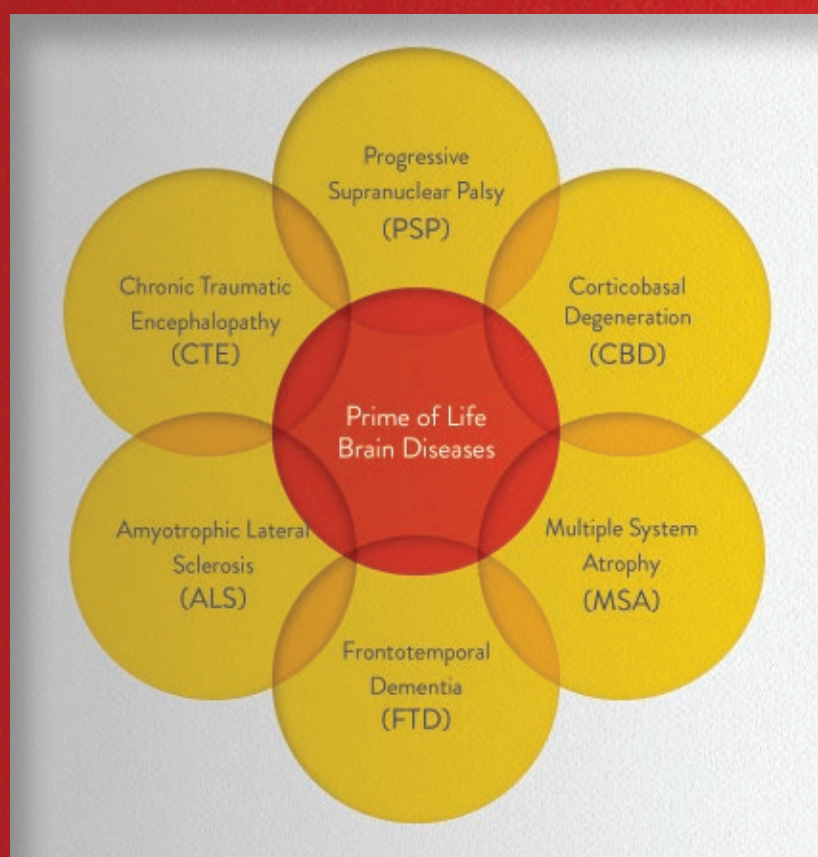


Guidebook



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

*Information you need to know
written by healthcare professionals*

2018 EDITION



This Guidebook is dedicated to the memory of

Pierre Richard Gervais,

April 28, 1950 - October 3, 2016.

Pierre Gervais became known by many as the leading advocate in the fight against progressive supranuclear palsy (PSP) with his contributions through videos and photographs, telling his story, sharing his journey, and his relentless support of researchers and physicians to find a cure for PSP.

Pierre always maintained his dignity, respect, and appreciation for those who cared for and supported him on his journey, leaving an amazing legacy that few can claim.

He was a great advocate for treatment and cure of PSP, creating the Pierre Gervais Fellowship grant at the Neural Stem Cell Institute, in 2016, and donating his brain to the Eloise H. Troxel Memorial Brain Bank, at the Mayo Clinic.

As many on this journey experience, his condition was misdiagnosed – first as Parkinson's disease, then as PSP. In autopsy, it was determined that Pierre's affliction was instead corticobasal degeneration (CBD), a disease closely related to PSP and often confused with it during clinical evaluation.

Pierre and the high school sweetheart he married, Jocelyne, raised three beautiful children, Eric, Leigh, and Sophie, who gave them eight grandchildren; Jackson, Emilia, Carter, Alexia, Gracie, Cooper, Madeline, and Catherine, who all cherished their Grandpa Pierre.

This dedication recognizes the generous contribution to the production of this guidebook by the Gervais family, a gift to Pierre's legacy in the hope that, in the future, no one else will have to suffer the same loss.



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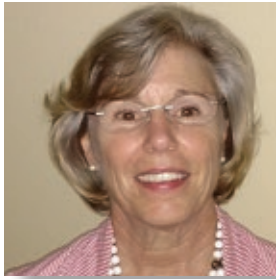
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Message from the Chair of CurePSP's Patient and Carepartner Advocacy Committee

Ileen McFarland

Those of us who have walked the walk as a patient or carepartner, associated with one of the rare prime of life brain diseases, know all too well the challenges we are confronted with on a day-to-day basis. Patients are stunned at their diagnosis and do not know where to turn and carepartners feel powerless and unqualified in their efforts to comfort and provide care for their loved ones. We here at CurePSP are sensitive to these circumstances and over the years have developed programs and support networks to assist you. The materials we have available to you from the onset of the disease through its course can be obtained by simply calling our office.

We have more than 50 support groups nationwide where patients and carepartners meet once a month to discuss their issues and ease their burden. These support groups are led by well-trained facilitators, some of whom have had a personal experience with one of the diseases. If a support group is not available in your area, we also have online support groups that are easily accessible. Periodically the support group leader will schedule professional speakers such as neurologists, physical therapists, occupational therapists, speech therapists, experts in legal matters, and hospice representatives, just to name a few. There are also online webinars that address specific subject matter related to the disease, such as managing carepartner stress, PT/ST/OT "how to" presentations, and others to come.

Twice a year, if not more often, we offer Family Conferences held at various locations nationwide. These conferences are for patients and carepartners and include a support group session the day before the conference. Additionally, we have approximately 80 Peer Supporters nationwide who play a very important role by making themselves available for patient, family, and carepartner phone calls. Most of our Peer Supporters have lost a loved one or a friend to one of the prime of life brain diseases and are familiar with the journey. They can provide useful information that will assist the patient and carepartners or just listen to your story.

Most of the above information can be accessed through our website www.curepsp.org. We continually work toward strengthening and expanding our advocacy support programs to provide the tools you need to ease the burden of patients and their families.

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

We are here for you... you are not alone. As research continues to search for a cure, we are sensitive to your daily challenges and needs and can provide resources and support during your fight against your neurodegenerative disease. Get in touch with us and let us know your concerns. We can't make it go away but we can help you persevere. Contact info@curepsp.org or 347-294-2871.



Message from the President

David Kemp

Unlocking the secrets of brain disease.

Thank you for contacting CurePSP. We offer this guidebook as the definitive resource to help you on your challenging journey. I know from talking with patients, families, and carepartners on a daily basis the struggles that lie ahead and have seen first-hand the tragic effects of PSP and related prime of life diseases at the conferences we hold for patients, families, carepartners, and medical professionals.

CurePSP is committed to three related areas of service and support:

Care: We provide a full range of services and support to patients, families, and carepartners;

Consciousness: We educate physicians and allied healthcare professionals about PSP and related diseases so they may provide earlier and surer diagnoses and refer patients to clinical trials and support groups;

Cure: We fund research that has the potential to lead us to treatments and a cure for PSP and, ultimately, other neurodegenerative diseases.

While we have not yet found treatments or a cure, there is great hope in the air. Research is accelerating and showing encouraging results. PSP, in particular, is now recognized as perhaps the most promising target for research and clinical trials that may lead to therapies for other neurodegenerative diseases. As such, it has the attention of both academic research institutions and pharmaceutical companies.

We are pleased to bring you this guidebook full of helpful information and advice from leading experts. We urge you to contact us if you require further information – or if you want to help with our cause. Meanwhile, we wish you the best as you embark on your journey. It will be difficult and challenging to be sure, but know that you are not alone and that others are there to stand by you and to help lessen your burden.



Message from CurePSP's Vice President-Scientific Affairs

Alex Klein, PhD

A world free of PSP, CBD, MSA, and related prime of life brain diseases: This is the vision for our work at CurePSP. To achieve this vision and to find a cure, we are working very closely with international researchers in academia and the pharmaceutical industry.

As the Vice President-Scientific Affairs, I am passionate about making the quest for a cure a success story and propelling neurodegeneration research to the next level in therapy development. Interestingly, prime of life brain diseases share some similar disease-causing pathways (the misfolding and accumulation of proteins); hence tackling those shared characteristics across the diseases may allow us to find cures not only for PSP, but also for other related brain diseases.

For more than 25 years, CurePSP has been actively supporting patients, and their carepartners and families, and also in funding cutting-edge research and developing novel therapies. With pride, we look back and report that we have funded more than 170 studies with an accumulated budget of around \$13 million! We have established a very prestigious research program and interact with international scientists at the forefront of PSP research and beyond. This includes related diseases, such as CBD, MSA, FTD, ALS, and CTE, but also — and that might surprise some — Alzheimer's and Parkinson's diseases.

As mentioned above, these diseases share protein misfolding as key a disease-causing trait, so patients suffering from those diseases could benefit from therapies developed for PSP. This has attracted interest not only from scientists, but also from pharmaceutical companies that have increased their research activities in these arenas, even though compared with approximately five and one-half million Alzheimer's patients, there are only about 20,000 PSP patients in the United States. However, clinical trials in rare diseases may require smaller numbers of patients and can be approved by the regulatory bodies using a fast-track program; this, in turn, allows more cost-effective trials, faster approval of the trials, and, we hope, faster cures for PSP and related disorders, including Alzheimer's and Parkinson's diseases.

CurePSP has become a reliable partner in brain science; we have developed a trusting relationship with the research community, which includes world-renowned scientists, other patient advocacy groups, and pharmaceutical companies from around the globe. The National Institute of Neurological Disorders and Stroke (NINDS) as part of the National Institutes of Health (NIH) in Washington, D.C., recently confirmed during the NINDS 10th Nonprofit Forum that the work of foundations like ours is of the highest importance not only in terms of offering patient support, but also in allowing researchers to experiment and search for novel therapies outside of government-funded projects. Their funding permits scientists to prove new ideas and concepts using this data as the basis for larger national projects.

The progress in PSP research can also be measured by the growing interest in our annual International CurePSP Research Symposium. From a small group just a couple of years ago, we have become a highly respected meeting with more than 150 attendees. We invite international opinion leaders across the disease spectrum to exchange their ideas and share data, which is a prerequisite for developing therapies for PSP. Only a worldwide team effort will make our quest for a cure successful.

Last but not least, I would like to thank our supporters. None of this work would be possible without the support of our generous and loyal donors whose contributions to PSP research have enabled great progress leading to more clinical trials and potential new therapies. We are confident that there is no better place to invest money than in the work of CurePSP. I encourage you to contact me with your thoughts and ideas at info@curepsp.org or 347-294-2872.

How to use this Guidebook

This guidebook can be used in various ways. It is a great learning tool so you can become more knowledgeable about prime of life diseases such as PSP, CBD, and MSA and it allows you to go back and refresh your memory on questions that come up over time. We also realize that this book has an emotional impact and sometimes reading sections that are most important at the time rather than reading the entire manual at one time is a good approach.

You'll find information regarding symptoms that will emerge over the course of the disease and this will help answer your questions about what to expect. There is sound, practical advice in these chapters to assist you in planning for and adjusting to these changes. Everyone will be confronted with decisions related to how much help their loved one will require over time and while these are difficult conversations to have, there is information that you need to know about available choices.

You will find some articles that speak about the challenges of being a carepartner and the impact of stress on both physical and mental health. We also acknowledge how families deal with a neurodegenerative disease, how to identify and understand the dynamics of family relationships under chronic stress, and what can be done to strengthen those relationships.

We recognize that no guidebook can adequately address all of the issues and questions that you will have on your journey, but we hope this will be a valuable resource. Please keep in mind that CurePSP is your primary resource and we are here to help in any way we can. Please feel free to contact us at 800-457-4777 or at info@curepsp.org.



PART ONE

The Diseases



PSP: SOME ANSWERS

Lawrence I. Golbe, MD

Professor of Neurology,

Rutgers Robert Wood Johnson Medical School

Director of Clinical Affairs and Scientific Advisory Board Chairman, CurePSP

What is progressive supranuclear palsy (PSP)?

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

Why has no one heard of PSP?

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

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

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

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

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

What are the most common early symptoms of PSP?

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

Are there other, rare types of PSP?

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

What happens next?

The name of the disease includes the word “progressive” because, unfortunately, the early symptoms get worse and new symptoms develop over time. After five or six years, on average, the imbalance and stiffness worsen to make walking very difficult or impossible. Trouble with eyesight eventually develops in almost all cases and can sometimes be as disabling as the movement difficulty. Difficulty with speech and swallowing are additional important features of PSP that eventually occur in most patients.

What does the name “supranuclear palsy” mean?

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

Is the visual problem the most important part of PSP?

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

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

PSP: SOME ANSWERS (continued)

The most common and characteristic eye movement problem in PSP is an impaired ability to move the eyes up or down. This can interfere with eating or with descending a flight of stairs, among other things. This problem is not usually as vexing for the patient and family as the inability to maintain eye contact or to coordinate eye movements while reading, but is much easier for the doctor to detect. Reduced vertical eye movement is usually the first clue that the diagnosis is PSP. Although other conditions, particularly Parkinson's disease and normal aging, can sometimes cause difficulty moving the eyes up, PSP is nearly unique in also causing problems moving the eyes down. This problem often takes the form of eye movement "apraxia," where the patient can move the eyes up or down only after several requests or with a delay after initiating the effort. In most people with PSP, the difficulty in downward eye movement starts out as a slowing of that movement. This can also interfere with vision, but can be very difficult for a physician to detect. Another eye movement problem that starts early in the illness is square wave jerks— rapid, involuntary, right-left movements that interfere with the ability to precisely aim the eyes at a target.

Yet another eye problem in PSP can be abnormal eyelid movement—either too much or too little—called blepharospasm. A few patients experience forceful involuntary closing of the eyes for a few seconds or minutes at a time, while others have difficulty opening the eyes, even though the lids seem to be relaxed. They may try to use the muscles of the forehead, or even the fingers, in an effort to open the eyelids ("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 three or four times per minute. This can allow the eyes to become irritated and react by producing extra tears, which in itself can become annoying.

What sort of speech problems occur?

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

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

What about the swallowing problems?

This is the source of the most important and dangerous long-term complications of PSP. Swallowing tough foods or thin liquids can become difficult because of throat muscle weakness or incoordination. This tends to occur later than the walking, visual, and speech problems, but can become very troublesome if the patient tends to choke on food or if food goes into the breathing passages. Usually, problems managing thin liquids precedes difficulty with solid food because the swallowing muscles have difficulty creating a watertight seal that separates the path to the stomach from the path to the lungs. This is true with many neurological diseases. For nonneurologic conditions, such as stricture of the esophagus, the difficulties start with solid

foods. Repeated, minor, often unnoticed episodes of small amounts of food and drink dripping into the lungs can cause pneumonia. Often, it is not apparent to the physician or family that the PSP patient's pneumonia is in fact the result of subtle aspiration. But aspiration pneumonia is the most common cause of death in PSP. The risk of aspiration is aggravated by the tendency to overload the mouth or to take big gulps of beverages due a loss of inhibition or a reckless impulsiveness that can be partly involuntary.

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

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

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

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

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

How is PSP different from Parkinson's disease?

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

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

PSP: SOME ANSWERS (continued)

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

What about treatment with medication?

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

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

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

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

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

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

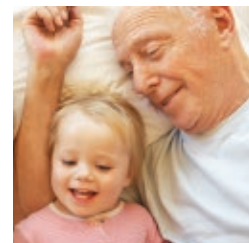
Antidepressants have also had some modest success in PSP, sometimes relieving the depression that can be part of the disease. There are many antidepressants and none has been shown to be superior to any others. The older, tricyclic antidepressants seem to be no less effective in PSP than the newer reuptake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

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

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

One possible success story is the dietary supplement Coenzyme Q-10 (CoQ10), which is available without a prescription, although the jury is still out. It helps the body's cells produce energy from sugar and oxygen and is a normal constituent of the mitochondria, the tiny compartments in our cells where that chemical process occurs. One small study gave modestly favorable results. Another gave similar benefit, but was too small to be sure that its outcome was not just the result of chance. The required dosage of the standard formulation of CoQ10 is probably at least 1200 mg per day and perhaps as high as 2400 mg. The newer liposomal form probably gives the same benefit at 300 mg per day. As a nonprescription item, CoQ10 is not covered by drug insurance and costs \$50 to \$100 month. Therefore, people with PSP should carefully consider the meager evidence to date for the benefit of CoQ10 before taking that long-term financial plunge. Their neurologists should do a careful exam upon starting the drug and repeat it two months later to determine whether the treatment is working, and if not, discontinue it.

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



PSP: SOME ANSWERS (continued)

What about other experimental drugs?

In the past 20 years, many drugs have been tested in patients with PSP. Some of these are intended to actually slow the long-term brain cell loss; i.e., slow progression of the disease. Unfortunately, none has helped. The most promising were riluzole, which modestly helps amyotrophic lateral sclerosis; davunetide, which is a fragment of a protein that helps maintain brain cells' internal skeletons; and tideglusib, which prevents the abnormal attachment of phosphate to the tau protein, preventing it from clumping up in the brain cells. The last drug did show a glimmer of promise; MRI scans showed less loss of brain mass in the patients on tideglusib than in those on placebo. This may justify further study of the drug, but it is doubtful that any drug company will want to pursue it after it failed to slow progression of the actual symptoms of the disease.

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

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

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

Is tube feeding advisable for advanced patients?

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

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

The PEG tube can be inserted with the patient awake but sedated, often as an outpatient procedure. The tube is clamped shut and hidden under the clothes when not in use. The feeding can 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 feeling “whole” or part of humanity. The issue of how a feeding tube will affect a patient’s quality of life must be considered carefully by the family, physician, and if possible, the patient and sometimes ethical or spiritual advisors. Some patients in the advanced stages of PSP may feel that their quality of life is so poor that prolonging that life by having a PEG installed is not what they want. It’s important to note that some nursing homes will advise PEG placement because it reduces the personnel time needed to feed the patients and because third-party payers often will pay an additional fee for tube feeding but not for the time-consuming task of hand feeding a patient by mouth.

Do any of the new brain operations for Parkinson’s work for PSP?

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

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

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

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

What about other nondrug treatment?

Formal physical therapy is worth a trial in PSP, especially with the goal of teaching the patient to use gait-assistive devices such as a walker. Certain exercises done in the home by oneself on a regular schedule can keep the joints limber. Exercise also has a clear psychological benefit that improves a sense of well-being for anyone with a chronic illness. For specific exercises, consult one of the books for patients with Parkinson’s disease or the pamphlets distributed by the national Parkinson organizations.

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

PSP: SOME ANSWERS (continued)

Probably the most important part of dealing with PSP is for the patient's family to understand that the problems with visual inattention and personality changes are part of the illness. The patient is not lacking willpower nor "faking." Furthermore, many of the problems in PSP are intermittent and can be aggravated by the patient's mental or emotional state. For example, walking, writing, and eating may be poor one hour and better the next. The family should understand that these fluctuations are not under the patient's control and that nagging and shouting usually 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 at another time.

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. People with such symptoms can fall if they move their body forward before the foot moves. In these cases, smooth soles could make it easier to slide the first foot forward. Handrails installed in the home, especially in the bathroom, may also be helpful. The difficulty looking down dictates that low objects such as throw rugs and low coffee tables be removed from the patient's living space.

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

What is the cause of PSP?

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

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

No one knows yet, but we have some clues. In the brain cells that are degenerating in PSP, there is an abnormal accumulation of the normal protein tau. These clumps of tau, once they reach a size that can be seen through a microscope, are called neurofibrillary tangles. The normal function of tau is to help support the microtubules, which have two important jobs: one is to form the internal "skeleton" of the brain cells and

the other is to serve as a “monorail” system, transporting nutrients around the cell. We don’t know whether the problem is that the tau is defective from the time of its manufacture, or if it is damaged later, or even if it remains normal but is produced in excess. If it is damaged, the nature of that damage could be the excessive attachment of phosphate (see the section on PSP preventive drugs). Or, the excessive phosphates on the tau molecules could simply be how the brain minimizes the effects of tau protein that is misbehaving for some other reason.

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

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

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

Is PSP genetic?

PSP only very rarely runs in families. Fewer than one in 20 people with PSP knows of even one other family member with PSP and detailed neurological exams of relatives of patients with PSP show no more definite abnormalities than exams of relatives of healthy people. However, two different variants in the gene on chromosome 17 that encodes the tau protein are more common in PSP than in the rest of the population. One of the variants is called the H1 haplotype. About 95 percent of people with PSP have this variant on both of their copies of chromosome 17, while this is true for only about 60-77 percent of the rest of us. So clearly, the H1 haplotype is (nearly) necessary but far from sufficient to cause the disease.

PSP: SOME ANSWERS (continued)

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

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

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

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

Could PSP be caused by toxins?

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

One important clue to a possible dietary factor in the cause of PSP comes from the island of Guadeloupe in the Caribbean. People there are far more likely to develop PSP and other “atypical parkinsonisms” than 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 foods in the Western diet, if any, may contain similar toxins. Research on that question is underway.

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

How can I help research?

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

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

Another way to help research and yourself is to participate in studies of PSP if 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. Joining the mailing list at CurePSP will allow PSP researchers to contact you regarding participating in new research studies.

Should I join a support group?

There can be great value in joining a group of other people with the same problem. You can exchange helpful tips on ways to cope physically and psychologically with the limitations of the illness and can learn more about the problem and its treatment from guest speakers. Many large medical centers have a Parkinson support group that welcomes members with PSP. While there are far fewer people with PSP than Parkinson’s in one geographical area, several dozen successful PSP support groups have been organized in the U.S., usually in more densely populated areas. All it takes is one organizer with some time and energy. See the chapter “**CurePSP Support Groups**” on page 98 of this Guidebook for information.

CBD: SOME ANSWERS

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What is corticobasal degeneration (CBD)?

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

What are its main signs and symptoms?

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

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

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

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

How does CBD start?

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

What happens later?

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

How common is CBD?

CBD is very rare: about five people per million population, which comes to about 1,600 people in the United States, most of whom have not yet been accurately diagnosed. For Parkinson's, the figure is about 700,000, and for Alzheimer's disease, about 5 million.

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

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

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

What happens in the brain?

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

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

CBD: SOME ANSWERS (continued)

What are the various subtypes of CBD?

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

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

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

Is CBD genetic?

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

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

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

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

How is CBD treated?

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

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

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

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

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

Is physical therapy useful?

Formal physical therapy is worth a trial in CBD, especially with the goal of teaching the patient to use gait assistive devices such as a walker. Certain exercises done in the home by oneself on a regular schedule can keep the joints limber. Exercise also has a clear psychological benefit that improves the sense of well-being of anyone with a chronic illness. For people with gait or balance problems, many useful exercises can be performed seated in a chair or lying on a mat. Using a stationary bicycle is usually feasible as long as there is help in mounting and dismounting safely. The best strategy is to have an evaluation and treatment plan from a physical therapist or physiatrist (a physician specializing in rehabilitation of chronic conditions). The same advice applies to the utility of occupational therapy in improving the apraxia and dystonia of the hand(s) that is so common in CBD.

CBD: SOME ANSWERS (continued)

Are there experimental treatments for CBD?

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

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

How long do people with CBD live?

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





MSA: SOME ANSWERS

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What is Multiple System Atrophy (MSA)?

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

How common is MSA?

MSA is rare, with about 13,000 sufferers in the United States, most of whom have not yet received a correct diagnosis. This compares with about 20,000 with progressive supranuclear palsy (PSP), which is a similar condition, about 500,000 with Parkinson’s disease, and five million with Alzheimer’s disease. New cases of MSA arise in about six persons per million per year. This means that in the U.S., about five people are newly diagnosed with MSA each day.

Are there different types of MSA?

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

What is the cause of MSA?

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

What’s going wrong in the brain and spinal cord cells?

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

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

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

At what age does MSA start?

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

What happens to someone with MSA?

All three kinds of MSA feature important balance problems that can eventually require assistance in walking or even a wheelchair. All three types can also display difficulty in the aspect of thinking called executive function. This is what allows us to organize information by categories, understand abstractions and instructions, create and follow a plan, and inhibit inappropriate actions. These 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 MSA that emphasizes the autonomic problem have a variety of symptoms including urinary urgency and incontinence, constipation, erectile disturbance in men, lightheadedness or even fainting upon standing, noisy or irregular breathing, sleep disruption, and difficulties with temperature regulation. Most people with MSA have some combination of the three types rather than a pure form.

Some other problems with movement can occur in MSA. The hands can gradually assume abnormal, fixed postures called dystonia. This can be prevented to some degree by stretching exercises and can be treated in some cases by botulinum toxin (Botox) injections into the hands or forearms. Some people with MSA hold their heads bent forward to an extreme degree, a condition called antecollis. This 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.

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

MSA: SOME ANSWERS (continued)

Is MSA a fatal disease?

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

How is MSA treated?

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

What happens eventually?

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

What research is being done?

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

How can I help research in MSA?

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

PART TWO

The Symptoms



PSP and the Bladder

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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

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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 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 composed of the nondigestible components of grains, fruits, and vegetables.

THE SYMPTOMS (continued)

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

Role of exercise

Inactivity is the enemy of regular elimination. Constipation often worsens after a period of marked immobility, such as a hospitalization or accident that reduces mobility. Regular walking aids regular bowel function. If walking is not possible because of balance or other problems, exercises such as pumping the legs, crawling, or waist bending can help. 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 1/2 lemon on waking each morning helps stimulate bowel action.
- Prune juice cocktail: Mix 1/2 cup applesauce, 1/2 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 another healthcare professional.

Complications

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

Pressure Sores

Pressure sores, sometimes called bedsores, decubitus 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 by a physician 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 and even skin grafting are required 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 healthcare 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 healthcare 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 Lyphazome.
- Good nutrition plays an important role in the prevention of and recovery from a pressure sore.

THE SYMPTOMS (continued)

Dry Eye Syndrome

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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.

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 nonpreserved 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 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

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Introduction

PSP affects the control centers in the brain that generate eye movements. Thus, patients progressively lose the ability to move their eyes. During the early stages of the disorder, the eyes still move fully but slowly, particularly in upward and downward directions. Most patients do not experience any eye symptoms at this stage. As time goes on, the range of eye movements decline, 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. Consulting with a neuro-ophthalmologist can be very helpful as she or he is more likely to be knowledgeable about how neurodegenerative diseases present in the eyes.

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 she or he 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 eyesight should see an eye specialist before attributing the loss of vision to PSP.

THE SYMPTOMS (continued)

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 three to four months. Repeated injections are necessary to keep the condition under control.

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

Nutritional Implications

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Introduction

Progressive supranuclear palsy (PSP) and corticobasal degeneration (CBD) are examples of neurodegenerative disorders that are considered under the heading of frontotemporal disorders. Although, each neurological disease can vary with respect to progression, severity, and manifestation of symptoms, they 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 carepartner, and the healthcare team caring for the patient.

Nutritional factors

Many different nutritional challenges can 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 the disease.

Oral and pharyngeal weakness

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

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. Not all healthcare providers may bring up the topic of the benefits and risks of a gastrostomy tube, but it's important to have the discussion when individuals show signs of significant difficulty or discomfort in swallowing, 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, and meal-related frustration, 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.

THE SYMPTOMS (continued)

Constipation

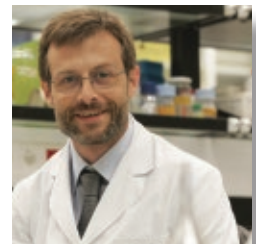
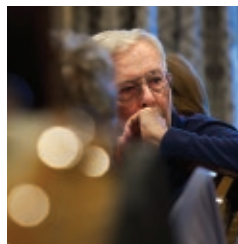
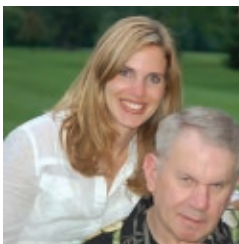
As disease progression continues, some form of constipation or slowed gastrointestinal motility will occur. Constipation is related to many varying factors including a reduction in physical mobility, side effect(s) of medications, decreased fluid intake, lower consumption of high-fiber foods, or a combination 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 carepartner. 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.

The ability to maintain adequate nutrition is likely to change throughout the course of PSP and related diseases. Recognizing, as well as adapting to change, with the help of a supportive physician and healthcare team, may provide ease in adjusting to these changes as they arise.





THE SYMPTOMS (continued)

Good Oral Hygiene

Nancy Brittingham, BS, RDH

Persons diagnosed with PSP/CBD/MSA and their carepartners face increasing challenges as the disease progresses. The carepartner's tasks continue to increase, and many times helping the patient 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 patient's self-image, 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 and 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, but maintaining good oral hygiene is something you can take charge of.

Tips

- Take action steps early after diagnosis to avoid extensive dental procedures later on. As the disease progresses exams and procedures are much more difficult to tolerate.
- Make an appointment with your dentist as soon as possible after diagnosis; make sure you have a list of ANY medical conditions you have and all the medications you're taking.
- This is an opportunity to educate the dental staff about your disease. Most professionals aren't knowledgeable about these diseases and it is important for them to understand your symptoms to be able to work most effectively with you.
- Although most dental insurances cover two cleanings a year, it would be a good investment to consider three/four cleanings a year. Prevention is key and a regular hygiene routine is important.
- Ask your dentist about prescribing a fluoride dental paste.
- Request a fluoride treatment.
- Ask your dentist to make sure partials or dentures are fitting correctly or that adjustments are made in a timely manner.
- Ask the dental staff if dental X-rays are up to date; talk with your dentist about completing the X-rays if necessary.
- Make sure the dental office is wheelchair accessible.

Brushing

- Make sure the patient'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.

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 the patient can 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 are still present in the mouth, and these recommendations are for patients who are tube-fed as well.

Conclusion

Good oral hygiene is very important. Take time out to take care of your well-being. Being able to prevent problems today is well worth it tomorrow.

THE SYMPTOMS (continued)

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:

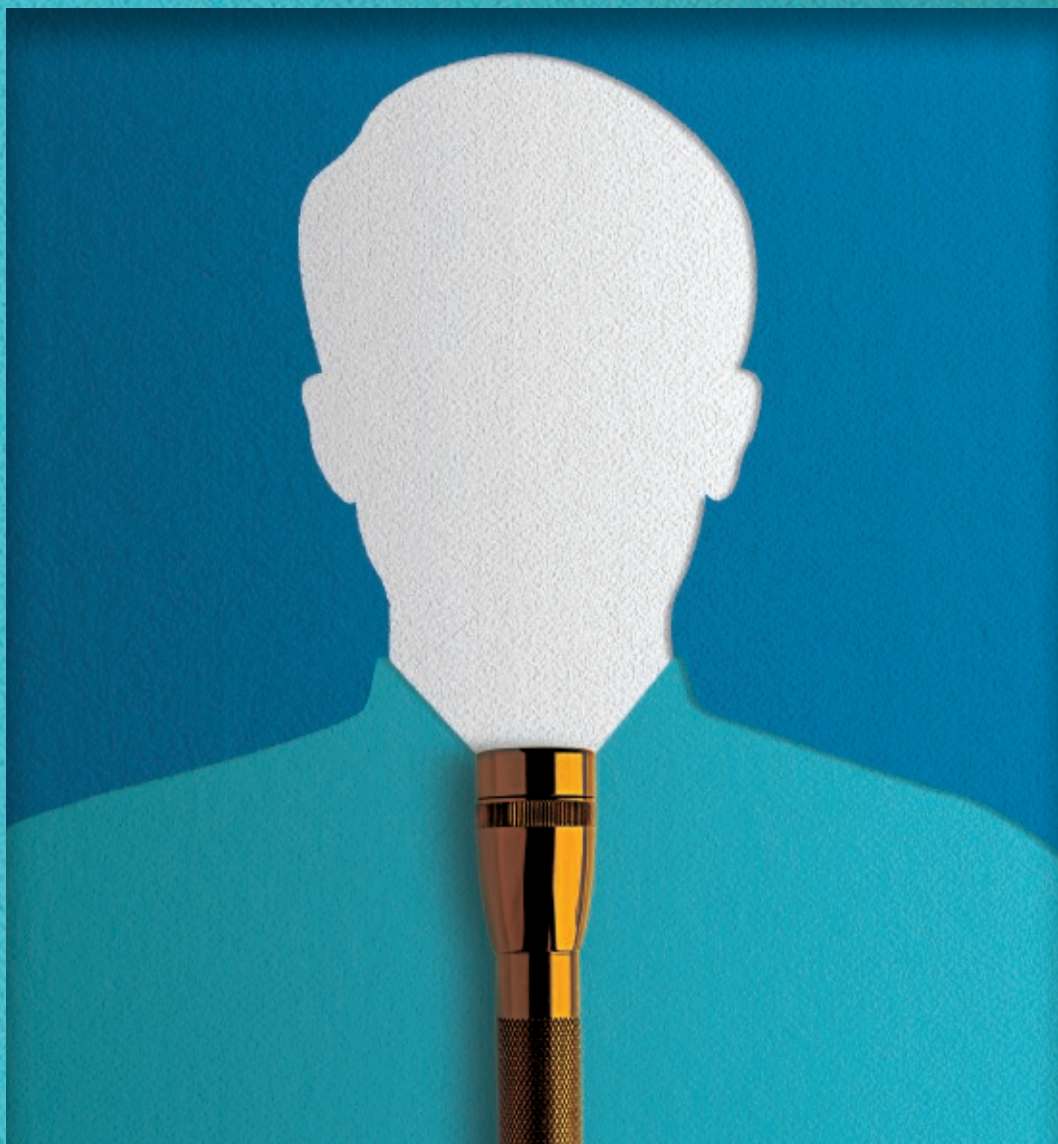
- Sit up straight when eating or drinking. Sitting in a chair is best.
- Eat slowly; take small bites of food and small sips of liquid.
- Stay sitting for about 30 minutes after eating, if you can.
- When you are in bed, keep your head raised with pillows; you should rest and sleep in a sitting position 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.

PART THREE

Managing Symptoms

and Getting Professional Support



Building Your Personal Advisory Team

Managing PSP, CBD, and related diseases is a continually evolving challenge. While they are degenerative diseases, 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 that is attuned to the specific, special needs of a person with one of these diseases.

Although fragmented healthcare services with limited communication among care providers can make it challenging to build an advisory team, the information below can help you understand the role of the different providers and what you can do to improve communications among them.

Your advisory team ideally should include the following:

Neurologist

A neurologist is a specialist in diseases of the nervous system. She or he will outline possible treatment options, work with you to identify specific concerns and needs, and refer you to therapists to help you find solutions to improve your day-to-day life. Your neurologist is also a good resource regarding any current research studies and can advise you as to how to find more information.

Movement disorder specialist

A movement disorder specialist is a neurologist who has received additional training in many types of movement disorders, including prime of life diseases. While a general neurologist may treat patients with any of more than 100 neurological conditions, a movement disorder specialist is often affiliated with a major university or teaching hospital and is likely to be on the cutting edge of knowledge and treatment of movement disorders. They are often best equipped to tailor a plan of care for you and your specific needs.

Nurse specialist

Every person living with PSP and other prime of life diseases should have a nurse specialist on their advisory team. Patients and families who have been on this journey attest to the value of having a nurse specialist to call on for guidance today and for advice down the road. These specialists often work with neurologists. These nurse specialists can provide you with tremendous guidance as you navigate the challenges of living with these diseases. They are registered nurses who specialize in the area of movement disorders. They are knowledgeable about the symptoms, medications, management strategies, research, and resources. They often facilitate support groups for those with a prime of life disease as well as their care partners, either independently or with social workers. Nurse specialists, in addition to helping the affected person along the disease trajectory, provide support, guidance, and resources to carepartners.

Physical therapist

Physical therapists (PTs) provide services that help maintain the highest level of function – improve mobility, reduce pain, and reduce the likelihood of falls that can lead to other physical disabilities. PTs examine a patient's medical history and test and measure strength, range of motion, balance, and coordination, posture, muscle performance, and motor function. They also determine a patient's ability to be independent and develop a treatment strategy with a clear purpose and anticipated outcome.

Speech-language pathologist (SLP)

As with any progressive neurological condition, early intervention is the key to maintaining or increasing communicative effectiveness and swallow function. As soon as a patient or carepartner 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 strengthen speech and swallowing functions.

Occupational therapist (OT)

Occupational therapists (OTs) can assist you in managing physical, functional, visual, and cognitive changes. They are trained to work from a person-centered perspective to build a supportive social and environmental context to help you manage the disease as well as possible. Your OT may address functional vision, physical skills, home safety, community independence, and cognition, working collaboratively with you and your family to build the best mix of supports to help your day-to-day life. Often, PTs and OTs work particularly closely together to ensure a patient's safety.

Social worker (SW)

You may also be referred to a social worker (SW) who can provide both practical and emotional support to help you and your family. A SW takes into consideration all aspects of one's life, including the emotional impact that the disease has on you, your family and your friends, and help address quality of life issues that impact your well being. A social worker can offer information about available community resources and acts as an advocate to assist people in accessing these resources. She or he also provides information on financial and legal issues and can assist 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.

What Every Social Worker, Physical Therapist, Occupational Therapist, and Speech-Language Pathologist Should Know About PSP, CBD, and MSA

A Comprehensive Guide to Signs, Symptoms, and Management Strategies

Disease summaries at a glance

Progressive supranuclear palsy (PSP)

- Rare neurodegenerative disease, the most common parkinsonian disorder after Parkinson's disease (PD).
- Originally described in 1964 as Steele-Richardson-Olszewski syndrome.
- Often mistakenly diagnosed as PD due to the similar early symptoms.
- Symptoms include early postural instability, supranuclear gaze palsy (paralysis of voluntary vertical gaze with preserved reflexive eye movements), and levodopa-nonresponsive parkinsonism.
- Onset of symptoms is typically symmetric.
- Pathologically classified as a tauopathy (abnormal accumulation in the brain of the protein tau).
- Five to seven cases per 100,000 people.
- Slightly more common in men.
- Average age of onset is 60-65 years, but can occur as early as age 40.
- Life expectancy is five to seven years following symptom onset.
- No cure or effective medication management.

Signs and symptoms

- Early onset gait and balance problems
- Clumsy, slow or shuffling gait
- Lack of coordination
- Slowed or absent balance reactions and postural instability
- Frequent falls (primarily backward)
- Slowed movements
- Rigidity (generally axial)
- Vertical gaze palsy
- Loss of downward gaze is usually first
- Abnormal eyelid control
- Decreased blinking with "staring" look
- Blepharospasms (involuntary eyelid spasms)
- Double vision
- Dystonia, commonly at neck and hands into flexion, but can also be into extension at neck
- Speech and swallowing changes
- Subcortical dementia (personality changes, slowness of thought)
- "Rocket sign," when patient jumps up quickly from seated position, often falling back in chair

Corticobasal degeneration (CBD)

- Rare neurodegenerative disease that affects the cortex (frontoparietal) and basal ganglia.
- Originally described in 1968 by Drs. Rebeiz, Kolodny, and Richardson, with earliest descriptions by Charcot (1888).
- Pathologically classified as a tauopathy (accumulation of the tau protein in the brain).
- Onset of symptoms is markedly asymmetrical.
- Diagnosis is difficult because clinical features often overlap with Parkinson's disease (PD), progressive supranuclear palsy (PSP), Alzheimer's, primary progressive aphasia, and frontotemporal dementia.
- Average age of onset usually between 60 and 80 years.
- Prevalence unknown; estimated to be less than one case per 100,000 people.
- Slightly more common in women.
- Life expectancy is seven to ten years following symptom onset.
- No known cure or medications to slow disease progression.

Signs and symptoms

- Asymmetrical presentation; symptoms begin on one side, which always remains worse
- Slowness and stiffness
- Shakiness
- Clumsiness in UEs or LEs
- Dysphasia, dysarthria, and dysphagia
- Changes in gait and balance
- Mild memory or behavior problems
- Increased parkinsonian symptoms (rigidity, bradykinesia, postural instability)
- Tremor
- Myoclonus
- Dystonia
- Blepharospasm (involuntary eyelid spasm)
- Sensory loss
- Increasing speech and swallowing difficulty
- Mild to moderate cognitive impairments
- Frontal dementia

Disease summaries at a glance (continued)

Multiple system atrophy (MSA)

- Rare neurodegenerative disease that affects multiple systems, particularly motor and autonomic nervous systems.
- First described in the 1960s as Shy-Drager syndrome.
- Pathologically classified as a synucleinopathy (accumulation of the protein alpha-synuclein).
- Subclassified by three core clinical features: parkinsonian, autonomic, and cerebellar.
 1. MSA-P (parkinsonian)-striatonigral degeneration implies parkinsonism with some degree of cerebellar dysfunction
 2. MSA-A (autonomic)-Shy-Drager syndrome reflects a predominance of autonomic failure
 3. MSA-C (cerebellar): olivopontocerebellar atrophy indicates primarily cerebellar defects with minor degrees of parkinsonism
- Three to four cases per 100,000 people.
- Average age of onset is usually after 50 years.
- More common in men.
- Life expectancy is seven to ten years after symptom onset.
- No cure or medications to slow disease progression.

Signs and symptoms

- Rigidity
- Action tremor: irregular, jerky, myoclonic movements
- Bradykinesia
- Freezing of gait
- Early postural instability and falls
- Orthostatic hypotension
- Erectile dysfunction
- Bladder dysfunction: urgency, frequency, incontinence
- Constipation
- Speech and swallowing difficulties: mixed dysarthria tends to emerge earlier in MSA than PD, is more severe and deteriorates more rapidly
- REM Behavioral Disorder (RBD): acting out dreams while sleeping due to lack of atonia
- Gait and limb ataxia
- Nystagmus and jerky pursuit
- Antecollis
- Difficulty with thermoregulation
- Cognitive impairment is typically mild

The role of social workers

Diane B. Breslow, MSW, LCSW

In order to successfully assess and treat people struggling with PSP, MSA, or CBD, social workers need to understand the symptoms, progression, and challenges these diseases pose. Social workers' understanding of their clients' biopsychosocial needs enable them to play a key role ensuring that patients and their families receive the best care, empathy, and guidance these chronic, progressive neurodegenerative diseases require.

Social workers can build ongoing, long-term relationships with their clients as well as provide valuable services to patients and families around specific needs or at periodic intervals, as particular challenges and changes develop.

Psychological support

Sadness and depression are intrinsic biochemical responses to these diseases, normal responses to the losses both patients and their families face. In addition, fear, uncertainty, and frustration are normal reactions to living with a neurodegenerative disease. It is important for social workers to help patients and family members to:

- Express emotions such as sadness, anger, worry, and frustration.
- Build a support team with healthcare providers, family, friends, volunteers, and clergy.
- Reach out to support groups, peer networks, or one-on-one peer support in order to gain knowledge, information, and resources; share feelings and experiences; receive understanding and encouragement.
- Develop attitudes and habits of flexibility, persistence, and adaptation.
- Cultivate a focus not on cure, but rather on living the best life possible and creating new meaning.

Social and family connections

Patients and families often feel, and actually are, alone and isolated. This is a result of the myriad of physical challenges (loss of balance and difficulty with mobility), emotional changes (loss of interest in previously enjoyable activities), self-consciousness (difficulty swallowing, slurred speech), and other difficulties brought on by PSP, MSA, and CBD. The social worker will help patients and families to:

- Prioritize their needs.
- Adapt their lifestyles to the changes incurred by living with someone with a progressive neurological disorder.
- Cope with caregiver stress.
- Build or maintain good communication about the impact of living with the disease throughout its progression.
- Resolve conflict in the family around family members' roles, ideas about care, and degrees of acceptance of the disease.
- Plan for the future, including understanding home care and housing, advance directives, insurance issues, and other concrete needs.
- Learn about and gain access to community resources, services, and programs.

MANAGING SYMPTOMS (continued)

The role of social workers (continued)

Education and resources

Management of these diseases requires input from multiple disciplines and community resources including physical therapy, speech therapy, occupational therapy, social work, psychiatry, and nutrition. Social workers can facilitate referrals, communication, and care coordination among multidisciplinary professionals and settings and educate family members and healthcare workers about the diseases and the needs of patients and families.

Resources to consider include:

- Associations like CurePSP
- Home safety evaluation and modification
- Workplace accommodation
- Social Security disability
- Legal assistance
- Exercise classes and videos
- Prescription drug programs
- Transportation
- Support groups
- Advance directives
- Caregiver support
- Home rehabilitation and home health agencies
- Adaptive and mobility equipment
- Respite
- Hospice

The role of physical therapists

Heather Cianci, PT, MS, GCS

While many of the signs and symptoms of PSP, CBD, and MSA appear similar, understanding the issues specific to each disease can help physical therapists develop and implement more successful treatment strategies.

Progressive supranuclear palsy (PSP)

Patients will benefit from continual exercise, physical activity, and social engagement throughout the course of the disease, including appropriate group exercise classes. Carepartners should be educated about the likelihood of the patient's increased movement impulsivity and decreased safety judgment as the disease progresses.

While the literature on rehabilitation for PSP is limited, it suggests that PT plays a role in managing balance and gait and will be needed at various times throughout the course of the disease.

The acronym “FIGS” will help to differentiate PSP from Parkinson’s disease (PD):

- F = Frequent, sudden falls (generally posteriorly; occurring early in the disease)
- I = Ineffective medication (antiparkinsonian meds do not work)
- G = Gaze palsy (vertical)
- S = Speech and swallow changes

Treatment strategies

- Patient and caregiver education about the disease
- Caregiver training in assistance techniques
- Dystonia: Botox (except for antecollis), stretching, positioning/bracing
- Blepharospasms: Botox, eye crutch
- Double vision: prism glasses
- Eye movement exercises, including tracking, searching for objects, reading words placed at various heights on paper on wall
- Aerobic, strength, and balance exercises along with fall prevention training
- Gait training, with a focus on large steps with adequate foot clearance and heel strike
- Appropriate assistive devices: generally, swivel-wheeled rollators with brakes work well; wheel chairs or scooters will eventually be needed
- Teach safe turns, i.e., U-turns in open spaces, avoiding pivot turns or crossing one foot over the other to turn
- Heel wedge in or on shoe to shift weight anteriorly (may help reduce posterior LOB)
- Home modifications
- Adaptive equipment/devices

Compensatory tips

- Wide, staggered stance with ADL performance
- Scanning environment before walking
- Tilting head down to assist with looking down
- Avoid bending low and standing up quickly to prevent posterior LOB

Research articles

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Winter Y, et al. Health-related quality of life in multiple system atrophy and progressive supranuclear palsy. *Neurodegener Dis*. 2011;8(6): 438-46.

The role of physical therapists (continued)

Corticobasal degeneration (CBD)

CBD presents with an “alien limb” phenomenon, arms and legs that seem to move on their own. Patients also experience increasingly worse apraxia and contractures. Encourage continual exercise, physical activity, and social engagement throughout course of the disease, with appropriate group exercise classes. While the literature on rehabilitation for CBD is limited, it suggests that PT plays a role in managing apraxia, balance, and gait and will be needed at various times throughout the course of the disease.

Treatment strategies

- Patient and caregiver education about the disease
- Aerobic, therapeutic, and balance exercises along with fall prevention training
- Encourage use and exercise of affected limb
- PROM, positioning and bracing to prevent contractures
- ADL training, adaptive devices, energy management, and fall prevention
- For parkinsonian-type gait, focus on large steps, and heel strike
- Teach safe turns, i.e., U-turns in open spaces, avoiding pivot turns or crossing one foot over the other to turn
- Break down tasks into smaller steps
- Appropriate assistive devices; swivel-wheeled rollators with brakes work well only in early stages; apraxia or alien limb often interfere with AD use
- Caregiver training in assistance techniques
- Home modifications and adaptive equipment/devices
- SLP consultation
- OT consultation
- Dystonia and blepharospasms may be helped with Botox

Research articles

Kawahira K, Noma T, Iiyama J, Etoh S, Ogata A, Shimodozono M. Improvements in limb kinetic apraxia by repetition of a newly designed facilitation exercise in a patient with corticobasal degeneration. *Int J Rehabil Res* 2009 Jun; 32(2):178-83.

Stamenova V, Roy EA, Black SE. A model-based approach to understanding apraxia in Corticobasal Syndrome. *Neuropsychol Rev*. 2009 Mar;19(1):47-63.

Multiple system atrophy (MSA)

Patients will benefit from ongoing exercise, physical activity, and social engagement throughout the course of the disease, including appropriate group exercise classes. While the literature on rehabilitation for MSA is limited, it suggests that PT plays a role in managing balance and gait. PT is likely to be needed at various times throughout the course of the disease.

Treatment strategies

- Patient and caregiver education about disease

For bradykinesia and rigidity:

- Levodopa and dopamine agonists may help initially
- Aerobic and flexibility exercises; large movements

For orthostatic hypotension:

- Fludrocortisone, midodrine
- Increase salt in diet
- Rising slowly and resting after position changes
- Support stockings
- Elevate head of bed
- Smaller, more frequent meals
- Avoid increased time in motionless positions
- Avoid warmer temperatures
- Avoid valsalva-provoking maneuvers

For meal time:

- SLP and OT consultations
- Upright posture in chair
- Alternate food and liquid swallows
- Softer foods
- Adaptive devices to help make self-feeding easier and safer

For antecollis:

- Stretching and positioning/bracing
- Tilt wheelchair
- Botox not often used due to possibility of further speech and swallow problems occurring

For parkinsonian-type gait:

- Focus on large steps and heel strike
- Teach safe turns, i.e., U-turns in open spaces, avoiding pivot turns or crossing one foot over the other to turn
- Appropriate assistive devices; generally, swivel-wheeled rollators with brakes work well; wheelchairs or scooters will eventually be needed

MANAGING SYMPTOMS (continued)

The role of physical therapists (continued)

For gait and balance:

- Therapeutic exercise, balance exercise, and fall prevention training

For freezing of gait:

- Don't "fight" the freeze

At the first sign of shuffling or freeze, remember the 4 S's:

Stop

Sigh – take a deep breath

Shift – weight side-to-side

Step – take a large step. Count "1 and 2 and 3..." and then step.

Imagine stepping over something on the ground and then actually step over it.

Place all of your weight on one leg while you swing the other leg back and forth a few times. On the last swing forward, take a step.

Other tips

- Caregiver training in assistance techniques
- Home modifications and adaptive equipment/devices

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Winter Y, et al. Health-related quality of life in multiple system atrophy and progressive supranuclear palsy. *Neurodegener Dis.* 2011;8(6):438-46.

The role of occupational therapists

Christine Robertson Roxberry, OTR/L

While many of the signs and symptoms of PSP, CBD, and MSA appear similar, understanding the issues specific to each disease can help occupational therapists develop and implement more successful treatment strategies.

Progressive supranuclear palsy (PSP)

Treatment strategies

- Patient and carepartner education about disease
- Carepartner training in assistance techniques
- Prism glasses for double vision

Fall prevention training

- Focus on the importance of always scanning the environment (via a downward head tilt) during all functional mobility and ADL in both in the home and outside in the community.
- Appropriate assistive devices; swivel-wheeled rollators with brakes generally work well.
- Teach safe turns during ADL training (i.e., in the kitchen during meal prep and in the bathroom during self-care).
- Using U-turns in open areas in the home or when out in the community.
- Always avoiding pivot turns or crossing one foot over the other to turn; instead move right foot first to turn right, left to turn left.
- Home modifications/adaptive techniques/compensatory strategies/AE/DME.

Compensatory tips

- Always use a wide and staggered stance while performing all ADL tasks.
- Use a shower bench with back and grab bars to eliminate LOB and to promote good posture and energy conservation.
- Use a hand-held shower to eliminate turning while removing soap/shampoo.
- Use a long-handled sponge to eliminate bending over and potential LOB.
- Use liquid instead of bar soap to compensate for decreased coordination.
- Install nonskid surface into tub/shower (nonskid strips).
- Tilt head down to assist with lack of downward eye gaze during all functional mobility and ADL, especially eating.
- Always dress in a seated position to eliminate LOB.
- Raising the height of plate/bowl to face level during meals to compensate for lack of downward eye gaze.
- Use of rocker knives, deep spoons, and food guards to assist with self-feeding skills.
- Reduce background distractions.
- Break down tasks to one step at time.
- Allow for increased response time.
- Try to face the patient when communicating and restate for clarification during conversations.
- PT and SLP consultations.

The role of occupational therapists (continued)

Corticobasal degeneration (CBD)

The role of exercise with this disease is to prevent contractures and disuse atrophy rather than improve coordination. There is no literature to support improvement in FMC via performing in the clinic or in the home.

Signs and symptoms

Initially

- Slowness and stiffness
- Shakiness/UE/LE clumsiness
- Dyphasia and dysarthria and dysphagia
- Changes in gait and balance
- Mild memory or behavior problems

As the disease progresses

- Increased parkinsonian symptoms (rigidity, bradykinesia, postural instability)
- Tremor/myoclonus/dystonia
- Sensory loss
- Increased speech and swallow difficulty
- Cognitive impairments/frontal dementia
- “Alien limb” phenomenon

Treatment strategies

- There is no treatment for “alien limb.”
- Avoid hand flexion/resistive exercise (e.g., Thera Putty®/foam squeezes); instead encourage hand/wrist extension to maintain an open hand for functional tasks.
- Educate/train patient and caregiver on how to improve safety during ADL via modifications to the patient’s routine/performance of tasks as well as to their home environment.

Recommendations

- Use a shower chair with a back and a handheld showerhead to eliminate turning and conserve energy.
- Install a grab bar to decrease risk of falls.
- As the disease progresses, employ PROM, positioning and splinting as necessary to prevent contractures; consider a hand therapist consultation for custom splint fabrication.
- ADL training – Adaptive techniques/compensatory strategies, adaptive devices (adaptive utensils), DME (bed rails/shower chairs), energy conservation, fall prevention.
- Family/caregiver education on how to assist the patient safely throughout disease.
- PT consultation for appropriate assistive device evaluation.
- Weighted utensils do not always help with tremor and self-feeding deficits.
- Generally, swivel-wheeled rollators with brakes work well in the early stages.
- Eventually unable to ambulate due to progressive rigidity/cognitive changes; use caregiver assisted wheelchair.

Cognitive/psychosocial strategies

- Break down tasks into smaller steps; keep instructions simple.
- Reduce background noise/eliminate distractions.
- Stress can increase symptoms; in stressful situations (e.g., out in the community) be sure to simplify tasks.

Multiple system atrophy (MSA)

Occupational therapy may improve functional abilities in patients with mild to moderate MSA.

Signs and symptoms

- Rigidity
- Action tremor, i.e., irregular, jerky, myoclonic
- Bradykinesia
- Freezing of gait/gait and limb ataxia
- Early postural instability and falls
- Orthostatic hypotension
- Erectile dysfunction
- Bladder dysfunction/constipation
- Speech and swallow difficulties; mixed dysarthria tends to emerge earlier in MSA than PD, is more severe, and deteriorates more rapidly
- REM Behavioral Disorder (RBD)—acting out dreams while sleeping due to lack of atonia
- Nystagmus and jerky pursuit
- Antecollis
- Difficulty with thermoregulation
- Cognitive impairment is typically mild

Treatment strategies

- No medications to slow disease progression

Patient and caregiver education about disease

For bradykinesia and rigidity:

- Levodopa and dopamine agonists may help initially.
- Avoid hand flexion/resistive exercise (e.g., Thera Putty®/foam squeezes).
- Encourage hand/wrist extension instead.

For orthostatic hypotension:

- Fludrocortisone, midodrine
- Increase salt in diet
- Rising slowly and resting after position changes
- Support stockings
- Elevate head of bed
- Smaller, more frequent meals
- Avoid increased time in motionless positions
- Avoid warmer temperatures
- Avoid valsalva-provoking maneuvers

Compensatory strategies for self-feeding skills with tremor:

- Use of adaptive devices (adaptive utensils and plate guards).
- Nonskid mat (Dycem®) to prevent the plate/bowl from sliding.
- Arm position—anchor elbow on table, then lower arm down to plate/bowl to retrieve food and then back up to mouth (using the arm as a lever, never taking the elbow up/off of the table).
- Use deep spoons to assist with scooping.
- Elevate the height of the dish bowl.

MANAGING SYMPTOMS (continued)

Functional mobility in the home bedroom:

- Use a bedrail to assist with difficulty rolling.
- Keep floors clutter free and walkways well lit.
- Wear warmer pajamas and use fewer blankets to decrease likelihood of getting tangled up in bed.
- Use a satin pillowcase/sheet under the bottom of the patient to assist with improving bed mobility.
- Always sit as much as possible when dressing to help decrease risk of falls (hypotension).
- Always dress the most affected limb first.

Functional mobility in the home bathroom:

- Grab bar/hand-held showerhead installation.
- Remove glass partitions in the shower/tub to make transfers in/out easier and safer (sit and swing technique vs. step in).
- Use a shower bench/tub chair with a back to decrease risk of slipping, promote good posture, and conserve energy.
- Always sit as much as possible when bathing; eliminate all turning in the shower.
- Avoid extremely hot showers to decrease dizziness/lightheadedness.

Functional mobility in the home kitchen:

- Never overreach; get as close as you can to the object that you are reaching for (e.g., in cabinets).
- Always support yourself with one hand when reaching (e.g., countertop).
- Always stand to the side of the dishwasher/oven/refrigerator when opening.
- Make U-turns in open spaces around the house (swivel-wheeled rollators with brakes work well).
- Avoid pivot turns or crossing one foot over the other to turn; instead move right foot first to turn right, left to turn left.
- Use a wide staggered stance when performing activities (this helps to reduce LOB and retropulsion).
- Side step when working at the counter-space or sink.

Research articles

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The role of speech-language pathologists

Laura Purcell Verdun, MA, CCC-SLP

Progressive supranuclear palsy (PSP)

Changes in swallowing and speech often occur early in PSP, typically with more profound and rapid deterioration compared with Parkinson's disease (PD). Management of swallowing and speech disorders requires changing intervention strategies as the disease progresses.

Dysphagia is one of the most common causes of mortality, with recurrent respiratory infections and respiratory deaths more common in PSP than PD. Early swallowing evaluation and treatment and frequent monitoring of swallowing function allow for problem anticipation and use of supportive measures to minimize complications such as aspiration pneumonia and malnutrition.

No efficacious approach to speech therapy has been documented for this patient population, but speech evaluation and re-evaluation may help to classify motor speech impairment type and clarify the neurodegenerative process. Therapy programs should focus less on speech outcomes alone and be simple and enhance functional communication as quickly and efficiently as possible. Strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline.

Problems with swallowing

Patients often lack awareness of swallowing difficulties including:

- Difficulty looking down at the plate
- Mouth stuffing and rapid drinking
- Tremor or stiffness interfere with self-feeding
- Restricted head and neck posture or hyperextension
- Delayed pharyngeal swallow onset
- Poor cough
- Occasional difficulty opening the mouth

Swallowing management: clinical evaluation should include mealtime observations and suggestions to promote easier and safer swallowing.

- Query the caregiver about swallowing symptoms, as patient may not recognize difficulties.
- Family should maintain a journal of observations to help define and adjust management strategies.
- VFSS (videofluoroscopic swallowing study), if conducted, needs to replicate the home eating environment and rule out non-neurogenic contributions.
- Decompensation and aspiration may not be observed in a controlled VFSS environment, thus severity of dysphagia may not be truly appreciated.
- Discussions regarding feeding tube options should take place sooner rather than later, and repeated frequently.
- Patient and family should agree in advance with a doctor about what can be accomplished with or without placement of a feeding tube.

The role of speech-language pathologists (continued)

Swallowing treatment strategies

- Optimize oral hygiene.
- Supervise mealtimes.
- Offer moist, soft, tender, and well-lubricated foods; avoid dry, particulate, textured foods.
- Keep the plate in the line of vision.
- Maintain head in a chin-tucked position.
- Restrict liquid and food bolus volumes.
- Make sure food is swallowed before taking more.
- Put cup and utensils down between bites and sips.
- Look for mealtime adaptive devices including cups, plates, and utensils to assist with self-feeding.
- Give medications with a pureed food.
- Ask the neurologist about anticholinergic drugs or botulinum toxin for management of secretions.

Speech problems

Mixed dysarthria, typically including hypokinetic and spastic dysarthria; ataxic features are less common.

- Strained voice, impaired speech fluency with slow rate of speech and palilalia (the compulsive repetition of utterances in context of increasing rate and decreasing loudness), and emotional lability.
- Language and cognitive deficits including frontotemporal dementia and progressive nonfluent aphasia have been observed and can interfere with therapy efforts.
- Progresses to anarthria.

Speaking strategies

Speaking must become a conscious effort and to enhance intelligibility. Patients must use compensatory strategies including:

- Breathing first and speaking loudly and slowly.
- Keep sentences short.
- Repeat entire sentence when necessary instead of isolated words.
- Say one sentence at a time without immediate repetition.
- Establish the context.
- Use gestures.
- LSVT® (www.lsvtglobal.com) techniques emphasizing increased phonatory effort may be of benefit but difficult to habituate.
- Investigate assistive forms of communication such as a communication board or speech-generating device to supplement natural speech, depending on an individual's visual, cognitive and motor limitations.
- Simple augmentative communication will be required in later stages.
- Consider personal portable amplifiers.

Listener strategies

- Keep comments and questions brief.
- Stick with familiar topics, and one topic at a time.
- Use “yes/no” question format.
- Ask for clarification, “Did you say...?”.
- Provide choices to ease decision making.

Research articles

Goetz CG, Leurgans S, Lang AE, Litvan I. Progression of gait, speech and swallowing in progressive supranuclear palsy. *Neurology*. 2003. 60(6):917-22.

Wenning GK, Litvan I, Tolosa E. Milestones in atypical and secondary Parkinsonisms. *Movement Disorders*. 2011. 26(6):1083-1095.

Corticobasal degeneration (CBD)

Changes in swallowing and speech often occur early in CBD and are typically more severe and deteriorate more rapidly than Parkinson's disease (PD). Management requires changing intervention strategies as the disease progresses.

No efficacious approach to speech therapy has been documented for this patient population. It may help, however, to evaluate and re-evaluate motor speech impairment type and clarify the neurodegenerative process. Therapy should be simple and enhance functional communication as quickly and efficiently as possible instead of focusing on speech outcomes alone and strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline.

Swallowing function should be evaluated early and monitored frequently to anticipate problems and provide appropriate supportive measures. Dysphagia is one of the most common causes of mortality, with recurrent respiratory infections and respiratory deaths more common in CBD than PD. The goal of swallowing therapy is to minimize complications such as aspiration pneumonia and malnutrition through early symptomatic treatment.

Problems with swallowing

- Impaired self-feeding
- Slow or incomplete chewing
- Oral and swallowing apraxia
- Slowed swallowing movements
- Patients may be aware of swallowing difficulties

Swallowing management

- Clinical swallowing evaluation should include mealtime observations and suggestions to promote easier and safer swallowing.
- Family should maintain a journal of observations to help define and adjust management strategies.
- VFSS (videofluoroscopic swallowing study), if conducted, needs to replicate the home eating environment and rule out non-neurogenic contributions.
- Decompensation and aspiration may not be observed in a controlled VFSS environment, thus severity of dysphagia may not truly be appreciated.
- Discussions regarding feeding tube options should take place sooner rather than later, and repeated frequently.
- Patient and family should agree in advance with a doctor about what is possible with or without placement of a feeding tube.

The role of speech-language pathologists (continued)

Swallowing treatment strategies

- Optimize oral hygiene.
- Supervision at mealtimes.
- Use less affected side for self-feeding.
- Maintain head in a chin-tucked position.
- Avoid highly textured, particulate foods.
- Blend multiple consistency foods.
- Proceed with caution with thin liquids.
- Alternate food and liquid swallows.
- Provide medications with pureed food.
- Use mealtime adaptive devices including cups, plates, and utensils to assist with self-feeding.
- Consider smaller, more frequent meals to shorten duration of mealtimes.
- Ask the neurologist about the role of anticholinergic drugs or botulinum toxin for management of secretions.

Speech problems

- Characterized by hypokinetic and spastic dysarthria, as well as progressive apraxia of speech and oral apraxia.
- Progressive nonfluent aphasia may be strongly associated with this diagnosis.
- Speech is hesitant and halting, with strained voice and slower speech production.
- Initially may have intact written language.
- Frontotemporal dementia may be present.
- Progresses to anarthria.

Patient speaking strategies

- Speech must become a conscious effort.
- Use compensatory strategies to enhance intelligibility.
- Use short phrases and simpler language because of increased errors with increased rate of speech, number of syllables, and complexity of language.
- Optimize use of written language.
- Use of gestures may be limited by apraxia.
- Be alert to “yes/no” confusion.
- Investigate assistive forms of communication such as a communication board or speech generating device to supplement natural speech, depending on patients’ cognitive and motor limitations.
- Simple augmentative communication will be required in later stages.

Listener strategies

- Eliminate distractions to reduce background noise.
- Face the speaker with CBD.
- Keep comments and questions brief.
- Stick with familiar topics, and one topic at a time.
- Use “yes/no” question format.
- Ask for clarification, “Did you say...?”.

Research articles

Josephs KA, Duffy JR. Apraxia of speech and nonfluent aphasia: a new clinical marker for corticobasal degeneration and progressive supranuclear palsy. *Current Opinion in Neurology*. 2008. 21:688-692.

Reich SG, Grill SE. Corticobasal degeneration. *Current Treatment Options in Neurology*. 2009. 11:179-185.

Multiple system atrophy (MSA)

The management of swallowing and speech disorders in MSA requires changing intervention strategies as the disease progresses. Mixed dysarthria is common and tends to emerge earlier in MSA than in PD, is more severe, and progresses more rapidly. No efficacious approach to speech therapy has been documented for this patient population. Up to one-third of individuals with MSA may have laryngeal stridor possibly caused by vocal fold abductor paresis or laryngeal dystonia. Therefore, a therapy program should be simple and enhance functional communication as quickly and efficiently as possible and strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline. Speech evaluation and re-evaluation may help to classify motor speech impairment and clarify the neurodegenerative process. Evaluate and monitor swallowing early and frequently to anticipate problems and maximize supportive measures.

Problems with swallowing

- Difficulty sitting upright at mealtimes.
- Tendency towards bolus holding in oral cavity, and discoordinated oral bolus formation and propulsion.
- Pharyngeal weakness and disruption of the cricopharyngeal segment.
- Excessive accumulation of pharyngeal secretions.
- Vocal fold motion impairment may compromise airway protection as the disease progresses.
- May have tracheotomy.
- Cough may also be compromised.

Swallowing management

- Clinical evaluation should include mealtime observations and suggestions for easier and safer swallowing.
- Family should maintain a journal of observations to help define and adjust management strategies.
- VFSS (videofluoroscopic swallowing study), if conducted, needs to replicate the home eating environment and rule out non-neurogenic contributions.
- Decompensation and aspiration may not be observed in a controlled VFSS environment, thus severity of dysphagia may not truly be appreciated.
- Discussions regarding feeding tube options should take place sooner rather than later, and revisited often.
- Patients and their families should agree in advance with a doctor about what can be accomplished with or without placement of a feeding tube.

Swallowing treatment strategies

- Optimize oral hygiene.
- Positioning/support to sit upright against the chair back.
- Maintain head in a chin-tucked position.
- Alternate food and liquid swallows.
- Restrict bolus volumes.

The role of speech-language pathologists (continued)

- Moist, soft, tender foods with lubrication; blend multiple consistency items.
- Use mealtime adaptive devices including cups, plates, and utensils to assist with self-feeding.
- Consider smaller, more frequent meals to shorten duration of mealtimes.
- Ask the neurologist about the role of anticholinergic drugs or botulinum toxin for management of secretions.
- There is the potential for lowered blood pressure following meal times (postprandial hypotension).

Speech problems

MSA-P: hypokinetic dysarthria is expected, sometimes mixed with spastic or hyperkinetic dysarthria, and hypophonia (parkinsonian)

MSA-A: often ataxic or hypokinetic dysarthria, but may be mixed with spastic dysarthria (autonomic)

MSA-C: ataxic dysarthria is most often expected, or in combination with spastic dysarthria (cerebellar)

- Typically, more changes in speech than voice.
- Cognitive impairment if present is typically mild.

Speaking strategies

- Speech must become a conscious effort.
- Emphasize taking a breath before speaking.
- Reduce rate of speech to improve coordination and accuracy.
- Intelligibility drills with exaggeration of articulation movements.
- Investigate assistive forms of communication such as a communication board or speech-generating device to supplement natural speech, considering motor and cognitive limitations.
- Simple augmentative communication will be required in later stages.
- Consider personal portable amplifier.

Listener strategies

- Eliminate distractions to reduce background noise.
- Face the speaker with MSA.
- Keep comments and questions brief.
- Stick with familiar topics, and one topic at a time.
- Use “yes/no” question format.
- Ask for clarification, “Did you say...?”.
- Provide choices to ease decision making.

Research articles

Tada M, Onodera O, Tada M, Ozawa T, Piao YS, Kakita A, et al. Early development of autonomic dysfunction may predict poor prognosis in patients with multiple system atrophy.

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Using Physical Therapy to Help Manage Mobility Issues

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Physical therapists (PT)

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 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 carepartners 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 neurodegenerative diseases are to:

- Maintain functional and safe mobility for as long as possible.
- Prevent and/or limit falls and injuries.
- Educate and train carepartners 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 she or he 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 her or his carepartner.
- Evaluate and treat walking problems, loss of balance, and falls.

MANAGING SYMPTOMS (continued)

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. 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 mobility.
- Teach carepartners proper body mechanics and techniques for assisting with mobility. When patients with one of these diseases can no longer safely move on their own, carepartners often have to assist with mobility. Learning proper mobility assistance techniques will help keep carepartners free from injuries such as back strains.
- Make referrals to movement and exercise programs in the community. Exercising with a group not only promotes mobility and fitness, but also helps with maintaining social interaction and limiting isolation.
- Make recommendations 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.”

Carepartner 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.)

Never pull partner by the neck or arms; rather, guide partner by placing your hands on her or his 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!

Adapting to Adaptability: An Occupational Therapist's Perspective

Dr. Tony Gentry

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 carepartners 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 one 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, 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. She or he 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 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.

MANAGING SYMPTOMS (continued)

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 carepartner 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 carepartner 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 carepartners 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
- Neuro-optometric Rehabilitation Association • www.nora.cc



Adapting to Swallowing Problems in PSP, CBD, and MSA

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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 causes of death are 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 toward 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 are 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.

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

MANAGING SYMPTOMS (continued)

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 including:

- 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 mealtime
- 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
- 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, multifactor 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 that may enhance the ease and safety of swallowing, assist in patient education, 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?

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

MANAGING SYMPTOMS (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 book or other stable object that will provide sufficient elevation (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
- Independence Spillproof Flo Tumbler: 888- 843-5287 • www.kcup.com
- Wedge Cup: 985-722-8269 • www.wedgecup.net
- Provale Cup: 423-265-3574 • www.meyerpt.com/provale-cup
- Maroon Spoons: 800-225-2610 • www.alimed.com/maroon-spoons.html
- Scooper Plate w/nonskid Base: 913-390-0247 • www.southpaw.com/non-skid-scooper-bowls-and-plates.html
- Skidtrol Non-Skid Bowl: 973-628-7600 • www.maddak.com/skidtrol-nonskid-bowl-p-27963.html
- Bruce Medical Supply: 800-225-8446 • www.brucemedical.com

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 on page 74. 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. 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.

MANAGING SYMPTOMS (continued)

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
- RESOURCE ThickenUp: 888-240-2713 • www.nestlenutritionstore.com
- SimplyThick: 800-205-7115 • www.simplythick.com
- Thick & Easy: 800-866-7757 • 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
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 noncooked 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
- Plak-Vac & Plak-Vac/Res-Q-Vac oral suction toothbrush system: 800-325-9044 • 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.
3. Make sure your toothpaste is fluoridated.
4. Use nonalcohol-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.

MANAGING SYMPTOMS (continued)

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 percutaneous 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

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Problems to anticipate 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 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's disease (PD) may provide 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 nonfluent 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.

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.

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

MANAGING SYMPTOMS (continued)

Some specific strategies for the speaker

It may be useful to consult with a speech pathologist who will recommend specific exercises to address limitations in speech and voice. Here are some strategies that may enhance the speaker's success:

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

Reports of successful speech intervention for individuals with PSP, CBD, and MSA are rare, and strategies beneficial to the voice and speech impairments of PD may have limited impact. Reasons for limited success include the presence of cognitive impairment, the relatively rapid progression of the disease, 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 that emphasizes good speaking habits before the underlying problems become so severe that new learning is difficult. It is critical to discuss the limitations and severity of the communication problem with the therapist, as well as the communication needs. Therapy efforts should focus on increasing loudness, articulatory precision, minimizing repetitions in the setting of palilalia, and enhancing awareness and participation in communication efforts. A therapy program called the Lee Silverman Voice Treatment® (LSVT®) (www.lsvtglobal.com) is frequently applied to the communication changes in 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 are low volume, rapid 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, it 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. Text can often 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.

Proloquo2Go: www.assistiveware.com/product/proloquo2go

Speak it!: itunes.apple.com/us/app/speak-it!-text-to-speech/id308629295?mt=8

How can I find someone to recommend a communication device?

The speech pathologist is responsible for evaluating patients and training them to use the communication systems. Ask if they have experience in working with augmentative communication devices and can determine which device best meets the needs and abilities of a particular patient. 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 patients and families to make informed choices 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:

ASHA (American Speech-Language-Hearing Association): 800-638-8255 • www.asha.org

ISAAC (International Society for Augmentative & Alternative Communication):
905-850-6848 • www.isaac-online.org

RESNA Technical Assistance Project: 703-524-6686 • www.resna.org

USSAAC (United States Society of Alternative & Augmentative Communication):
215-631-1877 • 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

Spokeman: 800-255-3408 • www.luminaud.com/spokeman.htm

Speech Enhancer: 314-467-0772 • www.speechenhancer.com

Communicating in an emergency

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

Managing Cognitive Changes

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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 carepartner will have an easier time dealing with symptoms).

Managing Difficult Behaviors

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Agitation

Potential causes or antecedents

Management strategies

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.

Sleep disturbance

Potential causes or antecedents

Management strategies

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.

MANAGING SYMPTOMS (continued)

Difficulty with personal care tasks

Potential causes or antecedents

Management strategies

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.

Incontinence

Potential causes or antecedents

Management strategies

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.

Wandering

Potential causes or antecedents

Management strategies

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.

Suspiciousness or paranoia

Potential causes or antecedents

Management strategies

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 SYMPTOMS (continued)

Inappropriate or impulsive sexual behavior

Potential causes or antecedents

Management strategies

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.



PART FOUR

Being A Carepartner



Make Meaning to Stay Positive

Janet M. Edmunson, MEd

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

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

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

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

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

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

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

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

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

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

Caregiver Stress and Burnout

Tips for Regaining Your Energy, Optimism, and Hope

<http://www.helpguide.org>

Caregiving

The demands of caregiving can be overwhelming, especially if you feel you have little control over the situation or you're in over your head. If the stress of caregiving is left unchecked, it can take a toll on your health, relationships, and state of mind – eventually leading to burnout. When you're burned out, it's tough to do anything, let alone look after someone else. That's why taking care of yourself isn't a luxury – it's a necessity. Read on for tips on how to rein in the stress in your life and regain balance, joy, and hope.

What you can do?

- Get out of the house and walk in the sunlight.
- Reach out and stay connected to people who support you.
- Join a caregiver support group to share your experiences.
- Get the amount of restful sleep that you need to feel your best.

Caregiver stress and burnout: What you need to know

Caring for a loved one can be very rewarding, but it also involves many stressors. Caregiver stress can be particularly damaging, since it is typically a chronic, long-term challenge. You may face years or even decades of caregiving responsibilities. It can be particularly disheartening when there's no hope that your family member will get better.

If you don't get the physical and emotional support you need, the stress of caregiving leaves you vulnerable to a wide range of problems, including depression, anxiety, and burnout. And when you get to that point, both you and the person you're caring for suffer. That's why managing the stress levels in your life is just as important as making sure your family member gets to his doctor's appointment or takes her medication on time.

Signs and symptoms of caregiver stress and burnout

Learning to recognize the signs of caregiver stress and burnout is the first step to dealing with the problem.

Common signs and symptoms of caregiver stress

- Anxiety, depression, irritability
- Feeling tired and run down
- Difficulty sleeping
- Overreacting to minor nuisances
- New or worsening health problems
- Trouble concentrating
- Feeling increasingly resentful
- Drinking, smoking, or eating more
- Neglecting responsibilities
- Cutting back on social outlets

BEING A CAREPARTNER (continued)

Common signs and symptoms of caregiver burnout

- You have much less energy than you once had.
- It seems like you catch every cold or flu that's going around.
- You're constantly exhausted, even after sleeping or taking a break.
- You neglect your own health.
- Your entire life revolves around caregiving.
- You have trouble relaxing, even when help is available.
- You're increasingly impatient and irritable with the person you're caring for.
- You feel helpless and hopeless.

Once you burn out, caregiving becomes a health problem often both physically and emotionally for the caregiver and has negative consequences for the patient as well. It's important to watch for the warning signs of caregiver burnout and take action right away when you recognize the problem.

Don't let caregiving take over your whole life. It's easier to accept a difficult situation when there are other areas of your life that are rewarding. Invest in things that give you meaning and purpose – whether it's your family, church, a favorite hobby, or other outlet. Read on for some additional tips to lighten the load:

Find ways to feel empowered

Feeling powerless is the number one contributor to burnout and depression. And it's an easy trap to fall into as a caregiver, especially if you feel stuck in a role you didn't expect or helpless to change things for the better. But no matter the situation, you aren't powerless. This is especially true when it comes to your state of mind. You can't always get the extra time, money, or physical assistance you'd like, but you can always work on your thoughts about the situation which, in turn, will affect your feelings.

Embrace your role as a caregiver. Acknowledge that, despite any resentments or burdens you feel, you have made an important decision to provide care. Focus on the positive reasons behind that choice. These deep, meaningful motivations can help sustain you through difficult times.

Focus on the things you can control. You can't wish the disease away or force your brother to help out more. Rather than stressing out over things you can't control, focus on the way you choose to react to problems. Celebrate the small victories. If you start to feel discouraged, remind yourself that all your efforts matter. You can't cure your loved one's disease but you are making a difference. Don't underestimate the importance of making your loved one feel more safe, comfortable, and loved.

Get the appreciation you need

Feeling appreciated can go a long way toward not only accepting a stressful situation, but enjoying life more. Studies show that caregivers who feel appreciated experience greater physical and emotional health. Caregiving actually makes them happier and healthier, despite its demands. But what can you do if the person you're caring for is no longer able to feel or show their appreciation for your time and efforts?

Imagine how your loved one would respond if he or she was healthy. If he or she wasn't burdened by a disease, how would your loved one feel about the love and care you're giving? Remind yourself that the person would express gratitude if he or she was able.

Applaud your own efforts. If you're not getting external validation, find ways to acknowledge and reward yourself. Remind yourself of the good you're doing. If you need something more concrete, try making a list of all the ways your caregiving is making a positive difference. Refer back to it when you start to feel low.

Talk to a supportive family member or friend. Positive reinforcement doesn't have to come from the person you're caring for. When you're feeling unappreciated, turn to friends and family who will listen to you and acknowledge your efforts.

Ask for help

Taking on all of the responsibilities of caregiving without regular breaks or assistance is a surefire recipe for burnout. Don't try to do it all alone. Look into respite care. Or enlist friends and family who live near you to run errands, bring a hot meal, or stay with your loved one so you can take a well-deserved break.

Tips for getting the caregiving help you need

Speak up. Don't expect friends and family members to automatically know what you need or how you're feeling. Be up front about what's going on with you and the person you're caring for. If you have concerns or thoughts about how to improve the situation, express them – even if you're unsure how they'll be received. Get a dialogue going.

Spread the responsibility. Try to get as many family members involved as possible. Even someone who lives far away can help. You may also want to divide up caregiving tasks. One person can take care of medical responsibilities, another with finances and bills, and another with groceries and errands, for example.

Set up a regular check-in. Ask a family member, friend, or neighbor to call you on a set basis (every day, weekly, or how ever often you think you need it). This person can help you spread status updates and coordinate with other family members.

Say “yes” when someone offers assistance. Don't be shy about accepting help. Let them feel good about supporting you. It's smart to have a list ready of small tasks that others could easily take care of, such as picking up groceries or driving your loved one to an appointment.

Be willing to relinquish some control. Delegating is one thing. Trying to control every aspect of care is another. People will be less likely to help if you micromanage, give orders, or insist on doing things your way.

Give yourself a break

As a busy caregiver, leisure time may seem like an impossible luxury. But you owe it to yourself – as well as to the person you're caring for – to carve it into your schedule. Give yourself permission to rest and to do something every day that you enjoy. You will be a better caregiver for it.

There's a difference between being busy and being productive. If you're not regularly taking time off to de-stress and recharge your batteries, you'll end up getting less done in the long run. After a break, you should feel more energetic and focused, so you'll quickly make up for your relaxation time.

BEING A CAREPARTNER (continued)

Maintain your personal relationships. Don't let your friendships get lost in the shuffle of caregiving. These relationships will help sustain you and keep you positive. If it's difficult to leave the house, invite friends over to visit with you over coffee, or a light meal. They may even be willing to bring in some take-out food!

Prioritize activities that bring you enjoyment. Make regular time for things that bring you happiness, whether it's reading, working in the garden, tinkering in your house, knitting, playing with the dogs, or whatever works for you.

Make yourself laugh. Laughter is an excellent antidote to stress – and a little goes a long way. Read a funny book, watch a comedy, or call a friend who makes you laugh. And whenever you can, try to find the humor in everyday situations.

Get out of the house. Seek out friends, family, and respite care providers to step in with caregiving so you can have some time away from the home.

Take care of your health

Think of your body like a car. With the right fuel and proper maintenance, it will run reliably and well. Neglect its upkeep and it will start to give you trouble. Don't add to the stress of your caregiving situation with avoidable health problems.

Keep on top of your doctor visits. It's easy to forget about your own health when you're busy with a loved one's care. Don't skip check-ups or medical appointments. You need to be healthy in order to take good care of your family member.

Exercise. When you're stressed and tired, the last thing you feel like doing is exercising. But you'll feel better afterwards. Exercise is a powerful stress reliever and mood enhancer. Any exercise is better than none at all. When you exercise regularly, you'll also find it boosts your energy level and helps you fight fatigue.

Meditate. A daily relaxation or meditation practice can help you relieve stress and boost feelings of joy and well-being. Try yoga, deep breathing, progressive muscle relaxation, or mindfulness meditation. Sometimes people who have a religious affiliation find strength in their beliefs and the use of prayer. Even a few minutes of quiet meditation in the middle of an overwhelming day can help you feel more centered.

Eat well. Nourish your body with foods that will fuel you with steady energy. Make eating as simple as possible and don't skip meals.

Don't skimp on sleep. Cutting back on time in bed is counterproductive – at least if your goal is to get more done. Most people need more sleep than they think they do. When you get less, your mood, energy, productivity, and ability to handle stress will suffer. If nighttime sleep is very difficult to arrange, take a nap during the day to regroup.

Join a support group

A caregiver support group is a great way to share your troubles and find people who are going through the same experiences that you are living each day. If you can't leave the house, many Internet groups are also available.

In most support groups, you'll talk about your problems and listen to others talk; you'll not only get help, but you'll also be able to help others. Most important, you'll find out that you're not alone. You'll feel better knowing that other people are in the same situation, and their knowledge can be invaluable, especially if they're caring for someone with the same disease as you are.

Local vs. online support groups for caregivers

Local support groups:

People live near each other and meet in a given place each week or month.

You get face-to-face contact and a chance to make new friends who live near you.

The meetings get you out of the house, get you moving provide a social outlet, and reduce feelings of isolation.

Meetings are at a set time. Therefore make a regular plan for someone to be with your loved one when you're at the group. You will need to attend them regularly to get the full benefit of the group. Since the people in the support group are from your area, they'll be more familiar with local resources and issues.

Online support groups:

You meet online, through email lists, websites, message boards, or social media.

You can get support without leaving your house, which is good for people with limited mobility or transportation problems. To find a support group it's often best to start by visiting the website or calling an organization dedicated to the problem. Please contact CurePSP's website www.curepsp.org for more information about support groups or call toll-free: 800-457-4777.



The 7 Deadly Emotions of Caregiving

Paula Spencer Scott

Caring.com

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

Caregiver emotion trap #1: guilt

Guilt is virtually unavoidable as you try to “do it all.”

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

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

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

Caregiver emotion trap #2: resentment

This emotion is still so taboo that many caregivers are loathe to admit it.

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

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

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

See also: Chapter on Managing Symptoms and Getting Professional Support.

Caregiver emotion trap #3: anger

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

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

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

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

Caregiver emotion trap #4: worry

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

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

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

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

Caregiver emotion trap #5: loneliness

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

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

BEING A CAREPARTNER (continued)

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

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

Caregiver emotion trap #6: grief

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

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

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

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

Caregiver emotion trap #7: defensiveness

Protecting yourself is good – to a point.

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

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

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

Keeping Families Strong

Patient and Carepartner Advocacy Committee, CurePSP

PSP, CBD, and related prime of life diseases are very challenging, and have significant impact on patients as well as their families. One important means of coping is to explore ways to grow in strength as a family while going through this difficult journey.

There are many factors that make families strong. We each have a role in our families – a role that develops over time, but one that helps to shape our interactions and to define our place regardless of the size of our family. An important characteristic of strong families is the expression of thoughts and feelings with one another, even when there are serious disagreements; it is through opening up to one another that the potential for greater closeness exists. Of course, as with all communication, it's important to be respectful, listen to one another, and try to understand even views that are contrary to our own. Time must be devoted to allow for a discussion to take place and privacy is essential.

Every family has its own history and traditions that relate to various stages of life and these memories of shared experiences can bring a wide range of emotions and a sense of togetherness. When a prime of life disease strikes, difficult issues that the family members have had all along can worsen; at the very least, it can be overwhelming to take on the responsibility of caring for a loved one. A medical diagnosis of PSP, CBD, or related disease changes everything for the patient and the family.

But in addition to the importance of communicating, one of the most significant characteristics of strong families is a willingness to help one another and to share responsibilities. It is often at times of illness that families bond in ways they couldn't have imagined.

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." The "new normal" is always changing as the disease worsens over time and it's the ability of the family to be flexible and dedicated to their loved one that makes it possible to have courage.

An additional complication for families is that it is common for family members to live in different areas of the country from where their parents live, and generally speaking, families are smaller in size than they were in the past. When a parent becomes ill, their adult children may be raising their own families in addition to coping with the stress of career demands. This "sandwich" generation is then often confronted with the multiple challenges of long-distance caregiving.

PSP and related neurodegenerative diseases often strike when people are in middle age. They have, perhaps, been looking forward to having more time to enjoy family and other activities, but their plans drastically change in anticipation of their future medical needs. It is also very common to have difficulty locating medical professionals who have experience with this ultra-rare disease. For many, there are significant financial constraints in terms of the ability to afford medical services, and social supports may be limited.

BEING A CAREPARTNER (continued)

The strength of a family is tested at diagnosis – although diagnosis is a process, not an event. Everyone is different and it's important to respect the ways in which people express their emotions, knowing that emotions will change over time. Especially with initial diagnosis, there is no right way to react.

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

The Leader. Often 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. Leaders manage their emotions by being active.

As with all roles we take on, there is also a negative side. The Leader can become overburdened 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 the Leader can grow to resent 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.

The Supporter. Another family role is the Supporter. Supporters are willing to help, but set 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 is likely more manageable than asking her or him 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 Leaders, who view themselves as doing the lion's share of what needs to be done.

The other side of the Supporter's struggle can be a sense of being poorly informed about the patient's disease and treatments. Just as their involvement tends to be task-oriented, Supporters may not have a full understanding of what is happening with their loved ones and may not feel able to ask for more information from the Leader. Supporters 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.

The Bystander. It seems that almost every family has one or more members who pull away during the course of the disease. The Bystander may reduce 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 Bystander's lack of involvement often makes the other family members feel taken advantage of, disappointed, frustrated, and/or angry. However, the Bystander may be expressing lot of emotions that others in the family are less comfortable expressing, albeit by indirect means. The Bystander often feels helpless and ashamed for not visiting or helping their loved one. They may feel unable to talk about their feelings, as they don't feel worthy of support. While this can be a risk for all family members, Bystanders may become depressed, which can be difficult to recognize due to their isolating behavior.

The Only One. It may be the case that only one person, perhaps the spouse/partner, an adult child or a sibling, is responsible for the care of a person with the disease. The enormity of this role clearly illustrates the need for additional supports, be they paid or unpaid, to help with day-to-day activities as well as to assist in emergency situations. The Only One will experience a wide range of emotions and will likely feel isolated over time. The physical and mental health of this person may not be well known to those around her or him, so if you or someone you know is the only carepartner, it is wise to seek outside supports such as support groups and other services offered by CurePSP. While there is no one formula for success, a sole carepartner will burn out without help.

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 under such difficult circumstances.

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 essential to acknowledge that everyone is human and everyone is dealing with multiple stressors all at the same time. Express your concerns and your fears and 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 important, stay close to your loved one who has the disease. Remember who they are, what you love about them, and that each moment you share is precious.

CurePSP Support Groups

Wendy M. Resnick, R.N., M.S., CS.

What is a CurePSP support group?

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

Who attends CurePSP support group meetings?

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

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

Professionals may be invited to attend support groups; for example, nurses, social workers, physicians – anyone who serves as a “paid helper” attempting to provide assistance to persons with these diseases and their families.

What happens at meetings?

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

What do people learn?

- Information about PSP and related neurodegenerative brain diseases.
- What are typical symptoms?
- What constitutes a thorough evaluation and diagnosis?
- How do these diseases vary from person to person?
- What can be expected as the disease progresses?
- What treatment options and/or clinical trials exist?

Information about available resources in the community

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

Information about identifying methods of coping

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

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

How do I find a CurePSP support group?

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

Caring From a Distance

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Adult children and other close relatives who live far away from loved ones coping with PSP and related diseases 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's 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 carepartner 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 relocating?

Perhaps some of these ideas contributed by support group members around the country will meet some needs for you and your family:

- 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 10-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.
- 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 example, for an 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.
- Send a surprise package to the frontline carepartner 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!

- Budget funds for regular trips to check on your family. Don't add to the primary carepartner's stress level by expecting her or him 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.
- Learn about the medications your person with PSP or a related disease takes. Encourage your family to communicate problems and changes to their physicians. Inquire about the health of the parent who is not ill. Is your mom's mammogram overdue? Does your dad neglect his own medical check-up because he's preoccupied 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.
- Strongly encourage and provide a three- to five-day respite for the primary carepartner once or twice a year. Plan to arrive a day or two prior to the carepartner's "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!
- If your parents are able to travel, arrange for them to visit you. For some couples living with a disease such as 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.
- Make a family pact that no one will make promises to the patient 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. 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

These practical tips for long-distance family members who are not primary carepartners barely touch the concerns of those of you who live far away from elderly parents yet serve as their primary carepartners.

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.

BEING A CAREPARTNER (continued)

Anna W. 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 carepartner 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 fact of people living longer lives has resulted in the phenomenon that many carepartners, 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 her or his services. Information to help locate a geriatric care manager can be found at www.aginglifecare.org.





BEING A CAREPARTNER (continued)

Travel Tips

Diane Breslow, MSW, LCSW

Consulting from Travel-On Travel Management Co.

Travel is accessible for many people with PSP, CBD, and other related neurodegenerative diseases. 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.

Before you start

- 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. (See “Insurance” below.)
- Ask your neurologist if she or 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 Parkinson’s disease 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.
- If traveling with a wheelchair, refrigerated medications, implanted medical devices, or other special needs, call TSA Cares (855-787-2227) at least 48 hours prior to departure.
- Wear a medical alert bracelet/necklace and also carry a back-up alert in your purse or wallet.
- Contact CurePSP for free wallet cards for PSP, CBD, and MSA.
- Carry a wallet card giving the name and contact for your doctor, insurance information, list of medications and dosages, list of medical conditions, allergies, and blood type.
- Carry a doctor’s letter identifying all medications – including generic name, dosage, frequency, and reason for taking each.
- Carry a doctor’s letter explaining any implanted medical devices.
- Ensure all doctor’s letters reference you by the exact same name as on your travel documents.
- Hydrate well before, during, and after air travel to help reduce the risk of deep vein thrombosis and the effects of jet lag.

Insurance

- Research insurance options carefully. Many will exclude pre-existing conditions. Any change in medications during the “look-back” period (varies, but often 60-120 days) will classify the condition as “pre-existing.” Thus, Type 1 diabetes, for which the insulin dosage changes constantly, will nearly always be considered a pre-existing condition, whereas a chronic heart condition for which there have been no changes to medication in the past six months may not be excluded.
- Ensure your medical insurance covers lost, stolen, or confiscated medications.
- Entitlement to emergency health care is not universal and in many countries will not be given without proof of insurance or other means to pay for treatment.
- Medicare/Medicaid does not provide overseas coverage.

About medications

- Be aware that some medications are not permitted in some countries. Most commonly these are narcotics and psychotropic drugs, but also include allergy medications and hormones. Some countries will allow foreign travelers to bring a limited supply into the country with a physician's documentation, but some countries will not. Many countries limit travelers to a 30-day supply of medications. Have a back-up plan. Know that it may not be possible to mail medications from the U.S., and some drugs may not be available at all, or in the same dosages, in other countries.
- 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 nongeneric 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 two watches: current time, and time at home.
- Liquid medications are subject to the TSA limit of 3.4 ounces per bottle. Multiple bottles are permitted.
- Notify the airline at least 48 hours in advance if traveling with a wheelchair or portable oxygen concentrator.
- All oxygen must be via an approved (specific models) portable oxygen concentrator (POC).
- With documentation of medical need, and with proper labeling, syringes are permitted on board.

At travel terminals

- If necessary, request wheelchair or electric cart service within terminals. (Your bags will be handled too.)
- Check in early – much better to relax at the gate than to rush to get there!
- 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.
- People who require a wheelchair or scooter must have physician's written "certificate of need." Medical devices in their own carrier may be counted as a carry-on item by some airlines. Devices in continual use (not packed in their own carrier), including wheelchairs, canes, walkers, insulin pumps, portable oxygen concentrators, etc., do not count against the carry-on allowance.
- People in wheelchairs can request private, rather than public, checkpoint screenings.
- Many airlines today limit preassigned seats and charge an additional cost for certain seats (aisles near the front, for example). Be aware that you may need to pay a fee to obtain an appropriate seat assignment.

BEING A CAREPARTNER (continued)

- Abide by TSA rules regarding bringing snacks and drinks through security. Purchase drinks after clearing security or bring an empty, refillable water bottle from home and fill it before boarding the aircraft.
- Service animals are permitted on aircraft with proper documentation and advance notice.

For more information about traveling by air with a disability, contact the Transportation Security Administration at www.tsa.gov.

Rail travel specifics

- Have valid photo identification.
- Amtrak trains can accommodate most wheelchairs; Amtrak may make random checks of wheelchairs.
- Most, but not all, Amtrak stations are wheelchair accessible.
- Reservations for wheelchair space and transfer seating are recommended on all Amtrak trains, including those where reservations do not normally apply.
- A service animal can travel with the passenger.
- Amtrak offers a discount on an accompanying attendant.
- For more information about traveling by train with a disability, contact Amtrak at www.amtrak.com.

Bus travel specifics

- Greyhound buses are equipped with wheelchair lifts.
- Greyhound assists with boarding, debarkation, luggage, transfers, and 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 bus with a disability, contact Greyhound at 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” 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 neurological condition hold you back from the trip of your dreams! Bon Voyage!

PART FIVE

Making Plans



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
- www.ssa.gov/dibplan/ (Disability Planner)

Eligibility and qualification

In order to be eligible for disability benefits, you must meet the medical and nonmedical criteria for the Social Security Disability Program (SSDI). For additional information about the SSDI program, please visit www.ssa.gov.

CurePSP will not be able to answer any questions regarding eligibility for disability benefits or qualification regarding 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/
- 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/

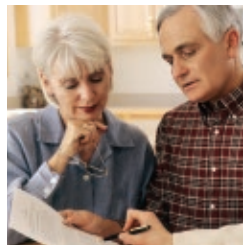
If you have already applied for disability benefits, the Compassionate Allowances fast-tracking process should be used during the initial application or at the reconsideration, hearing, or Appeals Council levels. It is advisable to also notify the disability examiner assigned to your claim that your 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 62-65), 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 nonmedical criteria based on income. For additional information about the SSI program, please visit this website: www.ssa.gov/ssi.



Support and Resources

Seeking help as a carepartner is not always an easy task. However, it is extremely important to take care of yourself to help those you love. If possible don't wait until you become overwhelmed as 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.

Resources that can help

Adult day care

Adult day care centers can give relief to the carepartner 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
- Socialization

For more information about adult day care centers across the U.S., please contact the National Adult Day Services Association (NADSA): www.nadsa.org or 877-745-1440.

Respite care

What is respite care?

Respite care is short-term care given to a hospice patient by another carepartner, so that a family member or friend who is the patient's carepartner can rest or take time off. This type of care was created to allow carepartners time away from administering care, with the goal to help the carepartners 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 nonprescribed treatments, but it does cover a certain amount of the respite care stay. 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 carepartner 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 agencies in your area. 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 carepartners 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 agencies will have a list of approved caregivers who provide in-home respite care. Home-based respite care programs are provided through a nursing agency; you can find these with an online search or by contacting senior services or social services agencies in your area. 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 carepartners 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 carepartner-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?

MAKING PLANS (continued)

7. What is the carepartner-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 nonmedical services. The purpose of the services is to keep people out of the hospital or nursing home and as independent as they can be for as long as possible. The services provided can be as simple as help with daily activities such as cooking, cleaning, and bathing or as complicated as medical care that requires a licensed and skilled healthcare 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 several studies conducted by AARP, 90 percent of older Americans want to stay in their own home and remain as independent as they can 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 can receive assistance and medical care in their own home.
- Research 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 kinds of services can I get in my home?

There are a 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 licensed health care providers

Please be aware that funding for these services and others are not guaranteed so be sure to understand your financial obligation for any services provided.

Who provides the services?

In most communities, there are several 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 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 psychological 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 supplemental-coverage 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 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 (800-633-4227) 24 hours, 7 days a week, including some federal holidays. TTY/TDD users can call 877-486-2048. However, the interactive phone system is available 24 hours every day of the year.

When to Hang Up the Keys

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

But when a person becomes a danger to her- or himself 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.

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 carepartners permit their loved ones to drive longer than they know they should, causing carepartner anxiety and fear of putting others at risk.
2. When it is time to "give up the keys," explain everything to your loved one and do not become critical of her or him. You are there to support the person and tell her or him you understand her or his feelings.
3. Never leave the keys out where they can be found.
4. Assure your loved one that she or he can depend on you for transport wherever and whenever she or he 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 retest 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 her or him each part of the re-examination as described above. In many instances, once the person with driving difficulties learns about the re-examination tests, she or he 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, but mainly for distance mobility. Many people believe that using a wheelchair reduces one's independence. Using this type of assistive equipment can do the opposite; it can increase one's independence. Visit an occupational therapy (OT), physical therapy (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. Look for a medical equipment supplier that is certified with the National Registry of Rehabilitation Technology Suppliers (NRRTS).

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 drugstore. 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 drugstore 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 her or his 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"), 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 categories 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, she or he 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 comprises:

- A consumer and maybe someone from her or his 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 OT or PT. These members should be registered in their profession or licensed by your state government. You can find out more about registration and licensing 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 she or he has completed additional training and passed a certifying examination from the Rehabilitation Engineering and Assistive Technology Society of North America (RESNA). (You can even use their web site at <http://www.resna.org/member-directory/individual> 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.

MAKING PLANS (continued)

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 she or he 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. 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.

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 because of having the equipment.

It is important that the items being requested are medically necessary. This might sound obvious, but the letter writer would be abusing the funding system or third-party payer if she or he 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 to avoid a costlier expense later. A letter of justification is an expert opinion about what is best for a 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 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 her or him. 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 she or he 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 yourself).

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 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 her or him. Be sure the person sits in the most neutral position possible and that her or his 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 her or him 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.

When Should Hospice Be Contacted?

Information provided by: www.medicare.gov

There are basically two separate Medicare benefit programs that may be available for people with PSP and related neurodegenerative conditions 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 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 related neurodegenerative 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 are 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 often asked is, "When is it the appropriate time to contact hospice?" People are sometimes taken aback by the most common response, which is often, simply, "today." 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 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 nondenominational pastor also can 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 to seven days a week, depending on need, and stay from one to one-and-one-half 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 the services and benefits hospice had to offer. Although there is no obligation to accept the involvement of all the different team members, I strongly encourage doing so. Each member has something different to offer that often can complement 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.

Hospice and Medicare benefits

Medicare Part A covers hospice care if you meet all 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 six months to live. 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 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.

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

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

MAKING PLANS (continued)

Estate Planning

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

There are several benefits to making an estate plan:

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

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

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

Provide for your family — An estate plan is especially important if you have minor children as it will name a guardian to care for your children and in many instances establishes a trust to help ensure their financial well-being.

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

Related to your final wishes

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

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

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

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

Provide for physical and mental incapacity

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

Steps to having an estate plan

Depending on your situation, creating an estate plan doesn’t have to be overly difficult or expensive.

Here are some practical steps to get you started:

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

MAKING PLANS (continued)

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

Frequently asked questions

Do I need to have an estate plan?

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

Do I need to see an attorney?

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

How often should I update my plan?

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

What if I have a plan, but want to change one thing?

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

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



MAKING PLANS (continued)

CurePSP Brain Tissue Donation Program

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Jacksonville, Florida

Brain tissue research gives patients and loved ones new hope.

CurePSP asks you to consider brain tissue donation when the clinical diagnosis is made. Pathologic diagnosis by direct examination of the brain tissue through the microscope is the only way to verify a clinical diagnosis.

By making this very generous donation, you improve the chances of finding a cure and treatment options for these diseases.

The final autopsy report provides information about other brain changes that may exist, including those that may have contributed to a clinical misdiagnosis. The Brain Bank will provide tissue samples to reputable scientists worldwide for investigation into the genetics and other aspects of the causes of neurodegeneration. Research is a tangible process that will provide loved ones with the answers they deserve and better serve patients currently suffering with these diseases.

Thank you for considering a brain donation to the Eloise H. Troxel Memorial Brain Bank hosted by the Mayo Clinic. CurePSP's Brain Tissue Donation Program started in 1998 and has been with the Mayo Clinic ever since. At the Mayo Clinic, Dr. Dennis Dickson oversees the operations of the brain bank. Dr. Dickson is a world-renowned scientist and pathologist who is very experienced in the field of prime of life brain diseases and diagnosing PSP, CBD, MSA, FTD, ALS, and CTE as well as many other neurodegenerative diseases.

Brain donations are an extremely valuable resource for science and for developing future therapeutic interventions for all neurodegenerative disorders including Alzheimer's and Parkinson's diseases. Researchers from all over the world benefit from brain donations to the Mayo Clinic as Dr. Dickson collaborates with many national and international scientists. By donating a brain to the Brain Bank, you create your own legacy in science that could be part of a future therapy. After the tissue collection, the next-of-kin will receive a comprehensive autopsy report that often offers closure after many years of suffering and caring for a loved one.

Once again, I would like to thank you for considering a brain donation.

Dr. Alex Klein

Vice President-Scientific Affairs

Phone: 347-294-2872

Email: klein@curepsp.org

Questions and Answers

Q. Why make your decision in advance?

A. For several reasons, CurePSP urges you to prepare all arrangements for a brain donation in advance. Your loved one can die suddenly, which is extremely stressful. A decision may have been made to donate the brain; however, without making prior arrangements, it is possible that the donation will not occur. The 24-hour window to perform the brain donation after death cannot always be met on short notice, especially on weekends and holidays.

- The patient, family members, and other loved ones should be involved in the decision. It may take time for everyone to come to an agreement. The Brain Bank Coordinator is available to answer any questions and assist in making the arrangements.
- It can be difficult to locate a pathologist to perform the tissue collection. It is most important to have someone lined up in advance to make sure this procedure is accomplished within 24 hours after death. The Brain Bank Coordinator can assist with finding a pathologist in your area.
- It is helpful to talk with family and friends, as well as the funeral home, about the planned arrangements. Planning in advance and the support of others will help during the time of grief.
- In addition to the brain donation, research efforts are aided by getting copies of your loved one's medical records showing the progression of the disease. These records will be correlated with the autopsy results; hence securing medical records in advance is a significant help to the pathologist conducting the examination. The Brain Bank Coordinator can advise and guide you through this process.

Q. Who may authorize a brain donation?

A. Legally, the patient and/or next-of-kin are the persons to sign the Autopsy & Research Consent Form (available in our brochure). If the patient's spouse is deceased, the oldest child will be considered next-of-kin. In some states, the patient or next-of-kin may sign this form prior to death. In other states, such as Texas, this is not legally binding unless signed after death.

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

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

Q. Where is the brain donation performed?

A. If death occurs in a hospital, the tissue collection will likely be performed in that facility if the procedure has been ordered by the attending physician. If death takes place in a nursing home or hospice, or at home, the body will have to be transported to the funeral home, crematorium, hospital, or medical examiner's office for the collection to take place. In that case, there may be additional costs for transport by the funeral home.

MAKING PLANS (continued)

Q. Can there be an open casket?

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

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

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

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

A. Please contact the Brain Bank Coordinator, Jessica Tranovich, at tranovich.jessica@mayo.edu or at (904) 953-2439. The Brain Bank has a list of pathologists in the U.S. and Canada and is available to help you locate a professional in your area. You can also ask a funeral director or the patient's neurologists.

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

A. While clinical diagnosis has been greatly advanced, there is no way to confirm a diagnosis for most of these diseases other than by examining brain tissue. That is why your brain donation provides invaluable material for developing less invasive diagnostic tests in the future, such as blood tests or brain scans that can be carried out during the lifetime of a patient.

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

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

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

A. Yes, the Mayo Clinic Brain Bank actively searches for healthy brains that serve as valuable control brains in research studies. It is very important to compare pathological changes in diseased brains with healthy brains; this helps the scientists to better understand disease processes and consequently to develop novel therapeutic strategies. Please contact the Mayo Clinic Brain Bank for more information on healthy brain donation.

Please note that CurePSP cannot provide financial assistance for the donation of healthy brains. If you have any questions, please do not hesitate to contact Dr. Alex Klein, Vice President-Scientific Affairs at CurePSP, at klein@curepsp.org or (347) 294-2872.

After your decision to donate has been made

Important paperwork is required to perform a legal tissue collection. Please follow the guidelines below and complete all four forms. You can download the brochure containing the forms at <http://www.psp.org/ineedsupport/braindonation/>. You may contact Joanna Teters at teters@curepsp.org or call 347-294-2871 and we will send you a printed copy of the forms.

STEP 1: Please contact the Brain Bank Coordinator, Jessica Tranovich. She will help you understand the next steps and procedures.

Phone: (904) 953-2439, Monday-Friday, 7 a.m. to 3:30 p.m. (U.S. Eastern time).

Email: jessica.tranovich@mayo.edu

STEP 2: Enroll in the Brain Bank by completing the Brain Bank Questionnaire (in the above-mentioned brochure) and mailing, faxing, or emailing it to the Brain Bank ahead of time. It is important to have this information on record once the brain arrives at the Mayo Clinic. It also helps to facilitate the process of the autopsy.

STEP 3: The Medical Release Form authorizes physicians who diagnosed and/or treated the patient's neurological disease to send copies of their clinical notes to the Mayo Clinic. Only the patient or the next-of-kin can authorize the release of these records, which are important to the Mayo Clinic's researchers. Please send copies of this form to all physicians and neurologists (1) who are listed on the Autopsy & Research Consent Form, (2) who have treated the patient for a neurodegenerative disease, and (3) whose clinical records could provide assistance to the researchers at the Brain Bank.

STEP 4: Complete the Autopsy Information Form. Make sure to have it placed in the patient's chart or medical files. The pathologist will sign and send the form along with brain tissue to the Mayo Clinic Brain Bank.

STEP 5: Complete the Autopsy & Research Consent Form, signed by the patient and/or next-of-kin. Make sure to have it placed in the patient's chart or medical files. This is the actual consent for donation of a postmortem brain. This form can be signed only by the following individuals in this order: the patient, spouse, oldest adult child, parent, adult sibling, guardian, or power-of-attorney. The Autopsy & Research Consent Form, with original signature(s), must accompany the deceased along with the Autopsy Information Form for the tissue collection to take place. Without a fully signed Autopsy & Research Consent Form, no brain donation is possible.

STEP 6: At the time of death, all family members and healthcare professionals need to know of the patient's wish to donate his or her brain. Please make sure that you have the pathologist's contact details available, so that she or he can be contacted immediately.

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

If the patient dies at home, in a nursing home, or with hospice, the funeral home or crematorium will be involved in arrangements for the tissue donation. At times, the procedure can be performed at the funeral home or crematorium. In other cases, it may be necessary to transport the body to a hospital or medical examiner's office for the procedure. Please note that extra costs might occur in case transportation of the body is necessary.

MAKING PLANS (continued)

If the patient dies in the hospital, be sure that the physician has placed an order in the patient's chart to have the tissue collected and sent to the Mayo Clinic Brain Bank.

Please note that it is required to have a pathologist or diener (a pathologist's assistant) in place to collect the tissue. Please remember that the brain tissue can only be harvested within 24 hours after death.

Locating a pathologist

Locating a pathologist can be a difficult task. Please contact Jessica Tranovich at the Brain Bank for help. The Brain Bank has a list of autopsy resources around the country and is available to help you locate a professional in your area. Please note, CurePSP does not provide a pathologist finder service.

The Brain Bank Coordinator is available to answer any questions you or the pathologist may have. The Coordinator will also work directly with the pathologist to ensure that the tissue arrives at the Mayo Clinic in a timely and safe manner.

If you have any questions with regard to the process of a brain donation, please call or email Jessica Tranovich, Brain Bank Coordinator at the Mayo Clinic.

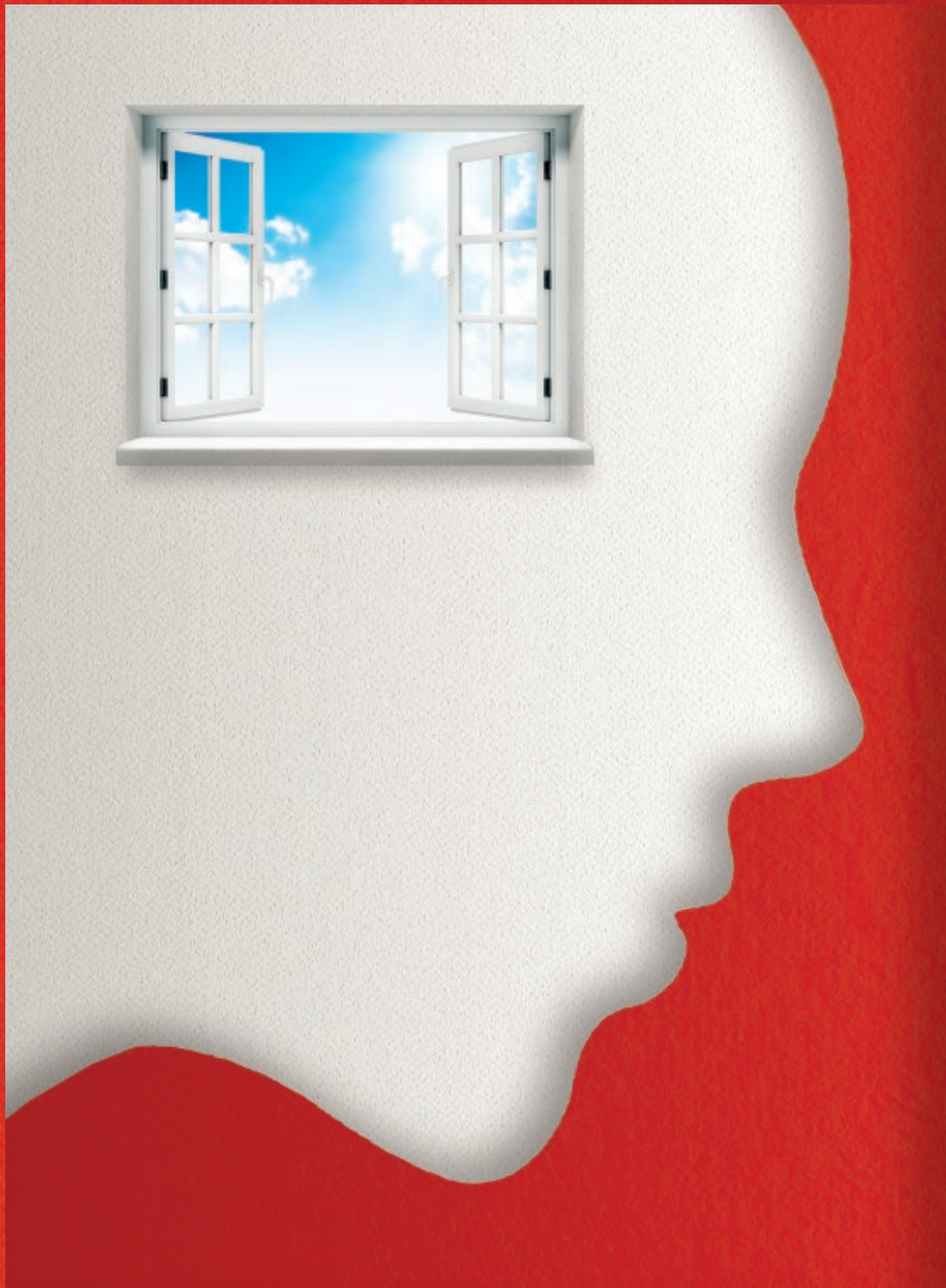
Cost of brain tissue donation

The cost of a brain donation (i.e., the cost for the pathologist and – if necessary – for transportation) is the responsibility of the family. The Brain Bank covers all other charges, including shipping the brain to the Mayo Clinic, as well as all costs associated with performing the autopsy and any research projects using the tissue.

At CurePSP, we recognize the fact that the cost of the brain donation can be prohibitive for some families. A generous donor has contributed funds to create the CurePSP Brain Tissue Donation Fund. This has allowed CurePSP to provide financial assistance of up to \$750 to families who wish to donate brain tissue but may have financial constraints. If your family needs assistance with the tissue collection costs, please call or email Office Manager, Joanna Teters, at 347-294-2871 or teters@curepsp.org.

PART SIX

Appendix



Glossary of Terms

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

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

Adult Day 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.

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

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

Ambulation: The act of walking.

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

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

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

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

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

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

Bilateral: Having or affecting two sides.

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

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

Bradykinesia: Slowness of movement.

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

Brainstem: The region of the brain consisting of the medulla oblongata, pons, and midbrain. This area of the brain form a connection 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 such as reflexes.

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

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

Case Managers: Responsible for 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.

APPENDIX (continued)

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

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

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

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

Certified Nursing 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.

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

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

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

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

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

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

Corticobasal degeneration (CBD): CBD is considered one of the "parkinsonian" disorders, or "parkinsonisms." There are about a dozen such diseases, all of which produce some degree of slowness, muscle stiffness, balance problems, and sometimes tremor. Most people with CBD have these things, but in addition they have unusual difficulty performing complex limb movements such as cutting food, buttoning,

or typing. There is often a tendency to hold part or all of a limb in a fixed posture called dystonia. There can also be very rapid, irregular, small movements of muscles called myoclonus. The unusual feature about CBD is that it is almost always very asymmetric, with one side affected much earlier and worse than the other. In half of people with CBD it's the left and in half, it's the right. The side affected has no relationship to the person's handedness.

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

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

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

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

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

Dopamine: A chemical that is known as a neurotransmitter. Neurotransmitters help relay messages from one nerve cell to another. Dopamine is especially important in relaying messages about movement. The most prominent brain area hosting most dopamine neurons is the substantia nigra, which is affected in Parkinson's diseases, and PSP, causing decreased levels of dopamine.

Dopamine Agonist: A drug that acts like dopamine on dopamine receptors to mimic its actions. Such medications stimulate dopamine receptors and produce dopamine-like effects. Often used in PD patients.

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

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

APPENDIX (continued)

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

Gait 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.

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

Gastrostomy Tube (G-Tube): A plastic tube inserted into the stomach through a surgical incision in the abdomen. A gastrostomy tube is used to deliver liquefied food to the digestive system when swallowing becomes dangerous or difficult. The most common G-Tube procedure is the percutaneous (through the skin) endoscopic gastrostomy (PEG).

Gene: The smallest units of heredity. The information from all the genes, taken together, makes up the blueprint or plan for the human body and its functions (genome). A gene is a short segment of DNA, which is interpreted by the body as a plan or template for building a specific protein.

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

Gland: A structure or organ that makes a substance, such as a hormone or chemical, that is used elsewhere in the body. Some of these hormones and chemicals are insulin, bile, growth hormone, estrogen, and testosterone.

Gray Matter: Nerve tissue that primarily consists of nerve cell bodies, dendrites, and unmyelinated axons, thus having a gray appearance. In contrast, white matter predominantly contains myelinated nerve fibers.

APPENDIX (continued)

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.

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 (nonmedical) services provide assistance with housekeeping, personal care, and Activities of Daily Living (ADLs), all of which help the older adult to remain safely at home. These services may also include light housecleaning, meal preparation, doing laundry, grocery shopping, and running errands. Additional nonmedical homecare services include feeding, bathing, bowel and bladder care, and dressing.

Homeopathy: A system of medicine that is based on the Law of Similars, a belief in which substances that cause healthy people to exhibit symptoms can, conversely, be used to restore the body to health. Taken from the NIH web site, please read the following key points carefully. More information and a general overview can be found at <https://nccih.nih.gov/health/homeopathy>.

Key Points: (a) There is little evidence to support homeopathy as an effective treatment for any specific condition. (b) Although people sometimes assume that all homeopathic remedies are highly diluted and therefore unlikely to cause harm, some products labeled as homeopathic can contain substantial amounts of active ingredients and therefore could cause side effects and drug interactions. (c) Homeopathic remedies are regulated by the U.S. Food and Drug Administration (FDA). However, the FDA does not evaluate the remedies for safety or effectiveness. (d) Several key concepts of homeopathy are inconsistent with fundamental concepts of chemistry and physics. There are significant challenges in carrying out rigorous clinical research on homeopathic remedies. (e) Tell all your health care providers about any complementary health practices you use. Give them a full picture of all you do to manage your health. This will help ensure coordinated and safe care.

Homeostasis: Homeostasis, from the Greek words for “same” and “steady,” refers to any process that living things use to actively maintain fairly stable conditions necessary for survival.

Hospice: A special way of caring for a person with a terminal illness. A person is eligible to receive hospice care when he or she has a life expectancy of six months or less and 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.

Huntington’s Disease (HD): A hereditary, progressive, neurodegenerative disorder primarily characterized by the development of emotional, behavioral, and psychiatric abnormalities; gradual deterioration of thought processing and acquired intellectual abilities (dementia); and movement abnormalities, including involuntary, rapid, irregular jerky movements (chorea) of the face, arms, legs, or trunk. HD may be inherited as an autosomal dominant trait (inherited only from one parent) or, less commonly, appear to occur randomly for unknown reasons (sporadically). The disorder results from

abnormally long sequences of information within a gene. Progressive nervous system dysfunction associated with HD results from loss of neurons in certain areas of the brain, including the basal ganglia and cerebral cortex.

Hypokinetic: Diminished movement and decreased motor function. Some movement disorders are hypokinetic, such as Parkinson's disease, PSP, or MSA.

Hyposmia: A decreased sense of smell.

Idiopathic: A disorder or condition of spontaneous origin; self-originated or of unknown cause. The term is derived from the prefix "idio-" meaning one's own and "pathos" indicating disease.

Inflammation: This is the body's first response to injury or irritation. The classic signs of inflammation are pain, heat, redness, swelling, activated immune response, and loss of function.

Inhibition: The restraint, suppression, or arrest of a process or the action of a cell or organ; the prevention or slowing of the rate of a chemical or an organic reaction. The term "reciprocal inhibition" refers to the restraint or "checking" of one group of muscles upon stimulation (excitation) and contraction of their opposing (antagonist) muscles.

Inhibitor: A substance that blocks, restricts, or interferes with a chemical reaction or other biologic activity.

Insidious: Of subtle, gradual, or imperceptible development; referring to the development of symptoms that may not be recognized by an affected individual until the disorder in question is established.

Jejunostomy Tube: Similar to a gastrostomy tube, although this tube is longer and is inserted through the abdominal wall into the jejunum, the middle section of the small intestine.

Lateral: Sideways; of, on, from, or toward the side.

Levodopa: A drug used to treat Parkinson's disease. It is also called L-dopa and, in the United States, is sold as Sinemet. Levodopa is converted by the body to dopamine, a neurotransmitter that is reduced in PD and PSP brains. However, levodopa does not work well in PSP patients.

Lewy Body Disease: Also called diffuse Lewy body disease or Lewy body dementia. Lewy body disease is a common cause of dementia, accounting for approximately 15%-20% of all cases. The age of onset is typically in the late 50s through the 70s. Lewy body disease is characterized by more daily fluctuations in symptoms than Alzheimer's disease, as well as more prominent psychosis. Patients are prone to have adverse reactions to antipsychotics. Patients also have parkinsonian features early in the disease, including slowed movements and rigidity, though usually without tremor. In contrast to PSP and CBD, Lewy bodies are not made of accumulated and misfolded tau protein, but consist of misfolded alpha-synuclein. In Lewy body disease, the Lewy bodies that cause neurodegeneration are most prominently found in the cortex, or surface of the brain, versus in the midbrain with Parkinson's disease.

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

Magnetic Resonance Imaging (MRI): A diagnostic scanning technique during which radio waves and an electromagnetic field are used to help create detailed, cross-sectional images of specific organs and tissues. MRI images can help to diagnose brain atrophy and other pathological changes seen for example in PSP, CBD, and MSA.

APPENDIX (continued)

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, provides some protection against the medically necessary costs of hospital and related healthcare. Medicare A is financed through Social Security payroll tax deductions. It covers institutional care in hospitals and skilled nursing facilities and care given by home health agencies and hospices. No premium is required from 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.

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

National Institutes of Health (NIH): The NIH is one of the world’s foremost medical research centers and the federal focal point for medical research in the United States. The NIH, comprising 27 separate Institutes and Centers, is one of eight health agencies of the Public Health Service that, in turn, is part of the U.S. Department of Health and Human Services.

Necrosis: Cell death.

Nervous System: The nervous system of the human body is divided into two interconnected systems: the central nervous system, which is made up of the brain and spinal cord; and the peripheral nervous system. The peripheral nervous system is further divided into the somatic nervous system (made up of peripheral nerve fibers that send sensory information to the central nervous system and motor nerve fibers that project to skeletal muscle) and the autonomic nervous system regulating the functions of our internal organs such as the heart, stomach, and intestines, but also muscles in the skin (around hair follicles and smooth muscles around blood vessels, in the eye (the iris), in the stomach, intestines and bladder, and of the heart (cardiac muscle).

Neurodegenerative: Marked by or pertaining to neurologic degeneration; deterioration of the structure or function of tissue within the nervous system.

Neuroimaging: The production of detail, contrast, and clearness in images of the brain and spinal cord (central nervous system) using computed tomography (CT) scanning, magnetic resonance imaging (MRI), positron emission tomography (PET) scanning, or other imaging techniques to assist in diagnosis, treatment decisions, or research.

Neuron: An individual nerve cell.

Neuropsychologist: Usually a state-licensed psychologist with expertise in evaluating and treating people who suffer from a brain disorder or organic problem. Neuropsychologists may play a pivotal role in the diagnosis and treatment of mental and emotional problems caused by brain dysfunction.

Neurotoxin: A substance that interferes with the functioning of nerve cells (neurons), preventing them from communicating with each other.

Neurotransmitter: A specialized substance (such as norepinephrine or acetylcholine) that transfers nerve impulses across spaces between nerve cells (synapses). Neurotransmitters are naturally produced chemicals by which nerve cells communicate. Neurotoxins can also lead to neurodegeneration, causing cell death and brain atrophy.

Nucleus: The part of the cell that contains the genetic material (the chromosomes); it is surrounded by a membrane called the nuclear envelope.

Nurses: People who care for individuals of all ages, families, groups, and communities, sick or well in all settings. Nursing includes the promotion of health, prevention of illness, and the care of ill, disabled, and dying people. Other important roles nurses assume include patient advocacy, promoting safe environments, conducting research, 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 states to provide skilled, 24-hour nursing and rehabilitation care. Generally, older adults who are bedridden, cannot feed themselves, or who require skilled nursing assistance are appropriate for this level of care.

Occupational Therapists (OTs): Primarily focused on the “occupation” – what people do each day. This means helping individuals with PSP, CBD, and MSA find new ways of doing the activities that mean the most to them. OTs help both patients and carepartners to problem solve and create new ways to make routine activities easier and safer.

APPENDIX (continued)

Orthostatic Hypotension: A sudden decrease in blood pressure that occurs when the affected individual sits up or stands. In some cases, it may occur as a side effect of certain medications.

Oxidative Stress: A process in which substances called free radicals (see above) build up in the cells as the cells convert nutrients into energy. The free radicals can damage different parts of the cells causing stress to the cell and impairing its functioning. The free radicals can be counteracted by antioxidants (for example, Coenzyme Q10 acts as a scavenger of free radicals).

Palliative Care: Specialized medical care for people with serious illness. This type of care is focused on providing relief from the symptoms and stress of a serious illness. The goal is to improve quality of life for both the patient and the family. Palliative care is provided by a specially trained team of doctors, nurses, and other specialists who work together with a patient's other doctors to provide an extra layer of support. It is appropriate at any age and at any stage in a serious illness. This type of care treats pain, depression, shortness of breath, fatigue, constipation, nausea, loss of appetite, difficulty sleeping, anxiety, and any other symptoms that may be causing distress.

Parkinson's Disease (PD): A slowly progressive neurodegenerative brain disorder characterized by slowness or poverty of movement (bradykinesia), rigidity, postural instability, and tremor primarily while at rest. Additional characteristic findings include a shuffling, unbalanced manner of walking; forward bending or flexion of the trunk; a fixed or "mask-like" facial expression; weakness of the voice; abnormally small, cramped handwriting (micrographia); depression; or other symptoms and findings. Such abnormalities are thought to result from progressive loss of nerve cells within a certain region of the substantia nigra of the brain and the associated depletion of the neurotransmitter dopamine.

Pathogenesis: The origination and development of a disease.

Peripheral Nervous System: The peripheral nervous system is that portion of the nervous system outside of the brain and spinal cord (see central nervous system).

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

Physical Therapists (PTs): 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.

Placebo: The sheer anticipation of new drug, surgery, or procedure that could potentially end the suffering of a patient can provide improvement of symptoms for a period of time. Hence in a clinical trial setting, a substance or procedure that appears to be identical to the treatment under study but that has no effects on the test subject, will be given to one group of study participants. At the end of a study when all data are analyzed, the placebo group are compared to the group who received the new active drug or procedure (called the treatment group). If only the treatment group improved, the drug or procedure had a unique positive effect on patients. If both groups improve to the same degree or do not change, the placebo group proved that the new drug or procedure is not effective.

Positron Emission Tomography (PET): An advanced, computerized imaging technique that uses harmless radioactive tracer substances (e.g., modified glucose) to demonstrate chemical and metabolic activities in the brain. PET images then display areas with normal, increased, or reduced activity that can be interpreted by radiologists to identify disease or normal function.

Postural Instability: Unsteadiness of gait or standing.

Progressive Supranuclear Palsy (PSP): PSP is a neurodegenerative disease (occasionally referred to as Steele-Richardson-Olszewski syndrome, after the three physicians who first described the disease in 1963). The most common first symptom, which occurs, on average, when a person is in her or his 60s, is loss of balance while walking. This may take the form of unexplained falls or of a stiffness and awkwardness in a person's gait that can resemble Parkinson's disease. Sometimes the falls are described by patients as attacks of dizziness. This often prompts the doctor to suspect an inner ear problem or hardening of the arteries supplying the brain. The second most common form of PSP is called PSP-parkinsonism. Its early stages more closely resemble those of Parkinson's disease, with less emphasis on balance problems and behavior changes and more on tremor. These typically have a better early response to antiparkinson drugs than is typical for PSP. PSP-parkinsonism comprises about a quarter of all PSP cases.

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

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

Psychiatrist: A board-certified psychiatrist has at least three years of specialty training after receiving an M.D. degree, and is a medical doctor who specializes in mental disorders. A psychiatrist is uniquely qualified to assess, diagnose, and treat mental and physical conditions.

Psychologist: A doctoral-level specialist in psychology, licensed by the state to practice professional psychology (e.g., assess and treat), teach psychology as a scholarly discipline, or conduct research. Psychologists are usually qualified to provide psychotherapy, administer psychological and educational tests, and diagnose and treat mental and psychiatric disorders.

Pulmonary: Referring to the lungs.

Range of Motion (ROM): The extent of a joint's free movement. The normal ROM of the elbow, for instance, carries the forearm through a half-circle. Passive ROM is tested while the limb is relaxed. Active ROM is movement controlled by the patient.

Registered Nurse (RN): A registered nurse (RN) acts upon the order of the physician, providing and directing nursing care services. Additionally, an RN utilizes agency and community resources to fulfill individual patient goals developed in coordination with patients and family.

Residential Care Facilities for the Elderly (RCFE), also known as Board and Care: Care in a setting that resembles a person's home. These are small facilities of about six to eight residents that care for older adults who need assistance with Activities of Daily Living (ADLs). Board and Care facilities are licensed by the state's department of social services.

APPENDIX (continued)

Respite Care: The temporary alleviation of the caregiver's responsibility and involvement with the patient. It can be obtained through adult day care or in-home agencies.

Retirement Home (Facility): Special complexes of apartments or private homes that provide a supportive environment for seniors, but allow residents to remain somewhat independent. Residents have their own living space (apartment or room) and services provided vary greatly. Examples of services are meals in a central location, laundry facilities, housekeeping, bathing assistance, dressing, and assistance taking medication.

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

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

Sialorrhea: Excess production of saliva, or increased retention of saliva in the mouth resulting from difficulty swallowing.

Side Effect: An effect of a drug that is not the main or intended effect. Side effects may be of no concern, or they may be bothersome or even dangerous, in which case they may limit the upper dose a patient can tolerate. Side effects are also called adverse effects and are closely monitored for through vigorous clinical testing.

Social Worker: A professional trained to work with individuals, couples and families around a wide variety of problems including those arising from medical and mental health concerns. Social workers are also knowledgeable about community resources and how to access them.

Striatum: See also basal ganglia for more information. The striatum is an area of the brain that controls movement and balance. It is connected to and receives signals from the substantia nigra.

Substantia Nigra: See also basal ganglia for more information. The substantia nigra is an area in the brain that controls movements and is the origin of most nerve cells that use dopamine as messenger to communicate with other nerve cells. Degeneration of cells in this region may lead to a neurologic movement disorder such as Parkinson's disease or PSP.

Tauopathy: A class of neurodegenerative disorders that have the pathologic aggregation of the tau protein as the common denominator. Examples for tauopathies are PSP, CBD, CTE, and Alzheimer's disease.

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

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

Resource Directory

A

AARP (American Association of Retired Persons) • 800-424-3410 • www.aarp.org
American Massage Therapy Association • 847-864-0123 • www.amtamassage.org
American Occupational Therapy Association • 301-652-2682 • www.aota.org
American Parkinson Disease Association • 800-223-2732 • www.apdaparkinson.org
American Speech-Language Hearing Association • 800-638-8255 • www.asha.org

B

Benign Essential Blepharospasm Research Foundation • 409-832-0788 • www.blepharospasm.org
Biotene (dry mouth) • 800-922-5856 • www.laclede.com
Botox • 800-44-BOTOX • www.botox.com

C

Centers for Medicare/Medicaid Services Regional Administrator • 800-633-4227 • www.cms.gov

D

E

Eldercare Locator • 800-677-1116 • www.eldercare.gov
Eldercare Web • www.elderweb.com

F

Family Caregiver Alliance • 800-445-8106 • www.caregiver.org

G

H

Healthcraft Products • 613-822-1885 • www.healthcraftproducts.com

I

International Society for Augmentative and Alternate Communication •
905-850-6848 • www.isaac-online.org

J

K

L

M

Meals-on-Wheels Association of America • 703-548-5558 • www.mowaa.org
Medicare, 800-633-4227 • www.medicare.gov
Movement Disorder Society • 414-276-2145 • www.movementdisorders.org

APPENDIX (continued)

N

National Academy of Elder Law Attorneys • www.naela.org

National Association of Professional Geriatric Care Managers • 520-881-8008 • www.caremanager.org

National Center for Complementary and Alternative Medicine • 888-644-6226 • www.nccam.nih.gov

National Council on Aging • 202-479-1200 • www.ncoa.org

National Family Caregivers Association • 800-896-3650 • www.nfcacares.org

National Hospice and Palliative Care Organization • 703-837-1500 • www.nhpco.org

National Institutes of Health • 301-496-4000 • www.nih.gov

National Institute of Neurological Disorders and Stroke (NINDS) • 800-352-9424 • www.ninds.nih.gov

National Long-Term Care Ombudsman Resource Center • 202-332-2275 • www.ltcombudsman.org

National Organization of Rare Diseases (NORD) • 203-744-0100 • www.rarediseases.org

Neuro-Optometric Rehabilitation Association • www.nora.cc

O

P

Q

R

S

Social Security Administration • 800-772-1213 • www.ssa.gov

T

U

V

Veteran's Helpline • 800-827-1000 • www.va.gov

W

Well-Spouse Foundation • 800-838-0879 • www.wellspouse.org

X

Y

Z

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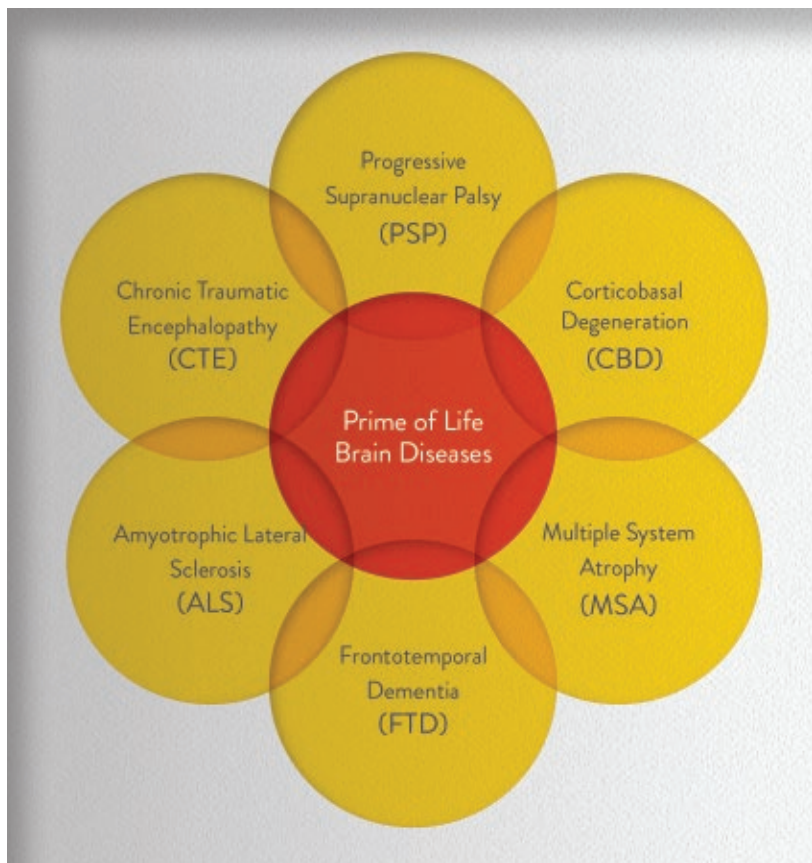
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