephalus, which is characterized by enlargement of cavities (ventricles) of the brain, with cerebrospinal fluid (CSF) pressure at the upper end of normal. Dementia in PSP and CBD, if it does occur, does not feature the memory problem that is so apparent in Alzheimer's disease. Rather, the dementia of PSP/CBD is characterized by slowed thought, difficulty resisting impulses, and difficulty synthesizing several different ideas into a new idea or plan. These mental functions are performed mostly by the front part of the brain (the frontal lobes). In Alzheimer's, on the other hand, the problem is mostly in the part of the brain just above the ears (the temporal lobes), where memory functions are concentrated.

**Differential Diagnosis:** Distinguishing between two or more diseases and conditions with similar symptoms by systematically comparing and contrasting their clinical findings, including physical signs, symptoms, as well as the results of laboratory tests and other appropriate diagnostic procedures.

**Dopamine:** A chemical that serves as one of the brain's most widespread neurotransmitters. Neurotransmitters help relay messages from one nerve cell to another. Dopamine is especially important in relaying messages about movement. The most prominent brain area hosting dopamine neurons is the substantia nigra, which is affected in Parkinson's disease, PSP, CBD, and MSA. In all of these disorders, many areas of the brain that do not use dopamine are also affected, and none of these disorders affects all of the dopamine-producing areas.

**Dopamine Agonist:** A drug that acts like dopamine on dopamine receptors to mimic its actions. Such medications stimulate dopamine receptors and produce dopamine-like effects. Often used in patients with Parkinson's disease.

**Dopamine Autoreceptor:** A type of dopamine receptor that acts like a thermostat, preventing excess dopamine release as levels rise.

**Dopamine Receptor:** A molecule on a receiving nerve cell (neuron) that is sensitive (or receptive) to stimulation (arousal) by dopamine or a dopamine agonist. Several types have been identified including D1, D2, and D3 receptors and the dopamine autoreceptor.

**Double-Blind Clinical Trial:** A double-blind trial is a clinical experiment in which neither the patients nor the researchers are aware of which patients are receiving the active treatment and which are receiving placebo.

**Durable Power of Attorney for Health Care (DPAHC):** A legal document whereby an individual grants authority to someone else to make healthcare decisions for her or him. These decisions are made if the individual becomes incapacitated and include the granting or withholding of life-sustaining treatment.

**Dysarthria:** Disordered or impaired articulation of speech due to disturbances of muscular control, usually resulting from damage to the central or peripheral nervous system.

**Dysesthesias:** Unpleasant sensations that are produced in response to normal stimuli.

**Dysphagia:** Difficulty in swallowing. Dysphagia may be associated with esophageal obstruction as well as certain neurodegenerative or motor disorders involving the esophagus.



**Dyspraxia:** Partial loss of the ability to coordinate and perform certain purposeful movements and gestures in the absence of motor or sensory impairments.

**Dystonia:** A neurologic movement disorder characterized by sustained muscle contractions, resulting in repetitive, involuntary, twisting, or writhing movements and unusual postures or positioning. Dystonia may be limited to specific muscle groups (focal dystonia), such as dystonia affecting muscles of the neck (cervical dystonia or spasmodic torticollis) or the eyes, resulting in closure of the eyelids (blepharospasm).

**Dystonic:** Referring to dystonia; sudden jerky or repetitive movements and muscle spasms due to impaired muscle tone and abnormal muscle rigidity.

**Elder Abuse:** Mistreatment of a person 65 years of age or older. Elder abuse may include physical abuse, neglect, intimidation, fiduciary abuse, abandonment, or other acts resulting in physical harm or mental suffering.

**Elder Law Attorney:** Attorneys who specialize in legal services affecting the elderly. These services include, but are not limited to, conservatorships, estate planning, decedent's estate, long-term care planning, Social Security, and elder abuse.

**Electroencephalography (EEG):** A noninvasive, diagnostic technique that records the electrical impulses produced by brain cell activity. An EEG reveals characteristic brain wave patterns that may assist in the diagnosis of neurologic conditions, such as seizure disorders, impaired consciousness, and brain lesions or tumors.

**Epidemiological Study:** Examination of the distribution of disease as well as the determining factors related to specific diseases or health-related problems in a specific population.

**Essential Tremor (ET):** A common, slowly and variably progressive neurologic movement disorder characterized by involuntary, rhythmic "back and forth" movements (i.e., tremor) of a body part or parts. In ET patients, tremor is primarily a "postural" or "kinetic" tremor or may be a combination of both types: i.e., tremor occurs while voluntarily maintaining a fixed position against gravity (postural tremor) and/or when conducting self-directed, targeted actions (kinetic intention tremor). In many individuals with ET, both hands are affected, although the condition may sometimes initially be noted in the dominant hand. ET also frequently affects the head. Less commonly, patients have tremor involving the voice, tongue, or roof of the mouth (palate), leading to impaired articulation of speech (dysarthria). ET may appear to occur randomly for unknown reasons (sporadically) or be transmitted as an autosomal dominant trait.

**Executive Function:** Executive function refers to a person's ability to establish a goal and then make decisions and put into action activities to meet that goal.

**Extended Care Facility:** A healthcare center (e.g., skilled nursing facility, long-term care facility, nursing home) that typically provides continuation of care after a hospital stay or when patients require care beyond that available in their own homes.

**Flexion:** The act of bending (as opposed to extending) a joint.

**Food and Drug Administration (FDA):** The FDA is an agency within the U.S. Department of Health and Human Services. The FDA is responsible for (a) protecting the public health by assuring that foods (except for meat from livestock, poultry, and some egg products, which are regulated by the U.S. Department of Agriculture) are safe, wholesome, sanitary, and properly labeled; ensuring that human and veterinary drugs, vaccines, other biological products, and medical devices intended for human use are safe and effective; (b) protecting the public from electronic product radiation; (c) assuring cosmetics and dietary supplements are safe and properly labeled; (d) regulating tobacco products; and (e) advancing the public health by helping to speed product innovations. The FDA's responsibilities extend to the 50 United States, the District of Columbia, Puerto Rico, Guam, the Virgin Islands, American Samoa, and other U.S. territories and possessions. This information was taken from the FDA's website.



**Free Radicals:** Free radicals are molecular species that are unstable, highly reactive, and can damage biologically relevant molecules such as DNA, proteins, carbohydrates, and lipids, thus causing cell damage and homeostatic disruption.

**Gait:** The style or manner of walking. Gait disturbances may be associated with certain neurologic or movement disorders (such as PSP, CBD, and MSA), orthopedic conditions, inflammatory conditions of the joints (i.e., arthritic changes), or other abnormalities.

**Gait Apraxia:** Loss of the ability to consciously sequence and execute the movements required to coordinate walking. Gait apraxia may result in unsteady walking patterns; "toe-walking;" a widely based, jerky gait; and balance difficulties.

**Gastrostomy Tube (G-Tube):** A plastic tube inserted into the stomach through a surgical incision in the abdomen. A gastrostomy tube is used to deliver liquefied food to the digestive system when swallowing becomes dangerous or difficult. The most common G-Tube procedure is the percutaneous (through the skin) endoscopic gastrostomy (PEG).

**Gene:** The smallest units of heredity. The information from all the genes, taken together, makes up the blueprint or plan for the human body and its functions (genome). A gene is a short segment of DNA, which is interpreted by the body as a plan or template for building a specific protein.

**Geriatrician:** A physician who has had basic postgraduate training in either internal medicine or family medicine with an additional one to two years training in medical, social, and psychological issues that concern older adults.

**Gland:** A structure or organ that makes a substance, such as a hormone or chemical, that is used elsewhere in the body. Some of these hormones and chemicals are insulin, bile, growth hormone, estrogen, and testosterone.

**Gray Matter:** Nerve tissue that primarily consists of nerve cell bodies, dendrites, and unmyelinated axons, thus having a gray appearance. In contrast, white matter predominantly contains myelinated nerve fibers.

**Half-Life:** The time required for the blood level of a drug to decrease by half after dosing is stopped. The half-life determines how frequently a drug has to be given to maintain a level in the tissues that is effective and safe.

**Hereditary:** Inherited; inborn; referring to the genetic transmission of a trait, condition, or disorder from parent to offspring.

**Home Health Care:** A skilled healthcare service provided to individuals who are home-bound and limited in their ability to leave their residence. Through intermittent home visits, nurses and therapists provide skilled nursing and physical, occupational, and speech therapy. Some personal care may also be covered in conjunction with the skilled care. A physician must order and monitor this service, which is reimbursed by Medicare and most private insurance plans for a limited period of time.



**Homecare Services:** Also known as companion care, domestic services, homemaker services, and personal care. Homecare (nonmedical) services provide assistance with housekeeping, personal care, and Activities of Daily Living (ADLs), all of which help the older adult to remain safely at home. These services may also include light housecleaning, meal preparation, doing laundry, grocery shopping, and running errands. Additional nonmedical homecare services include feeding, bathing, bowel and bladder care, and dressing.

**Homeopathy:** A system of medicine that is based on the Law of Similars, a belief in which substances that cause healthy people to exhibit symptoms can, conversely, be used to restore the body to health. Taken from the NIH website, please read the following key points carefully. More information and a general overview can be found at *https://nccih.nih.gov/health/homeopathy*.

*Key Points: (a) There is little evidence to support homeopathy as an effective treatment for any specific condition. (b) Although people sometimes assume that all homeopathic remedies are highly diluted and therefore unlikely to cause harm, some products labeled as homeopathic can contain substantial amounts of active ingredients and therefore could cause side effects and drug interactions. (c) Homeopathic remedies are regulated by the U.S. Food and Drug Administration (FDA). However, the FDA does not evaluate the remedies for safety or effectiveness. (d) Several key concepts of homeopathy are inconsistent with fundamental concepts of chemistry and physics. There are significant challenges in carrying out rigorous clinical research on homeopathic remedies. (e) Tell all your healthcare providers about any complementary health practices you use. Give them a full picture of all you do to manage your health. This will help ensure coordinated and safe care.*

**Homeostasis:** Homeostasis, from the Greek words for "same" and "steady," refers to any process that living things use to actively maintain fairly stable conditions necessary for survival.

**Hospice:** A special way of caring for a person with a terminal illness. A person is eligible to receive hospice care when he or she has a life expectancy of six months or less and chooses comfort care, over treatment, to cure the illness. A team of physicians, nurses, counselors, therapists, social workers, aides, and volunteers focus on the physical, emotional, and spiritual needs of patients and their families.

**Huntington's Disease (HD):** A hereditary, progressive, neurodegenerative disorder primarily characterized by the development of emotional, behavioral, and psychiatric abnormalities; gradual deterioration of thought processing and acquired intellectual abilities (dementia); and movement abnormalities, including involuntary, rapid, irregular jerky movements (chorea) of the face, arms, legs, or trunk. HD may be inherited as an autosomal dominant trait (inherited only from one parent) or, less commonly, appear to occur randomly for unknown reasons (sporadically). The disorder results from abnormally long sequences of information within a gene. Progressive nervous system dysfunction associated with HD results from loss of neurons in certain areas of the brain, including the basal ganglia and cerebral cortex.

**Hypokinetic:** Diminished movement and decreased motor function. Some movement disorders are hypokinetic, such as Parkinson's disease, PSP, and MSA.

**Hyposmia:** A decreased sense of smell.

**Idiopathic:** A disorder or condition of spontaneous origin; self-originated or of unknown cause. The term is derived from the prefix "idio-" meaning one's own and "pathos" indicating disease.

**Inflammation:** This is the body's first response to injury or irritation. The classic signs of inflammation are pain, heat, redness, swelling, activated immune response, and loss of function.

**Inhibition:** The restraint, suppression, or arrest of a process or the action of a cell or organ; the prevention or slowing of the rate of a chemical or an organic reaction. The term "reciprocal inhibition" refers to the restraint or "checking" of one group of muscles upon stimulation (excitation) and contraction of their opposing (antagonist) muscles.



**Inhibitor:** A substance that blocks, restricts, or interferes with a chemical reaction or other biologic activity.

**Insidious:** Of subtle, gradual, or imperceptible development; referring to the development of symptoms that may not be recognized by an affected individual until the disorder in question is established.

**Jejunostomy Tube:** Similar to a gastrostomy tube, although this tube is longer and is inserted through the abdominal wall into the jejunum, the middle section of the small intestine.

**Lateral:** Sideways; of, on, from, or toward the side.

**Levodopa:** A drug used to treat Parkinson's disease. It is also called L-dopa and, in the United States, is sold as Sinemet. Levodopa is converted by the body to dopamine, a neurotransmitter that is reduced in Parkinson's disease and PSP brains. However, levodopa does not work well in PSP patients.

**Lewy Body Disease:** Also called diffuse Lewy body disease or Lewy body dementia. Lewy body disease is a common cause of dementia, accounting for approximately 15–20% of all cases. The age of onset is typically in the late 50s through the 70s. Lewy body disease is characterized by more daily fluctuations in symptoms than Alzheimer's disease, as well as more prominent psychosis. Patients are prone to have adverse reactions to antipsychotics. Patients also have parkinsonian features early in the disease, including slowed movements and rigidity, though usually without tremor. In contrast to PSP and CBD, Lewy bodies are not made of accumulated and misfolded tau protein but consist of misfolded alpha-synuclein. In Lewy body disease, the Lewy bodies that cause neurodegeneration are most prominently found in the cortex, or surface of the brain, versus in the midbrain with Parkinson's disease.

**Long-Term Care (LTC):** An umbrella term referring to comprehensive healthcare delivered to people with functional impairments over an extended time period. An important goal of LTC is to provide care in the least restrictive environment (e.g., a person's home).

**Magnetic Resonance Imaging (MRI):** A diagnostic scanning technique during which radio waves and an electromagnetic field are used to help create detailed, cross-sectional images of specific organs and tissues. MRI images can help to diagnose brain atrophy and other pathological changes seen for example in PSP, CBD, and MSA.

**Medicaid:** A joint federal/state program of medical assistance for low-income individuals who are aged, blind, or disabled, or who are members of families with dependent children. Medicaid has no deductible or coinsurance. It typically covers inpatient hospital services, outpatient services, limited stays in skilled nursing facilities, limited home healthcare, lab tests, radiographs, family planning, early and periodic screening, diagnosis, and treatment.

**Medicare:** A federal health insurance program for people 65 years of age and over, people considered permanently disabled for purposes of the Social Security Act, and people with end-stage renal disease. Medicare is divided into two parts: Medicare Part A and Medicare Part B.

Part A, Hospital Insurance Benefits, provides some protection against the medically necessary costs of hospital and related healthcare. Medicare A is financed through Social Security payroll tax deductions. It covers institutional care in hospitals and skilled nursing facilities and care given by home health agencies and hospices. No premium is required from people entitled to retirement or disability benefits from Social Security or Railroad Retirement.

Part B, Medical Insurance Benefits, is financed through federal contributions and the monthly premiums of the enrollees. The monthly premiums are automatically deducted from Social Security checks, unless the beneficiary indicates that they do not want Medicare B. Medicare B covers outpatient services, physician visits, ambulance transportation, and durable medical equipment. It also covers some home healthcare. In addition to paying a monthly premium for Part B, Medicare beneficiaries are often required to pay a portion of the cost of the Medicare-covered services they receive. This cost-sharing takes the form of deductibles and co-insurance amounts. A beneficiary is responsible for a minimum of 20% of the Medicare-approved amount under Part B. These amounts may change annually. Older adults may find these changes in *The Medicare Handbook*, published annually and available at no charge through the Social Security Administration.



**Mental Health Services:** Provided by psychiatrists, psychologists, social workers, counselors, and other mental health professionals. Most clinical services may be provided on an outpatient basis. Crisis services are available. Occasionally, brief psychiatric hospitalization in a specialized unit for older adults may be required if the problem is more complex.

**Metabolism:** Refers to the ongoing chemical processes of cells of the body, including catabolism and anabolism. Catabolism or so-called "destructive metabolism" is the breakdown of complex chemical compounds into simpler substances, typically liberating or releasing energy. In contrast, anabolism or "constructive metabolism" refers to the "building up" or conversion of simple substances into more complex chemical compounds, requiring energy consumption (provided by catabolic processes).

**Multiple System Atrophy (MSA):** Multiple system atrophy is a disease of the brain and spinal cord. It is often classified as one of the "parkinsonian" conditions because it often resembles Parkinson's disease, at least for the first few years. The resemblance is in the general slowness, stiffness, and balance loss. But MSA usually has at least two other important categories of symptoms to some degree. One is impairment of the cerebellum, producing a coarse tremor, drunken-appearing walk, and slurred speech. The other is impairment of the autonomic nervous system, which maintains such things as blood pressure, sleep, bowel action, and bladder emptying. The result can be fainting, insomnia, constipation, and urinary urgency or incontinence. Many other less common symptoms can occur, too.

**National Institutes of Health (NIH):** The NIH is one of the world's foremost medical research centers and the federal focal point for medical research in the United States. The NIH, comprising 27 separate Institutes and Centers, is one of eight health agencies of the Public Health Service that, in turn, is part of the U.S. Department of Health and Human Services.

**Necrosis:** Cell death.

**Nervous System:** The nervous system of the human body is divided into two interconnected systems: the central nervous system, which is made up of the brain and spinal cord; and the peripheral nervous system. The peripheral nervous system is further divided into the somatic nervous system (made up of peripheral nerve fibers that send sensory information to the central nervous system and motor nerve fibers that project to skeletal muscle) and the autonomic nervous system regulating the functions of our internal organs such as the heart, stomach, and intestines, but also muscles in the skin (around hair follicles and smooth muscles around blood vessels, in the eye (the iris), stomach, intestines, bladder, and heart (cardiac muscle).

**Neurodegenerative:** Marked by or pertaining to neurologic degeneration; deterioration of the structure or function of tissue within the nervous system.

**Neuroimaging:** The production of detail, contrast, and clearness in images of the brain and spinal cord (central nervous system) using computed tomography (CT) scanning, magnetic resonance imaging (MRI), positron emission tomography (PET) scanning, or other imaging techniques to assist in diagnosis, treatment decisions, or research.

**Neuron:** An individual nerve cell.



**Neuropsychologist:** Usually a state-licensed psychologist with expertise in evaluating and treating people who suffer from a brain disorder or organic problem. Neuropsychologists may play a pivotal role in the diagnosis and treatment of mental and emotional problems caused by brain dysfunction.

**Neurotoxin:** A substance that interferes with the functioning of nerve cells (neurons), preventing them from communicating with each other.

**Neurotransmitter:** A specialized substance (such as norepinephrine or acetylcholine) that transfers nerve impulses across spaces between nerve cells (synapses). Neurotransmitters are naturally produced chemicals by which nerve cells communicate. Neurotoxins can also lead to neurodegeneration, causing cell death and brain atrophy.

**Nucleus:** The part of the cell that contains the genetic material (the chromosomes); it is surrounded by a membrane called the nuclear envelope.

**Nurses:** People who care for individuals of all ages, families, groups, and communities, sick or well in all settings. Nursing includes the promotion of health, prevention of illness, and the care of ill, disabled, and dying people. Other important roles nurses assume include patient advocacy, promoting safe environments, conducting research, and participating in shaping health policy, health systems management, and health education.

**Nursing Home:** Also known as skilled nursing facilities and long-term care facilities, nursing homes are licensed by the states to provide skilled, 24-hour nursing and rehabilitation care. Generally, older adults who are bedridden, cannot feed themselves, or who require skilled nursing assistance are appropriate for this level of care.

**Occupational Therapists (OTs):** Primarily focused on the "occupation"—what people do each day. This means helping individuals with PSP, CBD, and MSA find new ways of doing the activities that mean the most to them. OTs help both patients and carepartners to problem solve and create new ways to make routine activities easier and safer.

**Orthostatic Hypotension:** A sudden decrease in blood pressure that occurs when the affected individual sits up or stands. In some cases, it may occur as a side effect of certain medications.

**Oxidative Stress:** A process in which substances called free radicals (see above) build up in the cells as the cells convert nutrients into energy. The free radicals can damage different parts of the cells causing stress to the cell and impairing its functioning. The free radicals can be counteracted by antioxidants (for example, Coenzyme Q10 acts as a scavenger of free radicals).

**Palliative Care:** Specialized medical care for people with serious illness. This type of care is focused on providing relief from the symptoms and stress of a serious illness. The goal is to improve quality of life for both the patient and the family. Palliative care is provided by a specially trained team of doctors, nurses, and other specialists who work together with a patient's other doctors to provide an extra layer of support. It is appropriate at any age and at any stage in a serious illness. This type of care treats pain, depression, shortness of breath, fatigue, constipation, nausea, loss of appetite, difficulty sleeping, anxiety, and any other symptoms that may be causing distress.

**Parkinson's Disease (PD):** A slowly progressive neurodegenerative brain disorder characterized by slowness or poverty of movement (bradykinesia), rigidity, postural instability, and tremor primarily while at rest. Additional characteristic findings include a shuffling, unbalanced manner of walking; forward bending or flexion of the trunk; a fixed or "mask-like" facial expression; weakness of the voice; abnormally small, cramped handwriting (micrographia); depression; or other symptoms and findings. Such abnormalities are thought to result from progressive loss of nerve cells within a certain region of the substantia nigra of the brain and the associated depletion of the neurotransmitter dopamine.



**Pathogenesis:** The origination and development of a disease.

**Peripheral Nervous System:** The peripheral nervous system is that portion of the nervous system outside of the brain and spinal cord (see central nervous system).

**Physiatrist:** A physician with additional postgraduate training in physical medicine and rehabilitation.

**Physical Therapists (PTs):** Healthcare professionals with extensive clinical experience who examine, diagnose, and then prevent or treat conditions that limit the body's ability to move and function in daily life. They can help improve or restore mobility by applying research and proven techniques. Physical therapists provide care for people in a variety of settings, including hospitals, private practices, outpatient clinics, home health agencies, schools, sports and fitness facilities, work settings, and nursing homes.

**Placebo:** The sheer anticipation of new drug, surgery, or procedure that could potentially end the suffering of a patient can provide improvement of symptoms for a period of time. Hence in a clinical trial setting, a substance or procedure that appears to be identical to the treatment under study but that has no effects on the test subject, will be given to one group of study participants. At the end of a study when all data are analyzed, the placebo group are compared to the group who received the new active drug or procedure (called the treatment group). If only the treatment group improved, the drug or procedure had a unique positive effect on patients. If both groups improve to the same degree or do not change, the placebo group proved that the new drug or procedure is not effective.

**Positron Emission Tomography (PET):** An advanced, computerized imaging technique that uses harmless radioactive tracer substances (e.g., modified glucose) to demonstrate chemical and metabolic activities in the brain. PET images then display areas with normal, increased, or reduced activity that can be interpreted by radiologists to identify disease or normal function.

**Postural Instability:** Unsteadiness of gait or standing.

**Progressive Supranuclear Palsy (PSP):** PSP is a neurodegenerative disease (occasionally referred to as Steele-Richardson-Olszewski syndrome, after the three physicians who first described the disease in 1963). The most common first symptom, which occurs, on average, when a person is in her or his 60s, is loss of balance while walking. This may take the form of unexplained falls or of a stiffness and awkwardness in a person's gait that can resemble Parkinson's disease. Sometimes the falls are described by patients as attacks of dizziness. This often prompts the doctor to suspect an inner ear problem or hardening of the arteries supplying the brain. The second most common form of PSP is called PSP-parkinsonism. Its early stages more closely resemble those of Parkinson's disease, with less emphasis on balance problems and behavior changes and more on tremor. These typically have a better early response to antiparkinson drugs than is typical for PSP. PSP-parkinsonism comprises about a quarter of all PSP cases.

Balance difficulty, usually with falls, is the first symptom in a majority of people. Other common early symptoms can be misinterpreted as depression or even as senility. These include forgetfulness and personality changes, such as loss of interest in ordinary pleasurable activities or increased irritability. Less common early symptoms are trouble with eyesight, slurred speech, mild shaking of the hands, and difficulty driving a car. Freezing of gait can be a first and only symptom for several years and difficulty finding words, or aphasia, can be a first and most prominent issue.



**Prevalence:** Refers to the number of people in a given group or population who have a disease.

**Psychiatrist:** A board-certified psychiatrist has at least three years of specialty training after receiving an M.D. degree, and is a medical doctor who specializes in mental disorders. A psychiatrist is uniquely qualified to assess, diagnose, and treat mental and physical conditions.

**Psychologist:** A doctoral-level specialist in psychology, licensed by the state to practice professional psychology (e.g., assess and treat), teach psychology as a scholarly discipline, or conduct research. Psychologists are usually qualified to provide psychotherapy, administer psychological and educational tests, and diagnose and treat mental and psychiatric disorders.

**Pulmonary:** Referring to the lungs.

**Range of Motion (ROM):** The extent of a joint's free movement. The normal ROM of the elbow, for instance, carries the forearm through a half-circle. Passive ROM is tested while the limb is relaxed. Active ROM is movement controlled by the patient.

**Registered Nurse (RN):** A registered nurse (RN) acts upon the order of the physician, providing and directing nursing care services. Additionally, an RN utilizes agency and community resources to fulfill individual patient goals developed in coordination with patients and family.

**Residential Care Facilities for the Elderly (RCFE), also known as Board and Care:** Care in a setting that resembles a person's home. These are small facilities of about six to eight residents that care for older adults who need assistance with Activities of Daily Living (ADLs). Board and Care facilities are licensed by the state's department of social services.

**Respite Care:** The temporary alleviation of the caregiver's responsibility and involvement with the patient. It can be obtained through adult day care or in-home agencies.

**Retirement Home (Facility):** Special complexes of apartments or private homes that provide a supportive environment for seniors, but allow residents to remain somewhat independent. Residents have their own living space (apartment or room) and services provided vary greatly. Examples of services are meals in a central location, laundry facilities, housekeeping, bathing assistance, dressing, and assistance taking medication.

**Rigidity:** Stiffness and resistance to movement. May be a symptom of a neurologic movement disorder such as Parkinson's disease.

**Senior Center:** A community facility for the organization and provision of a broad spectrum of services, including health, social, recreational, and educational services and programs, for older people.

**Sialorrhea:** Excess production of saliva, or increased retention of saliva in the mouth resulting from difficulty swallowing.

**Side Effect:** An effect of a drug that is not the main or intended effect. Side effects may be of no concern, or they may be bothersome or even dangerous, in which case they may limit the upper dose a patient can tolerate. Side effects are also called adverse effects and are closely monitored for through vigorous clinical testing.

**Social Worker:** A professional trained to work with individuals, couples, and families around a wide variety of problems including those arising from medical and mental health concerns. Social workers are also knowledgeable about community resources and how to access them. Social workers are registered, certified, or licensed, depending on a state's accreditation designations.



**Striatum:** See also **Basal Ganglia** for more information. The striatum is an area of the brain that controls movement and balance. It is connected to and receives signals from the substantia nigra.

**Substantia Nigra:** See also **Basal Ganglia** for more information. The substantia nigra is an area in the brain that controls movements and is the origin of most nerve cells that use dopamine as a messenger to communicate with other nerve cells. Degeneration of cells in this region may lead to a neurologic movement disorder such as Parkinson's disease or PSP.

**Tauopathy:** A class of neurodegenerative disorders that has the pathologic aggregation of the tau protein as the common denominator. Examples for tauopathies are PSP, CBD, CTE, and Alzheimer's disease.

**Toxin:** A poisonous substance produced by a plant or animal or by humans as, for example, a by-product of some industrial, extractive, or agricultural activities.

**Tremor:** Rhythmic, involuntary, oscillatory (or to-and-fro) movements of a body part.

**Unified Parkinson's Disease Rating Scale (UPDRS):** A standardized set of questions and physical exam items used worldwide to evaluate Parkinson's disease. It can also be used in other parkinsonian disorders, such as PSP, CBD, and MSA, although those have their own, more specific scales. The UPDRS is useful both for ordinary clinical care and in research studies and is the standard measure accepted by the FDA as a measure of benefit of new treatments for Parkinson's.

**Urinary Tract Infection (UTI):** Such infections are almost always bacterial rather than viral or fungal. They can occur in the kidneys, bladder, or ureter (the tube from kidney to bladder). They can occur in otherwise healthy people but are far more likely in the presence of dehydration, diabetes, and immunosuppression and in any condition impairing the regular and complete emptying of the bladder. Such conditions include PSP, CBD, and MSA as well as benign prostate enlargement and tumors.

**Urgency:** Typically referring to urination: a sensation of having to urinate that appears more suddenly than normal, requiring an urgent trip to the bathroom to prevent incontinence. When incontinence does occur in this way, it is called "urge incontinence." This is the typical form of incontinence in PSP, CBD, and

MSA. The other common form is "stress incontinence," which is set off by a sudden increase in pressure in the abdomen, as during a cough, laugh, or a physical effort. This is typically a result of lax ligaments supporting pelvic organs such as the bladder or uterus.

**Ventilation:** The physical act of breathing. To be distinguished from "respiration," which technically is the complicated biochemical process by which oxygen and sugar are combined to produce energy for the body's use. Ventilation can be impaired slightly in the parkinsonian disorders if the general voluntary muscle stiffness is severe enough to limit movement of the chest wall. In MSA, ventilation control can be disordered, producing various gasping sounds or long pauses in breathing called "apnea."

**Vertebrae:** The bones of the spine. Each forms a ring through which the spinal cord passes. Between one vertebra and the next is a "disk" made mostly of flexible materials. Nerves called "roots" branch off from the spinal cord to emerge between the vertebrae to supply the sensory and voluntary motor functions of the body. These roots, often singly but sometimes in groups, can be compressed by disorders of the vertebrae, such as arthritis, to produce pain and sensory changes.

**Vertical Gaze Palsy:** "Vertical" means up and/or down. "Gaze" means the voluntary movement of the eyes. "Palsy" means weakness, or in this case, reduction of the range of eye movement. Vertical gaze palsy is a useful diagnostic sign of PSP but may not appear until years into the disease course, if at all. It also occurs in milder form in many people with CBD and MSA.

**Vertigo:** The illusion of movement of one's body or environment. This is typically caused by disorders of the inner ear but can also be a symptom of brain disorders like stroke, tumors, and multiple sclerosis. Vertigo is most commonly a spinning sensation, but the perceived movement can also be a rocking or swaying. The sensation of impaired balance caused by PSP, CBD, and MSA is often described by the sufferer as "dizziness" or "vertigo."



**Vestibular System:** The part of the inner ear used for perception of the position and movement of the body in space. Its disorders are called "vestibulopathies" and result in dizziness or vertigo.

**Wellness:** The state of being free of disease, or the measures one takes to achieve such a state. It differs from the tradition term "health" in emphasizing preventive measures such as diet, exercise, and various psychological techniques and encourages traditional and herbal approaches before prescription drugs.

**Whipple Disease:** A very rare, chronic bacterial infection which, when occurring in the brain, can mimic the vertical gaze palsy, balance loss, dementia, and dystonia of PSP. However, Whipple disease of the brain has many neurological features that do not occur in PSP. The infection usually starts in the intestine, where it causes diarrhea and other symptoms. It is curable with antibiotics if it has not progressed too far.

**Whole-Exome Analysis:** A laboratory test that reports the sequence of the genetic code of an individual. The term is also used for a research project that compares large groups of people such as those with and those without a certain disease on the results of such an analysis. The "exome" is (approximately) those sections of the chromosomes that are actually translated into proteins. Exons make up only about 1% of the chromosomes (the genome) but probably are the locations of 85% of defects that are major causes of disease. Whole-exome analysis is available commercially, but its interpretation can be difficult in individual cases because it may be hard to distinguish disease-causing changes from inconsequential, benign variants.

**Wilson's Disease:** A rare disorder of the brain and liver that includes cognitive and movement abnormalities and sometimes other features of PSP. The cause is a gene defect impairing the body's handling of copper. It usually starts during late childhood, but rare cases have started into the 40s. Treatment is very effective if started early. Left untreated, Wilson's disease results in severe disability and death. Most movement disorders specialists order the test for Wilson's disease in anyone with any sort of movement disorder under age 50.

# NOTES

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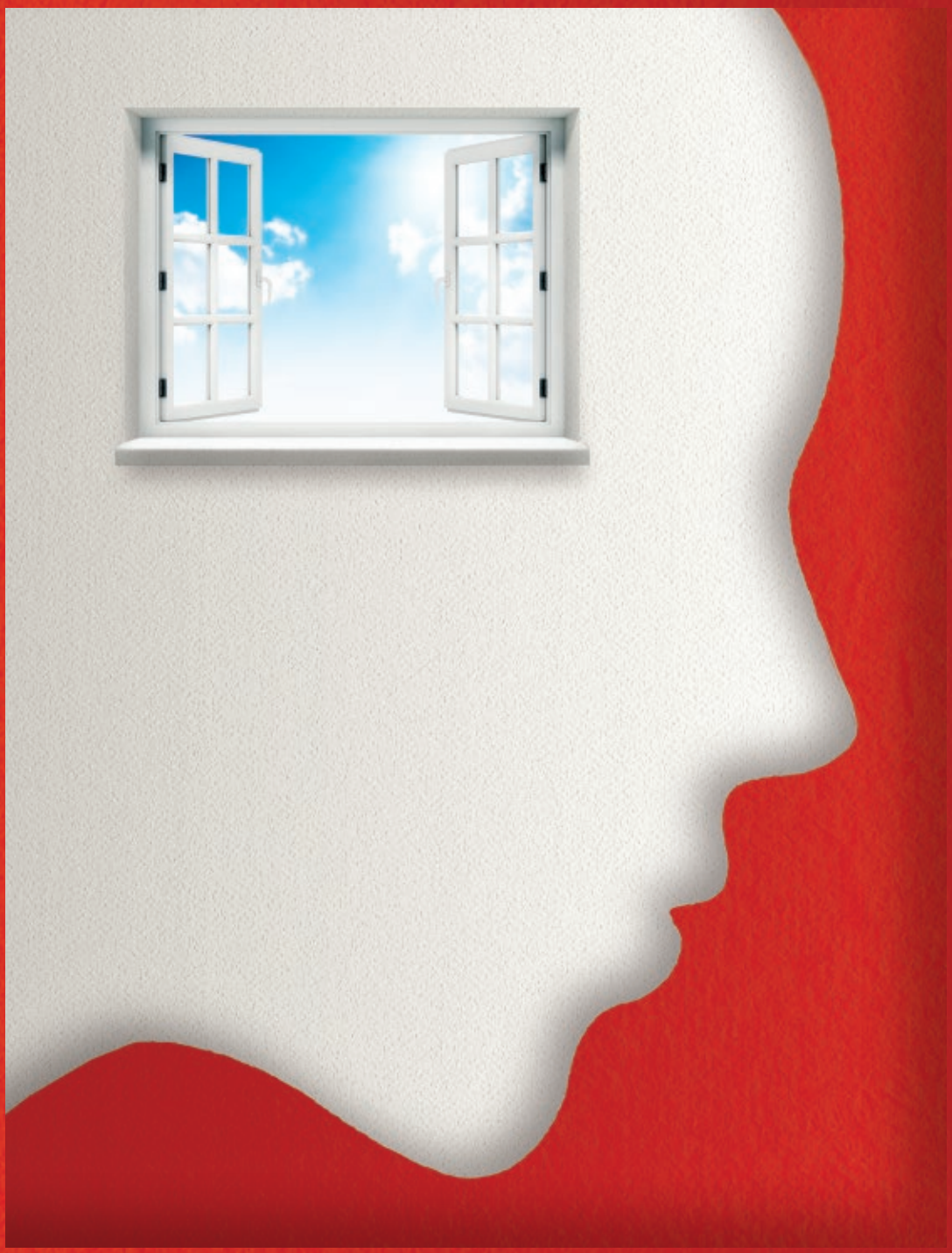

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