THE SOCIETY FOR
PSP
Progressive Supranuclear Palsy

Woodholme
Medical Building
Suite 5l5
1838 Greene Tree Rd.
Baltimore, MD 21208
1 (800) 457-4777
1 (410) 486-3330
FAX:
1 (410) 486-4283
email:
SPSP@erols.com
website:
www.psp.org

INSIDE

"The Society For PSP On The Move"

"Dudley Moore and PSP"

The PSP ADVOCATE

Fourth Quarter 2000, Vol. 11, No. 4

The Society for PSP on the Move Autumn, 2000

GOAL: To establish a strong national presence and increase education of PSP to persons with PSP, their families, carepartners, physicians, and allied professionals

"Visions and Opportunities"

STRATEGIC PLAN 2000-2003

The Society for Progressive Supranuclear Palsy

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**In 1963, Dr. J. C. Steele, together with Dr. J. C. Richardson and Dr. J. Olszewski, identified PSP as a distinct neurological disorder.*

The Society for Progressive Supranuclear Palsy, Inc. (SPSP) is a nonprofit 501-3(C) organization that exists to promote and fund research into finding the cause and cure for PSP. PSP is a rare, neurological disorder related to Parkinson's disease. SPSP provides information, education, support and advocacy to persons with PSP, their families, and caregivers. SPSP educates physicians and allied health professionals on PSP and how to improve patient care. The Society, Director, and staff are under the administration of the Johns Hopkins School of Medicine, Department of Neurology.

Phone 1-410-486-3330
Toll Free: 1 (800) 457-4777, FAX 1 (410) 486-4283
Ellen Pam Katz, Executive Director
SPSP, Inc.
Woodholme Medical Building
Suite 5l5
1838 Greene Tree Road
Baltimore, MD 21208
email-epkatz@erols.com
SPSP@erols.com
website-www.psp.org
List Serve: requests@hydra.welch.jhu.edu
Subscribe psp your name

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EDITOR
Nancy Ogiba Brittingham
NancyB501@cs.com
(757) 838-0777 • FAX (757) 838-6086
(In memory of Henry and Jane Ogiba)
Assistant to the Editor: Debra Thompson

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Director's Doings

I have been fortunate to drink from the tree of knowledge. I feel like a vessel swelled with new-found wisdom. However, I am frustrated that I can only serve small sips to others and that I cannot act immediately on all I have learned.

These feelings result from several events I recently attended on behalf of the Society. In mid-October, I participated in the annual conference and board meeting for NORD, the National Organization for Rare Disorders in St. Louis. As a board member for NORD, I was exposed to multidimensional issues facing persons with rare diseases. When I saw the breadth of the rare disease community and became involved in the decision-making process that affects millions of people, I learned that PSP cannot stand alone in this community. As persons affected by rare diseases, we have to strengthen the Office of Rare Disorders at the NIH, be advocates for many legislation initiatives affecting us and become knowledgeable on health issues that often seem remote and disjointed from our everyday life.

Other activities involved the St. Louis and Toronto symposia. How appreciative PSP families were that the Society for PSP was present to meet them! I brought a message of hope, noting how our research program had grown. While I increased my knowledge base on PSP, the most valuable experience was watching PSP families interact with each other.

However, the most important thing I have learned from my recent travels is that we are on the right path. We are altering the destiny of how PSP will be unraveled. We are expanding offerings of symposia and other educational services to families and health professionals. We are raising funds to bolster research. We are on the right path... I KNOW IT!!

Ellen Pam Katz

Denny Neagle Strikes Out PSP

Thank you, Denny and Jennifer Neagle for your gift of $14,800 representing 148 strikeouts Denny scored during the Y2000 professional baseball season.

Congratulations on Denny's new position as pitcher for the Colorado Rockies. We look forward to even more strikeouts next season!

The Power of Music

Dudley Moore's new CD, "Live From an Aircraft Hangar," was recently released after nine months of work, and has already attracted a lot of attention. Moore hopes to drawn on the power of music and the Internet to find a cure for progressive supranuclear palsy and to bring live music to people confined in institutions. "After nine months of work on the CD," he said, "labor pains and all, I have given birth to a big, round, shiny baby that I hope everyone in the world will want to own."

"I am asking my beloved public to join with me in supporting two important causes—the Dudley Moore Research Fund for PSP and Music For All Seasons," Moore said of his first major production since announcing his battle with the disease. The CD is available now through www.Amazon.com and on a new website that Moore helped to create, www.DudleyCD.com.

The new CD was produced by Martine Avenue Productions, Inc., Moore's production company. "We are very excited about releasing this CD. It was one of my all-time favorite concerts," he said.

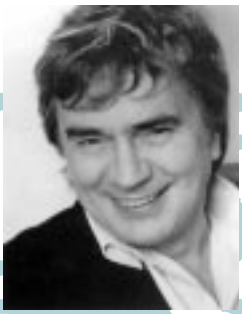
"I've had letters from thousands of people, encouraging me to keep going, and asking me for a new CD. People have been incredibly supportive. I felt it was important to let everyone hear my favorite concert. Join me online and let's see if we can top the charts."

The CD features Dudley Moore as pianist, composer and comedian, with the Dudley Moore Trio, and the BBC Concert Orchestra, in several comedy tracks and a couple of parodies featuring Moore as vocalist. Much of the music was written by Moore for films and for the stage, including three jazz tracks and his orchestral scores from the original "Bedazzled" and "Six Weeks." The 71-minute, 24-track CD was drawn from Moore's highly acclaimed 1992 concert at the Royal Albert Hall in London, a 5,000 seat hall he affectionately calls an "Aircraft Hangar."

According to Moore, the last year since announcing his condition has been a very difficult time of learning to deal with the effects of PSP, while finding a way to continue his involvement with music and reaching out to his audience. He continues with a regular program of physical, occupational and speech therapies designed to slow the progression of his condition.

For additional information, call Martine Avenue Productions, Inc. at 908-754-4190.

A Man For All Seasons



A Star-Studded Salute to Dudley Moore

Presented by Music For All Seasons

Monday, April 16, 2001, 7:00 PM

Carnegie Hall, New York

Barbara Walters will introduce this exciting evening
of music, comedy, and film
featuring
Julie Andrews Bo Derek Mary Tyler Moore
Jane Alexander Amy Irving Jill Eikenberry Tony Randall
Eric Idle Brooke Shields Angie Dickinson
and many other co-stars, friends, and associates…

Followed by a black-tie Dinner with the Stars
where his colleagues will pay informal tributes to Dudley

Performance and Dinner with the Stars $500 - $1000
Performance tickets only $25 - $95
Event Chairman: Michael Caine

Don't miss the event of the year!
CALL FOR TICKETS NOW ☑ 908 322 6300
Proceeds will benefit the work of Music For All Seasons and
the Dudley Moore Research Fund for Progressive Supranuclear Palsy

In PSP, what causes the brain cells to degenerate in the first place?

Lawrence Golbe, M.D.
Chairman, The Society for PSP Medical Advisory Board

No one knows - yet.

We can say that it seems not to be infectious. Brain tissue from patients with PSP has been shown to be free of prions, the type of infectious agent that cause "mad cow disease." Attempts to transmit PSP to chimpanzees by injecting the brains with brain material from humans with PSP have failed, fortunately. This is good evidence that PSP is not transmitted from one person to another or contracted from animal contact.

An important factor will probably turn out to be some yet-to-be-discovered and probably naturally occurring chemical in the food, air, or water that slowly damages certain vulnerable areas of the brain. A clue may lurk on the island of Guadeloupe in the Caribbean, where there is an unusually high prevalence of PSP and similar "atypical parkinsonisms." A survey there found that patients with PSP were disproportionately likely to have consumed two native tropical fruits called sweetsop and sour-sop. In fact, these fruits contain small amounts of certain chemicals known to be toxic to brain cells. Research supported by the Society for PSP is now in progress to give those chemicals to laboratory animals in an attempt to produce PSP. This would provide valuable insight into the human disease. Work is under way to attempt to identify foods in the diet in other countries such as the US that might contain similar toxins.

On the Pacific island of Guam, a common neurological disease occurring only there and on a few neighboring islands has some of the characteristics of PSP, Alzheimer's disease, Parkinson's disease and amyotrophic lateral sclerosis. We do not have the well-documented dietary clue there that we have for the Guadeloupe illness.

Another theory presently under investigation is that there may be some hereditary component to the cause. The medical literature has reported six families with autopsy-proven PSP in more than one member – two from the U.S. and one each from Canada, England, Japan and Spain. The last has seven affected members and a search for its culprit gene is under way. (The easiest way to find a disease gene is to examine the DNA of many members of a large family with and without the disease.) If that gene can be found, people with non-hereditary PSP can be tested for it to see whether the same defect in a non-inherited form may be the cause of PSP. Despite the promise of this line of research, the fact remains that PSP almost never occurs twice in one family. This stands in stark contrast to the situation in Parkinson's or Alzheimer's disease.

Recent research has found that the gene coding for a protein called "tau" is somehow abnormal in people with PSP. The tau protein is a normal component of brain cells, but is found in abnormal clumps as part of the neurofibrillary tangles mentioned above. The gene that codes for tau protein is located on chromosome 17. A particular version of the tau gene called "H1" is present in nearly every patient with PSP but in fewer than half of persons without PSP. A mutation in that gene, then, may be necessary but not sufficient to cause the disease. Perhaps developing PSP requires both the H1 form of tau and an environmentally acquired triggering factor such as unusual consumption of a certain food or exposure to a chemical.

Update from Guadeloupe

By Dominique Caparros-Lefebvre, M.D. with
Editorial Assistance of Lawrence Golbe, M.D.

The first publication of the unusual cluster of PSP and other atypical parkinsonian disorders on the Caribbean island of Guadeloupe appeared in early 1999. That paper reported that a majority of patients with parkinsonian disorders on Guadeloupe had PSP or similar "atypical parkinsonisms," a reversal of the ratio seen elsewhere. It also found that many of the patients with PSP or atypical parkinsonism on Guadeloupe had consumed tropical fruits that harbor certain chemicals known to be toxic to brain cells.

1. Clinical study: We have now expanded our group of parkinsonian patients to 180. Of these, only 40 (22%) have Parkinson's disease and the remaining 140 have atypical parkinsonism. Of these, 49 (35%) appear to have PSP. Another 75 (54%) have a condition that resembles PSP except for the absence of eye movement abnormalities - a sign ordinarily considered necessary for that diagnosis. This does not mean that they do not have PSP, as many such patients do not develop eye movement difficulties until many years into the illness.
2. Epidemiologic study: We plan to repeat and expand our questionnaire study of dietary and other risk factors related to parkinsonism on Guadeloupe. We will invite the participation of all of the general practitioners on the island, who would refer to us their patients with parkinsonism. We will examine and videotape the patients and administer a new questionnaire assessing dietary habits, including consumption of herbal tea and local tropical fruits.
3. Neuropathologic study: So far, we have had the opportunity to examine the autopsied brains of three Guadeloupean patients with the outward appearance of PSP during life. All three showed the microscopical changes typical of PSP, including neurofibrillary tangles containing tau protein. Biochemical analysis of the tau showed that it was identical to the tau that accumulates in classical PSP.
4. Toxicity study: Dr. A. Lannuzel of the Department of Neurology in our hospital has performed laboratory experiments with a mixture of chemicals extracted from sweetsop, one of the fruits frequently consumed by the PSP patients on Guadeloupe. He found that the mixture is toxic to rat brain cells growing in a dish. One of those chemicals, called corexime, is one of the most potent brain toxins known. It is produced by several plants, including some used as herbal remedies such as boldo, fumeterre, hydrastis and chelidonium. Hydrastis, which is also called goldenseal, is sold for the treatment of colds.
5. Relationship to Guamanian parkinsonism: A group of natives of the island of Guam in the Pacific have been known since the 1930's to have high rates of a parkinsonian disorder called the parkinsonia-amyotrophic lateral sclerosis-dementia complex of Guam. I had the opportunity in April 2000 to visit Guam, where I was able to confirm that those patients closely resembled those on Guadeloupe. In June, Dr. John C. Steele, who was one of the group of physicians who first brought PSP to the world's attention in 1963 and who now works on Guam, visited Guadeloupe. He was equally impressed by the similarity between the two diseases. In fact, autopsies show a deeper similarity between the two. Further study of both local disorders, especially regarding dietary risk factors, may shed light on the cause of PSP elsewhere in the world.

Brain Donation and How it Adds to What We Understand About the Cause of PSP

Shamaila Waseem, M.D. and Katrina Gwinn-Hardy, M.D.
Mayo Clinic Jacksonville, FL, USA

Some of us have heard about brain donation after death in PSP, but have not understood completely why this is done. Some may have wondered whether or not brain donation after death of a person with progressive supranuclear palsy would be worthwhile. Below, some of the issues involved in this are discussed.

Why consider donation of my brain for the study of PSP?

Neurological disease is the area about which we know the least despite its important consequences. PSP is no exception. Brain donation provides insight into conditions that may clinically masquerade as other diseases. For example, many of the features of PSP are similar to those of Parkinson's disease. Thus, many patients are initially given a diagnosis of Parkinson's disease before all the typical signs of PSP appear. There are many distinguishing signs; however, these may not be obvious early in the course. Many other distinguishing signs are seen upon examination of the brain after death. This allows a more sophisticated understanding of these differences during life. If the diagnosis is wrong, then, what we think we are learning about a given disease might not have anything to do with it after all. This is especially important in family studies. It makes sure that the researchers have correctly catalogued and considered the diseases they are studying. Better treatment can be developed as we continue to collect more information about these disorders. This includes clinical studies (including treatment trials), genetic and other molecular studies, imaging studies of the brain, and direct examination of the brain itself (which can only be done after death).

How can examination of a brain after death give you any more information that examining a person and their blood does when they are alive?

Examination of the brain is the most definitive way to make a diagnosis and to search for a cause. There are classic, reliable features of a given disease which are seen when the brain is examined. When someone is alive, we use terms like "Parkinson's disease," or, "PSP," but we aren't exactly sure until someone has died if the whole picture fits together: the clinical history (the medical records), the examination, and the blood tests are compared to the information obtained from the brain itself.

PSP is a "degenerative" disorder of the brain, affecting certain selected brain circuits. The term, "degeneration" implies that the selected brain cells prematurely age and die. The predominantly affected circuits in PSP are those involved in movement and balance. Why these certain brain circuits degenerate is not known. Other brain areas are spared, such as those involved with hearing and sensation. Because PSP is a disease of the brain, it is important to examine the brain directly in order to learn more about the disease. While MRI scanning and other research methods, such as PET, are useful for looking at the brain, they do not look at each area, nor do these tests look at the circuitry which is in place there, which would show the relationship between those regions. Thus, brain donation allows scientists to continue to learn about what occurs in PSP and how it differs from what occurs in normal individuals (those without any neurological disease) and how it differs from diseases which have major similarities as well as major differences (such as Parkinson's disease or lytico-bodig disease of Guam, which it closely resembles in some ways).

What does this mean for the future of patients and their families?

Although donating a brain does not help the person who donated it, it hopefully will contribute to helping a large number of other people, many of whom may even be in that person's family. As our knowledge increases, it is clear that some cases of PSP run in families. If genetic tests or other tests eventually become available, it is important to know that PSP is the actual diagnosis. If other family members might be prone to develop PSP, this information will be important for early diagnosis and treatment. A single brain donation will not answer all of these questions, but will add another essential piece of information to our solving of this puzzle.

Does donating my brain mean that I will have to be cremated? Will it be possible to have a normal funeral if the brain has been removed? Is the body taken away?

It is possible to remove the brain without disturbing a person's appearance for a funeral. The family members are free to choose any type of service that they might wish even if the brain has been collected. This is because the people who evaluate the person after death are highly trained to do this with care. The person's body may not need to be taken away from his or her hometown, because the procedure often can be done locally and sent to the researchers who are experts in a given disease.

Who evaluates the brain and what happens to the information?

The brain is evaluated by a neuropathologist (a person who is specially trained in making diagnoses from brain material). The case is then discussed with the doctors who took care of the person when he/she were alive, and with researchers who may have collected blood on a person. This information is taken into consideration to guide the further study in a given disease. The information becomes part of the person's medical chart, much in the way an X-ray or CT scan might. The information is also available to the family should they wish it. The information is shared with the family doctor. Also, donated brain material is used for world-wide research projects so that we can all share information and work together towards solving the mysteries of diseases like Parkinson's disease, tremor, and other neurological disorders.

Whom should I contact when I have questions about this matter or wish to arrange for myself or a loved one to make such a donation?

If you wish to donate your brain once you have passed away, ask your medical doctor to help arrange things far in advance, and identify contact people to help as well. Then, if you or a loved one were to pass away, things could happen smoothly and without additional difficulty to the family at a time when there are already enough emotional and personal issues at hand. The PSP Society has a Brain Bank, and the resources listed below may be very helpful to you in this regard.

Call the Society office to obtain additional informational packets and forms required for advance preparation of brain tissue donation: (800) 457-4777 or (410) 486-3330.

"Our Experience with Brain Tissue Donation"

By Mr. R.L. Mellers

We recently went through the experience of arranging a tissue donation to the PSP Brain Bank at the Mayo Clinic in Jacksonville, FL. It may be a service to others who are thinking about or who have made similar arrangements to hear our story.

My wife, Carolyn, was diagnosed with PSP about four years ago. We did not know much about the disease and it had taken several years for this diagnosis to be made. We found out about the PSP Society on the Internet and received the helpful literature that is sent out.

Our children wanted a confirmation of the diagnosis and also wanted to know more about how PSP might affect the downstream generations. One of the children, a professor at UC Berkeley, had a colleague in the UC Davis Medical School who was doing research in areas related to PSP. He confirmed the physician's diagnosis and made some suggestions to her regular doctor about medications and also used her as a subject in research he was conducting.

During one of our visits to his clinic, he approached me about making a tissue donation to his research program. He explained about the short half-life of some of the chemicals they are studying in the brain reaction area and emphasized the need for a quick deep freeze of the brain tissue. He also emphasized the scarcity of good tissue samples available for study in this field.

We had a difficult decision and we suffered over it for several weeks. Just about at this time, the news came out about the PSP Brain Program at the Mayo Clinic. We reviewed this alternate arrangement with our research doctor and he agreed that the PSP Program was the way to go. The family agreed that we salvage something from Carolyn's travails, so we sent for the tissue donation informational packet.

By this time, she had declined to the extent that we were put on the Kaiser Hospice program. I asked the Hospice nurse for help in making arrangements. She was also new to this procedure. We quickly found that the hospital pathology departments work a 40-hour week. They were not suitable for the quick round-the-clock response that we needed. Several years earlier, we had pre-selected a funeral home and the funeral staff gave us the name of a pathology service that would meet our needs.

We signed on, paid the fees and distributed copies of the Mayo packet to the funeral home; hospice and the pathology service. The pathologist warned us about one potential problem. We live in a different county from the location of the mortuary and he alerted us that if the coroner in either county put a hold on the autopsy, he could not meet the 12-hour time limit for freezing the tissue.

The hospice people rallied round. After several meetings, they decided the best way to satisfy either coroner was to create a signed death certificate that would accompany her at the time of death. Several Kaiser physicians volunteered to be called at any time to sign. Hospice set up a special file for Carolyn so that the nurse on duty would know how to proceed.

Carolyn lingered for another 15 months. We were in and out of hospice care three times. We found it necessary to check with all parties on a regular basis to be sure that the arrangements were still in place. Carolyn died at home in her sleep just after midnight on a weekend when our regular Hospice nurse was on vacation. Even so, I called the Hospice line and the situation worked like clockwork. Within twenty 20 minutes, a backup nurse was at the house and confirmed death. She was completely familiar with the procedures. She roused one of the doctors to sign the death certificate and returned within an hour. Carolyn's body was picked up at 3 a.m. and the tissue donation procedures were completed by the pathologist by 9 a.m.

To put it mildly, it was a most difficult night. Yet, it was satisfying to see that all the pre-planning had worked out. Once you decide to make a tissue donation, you want the process to work and be successful. I cannot emphasize enough the importance of making arrangements in advance and then reviewing them on a regular basis.

The Aricept Trial

Irene Litvan, M.D. (Member of the Society Medical Advisory Board) conducted a double-blind trial of Aricept (donepezil) for mental and movement symptoms in PSP. Twenty-one patients received placebo or donepezil (10mg.) for six weeks, separated by a one-month washout period in between. Donepezil led to mild improvement on only one of several neuropsychological tests, the Double Memory Test. There was also a mild improvement in two measures of overall ease in performing everyday activities: the Activities of Daily Living section of the Unified Parkinsonism Rating Scale and the Mobility Score of the Parkinson's Disability Questionnaire.

Unfortunately, the small magnitude and highly circumscribed nature of these improvements suggest that anticholinesterase agents such as donepezil should not be recommended for patients with PSP.

Society for Progressive Supranuclear Palsy Brain Donation Program

For Diagnosis and Research on PSP
Society for PSP Brain Bank
Supported by the
Eloise H. Troxel Memorial Fund
Mayo Clinic Jacksonville • Jacksonville, FL 32224

The purpose of the Society for PSP Brain Donation Program is:

1. To provide families with postmortem diagnostic evaluation for relatives suffering from PSP and related disorders.
2. To provide tissue for PSP research to scientists at medical institutions or other research centers.

To obtain informational packets about brain tissue donations, please contact the Society for PSP.
Phone: (800) 457-4777, (410) 486-3330 / E-mail: SPSP@erols.com
SPSP, Inc. Woodholme Medical Building, Suite 515
1838 Greene Tree Road, Baltimore, MD 21208

A Tribute to the Society for PSP
"A Decade of Caring"
1990-2000

The Society for PSP Founders David and Reba Saks, their daughter, Leila Saks Bowersox, grandchildren and great grandchildren joined others in Baltimore on October 21, 2000 to recognize the Society for PSP and the many individuals who helped to make it grow and prosper. The evening's event was a time of remembering and appreciating the generous and dedicated efforts of so many. The following Sunday, the office staff hosted an Open House at the newly expanded Society office. Everyone agreed the office was very impressive with careful planning to house the future growth of the Society's needs. Congratulations to the Executive Director, Ellen Katz! A beautiful job!

Joanne Armstrong, Honorary Board Member, offers a toast of appreciation to everyone who has worked during the past ten years to help make the Society successful.

George Jankiewicz, Chairman of the Society's Board of Directors, welcomes guests attending the Society's 10th year Tribute Dinner.

Guests at the Open House

Guests Attending the 10th Year Society Tribute.

Sherly Sagel, Eric Lemmon and Tammy Lemon, family of Reba and David Saks

Society staff, Adrienne Bantum and Wanda Perry

Ellen Katz, Executive Director and Nancy Brittingham, Advocate Editor

An office at the Society's headquarters

The St. Louis Regional Symposium

On October 15, 2000 the St. Louis PSP Support Group Network, in conjunction with the St. Louis Chapter of the American Parkinson's Disease Association, held its first Midwest PSP Symposium, "Focus on Function: Practical Applications for PSP." Speakers included:

Guests pick up information at the St. Louis Symposium

Welcome:
Mark Fontana, Registered Nurse
Amy Mandlman, Speech Pathologist
Patty Mueller, Human Resources

The Neurology of PSP and a Question and Answer Session
Brad Racette, M.D.,
Washington University School of Medicine

What You Need to Know: Legal, Swallowing and Dietary Issues
Craig Kallan, Attorney-At-Law
Jean Foster, Speech Pathologist
Jolynda Langston, Registered Dieticiain

Simplify Your Life: Practical Solutions You Can Implement Today
Sue Ekberg, Licensed Professional Counselor
Jacque Cornell, Physical Therapist
Janet Ferry, Occupational Therapist

These speakers taught functional and effective strategies to help those with PSP and their caregivers cope and simplify their lives. Each speaker prepared comprehensive handouts which will be available at a future date for further distribution. Separate videos for each topic will be available for your review. Watch for more details in the next Advocate.

The Society wishes to thank the St. Louis Support Group for PSP and the St. Louis chapter of the APDA for their tremendous efforts in organizing and presenting this program. Special thanks to Amy Mandlman, Patty Mueller and Mark Fontana for chairing this program.

Continued Page 12

The Second Toronto PSP Symposium

By Stephen Hamer
Member, PSP Society Board of Directors

Organizers and Speakers at the Symposium

The James Cameron Memorial Symposium was presented in Toronto, Ontario, Canada on Saturday, Nov.4, 2000 to a packed house. As Janice Stober, the Symposium coordinator, said, "PSP is like a 'Field of Dreams.' If you have a meeting, they will come." And so they did.

People were eager to meet other people and families with PSP and to hear from an outstanding group of professionals who had gathered to share their expertise. Everybody felt it was worth the drive to Markham Stouffville Hospital. While the medical perspective and research initiatives were ably covered by Mark Guttman, M.D. and Paul Fitzmaurice, M.D. respectively, the main emphasis was on the everyday challenges people with PSP and their families face.

The three-hour symposium was broken down into 20-minute segments. Questions and answers were left to the end of the program. Everyone received a folder containing the slide summaries of each presentation, along with brochures from the Society for PSP, including "Visions and Opportunities," which describes the mission and goals of the Society over the next three years.

The topics discussed included maximizing mobility, everyday living strategies, communication challenges, swallowing problems and nutritional issues. All of the speakers work at the Baycrest Centre for Geriatric Care, a unique facility that provides fully integrated services for the elderly in Toronto. Each segment was practical and useful, identifying specific workable examples.

Lynda Dunal, an occupational therapist, talked about everyday living strategies including tips on how to safely get in and out of the bathtub and bathrooms and the equipment available for that purpose. She also talked about practical ways to dress, including types of clothing that offered sensible dressing solutions, such as wearing slip-on shoes versus lace-ups, etc. Lynda stressed that balance is vital for keeping up spirits and staying nimble. She stressed the importance for both PSP patients and caregivers to conserve energy by paceing themselves, being well-organized and planning ahead. Leisure activities are essential for everyone, and there are many practical devices to use, such as large button remote controls for TVs and "postettes" with written messages of things we need to

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The Second Toronto PSP Symposium
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remember. Safety in the home was discussed. Motion sensors installed within the home can alert caregivers to the movement of the person with PSP. Voice-activated light switches are available. Lynda closed with some words of advice for each caregiver: Be gentle with yourself. You are an enabler, not a magician. It is okay to say "no." If you never say no, what does "yes" mean? Schedule time for yourself and change your routine as often as feels comfortable. Try to find someone you can talk to and learn to recognize the difference between what relieves stress and the complaining that reinforces stress. Give support, encouragement, and praise and learn to accept it in return.

How do we retain the ability to move easily and make it easier to do things like getting in and out of bed or a chair? Tanya Aggett, a physiotherapist, offered some helpful advice in talking about the role of exercise, how to move more easily in bed, tips for standing up and sitting down, and finally walking and walking aids. Tanya discussed the importance of trying to maintain flexibility. This helps people stay independent as long as possible. She pointed out that it is important to exercise and to use aids to compensate for loss of flexibility whenever possible. Heel lifts for balance, floor-to-ceiling poles for getting in and out of chairs, and swivel seats for cars were a few examples. She advised people to contact a physiotherapist as early as possible and make sure everyone is involved.

What about tips for swallowing and nutrition? We all know about the consequences of dysphagia (or difficulty with swallowing), such as malnutrition, dehydration and aspiration pneumonia. Nicole Shuckett and Lisa Sokoloff (speech therapists) handled this segment. They suggested that the person with PSP undergo a swallowing evaluation. It is vital to remain vigilant watching for uncoordinated tongue movements, accumulations of secretions in the throat, slowed oral movements, and material left in the mouth.

The goal is to manage feeding in order to maintain adequate nutritional levels while eating foods that are easy to chew and swallow. They emphasized the need to assume the best posture for eating, keeping the environment non-distracting, maintaining a slow rate of feeding, and keeping the plate of food in the line of vision of the eater.

They offered other helpful tips on swallowing and foods. Clean the person's mouth of secretions before and after eating. Alternate between foods and liquids when eating. Take small bites; and sip the drinks. Thicken liquids if they are hard to swallow, and finally, be alert to signs of pneumonia (i.e., chest congestion, chronic low-grade fever, cough, change in sputum).

Susan Romeril, a speech language pathologist, reviewed some simple solutions to address the speech difficulties that challenge a person diagnosed with PSP. Some of the difficulties include an inaudible voice, imprecise articulation, stuttering speech and forgetfulness. Speech pathologists can recommend techniques to increase loudness and ease of speaking. She demonstrated the use of an alphabet board which can be used for first letter cueing. She also showed us a "phrase board" with emergency items. One could also establish an attention-getting signal. Technology also makes its entry via the "Lightwriter." This is an amazing device that can speak. Descriptions alone will not do it justice.

Suffice it to say that you should immediately contact a speech pathologist who can get you on the right road and keep you going in the right directions. The last word should really go to Lynda Dunal, who pointed out that small things do make a difference. (For those who believe differently, try going to sleep with a mosquito in the room.)

The Society for PSP wishes to recognize these special Society friends and fundraisers!

- CAROLYN CHEEK, (Los Angeles Walk-a Thon) raised $4,000
- CHARLOTTE TRIPET, (Minnesota PSP Dinner) raised $2,700
- KEN TODD, (ran the New York Marathon) raised $3,000

Congratulations and thank you.

The publications of the PSP Advocate for the Year 2000 have been generously funded by Jay Troxel in memory of his beloved wife, Eloise H. Troxel

Dallas, Texas

"An Afternoon of Sharing and Caring for Persons with PSP, Families and Caregivers"

October 21, 2000

Malcolm Stewart, M.D. discusinng PSP

The Society for PSP hosted an organizational meeting of "Caring and Sharing for Persons with PSP, Families and Caregivers" on the afternoon of October 21, 2000 at the Marriott Suites Dallas Market Center. An invitation was mailed to families located within the Dallas/Ft. Worth area. Thirty-eight guests attended of which five persons were diagnosed with PSP. Nancy Brittingham, Editor of the PSP Advocate and former caregiver to her father who was diagnosed with PSP, represented the Society at this meeting.

The Society wishes to thank the special guest presenters who volunteered their time and efforts in addressing PSP issues at this meeting. Malcolm Stewart, M.D., a movement disorders specialist from Fogelson Neuroscience Center, Presbyterian Hospital of Dallas, presented an overview of PSP. He then welcomed an hour session of question and answers. Susan Imke, R.N., M.S., Gerontological Nurse Practitioner, former educational consultant to the National Parkinson Foundation and who currently maintains a clinical practice in Ft. Worth, described tools and skills needed to better manage the adjustments in day-to-day life with chronic neurological illnesses. Mary Cudly, R.N., Licensed Social Worker also from Fogelson Neuroscience, discussed resources in the area and the importance of investigating neurological outpatient rehab programs including the services of licensed speech and language pathologists, physical therapists and occupational therapists.

Families spent time meeting each other and sharing some of their PSP experiences and challenges. Plans began on establishing and maintaining communication between members of this new Dallas PSP Support Network.

Special thanks to Debra Stantec, Marketing Director, and all the staff at the Marriott Suites Dallas Market Center for their kindness and consideration as they acknowledged the special needs of those impacted by PSP.

John W. McDaniel with loving and supportive family

Some of the attendees at the Dallas meeting

The 104th Annual Meeting of the American Academy of Ophthalmology

Dallas, Texas • October 22-25, 2000

Over 14,000 physicians and health professionals attended the American Academy Of Ophthalmology's 104th Annual Meeting at the Dallas Convention Center in Texas last October. This meeting provides the highest quality of continuing medical education available to ophthalmologists today. There is opportunity to develop, maintain and enhance knowledge and skills in research and clinical advances through the various

PSP Volunteers Lynette Henery, Carol and Mel Miller

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Special Events

The 104th Annual Meeting of the American Academy of Ophthalmology

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learning formats. The latest and most advanced products and healthcare services available are housed in several exhibit halls stretching over 240,000 square feet of space.

The Society's informative exhibit at this annual meeting educated national and international ophthalmologists about the diagnosis of PSP and informed them about the services of the Society. Most persons with PSP begin their search for diagnosis with ophthalmologists. Physicians attending these types of meetings are given an electronic swipe card with their contact information which is imported into a mailing data base. There was an overwhelming number of ophthalmologists swiping their cards to obtain more information about PSP. The Society office staff immediately began mailing out Physician PSP Information Packets to these ophthalmologists.

The Society would like to thank special PSP friends: Lynette Henery, Carol Miller, Mel Miller, Jane-Lauren Brittingham and Kristen Brittingham for volunteering their time helping to distribute information at the Society exhibit.

Daughters of the Advocate Editor, Jane-Lauren and Kristen Brittingham, help to set up the PSP exhibit and distribute information

The St. Louis Regional Symposium

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The Society for PSP would like to thank the following St. Louis PSP Regional Symposium Corporate Sponsors:

COMMUNICATION AIDS
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St. Louis Symposium Organizers, Mark Fontana, Amy Mandlman,Brad Racette, M.D. and Patty Mueller

Corporate sponsors with demonstrations and product information

"METAMORPHOSIS"

Susan Hunt, Grief Counselor
Los Gatos, CA

PHASE I:

You walk into your doctor's office feeling hopeful. Your doctor has called to tell you that at last, she/he feels he knows what is wrong with you. He has a diagnosis. After months (years?) of questions, tests, disappointments, and discouragement, you are finally going to get an answer, a label for the symptoms that have plagued you for so long. You have been frustrated, angry, sometimes frightened by the medical profession's inability to diagnose what is happening to you. Now you will know and it will be such a relief!

Half an hour later, you walk out of the doctor's office dazed and shaken. Words like "chronic," "progressive," "degenerative," "possibly fatal," swirl in your mind. How can this be? Not ME! This can't be happening to ME. No! The doctor must be mistaken. What is this disease called PSP?! I didn't even know there was such a disease. It sounds so terrible. . . and I've never even heard of it before.

Those few moments in the doctor's office have changed your life and the lives of those who love you forever. And you simply can't take it in. It doesn't seem real. You feel like you're in a dream. Deep inside, the feeling of dread is already growing. What's going to happen to me? What is this rare diseas eall about? NO! NO! It can't be!

If you have been there, you remember. You may not recall all the details, but you will never forget the feelings. You may have been totally shocked, but you probably already suspected that something was terribly wrong. Now you know what it is (at least you have a name for it) and your world has suddenly turned upside down. Everything has changed. None of the rules by which you have lived your life apply any more... or do they? Which ones?

So many questions... How do I tell my family, my loved ones? What will they say and do? What will I say and do? How long will I live? What will my life be like? Am I going to become more and more limited, helpless, become a burden? NO! I can't let that happen. I can't do this! This isn't the way it is supposed to be, I have plans, dreams...

The reaction for each individual may be different, but the bottom line is you are going to be called upon to utilize every ounce of strength, courage, and integrity that you have. You and your loved ones are going to have to "reinvent" yourselves and your relationships. The metamorphosis has begun.

PHASE II

In the first few weeks after diagnosis, it is going to be very important for you and for your loved ones to be extremely kind and patient with yourselves and with each other. Often after a life-threatening diagnosis, particularly one that involves great physical and/or mental changes, there is a period of self-flagellation: "If only I hadn't smoked, or been overweight, or... I'd be OK." Or, if you have been very careful with diet and lifestyle, there can be anger and frustration and a feeling of being betrayed. "It's not supposed to happen to me. I took care of myself. It's not fair!" (You are right about that. It's NOT fair. But it's the way it is.) A diagnosis of PSP fills you with much uncertainty about your plans and dreams for the future. As the disease progresses, you fear the loss of feeling in control of your life. The disease has control, or at least that's how it seems. It dictates how you will live from now on, and how and when you will die. These feelings need to be acknowledged and expressed. They are the normal reactions to what is happening to you. And they will probably be with you, at least on some level, for the rest of your life. Be gentle with those feelings. Allow and express them. They are valid. And they change drastically how you view yourself and your relationship to others.

Other feelings may come up for you and your family that you have never experienced before: anger, fear, disappointment, hopelessness, helplessness. Typically, you will be encouraged to "be positive" and "look on the bright side." (What bright side, you wonder.) Depending on your "style" of dealing with loss, you may scream and throw things or you may withdraw silently to grieve. You may be able from the beginning to present a positive, courageous outlook, at least on the outside. Inside, it may very well be a different matter. Close loved ones and family members have their own reactions to the diagnosis. They are struggling with their own questions and grief. You don't want to let them know how deeply you are affected, yet you need their support and understanding. You may never have needed help before, but YOU NEED HELP WITH THIS NOW!

Friends try to support you. Some are devastated and show it. (This can be a help if you need validation and support for your grief process or a terrible ordeal if you are trying to be strong. Then, you need them to be strong with and for you.) Some may be unbelieving:" Have you gotten a second opinion?" (That's not always a bad idea.) Others are terribly cheerful and 'supportive,' "You're strong!" And you think, "Am I THIS strong?"

Relationships are really put to the test at this point. You've always been the one who helped others. Now you are going to be on the receiving end and you don't like this reversal of positions. You don't want to be "helpless" or a "burden."

Can you stop for a moment at this point? Can you take a deep breath and slowly, quietly recall the last time you helped someone you loved, just because they needed it? How did that make you feel? Did it give you a sense of being useful and valuable? Was there pleasure for you in being able to give something to, or do something for someone you cared about? Now you can turn that around and give someone who loves you the opportunity to experience being needed and appreciated.

Hard, hard, hard—but possible? There are so many changes all at once. Because PSP is so difficult to identify, many physical changes may have already occurred. More are coming and possibly very quickly. It's all so unexpected and unpredictable—and overwhelming. Learning more about the disease, talking to others who are going through the same experience, finding ways to express the feelings (most of us have never learned to do that) all can help. You will have different concerns than your loved ones and all of you need support at this time. Seek it out.

TALK with each other, with those who are knowledgeable about PSP and about dealing with illness and loss. Try to deal with one concern at a time, take one day at a time. Grieve and cry when you need to, separately and together. Then, take a deep breath and try to take one more step. There is no way to "accept" what is happening. It will never be "acceptable." There IS a way, however, to come to some accommodation, to

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Metamorphosis

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find a way to go on with your life. You are faced with the greatest challenge you have ever known. It's not a choice of WHETHER you are going to do this. It is HOW you and your loved ones are going to do it.

PHASE III

There is an aspect of this situation that is not always easy, at least in the beginning, to see. As you begin to "accommodate" to a new way of living, responding to the changes occurring in your body, it will be important to try to acknowledge the PERSON that you still are. You are not your disease! You are the living, loving, vital person inside that you always were. Your view of life and your place has changed, but you are still YOU!! And there ARE things over which you do still have control.

Can you learn to laugh at yourself, at some of the "ridiculous" things that are now a part of your life? Can you shift your focus for a little while, and again notice some of the beauty around you? Hear the birds singing their hearts out, notice the trees and flowers unfolding their beauty, watch a sunset and glory in the changing colors. Can you learn to accept, with loving gratitude and not guilt, the help and support of others? As a caregiver, can you learn to also take care of yourself so that you continue to have the physical and emotional energy to be supportive?

The greatest challenge of this terrible disease may also be its greatest opportunity: finding a way to change and grow inside; to love yourself and those around you more deeply; to see and experience more fully the simple, ordinary beauty that is all around you; to listen more consciously and see more clearly all the little things that are still good in your life. (They may be hard to see in the midst of all that's happening but they ARE there.)

Metamorphosis, change, is an inevitable part of our lives. It is not always comfortable, and sometimes, as with a diagnosis of PSP, it is terribly devastating. Choosing to explore the possibilities for LIVING with this disease, as fully as you can, for as long as you can, will bring healing for the whole person that you still are inside. Seek out the support and help that is available. Allow the grief. Look for the gifts. Be gentle and kind with you.

(Susan Hunt has been an educator, counselor, minister, and program developer. She has trained professional and volunteer grief counselors for The Centre for Living with Dying. She also was responsible for the Centre's Senior LINC Program that provides emotional support to older individuals in the community who are coping with loss and change. She specializes in grief, loss, and pain management.)

Diet Suggestions for the Person Diagnosed with PSP

Pat Beekman
Listserve

I used to give Ed 3-4 choices then repeat them and he would raise his hand to indicate which he wanted. I gave him anything he wanted: from steak to pizza, spaghetti,meatballs, fried chicken.(de-boned),roasts, Arby's or Burger King sandwiches, bacon-eggs-toast to BLT's. You name it, he ate it. I would fix it the way I always did before and then put it in the food processor and blended it to the consistency he could handle. If food was too thick, I added water or milk. If it was too thin and juicy, I added baby rice cereal or instant potatoes. Leftovers or extras went into zip lock snack-size baggies and into the freezer for another meal. He ate like this till the week before his death

If your loved one with PSP is not ready for blended foods, try soft things like cooked cereal or scrambled eggs, canned soup, apple sauce, puddings, jello, Instant Breakfast or homemade milkshakes with fruit. Ed liked angel food cake, crumbled, with jelly or pudding or fork-mashed fruit on top and then a little Cool Whip, and he could handle little bites of moist cake. Creamed corn was good and I made cream soup with spinach or broccoli and cheese.

Cooked prunes were an every morning item. It was odd but he could swallow them without a problem almost to the end and then I pureed a week's supply at a time and stored it in a covered bowl in the fridge.

The important thing was to give him a balanced diet with plenty of protein, fruits, veggies, milk products, (cheese, cottage cheese, eggs) and some type of starch. I used lots of instant potatoes, and noodles smashed with a fork. Casseroles like macaroni and tuna, or ground beef with macaroni and tomatoes also are easier to swallow as the sauce helps it slide down.

It's mostly a matter of trial and error. Try something; if it doesn't work out, try another. Usually, it just takes an adjustment in consistency. The most valuable thing in my kitchen was my food processor. With that, he could eat anything he wanted.

When he could no longer hold the spoon I fed him so he wouldn't get so tired he wouldn't eat. Make sure your PWPSP gets enough fluids. It does not have to be liquid—jello, pudding, ice cream, popsicles, thick juice that comes with canned fruit, watermelon—anything that has or is made with a liquid is considered a fluid. To get a higher calorie intake it might be wise to offer snacks between meals or 5-6 smaller meals at more frequent intervals

I found that a throwaway plastic coffee cup that bends when squeezed worked wonderfully. The bend let it adjust around his nose. I filled it about 2/3 full. Another thing I used was a quiche dish; the sides kept the food in the plate; or a baby's divided dish with hot water reservoir underneath to keep the food warm.

Use your imagination!!! To give pills, I put them in a spoon of applesauce and he took them without crushing. If they must be crushed, always check with your pharmacist first as some meds should not be crushed. Some are coated to protect the stomach or to delay the release of medicine into the system. Ask if they come in liquid form

I hope it is possible to keep your loved one at home. It is possible, but it is a lot of work! I don't regret it, though. I know I did everything I could for him as a 24/7 caregiver and he was happy being at home. The biggest thing you face is having enough dependable help for 24 hour care.

Pat B. Ohio

(Note from the Editor: I also purchased a portable hand-held electric food grinder as well as a hand cranked food grinder that I took with me on outings or other people's homes and restaurants. My dad had some favorite things he liked to order when we went out to eat at restaurants and no one ever objected to me going into the kitchen and grinding up his meal. Also, I used the crockpot to cook for many recipes and that was a big help in getting foods to the right consistency.)

"Ask a Doctor"

By Stephen G. Reich
Assistant Professor, Neurology
Johns Hopkins University School of Medicine
The Society for PSP, Chair Emeritus

PSP: WHAT'S IN A NAME?

It's bad enough having a disease no one's heard of and tougher still when it goes by a name that is not only incomprehensible but also a tongue twister. Such is the case with progressive supranuclear palsy. Mercifully, it is generally referred to as PSP. To confuse the situation even further, it is occasionally referred to by the initials SRO, short for the eponym Steele-Richardson-Olszewski after the three Toronto physicians who described it in 1964

What does progressive supranuclear palsy mean? The term progressive is straightforward - the disease does get worse with time, albeit very slowly. Palsy, another term for paralysis, refers to the impaired ("paralyzed") upward and downward movement of the eyes which is the most distinctive feature of PSP. While there is no actual "palsy" of muscles in PSP, the slowness and incoordination which characterize the disease often have a similar effect on functioning.

The word most people stumble over in PSP is supranuclear: To understand this term, you must first understand something about eye movements. Each eye is surrounded by six muscles which allow it to move in multiple directions. Two of the muscles control horizontal (side-to-side) eye movements and the other four control vertical (up-and-down) movements. These muscles are in turn controlled by three separate nerves, each with a nerve cell origin–known as a nucleus–located in the brain-stem. If the machinery for controlling eye movements was made up solely of the nerves and muscles, we would not be able to move our eyes in sync, and hence, would experience double vision (diplopia). In order to keep the nerves and muscles from functioning independently, they are controlled by groups of nerve cells, known as gaze centers, which ensure that the eyes move together, known as yoking. Since these gaze centers arise before or "above" (supra) the origin of the individual eye muscle nerves in the brainstem (the nucleus), they are referred to as supranuclear centers. The gaze center for coordinating vertical eye movements, upward and downward gaze, is located in the upper portion of the brainstem known as the midbrain, the area which bears the brunt of the damage in PSP. The center controlling horizontal or side-to-side eye movements is located in a lower portion of the brainstem known as the pons, which is almost always spared in PSP, accounting for the disparity between impaired vertical eye movements but normal or near normal horizontal movement.

Just why such selective regions of the brain and brainstem are affected in PSP remains a mystery but at the same time tells us where we should be searching for answers. What's in the name progressive supranuclear palsy? Maybe a clue about what causes it–since in neurology, like real estate, it's location, location, location!

Swallowing Problems in PSP

By Laura Purcell Verdun, M.A., CCC
Research Speech Pathologist

Feeding and swallowing problems are very common in progressive supranuclear palsy (PSP). Dysphagia is the medical term for disordered swallowing. There are many reasons for dysphagia in PSP: difficulty looking down may limit an individual's ability to see the plate during meals; loss or defective brain cells may affect control of the swallowing mechanism; there may be a change in behavior resulting in rapid drinking or mouth stuffing; poor self-feeding because of tremor or stiffness, extended head and neck posture and dementia.

The two primary concerns regarding swallowing are:

1. The ability to swallow safely and easily, to keep food and liquid from "going down the wrong way" into the trachea or "windpipe," known as aspiration, increasing the risk for developing pneumonia;
2. The ability to maintain an adequate intake of fluids and calories.

There are several warning signs of a potential swallowing problem including: drooling, food collecting in the mouth, apparent increased effort with swallowing, a "wet" voice, trouble swallowing medications, increased length of mealtime, low grade fever, chest congestion, trouble talking, coughing and choking with a red face, weight loss, need for the Heimlich maneuver, or pneumonia. Some individuals with PSP may not recognize that they have a feeding or swallowing problem. This is called "silent aspiration", and occurs when the individual does not cough or choke 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 who specializes in swallowing problems and is familiar with PSP. It is often beneficial to maintain a diary of the swallowing difficulties observed, including the setting in which the symptoms occurred, and with which foods or liquids. This will make the swallowing evaluation more thorough and the recommendations more specific to your needs.

In some situations, the speech pathologist's evaluation may determine that continued intake by mouth is unsafe because of the risk of aspiration, or may be inadequate resulting in weight loss. At this time a feeding gastrostomy tube may be considered. This is a very personal decision and should be discussed at length with family members and physicians. A feeding tube will provide adequate nutrition; however, it will not entirely eliminate the risk of aspiration.

Feeding Suggestions:

1. Sit upright, preferably in a chair, during meals.
2. Eliminate distractions such as television or conversation during meals.
3. Concentrate on maintaining a slow, steady rate of feeding.
4. Keep the plate of food in the line of vision, for instance, by placing it on a telephone book, if you have difficulty looking down.
5. Substitute a pair of reading glasses for bifocals, which are often ineffective in the setting of impaired downward gaze.
6. Experiment with different plates and utensils. Plates with a rim can be useful for keeping food on the plate. Using a spoon instead of a fork, or one with a modified grip, can make it easier to eat.

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Swallowing Problems in PSP

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7. Experiment with different cups and glasses. Many different cups are available to restrict how much liquid you can take in your mouth at once. Safe swallowing cups or dysphagia cups help maintain your head in a safe swallowing position, usually head neutral or chin down.

Swallowing Suggestions:

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-tucked position when eating or drinking.
3. Avoid extending the head back while eating, drinking or taking medications.
4. Avoid taking too large a bite, drinking too rapidly or placing more than one bite in your mouth before swallowing.
5. Alternate swallowing food and liquids to assist with clearance of drier or more textured foods.
6. Avoid highly textured foods (red meats, raw vegetables, etc.) or dry particulate foods (rice, pretzels, potato chips, crackers, cookies, etc.). Consider changing to soft, moist foods such as canned fruits or cooked vegetables, fish and chicken, complemented with gravies and sauces.
7. Avoid items of mixed consistencies, such as cold cereals, chicken noodle or vegetable soups and fruit cocktail. These items should be blended into one consistency.
8. Thin liquids may be difficult to swallow because they move faster through the mouth and throat, or because there is a slight delay in the onset of the swallow and closure of the airway. Thicken liquids with a commercial thickener (such as Thick-It, Thicken-Up, etc.), potato or banana flakes, or fruit purees.
9. It may be easier to take medications with a spoon of puree consistency such as pudding or applesauce instead of liquids. Do not crush mediations unless approved by a physician.

General Suggestions:

1. It is important to maintain good oral hygiene.
2. Dairy products often have a tendency to make secretions thicker, so minimize these products.
3. A portable suction machine may be of value to assist with clearance of secretion, particularly at meal times. Drinking more water, or sometimes use of carbonated beverages may help reduce or thin secretions. Your physician may have suggestions for medications to thin or reduce secretions. Avoid medications that dry up secretions, making them thick and ropey and more difficult to manage.
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 weight loss.
6. Monitor the medications being 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 pneumonia, including increased chest congestion, chronic low grade fevers, increased cough, and change in sputum.
8. The use of nutritional supplements such as Ensure, Carnation Instant Breakfast, Sustacal, etc may be of value to increase caloric intake quickly and easily.
9. Supervision during all meals is always a good idea.

There are cookbooks available for those who have difficulty swallowing.

Non-Chew Cookbook by J. Randy Wilson
Wilson Publishing, Inc.
P.O. Box 2190
Glenwood Spring, CO 81602-2190

MEALS
The Muscular Dystrophy Association
810 Seventh Avenue
New York, NY 10019

Pureed Food with Substance and Style
Imaginart Communication Products
307 Arizona Street
Bisbee, AZ 85603
800-828-1376

Attention California Residents

Available to help those dealing with PSP is an organization called "Coast Caregiver Resource Center." This group is a part of the statewide network of Regional Resource Centers serving caregivers of adults with degenerative disease of the brain. A wide range of services are available including financial assistance toward additional caregivers. For further information, please call 1-800-443-1236.

"I Am Not Alone Anymore"

The Advocate arrived yesterday and it was like a bright star in a dark sky. I am not alone anymore. There are others who understand PSP. However, no one close to reach out to. If I feel this way as a caregiver, how must the person diagnosed with PSP feel? We live in a good medical area but no one knows about PSP. We are the educators while we need to be educated ourselves. PSP consumes every inch of our lives and we are always grasping for things that could help with the frustration that has come with this disease.

As of now, the Advocate has been that straw that we seem to be needing. Thank you.

Edna K.

(Editor's note: Thank you to all the people who so graciously have contributed their unique talents through the years in helping to produce the PSP Advocate. A special thanks to Lawrence Golbe, M.D.; John Hargett at Gear Up Printing; and Jay Troxel, who has generously funded this past year's production of the PSP Advocate and has pledged to do the same for 2001.)

Support Groups

PSP Support Groups will encourage and organize activities that foster communication, exchange and interactions of comfort and mutual benefit to Support Group members who are family, friends, caregivers and persons with PSP. The Society would like to thank the following Support Group Leaders and Communicators who take their time and show their concern by sponsoring support groups, phoning and visiting PSP families. If you would like to help start a support group in your area, please call the SPSP office at 1-800-457-4777. For information on support groups in your area please contact:

ARIZONA:
Gale Kittle & Christina Watts
Muhammad Ali Parkinson Res Cent
500 W. Thomas Road, Suite 720
Phoenix, AZ 85013
(602) 406-4931
Maxine Faubion
Sun Lakes, AZ
(480)895-7646

Renae Valdes-Birch
Gilbert, AZ
(480) 632-1797

ARKANSAS:
Patsy Cunningham
Fort Smith, AR
(501) 648-1814
patphillip@prodigy.net

CALIFORNIA:
Carolyn Cheek
Torrance, CA
(310) 534-8623

Lila Kirkpatrick
Santa Clara, CA
(408) 244-5958

Carol Platt
Antelope, CA
(916) 332-6041

Mary Miano
Laguna Woods, CA
(949) 855-3972

Kathy Schweiger
Thousand Oaks, CA
(805) 496-7018

CANADA:
Sandie Jones/Janice Stober
Parkinson Foundation of Canada
4211 Yonge Street, Suite 316
Toronto, ON M2P 2A9
CANADA
(416) 227-9700;
stoberj@hotmail.com

Marie-Josee Fortin
The McGill Centre for Studies in Aging
Douglas Hospital
6825 LaSalle Blvd.
Verdun, Quebec H4H 1 R3

Carolyn Connors
The Parkinson Foundation of Canada
150 Newbold Count
London, Ontario N6E 1 Z7
CANADA
619-630-0430
parkinso@hotmail.com

COLORADO:
Dr. Lauren Seeberger/Kim Mantin
Movement Disorder Center
Colorado Neurology Institute
701 E. Hampden, #530
Englewood, CO 801101
(303) 788-4600

CONNECTICUT:
Frank Cadwell
Clinton, CT
(860) 664-9524

FLORIDA:
Paula John
Sarasota, FL
(941) 927-3955

Shirley Vlahakis
West Palm Beach, FL
(561) 969-9553

Virgie Saltzman
Nokomis, FL5
(941) 484-7259

John Arnold
Cape Canaveral, FL
(407) 784-5660

Bud Branson
Quincy, FL
(850) 627-6216

Linda Ives
DeBary, FL
(407) 668-7130

Helen Lovelle
Naples, FL
(941) 352-2909

FLORIDA (Miami)
Beatriz Chirino
(305) 270-8401

Yolanda Medina
(305) 553-4911

GEORGIA:
Kathy Thomas
Decatur, GA
(770) 939-2612

ILLINOIS:
Janet Kaiser
Gelnview, IL
(847) 729-1585

INDIANA:
Elizabeth Risk
Indianapolis, IN
(317) 786-8613

IOWA:
Esther Cooling
Cedar Rapids, IA
(319) 363-4752

KANSAS
Linda Smith
Salina, Kansas
(785) 825-6482

LOUISIANA:
Mary Schumann
New Orleans, LA
(504) 484-7840

Brenda Gremillion
Kenner, LA
(504) 467-6658

MAINE:
Faye Ryan
Whiting, ME
(207) 259-2152

MARYLAND:
Ruth Goldstein
Baltimore, MD
(410) 486-2822

MASSACHUSETTS:
Patti Ryan
Swampscott, MA
(781)595-4431 (H);599-1000 (W)

MICHIGAN:
Carol Ann Kiank
Commerce Township, MI
(248) 363-9064

Janet Maguire
Saint Claire Shores, MI
810-294-0071

Sally Smith
Zeeland, MI
(616) 722-4446

MINNESOTA:
Charlotte Tripet
Golden Valley, MN
(612) 546-1694

MISSISSIPPI:
Denver Flanagan
Jackson, MS
(601) 372-1784
denfian@aol.com

MISSOURI:
Pay Lynn
Jackson, MO
(573) 243-3964

Amy Mandiman
St. Louis, MO
(314) 432-5461

NEVADA:
Carol Upton
Las Vegas, NV
702-731-8329;
parkinsonslv@cs.com

NEW JERSEY:
Mary Gualandi
Toms River, NJ
(732) 505-1739

NEW YORK:
Mary Connolly
Canandaigua, NY
(716) 394-5306

Marcy Todd
Port Washington, NY
(516) 883-7455

OHIO:
Debbie Mills/Parkinson's Center
P.O. Box 670525,
Univ. of Cincinnati
Cincinnati, OH 45267
(513) 558-7312

Bernice Bowers
Canal Winchester, OH
(614) 833-2588

Jennifer P. Smith
Barnesville, OH
(740) 425-3253

OREGON/NORTH CALIFORNIA:
Betty Scoggins
Redding, CA
(530) 241-6663

OREGON:
Sandi Hansen
Clackamas, OR
(503) 698-8129

PENNSYLVANIA:
George Weaver
Mill Hall, PA
(570) 726-6164

George Weave
Mill Hall, PA
(570) 726-6164

PENNSYLVANIA (South Central):
Daniel K. Lake
Willow Street, PA
H: (717) 464-4057
W: (717) 397-9037
e-mail: dansr@desupernet. net

Ruth Nulph, R.N.
Butler, PA
(724) 287-8600

Jane Wright
PA Hospital, Neurology Department
330 S. 9th Street
Philadelphia, PA 19107
(215) 829-7273

RHODE ISLAND:
Kelley Harrison, Ph.D.
Providence, RI
(401) 621-8566

SOUTH CAROLINA:
Doris McCray
Myrtle Beach, SC
(843) 445-1647

TEXAS:
Karen Kennemer
Kingwood, TX
(281) 358-2282

Judy Brannen
Benbrook, TX
(817) 249-0824

VERMONT:
Janice Clements
Milton, VT
(802) 893-1263;
janclem@together.net

VIRGINIA:
Allen Rohifing
Virginia Beach, VA
(757) 426-5281

Kathy M. Sands
Vienna, VA
(703)242-9322; (703) 676-5319
e-mail: Kathy. rn. sands@saic. corn

WASHINGTON:
Roberta Hunt
Walla Walla, WA
(509) 529-1364

Fran MeHahon
Rochester, WA
(360) 273-9496
franmcdoll@aol. com

WISCONSIN:
Barbara Sharkey
Rhinelander, WI
(715) 362-1777

The Los Angeles County/ Orange PSP Walk

By Carolyn Cheek

The Los Angeles County/ Orange PSP Walk was a roaring success. As we walked along the beach in Redondo Beach, the sun was out, the ocean was blue and beautiful, our T-shirts were terrific. What more could we ask?

Caroly Cheek with daughter, Jan Bamrick and granddaughter Shannon Bamrick

The L.A. County/Orange County Support Group was joined by the Laguna Hills Group and the brand new Thousand Oaks Group in this event. This three-way effort was rewarding in many ways. The Laguna and Thousand Oaks groups mostly sent walkers - and that was what we needed most. Believe it or not, the planning was done in only one meeting. We raised $4,000 net.

The pastor of the Redondo Beach United Methodist Church, our HQ, was asked to give an inspirational short talk before the walkers took off. The Rev. Stan Ferguson said he was reminded of the story of a slave who told of the flax shirts that the slaves wore. They were made of the poorest and stiffest part of the flax and were agonizingly scratchy in the beginning. The slave said that the most loving thing anyone had ever done for him was when his brother took his new shirt and wore it until it was broken in. Rev. Ferguson went on to say that our T-Shirts were worn in that same spirit of love for our loved ones and all persons with PSP. What a fitting analogy.

Sights to behold:

Paul Wicorek and his three grandsons, ages 8 to 11, walking down to the beach. The three boys helping put up signs and ferrying items from the parking lot to the hall. Bravo to grandsons.

Carolyn Griffith, a foster parent, laying out donut holes with a baby in her arms and another one in the carriage. Bill Griffith, her husband, along with their eight year-old, recording all of the registrations and taking in the money. He and Carolyn both walked. What a team! Carolyn also designed and ordered 50 T-shirts that the walkers wore along with signs on their backs that read PSP WALK and underneath a line to say "honoring_____".

Norma Simon got there early – put up signs in the patio, delivered cookies, water and tote bags. Then she took water and manned a water stop halfway through the walk.

May Toll came very early and made REAL coffee and tea to go with the donuts and juice. May walked and then came back to help serve lunch.

Mary Miano, a ten-year PSP patient, and leader of the Laguna Hills Support Group, walked FOUR miles. Go Mary!

A young dentist and his wife with a twin stroller and his one-year-old and two-year-old walked down to the beach.

Carolyn Cheek's granddaughter, Shannon, and her daughter, Jan, pushed her around the two-mile walk in a wheelchair (she had knee surgery). They then proceded to walk the other four miles and then come back to play with the babies.

Penny Roberts brought five friends to walk. Penny lost her husband only four weeks before the event. Wendell Higbee helped with signs and stationed himself as greeter.

Wayne Stark and his wife, Margaret, walked the six miles. Wayne had laid out the course and served on the committee out of friendship for Carolyn and her late husband, Bob, a PSP patient.

Rhoda Cocca was everywhere, helping everybody and walking, too.

Our day was an outstanding example of teamwork and caring. All of the walkers expressed their joy at being able to help fight PSP in a concrete way. The stated purpose was to raise funds and raise awareness of PSP. I don't think we had counted on having a day of fellowship and friendship. But, we certainly did that!

MASSACHUSETTS PSP SUPPORT GROUP

On June 3, 2000, Liz Brisson, who is a member of the Board of Directors for PSP, addressed the MA group at Debra McConnell's church in Braintree. Discussion was given to fundraising events. Some members of the group expressed a desire to investigate different events and perhaps organize a fundraiser.

On August 5, Ginny Woodbury hosted a meeting at her home in Worcester. Janice Clements, also a member of the Society's Board of Directors, spoke with the group about several different PSP issues. On August 23, Patti Ryan, (MA Support Group Leader) organized a meeting with Dr. Allan Ropper at St. Elizabeth's Hospital in Boston. Dr. Ropper spoke about PSP and accented PSP research being conducted. There were approximately 25 people who attended this meeting. Dr. Ropper conducted a question and answer session after his presentation. Dr Hayes, also a neurologist, joined the discussion. Special thanks to Thelma Guarrusso, Anna Pellegrini, and David Stanger, along with their family members, for making the journey to St. Elizabeth's.

For information on future meetings, please contact Patti Ryan at (781) 599-4431.

News From Support Groups

NORTH OHIO PSP SUPPORT GROUP

The first North Ohio Support Group met on Oct. 14, 2000. Twenty-four people representing 10 PSP families attended. The group decided to meet monthly on the 2nd Sunday from 1-3 p.m. at Southwest General Health Center, Middleburg Hts. Pat Beekman, meeting organizer, reports that her publicity efforts were successful. There was a half-page article published in the Healthcare Monitor, a weekly paper mailed to all healthcare professionals. Five attendees saw or were told of the article

A bigger turnout is expected at the next meeting as several calls are coming in about the next meeting. Pat also received calls from physical, occupational and speech therapists volunteering to come to these meetings and talk to families and work with PSP individuals with some activities they may try. The weekly local paper has agreed to do public service announcements each a month. Pat says that "I know my 'Higher Power' is doing this as the ideas just pop into my head and are getting results without a tremendous amount of effort." For more information about these meetings, please contact Pat Beekman at (440) 234-0007. Planned meeting dates for 2001: Jan. 14, Feb. 11, March 11, April 8, May 13, June 10, July 8, Aug. 12, Sept. 9, Oct 14, Nov.11 and Dec. 9.

MICHIGAN SUPPORT GROUP NEWS

Sally Smith reports there was a Michigan support group meeting on October 21, 2000. This was the group's third meeting. They met at the local Senior Center in Holland, MI. Fifteen people attended the meeting; four were diagnosed with PSP. The next meeting is scheduled for January 13, 2001. The group decided that meetings will be informal, focused on PSP and how everyone is coping. The greatest distance someone traveled to this meeting was 100 miles to support a sister who was recently diagnosed with PSP. For more information, please call Sally Smith at (616) 772-4446.

HOUSTON, TEXAS PSP SUPPORT NETWORK

The Houston PSP Support Network meets the third Saturday of each month at 1:00 pm in the Learning Center B of the Memorial Hermann Southwest Hospital Professional Building II. Call Karen Kennemer (281) 358-2282 for more information.

PSP EUROPE

We are pleased to have a cooperative relationship with our sister organization, PSP Europe Association. For information on what is happening in the United Kingdom and throughout Europe, contact:

Michael Koe
The Old Rectory
Wappenham, Nr Towcester, Northamptonshire
NN12 8SQ
Telephone 0044 (0) 1327 860299
E-Mail psp.eur@virgin.net
Website http//www.pspeur.org

DISCLAIMER

Information, reference material concerning research being done in the field of PSP and answers to reader's questions are solely for the reader. It should not be used for treatment purposes but only for discussion with the patient's physician.

NEW ORLEANS PSP SUPPORT GROUP

On Saturday, October 28, 2000 at 2:00 p.m., the New Orleans PSP Support Group sponsored a meeting in honor of PSP Awareness Month and in tribute to the Society's 10th Anniversary of Caring at the home of Brenda Gremillion. Fifteen people attended and everyone brought food and drinks. It was a time of caring, laughter and support. Some guests drove over 3 hours, others 1½ hours to attend. They watched the PSP Baltimore Symposium on video as we enjoyed dishes like jambalya, fettucinne, green bean casserole, cake and "perfect" fudge. Brenda, support group leader, has scheduled two presenters for the next New Orleans meeting - a Hospice case worker and a speech pathologist from LSUHCS. For more information, call Brenda at (504) 467-6658.

Brenda Gremillion (1st row far right) and New Orleans Support Group

PHILADELPHIA SUPPORT GROUP

Philadelphia Support Group's 10th Year Tribute to the Society

Ellen Katz, Society Director, attends the Philadelphia Meeting

News From Support Groups

CONGRATULATIONS, ST. LOUIS!!

Dec., 1999–The First St. Louis PSP Support Group Meeting
October, 2000–The First St. Louis Regional PSP Symposium

In 1995, Amy Mandlman, a St. Louis speech therapist working in nursing homes, was treating a woman who later was diagnosed with PSP. But, at the time, the family did not have any idea about PSP. The diagnosis had not been made and the family did not know what was wrong with their loved one. Finally, the diagnosis was made. The family had no understanding of the disease and did not know what to do because their doctor told them that there was no known cause or cure. The family could not find any information. Amy researched PSP on the Internet and found a toll-free number and put the family in touch with The Society for PSP, which provided them with information and support to help them cope with the disease. Four years later, this family called Amy to thank her for the 800 number, but was in dismay that there was no one to talk with about their triumphs, tribulations, ideas and feelings. This prompted Amy to contact the Society's Executive Director, Ellen Katz, in January 1999. This was the beginning of the St. Louis PSP Support Group. Amy's name was placed in the next issue of the PSP ADVOCATE officially introducing a contact person in the area. By May 1999, a core group of 15 families called to express interest and verify that there was a need for the support group in the St. Louis area. The first meeting was held in December 1999 with 20 people in attendance. Currently, the support group's attendance has grown to 31.

Some topics of discussions at meetings include: medications, how people are feeling (from a caregiver's viewpoint as well as changes the person with PSP is experiencing), doctor visits, communication, news articles, magazines and websites found to be helpful, speaker topics, and ways to increase awareness of the disease. The support group, which has only been in existence for 10 months, organized a successful regional symposium presented on Oct. 14, 2000! The Society for PSP thanks all the members of this group for their commitment in increasing the awareness of PSP. (For more information about The Symposium, see the Special Events section of this Advocate.)

The St. Louis PSP Support Group

NEWS FROM THE MINNESOTA PSP SUPPORT GROUP

By Charlotte Tripet

I have to tell you about what a very special 10th Tribute Event for the Society for PSP that we had in Minnesota. First of all, thanks to the Society for SPSP for sending out the invitations to the event. This brought about numbers of people who have not been part of our support group: folks with PSP and their families as well as other extended family members who were asked to attend the gathering by the PSP family as they were not able to attend or lived too far away to come. We had one family who drove 125 miles to come to the event and another who drove 75 miles. Altogether, we had 70 people there, including four persons with PSP .

We were fortunate to have Paul Tuite, M.D., a neurologist from the University of Minnesota Department of Neurology, who specializes in movement disorders and John Anderson, M.D, from the University of Minnesota who specializes in eye movements. They spoke to our group and answered questions from the group. This was a very positive addition to the evening, and we are grateful for their attendance. We also had members of our support group share some experiences in traveling with their PSP family members — humorous experiences and challenges and how they dealt with them. Each family group and individuals introduced themselves, which allowed others who were living in a community near them to become acquainted. Our dinner was served by the Boy Scout Troop from Mayflower Congregational Church. They were great helpers and made the dinner and cleanup go very smoothly. Expenses for the dinner were covered by contributions from Ginny Smith, for the most part, and myself. The checks enclosed are proceeds from the dinner and additional contributions given that evening. The only amount we took out was the cost of using the church, which was $75. The total of the enclosed checks should amount to $2582.

It was a good experience. We all worked hard the night of the event, but it all came together well.

WANTED: VOLUNTEERS FOR PUBLIC RELATIONS COMMITTEE

The Society for PSP is hoping to expand its Public Relations Committee. Members are sought who are available to contact TV, radio stations and newspaper offices. Most projects have to be executed in the daytime so individuals that work from home or part-time may have more time available for these projects, but no one is out- especially those with good ideas and experience! An email address from the location from which they are helping is needed, and a fax machine would be a great help. The members need to be have experience with PSP so they are better able to answer questions reporters may have. Good communication skills, good telephone manners and an up-beat personality are essential. Please contact the Society office at 1-800-475-4777 or Public Committee Chairperson, Ceri Williams, through her email address: medianet@earthlink.net

A Tribute to the Love of Our Lives, Who Never Stopped Smiling

The love of our lives, Marie Hitt from Lake Odessa, Michigan, was a very special wife, mother, grandmother, great-grandmother, and friend. Her family and friends meant everything to her. She taught us all to laugh often, smile all the time, care for others, never complain and love unconditionally. She would always say, "You play the hand you are dealt and never question why." Wise words indeed for us all to live by.

Mom's life started falling apart in 1990 with a diagnosis of breast cancer. She endured a lumpectomy, chemotherapy, radiation and the loss of her beautiful hair. She was just 62 years old then and had just retired with her husband (our father, Lloyd) and seen the first of her five grandchildren get married. Through all her suffering, her greatest concern was not for herself, but that she may have genetically passed on this disease to her daughters.

Dad nursed her through all of this. He never once complained, just took care of her. He always brought her yellow carnations, her favorite flower, and just loved her through it all. Mom recovered fully from the breast cancer, and visits to the oncologist never showed any further signs of the cancer. But then other things started happening.

A fall inside her home in October of 1991 shattered her elbow into eight pieces, requiring six different surgeries, some to the joint and some for skin grafts.

Then other strange things started happening: stumbling, choking when eating, coughing, falling into a chair rather than sitting, more falls, trouble reading (one of her greatest passions).

Visits to the doctor brought NO answers; they just chalked it up to the aging process. We kind of thought maybe it was from the chemo and radiation.

In 1998, another fall onto her already crippled elbow required reconstructive surgery with a prosthetic device. By then, the vision, choking problems, and lack of strength in her legs worsened. We finally changed doctors, and after a series of tests, a diagnosis of PSP was given in September 1999 by a neurologist at Michigan State University. It was estimated she'd had PSP for approximately eight years.

It was at this point that we came to realize how long our father had been caring for mom, never letting on things were so bad. His own health had deteriorated to such a point that my sister and I became much more involved in both their lives, trying to help all we could.

In November of 1999, mom took another bad fall, breaking six ribs. After a brief stay in the hospital, she was transferred to a nursing home for six weeks. Upon her discharge, we found them an assisted living place so she and Dad could be together. While there, mom suffered two more bad falls, resulting in head injuries and hospital stays. After two months at the assisted living facility, it was obvious Mom needed more than what they could offer, and Dad's health was declining as well. By this time, Hospice became involved and we thought with them and home health care, we could move our parents back to their home, where they both truly wanted to be. Within days it was obvious that Mom needed care 24/7 and Dad could no longer be of much assistance.

In March 2000, we decided that placement in another highly recommended nursing home was the only alternative. We promised Mom the day she was admitted that not a day would go by where she would not have a family member there. We kept that promise. We would read to her; she loved Chicken Soup for the Soul books; we'd sing to her, paint her toenails, massage her body, climb in bed with her and just hold her and kiss her. She loved to be laid next to and held and kissed and told how much we loved her. We would wheel her down into the beautiful garden, with beautiful flowers and a flowing pond with Koi fish in it. She'd always ask "How many fish are in there today?"

Marie Hitt and granddaughters

If one of us could not be there, Mom's three granddaughters would take turns visiting her, bringing with them their children. Oh, how she loved to see them. This picture of mom with her three grandchildren was taken only six days before her death, the last time she saw them.

Mom always wanted to go back home. Even when she realized it was not going to happen she didn't cry, complain, or feel sorry for herself. She merely smiled every day at everyone she encountered, and "played the hand she was dealt." She fought PSP with every fiber of her being; she never gave up. Even in July and August, when she could barely speak and had to be spoon-fed with pureed food, she always ate every bite and never lost her will and still smiled at everyone.

In August, it was discovered that the skin on mom's bad elbow was so thin it had broken open, leaving the prosthetic elbow exposed to the open air, and it had become infected. They tried three very strong antibiotics, but did not heal the infection and the infection entered her blood stream.

We were able to celebrate her 72nd birthday on August 18. It was her last outing, as it had become impossible to take her out anymore and maneuver her into a car. We were going to take her to a park with a river because she loved being by water—Lake Michigan, a stream, any place by water. But all she wanted was to go and visit Dad as he was no longer able to drive to visit her.

On September 1, Mom passed away at 4:00 a.m. We were not by her side, where we wanted to be - holding her and telling her how much we loved her, even though we did this at every visit. We never got to say goodbye, which hurts the most.

Writing about Mom's experience with PSP is a healing process for our family and maybe in some way it is our way of saying goodbye to her, a beautiful lady on the outside, but even more beautiful on the inside. She was truly loved by everyone she ever came in contact with.

"You fought the good fight and have run the race. Thank you, mom, for all the life lessons you taught us until your last breath. You were not only a loving mother to us but also our very best friend. We miss you... we love you... you are in our hearts forever."

Your loving daughters,
Cathy and Susie

Support - Our PSP Stories

Dear Nancy,

In reading the stories of PSP victims in the last newsletter, I was moved to tears by what courageous, unselfish people they are. I was also impressed by the love and support expressed by their family members. I too, had a special father who was diagnosed with PSP. In March, he fell and broke his hip, necessitating surgery. Several days later, he quietly passed away. I'd like to share some of our PSP story.

Sincerely, Debra Holland

DAD'S EULOGY

As a psychologist, I constantly hear stories about how bad people's childhood's were—the fathers who were neglectful or abusive. When I listen to these stories, I always thank God for the father I had. A father who loved me dearly, who did things with me, proudly watched my activities or sports, taught me about life.

Through his example, I learned the value of hard work, importance of family, belief in God, moral behavior, courtesy, a healthy lifestyle, the importance of investing for the future. He shared with my brother Tim and me his love of skiing, tennis, swimming, and golf. Yet, he also encouraged us to try many activities and become well-rounded. He was always a giver–never a taker. It made him happier to provide for his family than to be given to.

I remember the strong Daddy who would carry me around, lift me up to touch the ceiling or float on his chest while he swam on his back in the pool. One of our special activities was horseback riding. On weekends he'd take me riding, and we'd ride the trails into the foothills of the mountains. I think if Mom had let him, he'd have bought me a horse – the fulfillment of any young girl's dream. And just knowing he might have done that for me made me feel special.

I was lucky to have the luxury of time with my father. As a teacher, he was home early and had the summers off. I have happy memories of spending time at our beach house or traveling and camping around the United States.

As you know, my father was an intellectual man, and he passed his love of reading and learning on to me. Yet for an academic man, he never pressured me about my grades. He let me know he valued my intelligence, and at the same time, he gave me the responsibility to do what I wanted to do with it. I think I surprised him, though, with what I actually did with my life. Getting a Ph.D from USC was something he almost did, completing the course work, but never writing his dissertation. But it was only a few years ago that I even learned this information. He never burdened me with trying to fulfill his own dreams.

On his own, my father would have lived a very quiet life, but he was also (as my mother puts it) "draggable," meaning he'd go along on trips or other activities that she wanted to do and usually end up enjoying himself. When my parents and I were on a trip last September, I listened to him recount with enjoyment the many different trips they'd taken.

It has been difficult and painful to watch my dad physically and mentally deteriorate this last year. And at the same time, I've admired how he maintained his dignity and courtesy as much as was possible. Aside from a disparaging comment or two when he had trouble maneuvering out of a chair, I never heard him complain. He still did his best to be a good parent to me, hugging me and telling me he loved me.

In December, we spoke of his illness. He felt when he was ready he'd be able to leave his body, and not go into that long final deterioration. I made him promise to wait to die until after Christmas and after my 40th birthday party in February. I said he had to wait until March. And he agreed–March. True to his word, March 1st he fell and broke his hip. I knew then that this was the beginning of the end. The next day, I reminded him of our conversation and asked him if he was ready to go. He told me almost, but not quite. I was able to tell him what a good father he'd been and how I'd always known I was loved. "Very much," he said, "and, Tim too." I also let him know that I'd be okay. The next few days, Mom, Tim, and I spent a lot of time with him–time that I will always be grateful for. Sunday, I spent all day at the hospital.

The next morning, he slipped away, quietly and quickly. A peaceful release. I am lucky to have had time to prepare and have no regrets — nothing neglected, nothing left unsaid. While I am sad that he is gone, I am also grateful for his release. I will always miss his loving presence in my life, and I know he has left me with a rich legacy of loving memories.

Nancy,

You asked for personal stories. My mother suffered from PSP and passed away in late January. Here is her story:

This is a story about my mother, Florence Butts, who was diagnosed with PSP in the summer of 1999. On January 28th of this year, Mother died from pneumonia. At the end, my sister and I held her hands and told her to stop fighting, that it was time to go and be at peace. You see, she had fought so long and hard that she didn't know when to stop, but it was that constant battle to win over the disease that typified my mother and her personality.

In 1993, Mother began to notice disturbing changes in her mobility. Although she was 75 years old at the time, she had been an avid golfer for years (in fact, she made a "hole-in-one" when she was 70!) and other than some occasional arthritis flare-ups, she was in good shape. As time passed, the rest of our family faced the fact that something was not right. She was starting to have a hard time getting up from a chair or out of a car, and within a few months, she was involved in two automobile accidents, both her fault. She complained of having a "terrible time" getting started in the mornings, feeling that she had suddenly aged quickly and irreversibly.

When she was told by a neurologist in 1994 that she had Parkinson's disease, my mother refused to believe the diagnosis. Even after a second neurologist, a specialist in Parkinson's, confirmed the diagnosis a few months later, Mother shook her head and said, "They don't know what they're talking about. I DON'T have Parkinson's!" This reaction didn't surprise us at all. Not only did we think that Mother was in denial, but as a former nurse (U.S. Army Nurse Corps during World War II), she was the worst patient ever, challenging the doctor's and nurse's techniques, and SHE always knew best!

Although she started on Sinemet immediately, she got progressively worse, and she soon needed the help of a walker to move around. She developed a shuffling walk and her voice began to get weaker. Her move to an assisted care home was the first of several moves to find the "right place." She fell several times, but amazingly she never broke a bone; there were several black eyes, a swollen nose, and a cut on her head that required stitches, but each time she would be up and walking again as soon as possible.

And this perseverance was my mother's way of dealing with the frustration, the embarrassment, the pain. She never gave up, even when a neurologist at Emory Neurology Clinic in

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Support - Our PSP Stories

Continued from Page 22

Parkinson's – she had a rare disease called progressive supranuclear palsy, and there was no cure. By that time, she was having difficulties swallowing food without choking, and she was in a wheelchair most of the time. During this period, my sister, who lived closest to Mother, tried every way she could to make her more comfortable and take her mind off her problems. She and her friend, Bob, took Mother on road trips, to ball games, to the lake, on shopping trips, and out to supper. Mother loved all this; she has always had a zeal for life and nothing was going to stop her! The workers and residents at the places where she lived were amazed by her "spunk" and spirited attitude.

Oh yes, there were rough days – very rough days for us and especially her – when the depression hit hard and nothing we did was right and she was upset with everyone. But there were also those days when we laughed and laughed (my mother and I shared the same of sense of humor; we were amused by the absurd), and somehow she could even joke about some of the things that were happening to her.

Mother loved to be around people, and people were drawn to her. Those of you who have been through serious illnesses with loved ones know that there are many angels on earth who touch you and your family member during the bad times and you never forget their kindness and compassion, no matter how brief the encounter. There were so many of these for Mother: Carol, Suzanne, Sheila, Ida, Dotty, Joe, others. They gave Mother joy and comfort in constant doses, and we loved them for it too.

As much as I miss my mother, I'm grateful that her struggle has ended. My heart goes out to all families who cope day by day with PSP, as a victim or a loved one. I know that there are others out there who persevere as my mother did, holding on to life as tightly as possible, and to you I say, "May God bless and keep you."

Barbara Beardsley

Dear Friends,

I am hoping to compile a broad cross section of personal stories that encompass the family of progressive supranuclear palsy. There will be similarities throughout all the stories, yet each story will also be unique. What may help one with the disease process may also help another. Please take this opportunity to share and care. Each of our stories will be filled with courage, creativity, and perseverance.

Please send your "stories" to me via: email: NancyB501@cs.com OR 6 Bramston Drive, Hampton, VA 23666

Denny Neagle Celebrity Golf Tournament

The Society for PSP and Denny Neagle of the Colorado Rockies have teamed up to host its first golf tournament in 2001 to raise money that will support funds for the Society. We will be forming a committee to work on this event and would welcome interested readers to contact the Society's office for more details.

Please call 800-457-4777 as soon as possible since we will need to begin planning for this event as soon as possible. Don't delay on being part of this exciting event.

Helpful Hints

CHECKERS FOR PSP:

I found a useful magnetic checkerboard with magnetic checkers in the Maxi-Aids catalogue (www.maxiaids.com / phone 516-752-0521). My husband, John, was able to enjoy playing checkers with our grandchildren. The magnet playing pieces helped with the clumsy hand movements of PSP and kept the pieces from slipping around to some extent. John had quite a few good games of checkers with his grandchildren. I wish I had found this a few years sooner!

COMMUNICATION AID:

When John began having extreme difficulty making himself understood, we used a cookie sheet with edges and our grandchildren's magnetic letters to make a spelling board. John was able to select letters and move them to make the main word he was trying to get out. There are many more sophisticated devices available through the Maxiaids catalogue but this one worked pretty well. Maxiaids @ www.maxiaids.com / 516-752-0521.

BRUSHING TEETH:

I asked my dentist for a spacer that is used in the dental office to use for my loved one with PSP so I can brush his teeth better. A mouth spacer slides between one's upper and lower teeth, keeping the mouth wide open so I can do a better job cleaning his teeth and not worrying about him clamping down on the toothbrush.

My dental hygienist suggested that I invest in a good electric toothbrush and as long as my dad can, have him rinse out twice daily with antiseptic Blue Mint Listerine (or the exact ingredient generics) is the only type of mouthwash that does have an antiseptic quality.

WALKER:

Dear PSP Friends,

I have researched walkers for my wife diagnosed with PSP for 2 years. I recently purchased a US-PC walker from a company called U-Step and am very pleased with the results. The US-PC's braking system seems unique to others because the brake is set when the unit is not in use, offering greater stability and safety when getting out of a chair or turning to use the walker's seat. I was puzzled at first when I read about the brake system always being on and the user having to press lightly on the mid-brake to keep it released as the walker is in motion. But, then it occurred to me that this type of brake control may reduce falls and it did for us.

Also, in October 1999, Medicare approved reimbursement at the full rate for the cost of the U-Step. Therefore, all but the $20.00 shipping charge is reimbursable for people with PSP who have Medicare and secondary coverage. U-Step's phone number is (800) 558-7837.

R.B.

Editor's Note: This information is provided as a service. The Society for PSP DOES NOT ENDORSE these or other products and neither the Society or its staff have financial interests in these products. Information is offered with the intention to inform others about products that may help to maintain independence and improve the quality of life. Please check with your physician. If you have any hints, medical devices, equipment or services you can recommend to other PSP families, please contact the Editor.

Report Of Gifts - August 1 thru October 31, 2000

The Society for PSP extends its thanks to our donors who have given so generously to help find the cure for PSP while helping families meet its difficult challenges.

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The Society for PSP is a non-profit 501-3(C) organization that exists to promote and fund research into finding the cause and cure for PSP. The Society is exempt from income taxes in the United States. We are able to sell donated securities and use every dollar to enhance our research funding, services for people with PSP and their families, educational programs, and advocacy for all affected by PSP

Transferring listed securities to the Society for PSP is quite easy. You simply have to mail the stock certificate or bond to our office. In a separate envelope, you must mail us the executed stock or bond power. Your generous gift will be valued on the postmarked date for tax purposes. If the stock or bond is with a brokerage account, inform the broker that you want your securities on an account for the Society for PSP. The broker must then call our office and the securities will be transferred by him/her. This usually occurs the same day that you instruct the broker to make the transfer.

Please consider a gift of securities. Each dollar will help people with PSP. We are the largest network of individuals dedicated to helping find the cause and cure for PSP and improving the quality of life for persons with PSP. Please join us in this challenge! Contact our office at 1-(8O0)-457-4777. Thank you friends of the Society and "ADVOCATES" from those suffering through the hardships of progressive supranuclear palsy.

Help With "Helpful Hints" Needed

Dear Friends,

This editor needs your help! In addition to the "Helpful Hints" featured in the PSP Advocate, I would like to put together a pamphlet of "Helpful Hints" for future distribution. Persons with PSP, their families and caregivers are very creative and adaptive when it comes to meeting the challenges of PSP. Several of these challenges are listed below. Please take the time to share your ideas, advice, possible solutions and accommodations with other families affected by PSP. Send your ideas to me on a separate sheet of paper and mail it to Nancy Brittingham, 6 Bramston Drive, Hampton, VA 23666, Fax (757) 838-6086 or email NancyB501@cs.com. Many times families impacted by PSP feel isolated and helpless. This is a positive opportunity to assist others and make a difference! I look forward to hearing from all of you and will put together all of your ideas so that you can share them with one another.

What advice of helpful hint do you have for:

- Anxiety
- Activities (especially in the later stages of PSP)
- Bathing
- Bladder Control
- Coughing
- Choking
- Depression
- Doctor's Questions After Visits
- Dry Mouth
- Eating
- Excessive Saliva
- Eye Moisture
- Gait/Balance/Falling
- Head Posture
- Impulsivity
- Spilling Food
- Toileting
- Tone/lack of
- Vision
- Wheelchair adaptations
- Other

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The Society for PSP, Woodholme Medical Building, Suite 515, 1838 Greene Tree Road, Baltimore, MD 21208
1 (800) 457-4777 1 (410) 486-3330
PLEASE MAKE ALL CHECKS/GIFTS TO "THE SOCIETY FOR PSP."

Send me copies of:

- ❏ #1 PSP Some Answers (Overall guide To PSP)
- ❏ #4 PSP ADVOCATE-Newsletter
- ❏ #6 Swallowing Problems
- ❏ #7 Personality Changes
- ❏ #8 Helping the Helpers Who Care for People with PSP
- ❏ #9 Eye Movement Problems with PSP
- ❏ #10 1999 National Symposium Video Tapes-3 pack $75 plus $7 shipping in US and $10 outside.
- ❏ #11 PSP Fact Sheet (1 page summary-can be duplicated and distributed)
- ❏ #12 Reprint of feature article, Baltimore Sun
- ❏ #13 Medical Professional's Journal Review/PSP Advocate
- ❏ #14 Brain Bank Information Packet
- ❏ #15 Physician's Referral Cards
- ❏ #16 Giving Envelopes
- ❏ #17 The Society for PSP/National Institutes of Health PSP Brainstorming Conference/Dr. John Steele meeting with the Maryland Support Group $25 + $3 shipping in US and $5 outside.
- ❏ #18 Beautiful Acknowledgment Card to someone special for any occasion and will personalize your message. By donation only.
- ❏ #19 Planned Giving Information
- ❏ #20 Information About PSP translated in Spanish
- ❏ #21 I Have Been Diagnosed With PSP (NEW)
- ❏ #22 Challenges in the Management of PSP (NEW)

Mail to: _____

_____

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Fax to : _____ Email to: _____

FOR PHYSICIAN'S ONLY:

- ❏ #2 Video "The Diagnosis of PSP" (Recommended for clinicians and faculty) $30 + $4 shipping in US and $7 outside
- ❏ Medical Professional Packet (Grant Award Information/PSP Rating Scale/copies of all other info.)

❏ I no longer wish to receive the PSP Advocate and by sending this will save expenses for the Society.

❏ My new address is: _____

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❏ Yes, I wish to be included on The Society for PSP's mailing list:

Name _____

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❏ Person w/PSP ❏ Family ❏ Physician ❏ Other _____

Enclosed, please find my gift to help support The Society for PSP and those impacted by PSP.

❏ $25 ❏ $50 ❏ $100 ❏ $250 ❏ $500 ❏ $500-$1000

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Thank you for your TAX-DEDUCTIBLE gift. A copy of financial statement available upon request.

You Can Help By Becoming A Volunteer

VOLUNTEER PROFILE

Name: _____ Connection to PSP _____
(Please include professional designations; i.e., MD, PhD, etc.)

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- ❏ Lead a Support Group
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THE SOCIETY FOR PSP
Progressive Supranuclear Palsy

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THE PSP ADVOCATE
Fourth Quarter, 2000.
PULL OUT MAILER FOR PHYSICIANS

Medical Professionals' Special Edition

To be filled out by PSP ADVOCATE reader.

Dear _____ ,

I am forwarding this information to you for your review. Thank you for taking the time to read it.

_____ _____
Signature of Person with PSP/caregiver/other Date

Research in PSP is expanding. These are summaries of some of the journal articles appearing over the past year that would probably be of most interest to physicians treating patients with PSP. Your neurologist/physician may appreciate your sending him/her this pull out section. There is space provided on the last page of this handout for address labeling.

Attention: Physicians and other Healthcare Professionals

The Society for Progressive Supranuclear Palsy is pleased to provide you the latest journal articles relating to PSP. The Society, through the support of its Medical Advisory Board, hopes you will utilize this information to expand your knowledge of PSP and share it with other health professionals. The Society also provides information about programs, services, professional publications, quarterly newsletters, and grant opportunities. Please fill in the enclosed form to receive this free information and to be placed on the Society's mailing list.

GRANT ANNOUNCEMENT:

The Society for PSP announces The Annual Eloise H. Troxel Memorial Fellowship, each of $50,000 for research in PSP.
Deadline, October 1.
Regular Grant Program to $50,000 - Deadlines are April 1 and October 1.
For information contact: Dr. Lawrence I. Golbe, Chairman of the Medical Advisory Board at golbe@umdnj.edu or Fax 732-235-7041.

Progressive Supranuclear Palsy Meets the Molecular Age

Published in Lancet September 9, 2000
(Lancet 356:870-871, 2000)

By Lawrence I. Golbe, MD
Professor and Acting Chair
Department of Neurology, UMDNJ-Robert Wood Johnson Medical School
New Brunswick, NJ USA 08901

Progressive supranuclear palsy (PSP) may be emerging from its state of scientific inscrutability. The clinical picture of PSP, still nearly untreatable, features early, prominent postural instability, supranuclear gaze abnormalities most severe in the vertical plane, predominantly frontal cognitive disturbance, erect or even opisthotonic posture, spastic or ataxic dysarthria, and dysphagia that is frequently fatal. Its prevalence of 5-6 cases per 100,000[1] is 3-5 percent that of Parkinson's disease (PD). The symptoms of PSP begin in the 60's in most cases, but PSP does not include the important younger-onset or familial components seen in PD. Median survival after symptom onset is only 6-9 years,[2-3] similar to that of PD before the levodopa era.

The variety of nuclei involved by PSP may account for its resistance to dopaminergic and other neurotransmitter therapy. Unlike PD, where the dopaminergic substantia nigra pars compacta is the principal affected area of basal ganglia, PSP additionally involves striatum, pallidum, subthalamic nucleus and thalamus. Like Alzheimer's disease (AD), PSP includes important degeneration of the cholinergic nucleus basalis of Meynert, which projects widely to frontal cortex. However, attempts to mimic the modest success of acetylcholinesterase inhibitors against the cognitive symptoms of AD have failed in PSP.[4] PSP also involves a neurochemically heterogeneous array of nuclei in the cerebral cortex, brainstem, cerebellum and spinal cord as well as important astrocytic glial pathology. Amitriptyline and amantadine, each with multiple pharmacologic actions, have been shown to benefit a few patients with PSP modestly and deserve a trial in most cases. More effective therapy may lie in trophic factors or neuroimmunophilin therapy presently in development.

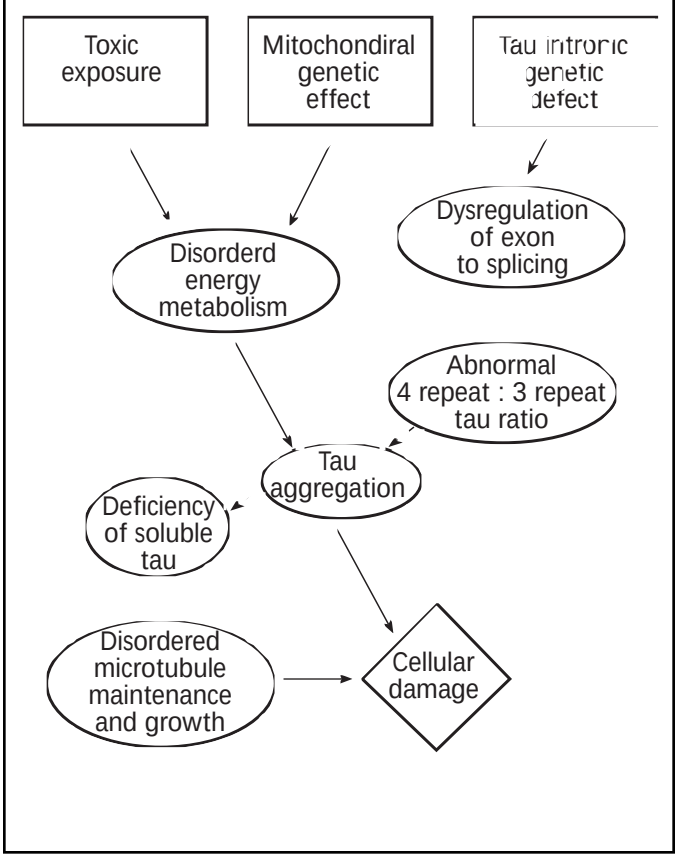
Like AD and a dozen other conditions, PSP features neurofibrillary tangles (NFTs) in degenerating neurons. NFTs are abnormal aggregates of tau, a protein that stabilizes microtubules.[5] In normal human brain, tau occurs in approximately equal proportions of two isoforms - with either three or four repeats of the microtubule-binding peptide domain.[6] The isoform is determined by whether the transcript of the *tau* gene's exon 10 is spliced in or out of the final tau protein product. In PSP, the ratio is at least 3:1 in favor of 4-repeat tau.[7] In some other "tauopathies" such as Pick's disease, 3-repeat tau predominates. Disordered regulation of exon 10 splicing may therefore explain NFT formation in PSP and other tauopathies.

A valuable clue to this sporadically occurring tauopathy arises from a dominantly inherited tauopathy, frontotemporal dementia (FTD). Here, the 3-repeat:4-repeat skew is similar to that of PSP.[8] Several mutations in and near the 5' splice site downstream of exon 10 have been described in FTD families.[9] The pathogenic mechanism of these mutations is probably disruption of a stem-loop structure in the RNA transcribed at the downstream end of exon 10. This stem-loop regulates splicing of the exon 10 transcript. Its dysfunction in FTD produces predominantly four-repeat tau. The cause then, of PSP and other sporadic tauopathies may be dysfunction of the same RNA stem-loop, but of nongenetic or nonmendelian genetic cause. This prospect has been strengthened by the recent description[10] of an Australian family with inherited, autopsy-typical PSP and a mutation at the downstream end of exon 10 close to those described in FTD. This single nucleotide substitution does not alter the tau amino acid sequence but would disrupt the RNA stem loop.

Following the lead of the FTD story, Conrad and colleagues[11] found a modest association between sporadic PSP and a genetic marker, the A0 allele, located in the intron upstream of exon 10. Subsequently, two haplotypes consisting of markers spanning most of the *tau* gene have been associated with PSP more strongly than the A0 allele alone.[12-13] Each haplotype is present in nearly all patients with PSP but also in a significant minority of controls. This suggests that a genetic variation necessary but not sufficient to cause PSP is located in or near the *tau* gene at chromosome 17q21. The occurrence of PSP in sporadic rather than familial fashion must then require an additional exogenous or genetic factor.

A case-control study in New Jersey found PSP to be associated with lesser educational attainment, but that study's inquiry into industrial or other known neurotoxins was negative.[14] Two geographic clusters of PSP-like tauopathies are known. Lytigo-bodig, or the amyotrophic lateral sclerosis-parkinsonism-dementia complex of Guam, has resisted multiple careful etiologic investigations since the 1950's. But an unusual concentration of a PSP-like tauopathy on the Caribbean island of Guadeloupe was described in 1999 by Caparros-Lefebvre, et al.[15] Their case-control survey found that illness to be associated with dietary or medicinal use of two indigenous plants, soursop and sweetsop. These species contain reticuline and corexime, which are dopaminergic toxins.[16] Discontinuing the use of these products resulted in marked and prolonged clinical improvement in some of the younger patients on Guadeloupe.[15]

Possible pathogenetic mechanisms in PSP



Evidence that deficiency of Complex I of mitochondrial genetic origin contributes to the cause of typical PSP is provided by recent observation of low Complex I activity in cultured neuronal cells in which native mitochondria were replaced by mitochondria from patients with PSP.[17] The possibility then arises that tau aggregation in PSP results from the combination of at least two factors: a mitochondrial defect of genetic or toxic origin producing an intracellular chemical environment conducive to tau aggregation; and a genetic defect in the *tau* gene causing overproduction of 4-repeat tau.

The next step is to understand how the function of the RNA stem-loop can be disrupted by toxic insults or by genetic defects. All of the exons and nearby intronic regions of tau have been sequenced in PSP with no consistent abnormality found. [G. Schellenberg; J. Hardy, pers comm] Therefore, the *tau* genetic markers known to be associated

with PSP are probably linked to a defect deep within an intron, where the normal sequence is unknown but where regulatory activity may lie. The task of sequencing the introns of tau will require years and new technology. But a realistic, short-term approach to preventive therapy for PSP may emerge from studying dietary or occupational toxin exposure, mitochondrial genetic defects and splicing regulation by RNA stem-loops.

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1. Mitochondrial dysfunction in cybrid lines expressing mitochondrial genes from patients with progressive supranuclear palsy
   RH Swerdlow, LI Golbe, JK Parks, DS Cassarino, DR Binder, AE Grawey, I Litvan, JP Bennett Jr, GF Wooten, WD Parker
   J Neurochemistry 2000;75:1681-1684

Mitochondria from PSP patients have reduced complex I activity, and host cells have increased markers of oxidative stress, according to this study.

Mitochondria from PSP patients were introduced into normal cells in culture. Resultant cybrid (cytoplasmic hybrid) cells showed a 12.4% decrease in complex I activity ($P<0.005$), but no change in complex IV activity. Cybrid cells also had significantly increased levels of several antioxidant enzymes. The authors note that since all the cybrid lines possess an identical nuclear background, "the origin of mtDNA should represent the only meaningful variable between cybrid lines." They suggest "increases in antioxidant enzyme activities indicate that oxidative stress is likely arising as a result of an mtDNA-encoded electron transport chain enzyme lesion."

2. Defects of brain and skeletal muscle bioenergetics in progressive supranuclear palsy shown in vivo by phosphorus magnetic resonance spectroscopy
   P Martinelli, C Scaglione, R Lodi, S Iotti, B Barbiroli
   Movement Disorders 2000;15:889-893

Five PSP patients underwent phosphorus MR spectroscopy of the brain and calf muscle. Compared to controls, patients showed significantly reduced levels of phosphocreatine and $Mg2+$, and increased levels of ADP and inorganic phosphate (means for each value differed from controls by 20% or more; $p<0.008$ or better for each). In calf muscle, $Mg2+$ and inorganic phosphate differed significantly between patients and controls, while phosphocreatine did not. The authors conclude, "Our present study shows a remarkable deficit of mitochondria oxidative metabolism in both brain and skeletal muscle in five patients with probable PSP, and supports the hypothesis that defective oxidative phosphorylation plays a role in the pathogenesis of brain neuronal loss in PSP."

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THE SOCIETY FOR PSP
Progressive Supranuclear Palsy
Woodholme
Medical Building
Suite 515
1838 Greene Tree Rd.
Baltimore, MD 21208

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