



Society for Progressive Supranuclear Palsy

PSP Advocate

Official Newsletter of the Society for Progressive Supranuclear Palsy, Inc. (SPSP)
David and Reba Saks, Founders

**FIRST INTERNATIONAL
BRAINSTORMING CONFERENCE
ON
PROGRESSIVE SUPRANUCLEAR PALSY
NATIONAL INSTITUTES OF HEALTH
MARCH 18-19, 1999**

The Society for PSP and the Office of Rare Diseases at the National Institutes of Health are sponsoring a Brainstorming Conference on "progressive supranuclear palsy" on March 18-19, 1999 under the Chairmanship of Dr. Irene Litvan, Chief of the Neuropharmacology Unit, Defense and Veteran Head Injury Program, Henry M. Jackson Foundation. The Brainstorming Conference will be held on the NIH campus in Bethesda, Maryland. The purpose of this conference is to identify data gaps and future research needed to understand the cause of neurodegeneration (why brain cells die) in this and related disorders. Search for gene-environment interactions (identify predisposing genetic and environmental factors for the development of this disorder), and critical issues regarding the development of experimental animal models, biomarkers for the diagnosis of these disorders as well as potential useful treatments to slow down the course of this disorder will be discussed. World-known international researchers in basic and clinical research in progressive supranuclear palsy and related neurodegenerative disorders will submit the results of their latest studies at this meeting. This will be a unique opportunity for cross-fertilization, design of new studies and new collaborative efforts. The results of this effort will be submitted for peer-review publication.

The overall plan is to have leading scientists identify those issues that could advance research in PSP and summarize what has been accomplished in these areas. Each presenter will bring a written document that will be the basis for discussion which will be given ahead of time so each panelist can prepare the discussion. After each presentation, a panel will discuss the presentations, followed by an overall group discussion. After finishing the presentations, the participants will be divided into smaller groups to write the summary of the discussions in each field paying particular emphasis to specific scientific issues identified, and to design future studies. The major points of the

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**PSP RESEARCH AT THE
FIFTH INTERNATIONAL CONGRESS
ON PARKINSON'S DISEASE AND
MOVEMENT DISORDERS
SPONSORED BY
THE MOVEMENT DISORDERS SOCIETY
OCTOBER 10-14, 1998**

The Movement Disorder Society holds an international Research Congress in alternate years. This year it was held in New York City. This year's congress included 1,141 presentations of original research, of which 17 were specifically on PSP and many others pertained indirectly to PSP. This summary and interpretation of the most important of these was compiled by Lawrence I. Golbe, MD of UMDNJ-Robert Wood Johnson Medical School, Chairman of the SPSP Medical Advisory Board:

Abnormal movement

It is known that patients with PSP do not have the normal startle response to sudden noises, whereas those with PD have a normal or slightly delayed response. Dr. P. Hollosi and colleagues from Innsbruck, Austria sought to determine whether startle testing could be used to separate PSP from multiple system atrophy (MSA) and dementia with Lewy bodies (DLB), two other degenerative disorders of the brainstem that, like PD, can be difficult to distinguish from PSP. They found that startle responses were absent in 90 of patients with PSP, but in only 13 of those with DLB and 43 of those with MSA. Startle testing may therefore be useful in differentiating among these conditions, although like any laboratory test, must be combined with data from many sources in making a final diagnostic decision.

Another movement response that can help distinguish PSP from healthy people of the same age is learning of certain automatic response, called motor conditioning. Dr. M. Sommer and colleagues from the National Institutes of Health in Bethesda, MD, found defects in this type of learning ability in patients with PSP. The severity of the defects did not correlate with PSP symptom duration, suggesting that very early PSP that may be difficult to distinguish from normal by other means

Continued Page 18

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*In 1963, Dr. J. C. Steele
 together with
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 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 PSI, their families, and caregivers. SPSP educates physicians and allied health professionals on PSP and how to improve patient care. The Chairman is Dr. Stephen Reich, Associate Professor of Neurology, at the Johns Hopkins Medical Institutions. The Society, Director, and staff are under the administration of the Johns Hopkins School of Medicine, Department of Neurology.

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(In memory of Henry and Jane Ogiba)

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documents might be detectable using this sort of test. The next step is to see if PSP differs from PD or other degenerative disorders in measures of motor conditioning.

Radiology

Dr. A. Tan and colleagues from Singapore attempted to distinguish between PSP, PD and healthy volunteers using magnetic resonance spectroscopy (MRS). This technique, which can be performed using an ordinary MRI machine with special software, has been found by other researchers to distinguish between PSP and normals and between PD and normals. The relevant abnormality is a change in the chemical composition of the lentiform nuclei, part of the brain that degenerates in PSP but not in PD. However, these researchers were unable to find a difference among those three groups' MRS images. Therefore, more work will be needed before the role of MRS in routine clinical diagnosis of PSP is clarified.

On the other hand, a different MRI technique, volumetry, was found by Dr. J.B. Schulz and colleagues from Tübingen, Germany to distinguish PSP from PD and from normals. This technique calculates the volume of certain brain areas that shrink to different degrees in various degenerative movement disorders. Unfortunately, multiple system atrophy of the parkinsonian type ("striatonigral degeneration"), which can be difficult to distinguish from PSP in the clinic, did not differ from PSP in this study.

Neurochemistry

Magnetic resonance spectroscopy (MRS) of a type different from that in the above study was used by Dr. N.C. Reynolds, JR and colleagues from Milwaukee to seek evidence for abnormal glutamate excitotoxicity or mitochondrial energy production. These two defects have been found in other neurodegenerative disorders. Neither occurred in the six PSP patients studied, although one or both occurred in the patients with Huntington's disease or idiopathic dystonia, the only other diseases in this study. This helps rule out those two mechanisms of brain cell degeneration in PSP.

Behavior and Intellect

There have been many studies of the mental decline in PSP, but until now none of these has studied the rate of decline by re-testing patients after time has passed. Dr. D. Monza and colleagues of Milan, Italy did just that, using a two-year interval, in 11 patients with PSP, 12 with PD and 14 with multiple system atrophy of the parkinsonian type. They found that the PSP patients deteriorated faster than the other two in two of the six tasks tested: paying attention to a task and performing hand gestures (apraxia). The other two groups did not decline at all, a somewhat surprising result. This study points out the importance of inattention and apraxia in PSP, two areas that have perhaps received less attention from researchers than they deserve.

The same Milanese group this time headed by Dr. P. Soliveri, compared the apraxia of PSP with that of corticobasal degeneration (CBD), a condition in which apraxia is one of the most important features. They found a slightly different profile of features, with the apraxia of PSP tending to reflect brain cell

Continued next column

loss in the frontal lobes and that of CBD reflecting loss in both frontal lobes, where movement is controlled and in other parts of the brain as well where sensory functions are served. This insight could be useful in rehabilitation planning. It has long been known from studies of autopsied brain that the levels of acetylcholine, an important chemical used as a form of communication between certain brain cells, was reduced in certain areas in PSP. Dr. S. Pappata and colleagues of Paris used positron emission tomography PET imaging to confirm this in living patients. They used a newly developed radioactive chemical, 11-C-physostigmine, which is taken up in brain areas where acetylcholine works. The PET machine creates an image of where the radioactivity ends up. This technique may allow the acetylcholine defect in PSP to be studied in living patients and could permit the effects of new treatments to be assessed in an objective way.

Neuropathology

The abnormality in the protein called tau, which makes up the neurofibrillary tangles of PSP and Alzheimer's disease and seems to lay at the root of PSP, can be studied in more detail in autopsied brain tissue as a result of a discovery presented by Dr. H. Mori and colleagues of Tokyo. It is standard practice to identify tau or other proteins in brain tissue by treating thin slices of the tissue with antibodies directed specifically against the protein being studied. This study found that an antibody directed against a specific abnormal component of the tau protein was more sensitive in detecting early-stage neurofibrillary tangles than the antibody routinely used. It could allow the location of early damage in the brain to be detected, thereby improving our ability to understand the pattern of damage produced by PSP.

Genetics

The fourth and fifth studies finding an association between PSP and a variant of the gene coding for tau protein were presented by Dr. D.J. Nicholl and colleagues of Birmingham, UK and Rome, Italy and by Dr. Morris and colleagues of London, UK. The variant, called AO ("A-Zero"), comprised 91-93 of all tau genes (two genes per person - one from each parent) in those with PSP but only 73-76 of the tau genes in healthy individuals. The chance of this occurring by chance was only 7 in 1000. This difference did not occur in patients with multiple system atrophy. Obviously, the correlation in PSP is not perfect, so either there are other abnormalities necessary to the development of PSP or this genetic variant does not itself contribute to the cause of PSP, but is only linked to i.e., some distance along the chromosome from an undiscovered causative gene. It is interesting that a defect in the same part of the same 17th chromosome is the cause of a highly hereditary neurodegenerative disease called "chromosome 17 dementia." Like PSP, this illness features neurofibrillary tangles with abnormal tau protein. The exact genetic defect underlying chromosome 17 dementia has not yet been identified, but when it is, it will shed light on the cause of PSP as well.

The idea that a simple genetic defect is capable of producing PSP was supported by the report of a family with three members with PSP. Dr. J.A. Van Gerpen and colleagues of the Mayo Clinic in Rochester, MN and Jacksonville, FL report a

Continued page 18

The Director's Doings . . .



Ellen Pam Katz
Director, SPSP Inc.

I am looking from the window of American Airlines, Flight 586, Anaheim CA to Dallas TX and am wondering about the remote towns I see throughout the Southwest desert. How many people in those isolated towns have PSP? How can they access support, appropriate medical services, and information? I have just attended two symposiums in Anaheim and Sunnyvale, CA, expertly coordinated by Carol Marchi and Pearllann Tabor of the

Parkinson's Institute in Sunnyvale. How fortunate those attendees were, as for the most part, they lived near a large, urban center where many other families lived. The breadth and diversity of our country require additional ways to inform, advocate and support for the needs of persons with PSP and their families.

The Board of Directors is approaching these challenges in a well-thought out plan. First, we have increased the number of directors from 10 to 15 and included persons with finely tuned skills to help the Society move to new levels. We

have engaged the services of Sue Dagurt, a certified fund raising executive, who has had a myriad of professional experience in helping with board development, fund and "friend-raising" and most importantly to help the Board of Directors begin the "Strategic Planning Process."

With the help of generous contributions, the ability to expand our donor base, and the increased office space and technology, we have reached a point from where we can make great strides forward. In January, the Board will begin that process with a retreat that will identify broad strategic issues and how they will be addressed.

This is a very exciting time for the Society and I am honored that I can be part of it.

As director, I am asking that you help nurture this vital organization. Please tell other people your story and ask them to contribute, ask a local newspaper to write a story about PSP and how it has affected you, contact your employers for matching gifts, or start a support group. YOU are the Society's most vital asset—and it is YOU who can do the most to ensure it can accomplish its mission.

Thanks for your support.

Best regards,

Ellen Katz

Director



Yes! I wish to be included on the Society for PSP's mailing list.

Name _____

Address _____

City _____ State _____ Zip _____ Country _____

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E-mail _____

Person with PSP _____ Family _____ Physician _____ Other _____

Name of Family member with PSP _____

Enclosed, please find my contribution to help support the mission of the Society.

\$25 Supporter _____ \$50 Sponsor _____ \$100 Patron _____ \$250 Silver Patron _____

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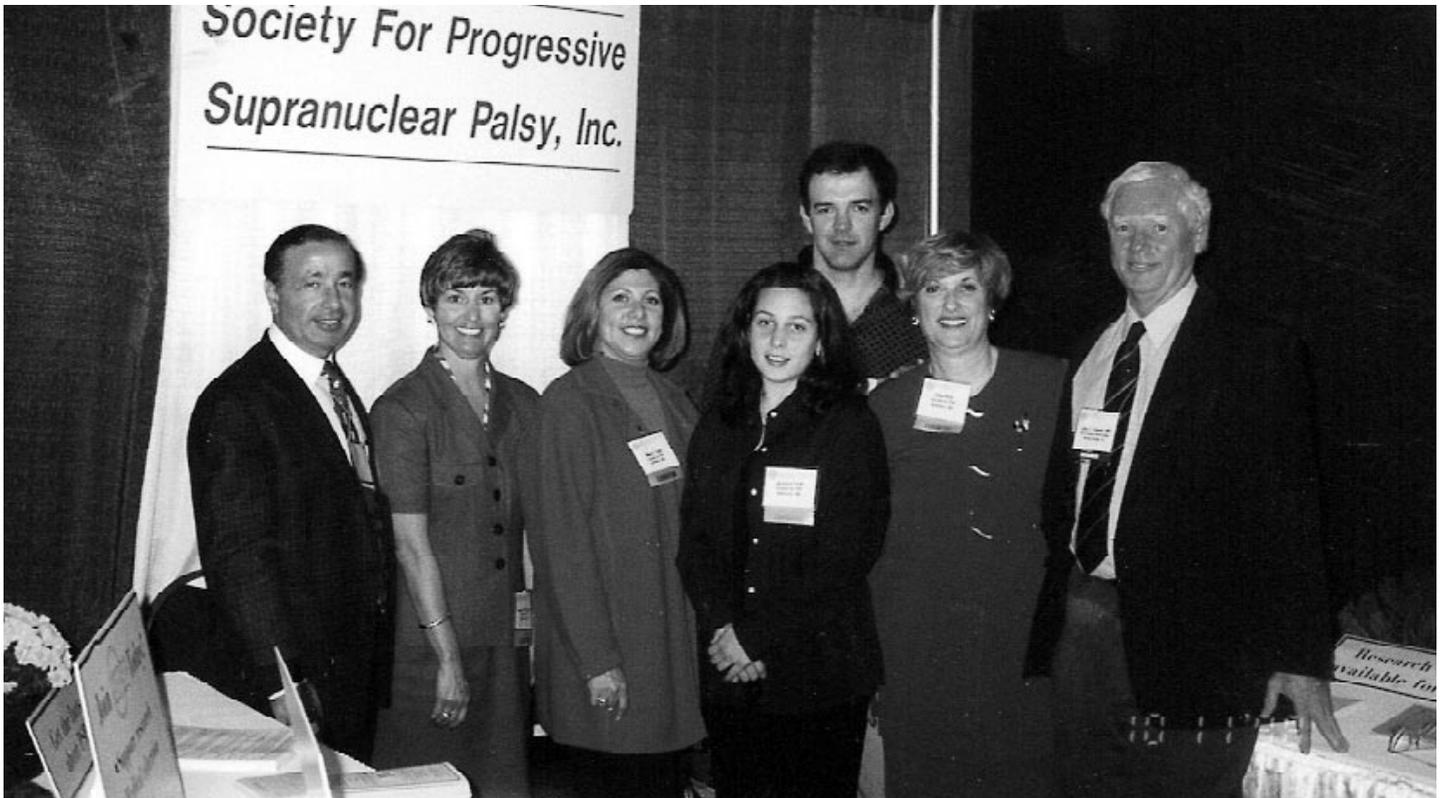
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Mail to The Society for PSP, Woodholme Medical Building, Suite 515, 1838 Greene Tree Road, Baltimore, MD 21208

Thank you for your TAX-DEDUCTIBLE contribution!

A copy of the Society's financial statement is available upon request.

International Congress of Parkinson Disease and Movement Disorders



Fifth International Congress of Parkinson's Disease and Movement Disorders in NYC.

Dr. Joseph Jankovic (Professor, Neurology, Baylor College of Medicine) Nancy Brittingham, Marcy Todd, Amy Todd, Larry Classon, Ellen Katz, and Dr. John Steele (Honorary Chairman, SPSP, Fellow, American Collage of Physicians, Core Director Micronesian Health Studies)

I am writing this in my hotel room in exciting New York City while attending the Movement Disorder Society's 5th International Congress of Parkinson's Disease and Movement Disorders. Ellen Katz (SPSP's dynamic Director) and I along with a host of wonderful PSP volunteers have spent the past few days distributing information about PSP and the Society to attending neurologists from all over the world. Many are the world's top experts in their field. This Congress is a five day meeting in which there are educational sessions, poster sessions, exhibits and breakfast seminars throughout the week. Selected topics are current and "hot" and selected speakers are actively engaged in the forefront of these scientific and medical advances. I have watched all the activity and I am in awe.

As we distributed educational pamphlets and newsletters to hundreds of neurologists today, I became so excited because our goal of raising the awareness level of PSP was happening!! Because it was a movement disorders symposium-we had a target audience! Many of the neurologists were specialists working in movement disorders clinics throughout the world. Several told me they had 15 to 20 people with PSP currently being managed through their clinics. They were so grateful to have educational materials to present to their patients. They were also very appreciative to learn about the Society. Now their patients with

PSP and families can seek support and services. I learned of two separate cases where each neurologist diagnosed a parent with PSP. Both specialists told me the diagnosis of PSP was not apparent to them for quite some time!!

It is a privilege for me to be at this meeting. This has been tremendously rewarding experience for for me. As I stood at our exhibit today, I realized more than ever that none of this would have been possible without your support. Through your contributions of time, talent and financial gifts, we have been able to greatly increase our efforts in finding the cause and the cure as well as developing better ways to manage and treat the disease. what better way is there to honor all the victims PSP! Thank you for letting me be a part of this most meaningful journey.

Nancy Ogiba Brittingham
Editor

Disclaimer

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

Your PSP Research Dollars At Work!!
\$209,000 Awarded
Part II

This has been a promising year for PSP research. The research grant proposals submitted to the SPSP's Medical Advisory Board were of particularly high caliber according to Medical Advisory Board Chairman Lawrence I. Golbe, MD. The Society is pleased to announce that nine PSP research grants have been awarded for 1998. Summaries of some of the research proposals awarded are included in this newsletter. The other proposals appeared in the PSP ADVOCATE, Third Quarterly, 1998, Vol. 9, No. 3.

**Molecular Studies of the Tau Gene In
PSP**

William G. Johnson, M.D.

University of Medicine and Dentistry of New Jersey-
Robert Wood Johnson Medical School

Progressive supranuclear palsy, PSP, is one of several disorders characterized by abnormal structures in brain cells, that contain tau, a protein that forms part of the cell structure in nerve cells and other cells. In some disorders, the tau abnormalities are clearly a secondary result of some other cause. However, recently some evidence has appeared, confirmed in our laboratory, that the primary cause of PSP may be a mutation in the tau gene or a gene located very close to it on chromosome 17.

We would like to study the tau gene to see if we can find such a mutation. This could greatly improve diagnosis and open up avenues for treatment. However, tau is a very large gene. Sequencing the whole gene would be a very large task. Also, some DNA variation always occurs from person to person and it would be difficult to tell if such a DNA sequence change in PSP were the causative mutation or just a normal variant.

Therefore, we propose studying the messenger RNA that the tau gene produces to make the tau protein. This study is much less time consuming. If the message has a different size, then the tau abnormality may be due to alternative splicing of the message. The tau gene is known to have extensive alternative splicing and the single tau gene normally produces at least six forms of the tau protein. If the tau message has a normal size but is increased or decreased in quantity, then a mutation in a regulatory part of the gene is implicated. If the tau message has a normal size and normal quantity, then a point mutation is implicated in the part of the message that codes for amino acids in the tau protein. Of course, these are guidelines not absolutes, but this approach can often save a lot of time and resources in locating a mutation.

**Mutational Analysis of the tau gene
in PSP.**

Joseph J. Higgins M.D.

Wadsworth Center for Research
NY State Dept. Of Health

Aberrant forms of a protein called tau are present in the brains of individuals with several neurodegenerative diseases including progressive supranuclear palsy (PSP), Alzheimer's disease, Pick's disease and corticobasal degeneration. In our investigations to find a genetic cause for PSP, we demonstrated that a variation in the tau gene was common in a group of unrelated individuals affected by PSP. These results suggested that mutations in the tau gene, or in a gene in close proximity, were responsible for causing the disease. Recently, a missense mutation in exon 13 of the tau gene was found in a family with a clinical phenotype that resembled Alzheimer's disease but with neuropathologic similarities to PSP. This finding lends credence to our proposed hypothesis that tau gene mutations may play a direct role in the pathogenesis of PSP. Although the specific mutation that was described in the family with atypical PSP was not found in our group of unrelated individuals, other mutations in the tau gene may be responsible for sporadic forms of PSP. The goal of the research proposed in this application is to conclusively identify mutations in the tau gene in patients with sporadic PSP by molecular analysis. The results of this research may have important implications in facilitating our basic understanding of how tau causes neuronal degeneration. Efforts to identify the genetic mutations in PSP and other related disorders with tau pathology are likely to refine diagnosis and target new treatment modalities.

SOCIETY RECEIVES BEQUEST OF \$135,000

The Society for PSP was the recipient of a bequest from the estate of Frank W. Weymouth. Mr. Weymouth, a victim of PSP, lived in Los Altos, CA. He was a talented artist who painted florals and seascapes while maintaining a florist business. Mr. Weymouth, who had no children, divided his estate among many charities he supported in his community. This generous gift will be well utilized to further the mission of the Society.

A Controlled, Randomized, Double-blind Trial of Donepezil In Patients With Progressive Supranuclear Palsy

Irene Litvan, M.D.
National Institutes of Health

We plan to use Donepezil (Aricept) to treat patients with PSP. PSP is a neurological disorder characterized by personality changes (e.g., severe apathy), problems with eye movements, speech, and walking. These problems occur because PSP impairs specific groups of brain cells cholinergic and dopaminergic neurons that use chemical substances such as acetylcholine and dopamine to communicate. Previous studies showed that Donepezil, a cholinergic drug, can stimulate the cholinergic brain cells to work more effectively. People with PSP will receive this drug in an attempt to improve the function of the cholinergic brain cells and, therefore, possibly improve mood and social behavior, memory, and thinking ability. Since neurons that regulate balance use acetylcholine, it is possible that balance may also improve. We will also test motor and eye disturbances to evaluate possible changes.

Study Plan: Following initial evaluations, patients will take either an oral dose of Donepezil or placebo (a sugar pill that looks like Donepezil) during the first month and a half of treatment. During the final month and a half of treatment, PSP patients will receive the drug they did not take before. In between the two treatments periods, there will be a month during which they will not receive any medications. Neither the patients nor their testers will know which drug patients are receiving.

PSP patients will be tested at the NIH Clinical Center four times, before and after the first month and a half of treatment and before and after the second treatment during this four-month study. Testing will include a medical evaluation, routine blood studies, and a series of personality and psychological tests. In general, these tests will ask questions about the patients' personality, the patients ability to think or remember (for example, they may be asked to remember a group of words), or will require them to perform simple motor tasks, such as tapping one key of a keyboard as fast as they can. Testing will ordinarily be performed in one or two days. The study will be conducted primarily on an outpatient basis.

Why Don't You Listen To Me?

Dear Editor,

I am a forty nine year old woman with PSP. As I am sure you can imagine, my life has changed radically over the past few years. I have found, however, that using my computer has helped me tremendously. It is a new way of being able to communicate with my family since my speech has become so impaired.

Last week, while being frustrated, I came to my computer to let it all out. The results I have enclosed to you at the request of my family. They all said that after reading my "poem", they understood a lot better. They felt that perhaps if it were published in the *ADVOCATE*, it might do some others some good as well. Therefore, I have enclosed a copy for you. Please free free to do with it as you see fit.

Sincerely yours,

Anna B.

Why Don't You Listen To Me?

Why don't you listen to me?

I have something important to say.

Make an effort, will you please

It's not my fault I have this disease.

At times I feel like I'm trapped inside,

Not being understood is not pleasant for me

But it is where I am today you see.

I am still the same person deep inside

With all the same emotions I have always had.

But, now you no longer listen to me

And does that make me sad.

So, why don't you listen to me?

I have something important to say.

I can no longer do the chores I once did

And that must cause you much fear.

But, have you ever stopped to think how I feel,

The frustration at times is too much to bear.

I was supposed to take care of you,

And now the reverse is true.

It makes me feel like a failure at times,

That is why I get depressed and feel blue,

Why don't you listen to me?

I have something important to say.

The simplest of chores have become an effort at times

And that takes a lot of getting use to

So, do not make it harder for me

By pretending that all is still fine.

I am still the same person deep inside.

The same person you once held so dear.

Though my exterior has changed quite a bit

I still need to be held very near.

So, please, please listen to me

I have something important to say.

Let us cherish each moment we have left.

Help me to laugh each day more and more,

For tomorrow will come soon enough

We never know for us what is in store.

So, let's try to talk while we still can

And continue to build each new day.

I love being part of your life,

And in your life I want to stay.

No time to waste with useless regrets

Help me instead, to be rid of my fears.

Continue to listen to me each day,

And there will be no need for tears.

You will know that you listened to me

Because I had something important to say.

The 1998 publications of the

PSP Advocate

have been generously funded by

John B. Ricker, Jr.

in memory of his loving wife,

JANE DARLING RICKER

SWALLOWING DIFFICULTIES IN PROGRESSIVE SUPRANUCLEAR PALSY: A PERSONAL PERSPECTIVE

*Bertrand L. Jaber, M.D.,
Division of Nephrology,
Department of Medicine,*

New England Medical Center Hospital, Boston, MA

My father was diagnosed with PSP in 1995 at the age of 70. He had been suffering from recurrent falls and visual difficulties for four years. At the time of diagnosis, a modified barium swallow study confirmed aspiration of clear liquids into his trachea, putting him at risk for pneumonia. At that time, the dietician recommended that all food and drinks have the consistency of thick-set yogurt. Soon afterward, I painfully discussed with my father the need to consider a feeding tube at some point. He deferred this decision to a later date. Over the following three years, the disease progressed and he became increasingly dependent on my mother who cared for him at home. It is a testament to my mother's determination and devotion that my father's health and weight were maintained for such a long period of time. Early this year, I urgently flew back home to find out that he had lost significant weight and feeding him by mouth had become an unbearable and inhumane ritual that neither of my parents could withstand. I felt compelled to revisit the need for a feeding tube, after having conferred with my mother and my two brothers and having obtained their support. I sadly succeeded in convincing him. The feeding tube was placed by a gastroenterologist under local anesthesia without any medical complications. However, this intervention was a major turn in my father's helpless resignation to this new physical handicap. In the next six months, his physical condition continued to deteriorate. He stopped ambulating and became bed-ridden, requiring total care. He continued to lose weight despite tube feeding. In fact, the feeding formula was often reassessed due to excessive diarrhea or constipation. He died at home on June 29, 1998 following an episode of aspiration.

Facing the dilemma.

Few people are equipped to deal with the painful ethical decisions that arise when a patient with PSP reaches an advanced stage of the illness. My experience both as a physician and as the son of a patient with PSP instigated me to review the issue of tube feeding. Whereas published literature on issues of tube feeding in elderly patients with dementia is quite elaborate, little is known of the role, timing and efficacy of tube feeding in PSP. The relationship between nutrition, aging and disease is quite complex. Indeed, the normal age-related decline in the host defense systems coupled to nutritional deprivation, may contribute to the increased susceptibility of the elderly individual to infection.

Whereas most physicians would agree that patients with PSP who have swallowing difficulties (or dysphagia) eventually require a feeding tube through which semi-liquid food has to be administered, few are clear on the timing, benefits and risks associated with this form of feeding, particularly in patients with advanced disease.

Continued next column

A priority one could argue that tube feeding should be reserved for patients with advanced stages of PSP, in whom there is a failure to insure minimal nutritional requirements due to severe dysphagia. However, this decision is often delayed due to ethical issues, in particular, the inability of the healthcare provider to confirm whether an intervention (such as the placement of feeding tube) will help or be distressing to a patient close to death. I propose that the need for a feeding tube should be discussed with the patient, and its placement pursued aggressively as soon as the diagnosis of dysphagia has been confirmed. If the patient is comfortable with this decision, I believe that early and intensive nutritional therapy can actually delay complications related to immobility, prevent infections, in particular, aspiration pneumonia.

Finally, malnutrition is often associated with vitamin deficiencies which may further weaken the host defense mechanisms against PSP, and contribute to the progression of this degenerative disease.. Indeed, free radicals are powerful substances that can damage cells. Recent research suggests that free radicals are associated with many degenerative conditions including the normal aging process, neurological disorders (such as dementia and Parkinson's disease) and cancer. Antioxidants are substances that inhibit the damaging effect of free radicals, and vitamin E is one such naturally occurring antioxidant. In the last issue of the PSP Advocate, there was a report on studies being launched in patients with PSP to examine the effect of free radicals on mitochondria, a cell component that is responsible for energy metabolism. The results of these studies may shed light on the role of free radicals in PSP, and help researchers design clinical studies that would target their harmful effects by using antioxidants. Although there is currently no evidence to support the use of antioxidants in PSP, I propose that vitamin E, at a daily dose of 400 I.U., is a safe and well tolerated dose, and may have beneficial effects in the treatment of this disease.

In conclusion, since aging and nutritional deficiencies may severely compromise an elderly individual's potential to confront PSP, it remains to be determined whether early and aggressive nutritional therapy, once acknowledged by a patient in the early stages of the disease, may delay its progression.

In memory of Toufic S. Jaber

Change In Membership Dues

The revised SPSP bylaws state that there will no longer be membership dues. As a nonprofit health organization, SPSP is solely dependent upon the financial contributions of our friends and supporters to sustain our research, education, and outreach programs. Your contribution is tax deductible.

If you wish further information about the SPSP bylaws, please call the SPSP office at 1-800-457-4777

SWALLOWING DIFFICULTIES IN PROGRESSIVE SUPRANUCLEAR PALSY: A SPEECH PATHOLOGIST'S PERSPECTIVE

By: Laura Purcell Verdun, M.A., CCC

I would like to start off by recognizing and thanking Dr. Jaber for sharing his personal experience with PSP and swallowing difficulties. My comments will reflect some of his, and expand somewhat on the type of problems that occur with swallowing and what resources are available to us. Additionally, I will incorporate some of the issues discussed at the Northern California PSP symposium in early November.

As many of you know, progressive supranuclear palsy (PSP) is a neurological disorder that predisposes individuals to swallowing difficulties. Therefore, it is important that aggressive efforts are made to evaluate and manage these difficulties to minimize complications such as malnutrition and aspiration pneumonia, earlier rather than later in the course of the disease. Dysphagia, or difficulty swallowing, can be life threatening particularly when it places an individual at risk for aspiration. Aspiration is when food, material, or saliva go down "the wrong way" into the airway, trachea, toward the lungs.

The swallowing evaluation usually consists of first a clinical examination. The majority of patients with PSP are referred to me by neurologists. I spend my time with patients and their care partners elucidating their concerns, their experiences and observations related to the potential swallowing problem. I find that when families maintain a diary of their observations that this is particularly useful for developing my management strategies. Generally during the clinical examination I will make suggestions to promote safer and easier swallowing. The suggestions are based on my understanding of normal swallowing physiology, and the changes I have observed in the numerous individuals with PSP with whom I have had the opportunity to work.

This clinical examination may be followed a modified barium swallowing study, or videofluoroscopic swallowing study. This is a dynamic radiographic examination of the swallowing passage during various consistency and volume presentations. Dr. Jaber notes that his father had this examination, which documented aspiration. I must admit that I do not standardly conduct this examination on all patients with PSP, because I do not believe that all patients need this examination. Clearly there are those individuals who are having such great difficulty with swallowing, that this examination would not provide us with any useful additional information. However, the decision to proceed with this examination is made on a case by case basis.

Dr. Jaber reported that they used a very common intervention, that is to modify the consistency of the foods and liquids his father was consuming. Thickening liquids will slow the transport through the mouth and throat. This is important if swallowing is not well coordinated, weak, or there is a disruption in timing events. So a patient may have trouble

Continued next column

swallowing grape juice, but making it a little thicker to a nectar consistency may be manageable. Soft and moist foods are also a common recommendation, avoiding dry and particulate foods such as nuts, and highly textured foods such as meats. These are generally easier to manage in the mouth, and may require less chewing. Another common suggestion is to take medications with a puree or pudding consistency, instead of with water of other liquids.

Clearly aspiration in and of itself can place an individual at risk for developing aspiration pneumonia. However, this is not likely exclusive. Over time, studies have shown that there are other contributing factors which can increase an individual's risk for developing aspiration pneumonia, for example, poor oral hygiene, bed dependence, and dependence for feeding. Additionally the presence of a feeding tube does not eliminate the potential for aspiration pneumonia, and it may potentially increase the risk. Clearly the development of aspiration pneumonia is a complicated, multifactorial process.

Dr. Jaber proceeds to share with us his personal decision making process regarding placement of a feeding tube for his father. This is a particularly difficult situation, and one that faces each family dealing with PSP. There is no right or wrong answer on how to make this decision, or what decision to make. However, I do believe that it is important policy for medical professionals and families to approach this topic sooner than later, so that the patient has the opportunity to communicate their interests. To some extent I believe this has the potential to minimize the stress of the family making the decision for their member with PSP. However, this does not necessarily mean that the tube should be placed sooner than later. Dr. Jaber addresses this issue perfectly. There is no research that would guide us on how to proceed. It is important to understand the risks for proceeding without a feeding tube, and alternatively, the risks and responsibilities associated with placement of a feeding tube. It is possible that early placement of a feeding tube may minimize the complications of malnutrition and dehydration, but this has yet to be proven, particularly in the PSP population.

I concur wholeheartedly with Dr. Jaber's conclusions. It is yet undetermined as to whether early and aggressive nutritional therapy may delay the progression of the disease. Perhaps it would allow that individual greater medical stability in with which to cope and minimize complications. My personal professional goal is that with a more prompt accurate diagnosis of PSP, patients will be referred on for swallowing intervention sooner rather than later, when perhaps little can be offered. I am very interested in determining if early intervention to address the anticipated feeding and swallowing difficulties associated with PSP, while an individual is still tolerating an oral diet, however with minimal or no evidence of impairment, could prolong safe, easy, and enjoyable mealtimes. Finally, I suggest that you refer to the new SPSP brochure on swallowing difficulties in PSP for additional information.

Laura Purcell Verdun is a Part Time Assistant Professor at Johns Hopkins University Department of Otolaryngology. She also works in the Voice and Speech Section, NINCD, National Institutes of Health. Laura is a member of the SPSP Board of Directors and has been active with PSP for over 6 years.

THE SOCIETY'S 1998 "GIVING CAMPAIGN" IS IN FULL SWING!

Thank you for your contributions to the annual "Giving Campaign.". If you have not sent your gift yet, an envelope is provided with this newsletter for your convenience. Many, many thanks in advance!

In addition to your gift, there are many ways to contribute to the Society.

- Initiate a letter writing campaign to friends and family.. Tell them your story. The Society will send you fact sheets and giving envelopes already addressed to include in your mailing. Gifts made be made to honor or remember someone special and the Society will be pleased to start a fund in someone's name.
- Ask your employer for a contribution or a matching gift. Ask them to publish an article in the company newsletter about PSP and ask fellow employees to contribute. *Consider a planned gift-a bequest, a gift of stock, insurance, creating a trust. The Society can help you design a plan.

**PLEASE HELP THE SOCIETY SUCCEED IN ITS' MISSION TO
FIND A CURE, PROVIDE INFORMATION AND SUPPORT SERVICES. GIVE TODAY!!**

**THE SOCIETY DEPENDS SOLELY ON YOUR
TAX DEDUCTIBLE CONTRIBUTIONS!**

New Online Newsletter Focuses on Neurology & Healthy Living

ST PAUL, MN Discover easy online access to the latest neurology health and wellness information. the American Academy of Neurology introduces its online newsletter NeuroVista.

The newsletter will feature articles on the latest advances in neurology and how they relate to maintaining a healthy brain. NeuroVista is an informative site that helps take the complexity out of complex issues.

This newsletter is for people who are committed to learning more about information impacting their health. By registering at the site, readers' will receive an e-mail alert highlighting the latest NeuroVista articles. Look for NeuroVista on the web at www.aan.com/neurovista.

Articles in the premiere edition of NeuroVista include:

- Exercise may reduce risk of Alzheimer's plus tips on how to begin an exercise program.
- Can you recognize warning signs of a stroke? Tips on lowering your risk for stroke.
- New dangers of smoking. Learn more about how to quit smoking.
- Improve children's attention deficit symptoms'.

The American Academy of Neurology, an association of more than 15,000 neurologists and neuroscience professionals, is dedicated to improving patient care through education and research.

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 / E-mail: SPSP@erols.com/ SPSP, Inc. Woodholme Medical Building, Suite 515 1838 Greene Tree Road, Baltimore, MD 21208

Grieving for the Lost SELF

Susan Hunt

A sense of self, a knowledge of who you are, the kind of person you are; pride in your uniqueness, in your abilities and achievements; all of these play crucial roles in helping you to value and enjoy your life.

You may have had to work very hard to develop this sense of self, this respect for who and what you are many hard-won achievements, some mistakes and "failures" overcome, are pieces of your "self" and help define the special person you perceive yourself to be. You may have to come to take pride in your skills your independence and your ability to contribute to the lives of others and to society. People do value themselves, very often, based on their ability to be productive, contributing members of society.

For the person with PSP all of that changes.

One of the most devastating aspects of PSP, is the immediate and ongoing, progressive, loss of pieces of "who" you really are. You cannot balance yourself, can't walk right. You may become stiff and clumsy. You may have double vision, have trouble swallowing, and/or have trouble controlling your eye movements. An enemy has entered your camp and is systematically robbing you of control of your body. The person you knew as "self" (healthy, independent, optimistic, in control) is disappearing and a different, unfamiliar "person" seems to be taking his place. Doctors tell you it will get worse, and there is very little you or they can do about it.

What a frustrating, terrifying, soul-searing condition to be in! People around you try to be encouraging and helpful. Your loved ones (and you) try to be hopeful and positive. (Maybe the doctors are wrong. Maybe the disease will stop progressing "go away". I don't have PSP after all. Maybe they will find a cure, -or at least an effective treatment.)

You may try to wall off the fear, yet as the weeks and months progress, -as the symptoms continue and/or new ones arise, you are gradually forced to acknowledge that things you cannot control are happening to you. You look in the mirror. The face is at least somewhat as you remember it, the PERSON behind the face is really still YOU, but so much about you has changed. Your actions, your appearance, even your behaviors occur often without your volition. You are changing and you cannot predict or control the changes. The feeling "I'm losing my SELF; I'm no longer who I was", builds within you and you find the wall you have built around the fear (and whatever other feelings you may have about this process -anger, frustration, helplessness, hopelessness) becomes harder and harder to maintain.

Take down the WALL. Acknowledge, allow the feelings. They are absolutely normal responses to the terrible situation you are in. Your fear is not cowardice. Your grief over losing parts of "who you are" is not unhealthy selfishness. You are a human being, whose normal, expected view of SELF would be positive and in control. It is your nature to value and care for yourself to see your body as a "tool", if you will, designed to do your bidding and give you your "place in the world. Valuing it and fighting for its survival is how you have survived so far. But the "rules" have changed. PSP is NOT a

normal process of life and is robbing you of parts of yourSELF. Honor the person (you) who is experiencing this loss and give yourself permission to grieve.

Look for someone who can be accepting and non-judgmental, who can sit with you as you grieve for the loss of the "self" you knew. You can grieve alone if you choose, but there is often great comfort in being able to share this deep and painful part of yourself with another caring person or with a group of individuals in similar situations. Find a way to speak the words, to cry the tears, to rage, if necessary. Holding the feelings inside takes energy and effort. Opening to an expressing the feelings, honoring the grieving part of you, is an important and valid part of the "protocol" if you will, for handling your disease. It can provide release. It can establish a connection to others, so you won't feel so completely alone. And, very often, once the feelings have been expressed, there is then room -and energy to take in a few of the good things that are happening around you TODAY, this moment. As difficult as they may be to see or to appreciate, they ARE there. Once you have released some of the grief, you may find it easier to see at least some of the beauty/love/pleasure around you.

You are special. You are a unique individual. And, you are dealing with a devastating disease. Be gentle with you.

Susan is a Clinical Hypnotherapist in private practice, specializing in grief, loss and pain management. She lives in Los Gatos, California.

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 Outbuildings, The Old Rectory
Wappenham, Nr Towcester, Northamptonshire
NN12 8SQ
Telephone 00 44 (0)1327 86029
E-Mail 10072, 30@compuserve.
Website [Http://www.ion.ucl.ac.uk/PSPeur](http://www.ion.ucl.ac.uk/PSPeur)

COMMUNICATOR'S LIST

The Society is updating its communicator's list. Communicators are persons who phone, write, or make friendly visits to persons and their families in their area or state. If you wish to be added or deleted from the list, please contact our office. If you would like a listing of the Communicators in your state, please call or write.

SPSP, Inc.
Woodholme Medical Building
Suite 515
1838 Greene Tree Road
Baltimore, MD 21208
1-800-457-4777



Support Group News

The Society would like to thank the following Support Group leaders who take their time and show their concern by sponsoring support groups, phoning and visiting PSP families. Please reach them at:

Carolyn Cheek
23009 Nadine Circle - Unit A
Torrance, CA 90505
(310) 534-8623

Janice Clements
49 Everest Road
Milton, VT 05468
(802) 893-1263

Janice Stober
NPF Center for Excellence
c/o Dr. Mark Guttman
377 Church Street, Suite 407
Markham, Ontario,
CANADA L6B1A1
905-472-7082

Charles and Janet Edmunson
17 Goodrow Lane
Framingham, MA 01701
508-872-9967

Maxine Faubion
9302 5. Citrus Lane
Sun Lakes, AZ 85248
(602) 895-7646
email: dmaxif@aol.com

Nancy Frederick
1131 Eden Avenue
San Jose, CA 95117
(408) 243-9987

Mary Gualandi
131 Ticonderoga Drive
Toms River, NJ 08755
(732) 505-1739

Kelly Harrison, Ph.D.
901 Southerly Road
Baltimore, MD 21204
410-337-9139
410-550-3167

Roberta Hunt
Route #1, Box 60A
Walla Walla, WA 99362
(509) 529-1364

Doris McCray
Rte. #2-Box 181C
Columbia, NC 27925
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June to Sept. (919) 796-0448

Fran McMahan
7148 191st Avenue, SW
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Ruth Nulph, RN
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Butler, PA 16001
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Margaret Prod-Homme, R.N.
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Dr. Lauren Seeberger
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701 E. Hampden, #501
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(303) 788-4600

Barbara Sharkey
5066 Evergreen Court
Rhineland, WI 54501
(715) 362-1777

Rosemary Taylor
25075 Poderio Drive
Ramona, CA 92065
(760) 789-5697

Charlotte Triplet
2440 Carvell Ave., North
Golden Valley, MN 55427
612-546-1694

Marcy Todd
21 Moorehead Oaks
Port Washington, NY 10050
516-883-7455

Kathy Thomas
3305 Francine Dr.
Decatur, GA 30033
770-939-2612

Dr. and Mrs. Robert Daft
527 Grovesnor Court
Sacramento, VA 95864
916-488-2608

Attention: Please notify the SPSP if you are in a PSP support group not listed above. Anyone interested in starting or joining a support groups, please call the society office at 1-800-457-4777.

Person interested in joining newly forming support groups near Cape Canveral, FL and Norfolk, VA, please call the office.

*Thank You, Thank You!!
For coordinating and organizing the California seminars in Sunnyvale and Anaheim — over 150 persons attended to receive valuable information - thanks to: The Parkinson's Institute, Anaheim Memorial Hospital, Sunnyvale Community Center, Carol Marchi, Pearlann Tabor, Leila Kirkpatrick, Nancy Frederick, Carolyn Cheek, Penny Roberts, Pat Kreuger and Wendell Higbee.*

**THE PSP ASSOCIATION (EUROPE)
CARERS AND THERAPISTS
CONFERENCE
OCTOBER 7, 1998
INSTITUTE OF NEUROLOGY
QUEEN SQUARE, LONDON**

*Reported By: John B. Ricker, Jr.
Caregiver's Caregiver, Ltd.
Society for PSP Board Member*

I arrived early and went to the Institute for Neurology and registered, received an agenda and order for the arrival of HRH The Duchess of Gloucester. We proceeded over to the Wolfson Theatre for the symposium's opening address given by Brigadier Michael R. Koe, Chief Executive, PSP (Europe) Association. Approximately 125 people attended the meeting. Michael Koe gave a brief history of the Association, a progress report that indicated strength, growth and contributions to research and support groups of which there are 75 in England. The support groups communicate with his staff daily. Ms. Grace Lewis, RGN, one of his contacts, told me that when they learn of someone with PSP they call immediately and offer help, assistance, love and affection. The new caregiver is urged to call anytime and they do just that; so the contacts and supporters are busy most of the time. They feed out information on a regular basis to the medical profession and the general public not only in the UK but across Europe as well. They support patients in Thailand, the Middle East and Australia. Brigadier Koe reported the PSP Association (Europe) has awarded the following grants for this year.

- | | |
|--|---------|
| 1. Dr. Huw Morris -Genetic Susceptibility | £32,00C |
| 2. Dr. Thomas Bak-Behavioral Effects of PSP | £4,000 |
| 3. Drs. Sue Daniel and Tamaz Revesz- The Tau Protein | £25,000 |
| 4. Dr. Uma Nath - A UK Wide Epidemiological Study | £93,000 |

This totals \$235,000

Dr. Andrew Lees, Chairman of the PSP Europe Medical Board, gave the introduction and spoke briefly mentioning the occurrences of PSP on the Island of Guadeloupe. Thirty one of sixty four patients have symptoms of PSP. Further research investigation is being conducted.



John Ricker in London attending the PSP Europe Association Symposium.



Over 125 attended the meeting.

Speakers and topics included the following:

- Dr. Huw Morris: PSP-clinical features, diagnosis and medical treatment
- Prof. Niall Quinn: MSA-clinical features, diagnosis and medical treatment
- Dr. Kaliash Bhatia: CBD-clinical features, diagnosis and medical treatment
- Mr. James Acheson: Eye movement and eyelid problems
- Dr. Clare Fowler: Bladder problems
- Prof Chris Mathias: Other "autonomic" problems
- Dr. Thomas Bak: Research-Neurobehavioral aspects
- Dr. Nick Wood: Research-Genetic aspects
- Dr. Anette Schrag and Dr. Peter Pramstaller: Research-Epidemiology
- Mrs. Sue McGowan: Speech and swallowing problems
- Ms. Jo Balcombe: Occupational Therapy
- Miss Cassie Gibson: Physiotherapy
- Open Forum: Questions to Panel of Speakers

During the afternoon of the symposium, HRH The Duchess of Gloucester arrived at Gowers Library. I was first in line as we were standing at attention and so she started off by inquiring about SPSP. I told her that we were moving forward faster in several directions, namely, fund raising, research, and caregiving under the direction of our relatively new director, Ellen Katz, who is innovative, energetic and untiring. HRH was pleased and wished us continued success. She is lovely and beautiful. We were able to get together again during the Tea Break.

I was pleased to meet face to face Dr. P. Pramstaller, from Italy, with whom I have corresponded over the years regarding his findings from familial link studies. I referred several of my PROFILE cases but he said that nothing developed from them.

I appreciated the opportunity to attend. I came away quite impressed with their work and accomplishments. I considered the meeting a huge success and from the many questions at the end I am sure that everyone was glad that they attended.

I suggest that SPSP and the PSP (Europe) Association maintain their close relationship and exchange of information.

L.A. AREA SUPPORT GROUP

By: Carolyn Cheek

There is something wonderful about the L.A. Area Support Group! As we share lunch once a month, we also share problems, laughs, hugs, tears, jokes, lawyers, information and hints. The group bonded instantly and we are proud to call one another friends.

At the beginning of each meeting, we share our current "story"-and every month it is different. The caregivers represent spouses with PSP that have suffered losses central to their lives. For example, a minister cannot speak, an athlete who cannot walk or throw, an engineer who finds it more difficult to follow directions, a social butterfly who has difficulty remembering directions to her friends' homes and a CEO of a large corporation that is robbed of his mathematical skills.

We got together for the first time last January. We decided that we would all wear red so that we could spot one another. Guess what!! It was Valentine's Day and everyone in the restaurant was wearing red. We shared our first laugh over that and then we shared our stories. What a relief to talk about what is going on in our lives with other people who understand. We shared ideas and helpful hints-even physicians. We also shed many tears-all the stories are heartbreaking and yet it is heartening to know that all of us are striving to make the life of our loved one with PSP as normal as possible. We all do whatever we can do to make our loved ones lives happier and interesting. Sometimes it is going out for dinner, reading the newspaper to them, going for walks, attending a class or going to church, or sharing books on tape.

Starting as support group in a city is easier than in a more sparsely populated area. I referred to the PSP ADVOCATE for lists of the names in my area and invited them to meet me for lunch. For a listing of people in your area, call the Society for PSP office. I called twelve people and five came to lunch. The original group was made up of intensely interested caregivers: Pat Krueger, Paul Wicorek, Penny Roberts, Jo

Michaels and me. At each meeting, we have added people. Joining us are Wendell Higbee, Gil Salazar, Barbara Miller, Alex Logan, Jeannette Muirhead, Rhoda Coco and Kay La Grandeur.

One of our first goals was to sponsor a West Coast PSP Symposium as several of us had seen tapes from the Baltimore Symposium in June 1997. We assumed it would be a do-it-yourself approach and proceeded to find a location and start thinking about speakers and publicity. The subject of postage came up and someone wondered aloud if the Society for PSP would underwrite the postage. The next day, I contacted Ellen Katz, Director, and posed the question. Ellen's response was very positive. We joined forces immediately. She contacted Carol Marcbi, Outreach Coordinator at the Parkinson's Institute in Sunnyvale and Society for PSP Board member, to work with us. The symposiums were planned for Nov. 6 in Sunnyvale and Nov. 7 at Anaheim Memorial Hospital in the L.A. area.

Our Support Group members are very excited about the November event and anticipated a large response. Our next Support Group meeting will include lunch with an attorney speaking and answering questions. We have created a flyer with information about the two events and have distributed it to interested parties. I now chair the Support Group and interface with Ellen and Carol. One of our members, Penny Roberts, created the Internet Support Group. She compiled a listing of the most frequently asked questions from the Internet group. Pat is a registered nurse and keeps us clear on medical terminology. Pat says that I need to say that I have excellent communication skills for which I thank her. I do know that any skills that I possess have been called into play.

The L.A. Area Support Group meets at Hof's Hut in Cerritos at 12:00 noon the second Friday of each month (except in November when a meeting was at the country club). We warmly welcome new participants.

For info: Call Carolyn Cheek (310) 534-8623 or
Wendell Higbee (714) 879-2620



The original support group (left to right) Jeannette Muirhead, Penny Roberts, Paul Wicorik, Pat Kreuger



*The July group (left to right) Seated: Jeannette Muirhead, Carolyn Cheek, Barbara Miller, Kathryn LeGrandeur
Standing: Paul Wicorik, Pat Kreuger, Wendell Higbee, Alex Logan and Penny Robert*

New York Support Group

By: Marcy Todd

I recently participated in the Parkinson's Disease and Movement Disorders Congress held at He New York Hilton. Our PSP executive director Ellen Katz accompanied by Nancy Brittingham our "PSP Advocate" editor put in endless hours of time preparing for this event. Together they organized an impressive information exhibit. Throughout the conference several volunteers assisted in manning the booth. Our thanks go to Larry Kasson, Jessica Todd, Teresa Arbeleta, Margaret Brent, Andy Todd, Beatrice Todd, Wendy Pankin and Kathy Voreg. Our PSP information booth attracted the attention of physicians with specialties in movement disorders. Those familiar with PSP shared some of their findings and experiences. Many physicians requested our informational packets and signed up to be included on our mailing list. With the support of these additional physicians, we hope to gain more awareness in the medical profession. Drug company representatives also visited the booth to share information regarding their updated drug trials.

It was wonderful to meet Dr. Steele. He is truly a man with a mission and a heart for people with PSP and their families. We are most fortunate to have him in our corner. Thank you, Dr. Steele, for all that you do!

On Sunday night we held an informal session for families and people suffering with PSP. Dr. Steele spoke to the group addressing all our questions. Dr. Joseph Higgins from the NY State Department of Health also presented his data drawing parallels with other neurodegenerative diseases that might be related. Our hope is that the researchers can discover the cure and work toward a treatment. Ellen Katz spoke about the growth of the Society and her passion was obvious.

At this meeting, the need for a support group in the New York area became apparent. This support group would be a forum for families and people with PSP to share information regarding equipment, care options and specialists that are available to help cope with the increasing demands of all.

On a personal note, my mother's battle with PSP has changed my life and that of my family. Never before was I faced with something that "could not be fixed" or that had no answers. This could not be happening to us. But it was and it did. Through my mom's strength of character, she influenced her family to think globally about this disease. As a result I have totally committed myself to being a voice and support in any way possible.

If you are interested in joining a support group, please contact me. My name is Marcy Todd and my phone number is (516) 383-7455.

The Society is starting support groups in Ohio, Kentucky and Indiana. Debby Mills of the Tri-State Parkinsons, will be the contact person. If you live in Cincinnati or the Tri-State area, please contact Debbie Mills at 513-558-7312.



APDA Coordinators meet in Baltimore
Celeste Duddleston, Deborah Mills,
Ellen Katz, Rebecca Dunlop



Dr. Mark Hirsch demonstrates on Bill Reed techniques for improving gait and balance. Dr. Mark Hirsch spoke at the September Baltimore Support Group meeting.

“A MOTHERS GIFT”

by Terry Sullivan



In 1994, I started a golf outing as a way to get together with my brothers as we live in different cities and states. The outing quickly grew from four people to twenty. We started raising money for SPSP in 1995 in memory of my mother, Jackie Sullivan. She died from complications of PSP in April 1994 at the age of 58. She left behind five sons ranging in age from twenty to thirty one.

This past July, we had our best outing ever by raising \$2,200 from the cash donations of players and other family and friends. It has become a tradition at the outing to donate winnings from individual and team contest to SPSP in the name of my mother.

My mother gave all her sons' the gift of golf by teaching us to play when we turned eight years old. Jackie was an excellent golfer and took pride in getting her sons involved in the sport. She started the junior golf program at the country club our family belonged to. She enjoyed teaching us immensely except when we started to out drive her and score better than she did. This usually started around age 12. We had great family golf outings and memories. There was time when my brother Tim had the opportunity to caddie for Jackie at the local club championship. On the 18th hole, she needed to chip into to win. Tim told her she was going to chip in and she did. Most of the family was there to watch.

Jackie was diagnosed with PSP in 1991 but had symptoms going back to 1988. Like most people with PSP, she was not properly diagnosed at first which made for a frustrating time for her and the family. She was very brave even when things were at their worst. She could not talk or show facial expressions in the later stages of the disease but was cognitively strong. There was a time when we had to ask her if she wanted to stop treating the pneumonia she developed with antibiotics and she strongly squeezed my hand twice for NO. Mom had a great faith life and deep respect for her life.

Whether you have PSP or are a family member supporting someone with PSP, you know the sacrifice and pain that this disease causes. Our outing does not raise a great deal of money but we have a great time doing it and I'm sure you too can find a way to help the society as well. It's been exciting to see all the society has done, with regard to research as well as developing family support materials and keeping people informed with new information about PSP. It is amazing what the human spirit can accomplish with the right focus and resources. We can all do more with the gifts our mothers gave us.

Helpful Hints

Two products are available that may help to improve the quality of life for people with Progressive Supranuclear Palsy.

The U-Step Walking stabilizer was designed with Parkinson's in mind and has also been helpful for people with PSP. It has a wide U-shaped base and a unique wheel formation that makes it extremely stable for those who are prone to fall. The walker is only 22 inches wide and conveniently turns in place and easily maneuvers in and out of tight places. The rolling speed of the walker can also be adjusted. If you have any questions, requests or need additional literature, contact In-Step Mobility Products, Inc., 8040 N. Ridgeway, Skokie, W. 60076 or call 1-800-558-7837.



The Uplift Seat Assist is a portable lifting cushion that can fit in any chair. The cushion provides the extra lift needed to get out of a chair. It is self-powered and portable. It weighs 8 pounds and is 17 inches wide. The Seat Assist is made by Daylight Technologies, Inc. For more information, contact Medassure at (602)-893-1662.

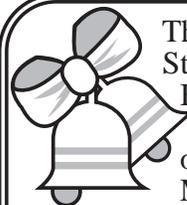
Editor's Note: This information is provided as a service. The Society for PSP does not endorse these or other products and neither the Society nor its staff have financial interests in these products. Information is offered with the intention to inform others about products that may help to maintain independence and improve the quality of life.

If you have any helpful hints, medical devices, equipment, or services you can recommend to other PSP families and caregivers, please write:

Nancy Brittingham, Editor
PSP Advocate

Wooholme Medical Building
Suite 515
1838 Green Tree Road
Baltimore, MD 21208

or
Fax: 757-838-6086
email:
103301.640@compuserve.com



The Society for PSP congratulates Stephen Reich, M.D., Chairman, Board of Directors, on his recent marriage to Dana Boatman, Ph.D. on December 6, 1998 in Baltimore, MD. The couple, who are both neurologists at The Johns Hopkins University School of Medicine, will reside in Baltimore. May you both have many years of happiness!!

The following people and organizations have generously contributed to the Society during the period August 1, 1998 through October 31, 1998. The Society is deeply appreciative of gifts from the following persons:

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The Society would like to thank the INTUIT Corporation for their donation of the multi user version of Quickbooks. This latest version of Quickbooks enables the Society to have more than one person access and log in information thereby improving the efficiency of our financial operations. Many thanks to the INTUIT Corporation.

5TH INTERNATIONAL CONGRESS

(Continued from page 3)

mother and her two sons, all three of whom developed PSP in their 40's. Autopsy in one of the sons confirmed the diagnosis. Such a family is too small to allow geneticists to find their culprit gene, but when combined with many other such families with the same genetic defect, could permit that defect to be found. There have been a handful of such families reported in the medical literature. The existence of such families does not imply that there is a high risk to the family members of everyone with PSP, but it does illustrate the point that a simple genetic defect can cause PSP and stimulates the search for weaker genetic factors that could contribute to PSP in general.

Clinical Features

PSP nearly always begins with right and left sides of the body equally affected, unlike PD or corticobasal degeneration (CBD), two conditions that can often be difficult to distinguish from PSP. Dr. H.H. Fernandez and colleagues from Boston University reported three patients with autopsy-proven PSP who each had as their initial symptoms definite abnormal limb posturing (dystonia) and difficulty performing familiar manual tasks (apraxia) on only one side of the body. This abnormality is the rule in CBD. Its presence in a few cases of PSP suggests an overlap in the causes of PSP and CBD (an overlap that was evident in any case from their very similar changes under the microscope) and shows that strict clinical criteria for the diagnosis of PSP may be unwarranted.

In the seemingly never-ending quest for a relatively convenient, objective test to distinguish PSP from PD, Dr. C. Brefel-Courbon and colleagues from Toulouse, France measured the reflex blood pressure response to being tilted head-up on a tilt table. Patients with PSP had the same ability to correct the drop as the normal volunteers, but those with PD who were on Sinemet demonstrated an important defect in this ability. Patients with PD in the early stages who were not on medication had a slightly deficient blood pressure response, but it did not differ significantly from that in normals or PSP. Once again, a test that can distinguish moderate or advanced PD from PSP, when a good neurologist can usually do the same thing, fails to distinguish the two diseases at an early stage.

Epidemiology

A remarkable concentration of people with PSP in the French West Indies was again presented by Dr. D. Caparros-Lefebvre and colleagues. Of 72 patients with various types of parkinsonism in their clinic, only 16 (24%) had PD, while 55 (75%) had probable or possible PSP. (In clinics everywhere else in the world, the figures are typically about 70% PD, 4% PSP, the remainder multiple system atrophy, multiple strokes, and other conditions.) The authors also note that all of the PSP patients habitually drank an herbal tea made from *Annona muricata* or *Annona squamosa*, while only a minority of the PD patients did so. These two plants contain benzyl-tetra-hydro-isoquinolines, which induce parkinsonism in experimental animals. Unfortunately, there seems to have been little progress in understanding these observations since they were first reported at a different research meeting early last year. There have been no independent confirmation of the

Continued next column

neurological diagnoses and no autopsies to confirm PSP. Another question is whether the tea drinking is merely a marker for a different, more important causative factor such as geographical or ethnic origin. We look forward to more information about this "cluster" of PSP with possibly huge scientific importance.

BRAINSTORMING CONFERENCE

(Continued from page 1)

written by each group will be discussed by the whole group with a final document being prepared. The final document will be submitted for publication.

The Brainstorming Conference on progressive supranuclear palsy will be a unique opportunity for the exchange of ideas and will lead to the development of research guidelines for the next years to come. In addition, we expect that this meeting will allow the possibility to increase collaborative studies among participating scientists.

Brainstorm Conference on PSP-Topics of study

I. Basic Mechanisms and Animal Models

- A. Studies evaluating the role of tau and astrocytes in PSP
- B. Search for animal models of the disease
- C. Studies searching for Biologic Markers
- D. Genetic Studies

II. Anatomoclinical Studies

- A. Basal Ganglia Circuits
- B. Oculomotor Studies
- C. Neurophysiologic Studies
- D. Neuroradiologic Studies

III. Clinical Characterization and Therapy

- A. Cognitive, Neuropsychiatric and Neuropsychologic Studies
- B. New Therapeutic Approaches
- C. Neuroepidemiologic Studies

The Society for PSP gives their thanks and appreciation to the family of Ruth Lindenbaum. Her daughter, Marcy Todd, along with her father and family, sponsored the first New York support group meeting at the New York Hilton on Oct. 11, 1998. Ruth Lindenbaum's family has also generously funded the updated educational pamphlet, *PSP SOME ANSWERS*, written by Dr. Lawrence I. Golbe, Professor, Robert Wood Johnson Medical School, NJ. The pamphlet is dedicated to the memory of Ruth and her courageous battle against PSP and her perseverance in acquiring the most current information available. Her family supported this brochure with the hope that others will receive it and learn more about PSP.

INFORMATION ON PSP AVAILABLE FROM SPSP

Learn more about PSP its symptoms and strategies for managing them. An informative booklet and several brief folders on PSP symptoms, written in nonmedical language, are available on request. There is no charge for booklet or folders.

Video tapes of symposia for people with PSP family members and caregivers. Provide scientific information as well as suggestions for day-to-day care-giving.

The booklet, folders and video tapes represent SPSP's effort to inform patients, family members, family-caregivers, and professionals about PSP and its management. **ORDER BY NUMBER at 1-800-457-4777.**

Video Tapes



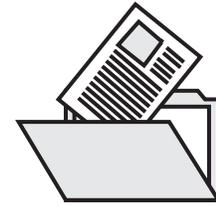
"The Diagnosis of PSP" (#2)

Made for and sold only to neurologists, neuroscience nurses and similar professionals. The tape depicts presentations on diagnosis of PSP at the international scientific conference in Barcelona. Recommended for clinicians and faculty. VHS format, U.S. \$30, prepaid.

1997 Symposium (#10)

Captures the presentations at SPSP/Johns Hopkins 1997 symposium for patients, family members and caregivers. See and hear Steele, Golbe, Reich, Zee, Litvan, Growdon and several other experts discuss various aspects of PSP. 3 tapes of complete program. VHS format, U.S. \$75 plus \$7 shipping outside U.S. \$10 shipping.

Pamphlets



"PSP: Some Answers" (#1)

This is a highly regarded, widely circulated discussion of PSP by Dr. Lawrence I. Golbe that is easy to read, and easy to understand. Recently revised, updated, and printed in booklet form. Specify English or Spanish.

"Helping The Helpers Who Care For People With PSP" (#3)

Dr. Stephen Reich discusses the vital role of the caregiver in PSP. He offers important advice for self care in the interests of the patient.

"The PSP Advocate" (Newsletter) (#4)

PSP Advocate, the popular quarterly published by SPSP to chronicle its activities, keep you up-to-date on PSP research, provide a support forum, inform readers on participation in research studies, and tell you about self-help mutual help opportunities through advocacy.

"Swallowing Problems in PSP" (#6)

This pamphlet deals with one of PSP's most critical problems. It offers recommendations for managing the symptoms and suggestions for avoiding its dangers.

"Personality Changes in PSP" (#7)

Some PSP patients undergo changes in personality and behavior that bewilder family members and caregivers. This pamphlet discusses the problem and offers suggestions.

PSP Fact Sheet (#11)

8 1/2 x 11 easy guide to PSP can be copied and distributed.

"Baltimore Sun" Article (#12)

Reprints of feature PSP article in Baltimore Sun written by Diana Sugg.

Brain Bank Information (#14)

Packet of information about the PSP Brain Bank. Details how to make preparations to make brain donations and help in the search for the cure for PSP.

PSP: Swallowing Problems (#15)

Feeding-Swallowing Difficulties General Suggestions. Written by Laura Purcell Verdun, M.A., CCC (Research Speech Pathologist).

ON LINE SERVICES

Website: www.psp.org

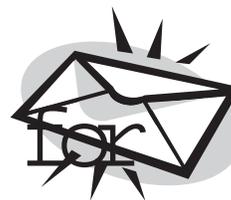
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Subscribe psp your name

Questions: Parkinson's Web <http://pdweb.mgh.harvard.edu>

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Please e-mail the addresses of any sites relating to PSP or other neurological disorders, caregiving issues, etc. to SPSP@erols.com and we will list them in the next *ADVOCATE*.



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