

Table of Contents pages
Medical Updates 3-8 Living
with PLS 8-10 Events 10-13
Caregiving 13-14 Funding
15

PLS Symposium By Thurza Campbell, editor At the California Connection in the fall of 2002, several PLSers met with Drs. Fink and Siddique. That meeting sprouted the seed for a symposium to define PLS clearly as an important step in better understanding our disease and eventually curing it. Dr. Siddique said in mid-January, "As some of you know, Dr. Fink and I are putting together plans for an NIH sponsored meeting that will include presentations and discussions by those practitioners and researchers with the most interest and experience in PLS. We envision having several sessions over a 2-day period that would address such topics as epidemiology, criteria for diagnosis, collaboration in future research, and relationship to other diseases of the motor neuron. This is an exciting prospect, and I think will bring together those with a serious interest in the disease so that we can move forward together to

better understand PLS."

Dr. Teepu
Siddique

Our responsibility, the PLS Community, is to raise \$25,000 as our share of the Symposium costs. Some of you responded in December, 2002 to this exciting gift opportunity and we thank you. As a matter of fact, we have raised 20% of our goal as of January 13, 2003 with 10 donations. Below is a letter, which serves as an example of how one PLS family member (my daughter) is trying to help raise money for the Symposium. You might like to adapt it yourself and send a similar one to your friends:

Winter 2003

**Serving the Primary Lateral Sclerosis Community
since 1997**

<http://www.als-pls.org/contribute.htm>

Meet Synapse Founder

As most of you know, in November of 2001, my mother, Thurza Campbell, was diagnosed with Primary Lateral Sclerosis (PLS). PLS is a rare motor neuron disease most characterized by progressive weakness and stiffness of voluntary muscles. Right now the main symptoms my mom exhibits are changes in her balance and her speech.

Dear Friends and Family, January, 2003

Unfortunately PLS is an “orphan disease.” There are still many unanswered questions and research to date has been incomplete to clearly define the disease.

My mom has been remarkable in dealing with this diagnosis; the daily challenges have been huge and she has handled them with her chin held high. Her strength and attitude are an inspiration to many.

Most recently, through her new role as editor of Synapse- an online PLS Newsletter, <http://synapse.home.attbi.com/>, my mom has been

made aware of the upcoming PLS Symposium, organized by the Northwestern University School of Medicine. An international meeting will take place in 2004 with the goal of defining the disease. Once this has happened, neurologists will be closer to a cure.

Needless to say, the need for fundraising is essential. In order for the symposium to complete its mission, \$25,000 must be raised by the PLS community. One of my New Years resolutions is to raise at least \$1000 for the symposium in honor of my mom. I am hoping that I can count on 50 friends and family members to each donate \$20. While the disease is not contagious, I hope that the fundraising will be...

If you are able to help, thank you. I can only hope you know how much your efforts mean to my family and me.

Thank you.

Sincerely,
y,

**Make tax-deductible donations to:
NUMS-PLS Symposium Mail donations
to: Office of Medical Development
Northwestern University Medical School
Abbot Hall, Suite 1312 710 North Lake
Shore Drive Chicago, Illinois 60611-3078 To
make a contribution by any major credit
card, use the following link to the
contribution page at the ALS-PLS**

Synapse - Winter 2003 Edition Page 2

Thanks to Joe, who brought me to Ed, who brought me to the computer, I now had all of you. Then we had the great Connecticut Connection and I put so many faces with

Website:

Joe Alberstadt and his wife Carol.

Joe began publishing **Synapse** in 1997. He actually sent it out on a monthly basis! Often he featured a different PLSer each month. We are all indebted to Joe for his inspiration to create **Synapse** and for his dedication to the task he set for himself.

Thank you, Joe Alberstadt

Contributed by Ronnie Grove My first contacts with other PLSers came as newsletters from Frank Levy and Joe Alberstadt. Those first newsletters were just that: letters with PLS news. Were they ever welcome! For me, this was pre-computer and the greatest thing happening in my world. Joe reported meeting other PLSers in Florida. Several of us asked Joe to consider a meeting a little farther north. I attended that first gathering Joe hosted in Vienna, VA in 1998. That's where I met Ed Ames who encouraged me to get a computer. By this time the newsletter was becoming quite savvy and had gotten itself named **Synapse**. Through **Synapse**, I was aware there was a lot happening that I was missing out on. Ed kept after me and I finally got on line in April 1999. Hallelujah!!! What a day!

names from the PLS Friends group and I can't begin to tell you how wonderful that was. Thus came the first thoughts of a West Virginia Connection, a.k.a., Spring Fling. I

wanted to share my PLS knowledge and maybe reach someone who was out there struggling all alone. Since that Connecticut meeting in October of 1999, there have been many regional connections, lunches, gatherings, meetings, dinners, creation of SP Foundation, and finally, this past September there was TeamWalk.

Above all, call them IMPORTANT. This is NETWORKING! Networking is something we can all do to be a part of the big picture. From Joe to Ed to computer to PLS-Friends to TeamWalk. We are making a difference! Thank you, Joe for starting the ball rolling.

MEDICAL UPDATES

Advances in HSP and PLS research.

Contributed by Dr. John Fink This past year has seen rapid advances in our knowledge of the causes of hereditary spastic paraplegia (HSP) and primary lateral sclerosis (PLS). Three genes (HSP60, kinesin heavy chain, spastin) have been reported in the past nine months. ALSin (a gene for both PLS and a familial form of amyotrophic lateral sclerosis (ALS), and genes for HSP (spastin, atlastin) were discovered in the previous year. There are at least 15 more HSP and PLS genes and additional gene discovery is underway. It will take some time to fully understand the function of each of these genes, their proteins, and the factors with which they interact. Nonetheless, each gene discovery provides another piece in the puzzle that will expose the biochemical processes that underlie these conditions. At present, gene testing for spastin and atlastin mutations will

diagnose more than 50% of individuals with dominantly inherited HSP. Discovery of additional genes extends the ability to diagnose

these conditions in the laboratory and not by clinical parameters alone. Over the next several years, the focus of HSP and PLS research will shift slowly from gene discovery to biochemical and cell biology investigations; and to explorations of potential therapies. Laboratories are already beginning to focus attention on potential disturbance of elements that give structure to neurons (microtubules, for example) and the process of transporting molecules down the long length of the nerve processes (axons) as possible mechanisms that underlie HSP and PLS. Animal models (mice) in which to explore the molecular causes and treatments of HSP and PLS are already being developed and showing promising results. There is very tangible progress and very real cause for hope that these advances will lead to real treatments for these conditions.

After Ground-Breaking Discovery, Siddique Digs further into ALS and Other Diseases

From The Northwestern University Institute for Neuroscience The work goes on for Teepu Siddique, who made national headlines last year when he and a team of international scientists discovered a second gene mutation that causes an inherited form of amyotrophic lateral sclerosis, more commonly known as Lou Gehrig's disease, Siddique, the Abbott Laboratories Duane and Susan Burnham

Research Professor and director of the Neuromuscular Disorders Program at the Feinberg School of Medicine, recently received a three-year \$1 million grant from the Dr. Ralph and Marian C. Falk Medical Research Trust, one of the country's largest foundations that awards grants for medical research. The grant will support

neurogenetics research, which includes ALS and other neurodegenerative diseases, Siddique also serves as director of the Les Turner ALS Research Laboratory at the medical school. Also, in April he received a research grant from the National Organization for Rare Disorders to

Synapse - Winter 2003 Edition Page 3

study the molecular genetics of Primary Lateral Sclerosis. Because more than one gene is involved in the development of PLS, the group is investigating the disease in families not linked by the same locus described by Siddique and colleagues on chromosome 2. Work in progress includes making mice with the ALS2 genetic defect and identifying the chromosome that causes the genetically unique occurrence of dominant ALS. He and his team are also looking into the genetic contribution to the sporadic-or non-inherited-form of ALS, environmental interaction with the genes that lead to the disease, and additional loss of function genes, which includes an ALS gene on chromosome 15. Yet another project is screening 1,000 FDA- approved drugs in a tissue culture system derived from the transgenic ALS mouse model developed in 1994 to see if any of them reverse the disease. "If you can reduce the amount of mutant protein, you can cure the disease or postpone the disease in ALS caused by mutations in SOD1," he said. "The idea is to find a drug that will dampen down this particular protein and we are testing that in the test tube right now." If successful, mouse

materials are next, and then determining the correct dosages for patients. The path to human use is expected to be a smooth one because the drugs are already approved, he said. As reported in the October 2001 issue of Nature Genetics, the newly identified gene mutation, ALS2, is responsible for a rare, slowly progressive, early-onset form of the disease called juvenile inherited ALS. The gene is transmitted in an autosomal recessive pattern; that is, the individual inherits copies of the same recessive gene from both parents. It was discovered on chromosome 2q33 in highly inbred families from Tunisia and Saudi Arabia. Equally as stunning was the finding that mutations in the same gene also cause childhood-onset PLS, thus ending a century-old debate as to whether PLS is a form of ALS or not. Because of this investigation, it is now known that the two diseases have the same genetic origin, although they arise from different mutations. In 1991 Siddique discovered the location of the first gene, ALS1, on the smallest human chromosome, 21. Two years later he and collaborators from Massachusetts General Hospital identified the gene to be Cu, Zn superoxide

dismutase. They discovered the location of the second gene in 1994, and also that year, developed a transgenic mouse model that mimics ALS1. These mice are now available to researchers worldwide. "For the future, what we want to do is understand what pathway or pathways in the cell go wrong," he said. "The molecular architecture of the nervous system, spinal cord and brain stem are still totally inaccessible by imaging or any other way we know in live patients. We have to understand these diseases by using animal models to come out with predictable therapies and treatments."

MGH ALS Research Program

Progress Report Dr. Robert H. Brown, Jr., D.Phil, M.D. **Introduction** Created in 1984, the Day Neuromuscular Research Laboratory and Neuromuscular Clinic at the Massachusetts General Hospital have been

focused on finding ways to understand and treat paralyzing disorders of nerve and muscle, with a particular focus on ALS. This effort has entailed the development of a strong clinical program and, in parallel, a basic research program. The clinic is now an active, full-service clinic with multiple staff members participating. Central to the clinic is a team engaged in implementing drug trials in ALS (see below), simultaneously with laboratory-based studies of drug therapy in ALS mice. The laboratory has actively pursued causes of several neuromuscular diseases since its founding in 1984. However, the primary focus has been ALS because ALS is clinically so devastating. Since the early 1980's, members of the laboratory have elected to study inherited forms of neuromuscular disease, primarily because the technology to do so emerged in the early and mid-1980's. With the

Synapse - Winter 2003 Edition Page 4
method of genetic linkage analysis, the laboratory was honored to discover gene defects that cause two forms of familial ALS (adult onset and juvenile onset), hyperkalemic paralysis, a type of sensory nerve degenerative disease, and a type of muscular dystrophy. **Stem Cell Transplantation Studies** Over the last two years, we have devised a stem cell transplantation project to test the hypothesis that stem cell and fetal neuronal cells delivered both into the spinal cord and systemically will inhibit motor neuron cell

death in a mouse model of ALS. The premise of this effort is that stem cells may be beneficial in ALS in at least three ways: ~At least in theory, the stem cells might become new motor neurons. This possibility has yet to be validated but nonetheless holds out the important promise of restoring lost neurological function to the paralyzed spinal cord. ~ There is extensive literature suggesting that stem cells may be therapeutic by secreting growth factors that help the spinal cord retard the process of motor neuron cell death in a disease like

ALS. ~ There are data arguing that stem cells can differentiate into types of cells that can benefit motor neurons even if there is no differentiation into motor neurons. With this as a rationale, we have embarked on two type types of studies of stem cells. In one arm of the project, we have administered cells intravenously. In the second arm of the project, we have administered the cells directly into the spinal cord via a microscopic surgical procedure. **Genetic Investigations** The Day Lab has continued the program to discover new ALS genes in the expectation that this information will provide fresh insight into pathways and mechanisms for the cause of ALS. Moreover, as in the case of SOD 1, new genes may be powerful routes to developing new animal and cell models of ALS. Our collection of families with inherited ALS now numbers nearly 500. Of these, only about 25% arise from mutations in the SOD1 gene.

This fall (2002) we were part of a collaborative team that reported finding a gene whose deficiency causes a childhood onset form of motor neuron disease. In the other 14 non-SOD 1 ALS families we have almost completed a full screen of the human genome for more linkage. We believe we have found at least two new ALS loci during this process. Unfortunately, each is linked to only one family. On the other hand, the silver lining on this possible cloud is that we have shared the information about these new ALS gene addresses. To our surprise, three of

our collaborators (in Europe) have confirmed that they have ALS families whose genetic address is linked to the same region as in one of our ALS families. Accordingly, as we go forward we will focus on this region. **ALS Biomarkers** With the exception of screening for SOD 1 gene mutations, there is no definitive diagnostic test for ALS. With this in mind, we have engaged, with collaborators, in a search for molecules that are uniquely present (or absent) in ALS tissues and fluids as compared to controls. Such molecular markers, or "biomarkers", offer two important research advantages. They may provide clues about what causes ALS. Also, they may be useful in the process of monitoring drug therapy in ALS. It may be that one examines the effect of a possible ALS therapy on the biomarker in a matter of a few days or weeks whereas many months are required to see if that therapy alters survival in ALS patients. We have looked for types of DNA that have been modified by free radicals and found clear evidence of such modified DNA in ALS blood, urine and spinal fluid. More recently, we have begun to use very sophisticated methods to look at the levels of hundreds of small molecules in the body that reflect on-going cellular metabolism. This search for biomarkers is not likely to provide short-term answers. The studies are long and expensive. However, the potential benefit to understanding and monitoring cell death in ALS

are enormous, and amply justify the long-term commitment to the hunt for biomarkers. The Community of ALS Scientists and Partners We acknowledge that finding a cure for ALS will continue to be a team effort. Multiple types of resources must be mustered to fund the research in a complementary, non-duplicative manner. At the same time, ALS investigators in diverse centers with varied programs must interact closely to enhance rather than compete with each other. We are fortunate to receive support for our research from both federal and private sources. We are also committed to close interactions with multiple collaborating ALS researchers. I have been honored to chair an NIH research grant committee (related to all neurological diseases) and to serve on the boards of ALS support groups such as the Angel Fund, ProjectALS, ALS Family Charitable Foundation, ALS Therapy Alliance, and the Al Athel ALS Therapy Program. We will continue to foster and maintain interactions like this to guarantee that our research and therapeutic programs are as focused, original, and productive as possible. **Note:** Information on Dr. Brown's Drug Discovery Programs and Human and Mouse Drug Trials will be in the Spring, 2003 issue of Synapse.

Medical Terms that are often associated with ALS & PLS

Contributed by Jennifer Thomson **Brisk Reflex** - a condition that describes the deterioration of the upper motor nerve cells (neurons). **Bulbar Muscles** - the muscles

that control the speech, chewing and swallowing. **Classical ALS** - a progressive neurological disease characterized by a deterioration of upper and lower motor nerve cells (neurons). This type of ALS affects over two-thirds of all people with ALS. **Dysarthria** - impaired speech and language due to weakness or stiffness in the muscles used for speaking. **Dysphagia** - impaired chewing and swallowing.

Exertional Dyspnea - a condition characterized by shortness of breath during physical activity. **Familial ALS (or FALS)** - a progressive neurological disease that affects more than one member of the same family. This type of ALS accounts for a very small number of people with ALS in the United States (5-10%). **Fasciculations** - non-painful, rapid and involuntary contractions or twitchings of groups of muscle fibers. This is often described by people with ALS as "a persistent rolling beneath the skin." **Flaccid, Weak Muscles (also Hypotonicity)** - a condition characterized by a decrease or loss of normal muscle tone due to the deterioration of the lower motor nerve cells. **Hyperreflexia** - excessive response of muscle reflexes when a normal stimulus is applied. **Hyporeflexia** - weak or absent muscle response when a normal stimulus is applied. **Lower Motor Neurons** - nerve cells (neurons) situated in the spinal cord and brain stem and their projections (axons) forming nerves that end in the muscle fibers. **Motor Neuron Diseases** - a group of disorders in which motor nerve cells (neurons) in the spinal cord and brain stem deteriorate and die. ALS is the most

common motor neuron disease. **Muscle Atrophy** - loss of muscle fiber volume characterized by a visible decrease in muscle size. This occurs because muscles no longer receive impulses, or "messages," from nerve cells (neurons). **Muscle Cramps**, Unexpected - involuntary, painful shortening of muscles. Usually, a knotting of the muscles is visible. **Muscle Weakness** - loss of strength, increased fatigue, loss of

coordination and difficulty with motor skills and lack of ability to carry out certain skills. **Primary Lateral Sclerosis (PLS)** - a progressive neurological disease in which the upper motor nerve cells (neurons) deteriorate. If the lower motor neurons are not affected within two years, the disease usually remains a pure upper motor neuron disease.

Synapse - Winter 2003 Edition Page 6

Progressive Bulbar Palsy (PBP) - a condition that starts with difficulties speaking, chewing and swallowing due to lower motor nerve cell (neuron) deterioration. This disorder affects about 25% of all people with ALS. **Progressive Muscular Atrophy (PMA)** - a progressive neurological disease in which the lower motor nerve cells (neurons) deteriorate. If the upper motor neurons are not affected within two years, the disease usually remains a pure lower motor neuron disease. **Pseudobulbar Palsy** - a condition that is characterized by difficulties with speech, chewing and swallowing. These symptoms resemble those of bulbar palsy, but this condition is also characterized by spontaneous or unmotivated crying and laughing. **Sialorrhea** - drooling resulting from the lack of spontaneous, automatic swallowing to clear excessive saliva in the mouth. **SOD1 positive FALS** - the copper-zinc SOD (SOD1) is the first gene found to have mutations resulting in approximately 20% of FALS, or approximately 1-2% of all ALS. **Spasticity** - a state of increased muscle

ension when the muscle is lengthened. Often involves an exaggeration of the tendon reflexes. **Spinal Muscular Atrophy (SMA)** - a hereditary neurological disease in which only the lower motor nerve cells are affected. **Upper Motor Neurons** - nerve cells (neurons) originating in the brain's motor cortex and their projections (axons) descending through the brainstem and spinal cord.

Study Seeks PLS Patients

Columbia-Presbyterian Medical Center in New York seeks PLS patients for study to help differentiate diagnosis between PLS and ALS. Uses magnetic resonance spectroscopy.

Email:

cmd9@columbia.edu or
call (212) 305-5105

The Physician's Guide To Primary Lateral Sclerosis Is Going Over

Very Well Contributed by Linda Gentner
To quote NORD, "we're receiving MANY requests for the PLS booklet, and some nice

notes from physicians indicating that they are finding the booklets helpful". (Hooray, that was our goal!). I have mailed a few copies to my "other" doctors and they, too, received them with gratitude. I know that others of you have gladly taken them to your physicians as well.

PLS Education Fund -- approx. \$36,000 -- of which we used \$14,000 in the initial mailing to the neurologists and teaching hospitals. Since we have a large amount left, we are doing an additional mailing to 2,600 health science libraries at hospitals. Recap: brochures mailed to approx. 14,000 neurologists, plus approx. 400- 500 teaching hospitals and now an additional 2,600 to health science libraries. NORD also stated that Angela Dixon saved our Education Fund **at least** \$1,500 by providing the art work, and her expertise. Kudos, once again, to Angela. (Note: \$36,000 was total before any

mailings and the transferring out of \$3,000 to our Research Fund.) **PLS Research Fund** -- approx. \$32,000. I transferred \$3,000 from the Education Fund to the Research Fund in order to bring the Research Fund up to \$35,000, which is the minimum amount needed for the Request for Proposal (request for the grant money) since NORD's next cycle date is January. Most of us were frustrated with the amount of turn around time it took from our raising the over \$120,000 to the actual granting of the awards, but at least it finally happened much to the delight of our recipients Drs. Teepu Siddique and John Fink. As a reminder, if you need additional booklets, e- mail Mary at <http://www.rarediseases.org> and if you need the postal address, it's: Mary Dunkle Vice President of Communications National Organization for Rare Disorders 55 Kenosia Avenue

Synapse - Winter 2003 Edition Page 7
PO Box 1968 Danbury, CT 06813-1968 --
or Phone: 203-744-0100 Fax:
203-798-2291 We have the booklets
available free of charge, please use
them!

LIVING WITH PLS

Free Admission to all US National Parks with Handicapped

Plate/Permit Anyone who has a Handicap
plate, permit or is permanently disabled can
get in free at all US National Parks. Go to the

information center at any National Park and
ask for a handicap form. They will give you a
'Golden Access Passport', A Lifetime
Admission Permit. Any citizen or person
domiciled in the United States medically
determined to be permanently disabled for
purposes of receiving benefits under Federal
Law can get one. Everyone in your private
car is admitted free, also.

Subject: Age and other stuff...

Contributed by ? Eventually you reach a
point when you stop lying about your age
and start bragging about it.

----- The

older we get, the fewer things seem worth waiting in line for.

----- Some people try to turn back their odometers. Not me, I want people to know "why" I look this way. I've traveled a long way and some of the roads weren't paved.

When you are dissatisfied and would like to go back to youth, think of algebra.

----- I don't know how I got over the hill without getting to the top.

----- - One must wait until evening to see how splendid the day has been.

Yah, being young is beautiful, but being old is comfortable.

----- Old age is when former classmates are so gray and wrinkled and bald, they don't recognize you.

----- If you don't learn to laugh at trouble, you won't have anything to laugh at when you are old.

Cracked pots are we Contributed by Kathi Geisler A water bearer in India had two large pots, each hung on the ends of a pole which he carried across his neck. One of the pots had a crack in it while the other pot was perfect and always delivered a full portion of

water. At the end of the long walk from the stream to the house, the cracked pot arrived only half full. For a full two years this went on daily, with the bearer delivering only one and a half pots full of water to his house. Of course, the perfect pot was proud of its accomplishments, perfect for which it was made. But the poor cracked pot was ashamed of its own imperfection, and miserable that it was able to accomplish only half of what it had been made to do. After 2 years of what it perceived to be a bitter failure, it spoke to the water bearer one day by the stream. "I am ashamed of myself, and I want to apologize to you. I have been able to deliver only half my load because this crack in my side causes water to leak out all the way back to your house. Because of my flaws, you have to do all of this work, and you don't get full value from your efforts," the pot said. The bearer said to the pot, "Did you notice that there were flowers only on your side of the path, but not on the other pot's side? That's because I have always known about your flaw, and I planted flower seeds your side of the path, and every day while we walk back, you've watered them. For two years I have been able to pick these beautiful flowers to decorate the table. Without you being just the way you are, there would not be this beauty to grace the house."

Synapse - Winter 2003 Edition Page 8

Moral: Each of us has our own unique flaws. We're all cracked pots, but it's the cracks and flaws we each have that make

our lives together so very interesting and rewarding.

On Being Differently Abled

Contributed by Galen Hekhuis NpD, JFR, GWA Sometimes I think there is too much emphasis given to the "I can't" rather than the "I can" aspects of this disease. Without being too Pollyanna-ish or "if life gives you lemons, make lemonade" (I think wine might be more fun anyway), there is something to be said about considering ourselves "differently abled" rather than "disabled. There are positive things about this viewpoint. In my case, it was kayaking. Granted, I can't walk too well anymore, but my arms still work pretty well, so I paddle. I'm not going to see the Mohave Desert or anything that way, but it is amazing how many other things have some kind of water access. You sit down and don't have to do any walking. I wear a life jacket (personal floatation device) so I don't even have to swim if I go in the water, but I never have even come close to going in the water. *I doubt if I would ever have given boating a second thought if hiking had still been an option.*

Exercise Video Recommended by Julia Walker Wellness in Motion is a video I recommend for PLSers. Contact: Betsy Lindsey, P.T. University of Arizona Dept. of Neurology 1501 N. Campbell Ave. Tucson, AZ 85724

Make out the check for \$6.00 to University of Arizona.

The Smile Starter

Contributed by Jane Langdon

Smiling is infectious, You catch it like the flu. When someone smiled at me today, I started smiling too.

I passed around the corner, and someone saw my grin. When he smiled I realized, I'd passed it on to him.

I thought about that smile, then I realized its worth. A single smile just like mine, could travel 'round the earth.

So, if you feel a smile begin, don't leave it undetected. Let's start an epidemic quick, and get the world infected!

Stretching exercises from the Mount Sinai School of Medicine Contributed by Dolores Carron **NECK**--1) look up to the ceiling, then extend your chin downward toward your chest, 2) tilt your head sideways, moving the right ear toward the right shoulder, then tilt your left ear toward your left shoulder, 3) rotate your head to the right, then to the left. **SHOULDER**--put your hand, thumb pointing up, in the middle of your lower back and raise your hand toward your shoulder blades, then repeat with the other arm. **ELBOW**--starting with one arm outstretched, draw your hand toward your shoulder and then straighten it again, then

repeat with the other arm. **WRIST**--use your index finger as a pointer to spell the capital letters of the alphabet, then repeat with the other wrist. **LOWER BACK**--1) from a

standing position, bend down slowly and reach down toward your toes (don't overextend or bounce in an attempt to touch your toes), 2) straighten up, place your open

Synapse - Winter 2003 Edition Page 9

palms on your buttocks, and arch backward slightly, 3) rest your hands on your hips and slowly bend sideways, while sliding your left hand, with palm open, down your left thigh toward your knee and repeat on the right side. **HIPS**--1) while standing, hold onto a chair or table with your left hand to brace yourself and bring your right knee straight up, then lower it, 2) now lift the knee again and gradually lower and sweep the leg backwards without letting your foot touch the ground, and return to standing position, 3) switch position and repeat with the left leg. **KNEE**--while sitting, extend your right leg fully, then repeat with the left leg.

ANKLE--using your big toe as a pointer, spell the capital letters of the alphabet, then repeat with the opposite leg. ****GENERAL RECOMMENDATIONS: start low, go slow, performing each exercise a few times only and then increase to a comfortable number or repetitions. Don't force it. Back off if you feel pain. Range of motion decreases if you don't use your major joints. With disuse, muscles and tendons contract and make the joints feel stiff. The longer the joint remains fixed, the harder it is to reverse the effect.

EVENTS

Autumn in Carolina 2 Contributed by

Don Wilson Gatherings of PLSers and caregivers began small a few years ago with lunch at Fudrucker's with Joe Alberstadt in Vienna, Virginia. Gatherings have been held in New Hampshire, Connecticut, Ohio, West Virginia, Tennessee and North Carolina. Results of these have been positive. PLSers and caregivers have been able to meet and learn that they are not alone, to exchange ideas and "tricks of the trade", and to form a foundation of support, likely the most important function. With PLS in common, and sometimes PLS- FRIENDS, when a few come in contact, it is as if all had been long-time friends. That was the case when the second Autumn in Carolina hosted by Don and Bettie Jo Wilson began on October 11, 2002. Participants began arriving in Winston-Salem in the early afternoon. By early evening Martin* and Mary Ann Beckner (Wilmington, NC); Pat* and Buzzy Croom (Garner, NC); Ronnie* Grove (Beckley Springs, WV) along with friend Carla Fike; Vaughn* and Neal Hickman (Knoxville, TN); David* and Lois Lehman (Kidron, OH); Jeanne Ann* and Chris Neff with daughter Julie and her friend, Allison Tucker, were joining Don and Bettie Jo, taking over a corner of the Mayflower Restaurant in Rural Hall, NC. (Names marked * are PLSers.) By mid-morning Saturday, Donna* and Lee Isenhour (Concord, NC); Don* and Pat

Penny (Buies Creek, NC); and Jean Mills* with sister Doris White and caregiver Mable Lee (Greensboro, NC) arrived to complete the gathering. Conversations spring up immediately all over the room. It is nearly uncanny how easy it is to talk to total strangers when the common thread is PLS. Doug Somers was the speaker for the morning session. Doug has worked as a rehab specialist, a salesman for assistive equipment, and is presently a teacher at Horizons Residential Care Facility, working with severely handicapped children. He spoke on the importance of proper lifting techniques in moving and transferring individuals. He noted that nearly all PLSers

attending already were using some assistance in the way of canes, wheelchairs or power chairs, and urged everyone to make use of such equipment before chancing a fall from which recovery would be extended. Doug also spoke on different types of lifting equipment presently available. The bright sunshine drove the temperature to the low 80's, which afforded an opportunity for everyone to check out different vehicles from fully converted mini-vans to standard vans with rear loaders for power chairs or scooters. Jeanne Ann, Jean and Pat decided that the fastest power chair/scooter should be recognized so they lined up for a race in the parking lot. Jean pulled a neat

Synapse - Winter 2003 Edition Page 10
maneuver on Jeanne Ann cutting her off, and then raced to victory, grinning ear to ear. Don and Bettie Jo teamed to do an afternoon presentation on adaptive gardening. Bettie Jo wrote the presentation, which Don read, and then Don demonstrated tools that work very well for adaptive gardening. Bettie Jo presented each PLSer with a plant that she had nurtured, in a clay pot that she had painted then added sand and glitter while the paint was still wet. The afternoon began to wind down and some good-by's were said to those returning home. Those remaining in town overnight shared an evening meal, and a few folks were given an opportunity to sample Lexington style barbecue and barbecue slaw. Again, everyone talked and laughed as if we were family, and indeed we had become so by the end of the weekend.

SPF TeamWalk Report Contributed by Mark Weber It's hard to believe it, but it's been over four months since TeamWalk. I'd like to recap that event, and update you on the progress of the Spastic Paraplegia Foundation. First, the SPF TeamWalk raised over an incredible \$95,000. Families came through in droves to further our mission of finding the cures for these two disorders (HSP & PLS). Thank you all so much for your tremendous show of support TeamWalk Chairman Shellie Fischer, her assistant Adam Roach, and a number of volunteers did an outstanding job organizing the weekend. The walk itself was superbly organized, Elvis was a scream, and Dr. Fink inspired all by walking with us. It doesn't get any better than this. See photos: <http://sp->

foundation.org/Teamwalk_Summary.htm So, what are we going to do with that money? Most of it will go to fund medical research on PLS and HSP. The SPF also provides information about the disorders and creates support initiatives. We are currently in the process of establishing the SPF medical research grant program so we can distribute funds to research. Once this process is in place, we'll be able to award our initial grants.

Then, we'll be working to raise funds year-round and we'll award grants annually. Our medical research grant program will work as follows: Later this winter, the SPF will announce the availability of research monies to study PLS and HSP. Interested scientists will reply with a short proposal. Our Medical Advisory Committee (currently being established) will review the proposals, and request more detailed proposals and budgets from the scientists who submitted the best proposals. The Committee will then review the more detailed proposals, and select the very best for funding. Then the grants will be awarded. Once again, I want to thank everyone who made the SPF's TeamWalk

such a success. It was our first major event and put the Spastic Paraplegia Foundation on the map. Remember, we are a volunteer-run agency, and we will move as quickly as our volunteer force (all of us) can take us. Working together, we will help find our cures. Count on it.

Upcoming Events in 2003 for the PLS and HSP Communities

<http://sp-foundation.org/events.htm>

***FEBRUARY* Southern Florida Patient Connection Luncheon** Sunday, February 2, 2003; 11 a.m.-3 p.m.; \$20 ea. Registration due by January 24 Holiday Inn Ft. Lauderdale Plantation/Sawgrass 1711 N. University Dr. Plantation, FL, 954-472-5600 Checks payable to: Spastic Paraplegia Foundation c/o Kathi Geisler 209 Park Rd. Chelmsford, MA 01824 KathiPro@aol.com

Vancouver, BC Conference Saturday, February 15, 2003 Featuring Dr. Siann Spacey, Neurogenetist, and other programs.

Synapse - Winter 2003 Edition Page 11
Best Western Richmond Inn Contact Jean Chambers at 604-990-1060 for more details
jchambers@intergate.bc.ca

***MARCH* Boston MA area Lunch Conference** March TBD Details to follow or contact Kathi Geisler 978- 256-2673
KathiPro@aol.com

Atlanta, GA Connections Coffee Saturday, March 8
Sheraton Gateway Hotel
Contact Kay Bell, (714)
342-6401
KAY_BELL@PRODIGY.NET

APRIL Berkeley Springs, West Virginia

Spring Fling Weekend – 3rd Annual PLS

Gathering April 4-6 Friday, April 4: Gather at Cacapon State Park Lodge for lunch at 12:00 PM on Friday. The speaker will be an MDA spokesperson, followed by free time and a group dinner. Saturday, April 5; We will meet by 9:00 AM at the Best Western Conference Room for our all day meeting followed by a group dinner. There will be lots of time during this relaxed weekend to get to know each other. Lodgings are available at Best Western – Berkeley Springs Inn 866-836-9330 at a group rate (20 rooms have been set aside). There are other options for lodgings, too. Check out <http://www.berkeleysprings.com/> to learn all about the town and its accommodations. There is no registration fee for Spring Fling, but please let Ronnie know your plans so she can make dinner reservations: Ronnie L. Grove, 13 Erin Lane, Berkeley Springs, WV 25411. frogrove@intrepid.net

Chicago IL Conference April 11 @ 5:30 p.m. Dinner, \$30 ea "Dinner With The Experts" featuring Teepu Siddique, M.D., world renowned researcher for

ALS/PLS/HSP. To register for the dinner, click here: [http://sp-](http://sp-foundation.org/pdf/ChicagoDinner.pdf)

[foundation.org/pdf/ChicagoDinner.pdf](http://sp-foundation.org/pdf/ChicagoDinner.pdf)

Registration deadline is April 1. The dinner will be held at: Holiday Inn Chicago-O'Hare Kennedy, Rosemont Room 8201 West Higgins Road, Kennedy/O'Hare Airport, CHICAGO, IL, 60631

<http://www.holidayinnchicagoil.com/hotels/chioh.html> Tel: 773 693 2323. It is 3 miles from the airport and there is shuttle service. There is free parking. Accessible.

Edison, NJ Outing and Lunch to the annual Abilities Expo April 12 Contact Sue Meholick at: (814) 653-8566 momofboysonly@yahoo.com.

Salt Lake City, Utah Patient Connections Dinner April 6 – Patient Connections Luncheon Contact Kathi Geisler, 978-256-2673 KathiPro@aol.com

MAY Austin, TX Patient Connections Dinner May TBD Contact Marlene Doolen, 512-331-1953 MDoolen512@aol.com

The two wisest remarks that were made to me as a caregiver when my husband was first diagnosed were: 1. Don't run too far down the road. In other words, don't waste any energy anticipating what's going to happen. Get practical details taken care of and then stay in the present. Promising research is going on. 2. A lot of people will be taking care of the patient. No one will be taking care of the caregiver. If you are going to be helpful, then you have to arrange for some caregiving for yourself. The big thing to remember is that you as a patient and caregiver are consumers. You have the right to have your questions answered as fully as possible. If your doctor seems rushed, ask him or her to schedule your appointment at the end of the workday when he or she will be able to give you as much time as you need. This is not a favor, but your right as a consumer of medical services. You are entitled to a physician who is going to help you manage your disease. Finally you shouldn't accept statements like these easily, if ever: 1. There's nothing more I can do for you. 2. It's impossible. We can't do that/we don't do that. 3. Your request has been denied. 4. That's never been done before.

10 Warning Signs Of Caregiver

Stress Contributed by Don Wilson 1.

DENIAL about the disease and its effects on the person who's been diagnosed. I know he/she is going to get better. 2. **ANGER** at the person with the disease: that no effective treatments or cures currently exist, and that people don't understand what's going on. If he/she calls me one more time, I'll Scream.

3. **SOCIAL WITHDRAWAL** from friends and activities that once brought pleasure. I don't care about getting together with the neighbors anymore. 4. **ANXIETY** about facing another day and what the future holds. What happens when he/she needs more care than I can provide? 5.

DEPRESSION begins to break your spirit

CAREGIVING

Hints and Tips From Barbara Dickinson, ALSA National Chapter Trustee Caregivers have questions and should expect physicians or other professionals to take the time to answer these questions completely. 1. What kind of equipment is available to help me and the patient? 2. Will the patient become incontinent? How should I handle bowel and bladder problems? 3. Will the patient have pain? Other discomfort? What can I do? 4. Do you know different ways I can communicate with the patient? 5. What should I do about nutrition? 6. How can I control choking and other swallowing difficulties? 7. How can I manage skin care? Mouth care? 8. What should I look for to indicate that the patient is in trouble? 9. What other problems can I expect to encounter? 10. HOW CAN I GET SOME HELP? Ideally, several professionals should be involved in the care of a patient with ALS/PLS: a neurologist, a pulmonologist, a physical therapist, an occupational therapist and a social worker. Consults with a nutritionist and a dentist can be helpful. The caregiver also needs some professional help: a mental health professional and/ or a member of the clergy are essential. Support groups are also

often very helpful. Most important is that the caregiver MUST find some way to get some respite. The two smartest things that were said to my husband when he was first diagnosed were: 1. Don't read any statistics. This disease is now your personal

Synapse - Winter 2003 Edition Page 13

and affects your ability to cope. I don't care anymore. **6. EXHAUSTION** makes it nearly impossible to complete necessary daily tasks. I'm too tired for this. **7. SLEEPLESSNESS** caused by a never-ending list of concerns. What if he/she falls and is seriously injured? **8. IRRITABILITY** leads to moodiness and triggers negative responses and reactions. Leave me alone! **9. LACK OF CONCENTRATION** makes it difficult to perform familiar tasks. I was so busy, I forgot we had an appointment, **10. HEALTH PROBLEMS** begin to take their toll, both mentally and physically. I can't remember the last time I felt good.

On Being a Caregiver Contributed by Doug Brand In a nutshell, it's very difficult and often heartbreaking being a spouse of a PLSer and seeing how your partner struggles. Nothing in life can prepare a person to be a "caregiver". It's totally foreign. With the sincerest intent to help, common sense collides with emotion, thus blurring the correct course of action. I experienced this in the very beginning of Flora's decline. I jumped in with both feet, determined to make life safe, secure and easy for her while she worked through the mental and emotional

possession. It will take its own course with you. 2. Don't concentrate on the big picture; it's out of your control. Take as much pleasure as you possibly can in every moment of your life. The small, sweet things will be your consolation.

stain of acceptance. And, I must admit, it was no easy job. Laundry, meal planning and prep, house cleaning and a myriad of other activities she lovingly performed so well, were now my responsibility...or so I thought. It wasn't until she exclaimed "enough is enough", that I realized I was doing more harm than good. PLS was eroding her self esteem and independence fast enough, without me hurrying it along. Flora made it very clear that if and when she needed help, she'd ask for it. My mission in the process was relegated to remaining on the periphery of her world, thus allowing her to take on tasks and challenges at her own pace and in her own time.

This was very difficult to accept in the beginning, but it delivered a very needed wake up call. Over the years, I have marveled at her accomplishments and determination not to give up ground. Be assured, anything Flora has to give up comes with claw marks! I have come to understand that spouses need to come to terms with the givens of the disease: (A) Your partner has it (B) It's not going to go away (C) At present there is not cure (D) It most likely will get worse with time. As harsh as it may seem, those are the cold hard facts. We also need to admit to our

limitations. We can't relieve the pain, straighten the spastic gait, clarify the speech, steady the tremors or have a positive effect on any of the many other conditions of PLS. So, where does that leave us? We need to be there for them and "on call" when needed. We need to exhibit restraint and understanding when tensions cause irrational behavior or cutting words. We need to show strength when we see them struggle while our instincts tell us to take control. We need to see that no one or set of circumstances

compromise their dignity and rights. We need to be there. Cultivating an atmosphere that breeds self pity is the last thing PLS patients need to surround themselves with. That can only lead to depression, despair and surrender. These are brave, courageous people who God has chosen to challenge. That fact alone makes them very special in my eyes. We have been singled out as well to help them with those challenges by moving forward one day at a time. Panic and impulsive thinking do more harm than good.

And the Winner Is. . . . Jeanne Ann Neff is pictured here with Santa. She was the winner of this wonderful Santa, which was crafted by Ronnie Grove. Ronnie also organized and conducted the raffle which netted \$631 for PLS!! Congratulations to Jeanne Ann and many thanks to Ronnie!

Board Member Sought for SP

Foundation The Spastic Paraplegia Foundation Board has been structured to include one PLS person. As you know, the SP Foundation raises money for both HSP and PLS. This PLS member would agree to attend Board meetings two times a year at his/her personal expense. We need representation on this Board. Anyone interested in finding out more details about this position, and possibly volunteering for it please contact: Mark Weber 11 Pepper Pond Rd. Sherman, CT 06784

