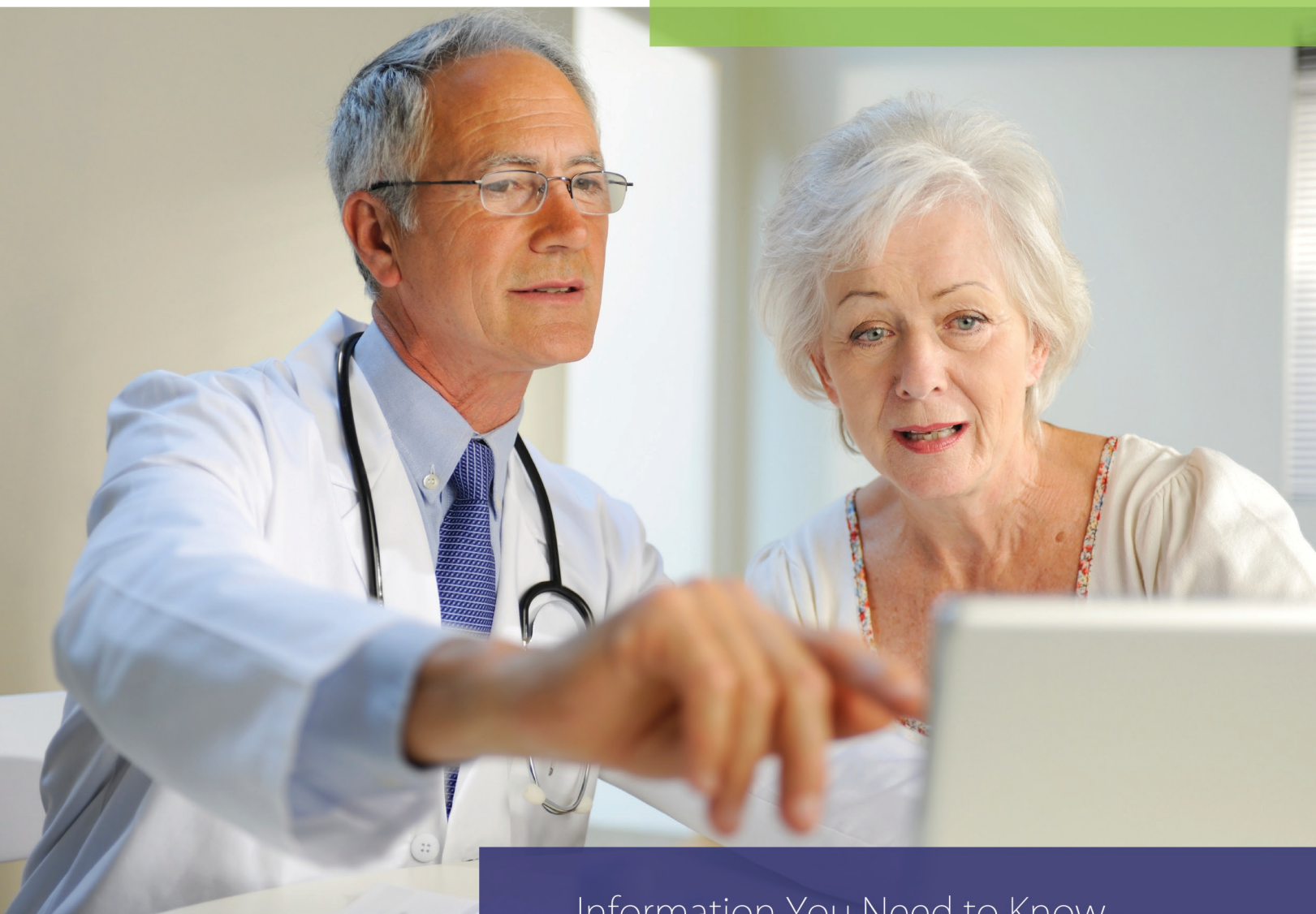




Foundation for PSP | CBD and Related Brain Diseases

*because hope matters*

# A Guide For People Living With PSP, CBD, and Other Atypical Parkinsonian Disorders



Information You Need to Know

Written by Healthcare Professionals

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## *Because Hope Matters*

I'm searching for hope, so is every other patient in my opinion, as well as their family, friends, and potential contributors to PSP research! Hope, I've come to believe, is as vital to our lives as the very oxygen we breathe. If I were to believe that I couldn't exert any level of control over my circumstances, I would have already lost the game! I understand that physicians have to continue to concentrate on understanding the nature of PSP – but, by the same token, everyone connected with PSP has to find ways to better understand the connections between emotions and how the brain and body biologically talk to each other. This may be harder to accomplish than it would appear, since for any physician to effectively impart real hope, they have to believe it themselves.

Each disease and each case of each disease is uncertain in its outcome, and within that uncertainty, we can find hope! The disease and the brain cells affected have not read a textbook or research paper. This is the great paradox of true hope because nothing is absolutely determined. There are more reasons for hope than fear! So we must find ways to bridle fear and give great rein to hope. We all must believe that science will progress and in time new therapies will make PSP curable!

As you can tell, I see hope at the very heart of healing. For those of us who have hope, it may help us live longer. And if it doesn't, it will certainly help us to live a better life!

*Chuck Peebler*

*Charles D. Peebler, Jr. served as a member of the Board of Directors of CurePSP (Foundation for PSP | CBD and Related Brain Diseases.) He passed away on April 18, 2009 at his home in Palm Desert, California. Chuck was in his sixties when he was diagnosed with progressive supranuclear palsy.*

# Message from the President-CEO

A guide is one who leads another. The purpose of this guide is to assist at least two people—the patient and his/her caregiver—along a challenging path. This path, which is just one part of life’s journey, is not without its winding curves and sharp turns. But it is also not a path devoid of hope or meaning.

The people who have put together this guide are individuals who have traveled down this difficult path of progressive supranuclear palsy (or other “atypical Parkinsonian disorder”) with a loved one. While each has, no doubt, experienced some darkness and pain on the road, each has also experienced personal growth and purpose in their own personal journey.

The purpose of this guide is not to offer ultimate answers or reasons for this disease, as helpful as that might be, because at this time the scientific community doesn’t have all the answers. The purpose of this guide is to meet those individuals—both persons with PSP and their caregivers—face-to-face while they are walking down their personal paths of living with PSP.

While the person with the disease is generally thought of as the one who receives the services of the caregiver, the caregiver is also one who receives a gift from the ill person. The gift may come in various forms such as increased patience, a more loving attitude, a clearer understanding of one’s capabilities, problem solving, consoling, improved listening skills, better understanding of one’s emotional and spiritual strengths, and especially hope. Many of those who have provided years of caregiving have gone on to serve others with PSP and to serve as models of strength, service, and wisdom. CurePSP is indebted to those who have served as caregivers and who are now serving as beacons of light to those who are struggling with the disease.

We hope that you will find this guide to be very useful and we hope that in the very near future we may have the opportunity to revise it with the words; we have now found a cure!

*Richard Gordon Zyne, DMin*  
*President – CEO*  
*Foundation for PSP | CBD and Related Brain Diseases*

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# Introduction

When I was caring for my husband, the practical information available from CurePSP was critical and helped us deal with many issues like incontinence and rigidity. We hope this newly revised Guide serves you in that same way.

As Chair of the Board, I often get questions from families about the day-to-day challenges they face. Simple solutions are sometimes available. And when not, it is good to know what else might be recommended. This Guide will lead you through those issues.

I hope you use this Guide as your reference book. You probably won't read it from front to back, although you are welcome to. But it will be there as a great resource when symptoms change and new solutions are needed.

This is the first time we've added MSA (multiple system atrophy) to the Guide. Many of the recommendations for those with PSP and CBD apply to people with MSA who are having similar symptoms. However, some of the issues and needs of people with MSA and their caregivers is unique to that disease, and this Guide addresses those.

To ensure that people dealing with these diseases are getting the best care and support, please share this resource with your full family unit as well as professional caregivers involved in his/her care.

We probably have missed some issues that need to be addressed. So let the CurePSP staff know of other concerns that you'd like them to write about and make available. And please keep an eye on our website at [www.curepsp.org](http://www.curepsp.org) to view new articles and educational information, as they will be put on the website first.

May this educational resource ease your efforts—if even just a little bit.

*Janet Edmunson*  
*Chair, Board of Directors*  
*Foundation for PSP | CBD and Related Brain Diseases*



# PSP: Some Answers

## **What is progressive supranuclear palsy?**

PSP is a brain disease in the category of “neurodegenerative” diseases. Alzheimer’s, Parkinson’s and Lou Gehrig diseases are in the same category – where brain cells cumulatively break down for unclear reasons. PSP affects cells that control walking, balance, mobility, vision, speech, swallowing and behavior. Five to six people per 100,000 have PSP, a number similar to that of Lou Gehrig disease, but only about a third of these have received the correct diagnosis. Symptoms begin, on average, when an individual is in the early 60’s, but may start as early as in the 40’s. It is slightly more common in men than women, but PSP has no known geographical, occupational or racial preference.

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

PSP is rare - only about one percent as common as Parkinson’s disease - and because even when it does occur, it is often misdiagnosed. This is gradually changing. As more doctors become familiar with PSP, it will be diagnosed more readily. No one even realized it existed until 1964, when several patients were first described at a national neurology research convention and the disease received its name. In retrospect, at least 12 cases of PSP had appeared in the medical literature since 1909, but because of its resemblance to Parkinson’s disease, no one had recognized it as a distinct disease until the 1960’s.

The rarity of PSP is not the only reason it is not widely known. PSP is a bit more common than the very well-known disease amyotrophic lateral sclerosis (ALS; called Lou Gehrig disease in the US and motor neuron disease elsewhere). But ALS is easier to diagnose than PSP and affects much younger people in the prime of life. It is unlikely that any of the approximately 4,500 people in the United States who have been diagnosed as having progressive supranuclear palsy had ever heard of the disease before. In fact, many patients with PSP report that their family doctors knew nothing about PSP until a neurologist made the diagnosis. Moreover, the neurologist probably thought the diagnosis was Parkinson’s disease until several years into the illness. For every person with a diagnosis of PSP, there are three with PSP that could be diagnosed if their doctor suspected it and performed the appropriate examination. Recently, more and more has appeared in medical journals to help doctors remedy their unfamiliarity with PSP.

## **What does the name “supranuclear palsy” mean?**

In general, a “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 themselves. These “higher” control areas are what the prefix “supra” in “supranuclear” refers to.

Sometimes complicated disease names are avoided by the use of the name of the physician who discovered the disease. However, for PSP, there were three such physicians and the string of names - Steele, Richardson, and Olszewski (ol-SHEF-skee) – was even less convenient than the descriptive name. “Steele-Richardson-Olszewski syndrome” is rarely used these days as a synonym for PSP.

Incidentally, although Drs. Richardson and Olszewski are deceased, Dr. John C. Steele, who was a neurology resident (i.e., a trainee) when he collaborated in the original description of PSP, still does neurological research and serves as Honorary Chairman of CurePSP.

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

The most common first symptom, occurring, on average, in the 60's, is loss of balance while walking. This may take the form of unexplained falls or of a stiffness and awkwardness in the walk that can resemble Parkinson's disease. Sometimes the falls are described by the person experiencing them as attacks of “dizziness.” This often prompts the doctor to suspect an inner ear problem or hardening of the arteries supplying the brain.

Other common early symptoms are forgetfulness and changes in personality. The latter can take the form of a loss of interest in ordinary pleasurable activities or increased irritability. These mental changes are misinterpreted as depression or even as senility. Less common early symptoms are trouble with eyesight, slurring of speech, and mild shaking of the hands. Difficulty driving a car with several accidents or near misses is common early in the course of PSP. The exact reason for this problem is not clear.

There is a form of PSP, called “PSP-parkinsonism,” in which the early stages more closely resemble those of Parkinson's disease, with less emphasis on balance problems and behavior changes and more on tremor, with a better early response to antiparkinson drugs than is typical for PSP. PSP-parkinsonism comprises about a third of all PSP.

### **What happens next?**

The term “progressive” was included in the name of the disease because, unfortunately, the early symptoms get worse and new symptoms develop sooner or later. After 5 to 6 years, on average, the imbalance and stiffness worsen to make walking very difficult or impossible. If trouble with eyesight was not present early on, it 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 occur eventually in most patients.

### **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 appear, on average, until 3 to 5 years after the walking problem. Because the main difficulty with the eyes is in aiming them properly, reading often becomes difficult. The patient finds it hard to shift down to the beginning of the next line automatically after reaching the end of the first line. This is very different from just needing reading



## PSP: Some Answers—*continued*

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 can interfere with driving a car.

The most common 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. This reduction in vertical eye movement is usually the first clue to the doctor that the diagnosis of the difficulty is PSP. Other conditions, particularly Parkinson's disease and normal aging, can sometimes cause difficulty moving the eyes up. However, PSP is nearly unique in also causing problems moving the eyes down.

In most people with PSP, the difficulty in downward eye movement starts out not as a restriction of the degree of downward movement, but as a slowing of that movement. This can interfere with vision also, but can be very difficult for a physician to detect. Another eye movement problem that starts early in the illness is "square wave jerks." These rapid, involuntary, right-left movements interfere with precisely aiming the eyes at a target.

Yet another eye problem in PSP can be abnormal eyelid movement -- either too much or too little. A few patients experience forceful involuntary closing of the eyes for a few seconds or minutes at a time, called "blepharospasm." Others have difficulty opening the eyes, even though the lids seem to be relaxed, and will try to use the muscles of the forehead, or even the fingers, in an effort to open the eyelids ("apraxia of lid opening"). About 20 percent of patients with PSP eventually develop one of these problems.

Others, on the contrary, have trouble closing the eyes and blink very little. While about 15 to 25 blinks per minute are normal, people with PSP blink, on average, only about 3 or 4 times per minute. This can allow the eyes to become irritated. They often 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 3 or 4 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 features of speech in Parkinson's disease. Most commonly, there is a combination of at least two of these three features in the speech of PSP.

The speech difficulty of PSP, in combination with the forgetfulness, slow (albeit accurate) 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?**

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. Unlike the other difficulties in PSP, this one can sometimes pose a danger for the patient - the danger of food going down the wrong pipe into the breathing passages, termed "aspiration." Usually, difficulty managing thin liquids precedes difficulty with solid food. This is because in PSP, the swallowing muscles have difficulty creating a watertight seal separating the path to the stomach from the path to the lungs. The same is true for the swallowing difficulty of many neurological diseases. For non-neurologic conditions such as stricture of the esophagus, however, 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," in fact, 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. In many people with PSP, there a loss of inhibition or an impulsiveness that the ill person recognizes and promises to resist, but these risky behaviors can be partly involuntary.

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

Although mental confusion in patients with PSP is more apparent than real, most patients do eventually develop some degree of mental impairment. Some are mislabeled as having Alzheimer's disease. This is not very different from the situation in Parkinson's disease. In PSP, the dementia, 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 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 previous familiar environment. Fortunately, these symptoms almost never occur in PSP. Nevertheless, the "frontal" problems of PSP 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.

## PSP: Some Answers—*continued*

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 minutes. Probably the most important aspect of the dementia of PSP is apathy. People with PSP seem to lose interest in their surroundings, again creating the impression of loss of thinking ability and interfering with family interactions.

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

Both PSP and Parkinson's disease cause stiffness, slowness, and clumsiness, a combination called "parkinsonism" (with a small p). This is why early on, PSP may be difficult to distinguish from Parkinson's disease. However, shaking ("tremor"), while prominent in about two-thirds of people with Parkinson's disease, occurs in only about one in twenty people with PSP. A more common type of tremor occurring in PSP is irregular, mild, and present only when the hand is 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 of 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 problem with vision and swallowing and to respond better to drugs for Parkinson's.

The mainstays of drugs for Parkinson's disease are those that enhance replace or mimic a brain chemical called dopamine. Parkinson's responds better to such drugs than does PSP because in PD, deficiency of dopamine is by far the most important abnormality, while 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 medications, all available only by prescription, can help PSP in some cases.

**Sinemet** – this is the brand name for a combination of levodopa and carbidopa. Levodopa is the component that helps the disease symptoms. Carbidopa simply helps prevent the nausea that levodopa alone can cause. When levodopa came along in the late 1960's, it was a revolutionary advance for Parkinson's but, unfortunately, 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. It usually loses its benefit after two or three years, but a few patients with PSP never fully lose their responsiveness to Sinemet.

Some patients with PSP require large dosages, up to 1,500 milligrams of levodopa (with carbidopa) per day, to see an improvement, 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 Sinemet in patients with 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 Sinemet dosages.

Patients with PSP should generally receive the standard Sinemet (or generic levodopa/carbidopa) preparation rather than the controlled-release (Sinemet CR or generic levodopa/carbidopa ER) form. The CR form is absorbed from the intestine into the blood slowly and can be useful for people with Parkinson's disease who respond well to Sinemet but need to prolong the number of hours of benefit from each dose. In PSP, however, such response fluctuations almost never occur. Because Sinemet CR is sometimes absorbed very little or erratically, a poor CR 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.

A new formulation of levodopa-carbidopa is Parcopa, which dissolves under the tongue. For people with PSP who cannot swallow medication safely, this could be useful. Another approach for such patients 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 new formulation of levodopa-carbidopa combines those two drugs with a third drug, entacapone, in the same tablet. This is called Stalevo. The entacapone slows the rate at which dopamine is broken down. It is useful for patients with Parkinson's whose levodopa-carbidopa works well but only for a few hours per dose. This situation rarely, if ever, occurs in PSP.

**Dopamine receptor agonists** – there are three such drugs on the market for Parkinson's – Parlodel (generic name, bromocriptine), Permax (pergolide), Mirapex (pramipexole) and Requip (ropinirole). A fifth drug in this class, rotigotine, will soon be available in the US as a skin patch called Neupro (it is already available in Europe). For PSP, these rarely give any benefit beyond that provided by carbidopa/levodopa. One careful trial of Mirapex showed no benefit at all in PSP.

The main possible side effects of the dopamine receptor agonists are hallucinations and confusion, which can be more troublesome for PSP than for Parkinson's. They can also cause excessive involuntary movements, dizziness and nausea.

**Antidepressants** – another group of drugs that has been of some modest success in PSP are the antidepressant drugs. The anti-PSP benefit of these drugs is not related to their ability to relieve depression. The best antidepressant drug for the movement problems of PSP is probably amitriptyline (brand name, Elavil). It has been used against depression since the early 1960s. The dosage should start at 10 mg once daily, preferably

## PSP: Some Answers—*continued*

at bedtime. It can be increased slowly and taken divided into at least two doses per day. Past 40 mg per day, the likelihood of side effects increases to an unacceptable level for most patients. Amitriptyline is also a good sleep medication for some elderly people and may provide this benefit in PSP if taken at bedtime. One important side effect in some people is constipation. Others are dry mouth, confusion and difficulty urinating (in men). Unfortunately, some patients with PSP find that their balance difficulty worsens on amitriptyline.

**Symmetrel** – this drug (generic name, amantadine) has been used for Parkinson's since the 1960s. Because it affects more than just the dopamine system, it can be effective in PSP even if Sinemet is not. It seems to help the gait disorder more than anything else. 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."

**Drugs for dementia** – Cognex (tacrine), Aricept (donepezil), Reminyl (galantamine) and Exelon (rivastigmine) are drugs that 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. But they do not help the mental difficulties of PSP. A fourth anti-Alzheimer drug, Namenda (memantine) acts on a different brain chemical, glutamate. It works no better for PSP than the others and in addition can cause confusion and agitation in those patients.

**Botox** – a different sort of drug that can be useful for people whose PSP is complicated by blepharospasm is Botox or Myobloc (two types of botulinum toxin). This substance is produced by certain bacteria that can contaminate food. Its poisonous action occurs because it weakens muscles. A very dilute solution of the toxin 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 injection of Botox into the neck muscles can sometimes cause slight weakness of the swallowing muscles, which are nearby. In PSP, where swallowing is already impaired in many patients, caution should be used when considering use of Botox in neck muscles.

**Experimental drugs** – in the past 20 years, many drugs have been tested by researchers in patients with PSP. Some of these are intended to actually slow the long-term brain cell loss and therefore slow the progression of the disease. Perhaps the largest trial was of the drug riluzole, which helps amyotrophic lateral sclerosis modestly but was found not to help PSP.

One possible success story, though the jury is still out, is the dietary supplement coenzyme Q-10. That drug is available without a prescription and helps the body's cells produce energy from sugar and oxygen. It is a normal constituent of the mitochondria, the tiny compartments in our cells where that chemical process occurs. In one

small, brief study in Germany, it was found to improve the signs of PSP, both by neurological examination and by a high-tech measure of energy production by the brain. Two other trials of coenzyme Q-10 in PSP and one large trial in Parkinson's are in progress. The dosage of this supplement needed to give benefit is probably at least 1200 mg per day and perhaps as high as 2400 mg. Even the lower amount costs \$200 per month and is not covered by prescription insurance. Therefore, people with PSP should carefully consider the meager evidence to date for the benefit of coenzyme Q-10 before taking that long-term financial plunge.

Another experimental approach to slowing of disease progression in PSP attempts to prevent the abnormal attachment of phosphate groups to the tau protein. We are still unsure why phosphate groups become abnormally attached to tau in PSP. But in animal "models" such as mice genetically engineered to develop tau tangles in the brain, such drugs do slow that process. These drugs inhibit an enzyme called GSK3-beta.

One such GSK3-beta inhibitor, lithium, has been used in psychiatry for bipolar disorder for many years for reasons unrelated to its action on tau. Unfortunately, patients with PSP were unable to tolerate it because of a variety of side effects such as nausea and dizziness.

Another, called tideglusib (brand name, Zentylor; formerly called Nypta) has completed enrollment and is expected to announce results in early 2012. It appears to be better tolerated than lithium, at least so far. If this trial demonstrates successful slowing of PSP progression, Zentylor will probably be approved by the FDA for commercial sale.

A different approach to slowing the progression of PSP is to enhance the growth and maintenance of the microtubules, ordinarily the job of the tau protein. The drug davunetide, which is taken as a nasal spray, does this. It is being tested at present in 44 sites in Europe and North America and will continue to recruit new patients into the second half of 2011.

Fortunately for PSP sufferers, drug companies have started to act on the realization that a prevention for PSP, where the market is tiny, could work also in Alzheimer's disease, where the market is huge.

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

An operation that may be advised for extreme cases of poor swallowing where choking is a definite risk is the placement of a tube through the skin of the abdomen into the stomach ("gastrostomy" or "percutaneous endoscopic gastrostomy" or "PEG") for feeding purposes. 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 who is 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: aspiration pneumonia; small amounts of aspiration with each swallow; significant weight loss from insufficient feeding; or when a meal requires so much time that the functioning of the household is disrupted.



## PSP: Some Answers—*continued*

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 easily be managed 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 easily be provided 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 removed.

One potential downside of tube feedings for some patients is a loss of the feeling of "wholeness" or humanity. The issue of how much additional quality will be introduced into the patient's life must be considered carefully. The family, physician and if possible, the patient must all voice their opinions. Another downside is that some patients who are 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 may be useful 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 payors 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 output of the basal ganglia (the group of nuclei that control movement) to the rest of the brain is overactive in Parkinson's. The operations dampen down this overactivity. The main operations for this purpose are pallidotomy, which is rarely performed nowadays, and subthalamic nucleus stimulation, which is the most common Parkinson's operation at present. In PSP, the area of the basal ganglia from which the output comes is itself damaged, so its activity is already dampened down. The operations would only make things worse.

However, there are trials now in progress to test stimulation of an area of the brain serving balance, the "pedunculo-pontine nucleus" (PPN), in people with PSP. The PPN is in the brainstem, which is an area tightly packed with critical circuitry. The procedure seems to be acceptably safe, but the overall improvement in the patients is still undetermined. The procedure does seem to help the balance problem in some patients with Parkinson's disease.

The other category of operation for Parkinson's attempts to replace the lost dopamine-producing brain cells. The reason this is unlikely to work for PSP is that while in Parkinson's, most of the movement problem is caused by loss of the main dopamine-producing nucleus, the substantia nigra, while in PSP, the movement problems are caused by loss of many other nuclei in addition. Many of those other nuclei receive their input from the substantia nigra, so replacing only the first "link in the chain" will not help much. It would be impractical to replace cells in all of the nuclei involved in PSP – it would require too much trauma to the brain.

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

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 will power 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 conscious control and that nagging and shouting usually just 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 during the "bad" times.

Walking aids are often important for patients with PSP. Because of the tendency to fall backwards, if a walker is required it should be weighted in front with sandbags over the lower rung. 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. Such people can fall if they move their body forward before the foot moves. In these cases, a smooth sole 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 in 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 glasses called prisms are sometimes prescribed for people with PSP. These are sometimes worth trying, but are usually 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. If this additional problem exists, special glasses would not help.

Formal physical therapy is of no proven benefit in PSP, but certain exercises done in the home by oneself on a regular schedule can keep the joints limber. Exercise also has a clear psychological benefit that improves the sense of well-being of 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.

## PSP: Some Answers—*continued*

### **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”, is also affected in Parkinson’s disease and damage to it accounts for the symptoms that PSP and Parkinson’s have in common. However, several important areas are affected in PSP that are normal in Parkinson’s (and vice-versa). Moreover, under the microscope, the appearance of the damaged brain cells in PSP is quite different from those in Parkinson’s and resembles, rather, the degeneration in Alzheimer’s disease. However, the location of the damaged cells is quite different in PSP and Alzheimer’s. Furthermore, in PSP there are no amyloid plaques, 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 a normal protein called “tau.” These clumps of tau are called “neurofibrillary tangles.” The normal function of tau is to help support the internal “skeleton” of the brain cells whose long extensions make contact with other brain cells. 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 produced in excess. If it is damaged, the nature of that damage could be the excessive attachment of phosphate (see above). Or, the excessive phosphates observed could simply be the brain’s normal response to minimize the effects of tau protein that is misbehaving for some other reason.

A clue to what is going wrong with tau protein is that most of the tau protein in the neurofibrillary tangles of PSP is of one type called “four-repeat” tau. In the normal brain cells, there are equal amounts of four-repeat and three-repeat tau. The “repeat” number refers to the number of copies of the part of the protein that binds it to another component of the cell’s internal skeleton, the microtubules. So in PSP, the problem may be that too much four-repeat tau is made, or that too little three-repeat tau is made, the result being clumps of four-repeat tau.

It is looking more likely that at least one important part of the cause of the misbehavior of the tau protein is some sort of genetic defect in or near the tau protein’s gene on chromosome 17. But the nature of that defect remains elusive.

### **Is PSP genetic?**

PSP only very rarely runs in families. Fewer than 1 in 100 people with PSP knows of even one other family member with PSP. However two different variants in the gene on chromosome 17 that encodes the tau protein are more common in PSP than in the rest of the population. One of the variants is called the “H1 haplotype.” About 95% of people with PSP have this variant on both of their copies of chromosome 17, while this is true for only about 60% of the rest of us. So clearly, the H1 haplotype is (nearly) necessary but far from sufficient to cause

the disease. There is evidence that what this variant is doing wrong is directing the brain cells to produce too much tau protein. The evidence suggests the tau starts to aggregate into clumps and that the damage is caused by an early stage of these that is still too small to be seen through the microscope. The larger, mature neurofibrillary tangles may simply be the result of the cell's attempts to pack the small, toxic, clumps of tau into a harmless mass that cannot interact with normal cell constituents.

The other PSP-related variant in the tau gene was discovered in 2010 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. This will be the subject of intense research over the next few years.

The same Peebler Genetics Program of CurePSP has discovered several other genetic variants that are more common in people with PSP than in those without PSP. Those genes make enzymes that help control the brain's system for disposing of mis-folded proteins. This could explain why the clumps of tau protein form. Other genes newly implicated in PSP help 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.

The insights from this new genetic study 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.

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

There is also evidence that chemicals in the environment or diet may contribute to the cause of PSP. Surveys of PSP patients have hinted at a predilection for rural living and, on average, lesser educational attainment in people with PSP. This suggests that part of the cause of PSP is certain occupational factors exposing people to different chemicals than are encountered by non-rural people or those with more sedentary, office-bound occupations.

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 are people elsewhere. A questionnaire survey on Guadeloupe revealed that people with PSP-like illnesses there 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, if any, foods in the Western diet may contain similar toxins. Research on that question is under way.

## PSP: Some Answers—*continued*

### **What research is being done?**

Research is proceeding in the areas of genetics, epidemiology, treatment trials, and molecular studies. CurePSP's genetics program has searched the entire genome for genes related to PSP and corticobasal degeneration (CBD). The results, which showed at least four genes not previously suspected of a relation to these diseases, will be published in 2011. In addition to working with researchers at the University of Louisville to study environmental, occupational, and genetic risk factors associated with PSP, CurePSP and other funding agencies are presently sponsoring exciting and promising research into many aspects of PSP, including ways to prevent the brain cells from dying and treatment of the symptoms with drugs, magnetic stimulation, and deep-brain electrical stimulation. Two drug companies are presently running large, formal trials of new medications designed to slow progression of PSP.

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

Of course, CurePSP welcomes donations to its research grants program. Since its inception in 1997, it has provided over \$9 million to researchers' institutions to support their work. Some of the support has gone to senior researchers with excellent track records of productivity and some has gone to junior scientists with original ideas and first-rate training. CurePSP does not restrict its grants to any country or continent. It 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.

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, too.

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

### **Should I make arrangements to donate my brain after death?**

Another very important way to help PSP research is to make arrangements to donate your brain after death. CurePSP sponsors the Eloise H. Troxel Memorial Brain Bank located at the Mayo Clinic in Jacksonville, FL. Brains donated there are stored and used only for research in PSP by legitimate researchers after their proposals are examined and approved by the CurePSP Scientific Advisory Board. Donating to a brain bank does not interfere with funeral arrangements and costs the family a few hundred dollars for expenses of brain removal and transporta-

tion. The family will receive at no charge a full diagnostic report from the Mayo Clinic pathologist, Dennis W. Dickson, MD, who is one of the world's foremost authorities on PSP and related disorders. Further information is available from the CurePSP. There are many other brain banks throughout the country, generally located at major university hospitals.

### **Should I join some sort of support group?**

The value of membership in a group of other people with the same problem is tremendous. 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 PD 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. Contact CurePSP for help.

A major goal of CurePSP is to increase awareness of PSP among the public and the medical profession in order allow its correct diagnosis. If, as we suspect, PSP proves to be much more common than has been assumed, improved diagnosis may allow local support groups to flourish, will foster the growth of CurePSP, and will draw the attention of more researchers to finding the cause and cure of this unique and puzzling illness.

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# CBD: Some Answers

*Synonyms:* corticobasal syndrome (CBS), corticobasal ganglionic degeneration (CBGD), cortical-basal ganglionic degeneration

## **What is corticobasal degeneration?**

Corticobasal degeneration is a progressive neurological disorder characterized by nerve cell loss or deterioration and atrophy (shrinkage) of multiple areas of the brain. The cortex, or outer layer of the brain, is severely affected, especially the fronto-parietal regions, located near the center-top of the head. Other, deeper brain regions are also affected, including parts of the basal ganglia, hence the name “corticobasal” degeneration. The combined loss of brain tissue in all these areas causes the symptoms and findings seen in people with CBD. CBD was first described in 1968 by Rebeiz and colleagues.

Historically, CBD patients have been diagnosed on the basis of movement problems which sometimes appear similar to Parkinson’s disease (PD). Unlike PD, however, CBD patients typically do not respond significantly to PD medicines, such as levodopa/carbidopa (Sinemet). Also, many symptoms of CBD are not found in PD patients. For this reason CBD is often referred to as a ‘Parkinson’s-plus’ syndrome.

The Parkinsonism is generally an asymmetric akinetic rigid syndrome, unresponsive to levodopa, similar to that of multiple system atrophy and PSP. Eye movement abnormalities are common, as in PSP, and a supranuclear gaze palsy can be seen as in PSP. Given the genetic similarities between CBD and PSP, it seems possible that they are simply two “faces” of the same disease.

## **What causes degeneration of brain tissue in CBD?**

Unfortunately, the cause of CBD is entirely unknown. There is currently no strong evidence to suggest CBD is an inherited disease, and no other risk factors, such as toxins or infections, have been identified. Studies of brain tissue of individuals with CBD show certain characteristic cell changes. Similar, although not identical, changes are observed in two other neuro-degenerative diseases, Pick’s disease and progressive supranuclear palsy. These changes, involving a brain protein called tau, have provided researchers some initial clues in their search for the causes of CBD. While CBD patients have normal saccadic velocity, this may be an artifact of case definition. If PSP and CBD share the same pathologic mechanism, they may simply be two different presentations of the same disease.

Tau protein accumulates in this disorder and it may be related to a mutation in the tau gene. Tau is a microtubule-binding protein that is normally abundant in neurons. Other “tauopathies” include Alzheimer’s disease, Picks disease, frontotemporal dementia and parkinsonism, ALS-parkinson dementia complex of Guam, and progressive supranuclear palsy (PSP). CBD shares the same tau haplotype as do PSP patients suggesting that both CBD and PSP share the same genetic background and possibly the same pathologic mechanism.

**What are the symptoms of CBD?**

Symptoms of CBD usually begin after age 60. The initial symptoms of CBD are often stiffness, shakiness, jerkiness, slowness, and clumsiness, in either the upper or lower extremities. Other initial symptoms may include dysphasia (difficulty with speech generation), dysarthria (difficulty with articulation), difficulty controlling the muscles of the face and mouth, or walking and balance difficulties. Symptoms usually begin on one side of the body, and spread gradually to the other. Some patients (probably more than commonly recognized in the past) may have memory or behavioral problems as the earliest or presenting symptoms.

CBD is a progressive disease, meaning the symptoms worsen over time. Over the course of one to several years, most people with CBD gradually worsen, with symptoms progressing to involve upper and lower extremities and other body regions. Symptoms of advanced CBD include:

- Parkinsonism (rigidity, slow movements, postural instability)
- Tremor
- Myoclonus (sudden, brief jerky movements)
- Dystonia, including blepharospasm
- Speech difficulty
- Acalculia (difficulty carrying out simple calculations, such as adding and subtracting)
- Personality changes
- Inappropriate behavior
- Repetitive and/or compulsive activities
- Short-term memory problems (such as repeating questions or misplacing objects)
- Sensory loss
- “Alien hand/limb” phenomenon (difficulty controlling the movements of a limb, which seems to undertake movements on its own)

There is mild-to-moderate cognitive impairment (memory loss, difficulty planning or executing unrehearsed movements, dementia). Patients with CBD who present with cognitive difficulties are usually initially diagnosed with frontotemporal dementia or Alzheimer’s disease.

It is only after a patient develops movement symptoms that the diagnosis of CBD is entertained. Occasionally, a diagnosis of CBD is not apparent until a patient’s brain is examined at autopsy.

Progressive difficulty with language is a common cognitive complaint in CBD. This most commonly involves difficulty with expression of language, such as word finding difficulty or naming problems. Reading, writing and simple arithmetic may also be impaired.

## CBD: Some Answers—continued

### **How is CBD diagnosed?**

Early in the disease course, it is often difficult to distinguish CBD from similar neurodegenerative diseases. Diagnosis of CBD involves a careful neurological exam, combined with one or more types of laboratory evaluations. Electro-physiological studies, including an EEG (electroencephalogram), may show changes in brain function over time that are consistent with the neurodegeneration. CT or MRI scans can also be used in this way, providing images of asymmetric atrophy of the fronto-parietal regions of the brain's cortex, the regions most frequently involved in the disease. Imaging that shows the function, as opposed to the structure, of the regions of the brain would show asymmetric abnormalities, especially in the frontal and parietal regions. The most commonly available such imaging procedures are SPECT (single photon emission computed tomography) and PET (positron emission tomography) scans.

It is also important for the neurologist to look for, and rule out, other explanations for asymmetric loss of brain tissue. The most common such mimic that requires very different treatment is stroke.

Unfortunately, there are no drugs or other therapies that can slow the progress of the disease, and very few that offer symptomatic relief. Tremor and myoclonus (sudden, brief jerky movements) may be controlled somewhat with drugs such as clonazepam. Baclofen may help reduce rigidity somewhat. Levodopa and other dopaminergic drugs used in Parkinson's disease are rarely beneficial, but may help some CBD patients.

Physical therapy exercises may be useful to maintain range of motion of stiff joints. This may prevent pain and contracture (muscle shortening) and help maintain mobility. Occupational therapy may be used to design adaptive equipment that supports the activities of daily living, thus helping to maintain more functional independence. Speech therapy is used to improve articulation and volume.

### **What is the usual course of CBD?**

A person with CBD will usually become immobile due to rigidity within 5 years of symptom onset and may have to make a decision about a feeding tube at some point before that. Most often, within 10 years of onset, pneumonia or other bacterial infections may lead to life-threatening complications.

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*Courtesy of wemove.org*

# MSA: Some Answers

## **What is multiple system atrophy?**

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.

## **How common is it?**

MSA is rare, with about 13,000 sufferers in the US, most of whom have not yet received a correct diagnosis. This compares with about 20,000 with progressive supranuclear palsy (a similar condition), about 500,000 with Parkinson’s disease and 5 million with Alzheimer’s disease.

New cases of MSA arise in about 6 persons per million population per year. This means that in the US, about 5 people are newly diagnosed with MSA each day.

## **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 causes 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 (abbreviated 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 events that result in damage to certain brain cells are called the “pathogenesis” of a disease. In MSA, this process centers around the accumulation of alpha-synuclein protein into clumps called “glial cytoplasmic inclusions.” We do not know why the protein clumps, but we do know that the same protein clumps up in a different set of brain cells in Parkinson’s disease. (In most other neurodegenerative diseases, other proteins clump up in different sets of cells.) MSA, like many other neurodegenerative diseases, includes abnormalities

## MSA: Some Answers—*continued*

in disposal of abnormal or excessive protein and in the having an excessive amounts of damaging chemicals called “oxidation products.”

The cells that are involved first in MSA appear to be the “glia,” the cells that are thought to be electrically inactive and provide nutritional and physical support for the electrically active cells, the “neurons.” This is different from most other neurodegenerative diseases, where the problem seems to originate in the neurons.

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

There are three types that grade into one another. Cases emphasizing slowness and stiffness were once called “striatonigral degeneration”, or MSA-P. Cases emphasizing cerebellar problems were called “sporadic olivopontocerebellar atrophy”, or MSA-C. Cases emphasizing autonomic problems were once called “Shy-Drager syndrome.” The commonality consists of a type of protein that forms clumps in the same types of cells in the three. The differences among the three types of MSA were dictated by which part of the brain or spinal cord that was most involved. These terms were discarded in 1989, when all three were found to be variants of the one disease, which then received its current name. Most people with MSA have elements of all three types, but most have one or the other as their first and most important element.

### **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 things often decline to some extent in people with MSA.

Some 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 can also have difficulty 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 with the type of 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 head bent forward to an extreme degree, a condition called “antecollis.” This 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 that is annoying but almost never large enough to interfere with normal movement. This 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 occurs when the limb is in use rather than at rest as in PD. It may respond to medication, but usually is too mild to require treatment.

### **How fast does MSA progress?**

Unfortunately, MSA progresses rapidly. It starts at an average age of 54 and within only 2 or 3 years produces important disability with regard to walking and balance. Most people with MSA are wheelchair-bound by the 4th year and eventually become bedbound because of general stiffness. This state increases the risk of dangerous complications such as pneumonia, bladder infections, skin breakdown and blood clots in the legs that float up to the lungs. The average person with MSA lives only about 7 years after the initial symptoms start, but some live much longer than that average.

### **How is MSA treated?**

There is still no cure for MSA nor any way known to slow the progression of its underlying brain cell loss. However, there are many ways to lessen the severity of the symptoms and to improve both the quality and quantity of life. These are called “palliative” measures.

**Antiparkinson treatment:** Many people with MSA who have stiffness of limbs and slowness of movement find that carbidopa/levodopa, the main drug used for Parkinson’s disease, can help those symptoms. The duration and intensity of the drug’s effect is usually less than in Parkinson’s.

**Low blood pressure:** There are many ways to treat low blood pressure. If one is taking drugs or limiting salt intake to treat what once was high blood pressure, these measures could be reduced, but only under the supervision of a physician. Drugs that can increase the blood pressure include fludrocortisone (Florinef), midodrine (Pro-Amatine) and pyridostigmine (Mestinon). There are a handful of other drugs that are often worth a try. Non-drug measures include increasing the salt and fluid intake (if there are no heart or kidney problems that would make that risky), elevating the head of the bed by putting six-inch blocks under the legs at that end and using pressure stockings.

**Bladder problems:** The need to urinate frequently can be reduced by drugs that inhibit the muscle that empties the bladder. These are called “peripherally acting anticholinergics” and are widely advertised in the popular media.

**Constipation:** This symptom in MSA is treated as in any other setting. It is best to start with a stool softener (docusate; Colace) or a bulk-forming agent (Metamucil).



## MSA: Some Answers—continued

**Swallowing problem:** This is best treated by changing one's habits regarding choice of foods and food textures, chewing technique and swallowing technique. This is best assessed by a trained speech/swallowing pathologist, often guided by an x-ray video of the person swallowing a range of food textures, called a "modified barium swallow."

For severe swallowing difficulties that present a high risk of "aspiration" (food going down the wrong pipe into the lungs), a soft rubber tube can be placed through the skin of the abdomen directly into the stomach ("percutaneous endoscopic gastrostomy" or "PEG"). But this step should not be undertaken lightly. If such a technique becomes necessary to prevent aspiration, a careful decision should be reached by the patient, family, physician and other available advisors. They may decide that while the PEG may prolong life, the quality of life that the other aspects of MSA allow at that point for that individual could make the PEG a poor choice.

### **What research is being done on MSA?**

Fortunately for MSA sufferers, the protein that clumps abnormally in the brain cells is the same one that clumps in Parkinson's disease. This means that much of the research that sheds light on the basic cellular abnormality of Parkinson's does the same for MSA. But there is plenty of research that directly addresses MSA. The number of mainstream biomedical journal articles that prominently mention MSA increased from 113 in 1990 to 251 in 2000 and to 661 in 2009. Furthermore, the basic problems in many of the neurodegenerative disorders are turning out to be very similar. That means that advances against Alzheimer's disease, Lou Gehrig disease, to name two well-funded examples, are useful in the fight against MSA. When the fine day comes that one neurodegenerative disease falls, the others will not be far behind.

### **How can I help with research efforts on 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 (NIH): [www.clinicaltrials.gov](http://www.clinicaltrials.gov). Simply enter "multiple system atrophy" into the search box, and a number of clinical trials focusing on MSA will appear. Participants in clinical trials not only may benefit from a new treatment that is not generally available, they also often receive detailed care and attention that is not part of an average routine, even at excellent medical centers. They also get the satisfaction of helping the fight against their illness. Other trials may look for new genetic or environmental contributors to the cause of other related diseases. Discovering these could also point to potential preventative measures or treatments in MSA.

More information on research is available from CurePSP ([www.psp.org](http://www.psp.org) or 1-800-457-4777), which includes MSA among the disorders for which it provides education and support for patients and their families.

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# Building an “Advisory Team”

Managing PSP, CBD and other related atypical Parkinsonian disorders is a continually evolving challenge. While it is a degenerative brain disease, the rate of the degeneration varies from person to person. It is important to remain independent and functional for as long as possible. You will need a supportive, patient-oriented healthcare team who can work with the special needs of a person with these diseases. Your “advisory team” should include the following:

## **Neurologist**

A neurologist is a specialist in diseases of the nervous system. He/she will outline possible treatment options available, help you to preserve a positive self-image, work with you in identifying specific concerns and needs, and refer you to therapists to help you find solutions to those needs.

## **Physical Therapist**

Physical therapists (PTs) provide services that help maintain function, improve mobility, relieve pain and prevent or limit permanent physical disabilities. PTs examine a patient’s medical history and test and measure the patient’s strength, range of motion, balance and coordination, posture, muscle performance, respiration and motor function. They also determine a patient’s ability to be independent and develop treatment plans describing a treatment strategy, its purpose and its anticipated outcome.

## **Speech-Language Pathologist**

As with any progressive neurological condition, early intervention is the key to maintaining or increasing communicative effectiveness and swallow function. As soon as an individual with PSP, CBD or other related atypical Parkinsonian disorders or a caregiver notices changes in speech and swallowing, it is time to seek referral to a 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 mechanism highly functional than it is to rebuild what may be lost. However, it is never too late to see an SLP and get help to restore functions regarding speech and swallowing.

## **Occupational Therapist**

Occupational therapists (OTs) can assist you in managing physical, functional, visual and cognitive changes related to these disorders. They are trained to work from a person-centered perspective focused on building a supportive social and environmental context to help you fight the disease. Your OT may address functional vision, physical skills, home safety, community independence, and cognition, always working collaboratively with you and your family to build the best mix of supports to help you.

## Building an “Advisory Team” *–continued*

### **Other Team Members**

You may also be referred to a social worker whose tasks include both practical and emotional support to help people with PSP, CBD and other related atypical Parkinsonian disorders and their families. The social worker also offers information about available community resources and acts as an advocate to assist people in accessing these resources. He/she also provides information on financial and legal issues and assistance in accessing these resources. You may also call on a pastoral care worker, priest, minister, rabbi or chaplain to assist in making decisions, facilitate spiritual reflection and offer support during emotional and physical crises. There may be other healthcare professionals on your team, such as a wheelchair seating expert or a psychologist, but the above mentioned professionals are the core members of the “advisory team.”

## 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 stretching 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 in the bladder excessively, 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 fluid, 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 oxybutinin (Ditropan) and tolterodine (Detrol). Both drugs can also cause dry mouth and constipation. The latter is often already a problem in PSP and can be treated by fluids, fiber and 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. 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. 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 and for 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 and gives them a surface in the bladder on which to grow and cause infection.

## PSP and Constipation

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, multiple system atrophy, corticobasal degeneration and Parkinson's disease.

### **What is Constipation?**

Constipation is defined by both frequency and quality of bowel movements. There is no "normal" bowel movement frequency that 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. Reduction in bowel movement frequency or even constipation may precede the limb movement problems in such disorders by many years. There is even a respectable theory that

## PSP and Constipation—*continued*

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.

Food is primarily liquefied by strong digestive juices in the stomach so 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, there are thickening products readily available at your pharmacy. Fluid intake can be enhanced using treats like jello, 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 comprised 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 healthy 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. 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 oz with 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 and 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 actually 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 other health care 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.



# Pressure Sores

Pressure sores, sometimes called bedsores, decubitis ulcers or pressure ulcers, are red areas or sores on the skin. A pressure sore is a skin and underlying tissue injury that is usually caused when a small patch of skin suffers prolonged contact and pressure. Pressure occurs if a person lies or sits in one position too long. Unrelieved pressure squeezes the blood vessels that supply nutrients and oxygen to the skin. When skin is deprived of nutrients and oxygen for too long, tissue can die and a pressure sore can form. This can occur in less than two hours. Pressure sores appear most frequently on thighs, buttocks, the coccyx (tailbone), heels or any part of the body that is in constant contact with another object, such as a bed or wheelchair. Because pressure sores often begin as a blistered or reddened area on the skin, the sore will often develop unnoticed. These sores infect easily, and if left untreated, can rapidly become a large hole reaching to the bone and carrying infection with them.

## **Early treatment of a pressure sore**

When a pressure sore is discovered in an early stage, the first step is to eliminate all pressure from the area. The sore must be dried and kept dry. Periodically clean the area with hydrogen peroxide or a saline (salt and water) solution. Expose the clean sore to a 100-watt electric light bulb held two feet away from the sore for about 10 minutes. After the sore dries, dust it with cornstarch. Applying Benzoin can toughen the healthy skin around the wound. Using a polyurethane film dressing that creates an artificial skin can be useful for a minor skin pressure sore.

## **Treatment for more advanced sores**

Trimming away tissue that is dead (debridement) is required when treating advanced sores. Antibiotics are prescribed when an infection is present. An infection can develop rapidly without debridement. After debridement procedures, dressings are changed frequently. Sometimes more involved surgery is required and even possible, skin grafting to protect the tissue and allow it to heal. Lack of proper care for an advanced sore can result in an ulcer, which can become life-threatening if permitted to advance to a larger size.

## **Prevention tips:**

- Skin should be inspected every day to find and correct problems before a pressure ulcer forms; pay particular attention to the bony prominences
- Clean skin with a mild soap, warm water and minimal friction
- Move or change positions every two hours in bed and every hour if in a chair
- Alternating pressure mattresses, foam wedges and pads helps reduce friction; consult your health care provider about the use of a special mattress or cushion that contains air, gel, foam or water; the use of “doughnut” cushions and egg crates is discouraged
- Placing sheepskin under vulnerable areas will cushion the entire body; sheepskin reduces pressure and friction and absorbs moisture keeping the patient dry

- Reduce friction or rubbing when moving by lifting rather than dragging
- Do not use a trapeze to lift if possible
- Use moisture absorbent underpads or briefs if necessary to reduce moisture exposure from urine and perspiration; an ointment or cream to protect skin from urine or stool drainage may be helpful
- Raise the head of the bed as little as possible
- Check with your health care provider about the use of lotions and medicines; routinely use a good quality skin cream; ask your physician about products such as Barri Care, Aquaphor, Lac-Hydrin, Care Crème and Lypha Zome
- Good nutrition plays an important role in the prevention of and recovery from a pressure sore

## Dry Eye Syndrome

### **What is Dry Eye Syndrome?**

Many patients develop dry eye syndrome. Left unattended, it can cause damage and roughness to the surface of the eyeball. The artificial tears are not the same as common eye drops. If dry eyes are a problem, it is always best to consult an eye professional for advice and treatment. Dry eye syndrome (also referred to as keratitis sicca) is a common condition thought to affect approximately 60 million Americans. In some instances, it is characterized by dry, irritated eyes due to a lack of lubricating tears caused by an imbalance between tear production and tear volume drainage via the nasolacrimal ducts (NLD). It can also result from excessively watery eyes due to tears lacking the proper balance of mucous, water and oil to coat the eyes properly. The tear film is made up of a mucous 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 is 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 due to smoke, smog, irritants like onions, or when the eyes lack proper protection from lubricating tears.

### **What are the 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 pebble of sand, in the eye.

## Dry Eye Syndrome—continued

### How is Dry Eye Syndrome Diagnosed and Treated?

Dry eye syndrome can usually be diagnosed by an ophthalmologist with the patient's history alone, though the exam confirms the diagnosis. On exam, the patient typically shows a reduced tear volume and rapid tear break-up time (the time for dry spots to occur on the cornea). Placement 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."

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 ointment in the eye on a regular basis, perhaps four times a day. If the condition is to be 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 with your eye doctor. Excessive or prolonged use of artificial tears can disrupt the eyes' 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. Due to hard tap water in most areas, however, distilled water is usually required. Hard water in many areas will create an airborne mineral dust, which may make the humidifier less effective for its intended purpose.

## Visual Issues and PSP

*PSP is a degenerative disorder of the brain with no known cause. The disorder was named as such because of characteristic eye movement abnormalities that have become the signature of the disorder.*

### Introduction

PSP affects the control centers in the brain that generate eye movements. As a result, 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, and eventually total paralysis of eye movements ensues. Typically, upward and downward gaze is more impaired than side-to-side gaze.

### Visual Disturbances of PSP

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. It is not unusual to hear from family members that patients unknowingly leave food on their plates. The inability to look down impairs the ability to go down stairs or 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. 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 see ophthalmologists with 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.

Another frequent cause of reading difficulty is convergence insufficiency and paralysis. Besides not being able to look down, the patients can't converge and focus their eyes 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 in frustration.

Eyelid functions can also be affected by PSP. Patients 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 called Apraxia of Eyelid Opening. The patient has preserved ability to blink but from time to time he/she can't open the eyelids after blinking. Often the person is forced to use his fingers in order to manually lift the eyelids.

Dry eyes are a frequent complaint of PSP patients. However, this is not the direct result of the disorder. PSP often starts during 6th and 7th decades, when, as a result of physiologic aging, tear production declines. But reduced rate of blink can augment the dry eye syndrome.

PSP does not lead to blindness. Any patient with PSP, who begins to lose his /her eyesight, should see an Eye specialist before attributing the loss of vision to PSP.

### **Treatments**

There are several ways of managing the eye symptoms. Unfortunately, however, there is no satisfactory treatment of paralysis of eye movements. Eye exercises are futile.

Those patients who can't look down and read through their bifocals are recommended to hold the reading material higher to the level of the eyes, 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.

Patients who have convergence paralysis are best helped by special prism glasses. 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 Botox injections. The results are good. Botox does not cure the condition but improves it for 3-4 months. Repeated injections are necessary to keep the condition under control. An eyelid crutch is an alternative to Botox, but not well-tolerated by all patients. Lid crutch-

## Visual Issues and PSP—*continued*

es are metallic bars that are attached to the rim of the glasses. The position of the bar is so arranged that it sits right at the level of the eyelid and mechanically lifts the lid. Some optical shops carry these crutches and fit the patients. Taping the eyelids to the forehead is another simple method to keep the lids open. Finally, in the worst case scenario, surgery may be necessary to remedy the 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.

Dry eye are easily treated by liberal use of lubrication eye drops that are available over the counter.

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## Good Oral Hygiene

Persons diagnosed with PSP/CBD/MSA and their caregivers face increasing challenges as the disease progresses. The caregiver's tasks continue to increase, and many times, helping the care receiver to maintain good oral hygiene becomes a low priority. Yet, maintaining good oral hygiene will help prevent future problems with infection and pain. It will also promote the care receiver's integrity and comfort. There is another important reason for the person with PSP to maintain good oral hygiene. As the disease progresses, swallowing problems develop because of throat muscle weakness and coordination. Problems with swallowing thin liquids usually occur before difficulty with solid food.

These weakened throat muscles have difficulty forming the watertight seal that separates the pathway to the stomach from the lungs. 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 pneumonia, a serious infection or inflammation of the lungs. Aspiration pneumonia is a bacterial infection 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 could be aspirated during coughing or choking spells. There are many difficulties in PSP/CBD/MSA that are out of one's control. Maintaining good oral hygiene is something you can take charge of.

### **Tips**

- Take action steps early after diagnosis to avoid extensive dental procedures later when they will be much more difficult to tolerate
- Make an appointment with your dentist as soon as possible after diagnosis; make sure you have a list of all medical conditions and medications on hand
- Take this opportunity to educate the dental staff about the disease
- Although most dental insurances cover two cleanings a year, it would be a good investment to consider three/four cleanings a year
- Ask your dentist about prescribing a fluoride dental paste that will meet your needs
- Request a fluoride treatment at your appointment
- Ask your dentist to make sure partials or dentures are fitting correctly
- Ask the dental staff if dental X-rays are up to date; make sure the dental office is wheelchair accessible

### **Brushing**

- Make sure your care receiver's teeth are brushed at least twice a day with the last brushing being after meal/snack in the evening
- Purchase an electric toothbrush
- After eating, rinse mouth with water, and use a wet wash cloth to sweep through the folds of the cheek to remove food that may be tucked away in these areas
- Make sure your toothpaste is fluoridated; over-the-counter fluoride rinses are also available
- If teeth are sensitive, try using desensitizing toothpaste
- If dentures are worn, remove dentures and use soft bristled toothbrush to scrub gums
- If a partial is worn, make sure it is taken out so all areas of the teeth and gum can be cleaned

### **Flossing**

It is difficult enough to floss one's own teeth. Flossing another's teeth is even more difficult and frustrating. Truly, there are more important matters to worry about, so brush and rinse well with an antiseptic after brushing in the morning and evening. Make sure the rinse is an antiseptic as not all mouthwashes are. Don't rinse with the antiseptic if you suffer from dry mouth.

### **Dry Mouth**

There are artificial over-the-counter products that mimic natural saliva. Artificial saliva can help to relieve the soft tissues of the mouth as well as help with decay control.



# Good Oral Hygiene—*continued*

## **Water Irrigating Devices**

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

## **Denture/Partial Care**

Dentures and partials are very delicate and may break if dropped even a few inches.

Always stand over a folded towel or a basin of water when handling dentures. 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 on dentures. 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, which are both acceptable for cleaning dentures. Do not use other powdered household cleansers, which may be too abrasive, and avoid using 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 thoroughly. Moisten the brush and apply denture cleanser. Brush every surface, scrubbing gently to avoid damage. Rinse the denture with water or an antiseptic rinse after scrubbing. 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.

## **Tube Feeding and Dental Care**

Bacteria is still present in the mouth, and these recommendations are for the tube-fed care recipient as well.

## **Conclusion**

Good oral hygiene is very important. Take time out to take care of your well-being.

*Nancy Brittingham, BS, RDH*

# Nutritional Implications

## **Introduction**

Progressive supranuclear palsy (PSP) and corticobasal degeneration (CBD) and other related atypical Parkinsonian disorders can be considered combined under the umbrella of what is generally known as atypical Parkinsonian disorders. Although, each of these neurological diseases can vary as far as diagnoses, progression and severity/timing of symptoms, these disorders have many similar nutrition-related issues throughout their course. Maintaining adequate nutrition and a healthy body weight as these diseases progress, may present many challenges to the individual, the caregiver and the primary team caring for the individual.

## **Nutritional Factors**

Many different nutritional implications occur as the disease progresses that can impact overall nutrition and hydration. These factors may include oral and/or pharyngeal muscle weakness, upper and/or lower extremity weakness, depression, cognitive dysfunction, decreased or altered appetite, and constipation. The following breakdown of each of these nutritional implications should be routinely addressed and monitored by your physician and/or healthcare team throughout the course of any of the atypical Parkinsonian disorders.

### **Oral and Pharyngeal Weakness**

If difficulty chewing or swallowing is present, a formal swallow evaluation should be completed by a speech-language pathologist. The swallow evaluation can help to assess the safest consistency for food and/or liquids to possibly decreased aspiration risk. As eating or drinking may become more difficult, mealtimes may be extended, contributing to frustration, exhaustion and possibly anxiety by the individual as well as the caregiver. Eating and/or drinking may be seen more as a “chore” than a pleasurable experience.

Trialing smaller, more frequent, calorically dense meals should be offered. High calorie foods/liquids should be discussed and encouraged if weight loss is present. Modifications to food/liquid consistency may also need to be implemented to maintain optimum safety during meals. Although, these compensatory measures may be effective for a while, more invasive measures may need to be discussed as the disease progresses. A gastrostomy tube should be discussed when individuals show signs of significant dysphagia and/or continue to lose weight.

### **Upper Extremity and/or Lower Extremity Weakness**

Nutritional intake may be compromised when patients suffering from Parkinson’s plus syndrome have hand and arm weakness or lack of coordination even if no dysphasia is noted. Patients may have increased difficulty with meal preparation and self-feeding, thus possibly promoting extended mealtimes, meal-related frustra-

## Nutritional Implications—*continued*

tion, which in turn may lead to smaller portions and decreased oral intake. An occupational therapist may offer adaptive equipment to ease with self-feeding. Offering liquid nutritional supplements may help with alleviating frustration of utensil-to-food-to-mouth coordination.

Lower extremity weakness or discoordination causing decreased mobility can contribute to constipation and even dehydration—which can affect overall intake of food/liquid. Often when patients have difficulty ambulating, fluid intake may be decreased subconsciously or consciously to avoid frequent trips to the bathroom. Decreasing fluid intake can lead to dehydration as well as constipation. Limited mobility may contribute to constipation possibly leading to a reduction of oral intake.

### **Depression**

Due to progressive nature of these diseases, the ever worsening physical deterioration presents many challenges, including depression, from the time of diagnosis to the end stages of these diseases. A common sign of depression is a reduced oral intake. Medications can be prescribed to treat depression. However, if untreated, depression may contribute to poor nutritional status. Mood and depressive symptoms should be reported to the physician or team to ensure proper treatment is in place.

### **Loss of Appetite**

Although loss of appetite may be related to depression, side effects of medications may be a culprit as well to suboptimal oral intake. If patients desire, appetite stimulants may be offered. Medications categorized as prokinetics, may alleviate symptoms of early satiety or feelings of fullness soon after mealtimes. The benefit of these medications may vary from patient to patient.

### **Constipation**

As disease progression continues, some form of constipation or slowed gastrointestinal motility will occur. Constipation is related to many varying factors including a reduction in physical mobility, side effect(s) of medications, decreased fluid intake, lower consumption of high-fiber foods or a combination of one or more of these factors. As mentioned earlier, consciously or subconsciously, decreased overall fluid intake related to dysphasia or limited physical mobility, can greatly increase one's susceptibility to constipation. Supplemental fiber, along with adequate hydration may lessen constipation. Over-the-counter medications such as stool softeners, laxatives, suppositories, and enemas may be used if constipation persists. Maintaining bowel regularity may promote more consistent nutritional intake.

### **Alternate Nutritional Interventions**

Malnutrition may still present itself due to disease progression despite one's best efforts. The implementation of adaptive equipment, compensatory swallowing strategies and changes in diet or liquid consistencies may not be enough to decrease the incidence of malnutrition and/or dehydration. A feeding tube may be introduced as an alternate route for providing nutrients, medications and water for individuals with progressive weight loss, dysphasia or inability to maintain adequate hydration.

Gastrostomy tubes, which are the most commonly placed feeding tubes for individuals with neurological disorders, can be placed in primarily three different ways: surgically, endoscopically or radiologically. Surgical placement of a gastrostomy tube involves general anesthesia, thus, the endoscopic or radiologic placement of gastrostomy tubes are often the preferred placement methods. Regardless of how gastric access is obtained, the delivery of nutrition, management and follow-up are essential for the patient and caregiver. The recommended amount of nutrition and water administered via the gastrostomy tube differs for each individual. Whether the feeding tube is for supplemental use, or is to provide all nutrition and hydration for an individual, a comprehensive nutritional assessment should be completed by a dietitian. The appropriate nutrition and fluid recommendations should be conveyed to the individual/caregiver. Then placement of a feeding tube can possibly alleviate certain "stressors" surrounding mealtimes. Although, there may be many benefits to getting a gastrostomy tube placed, it may not be appropriate for everyone. As with any procedure, certain risk factors should be identified, evaluated and discussed. It is best to review all options with your physician and healthcare team to assure a feeding tube is the appropriate choice for you.

Maintaining adequate nutrition is likely to change at some point, and to some degree, throughout the course of atypical Parkinsonian disorders. Recognizing, as well as adapting to change, with the help of a supportive physician and healthcare team, will hopefully provide ease in adjusting to these changes as they may arise.

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# Aspiration Pneumonia

Aspiration pneumonia is an infection in the lungs. It can develop when food or liquid goes down the windpipe into the lungs rather than into the stomach. This can happen when a person develops problems with swallowing. Aspiration pneumonia can also happen when food, liquids or tube feedings are vomited into the lungs and not coughed out properly. Everyone sometimes has a small amount of food “go the wrong way.” In most cases, these small amounts are removed by strong coughing and usually do not cause pneumonia.

## **How Is Aspiration Pneumonia Treated?**

Treatment for aspiration pneumonia usually includes:

### **Chest Physical Therapy**

Therapists work with the patient to encourage deep breathing and coughing. They also use a “clapping” treatment on the side of the chest. This helps loosen the secretions from the pneumonia.

### **Suctioning**

Nurses and therapists sometimes use a small tube to remove the secretions in the lungs caused by the pneumonia. The tube is placed in the nose or mouth and moved into the lungs. The tube is attached to a suction machine that removes the secretions.

### **Intravenous Antibiotics**

These are medicines used to treat the infection. They are given through the intravenous (IV) line.

### **Oxygen**

Sometimes, people need oxygen while the infection heals. Oxygen is given through the nose or a face mask.

## **How Can Aspiration Pneumonia Be Prevented?**

Some people who have had aspiration pneumonia are at risk for getting it again.

*To reduce your risk, you can:*

- Sit up straight when eating or drinking. Sitting in a chair is best
- Eat slowly; take small bites of food and small sips of liquid
- Stay sitting for about 30 minutes after eating, if you can
- When you are in bed, keep your head raised with pillows; you should rest and sleep in a sitting position in bed as much as possible
- See if changing the consistency and temperature of the food makes it easier to swallow; it may be easier to swallow thicker liquids than thin liquids
- You can also ask your doctor for a referral to a swallowing therapist

# Using Physical Therapy to Help Manage Mobility Issues

## Physical Therapists

Physical therapists (PTs) provide rehabilitation services that help to restore or maintain function, improve mobility and safety, relieve pain, and prevent or limit disabilities. PTs perform physical examinations that can include testing the strength and flexibility of muscles, and also testing balance, coordination, posture, and mobility. PTs will also perform an assessment of your home environment and your daily routine. With this information, PTs then develop a plan of care to meet your individual needs. The plan of care can include an exercise program, mobility training and adaptive device recommendations.

Physical therapy plays an important role in helping people with these diseases and their care partners cope with mobility changes that occur as the disease progresses. Commonly, people are referred to physical therapy due to problems with muscular tightness, slowed movements, mobility difficulties, loss of balance and falls.

### **The goals of physical therapy in the treatment of PSP, CBD and other atypical Parkinsonian disorders are to:**

- Maintain functional and safe mobility for as long as possible
- Prevent and/or limit falls and injuries
- Educate and train care partners to safely help the person with the disease who are no longer able to move independently

It is recommended that everyone with these diseases have a physical therapy evaluation as soon as he/she is diagnosed. Establishing a relationship early on with a therapist can help ease the physical changes that occur throughout the disease progression.

### **Physical therapists can:**

- **Design an exercise program to meet your particular needs**  
A home exercise program can be established to help maintain and/or improve strength, flexibility, coordination and balance; exercise can help maximize a person's function even with disease progression
- **Evaluate and treat mobility problems**  
Learning the proper ways to move and walk helps ensure the safety of the person and his/her care partner
- **Evaluate and treat walking problems, loss of balance, and falls**  
Due to problems with eye movements, particularly the downward gaze, people with PSP often trip or fall on uneven floor surfaces or objects on the floor; using head movements to help move the eyes, specific eye movement exercises, balance exercises, and new walking techniques can all help with fall prevention; people with PSP tend to fall backwards, so rolling walkers with weighted fronts and heel wedges in shoes can often limit falls by helping to keep the center of gravity forward;



# Using Physical Therapy to Help Manage Mobility Issues—continued

generally, rolling walkers with swivel wheels work nicely to help with improving walking safety; many types are available, from 2- and 3-wheeled to 4-wheeled, with brakes and seats to rest on

***(It is very important to have a PT help determine which device is best for your particular needs. With disease progression, different gait and mobility devices will be needed at different times.)***

- **Evaluate and treat joint or muscle pain that interferes with activities of daily living (ADLs)**  
When left untreated, pain associated with muscular tightness and injuries from falls, can lead to less mobility and independence; PTs use many different techniques to reduce pain and improve
- **Teach care partners proper body mechanics and techniques for assisting with mobility**  
When the person with one of these diseases can no longer safely move on their own, care partners often have to assist with mobility; learning proper mobility assistance techniques will help keep care partners 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 for adaptive devices to assist with performing Activities of Daily Living (ADL) such as working in the kitchen**  
Many devices are available to make ADLs easier and safer; PTs can work with you to find the best device for your needs and provide education and training in how to safely use the device

*Some examples are:*

- Bed rails
- Chair and bed risers to make surfaces higher
- Motorized lift chairs
- Specialized wheelchairs
- **Perform home safety evaluations**  
Small changes such as removing throw rugs, adding nightlights, and installing grab bars near the toilet make big differences in safety
- **Teach you new ways of performing old tasks**  
PTs can show you new ways of getting in and out of the car, in and out of a chair at a table, and many others

- **Recommend braces and proper positioning techniques to limit muscle tightness and prevent contractures**

### **Transfer Tips for Chairs and Beds**

NOTE: All techniques should first be practiced with a therapist to ensure they are being performed properly and are the best techniques for your needs

#### **Chair Tips**

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

#### **Chair Transfer Technique**

- 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”

#### **Care Partner Technique/Tips**

- Assist partner with sliding to the front of the chair and properly positioning feet
- Stand to the side of partner
- Place one arm under partner’s arm and your other arm on partner’s back
- Keep your feet wide apart and knees gently bent
- Gently guide partner forward and up

#### **Bed and Bedroom Tips**

- Avoid flannel sheets
- Avoid heavy, bulky blankets
- Make sure bed is at the correct height for your needs
- Equip bed with hand rails
- Use hospital beds with elevating head and feet options
- Use nightlights
- Keep a clear path from the bed to the bathroom, or use a urinal or commode

#### **Bed Transfer**

(It is recommended that you try various techniques with the assistance of a therapist to find out which technique works best.)

# Using Physical Therapy to Help Manage Mobility Issues—continued

Never pull partner by the neck or arms; rather, guide partner by placing your hands on his/her shoulders, trunk or hips. Take your time and break the task down into small steps; do not try to do it all at once.

## **General Exercises**

Performing regular exercise helps with strength, flexibility, posture and mobility. Exercise also promotes cardiovascular fitness, 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) are all forms of 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. Aquatic therapy is well worth it for those who are comfortable in the water. Contact your local YMCA, community center or health club to see what programs they offer. The trick to staying with a program is to find something that is enjoyable for you.

For a more detailed and individualized program, see your physical therapist. *Happy exercising!*

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# Adapting to Swallowing and Communication Problems in PSP, CBD, and MSA

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 (PD). The intent of this chapter 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. Consultation with your physician and speech pathologist are recommended to tailor a program to your specific needs.

## **Swallowing**

PSP, CBD, and MSA are neurodegenerative disorders that result in swallowing difficulties. The primary cause of death in patients with PSP, CBD, and MSA is infection and pulmonary complications in the setting of immobility, feeding dependence, and swallowing difficulties. Therefore, it is important that aggressive efforts are pursued to evaluate and manage these symptoms in an attempt to minimize complications such as malnutrition 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, or saliva “go down the wrong way”, into the trachea toward the lungs, instead of towards the stomach. This can lead to the development of aspiration pneumonia, an infection that develops in the lungs because material is aspirated, whether saliva, food, liquids, or refluxed contents from the stomach. 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, limiting the ability to see the plate during meals; behavioral changes such as disinhibition, resulting in rapid drinking or mouth-stuffing; and stiffness, interfering with self-feeding. The control and timing of the swallowing mechanism may be affected by an extended head and neck posture altering the proportional relations of the mouth and throat, dementia, or lack of awareness of deficits. Slow and incomplete chewing as well as swallowing apraxia is observed in CBD. Apraxia is the inability to perform purposeful movements, so they may hold foods in their mouth and not swallow. The apraxia can also affect the hands and interfere with the ability to feed themselves. In MSA, there may be difficulty sitting upright at mealtimes, and a tendency towards holding foods or liquids in the mouth. Cough ability may also be impaired.

# Adapting to Swallowing and Communication Problems in PSP, CBD, and MSA—*continued*

## **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, 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 a potential problem with swallowing: drooling, food collecting in the mouth, apparent increased effort with swallowing, a “wet” voice quality, difficulty managing secretions, abnormal posture, trouble swallowing medications, increased length of meal-time, low-grade fever, chest congestion, trouble talking, coughing and choking with a red face, coughing more during meal times than at other times of the day, unintentional weight loss, need for the Heimlich maneuver, need for diet modifications, and pneumonia. Some patients with PSP may not recognize that they have an eating or swallowing problem, whereas persons with CBD and MSA do tend to be aware. Coughing is a normal response, and “silent aspiration” occurs when the individual does not cough when food, liquids, or saliva go down the wrong way. If any of these signs occur, you should notify your physician. A referral should be made to a speech pathologist specializing in swallowing problems, preferably one who is familiar with PSP, CBD, or MSA, or related movement disorders.

## **What Is Aspiration Pneumonia?**

Aspiration pneumonia is an infection that forms in the lungs following 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, including 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, and it may potentially increase the risk. The development of aspiration pneumonia is a complicated, multi-factorial process.

## **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 to help loosen the secretions
- *Suctioning:* Nurses use a small tube to remove the secretions in the lungs caused by the pneumonia, via a tube attached to a suction machine and placed in the nose or mouth and briefly advanced into the lungs
- *Antibiotics:* Medications used to treat the infection
- *Oxygen:* Occasionally people need oxygen administered through a face mask or nasal cannula while the infection resolves

### **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 what the surrounding circumstances were. 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. 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 dynamic, video recorded radiographic 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 clinic, but this is typically of less benefit as it does not visualize the mouth, but only the throat during the swallow. A swallowing study is used to document current swallowing ability, rule out non-neurogenic causes of dysphagia, identify strategies which may enhance the ease and safety of swallowing, assist in patient education to the extent of dysfunction methodology for strategies, and perhaps 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 monitored frequently.

### **Will Swallowing Therapy Help?**

Presently, there are few reports in the literature as to the role of direct swallowing exercises in neurodegenerative diseases, much less 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, reduce the risk of choking and aspiration pneumonia, and to continue oral intake as long as possible for enjoyment and independence.



# Adapting to Swallowing and Communication Problems in PSP, CBD, and MSA—*continued*

## **What Can Be Done to Improve the Mealtime Success?**

1. Sit upright, preferably in a chair, during meals. For MSA in particular, support to sit upright against the chair back.
2. Eliminate distractions, such as television or conversation during meals.
3. Concentrate on maintaining a slow, steady rate of self-feeding or feeding assistance (PSP). Make sure food/liquid is swallowed before the next bite or sip.
4. Keep the plate of food in the line of vision by placing it on a telephone book (PSP).
5. Substitute a pair of reading glasses for bifocals, which are often ineffective in the setting of impaired downward gaze (PSP).
6. 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 one with a modified grip, to make it easier to hold.
7. 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.
8. 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 & Provale Cup*: 800-225-2610 • [www.alimed.com](http://www.alimed.com)
- *Independence Spillproof Flo Tumbler*: 888- 843-5287 • [www.kcup.com](http://www.kcup.com)
- *Wedge Cup*: 985-722-8269 • [www.wedgocup.net](http://www.wedgocup.net)
- *Provale Cup*: 423-265-3574 • [www.magistercorp.com/provale-cup.html](http://www.magistercorp.com/provale-cup.html)
- *Maroon Spoons*: 800-225-2610 • [www.alimed.com/maroon-spoons.html](http://www.alimed.com/maroon-spoons.html)
- *Scooper Plate w/non-skid Base*: 913-390-0247 • [www.bindependent.com/product/SNK107/Scooper-Plate](http://www.bindependent.com/product/SNK107/Scooper-Plate)
- *Skidtrol Non-Skid Bowl*: 973-628-7600 • [www.maddak.com/skidtrol-nonskid-bowl-p-27963.html](http://www.maddak.com/skidtrol-nonskid-bowl-p-27963.html)
- *Bruce Medical Supply*: 800-225-8446 • [www.brucemedical.com](http://www.brucemedical.com)
- *WisdomKing.com, Inc.*: 877-931-9693 • [www.wisdomking.com/line250.html](http://www.wisdomking.com/line250.html)

## **What are Some Strategies That Might Make It Easier and Safer To Swallow?**

1. Make sure the mouth and throat are clear of excessive secretions/saliva prior to eating or drinking.
2. Maintain the head in a slightly chin down position while eating, drinking and taking medications, specifically avoiding extending the head backwards.
3. Avoid taking too large a bite, drinking too rapidly, or taking more than one bite at a time.
4. Alternate food and liquid swallows to assist with clearance of dry or more textured foods.
5. 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.

6. 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.
7. Thin liquids may be difficult to swallow because they move faster through the mouth and throat. Either restrict the amount per swallow, or thicken liquids. Information on commercially available thickeners is provided below. Potato or banana flakes, fruit purees, tofu, tapioca, and oatmeal can also be used.
8. Carbonated beverages may reduce frequency of aspiration and result in less residue in the throat after the swallow.
9. Try cold liquids versus liquids that are body temperature.
10. It may be easier to take medications with a spoonful of a puree consistency such as pudding or applesauce instead of liquids. Do not use gelatin. Do not crush medications unless approved by a physician. Take one medication at a time.
11. Consider more frequent, smaller meals.
12. Use less-affected side for self-feeding (CBD).
13. 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?**

1. It is important to maintain good oral hygiene to minimize risk of aspiration pneumonia and enhance ease and enjoyment of oral intake.
2. Minimize use of dairy products, which often have a tendency to make secretions thicker, thereby interfering with swallowing.
3. A portable suction machine may be of value to assist with clearance of secretions, particularly at meal times. Drinking more water, or sometimes use of carbonated beverages, may help break up secretions. Your physician may have suggestions for over the counter or prescription medications to thin or reduce secretions.
4. It is important that the caregiver be educated in the use of the *Heimlich maneuver*. Ask your physician, nurse or therapist for instructions.
5. Monitor for unintentional weight loss.
6. Evaluate the medications prescribed, because some of them may cause the swallowing problem or make it worse.
7. It is important to be alert to the signs of aspiration pneumonia, including increased chest congestion, chronic low grade fever, increased cough (particularly with meal times) and change in sputum. Do they cough more at meal times than at other times of the day?
8. 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.

# Adapting to Swallowing and Communication Problems in PSP, CBD, and MSA—*continued*

Consider the “plus” version of these products for more concentrated calories and nutrition.

9. Supervision during mealtimes is always a good idea.
10. Consult with a dietitian to ensure that your meal plans contain all the nutrients that you need.

## **Commercially Available Food and Liquid Thickeners:**

Thickening liquids is sometimes used to slow the rate the 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. 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*: 800-333-0003 • [www.thickitretail.com](http://www.thickitretail.com)
- *RESOURCE ThickenUp*: 888-240-2713 • [www.nestlenutritionstore.com](http://www.nestlenutritionstore.com)
- *SimplyThick*: 800-205-7115 • [www.simplythick.com](http://www.simplythick.com)
- *Thick & Easy*: 800-866-7757 • [www.hormelhealthlabs.com](http://www.hormelhealthlabs.com)

## **Swallowing Cookbooks:**

Other cookbooks are on the market, however, these books are recommended because they are more readily available.

1. Achilles E & Levine T. *The Dysphagia Cookbook*. 2003. Cumberland House Publishing.
2. *MEALS for Easy Swallowing*. 2005. Muscular Dystrophy Association Publications.  
[mda.org/sites/default/files/publications/Meals\\_Easy\\_Swallowing\\_P-508.pdf](http://mda.org/sites/default/files/publications/Meals_Easy_Swallowing_P-508.pdf)
3. Weihofen D, Robbins J, Sullivan P. *Easy-to-Swallow Easy-to-Chew Cookbook*. 2002. John Wiley & Sons, Inc.
4. 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.
5. Woodruff S & Gilbert-Henderson L. *Soft Foods for Easier Eating Cookbook: Recipes for People who have Chewing and Swallowing Difficulties*. 2007. Square One Publishers.

## **Why Is Oral Hygiene So Important?**

Poor oral hygiene, including dry mouth and thickened secretions, can interfere with the ease and safety of swallowing, as well as pulmonary health. It can affect chewing, the ability to start a swallow and the duration of a swallow, as well as the perception of swallowing abilities. Dysphagia may be an important risk factor in the development of aspiration pneumonia, but generally is not sufficient to cause pneumonia unless other risk factors are present. Other risk factors include dependence for feeding assistance, dependence for oral care, presence of a feeding tube, more than one medical diagnosis, smoking, and poor oral hygiene. So, it is important to optimize oral hygiene.

**How Can I Improve Upon Oral Hygiene?**

1. Maintain scrupulous dental care.
2. Use soft toothbrushes, and electric toothbrushes are even better. Toothettes may be better than nothing, but tend not to be abrasive enough.
3. Avoid smoking.
4. Avoid alcohol, caffeine and citrus containing drinks.
5. Drink plenty of water.
6. Use sugar-free hard candies.
7. Minimize non-cooked dairy products.
8. Avoid overly salty foods.
9. Meat tenderizer or papase (papaya enzyme found in fresh/frozen papaya) may thin secretions, but should be used with caution.
10. Club soda or sparkling water can be used to cut through thickened secretions.
11. Avoid mouthwashes and lozenges that contain menthol or alcohol.
12. Consider use of a night-time humidifier at bedside.
13. Make sure dentures are clean and well fitting.
14. Consult with a dentist.

**Oral Hygiene Products:**

- Biotene oral care products contain moisturizers: 800-922-5856 • [www.biotene.com](http://www.biotene.com)
- Plak-Vac & Plak-Vac/Res-Q-Vac oral suction toothbrush system: 800-325-9044  
[www.trademarkmedical.com/personal/personal-oral.html](http://www.trademarkmedical.com/personal/personal-oral.html)

**Should I Make a Dental Appointment?**

- Take action steps early after diagnosis to avoid extensive dental procedures later when they will be much more difficult to tolerate
- Take this opportunity to educate the dental staff about PSP, CBD, and MSA
- Although most dental insurances cover two cleanings a year, it would be a good investment to consider three/four cleanings a year
- Ask your dentist about prescribing a fluoride dental paste that will meet your needs
- Request a fluoride treatment at your appointment
- Ask your dentist to make sure partials or dentures are fitting correctly
- Ask the dental staff if dental X-rays are up to date
- Make sure the dental office is wheelchair accessible

**What Do I Need to Know about Brushing Teeth?**

1. Make sure teeth are brushed at least twice a day with the last brushing being after meal/snack
2. After eating, rinse mouth with water, and use a wet wash cloth to sweep through the folds of the cheek to remove food that may be tucked away in these areas.

# Adapting to Swallowing and Communication Problems in PSP, CBD, and MSA—*continued*

3. Make sure your toothpaste is fluoridated.
4. Use non-alcohol based mouthwashes, such as Listerine Zero or Crest.
5. If dentures are worn, remove dentures and use soft bristled toothbrush to scrub gums.
6. If a partial is worn, make sure it is taken out so all areas of the teeth and gum can be cleaned.

## **What Do I Need to Know About the Care of Dentures and Partial Plates?**

Like natural teeth, dentures must be brushed daily to remove food deposits and plaque. Use a brush designed for cleaning dentures or a toothbrush with soft bristles. Brush every surface, scrubbing gently to avoid damage. Rinse the denture with water or Listerine after scrubbing. 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.

## **Is It Important to Maintain Oral Care while Using a Feeding Tube?**

Bacteria are still present in the mouth, so oral hygiene must be maintained even when using a feeding tube.

## **Alternative Nutrition**

### **What Do I Need to Know about Alternative Nutrition?**

All or most of nutrition and hydration can be provided in liquid form by a feeding tube. In some situations, a feeding tube may be indicated if there is evidence of recurrent aspiration pneumonia, reduced enjoyment of mealtimes, increased duration of mealtimes, progressive weight loss or dehydration despite efforts to optimize the feeding situation, trouble swallowing coexisting with depressed alertness, or clinical evidence of frequent aspiration or significant silent aspiration on the swallowing study. Little is known of the role, timing, and benefits of tube feeding in PSP, CBD, and MSA, particularly in persons with advanced disease. Since aging and nutritional deficiencies may severely compromise a person's potential to confront PSP, CBD, and MSA, it remains to be determined whether early and aggressive nutritional therapy may delay its progression. Placement of a feeding tube does not eliminate the potential for aspiration pneumonia, as gastric contents and saliva can still be aspirated. Placement of a feeding tube does not preclude ongoing oral intake.

### **What Is a Feeding Tube?**

A feeding tube is a soft plastic tube with a cap on it, sealed when not in use, 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, but for those patients who require longer use, the tube is placed directly into the stomach through the abdominal wall. The tube is usually placed in the stomach, but can be placed further down, in the jejunum. The type of tube and location of placement will be determined by the physician. The tube can either be placed surgically, a gastrostomy tube (G-tube), or endoscopically, a percutane-

ous endoscopic gastrostomy (PEG). The majority of tubes placed today are PEGs. Feeding tubes are not painful and are not easily visible when wearing normal clothes. When not in use, it can simply be taped or bound to the belly to prevent it from 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 or not to have a feeding tube placed. This discussion should take place sooner rather than later, and be repeated frequently. Prior to determining that a feeding tube is the right course of action, the facts need to be reviewed, and discussions should be initiated prior to a health crisis. The person with PSP, CBD, or MSA and family should agree in advance with the doctor about what is hoped to be accomplished from placing a feeding tube. Decisions must revolve around the assessment of burdens and benefits. This requires value judgments and consideration of quality of life.

## **Communication**

### **What Problems Can I Expect with Talking?**

Problems with communication tend to be present and progressive in all individuals with PSP, CBD, and MSA. Dysarthria is the term used for the speech disorders that result when the muscles cannot move with the correct range of movement, strength, speed, force or coordination, causing slurred speech, slowed or rapid speech, etc. Dysphonia is the term used for disorders of voice, affecting the sound that comes from the voice box (larynx) causing hoarseness, weakness, monotone, vocal strain, etc. To address the changes in communication, some comparisons with Parkinson disease (PD) may provide some additional insight.

Speech characteristics may ultimately distinguish PSP, CBD, and MSA from PD. In PSP, CBD, and MSA, the dysarthria is often mixed, whereas in PD, hypokinetic dysarthria predominates. The speech and voice of PSP is characterized by strained voice, slowed rate of speech, emotional lability, and palilalia. Palilalia is the compulsive repetition of utterances, often in the context of increasing rate and decreasing loudness. Changes in speech versus voice are typically more apparent in MSA. "Yes-no" confusion, apraxia, and non-fluent aphasia are common in CBD. Apraxia is the inability to complete a motor movement on command, in the absence of weakness. Dysarthria in PSP, CBD, and MSA can be severe even in earlier stages, and anarthria, or the inability to speak, may ultimately result in later stages of the disease.

### **What Are Some 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.

1. Eliminate distractions, and reduce background noise (TV, radio, newspaper, large groups of people, close the door, etc.)
2. The listener should face the speaker and be an active listener.
3. Keep questions and comments brief.
4. Ask one question at a time, allowing time for a response.



# Adapting to Swallowing and Communication Problems in PSP, CBD, and MSA—*continued*

5. Stick with one topic at a time.
6. Ask for the topic of the message so you can use context cues to help with interpretation.
7. Ask targeted “yes/no” format questions.
8. Pay attention to gestures and facial expressions.
9. 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. . . ?”
10. Try to keep to familiar topics.
11. Allow enough time for the person to convey his/her message.
12. Give the person choices to ease decision making, such as, “Do you want coffee or tea?” rather than, “What do you want to drink?”
13. Be patient.

## **What Are Some Specific Strategies for the Speaker?**

Consult with a speech pathologist who will recommend specific exercises to address limitations in speech and voice. Here are some strategies which may enhance the speaker’s success:

1. Keep sentences short.
2. Repeat the entire sentence when necessary, versus an isolated word, providing a context for the listener.
3. Exaggerate and be deliberate with all speech sounds.
4. Take a deep breath before speaking, and breathe often.
5. Use gestures.
6. Speak one sentence at a time without immediate repetition, remembering potential for palilalia.
7. Speak loudly and slowly.

## **Do Speech Therapy Programs for PD Help PSP, CBD, or MSA?**

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

A speech pathologist can be helpful at all stages of the disease, preferably with early involvement and intervention emphasizing good speaking habits, prior to the underlying problems becoming so severe that new learning is difficult. It is critical to discuss with the therapist 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](http://www.lsvtglobal.com)) is frequently applied to the communication changes in PD. 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 PD communication impairment, because the primary problems experienced are low volume, rapid rate of speech and imprecise articulation. It may not have the same impact in PSP, CBD, or MSA given that the speech problem is more pervasive, and 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 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 can be used to facilitate conversation. Text to speech options with tablets and smart phones are of good quality and easy to access, using apps such as *Speak it!* for iPad for example. Often text can be scripted and saved to facilitate conversation and expression of daily needs. Augmentative communication systems with more diverse means of access are available as well.

- *Lightwriter*®: 877-258-4335 • [www.toby-churchill.com/products/text-speech/keyboard-lightwriters/sl40](http://www.toby-churchill.com/products/text-speech/keyboard-lightwriters/sl40)
- *Dynavox*: 866-396-2869 • [www.dynavoxtech.com/products/](http://www.dynavoxtech.com/products/)
- *Proloquo2Go*: • [www.assistiveware.com/product/proloquo2go](http://www.assistiveware.com/product/proloquo2go)
- *Speak it!*: • [itunes.apple.com/us/app/speak-it!-text-to-speech/id308629295?mt=8](https://itunes.apple.com/us/app/speak-it!-text-to-speech/id308629295?mt=8)
- *Word Power™ OnBoard*: 800-588-4548 • [www.mayer-johnson.com/word-power-onboard](http://www.mayer-johnson.com/word-power-onboard)

### **How Can I Find Someone to Recommend a Communication Device?**

The speech pathologist is responsible for evaluating the patient and training them to use the system for communication. Ask the therapist if they have experience in working with augmentative communication devices. An evaluation determines which device best meets the needs and abilities of the person with PSP, CBD, and MSA. Evaluations usually are conducted by a team of professionals, including a speech pathologist, and perhaps an occupational therapist, a physical therapist, and a rehabilitation engineer. The evaluation will include an assessment of speech, language, visual, and physical motor abilities. An evaluation by objective professionals will allow one to make an informed choice from firsthand experience with a variety of devices. These teams usually work in centers specializing in assistive technology located mainly in rehabilitation hospitals, university clinics, and not-for-profit organizations. These centers typically have a variety of equipment available so that the person can try a number of devices that might meet their needs. Some even allow for a loaner period to further confirm the 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:

# Adapting to Swallowing and Communication Problems in PSP, CBD, and MSA—*continued*

- ASHA (American Speech-Language-Hearing Association): 800-638-8255 • [www.asha.org](http://www.asha.org)
- ISAAC (International Society for Augmentative & Alternative Communication): 905-850-6848  
[www.isaac-online.org](http://www.isaac-online.org)
- RESNA Technical Assistance Project: 703-524-6686 • [www.resna.org](http://www.resna.org)
- USSAAC (United States Society of Alternative & Augmentative Communication): 215-631-1877  
[www.ussaac.org](http://www.ussaac.org)

## **What Is a Voice Amplifier, and Will That Help Me?**

The muscles involved with voice production and breathing can be affected, resulting in a weak voice. A voice amplifier can increase the loudness of speech, and 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 also. There are many devices available - below are some suggestions.

## **Personal Voice Amplifiers:**

- *Chattervox*: 847-816-8580 • [www.chattervox.com](http://www.chattervox.com)
- *Spokeman*: 800-255-3408 • [www.luminaud.com/spokeman.htm](http://www.luminaud.com/spokeman.htm)
- *Speech Enhancer*: 888-463-7353 • [www.speechenhancer.com](http://www.speechenhancer.com)

## **What about Communicating within an Emergency Situation?**

There should be a mechanism in place within the home to communicate with one another, as well as a means to alert to the need for medical attention. Within the home, things such as walkie-talkies or a bell may be sufficient. A medical alert system is often a wise consideration to request outside medical attention.

Consultation with the physician and speech pathologist should help tailor a swallowing and speech therapy program appropriate to the needs of the person with PSP, CBD, and MSA.

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# Adapting to Adaptability: An Occupational Therapist's Perspective

*Occupational therapists (OTs) are primarily focused on the “occupation” of someone and what they 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 care partners to problem solve and create new ways to make routine activities easier and safer.*

As anyone with PSP knows, the disease is relentless and sneaky. Over time, it whittles away at your ability to carry out the daily activities that matter to you. Vision may begin to change, posture may stiffen, and your legs may seem to have minds of their own. The easiest things, such as speaking and swallowing, may become difficult. With each change in your abilities, you may feel as though you are gradually losing the skill to live a safe and fulfilling life. Many people retreat from the things they love most, afraid to fail. The disease tries to rob you of mobility, social relationships and work skills. In this way, it attacks your very sense of identity and purpose in the world. You can fight back. You can learn new ways to do things. You can make the environment around you safer and more functional. You can use the many tools available to help you stay active and independent for as long as possible.

Occupational therapists (OTs) can help you along this path in a variety of ways, teaching you and your loved ones compensatory strategies for managing everyday tasks in new ways, home adaptations to make your environment safe and more functional and how to make wise choices from the assortment of assistive devices available on the market today. OTs can assist you in managing physical, functional, visual and cognitive changes related to PSP. They are trained to work from a person-centered perspective, focused on building a supportive social and environmental context to help you fight the disease. Your OT may address functional vision, physical skills, home safety, community independence and cognition, always working collaboratively with you and your family to build the best mix of supports to help you. Though every person with PSP has a different situation and requires an individualized approach, let's look at some of the most common problems you may face and some of the recommendations your OT may suggest:

## **Functional Vision**

Vision is our most far-reaching sensory ability. Any visual dysfunction can prove crippling, and in PSP, several visual skills are under attack. Characteristic visual changes associated with PSP include difficulty in down gaze, blurred or double vision (diplopia), reduced blink rate (leading to painful, dry eyes), photosensitivity (especially in bright sunlight), interrupted smooth tracking and steady gaze, and a reduced ability for the eyes to converge while watching objects move closer. Family members often say that their loved ones with PSP seem to stare off into space. This is probably an unconscious compensatory strategy intended to ease eye strain. If you have begun to experience visual difficulties, by all means see a neuro-ophthalmologist. These eye doctors specialize in managing visual changes wrought by neurological processes. But don't stop there. Seek out a neuro-optometrist too. These eye care professionals can fit you with glasses that combat double vision and

provide other treatments to address photosensitivity and tracking difficulties. Neuro-optometrists (sometimes called behavioral optometrists), like neuro-ophthalmologists, specialize in working with individuals who have neurologically-based visual problems.

You will want to discuss your visual problems with your occupational therapist also. He/she may recommend home improvements, task adaptations and assistive devices to help you optimize your visual skills. For instance, it is important to make sure that your lighting at home is bright, but without glare, especially in the areas where you like to read, cook, groom, work on crafts or hobbies, etc. Spend a weekend clearing out clutter from floor areas, cabinets and work surfaces so your overtaxed eyes can find things more easily and avoid pathway obstacles.

Wherever possible, improve visual contrasts in the home. For instance, you may want to replace wall switch faceplates and doorknobs to make better visual targets. Florid wallpaper can play havoc with already-challenged eyesight. Flat colors and clearly marked surfaces work better. If you have down gaze difficulty, you may find that a downward-tilted mirror at the bathroom sink can help. You see items below your visual field. Some people even install downward-tilted mirrors above their kitchen stoves. You can purchase mirrored prism glasses that work in the same way as downward-tilted mirrors, allowing you to hold a book in your lap and read as you always have. Bookstands are available that hold reading materials at eye level. If text seems blurred, magnifying glasses may help. If you use a computer, you can easily increase print size and contrast on the screen using software that comes already installed on home computers. Many internet web pages offer a "print only" version that is more easily read than a graphics-laden page.

If you suffer from double vision, your occupational therapist may show you how to tape a pair of glasses to compensate. To do this, you apply a finger's width of white tape to the nasal half of one lens of your glasses, blocking the vision from one eye when staring straight ahead. Since it is most important to have sharp vision when staring straight ahead (as when reading), this inexpensive strategy can help reduce double vision, at least until your eye doctor offers a more permanent solution.

Visual difficulties can make community outings a chore. Gray-tinted wraparound glasses are good choices for reducing photosensitivity in bright daylight. You may want to use a cane as well, to help demark surface changes as you walk. Always allow extra time for any activity in the community. Seek quieter outings at less busy times, so as not to rush or overtax your visual abilities. Finally, your occupational therapist may help you learn a systematic head-bobbing strategy that can help you scan your visual environment as you walk.

### **Home Adaptations for Fall Safety**

If you have fallen, or if your legs feel weak, definitely talk to your occupational therapist about ways to make everyday tasks safe. If you do not have a tub bench or shower seat, you should seriously think about getting one. These sturdy, waterproof furniture items are available at medical equipment houses and larger pharmacies, and the Salvation Army often offers them used at a sharp discount. They allow you to get in and out of the tub safely, sitting down first, and then swinging your legs in. You shower sitting down, so there is no

risk of falling. You may want to add a shower mat, a clamp-on grab rail and maybe a shower hose as well. Your occupational therapist can recommend exactly the right combination for your needs and bathroom. For those of you who prefer baths to showers, some newer bath chairs actually raise and lower you safely into the tub. Various models are available, but none are cheap (the least expensive currently runs about \$1,000). Getting on and off the toilet, or any chair in the house, can be a cumbersome task. Three-in-one commodes are covered by Medicare and most insurance plans. These items can fit over your toilet, serving as a bedside commode or even as a shower chair. They have armrests, and you can raise or lower their height to fit yours. One version even comes with a spring system that slowly lifts the seat to help you to your feet.

The same principle works on spring-based boost-up chair pads, available at medical supply houses. You can use these pads with any chair in your home. 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 your legs into the car. When getting out, bring your legs out first, then lean forward to come to a stand. If you have trouble pivoting on the car seat, frisbee-shaped pivot-disks are available to sit on. Or, you may choose to place a plastic garbage bag on the seat to help you slide more easily when getting in and out.

### **Electronic Assistive Devices**

All sorts of new portable devices are available to help you manage everyday tasks and communicate with caregivers. Personal pagers and walkie-talkies can serve as emergency call bells for when your caregiver is out of earshot. Miniature computerized reminder systems now come built into wrist watches and handheld devices. Newer pill boxes have timer alarms, and some even dispense medication or send messages over the Internet to a caregiver or medical provider.

There are many varieties of adapted telephones today, some with extra large keys, photo identification screens or one-touch dialing. For as little as \$100, you can purchase plug-in devices that smarten up your home, making control of lights and other appliances automatic or managed by a TV remote control. With these items, you can maintain your independence, relying less on caregivers to help you through your day. Your OT can help you choose the right combination of electronic assistive devices for your situation. In cases where assistive technology is not covered by insurance, you may be able to acquire a low-cost assistive technology loan. Many states have set up low-cost loan programs and have web sites designed to guide you in the process. PSP is a relentless disease, but there are many ways that you can fight back to maintain your functional abilities in the face of this sneaky opponent. Some of the options available for managing everyday tasks have been discussed above. Talk to your occupational therapist about the best ways to address your own special issues.

### **Resources:**

- *American Occupational Therapy Association* • [www.aota.org](http://www.aota.org)
- *Neuro-optometric Rehabilitation Association* • [www.nora.cc](http://www.nora.cc)

*Tony Gentry, MA, OTR/L BCN  
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# Alternative Exercise Options in the Home: The Xbox Kinect

The Xbox Kinect is one of a class of emerging consumer technologies referred to as a NUI or a Natural User Interface. NUIs are devices that recognize visual, audio and other kinds of user input beyond the keyboard, mouse and game controller. This device for the XBOX 360 game console provides individuals with a unique opportunity to work or exercise in a virtual environment that is safe and fun while being in the comfort of their own home with family and friends.

There are a variety of games to choose from depending on your interest and level of function. Games for the Kinect include sports simulations, exercise tutorials and fun athletic adventures. While having fun, you can simultaneously be working on things like improving arm movement, balance and even cognition. We all know that when we enjoy something we are more motivated to want to do it more often and the Kinect is an excellent way to compensate for loss of the ability to participate in similar activities outside of the home.

Recent studies have demonstrated that moving your body works your brain more than regular brain exercises. An example of integrating body and brain functions can be found in a collection of exercises found in Body and Brain Connection for the Kinect. These exercises were all supervised and approved by neuroscientist, Dr. Ryuta Kawashima, and require simple movements so that almost everyone can participate. Body and Brain Connection is available for purchase online or at most major retail stores.

Beyond the consumer focused XBOX 360 market and games like the Body and Brain Connection, Microsoft Research has just provided the Kinect programming interfaces for anyone interested in developing their own NUI applications. The potential of expanding the Kinect for use as an adaptive technology in clinics, at home and wherever else opens up an exciting avenue for continued wellness programs. Healthcare in the near future will benefit from the creativity of the many developers who are dedicated to expanding the reach of the Kinect and other NUIs.

As with any exercise or physical activity, you should always consult with your neurologist and/or physical and occupational therapist before beginning a new program. It is important to note, that not all exercise and activities are appropriate for every individual. Some individuals may need environmental modifications and/or assistance from a caregiver when playing one of these games. We are all different and it is important to recognize and to respect your own limitations to ensure a safe and satisfying experience.

*Christine Robertson Roxberry, OTR/L*



# Managing Cognitive Changes in PSP, CBD and Other Related Atypical Parkinsonian Disorders

*Ask patient/care partner what is most bothersome, rank them, and then deal with them:*

1. Cognitive
2. Neuropsych
3. Motor
4. Speech/Swallowing
5. Sleep

## **Cognitive:**

- Stay physically, mentally, and socially active
- Establish a ROUTINE and KEEP a daily routine
- RELIGIOUS use of a daily planner

## **Neuropsych:**

- Develop tolerance
- Choose battles wisely
- Acknowledge and redirect
- Refrain from arguing
- Use “therapeutic fibs” when necessary
- Support group involvement
- Work closely with local psychologists
- Some medications help with apathy
- Other behavior-modifying medication use is controversial

## **Motor:**

- Physical and occupational therapy
- Assistive technology

## **Speech and Swallowing:**

- Speech therapy
- Begin discussion on PEG tube sooner, rather than later

## **Sleep:**

- Sleep study
- Sleep is a quality of life issue and should be dealt with  
*(a well-rested patient and care partner will have an easier time dealing with symptoms)*

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# Managing Difficult Behaviors

<b>Agitation</b>	
<b>Potential Causes or Antecedents</b>	<b>Management Strategies</b>
Discomfort, pain	Determine source of pain and provide appropriate remedy; check clothing for comfort
Physical illness	Obtain medical examination and treatment of illness
Medication/Substance effects	Assess and monitor medication, eliminate caffeine, alcohol and other stimulants
Fatigue	Schedule adequate rest, monitor activity level and adjust as necessary
Overstimulation—excessive noise, people, radio, television	Remove patient from situation, provide quiet and safe setting, play soothing music
Caregiver becomes upset/angry	Remain calm in interactions, use low tone and slow rate of speech, control emotions
Overextending capabilities (resulting in failure)	Do not put patient in failure-oriented situations, maintain appropriate expectations

<b>Sleep Disturbance</b>	
<b>Potential Causes or Antecedents</b>	<b>Management Strategies</b>
Illness, pain, medication	Medical evaluation, change or eliminate medications as appropriate
Depression	Have patient evaluated for use of antidepressant medication or bedtime sedative
Less need for sleep	Increase daytime activity, schedule later bedtime, provide safe evening activities
Uncomfortable	Adjust temperature, lighting, determine if patient is hungry or needs to use bathroom
Excessive daytime sleepiness	Limit or eliminate naps, provide activity during the day, increase exposure to light

<b>Difficulty with Personal Care Tasks</b>	
<b>Potential Causes or Antecedents</b>	<b>Management Strategies</b>
Task too difficult or overwhelming	Divide task into small successive steps, provide assistance as needed
Caregiver impatience, rushing	Be patient, allow ample time, try again at a later time, obtain caregiving assistance from others
Cannot remember task	Demonstrate action or task, allow patient to perform parts of task that can still be completed
Cannot understand or follow caregiver instructions	Re-state instructions more simply, break down task, provide step-by-step instructions
Fear of task--cannot understand need for task or instructions	Reassure, comfort, distract with music or conversation, have patient help perform task
Inertia, lack of motivation; difficulty initiating tasks	Set up task sequence by arranging material (i.e., clothing) in order to be used, help initiate task
Pain involved with movement	Have physician evaluate; consider medication that will not further affect mental functions

<b>Incontinence</b>	
<b>Potential Causes or Antecedents</b>	<b>Management Strategies</b>
Infection, chronic illness, medication side effect, stress	Evaluate and treat medically
Difficulty finding bathroom	Place signs, picture on door, ensure adequate lighting
Difficulty seeing toilet	Use contrasting colors on toilet and floor
Difficulty undressing or impaired mobility	Simplify clothing, use elastic waistband, provide a commode, treat associated pain
Dependence created by socialized reinforcement	Allow independence when possible, even if inconvenient
Cannot express need	Schedule toileting, reduce bedtime liquids when possible
Task overwhelming	Simplify; establish step-by-step routine

<b>Wandering</b>	
<b>Potential Causes or Antecedents</b>	<b>Management Strategies</b>
Stress - noise, clutter, crowding	Reduce excessive stimulation, remove patient from stressful situation
Lost - looking for someone or something familiar	Provide familiar objects, offer assistance, reassure
Bored- restless, no stimulation	Provide meaningful activity (at appropriate level of difficulty so as not to be frustrating)
Medication side effect	Contact physician to review, change, reduce or discontinue medication
Physically active personality style, means of coping with anxiety	Provide safe area for moving about, address underlying mood, relaxation aids (warm bath, etc.)
Needing to use toilet	Institute toilet schedule, place signs or pictures on bathroom door
Responding to environmental stimuli (exit signs, doorway)	Remove or camouflage environmental stimuli, provide identification or alarm bracelet

<b>Suspiciousness or Paranoia</b>	
<b>Potential Causes or Antecedents</b>	<b>Management Strategies</b>
Forgot where objects were placed	Offer to help find, have more than one of same object, learn favorite hiding places
Misinterpreting actions or words	DO NOT argue or try to reason with patient, distract, do not take personally
Misinterpreting who people are, suspicious of their actions	Introduce self and role routinely, draw on old memory, connections; do not argue or quiz
Change in environment or routine	Reassure, provide familiar objects, maintain stable and consistent routine
Misinterpreting environment	Assess vision, hearing; modify environment, provide simple explanation, distract
Social isolation	Encourage and provide familiar social opportunities
Physical illness	Medical evaluation and treatment
Actual theft	Verify the situation, intercede when appropriate

## Managing Difficult Behaviors—*continued*

<b>Inappropriate or Impulsive Sexual Behavior</b>	
<b>Potential Causes or Antecedents</b>	<b>Management Strategies</b>
Decreased judgment and social awareness	Do not overreact or confront; respond calmly and firmly; distract and redirect
Misinterpreting caregiver's interaction	Do not give mixed sexual messages—even in jest, distract while performing personal care
Uncomfortable—too warm, tight clothing, genital irritation	Check room temperature, ensure elimination needs are met, examine for medical problems
Need for attention, affection, intimacy	Meet basic need for touch and warmth, offer soothing objects, hand or back massage
Self-stimulating, reacting to what feels good	Offer privacy, remove from inappropriate places

*Attribution to: Cognitive Neurology and Alzheimer's Disease Center  
 Northwestern University*

## Make Meaning to Stay Positive

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

## Make Meaning to Stay Positive—*continued*

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.

*Janet M. Edmunson, MEd*

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

# Long Distance Caregiving

## Caring from a Distance

Adult children and other close relatives who live far away from loved ones coping with PSP often feel uncertain how to provide help in ways that are meaningful and possible. Some feel guilty that they cannot be available to help meet everyday needs, and fall into the temptation to compensate by offering lots of advice regarding Mom or Dad's care!

As a nurse practitioner, I sometimes facilitate family conferences when it is time to make important decisions about care for people with PSP or other neurological disorders. Curiously, it is often the son or daughter who lives farthest away who voices the strongest opinion regarding what needs to happen next. It is probably best to avoid coming for a "whirlwind" visit, and dictating changes in the plan already established and supported by other siblings. Even the best-intentioned advice, without the advisor in place to monitor changes, can leave the patient and local caregiver confused and upset.

It is true that relatives not struggling with issues of everyday care often do have an ability to "see the forest instead of the trees". However, this objective insight must be expressed with caution and without criticism for family members who live close by and carry the greater responsibility for caregiving.

So what can long-distance relatives do to be helpful, short of "moving back home"? Perhaps some of these ideas contributed by support group members around the country will meet some needs for you and your family:

1. Set a designated day and time to call home each week. Don't assume that "no news is good news". Many older parents keep their own counsel even during extremely tough times, not wanting to bother adult children who are "busy with their own lives". Make the call faithfully. Two ten minute check-in visits may have more meaning than a longer call less often. If you are the son, speak for yourself. While your parents may dearly love their daughter-in-law, they want to talk to you personally.
2. Inquire tactfully whether financial help is needed. Many parents won't ask for help of a monetary nature, even if the limitations of a fixed income and medication costs mean doing without vital supplies. If a regular cash subsidy is unacceptable, offer something specific: For the incontinent patient, someone picking up the tab for pads and diapers is useful. When health insurance does not cover medication costs, offer to pay co-pays, or perhaps for dental work they've neglected.
3. Send a surprise package to the frontline caregiver once a month. This might be a bouquet of flowers, a restaurant gift certificate, or a pamper basket filled with elegant bath supplies. Be creative. Even a card with an encouraging hand-written message can mean a lot in the middle of a trying day!

## Long Distance Caregiving—*continued*

4. Budget funds for regular trips to check on your family. Don't add to the primary caregiver's stress level by expecting him or her to take care of you too! Get your own ride from the airport. If crowded quarters will be a strain, reserve a nearby hotel room. You are not there as a guest to be entertained, but to provide a listening ear and a helping hand.
5. Learn about the medications your person with PSP takes. Encourage your family to communicate 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 check-up because he's pre-occupied with caregiving? Ask them to schedule a visit to their neurologist when you can accompany them to the appointment. The doctor will benefit from hearing your observations, and you will learn much about your parent's communication style and be able to reinforce the doctor's instructions later.
6. Once or twice a year, strongly encourage and provide a 3-5 day respite for the primary caregiver. Plan to arrive a day or two prior to the caregiver "leave of absence" so you can master the necessary skills to help your care recipient. If filling in personally is simply not possible, offer to pay for a respite stay in an extended care facility. You and your family may be saving for a rainy day when it's pouring outside!
7. If your parents are able to travel, arrange for them to visit you. For some couples living with PSP, it is easier to travel and be a guest than to host family get-togethers in their home. As an added benefit, while parents are visiting your family, other relatives get a welcome break from their usual caregiving responsibilities.
8. Make a family pact that no one will make promises to the person with PSP that may be impossible to keep. Rather than saying, "Oh Dad, of course we'd never put you in a nursing home", it is better to express your empathy for a parent's preference to live at home, and affirm your willingness to discuss alternatives if the need arises. If and when the time comes that a skilled nursing care facility is the best choice or hospice care is a good alternative, exercise your strength as the "out-of-towner" with less caregiving burden to affirm the difficult decision to place the loved one with PSP 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's best for the key players whose lives are most affected.



## The Hardest Job of All

The practical tips for long-distance family members barely touch the concerns of those of you who live far away from elderly parents yet who are the primary caregivers.

Few of us would move away from infirm parents to avoid the trials of caregiving, but it is a much more agonizing decision whether to move back to help them in later years. Most people have established families and careers that preclude moving back to the hometown or current location of ailing parents to provide daily care. One family's story illustrates this point.

Anna Wright is a recently widowed 75-year-old homemaker who lives in a lakeside community near San Antonio. Despite dealing with the sudden loss of her husband two years ago and the fact that her three adult children live in cities scattered across the U.S., Anna maintains her own home and yard, and leads a very active life in her small town. This rosy picture of an active senior enjoying a productive existence leaves out one poignant reality: She is the sole caregiver for her mother, age 99, who resides in an assisted living facility 400 miles away from this only daughter. This means making the long drive every month, telephone calls from frustrated nursing staff in the middle of the night (the elder lady is not always a joy to deal with), arranging last-minute flights when emergencies occur, and the constant nagging worry of, "What will happen next?"

The odyssey that people are living longer has resulted in the phenomenon that many caregivers who themselves are seniors, are called on to take care of even older relatives. Neither the healthcare nor the social work system in this country has yet come up with practical solutions to help this most forgotten group of care providers.

One encouraging trend is the emergence of geriatric care managers (also known as geriatric case managers). This is a rapidly growing professional group, many coming from the fields of nursing, social work, and gerontology. Geriatric care managers provide a variety of services such as arranging financial, legal, and medical services, in-home care providers, and transportation. They regularly communicate with family members, and may also help with comprehensive needs assessment, family conflict mediation and crisis intervention.

Geriatric case management fees vary, depending on geographic location and what services are needed. It's important to know that geriatric care managers are required to be certified; it is also recommended that you personally interview the prospective care manager prior to engaging his or her services. Information to help locate a geriatric care manager can be found at [www.caremanager.org](http://www.caremanager.org).

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## Keeping Families Strong

PSP, CBD and other related atypical Parkinsonian disorders are very challenging diseases which have a significant impact on patients and their families. One important means of coping is to explore ways to grow in strength as a family while going through the disease.

There are many factors that make families strong. We each have a role in our family – a role that develops over time, but one that helps to shape our interactions and to define our place. Strong families express their thoughts and feelings with one another, even when there are serious disagreements and through opening up to one another discover greater closeness. Every family has its own history and traditions which relate to the stages of life and these memories of shared experience bring a wide range of emotions and a sense of togetherness. Perhaps one of the most important characteristics of strong families is a willingness to help one another and to share in that responsibility.

Any time we talk about strength, we are also talking about resilience. To be strong is a component of resilience but resilience is something more, it's the capacity to recover from hardship often with a new found sense of courage and resolve to continue to move forward. Resilience is the ability to adapt to change, to find creative solutions to complex problems and to accept what has become the 'new normal'. In diseases like PSP, CBD other related atypical Parkinsonian disorders working within this reality is something that families can help one another do.

There are many complications for family members when a loved one is diagnosed with one of these diseases. It has become a part of our culture for family members to live in different areas of the country from where they once grew up, and families are smaller in size than they were in the past. Since a person may be diagnosed in their 60's, their plans for retirement may be affected by the anticipation of their future medical needs. They may also find it difficult to locate medical professionals in their community who have experience with atypical Parkinsonian disorders. There may be financial constraints in terms of the ability to afford services, while social supports may be limited. When a parent becomes ill, their adult children may be in the midst of raising their own children. This 'sandwich generation' has the additional stress of career demands and childcare responsibilities as they also care of their affected parent.

There is a process that families go through, when they are made aware that their loved one has one of these disorders. Especially with initial reactions to the diagnosis, there is no right way to react. Everyone is different and it's important to respect the ways in which people express their emotions, knowing that they will change over time.

You may notice that family members take on certain roles once the diagnosis is made. While this is not an exhaustive list, the descriptions may help you gain a better understanding of what is happening in your family and also help you feel less alone.

Usually one family member becomes 'The Leader'. The Leader takes control of the situation, tries to obtain as much information about the disease as possible and initiates action by setting up appointments, helping maintain communication with other family members, and offering them support. The Leader manages their emotions by being active.

As with all roles we take on, there is also a negative side. The Leader can become over-burdened and feel burned out. They may find it hard to express their emotions as they see themselves as the person who needs to be strong for others. The Leader may not know how to ask for help and may fear taking a less active role because others may not be as diligent. The other negative side to this role is that it can lead to resentment of family members who are less involved. Often, this is an emotion that's not directly expressed through words, but can certainly be expressed through behavior.

Another family role is "The Supporter". The Supporter is willing to help, but sets clear boundaries. They are most comfortable taking on specific tasks that have a clear beginning and end. Asking The Supporter to take Dad to a doctor's appointment may be more manageable than asking him or her to provide companionship for an evening. The supporter may be reluctant to provide hands-on help and may seem distant from other family members. Feelings of anger or resentment may be expressed more openly by this family member, which can be particularly hard for The Leader, as they view themselves as doing the lion's share of what needs to be done.

The other side of The Supporter's struggle is a sense of being poorly informed about the patient's disease and treatments. Just like their involvement tends to be task-oriented, The Supporter may not have a full understanding of what is happening with their loved one and may not feel able to ask for help from others. The Supporter may feel undervalued because they are setting limits about what they will and won't do. They may also feel as if they should be doing more, which can bring up many complex emotions.

It seems that almost every family has one or more members that pull away during the course of the disease. "The Bystander" reduces overall communication with the family and may avoid having direct contact with the person who is ill. They are very unlikely to take on any tasks to help with care and their emotions can be suppressed or expressed in ways that make others uncomfortable. The lack of involvement of The Bystander often makes the other family members feel taken advantage of, disappointed, frustrated and/or angry. The Bystander, however, may be feeling a lot of emotions that could potentially bring them back into the family, if they were able to express themselves. The Bystander usually feels helpless and ashamed for not visiting or helping. They may feel unable to talk about what's going on and not feel worthy of support. While this can be a risk for all family members, The Bystander may become depressed, which can be difficult to recognize due to their isolative behavior.

## Keeping Families Strong—*continued*

### **So what can families do to stay strong?**

One of the first things to do is to get back to basics. One thing that naturally happens when a family member is very ill is that the focus becomes exclusively on that person, their treatment, and their well-being, but it's important to strike a balance. Consider the following:

- *Positive Emotions*: openly express love, affection and appreciation for one another
- *Acceptance*: recognize the frailty in one another and respect each person's efforts to cope
- *Definition of Roles*: make decisions as a family about who can take on a particular task, recognizing that roles will need to change over time
- *Communication*: set up routines to stay in touch, express feelings and attempt to resolve conflicts quickly
- *Willingness to Help*: be flexible, develop solutions to new and ongoing problems by combining ideas, seek out support groups for ideas
- *Sense of Togetherness*: take time out to decompress and simply enjoy one another and include your loved one who has the disease
- *Traditions*: maintain meaningful activities and celebrations as they provide continuity and are a reminder of growth and change

It's paramount to acknowledge that everyone is human and everyone is dealing with multiple stressors all at the same time. It's imperative to express your concerns and your fears and to ask each other for support. Remember to 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. Perhaps most importantly, stay close to your loved one who is experiencing the disease. Remember who they are, what you love about them, and that each moment you share is precious.

*Trish Caruana, MSW  
Vice President of Programs and Education  
CurePSP*

# Joining a Support Group

***“No, thanks. That’s not for me.”***

Does this sound familiar? Perhaps it was your own reaction when someone first suggested that you join, or start, a PSP, CBD or other related Parkinsonian disorders support group. The idea of a “support group” may have made you uneasy. You may have objected to the idea of sharing your problems with others and, besides, how can hearing about other people’s problems help?

A support group is not a problem exchange. Nor is it a place to go in order to feel more burdened. Let’s be clear from the start about what a support group is not:

- A support group is not a therapy group
- A support group is not a “12-step” program
- A support group is not a substitute for medical treatment or health counseling
- A support group is not a replacement for family and friends, or other close relationships

This is not to deny the need for family or personal counseling and other professional support, in fact, a support group can also be a resource for finding these services.

## **What is a Support Group?**

A support group is a self-help group. It is run by and for people who have a particular challenge or life situation in common. It’s a place where people are willing to talk freely and to trust openly in the goodwill of the other group members. A support group is a place to be yourself, a place where people are welcomed and accepted. Being part of a group means knowing that your privacy and confidentiality will be respected.

A support group is a resource for patients and for families. Support group members can offer a wealth of practical experience and suggestions to help you manage the day-to-day challenges of the disease. Groups often share articles, newsletters, books and websites that can also help. While no one is an “expert,” everyone has experiences to share.

A support group is an added circle of friends. The group is the place to talk and share emotions about life with these disorders with people who will listen, laugh, and cry. It is also a place where other family members and friends can feel welcome and supported. Each group is as unique as its membership and those who attend the group can change over time.

CurePSP offers both face-to-face support groups and “online” support groups. Online groups allow people to use their computer (or their telephone) to connect with people from all over the country; these groups

## Joining a Support Group—*continued*

are especially important in areas where there aren't any face-to-face groups or when travel is an issue. Face-to-face support groups meet in the community giving people the chance to sit together and get to know one another in a social setting.

### **Have you thought of becoming a support group leader?**

Many people have volunteered to run support groups and, while the reasons may vary, there are often two primary motivations for starting a group:

- There aren't any groups in your area and you want to connect with people dealing with the disease
- You feel like you want to help others

Of course, only you can decide if becoming a support group leader is something you want to do, so here are a few things to consider:

- Starting a support group takes time and energy
- Starting a support group requires organization
- Starting a support group is an ongoing commitment
- Starting a support group is not a one-person job
- Starting a support group may change your life

*Carol Mackenzie Jackson, PhD*

*Adapted for PSP from:*

*The National Parkinson's Foundation Support Group Leaders Manual*

# Ten Commandments for Family Caregivers

## **I. Thou Shalt Love Thy Self as Thy Neighbor**

Admittedly, a reversal from the original ten in Judeo-Christian tradition! Many caregivers become quite skilled over time at meeting the needs of others -- not just their primary care recipients, but those in the family and periphery who always seem to need help. This is an admonition to reserve some time and caring for your own physical and mental health in addition to what you devote to that deserving "other".

## **II. Thou Shalt Take Thine Own Advice**

This commandment is like unto the first. Seasoned caregivers not only have advice to give, but it's often quite good advice, based on a wealth of experience. Would you not encourage a neighbor or good friend to make sure her mammogram is done on time or get a flu shot? Do you treat yourself as kindly?

## **III. Thou Shalt Not Take Ridiculous Risks**

David Letterman on late night television once had a segment called, *Stupid Human Tricks*. This refers to things like climbing on ladders ten years after that practice was safe for you, moving furniture by yourself, mowing the lawn at high Noon in August, etc. You get the idea. People are lured into doing these things because their care partner is no longer able to do them, or they don't know who to call, or don't want to pay someone else to do the job. I often remind caregivers that they are just one broken hip away from a family disaster. If a house or yard task is out of your reach or above your pay grade, just say no!

## **IV. Thou Shalt Not Take Thy Doctors Too Seriously**

Not to demean my colleagues in the medical profession, but sometimes physicians, patients, and caregivers indulge in a small group hallucination regarding who is in charge of the patient's regimen. Rather than consider the neurologist or other doctor as the "boss of you", consider reframing the relationship into one where the doctor is the head coach. Even the best medical coach cannot be out on the field, at home with you every day observing subtle changes in the patient, responses to medications, etc. An educated caregiver is in the best position to collaborate with the patient to take the lead on the healthcare team.

## **V. Thou Shalt Claim Both Rest and Respite**

It's helpful to differentiate the two terms. Rest means literally taking time during the day to sit down and put your feet up, or maybe even grab a 30-minute power nap. It means getting a good night's sleep, even if doing so requires occupying a different bedroom part of the time. Respite is better defined as those lovely little slices of time when a caregiver permits a break from the usual responsibilities.



## Ten Commandments for Family Caregivers—*continued*

I urge caregivers to think in terms of one hour all to themselves every day, one day out of every week (remember Mother's Day out when your children were small?), and one long weekend away every month. For full-time primary caregivers, this is the tricky one. You have to train and trust a back-up caregiver, and you have to define away. Being available by cell phone with no boundaries hardly qualifies! But with advanced planning and deliberate attention to expanding your circle of support, it is possible.

### **VI. Thou Shalt Not Roll Thine Eyes**

Long-term caregiving puts you at risk of becoming weary and cynical. The unthinkable question, "can I continue to do this indefinitely" creeps into one's consciousness. There is a very thin line between constructive coaching of your loved one ("honey, why don't you try it this way?") and criticism-sliding-into-contempt ("you've got to quit jumping up to get the phone; you're driving me crazy!"). Contempt is never a good thing in a committed relationship.

### **VII. Thou Shalt Observe Regular News Breaks**

I recently toured the CNN Studios in Atlanta, and despite the operation being quite impressive, I was struck with the reality that the 24/7 news addiction in this country is truly bad for our mental health! People who are at home a great deal during the day with TV for companionship are particularly at risk. Watching the network and local news before bedtime is hardly a prescription for feeling at peace with the world and ready for a restful night's sleep! Remind yourself and your care recipient that the most important feature of the remote control is the "off" button.

### **VIII. Thou Shalt Know When Thy Warranty is Up**

Bluntly, if you're over 70, your warranty is up, and you could begin to have parts fail at any time! Why dwell on this sobering fact? First, it solidly reminds caregivers to take good care of themselves (like the quip, "if I'd known I was going to live this long, I'd have taken better care of myself!") Second, but perhaps even more important, the warranty analogy reminds us to get busy doing the things we really want to accomplish or experience in our lifetime. I often urge caregivers to see the movie *Bucket List*, and begin thinking about their own wish lists.

**IX. Thou Shalt Cultivate Thy Sense of Humor**

Humorist Erma Bombeck was speaking to an audience of professional speakers when I was practicing in Phoenix a few years ago. She taught us that a sense of humor is not the art of being funny, it's simply recognizing what's truly funny all around us. Fine-tuning this skill can immensely improve quality of life for family caregivers. We can learn to take our caregiving mission seriously without having to take ourselves too seriously. Laughter really is the best medicine!

**X. Thou Shalt Hope Without Ceasing**

Working with the hospice movement the past few years has taught me incredible things about the power of hope. For instance, there is no such thing as "false" hope; there is just hope. Whether it is hope for world peace, hope for life after death, hope that lunch will be on time, or hope that a visitor won't drop in during your favorite TV program, HOPE is all powerful and it stands alone. Emily Dickinson's famous poem about hope begins with this inspiring stanza:

***Hope is the thing with feathers  
That flutters in the soul  
And sings the tune without the words  
And never stops at all***

*Susan C. Imke, FNP, GNP-C  
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# Travel Tips

Travel is accessible for many people with PSP, CBD and other related atypical Parkinsonian disorders. The idea of planning and taking a trip may be daunting and stressful for some. However, once broken down into specific categories and questions, the task is not only manageable, but also produces fruitful and satisfying results.

## **For Starters:**

- Plan your trip carefully and in advance
- Check your medical insurance policy to be certain that you are adequately covered and be aware of services provided (or not provided) in other countries
- Ask your neurologist if s/he can give you the name of a doctor in the area to which you are traveling
- Find out if there is a CurePSP support group or National Parkinson support group in the area you are visiting
- Don't forget to rest the day before your trip AND the day (or day after) you arrive

## **About Medications:**

- Carry all medication in original bottles, with the name of the drug and your doctor's name on the label
- Bring a copy of your prescriptions (generic and non-generic names) and medication regimen, including your physician's name and contact information
- Bring all your medication, for your entire trip, in your carry-on bag, and include snacks, water, or juice to take with meds
- Bring a replacement supply of prescription medications in case you are detained or your supply is lost
- If you are changing time zones, continue to take your medications as prescribed, with the same intervals between doses; consider wearing 2 watches: current time, and time at home

## **At Travel Terminals:**

- If necessary, request wheelchair or electric cart service within terminals (Your bags will be handled too)

- Check in early
- Utilize early boarding privileges, and, if necessary, special accommodation to get to your seat
- Request an aisle seat, and as close as possible to bathroom

**Air Travel Specifics:**

- Airline carriers must provide meet-and-assist service (e.g. assistance to gate or aircraft) at drop-off points
- Personal care assistants of passengers with disabilities are allowed beyond screener checkpoints
- The limit of one carry-on bag and one personal bag (purse) per traveler does not apply to medical supplies and/or assistive devices
- Assistive devices such as canes and wheelchairs are permitted on board
- People who require a wheelchair or scooter must have physician's written "certificate of need"
- People in wheelchairs can request private, rather than public, checkpoint screenings
- With documentation of medical need, and with proper labeling, syringes are permitted on board

For more information about traveling by air with a disability, contact the Transportation Security Administration at [www.tsa.gov](http://www.tsa.gov).

**Rail Travel Specifics:**

- Have valid photo identification
- Amtrak trains can accommodate most wheelchairs; Amtrak may make random checks of wheelchairs
- A service animal is allowed to travel with the passenger

For more information about traveling by train with a disability, contact Amtrak at [www.amtrak.com](http://www.amtrak.com).

**Bus Travel Specifics:**

- Greyhound buses are equipped with wheelchair lifts

## Travel Tips—continued

- Greyhound provides assistance with boarding, de-boarding, luggage, transfers, stowing and retrieving mobility equipment.
- Greyhound allows personal attendant to travel one-way at no charge (arrangements for a return ticket are made at the returning location)
- Service animals and oxygen and respirators are permitted

For more information about traveling by train with a disability, contact Greyhound at [www.greyhound.com](http://www.greyhound.com).

### **Ocean Cruise Specifics:**

- Ocean liners offer scooters for rent during cruises
- Determine in advance whether any ports of call require a license for a motorized wheelchair

### **Hotels:**

Ask specific questions. For example, what does “accessible room” actually mean? Is there a walk-in shower? Grab bars? What is the proximity to elevators?

### **General Reminders:**

- See fewer sites, enjoy them more
- Give yourself extra time for everything

*Don't let PSP, CBD or any other related atypical Parkinsonian disorders hold you back from the trip of your dreams! Bon Voyage!*

*Diane Breslow, MSW, LCSW  
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# Compassionate Allowances for PSP, CBD, and MSA

## **Expediting Disability Benefits**

The Social Security Administration (SSA) has added PSP, CBD, and MSA to its list of Compassionate Allowances (CAL) conditions.

CAL is a way of quickly identifying diseases and other medical conditions that meet SSA's disability standards, based on minimal objective medical information. Individuals diagnosed with PSP/CBD/MSA will receive a decision on their disability claim in a matter of weeks, rather than the months it normally takes.

The links below provide general information regarding the Compassionate Allowances initiative and SSA's disability programs:

- [www.ssa.gov/compassionateallowances](http://www.ssa.gov/compassionateallowances)
- [www.ssa.gov/dibplan/index.htm](http://www.ssa.gov/dibplan/index.htm) (Disability Planner)

## **Eligibility and Qualification**

In order to be eligible for disability benefits, you must meet the medical and non-medical criteria for the Social Security Disability Program (SSDI). For additional information about the SSDI program, please visit this website: [www.ssa.gov/dibplan/dqualify.htm](http://www.ssa.gov/dibplan/dqualify.htm)

CurePSP will not be able to answer any questions regarding eligibility for disability benefits or qualification via Compassionate Allowances. Questions regarding these matters must be directed to the SSA.

## **Applying for Benefits**

Individuals can apply for disability benefits immediately. As of December 2011, SSA's computer systems automatically identify applicants with these diseases and expedite applications.

### **There are three ways to apply for disability benefits:**

- Fill out and submit an online application: [www.ssa.gov/applyfordisability/](http://www.ssa.gov/applyfordisability/)

## Compassionate Allowances for PSP, CBD, and MSA—*continued*

- Call the SSA's toll-free telephone number at 1-800-772-1213; if you are deaf or hard of hearing, you can call the SSA at TTY 1-800-325-0778
- Call or visit your local Social Security office: [www.ssa.gov/locator/](http://www.ssa.gov/locator/)

If you have already applied for disability benefits, the Compassionate Allowances fast-tracking process should be used during the initial application or at the reconsideration, hearing or Appeals Council levels. It is advisable to also notify the disability examiner assigned to your claim that your condition has been selected for inclusion in the Compassionate Allowances program.

If you are planning to appeal a denial for disability benefits prior to December 10, 2011, you should include a statement on the reconsideration form that your condition has been selected for inclusion as a Compassionate Allowance. After December 10, 2011, your claim will be automatically identified and expedited.

After being approved for disability benefits and receiving benefits for 24 months, you will begin receiving Medicare benefits. SSA will automatically enroll you in Medicare. SSA begins "counting" the 24 month period based on the month you were first entitled to receive disability - not the month when you received your first check.

### **Retirement Benefits and Supplemental Security Income**

If you are 65 or older, you are eligible for Social Security retirement benefits, which are comparable to Social Security disability benefits. There is no reason to apply for disability benefits if you are in this classification. However, if you are receiving early retirement benefits (age 62-age 65), you should apply for SSDI because your benefits could increase and you may become eligible for Medicare before you turn 65.

You may also apply for Supplemental Security Income Program (SSI) benefits, which has different non-medical criteria, which are based on income. For additional information about the SSI program, please visit this website: [www.ssa.gov/ssi](http://www.ssa.gov/ssi)



# Support and Resources

Seeking help as a caregiver is not always an easy task. However, it is extremely important to take care of yourself in order to help those you love. Once you have realized that you are becoming overwhelmed, there are services that can provide respite for you.

## **Support Groups**

Support groups can make a significant difference in how you cope with stress as a caregiver. People involved in support groups are going through the challenges of the disease and are likely experiencing the same emotions. Being a part of a support group provides a sense of relief for many people and is a great way to share information as to what products, services and other resources that can help you and your loved one.

*For a complete listing of both online and face-to-face support groups, please visit [www.curepsp.org](http://www.curepsp.org).*

## **Resources That Can Help**

### **Adult Day Care**

Adult day care centers can give relief to the caregiver and provide their loved one with a safe place to engage in activities and socialize with others.

Adult day care centers are community-based group programs designed to meet the needs of functionally and/or cognitively impaired adults through an individual plan of care. These structured, comprehensive programs provide a variety of health, social, and other related support services during certain hours of the day (schedules vary) but they do not provide 24-hour care. Adult day centers generally operate programs during normal business hours five days a week. Some programs offer services in the evenings and on weekends.

*What are the benefits of adult day care?*

- A safe, secure environment
- Assistance with eating, walking, toileting, medications
- Exercise
- Nutritious meals, snacks or special diet

For more information about adult day care centers across the US, please contact the National Day Care Services Association:

[www.nadsa.org](http://www.nadsa.org)

877-745-1440

# Support and Resources—*continued*

## **Respite Care**

### **What Is Respite Care?**

Respite care is short-term care given to a hospice patient by another caregiver, so that a family member or friend who is the patient's caregiver can rest or take time off. This type of care was created to allow caregivers time away from administering care, with the goal to help the caregivers have lower stress and at the same time fill the needs of the individual receiving care. According to the Centers for Medicare & Medicaid Services, respite care comes from Medicare-approved facilities, like a hospital, nursing home, or hospice inpatient facility.

### **What Respite Care Does Medicare Cover?**

Respite care is covered in the same manner as hospice care; it includes medical supplies and medical care under a short-term stay at a facility. There are specific respite care facilities that serve persons receiving hospice care at home. The provisions of care that are normally paid for do not change for respite care. This means that the prescription and medical coverage stay the same. As in hospice care, respite care provided through Medicare does not include room and board for the individual's home or non-prescribed treatments, but it does cover a certain amount of the respite care stay. According to the Centers for Medicare & Medicaid Services, the person receiving respite care may be responsible for 5% of the Medicare-approved amount for respite care. For example, if Medicare pays \$100 per day for inpatient respite care, you will pay \$5 per day. Each time a person receives respite care, Medicare covers up to five days. There is no limit to the number of times that a person can receive respite care. The amount that you will pay for respite care can change each year.

### **Can I Get Respite Care for Any Person Who Needs Continuous Live-In Care?**

Medicare does not pay for respite care for the caregiver unless the patient is in hospice. However, other help may be available. Contact your local Area Agency on Aging (AAA) for possible respite care sources in your area. You can also contact Senior Services and Social Services. Senior volunteer services and private nonprofit agencies are the common providers of home-based respite care programs. The U.S. Department of Health and Human Services Eldercare Locator Services helps place caregivers with agencies and services in their area. Their telephone number is (800) 677-1116.

## **Types of Respite Care Programs**

### **In-Home Respite Care**

In-home respite care is temporary care provided in the person's home. This allows the family and patient to be comfortable and saves them from having to adjust to a new environment. Local Senior Services will have a list of approved caregivers who provide in-home respite care. Home-based respite care programs are pro-

vided through a nursing agency; you can find these in the telephone book or by contacting Senior Services and Social Services. Senior volunteer services and private nonprofit agencies are the common providers of home-based respite care programs.

### **Out-of-Home Respite Care**

Out-of-home respite care programs provide the opportunity for the family or caregivers to leave the person needing care at a facility, such as an assisted living center, nursing home, or hospital, depending on the level of care needed. It is important to remember that the person requiring care will have to be transported, and special medical equipment may also have to be moved. Contact senior services and ambulatory services to find out about special transportation needs, such as a wheelchair van. Residential facilities are most common for this type of respite program. Senior day-care facilities may be available in the area, but these are not as common or as available. You will want to find out the caregiver-to-patient ratio as well as the facility's licenses and bonds. Senior foster care homes may also be available; these are generally an individual's home that cares for seniors and disabled persons. Hospital-based respite care is also available depending on the local hospital's programs. Contact the hospital in your area to find out if they offer respite care.

### **How To Choose Respite Care**

The following offers a guideline of questions to ask when seeking respite care:

1. What types of special services are needed to provide care for the individual?
2. Does the patient/family prefer in-home or facility care?
3. What is the cost of care at the facility (or by the individual caregiver)?
4. What type of training does the respite care provider have?
5. Is the respite care provider/facility receptive to receiving specific instructions written or verbal?
6. Is the person/facility that provides care licensed and bonded for Medicare and other health insurance coverage?
7. What is the caregiver-to-patient ratio?
8. How can the person/facility provide care for special needs (medication, diabetes, physical requirements)?
9. Does the person/facility have experience providing the type of care necessary?
10. What activities/entertainment can the person/facility provide?

### **Home Health Care**

#### **What Is Home Health Care?**

The term "home health care" is used to describe a wide range of services provided to seniors in their home. They include medical and non-medical services. The purpose of the services is to keep people out of the hospital or nursing home, and as independent for as long as possible. The services provided can be as

## Support and Resources—*continued*

simple as help with daily activities such as cooking, cleaning, and bathing or as complicated as medical care that requires a licensed and skilled health care provider. Most people who require these services are recovering from an illness or injury, or are living with a long-term medical condition.

### **Is There an Advantage to Receiving Care in My Own Home?**

The good news is that today many types of medical treatment can be done in your own home. Home treatment is less expensive, more convenient, and just as successful as the care you get in the hospital or nursing home. In addition, it is better for you if you can stay in your own home.

- According to a 2006 study by AARP, ninety percent of older Americans want to stay in their own home and remain as independent for as long as possible
- According to the American Association of Homes and Services for the Aging in Washington, D.C., older Americans report they feel better and younger when they are able to receive assistance and medical care in their own home
- A 2008 article published by the Agency for Healthcare Research and Quality showed assisted living care in the home reduced the use of more expensive hospital services

### **What Kind of Services Can I Get in My Home?**

Today there are a full range of services that you may be able to have provided in your home. Some of the most common services used are:

- Help with personal care and mobility
- Transportation
- Meals On Wheels
- Case management
- Medical care in the home by a licensed health care providers

#### *Who Provides the Services?*

In most communities, there are a number of licensed home health care agencies that can provide any level of home care needed. Many people use a combination of family, friends, and professional services for their home care.

### **How Much Does Home Care Cost?**

Home care costs can vary depending on where you live, the type of care you need, and how often you need care.

### **Is Home Care Covered by Medicare?**

Medicare will cover some of the costs of home care. To receive coverage:

- The home health agency you choose must be Medicare-approved
- You must meet the qualification of your Medicare, Medicare Advantage or MediGap/Medicare Supplemental plan

In addition to Medicare, you may be eligible for other state and federal sources for coverage. For example, people with low income may be covered under their local Medicaid program. For those with limited income, there may also be local community programs that assist with daily living needs for seniors or individuals with certain disabilities. When contacting the provider be sure to ask if they can help you apply for the coverage for which you qualify.

### **How Do I Qualify For Medicare Coverage?**

To qualify for home health care, your doctor must write a plan for your home care. In addition:

- You must need at least one or more of the following:
  - Part-time nursing care
  - Physical therapy
  - Speech language therapy
  - Occupational therapy
- You must be homebound. This means that leaving your home is a major effort and you seldom do unless, for example, you:
  - Attend religious services
  - Leave to get medical treatment, including therapeutic or psychosocial care
- You must receive the services from a home health agency that is approved by the Medicare program
- You may also get care in an adult day-care program that is Medicare approved and state certified to provide adult day care services

If you have a Medicare Advantage Plan or MediGap/Medicare Supplemental policy, call your plan and ask about your coverage. You may have to use one of the home agencies that they have on contract.

### **What Do I need To Consider When Choosing a Home Health Agency?**

It is important to:

- Choose a home health agency that will meet your personal and medical needs and is affordable; make sure you choose an agency that is approved by Medicare or is

## Support and Resources—continued

- contracted with your Medicare Advantage or MediGap/Medicare Supplemental plan
- Include your doctor and family members or trusted friends in the process of picking an agency; ask your doctor, other health care providers, friends, and family for recommendations

Make a point of checking the health and safety record of the agency before making a choice. Call Medicare and ask them for any reports on the agencies in which you are interested: 1-800-MEDICARE (1-800-633-4227) 24 hours, 7 days a week, including some federal holidays. TTY/TDD users can call 1-877-486-2048. However, the interactive phone system is available 24 hours every day of the year.

## Advanced Planning: Business Everyone Should Take Care Of

Because we never know the circumstances when information might be needed, be sure to inform someone of the existence and location of your important documents. You may also wish to give photocopies of the documents to a trusted family member or your attorney for safekeeping.

### **Family Section**

The family section should contain addresses and telephone numbers for your home and office; the make, model, color and license number of automobiles; names, addresses and phone numbers of neighbors and nearest relatives who should be notified in case of an emergency. Include the location of an extra house key, electrical breaker box, water cutoff, thermostat, alarm system box and any instructions that would be helpful. List the names and phone numbers for each of the following: accountant, heating and air conditioning, ambulance service, insurance agents, appliance repair, investment counselor, attorney, lawn/yard care, banker, pharmacy, clergy, plumber, dentist, realtor, doctors and hospitals, security (alarm system), electrician, utility companies and veterinarian.

This section should include a brief personal history of each household member, providing information that would be needed in the event of emergency medical treatment or a death. Consent for emergency medical treatment and advance directives to physicians also belong in this section. Family members may wish to leave written instructions regarding funeral services, burial or cremation, etc., in this section.

Copies (not originals) of the following documents should be stored in the pocket dividers or envelopes in the family section: wills, trusts, military papers and discharge, disability verification, marriage licenses, any divorce or separation papers, social security cards, passports and birth certificates. Write the location of the original documents on the pocket divider. For wills and trusts, include the dates, names and telephone numbers for the attorneys who drafted them.

### Finance Section

The finance section should include: All bank accounts, including the name, number and type of each account. Note the name of the bank, a bank officer to contact, persons who are authorized to sign on the account, and where to find the checkbook or passbook; location and number of any safe deposit boxes; location of keys and persons authorized to sign for entry into the box and its contents; credit cards, including the bank or company issuing each card, a telephone number, and who is allowed to sign on the card; investments firms and stockbrokers, including addresses and telephone numbers. List each account name and number with the firm, the names of the stocks, bonds, certificates of deposit (CD) or other investments handled by the firm, the number of shares and the location of the certificates. Miscellaneous assets such as retirement plans, annuity contracts, gold coins, antiques or jewelry should be included.

The insurance section of your notebook can be divided into types of insurance, such as life, health, disability, automobile and homeowner. For each policy, you will need the name of the insured, the company, agent, policy number, where the policy is located and details of coverage and premium payments.

The property section should include a list and location of deeds and mortgage papers on any real estate you own. Titles and license receipts for automobiles, boats or other vehicles can also be listed in this category. As with other important legal documents, the originals should be stored in a fireproof place, and only copies placed in the pocket divider. Receipts for major property improvements should be stored in the envelopes in this section, as well as important information related to any vehicles owned. Some people may prefer to combine property and insurance into one category and folder.

### Business Section

The business section is reserved for information about any businesses you own or in which you have a financial interest. Information might include:

- Name and type of business
- Structure for legal and tax purposes (whether it is a sole proprietorship, partnership or corporation)
- Names, addresses and phone numbers for partners or associates
- Names, addresses and phone numbers for accountants, attorneys and other key consultants
- List of business bank accounts and numbers and contact information for an appropriate bank officer
- List and location of important business documents such as tax records, income and property records, canceled checks, receipts, keys, employee files
- Copies of employment contracts, partnership agreements, buy/sell agreements, etc.  
(Again, originals need to be more safely stored)

### Legal Documents

Among the important documents listed in the family section of your notebook are wills, living trusts, advance directives (living wills) and durable powers of attorney. A will allows you to legally define who your heirs are and what they will inherit upon your death. If you should die without a valid will, the law of the state in which you have permanent residence will determine how your property will be distributed. A valid will can also minimize the cost and hassle of distributing your estate for your heirs. Because laws change and circumstances change,



## Advanced Planning: Business Everyone Should Take Care Of<sub>—continued</sub>

your will should be reviewed periodically. A living trust is a revocable plan that provides for the management of your assets if you become disabled. It can also provide for distribution of your assets when you die and frequently avoids the delays and expense of probate. The trust becomes the legal owner of your property, and you manage the assets as the trustee. You can also name an alternate trustee to take charge if something happens to you. A living trust is revocable at any time, and can be amended when necessary.

A living trust can be used to manage most of your finances; however, a durable power of attorney may still be needed so that your designated agent can sign related legal documents such as insurance, retirement and pension forms. A durable power of attorney (POA) for finance allows you to appoint someone to act as your representative for financial and business matters. This representative is called your agent or attorney-in-fact. A “durable” power of attorney continues after the incapacity of the creator (you), whereas a “regular” power of attorney ceases upon the creator’s incapacity. Therefore, you should specify that the powers given to your agent continue even if you should become incapacitated.

The powers of your agent are determined by the terms of the durable power of attorney itself. A durable power of attorney is a valid document on its face, and the person in possession of it can present it to any authority, such as a bank or title company, and transact business on your behalf. It is wise to consider very carefully whom to appoint as your agent. Because the durable power of attorney gives your agent so much power, you may wish to keep it in your notebook to be accessed if needed rather than give it to the individual in advance. In addition to rectifying what powers your agent will have, you may also be able to specify special circumstances under which your agent’s powers take effect. For example, the durable power of attorney may specify that it does not go into effect unless a doctor certifies that you have become incapacitated.

Advance directives are of two types: a living will and a medical power of attorney (or durable power of attorney for health care). The living will, also called a directive to physicians, allows you to put into writing your wishes about medical treatment should you be unable to communicate your preferences near the end of your life. It allows you to state that you do not want artificial life support if you have a terminal condition. A living will can be revoked at any time, orally or in writing. As long as you are able to express your own decisions about your medical care, your advance directive will not be used. You can accept or refuse any medical treatment. Both state and federal law govern advance directives, but all 50 states and the District of Columbia have laws recognizing them.

A medical power of attorney is designed to allow a representative of your choosing to make routine medical decisions for you decisions. This representative, as your agent, can generally make health care decisions only after a physician certifies that you are unable to make these decisions yourself. Your agent is authorized to

make health care decisions at any time you are unable to do so, not just at the end of your life. Your agent is required to make health care decisions according to your wishes. If the agent is uncertain of your wishes, then decisions should be in your “best interest.” You can veto the doctor and the agent’s decision at any time, regardless of your mental state.

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## When to Hang Up the Keys

Our society values independence and the ability to drive is a sign of this independence.

But when a person becomes a danger to him/herself or others, it is time to consider giving up this privilege. Different physical and mental conditions may impair driving, and 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. Try to determine a person’s attention span, ability to process information and distance perception. Some of the warning signs of driving problems include:

- Hitting curbs
- Dents or scrapes on the car or garage
- Not anticipating danger
- 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
- Running stop signs
- 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 needs to remain focused on the self-respect of their loved one and the safety of others on the roadways.

# When to Hang Up the Keys—*continued*

## What to Do?

1. Begin discussions and planning early when there is a physical or mental impairment. Be sure to involve the driver in these conversations. Open and early communication can help to avoid a crisis later. Many caregivers permit their loved ones to drive longer than they know they should, causing caregiver 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 him/her. You are there to support the person and tell him/her you understand his/her feelings.
3. Never leave the keys out where they can be found.
4. Assure your loved one that he or she can depend on you to transport him/her wherever and whenever he/she needs to go.

## Re-Examination

If the impaired driver resents or fights turning over the keys, one option is to talk to the neurologist or family doctor. Each state differs in its procedures, but you or the neurologist can contact your Division of Motor Vehicles and supply the DMV with a Driver Medical Evaluation form. The driver will be notified of the need for re-examination. An in-person contact meeting is then needed to assess cognitive processes, perception and awareness. The driver will be asked questions concerning health, medical treatment, daily routine and need for assistance with daily activities. Further examination will include a test used to determine the driver’s mental competency as well as language and cognitive skills. A special driving test is next to assess the driver’s ability and competency in concentration, attention, perception and judgment. If the tests are all satisfactory, the driver will be issued an appropriate license and scheduled to re-test in one year.

For information on physical and mental impairments and driving, contact your local DMV and ask for the Regional Driver Safety Office. Explain the need for re-examination to your loved one. Discuss with him/her each part of the re-examination as described above. In many instances, once the person with driving difficulties learns about the re-examination tests, he/she opts not to take the tests for fear of not passing and then more willingly gives up the car keys, thus taking ownership in the decision to stop driving.

# When Is It Time to Get a Wheelchair?

## **A Wheelchair Can Be a Wonderful Liberator**

It is time to get a wheelchair when you are in danger of falling and being injured and you are no longer going on routine outings. When you get a wheelchair, it does not mean that you must use it all the time; use it for distance mobility. Many people believe that using a wheelchair reduces one's independence. Using this type of assistive equipment can actually do the opposite; it can increase one's independence. Visit an OT, PT or rehab specialist to be evaluated for the proper fitting of a wheelchair and appropriate accessories. You will be measured and weighed to determine seat depth, width, height, back height and wheel placement. Get your wheelchair from a medical equipment supplier that is certified with the National Rehab Tech Supplier.

## **The Wheelchair Prescription**

The process of getting a wheelchair usually starts with getting a prescription for a wheelchair from a physician; most of the time this prescription comes from your family doctor or internist. You are familiar with a prescription for a medication that you take to the drug store. These prescriptions are precisely written instructions about the quantity, the dose and the brand that your doctor wants you to take. You take this prescription to the drug store and you get exactly what the doctor ordered. The prescription for a wheelchair is usually very different. Instead of getting very specific, the doctor usually writes a prescription for "a wheelchair." Since there are many options and characteristics of wheelchairs, and since most physicians do not know much about wheelchairs, this leaves the process wide open for other people to get involved and, possibly, for you to be less than satisfied with the outcome.

## **The Wheelchair Evaluation Process**

A wheelchair evaluation can be a good solution to this problem. An evaluation should start with the consumer and his/her needs, as well as some questions about daily routines and lifestyle. It should involve the consumer all the way through the process. It should conclude with a recommended wheelchair and seating system. Evaluations may be required by a reimbursement source, whether that is Medicare or private health insurance, as a condition for payment. Because the outcome can be so much better, many funders are starting to require them.

Occasionally, physicians who are specialists in physical medicine and rehabilitation (often called "physiatrists"), actually know quite a bit about wheelchairs and can write a more detailed prescription or may get more involved in the selection process. Otherwise, if the service is available, many physicians refer their patient to a rehabilitation professional (like an OT or a PT). This professional takes the lead in helping to determine exactly what kind of wheelchair is right for you. They are skilled at assessing your strengths and limitations and finding out about your lifestyle and your expectations for activities. They are also familiar with the brands and cate-

## When Is It Time to Get a Wheelchair? *—continued*

gories of wheelchairs that match your needs. If you will be spending long periods of time in your wheelchair, or if you have specific posture needs, your prescription may need to include some additional information. If you are working with a rehabilitation professional, he/she can request that the physician be more explicit in writing your prescription. If special features are needed in a wheelchair, most funding sources require that the prescription describe them explicitly. Some examples of additional things that may need to be written into a prescription are a pressure relieving cushion, a solid seat or back, brake extensions, special push rims, a one-arm drive, or being low to the floor to enable pushing with the feet.

### **The Evaluation Team**

A wheelchair and seating team is usually comprised of:

- A consumer and maybe someone from his/her family or an advocate
- An occupational therapist or a physical therapist
- A rehabilitation technology supplier
- Sometimes, a rehabilitation engineer

### **The Consumer as the Focus**

You, the consumer, should be the person who guides the direction of the evaluation team. The whole point of the evaluation process is to get the right or the best wheelchair into the hands of the person who will use it every day. It is something that must function in many environments: the house, the grocery store, the neighborhood, at church and out in the community.

A wheelchair, however, is not a car or a bicycle that will be parked somewhere and used only to move beyond the house. It must be appropriate for full-time use because it must either substitute for, or augment, moving around on legs. It needs to just right for you. It needs to be easy to maneuver, well-fitting, comfortable, durable and safe, and it must not contribute to future problems, like shoulder injuries. The wheelchair that is chosen at the end of the process should be something that allows you to do the things you want to do in your everyday life. For that reason, it is good to think of a few things ahead of time. It is the consumer's job to be as clear as possible about what he/she wants from a wheelchair.

- Where will I use my wheelchair most?
- When will I use my wheelchair occasionally?
- What kinds of activities that I do (or did) everyday are most important to me to get back to doing?
- How will I get my wheelchair (and myself) from place to place?
- How much of the day will I be spending in this wheelchair?
- How will I transfer from the wheelchair to other surfaces?

- If I will need help with my wheelchair, who will that come from, and what features about my wheelchair are important to them?
- How will I get my wheelchair around my neighborhood or yard?
- What kind of surfaces or slopes are involved?

The next member of the wheelchair team would be the occupational therapist (OT) or the physical therapist (PT). These members should be registered in their profession or licensed by your state government. You can find out more about registration and licensure at the American Occupational Therapy Association or the American Physical Therapy Association. If your clinician is certified as an Assistive Technology Provider (ATP), this is an excellent credential and means that he/she has completed additional training and passed a certifying examination from RESNA. (You can even use their Web page at [www.resna.org/cert/index.htm](http://www.resna.org/cert/index.htm) to possibly find a credentialed provider in your area.) Years of experience and the “word of mouth” reputation of a therapist among people who use wheelchairs can also be helpful indicators about expertise. Besides helping to determine what type of wheelchair is right for you, it is the therapist’s job to write the letter of justification that makes sure the insurance company understands the relationship between your mobility needs and the equipment that is recommended for you.

The other important team member is the rehabilitation technology supplier. This person is sometimes called a “vendor” or a “durable medical equipment” dealer. These names do not give credit to the high level of professionalism and knowledge of some rehabilitation technology suppliers (RTS) professionals. Like therapists, it is possible and desirable for an RTS to be credentialed by RESNA or by NRRTS. This means that they he/she has received additional training and is committed to a code of ethics. It is the job of the RTS to know all about specific kinds of wheelchairs, to know the equipment features and manufacturers, and to be able to compare the characteristics of wheelchairs or cushions.

Occasionally, a rehabilitation engineer is on the team, especially in an evaluation center that works with clients who have more complex physical disabilities. Rehabilitation engineers are experts at customizing equipment that is commercially available or at fabricating something completely unique. It is their job to know about loads, forces, torques, movements and the interaction of those things with your body and your wheelchair. All of these things affect how your wheelchair will perform for you over time.

### **The Evaluation Summary**

The end product of the team’s evaluation should be some type of evaluation report that summarizes the findings and the recommendations from the evaluation team. It will probably be sent to your physician, with a copy kept in the evaluation team’s files as well. Then, one of two things can happen. The doctor can write a letter of medical necessity, or the therapist can write a letter of justification.

# When Is It Time to Get a Wheelchair? *—continued*

## **Letter of Medical Necessity or Justification**

A letter of medical necessity is usually written by a physician and is addressed to the third-party payer. It tells the payer that a piece of equipment (usually some kind of medical equipment) is needed because of an authentic or verifiable medical condition or impairment. A letter of justification is usually written by a person very familiar with the consumer/client and the product recommended. Usually, it is a therapist, but in some cases, experienced rehabilitation technology suppliers write them. This kind of letter correlates the recommendations that come out of the evaluation to the features of a recommended wheelchair or seating system to “paint a picture” for the payer. The letter helps the third-party payer understand why certain features or characteristics of the recommended equipment are important. It describes the relationship between product features and the anticipated functional outcome for the individual or the consumer. It should tell what the consumer will be able to do as a result of having the equipment.

It is important that the items being requested really are medically necessary. This might sound obvious, but the letter writer would be abusing the funding system or third-party payer if he/she requested equipment that a client just wanted to have but did not actually need. A letter of justification also helps third party payers realize why it might be better to spend a little more money for a certain feature on a wheelchair now in order to avoid a more costly expense later. A letter of justification is an expert opinion about what is best for a particular consumer. Having a therapist or supplier who is good at writing this kind of letter provides a valuable resource for helping you get the kind of equipment that you really need.

## **Caring for a Person in a Wheelchair**

“Walking people” often “down-talk” to people sitting in a wheelchair. Try to be more aware of your body language. Kneel or stoop down to meet the person’s eye level when talking to him/her. Also, try to be more aware of the needs of wheelchair riders, especially if they cannot propel themselves and/or if they have language problems, sensory impairment (lack of sight, hearing and feeling) or memory problems, or are unable to mentally understand instructions. Check the position of the person in the wheelchair to prevent pressure sores and injuries. The person should sit in the most neutral position possible. Ensure that both feet are placed properly on the foot plates. Make sure that the fingers cannot be caught in the wheels (place the hands into the lap or use a table top for those persons who have no control over their upper limbs). Ask the person in the wheelchair whether he/she is comfortable.

## **How to use a wheelchair**

Unfold wheelchair: Most wheelchairs are unfolded by pushing down and out on the side edges of the seat, keeping the fingers inwards (never put your fingers between the frame and the seat; you could injure your-



self). Fold wheelchair: Turn the footplates upright (if detachable model, rotate or remove footplate). Pull the seat upward or use the grab handles. Before using wheelchair, always make sure that the chair is in good working condition (brakes are okay, tires are firm, etc.). Before transferring someone into the chair, make sure that the brakes are locked, and lift up or swing away the footrests to prevent the person from falling over the footplates. If you have a detachable model, take out the armrest nearest to the person for an easier transfer. Encourage the person to sit back into the chair or assist him/her. Be sure the person sits in the most neutral position possible and that his/her fingers cannot be caught in the wheels. When approaching a narrow doorway, also ensure that the person's elbows are inside the wheelchair to prevent them from knocking the door frame.

### **Going Over Curbs**

Place the wheelchair at a right angle facing the curb. Stand behind the wheelchair while holding onto the push handles. Step onto the tilting bar at the rear of the wheelchair, and tilt the wheelchair carefully backward (towards you). Lift the front casters onto the curb, and push the chair forward on its back wheels until they touch the curb. Gently lower the chair until the front casters touch the floor, and lift the back wheels onto the curb. To go down a curb, just do the opposite – stand with the back of the wheelchair at a right angle to the curb. Step down, and again tilt the wheelchair by stepping onto the tilting bar. Wheel the chair backward down (on its back wheels), and again gently lower the chair until its front casters touch the floor.

### **Ascending Stairs**

Two persons are required for safety. When going up stairs, place the wheelchair backwards at a right angle to the stairs. Tilt the chair by stepping onto the tilting bar. The person holding onto the push handles is the leading person. The second person holds onto the wheelchair frame (do not hold onto any detachable parts). The leading person gives the command. Step by step, the leading person pulls the chair up, and the second person pushes the chair up, preventing any unwanted downward movements.

### **Descending Stairs**

Place the wheelchair backwards at a right angle to the stairs. Now, the lead person is the one going down first (the person holding onto the wheelchair frame), and the person at the push handles moves forward with a pulling action to prevent the chair from “running” down. Here again, tilt the chair and move forward, step by step, going down.

*Attribution to [www.wheelchairnet.org](http://www.wheelchairnet.org).*

## When Should Hospice Be Contacted?

There are basically two separate Medicare benefit programs that may be available for people with atypical Parkinsonian diseases and their families. These include the Medicare Home Health Benefit and the Medicare Hospice Benefit. Many private insurances have guidelines for qualifying for their own programs, but quite often, they are virtually identical to those offered through Medicare. It is usually worthwhile to review the publications available from the insurer and then speak with the benefit administrator to see what is available.

Each of the two plans has separate criteria which need to be met in order to qualify for the program. For the Medicare Home Health Benefit there must be a need for skilled care (custodial care alone, such as would be provided by a nurse's aide, generally would not qualify), and the patient must be home bound. In the case of the Medicare Hospice Benefit, both the admitting physician and the hospice medical director must certify that they believe if the disease runs its normal course, the patient has a prognosis of six months or less. With many diseases that have an unpredictable rate of progression, and PSP, CBD and other related atypical Parkinsonian disorders are no exception, determining a six-month prognosis with any true accuracy is extremely difficult. In consideration of this, the Medicare Hospice Benefit provides for unlimited renewals. Basically, this means that provided the admission criteria is still met, a person could potentially be eligible to receive all the care and benefits that hospice provides for well beyond the original six-month prognosis.

Another question I am often asked is, "When is it the appropriate time to contact hospice?" People are sometimes taken aback by my most common response, which is often, simply, "today." The reason I feel this is the most accurate answer is that by contacting hospice today, you have absolutely nothing to lose, but a priceless amount of information, support and services to gain. When contacted, many hospices will give you the option of having a nurse come to the home (or nursing home if that is where the patient resides) and explain the benefit. The nurse can often tell you on the spot whether the hospice benefit may be available as an option now, or, if not, what criteria would need to be met in order to qualify. Upon accessing the hospice benefits, a registered nurse will be assigned whose focus will be on controlling the symptoms of the disease and helping to promote the best quality of life possible.

The nurse will come to the home (usually from one to seven times per week, depending on need) for ongoing symptom management. There is also a registered nurse available 24 hours a day by phone for the hours that the assigned nurse is not available. A social worker will also be assigned who can assist in obtaining any available community resources, as well as helping both the person with the disorder and the family deal with the emotional aspects of the losses this disease can bring. A non-denominational pastor can also be assigned who can work alone or in conjunction with community clergy to help cope with the spiritual aspects of dealing with the disease. In addition, nurse's aides can be included to assist with personal care, such as bathing and dressing. Nurse's aides generally visit from two-seven days a week, depending on need, and stay from 1-1 1/2 hours per visit. Trained volunteers can also become involved. They can help by making friendly visits to sit and read to the patient, running errands, assisting with rides to appointments or helping in any other way possible.

Other services, such as speech or physical therapy, can also be included as part of the hospice plan of care.

By invoking the benefit, you gain access to a team of well-trained professionals whose focus will be on providing the person with the absolute best quality of life possible. In addition to the professionals involved in the care, hospice also covers related medications as well as home medical equipment, such as walkers, wheelchairs, commodes, hospital beds and other equipment. An additional positive aspect of the hospice benefit is that it can be provided not only in the home setting, but also in nursing facilities and hospitals.

Often, people have other insurance in addition to Medicare, such as Medicaid or long-term care insurance. If this is the case, the additional insurance can sometimes be used to cover the cost of being in a nursing facility, while Medicare is used for the hospice services. Some patients choose to use hospice houses, which are facilities that deal exclusively with hospice patients and often strive to create a more home-like environment as opposed to a medical one. Of all the families I have had the pleasure and privilege of being involved with, the ones who have gained the most from the program all had one basic thing in common: they accepted all of the services and benefits hospice had to offer. Although there is no obligation to accept the involvement of all of the different team members, I strongly encourage doing so. Each member has something different to offer that often can compliment what the others provide.

Hospice is a benefit that is available much sooner than most people realize. Referrals for hospice evaluations can be made by patients, friends or family members, and can be called in directly to any hospice in your area. The service does not need to be initiated by a physician's office, but it is often helpful to find out which hospices your doctor recommends.

*William Carroll, RN, CHPN  
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### **Hospice and Medicare Benefits**

Medicare Part A covers hospice care if you meet all of the following conditions:

- You are eligible for Medicare Part A
- Your doctor certifies that you're terminally ill and are expected to have less than 6 months to live\*
- You accept palliative care (for comfort) instead of care to cure your illness
- You sign a statement choosing hospice care instead of routine Medicare-covered benefits for your terminal illness

## When Should Hospice Be Contacted? *—continued*

Medicare will still pay for covered benefits for any health problems that aren't related to your terminal illness.

*\* In a Medicare-approved hospice, nurse practitioners aren't permitted to certify the patient's terminal diagnosis, but after a doctor certifies the diagnosis, the nurse practitioner can serve in place of an attending doctor. You can continue to get hospice care as long as the hospice medical director or hospice doctor recertifies that you're terminally ill.*

Hospice care is usually given in your home. It includes the following services when your doctor includes them in the plan of care for palliative care (for comfort) for the terminal illness and related conditions:

- Physician and nursing services
- Social work services
- Counseling services
- Hospice aide or homemaker services
- Physical, occupational, or speech-language pathology therapy services
- Drugs and medications for pain or other symptoms
- Medical supplies and durable medical equipment
- Short-term inpatient care for symptom relief, or for respite care\*
- Any other services normally covered by Medicare to provide care for the terminal illness and related conditions

\* Respite care is inpatient care given to a hospice patient so that the usual caregiver can rest. You can stay in a Medicare-approved facility, such as a hospice facility, hospital, or nursing home, up to 5 days each time you get respite care.

In 2011, YOU pay \$0 for hospice care. You may need to pay a copayment of up to \$5 per prescription for outpatient prescription drugs for symptom control or pain relief (always check with Medicare for any changes in this benefit). Medicare doesn't cover room and board when you get hospice care in your home or another facility where you live (like a nursing home). If your attending doctor isn't employed by the hospice, you pay your usual Part B deductible and copayment for his or her services.

If the hospice staff determines that you need short-term inpatient care in a hospice facility, hospital, or nursing home, or if your caregiver needs a short period of respite, Medicare covers the costs for room and board. You pay 5% of the Medicare-approved amount for inpatient respite care.

*Information provided by: [www.medicare.gov](http://www.medicare.gov)*

# The Importance of Brain Tissue Donation

Well-known actor Dudley Moore announced in 1999 that he had PSP and he died with that diagnosis in 2002. Since then, CurePSP's website and their multi-faceted efforts to educate the public have increased the awareness of PSP, along with CBD and MSA.

In recent years, researchers have learned a great deal about these diseases, but there is still much more to discover. At this time we still don't know what causes the diseases or how they are triggered. Investigators are searching for the answers to these and many other puzzling questions about neuromuscular diseases in their efforts to find effective treatments and ways to prevent PSP, CBD and MSA. When tissue from people living with a disease is compared to complete information about the donor's medical history, researchers are able to piece together information that will eventually provide some of the answers.

Persons with these diseases and their families often realize the importance of research because understanding the cause of the diseases may offer the best opportunity to find and produce effective therapies to treat and/or prevent disease in the future. For some people, the decision to donate tissue at death may be difficult, especially when coping with the day to day challenges of living with the disease. However, many also realize that being a tissue donor provides a sense of purpose in knowing that the donation will contribute to the quest for knowledge. And since the only way to definitively confirm a clinical diagnosis is through autopsy, we strongly encourage you to consider brain tissue donation. And we recommend just as strongly that you make this decision and execute your plans well before death is imminent! It is not always easy to find a medical facility to harvest the tissue in a timely manner.

The CurePSP Brain Tissue Donation Program brochure at [www.curepsp.org](http://www.curepsp.org) will help you to understand the process involved in tissue donation as well as answer some of your general questions.

Tissue sent to us is first used to provide the family with an autopsy report confirming the clinical diagnosis as well as detailing any other changes that were happening in the brain of the patient. This report is sent to the next-of-kin within 4-6 weeks after the tissue has been received at the brain bank. The tissue is then used by researchers as they develop protocols to determine the causes and find the cures for some of these diseases. Research is how we hope to aid other families and individuals who will face the challenges of living with neurological diseases in the future.

## **There are three important parts of the tissue donation process:**

### **Collecting the tissue is the most critical step because it must be done within less than 24 hours of death.**

All concerned parties need to be aware of the family's plan at the time of death. This includes the nursing home or hospital, hospice, the funeral home and the pathologist. If arrangements are made well in advance of the anticipated death, it will prevent such problems as the availability of a pathologist, and the family will be able to grieve their loss without even having to think about the process.

Locating a pathologist or diener (pathologist's assistant) in your area that will be able to collect the tissue within

## The Importance of Brain Tissue Donation—*continued*

less than 24 hours of death, even after business hours or over the weekend, can be difficult (see the instructions in CurePSP's Brain Tissue Donation brochure). If there is a large hospital near you that has a pathology department or a hospital that the patient has used, someone there may be able to perform the procedure. The patient's neurologist may be able to locate a pathologist or the coroner's or medical examiner's office may be able to help—they will sometimes do what they call a private autopsy, although this varies from state to state. If there is a nearby university with a medical school, their staff may be able to help. If all else fails, we at the Brain Bank can try to help locate someone.

**The second issue is the cost of the tissue collection.** There are some agencies that do not charge for the procedure because they realize how valuable the tissue is for research. But there are also some agencies who will request an unreasonable payment. We have found that \$500 is the normal fee for the service. Should that cost be an issue that may prevent the donation from happening, CurePSP has a very generous donor who has made funds available for such instances.

**The third issue is the importance of having copies of the patient's neurology medical records available for the autopsy session and to help in the development of research protocols.** We maintain Advance Records files to help make the process easier for the family. As soon as the decision has been made to donate, we encourage you to complete the forms in the brain donation packet including the Release of Information form which should be sent to the physician who diagnosed and followed the progression of the disease. Once the paperwork has been completed, you are free to enjoy the time left with your loved one.

Once the tissue collection has been accomplished, we take over at the Brain Bank. We ship containers and all necessary materials to the pathologist along with Federal Express labels so we can track the tissue as it is being transported to Mayo Clinic in Jacksonville, Florida. Dennis W. Dickson, M.D. performs all autopsies once the tissue has been prepared for the procedure (normally after about two weeks). Then slides are prepared, stains are completed and analysis of the tissue is done by Dr. Dickson who then writes the final autopsy report. A copy of that report is sent to the legal next-of-kin confirming the clinical diagnosis and reporting any other changes that may have been occurring in the brain. This process is normally completed within 4-6 weeks after the autopsy.

When the autopsy report has been completed, the tissue will continue to be used in support of our educational and research activities. The Human Genome Project makes genetic research possible in addition to our ongoing cellular and biological studies.

**The bottom line for us:** There will never be a cure for these unfair diseases without tissue donation. CurePSP is making it possible for world-renowned researchers to ultimately uncover the mystery of PSP, CBD, and MSA.

*Beth M. Marten*  
*Brain Bank Coordinator*  
*Mayo Clinic, Jacksonville, Florida*

# 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, and other functions of daily living.

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

**Akinetic:** Referring to absence 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 "senile plaques" and "neurofibrillary tangles") and reduced activity of acetylcholine and other 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 bodily cells. 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, or infection. It may also occur as a result of impaired development of the cortex as in certain neuro-developmental disorders, including Rett syndrome. Apraxia may affect almost any voluntary movements, including those required for proper eye gaze, walking, speaking, or writing.



## Glossary of Terms—*continued*

**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:** Several large clusters of brain cells, including the striatum and the substantia nigra, deep in the cerebral hemispheres.

**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. BTX-A is currently the only form (i.e., serotype) of botulinum toxin approved for clinical use. (BTX-A [BOTOX<sup>®</sup>] is produced by Allergan, Inc. and used in the United States and many other countries. Outside the U.S., it is available as Dysport<sup>®</sup> from Ipsen, Ltd.) 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. BTX-A is now increasingly being used as a therapeutic option for selected patients with other disorders characterized by severely increased muscle activity (hyperactivity), such as tremor, other focal dystonias, and spasticity. BTX-B is currently under investigation (by Athena Neurosciences, Inc.) for patients with cervical dystonia.

**Brainstem:** The region of the brain consisting of the medulla oblongata, pons, and midbrain. The brainstem primarily contains white matter interspersed with some gray matter. This area of the brain serves as a two-way conduction path, conveying nerve impulses between other brain regions and the spinal cord. In addition, most of the 12 pairs of cranial nerves from the brain arise from the brainstem, regulating breathing, digestion, heartbeat, blood pressure, pupil size, swallowing, and other basic functions.

**Bradykinesia:** Slowness of movement.

**Bradyphrenia:** Slowness of thought process.

**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.

**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), plays an essential role in the coordination and control of the entire body.

**Cerebellum:** A large structure at the lower back part of the brain responsible for the coordination of movement and balance.

**Cerebral Cortex:** This information is processed and then relayed (by way of the thalamus) to areas of the brain responsible for controlling complex motor functions. The caudate nuclei are specifically thought to process and transmit cognitive information that influences the initiation of complex motor activities.

**Cerebrospinal Fluid (CSF):** The fluid that flows through and protects the 4 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 particular 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).

**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 comprised of DNA. Deoxyribonucleic acid or DNA carries genetic information involved in directing cellular activities, thus controlling bodily growth and functioning and determining the expression of inherited traits.

**Cognition:** This involves thinking skills 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 health-care systems, practices, and products that are not presently considered to be part of conventional medicine.

**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.

**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 state of frenzied excitement or wild enthusiasm.

**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 due to 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

## Glossary of Terms—*continued*

**Dementia—*continued*:** 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. (CSF flows through and protects the ventricles of the brain, the spinal canal, and the space between layers of the membrane [meninges] enclosing the brain and spinal cord.) Normal-pressure hydrocephalus is associated with dementia, gait disturbances, and an inability to control urination (urinary incontinence).

**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 is known as a neurotransmitter. Neurotransmitters help relay messages from one nerve cell to another. Dopamine is especially important in relaying messages about movement.

**Dopamine Agonist (DA):** A drug that acts like dopamine. DAs combine with dopamine receptors to mimic dopamine actions. Such medications stimulate dopamine receptors and produce dopamine-like effects.

**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. At least five types have been identified including D1, D2, D3 receptors and the dopamine autoreceptor.

**Double-Blind 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.

**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.

**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 particular 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, with tremor occurring in a “no-no” horizontal pattern in about three quarters of patients and the remainder affected by vertical “yes-yes” tremors. Less commonly, patients have tremor involving the voice, tongue, or roof of the mouth (palate), leading to impaired articulation of speech (dysarthria). Rarely, tremor may affect the trunk or lower limbs, particularly with advanced stages of disease. 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.

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

**Food and Drug Administration (FDA):** A federal agency charged with ensuring that the food supply in the United States is safe and wholesome, that cosmetics are not harmful, and that medicines, medical devices, and radiation-emitting consumer products are safe and effective.

**Free Radicals:** Unstable molecular fragments that can damage cells. One hypothesis holds that free radical formation in the substantia nigra causes the loss of nigral cells (leading to Parkinson’s disease).

**Gait:** The style or manner of walking. Gait disturbances may be associated with certain neurologic or neuromuscular disorders, 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:** A plastic tube inserted into the stomach through a surgical incision in the abdomen. A gastrostomy tube is used to deliver liquified food to the digestive system when swallowing becomes dangerous or difficult.

## Glossary of Terms—*continued*

**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. A gene is a short segment of DNA, which is interpreted by the body as a plan or template for building a specific protein.

**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 half life of a drug is the time it takes for the blood level to decrease by half after a drug is stopped.

**Hereditary:** Inherited; inborn; referring to the genetic transmission of a trait, condition, or disorder from parent to offspring.

**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.

**Homeostasis:** This exists when an organism's internal state of being is in balance even when the external environment is changing.

**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 or, less commonly, appear to occur randomly for unknown reasons (sporadically). The disorder results from abnormally long sequences or "repeats" of certain coded instructions (i.e., unstable expanded CAG repeats) within a gene (located on chromosome p16.3). 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.

**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, and loss of function.

**Inhibition:** The restraint, suppression, or arrest of a process or the action of a particular cell or organ; the pre-

vention 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 particular 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 crosses the blood-brain barrier and is converted by the body to dopamine. A loss of dopamine-producing nerve cells in the part of the brain that controls movements leads to the symptoms of Parkinson’s disease.

**Lewy Body Disease:** Also called diffuse Lewy body disease, 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. The Lewy body is a protein aggregate found in dying neurons in the brain. In Lewy body disease, the Lewy bodies are most prominently found in the cortex, or surface of the brain, versus in the midbrain for Parkinson’s disease.

**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.

**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:** A neurodegenerative disorder characterized by parkinsonism, ataxia, and dysfunction of the autonomic nervous system.



## Glossary of Terms—*continued*

**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.

**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) through the use of 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.

**Neurotoxin:** A substance that interferes with the electrical activity or 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.

**Nucleus:** The part of the cell that contains the genetic material; it is surrounded by the nuclear envelope.

**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 build up in the cells as the cells convert nutrients into energy. The free radicals damage different parts of the cells in a process that is similar to the way in which rust builds up on metal. The free radicals can be counteracted by antioxidants, but if your cells do not have enough antioxidants, the free radicals accumulate and cause damage. Coenzyme Q10 acts as a scavenger of free radicals.



**Parkinson's Disease (PD):** A slowly progressive degenerative disorder of the central nervous system 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 (central nervous system).

**Placebo:** A substance that appears to be identical to the treatment under study but that has no effects on the test subject. The placebo is given to members of the control group during experimental trials that test the effects of a drug or other substance.

**Positron Emission Tomography (PET):** An advanced, computerized imaging technique that uses radioactively-labeled substances (e.g., glucose) to demonstrate chemical and metabolic activities in the brain as well as track other brain functions.

**Postural Instability:** Unsteadiness of gait or standing.

**Prevalence:** Refers to the number of people in a given group or population who have a disease.

**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.

**Rigidity:** Stiffness and resistance to movement. May be a symptom of a neurologic movement disorder such as Parkinson's disease.

**Sialorrhea:** Excess production of saliva, or increased retention of saliva in the mouth, due to 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.

**Striatum:** An area of the brain that controls movement and balance. It is connected to and receives signals from the substantia nigra.

## Glossary of Terms—*continued*

**Substantia Nigra:** A dark band of gray matter deep within the brain where cells manufacture the neurotransmitter dopamine for movement control. Degeneration of cells in this region may lead to a neurologic movement disorder such as Parkinson's disease.

**Tauopathy:** Refers to any group of diseases that cause dementia related to a problem with tau, a protein that is important in maintaining the structure of brain cells.

**Toxin:** A poisonous substance that is produced by a plant or animal.

**Tremor:** Rhythmic, involuntary, oscillatory (or to-and-fro) movements of a body part.

*Attribution to WeMove.org*

## Caregiver Glossary

**Activities of Daily Living (ADLs):** Include bathing, dressing, transferring, toileting, eating, walking, and control of bowel/bladder. They are descriptions of physical functions that are useful tools when planning helping services for older persons.

**Adult Day Health Care Centers:** Adult day health care 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.

**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 persons 60 years and older.

**Assisted Living:** Provides 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. [www.ataporg.org/atap/index.php](http://www.ataporg.org/atap/index.php)

**Case Management:** The primary goal of case management is to maintain the highest functioning, safety and independence of each person by linking him/her 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 coordinating and monitoring services, as well as intermittent reassessment of the client's situation. Case management may be licensed or certified by various groups depending on the funding agency's standards.

**Certified Nursing Assistants (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.

**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.

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

**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.

**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.

**Geriatrician:** A physician who has had basic postgraduate training in either internal medicine or family medicine with an additional 1-2 years training in medical, social and psychological issues that concern older adults.

**Home Health Care:** A skilled health care 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 (non-medical) services provide assistance with housekeeping services, personal care, and Activities of Daily Living (ADLs), all of which help the older adult to remain safely at home. These services may

## Caregiver Glossary—continued

also include light housecleaning, meal preparation, doing laundry, grocery shopping and running errands. Additional non-medical homecare services include feeding, bathing, bowel and bladder care and dressing.

**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 choose 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.

**Long-Term Care (LTC) :** An umbrella term referring to comprehensive health care 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).

**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 health care, lab tests, radiographs, family planning, early and periodic screening, diagnosis and treatment.

**Medicare:** A federal health insurance program for persons 65 years of age and over, persons considered permanently disabled for purposes of the Social Security Act, and persons with end-stage renal disease. Medicare is divided into two parts: Medicare Part A and Medicare Part B. Part A, Hospital Insurance Benefits, provided 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 also care given by home health agencies and hospices. No premium is required from persons 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 he or she does not want Medicare B. Medicare B covers outpatient services, physician visits, ambulance transportation and durable medical equipment. It also covers some home health care. 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 and 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.

**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.

**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, participating in shaping health policy, health systems management, and health education.

**Nursing Home :** Also known as skilled nursing facilities and long-term care facilities, are licensed by the State Department of Health Services 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” of who someone is and what they 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 care partners to problem solve and create new ways to make routine activities easier and safer.

**Physiatrist:** A physician with additional postgraduate training in physical medicine and rehabilitation.

**Physical Therapists (PTs):** Health care 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.

**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.

## Caregiver Glossary—continued

**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 6-8 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.

**Senior Center:** A community facility for the organization and provision of a broad spectrum of services, including provision of health, social and educational services, and recreational activities for older persons.

## Resource Directory

### A

- AARP ( American Association of Retired Persons) • 800-424-3410 • [www.aarp.org](http://www.aarp.org)
- American Massage Therapy Association • 847-864-0123 • [www.amtamassage.org](http://www.amtamassage.org)
- American Occupational Therapy Association • 301-652-2682 • [www.aota.org](http://www.aota.org)
- American Parkinson Disease Association • 800-223-2732 • [www.apdaparkinson.org](http://www.apdaparkinson.org)
- American Speech-Language Hearing Association • 800-638-8255 • [www.asha.org](http://www.asha.org)
- Attainment Company, Inc. (specialized clothing/dressing aids) • 800-327-4269  
[www.attainmentcompany.com](http://www.attainmentcompany.com)

### B

- Benign Essential Blepharospasm Research Foundation • 409-832-0788 • [www.blepharospasm.org](http://www.blepharospasm.org)
- Biotene (dry mouth) • 800-922-5856 • [www.laclede.com](http://www.laclede.com)
- Botox • 800-44-BOTOX • [www.botox.com](http://www.botox.com)
- Bruce Medical Supply • 800-225-8446 • [www.brucemedical.com](http://www.brucemedical.com)

### C

- Calmoseptine, Inc. • 800-800-3405 • [www.calmoseptineointment.com](http://www.calmoseptineointment.com).
- Caregiver's Marketplace • 414-355-1330 • [www.caregiversmarketplace.com](http://www.caregiversmarketplace.com)
- Centers for Medicare/Medicaid Services Regional Administrator • 800-633-4227 • [www.cms.gov](http://www.cms.gov)

- Children of Aging Parents • 800-227-7294 • [www.caps4caregivers.org](http://www.caps4caregivers.org)
- Comfort Cape • 843-347-0167 • [www.comfort-cape.com](http://www.comfort-cape.com)
- Compassion in Dying • 800-247-7421 • [www.compassionandchoices.org](http://www.compassionandchoices.org)

### **D**

### **E**

- Eldercare Locator • 800-677-1116 • [www.eldercare.gov](http://www.eldercare.gov)
- Eldercare Web • 309-451-3319 • [www.elderweb.com](http://www.elderweb.com)

### **F**

- Family Caregiver Alliance • 800-445-8106 • [www.caregiver.org](http://www.caregiver.org)
- *The Fearless Caregiver: How to Get the Best Care for Your Loved One and Still Have a Life of Your Own*, Barg, Gary, Capital Books, Inc. • 800-829-2734
- Five Wishes, 888-594-7437 • [www.agingwithdignity.org](http://www.agingwithdignity.org)

### **G**

### **H**

- Healthcraft Products • 613-822-1885 • [www.healthcraftproducts.com](http://www.healthcraftproducts.com)

### **I**

- International Society for Augmentative and Alternate Communication • 416-385-0351 • [www.isaac-online.org](http://www.isaac-online.org)

### **J**

### **K**

### **L**

- Luminaud (speech equipment) • 800-255-3408 • [www.luminaud.com](http://www.luminaud.com)

### **M**

- Maxi Aids (medical supply/visual assistance store) • 800-522-6294 • [www.maxiaids.com](http://www.maxiaids.com)
- Meals-on-Wheels Association of America • 703-548-5558 • [www.mowaa.org](http://www.mowaa.org)
- Medicaid, 800-633-4227 • [www.cms.gov/medicaid](http://www.cms.gov/medicaid)
- Medicaid Fraud Control Unit • 202-326-6000 • [www.naag.org](http://www.naag.org)
- Medicare, 800-633-4227 • [www.medicare.gov](http://www.medicare.gov)
- Menu Direct Corporation (pureed meals) • 888-MENU-123 • [www.menudirect.com](http://www.menudirect.com)
- Mercy Medical Airlift National Patient Travel Center • 800-296-1217 • [www.patienttravel.org](http://www.patienttravel.org)
- MOMS (medical supplies) • 800-232-7443
- Movement Disorder Society • 414-276-2145 • [www.movementdisorders.org](http://www.movementdisorders.org)

### **N**

- National Academy of Elder Law Attorneys • 520-881-4005 • [www.naela.org](http://www.naela.org)
- National Association of Professional Geriatric Care Managers • 520-881-8008 • [www.caremanager.org](http://www.caremanager.org)



## Resource Directory—continued

- National Center for Complementary and Alternative Medicine • 888-644-6226 • [www.nccam.nih.gov](http://www.nccam.nih.gov)
- National Council on Aging • 202-479-1200 • [www.ncoa.org](http://www.ncoa.org)
- National Family Caregivers Association • 800-896-3650 • [www.nfcacares.org](http://www.nfcacares.org)
- National Hospice Foundation • 800-338-8619 • [www.hospiceinfo.org](http://www.hospiceinfo.org)
- National Hospice and Palliative Care Organization • 703-837-1500 • [www.nhpco.org](http://www.nhpco.org)
- National Institutes of Health • 301-496-4000 • [www.nih.gov](http://www.nih.gov)
- National Institute of Neurological Disorders and Stroke (NINDS) • 800-352-9424 • [www.ninds.nih.gov](http://www.ninds.nih.gov)
- National Library of Medicine • 888-3463656 • [www.nlm.nih.gov](http://www.nlm.nih.gov)
- National Long-Term Care Ombudsman Resource Center • 202-332-2275 • [www.ltcombudsman.org](http://www.ltcombudsman.org)
- National Organization of Rare Diseases (NORD) • 203-744-0100 • [www.rarediseases.org](http://www.rarediseases.org)
- National Parkinson's Foundation • 800-3274545 • [www.parkinson.org](http://www.parkinson.org)
- No-Rinse Bathing • 800-223-9348 • [www.norinse.com](http://www.norinse.com)

### **O** **P**

- Patterson Medical (medical supply/assistive devices) • 800-323-5547 • [www.pattersonmedical.com](http://www.pattersonmedical.com)
- Plak-Vac • 800-325-9044 • [www.trademarkmedical.com](http://www.trademarkmedical.com)
- Prentke Romisch (communication) • 800-262-1984 • [www.prentrom.com](http://www.prentrom.com)
- Pride Mobility Products Corporation • 800-800-8586 • [www.pridemobility.com](http://www.pridemobility.com)
- PSP Europe • +44 (0) 1327 861077 • [www.pspeur.org](http://www.pspeur.org)

### **Q** **R**

- RESNA (technology-assistive devices) • [www.resna.org](http://www.resna.org)

### **S**

- Saltillo (communications) • 800-382-8622 • [www.salttillo.com](http://www.salttillo.com)
- Simply Thick (thickeners) • 800-205-7115 • [www.simplythick.com](http://www.simplythick.com)
- Social Security Administration • 800-7721213 • [www.ssa.gov](http://www.ssa.gov)

### **T** **U**

- U-Step Walking Stabilizer • In-Step Mobility Products • 800-558-7837 • [www.ustep.com](http://www.ustep.com)

### **V**

- Veteran's Helpline • 800-827-1000 • [www.va.gov](http://www.va.gov)

### **W**

- Well-Spouse Foundation • 800-838-0879 • [www.wellspouse.org](http://www.wellspouse.org)
- WE MOVE • [www.wemove.org](http://www.wemove.org)

### **XYZ**



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Fax: 410-785-7009  
[www.curepsp.org](http://www.curepsp.org)