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2005**

National - Ohio October 2 Northeast September 11

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Autumn 2005

**Serving the Primary Lateral Sclerosis Community
since 1997 Welcoming the SP Foundation since 2003**

and Carrol Doolen at the Red Lobster. We had a great meal and visit. Marlene was one of the founding board members of the SPF and it was indeed a pleasure to meet her. On Saturday, it was very nice to be able to place faces with e-mail contacts and also to visit with old friends at the same time. After visiting a while, we walked and rolled along the beautiful Legacy Trail. My daughter led the walk wearing her roller blades because,

when a cure is found, I will be roller blading right along side of her, holding her hand. Norman Regional Hospital brought out lunch for us to enjoy after the walk. They also sent Chaplin, Mike Bumgarner to lead us in a very informative discussion about depression. His discussion was based on our comments to him about depression related to a variety of situations including loss of jobs due to disability, losing the

ability to do things we used to do and being dependent on someone else. His main point was to find a sounding board, an impartial person you can talk with about how you feel and what you are going through. Several of us met at the Santa Fe Grill that evening and enjoyed talking and eating together. We then went to the hotel guest area and continued to visit until about 10:30. All in all, it was a great weekend and an experience we will never forget. **SP National**

Conference and TeamWalk Columbus, OH September 30-October 2 Contributed by Thurza Campbell, editor The 5th annual SP National Conference and TeamWalk was masterfully organized by Margie and Paul Brockman. Their energy, enthusiasm and youth reached beyond our patient community to include the wider community in the weekend. The event began Friday evening with, casual get acquainted time, then a buffet followed by introductions and questions to the SP Board members. The weekend full of special activities for kids began this evening. Saturday was the National Conference, attended by 112 people, our largest yet! **National**

Conference Speakers *Ed. Note: The paragraphs below are my attempt to capture some of the points made in each talk. The talks were videotaped. Please watch for the announcement of their availability so you can see and hear the complete talks.*

**EVENT
REPORTS**

SP Patient Connection Luncheon Austin, Texas. Saturday, September 3, 2005
Contributed by Marlene Doolen The fourth

annual Austin Patient Connection was held today. "We are the Experts" was the theme. After a tasty lunch, we held a round table discussion about a variety of subjects dealing with PLS and HSP. Before the discussions, a Certificate of Appreciation was presented to Mike Schultz representing the Austin Windsurf Club (AWC). For the past four years, the AWC has designated the Spastic Paraplegia Foundation (SPF) to receive the funds generated from the club's "Learn to Windsurf Day Festival". It was good to see returning Austin Patient Connections participants. One couple drove in from Houston. Some new things were earned by everyone as we each shared information.

New England TeamWalk Newport, Rhode Island. September 11, 2005 Contributed by Kathi Geisler The first 2005 TeamWalk took place today in Newport, RI. A perfectly glorious fall day was enjoyed by all. We had 32 people from MA, CT, NH and NY. Some of us got to the picnic area on the wharf early so we could make claim to a bunch of picnic tables. By 11:30, all of us had arrived and were having a great time connecting with old friends and meeting some new ones. Some walked before lunch, most afterwards. We had a Subway lunch at 12:00 with more socializing and then began our TeamWalk at 1:00. I think we all looked pretty smashing in our TeamWalk T-shirts!

Mid America TeamWalk Norman, Oklahoma. September 17, 2005 Contributed by Mark Dvorak We had a wonderful time at the Mid America TeamWalk and Connection

for Our Cures held in Norman, Oklahoma. The Event was attended by 32 people and they came from these various cities and states: Austin, Allen, and Bulverde, Texas;

Topeka, Kansas; Norman, Purcell, Oklahoma City, Stillwater, Fairmont, and Tulsa, Oklahoma. On Friday evening, I enjoyed meeting Marlene

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disorder? SOD1 mice and ALS mice have already advanced understanding of the processes of the disease, e.g. how proteins malfunction. Environmental factors may also play a part in disease development. Both the disorders PLS and HSP are disorders of the same systems – upper motor neurons.

Understanding the process of transport of messages from brain through the spinal cord is the mystery needed to be solved in both. Nailah Siddique explained that one person has been hired part time at NWU using the money from the SP grant to collect and organize samples and data. As a result, they are making progress collecting data.

The number of patients in data base has doubled: 152 patients of these, 22 had both parents living (the best kind of sample); 450 community controls with no disease have also given samples. **Spasticity**

Management and Botulinum Toxin Dr. William Pease With slides, a talk and answering questions, Dr. Pease explained how the leg muscles are impacted by HSP. Spasticity is caused by over- active stretch reflex. The muscle fails to relax when it should; an antagonistic interference of opposite muscles. He explained how surgery is an option for some, and periodic shots of Botulinum Toxin are effective for many. Each series of injections can last several months. **Dealing with Disability**

Stacy James Stacy was paralyzed in a

swimming pool accident at age 19. Through rigorous therapy she has reclaimed 25% use of her arms, and can actually walk with crutches. She told of her gold medal Olympic wins, and of her marathon participations. In her inspiring talk, she shared with the audience her five tools to cope with disability: 1.Attitude- a positive attitude will let you triumph 2.Find your significance – for Stacy her significance is through Christ and God. Find who you are on the inside. 3.Courage – being afraid but doing something anyway. 4.Perseverance – don't give up on your dreams. She completed her first marathon in 11 1/2 hours!

Research Advances in HSP Dr. John Fink HSP is a group of conditions. HSP is genetic, it is usually progressive, and usually only affects the body from the waist down. At least 29 different genes can cause HSP. It affects axons in the spinal cord. Motor and sensory nerves are involved. Some of the genes are dominantly inherited, others recessively. A dominant gene only needs to be in one parent for a child to have a 50% chance of inheritance. Even if both parents have recessive genes the chances of HSP developing is low risk. There are many variables as to age of onset and how the disease presents. There are complicated and uncomplicated forms. Complicated can involve muscle wasting. Often very early onset childhood HSP does not progress.

Genetic testing is available for some of the dominantly inherited forms of HSP. There may be other modifying environmental and/or genetic factors which explain the variability. Stretching and strengthening exercises are important as part of a lifestyle regime, to maintain cardiovascular health. Genetic testing is not part of the diagnostic process, but can be used to confirm. Dr. Fink created an analogy of a road between two cities, describing various cars and cargo in the vehicles to explain motor and sensory nerve dysfunction. **Insight into the Syndrome of PLS** Dr. Teepu Siddique Dr. Siddique began his talk by reviewing the history of the study of ALS and PLS. There has always been difficulty in determining exactly what PLS is. The diagnosis takes

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5. Have faith in yourself – we disabled are people first. She has a personal relationship with God, and faith in Christ. *Ed. Note: There were three other break-out sessions, two led by Paul Brockman on finding a medical supplier and selecting proper equipment, and one led by Robert Weld on fundraising. Unfortunately I could only be in one session at a time, so please rely on the upcoming videotape for the content of these sessions.* Dinner or a Columbus Crew soccer game rounded out the day. Sunday was the TeamWalk. Another 123 people joined the 112 from the Conference to hear the mayor of Columbus proclaim the day HSP/PLS Day! We rolled, strolled and walked down High St. to beautiful Goddard Park. (I brought up the rear. . . but I finished!) We were greeted by a band, life sized cartoon characters, drinks, snacks,

three to five years. The brain/spinal system of humans are much more complex than that of mouse, meaning difficulties in using a mouse model for study. If you have PLS or HSP you are still susceptible other diseases, such as cancer or cardiovascular disease. To think that stem cells could be put into the motor cortex and have them go into the spinal cord to replace damaged axons is a unrealistic. He went on to say he feels that research should focus on learning the biology of the disease and then realistic ways to repair and reverse the symptoms. Some forms are inherited. Analysis of the human genome will help determine what genes might cause PLS. Technology is speeding up genetic analysis; if we don't know what's wrong, how can we fix a

and activities continuing for the kids. All too soon it ended, and everyone said, 'good-bye 'til next year'.

NW Connection & TeamWalk Pleasanton, CA October 7-8, 2005 Contributed by Linda Gentner Friday evening, 10 couples gathered for dinner at the Hilton where the out-of-towners were staying. The couples were: Angela & Jim Dixon from Arizona, Sue & John Dutton from Placerville, CA, Don & Gayle Gould from Oregon and Jane & John Mitchell from Escondido, CA and Linda & Craig Gentner who coordinated the weekend festivities. Conversations ranged from typical PLS/HSP discussions to how couples met and to square dancing! Saturday, 60 people were in attendance. We had 13 people with PLS and HSP, a good turnout for our SPF Community. We

gathered for registration, and Continental breakfast at the church before we started out on our Walk through downtown Pleasanton. Most walkers seemed to enjoy the walk through quaint downtown Pleasanton and especially the Farmer's Market. Then we went back to the church for lunch. After lunch, Dr. Lomen-Hoerth spoke briefly about PLS and HSP and current research findings about familial ALS which was very exciting. She was available for one-on-one questions after we broke for the day. Following

was the announcing of the raffle prizes. We ended up with lots of raffle prizes thanks mainly to my daughter-in-law's father - 4 cases of wine! also dinners for local restaurants and some golf packages and 3 fun holiday decorations and an educational toy . We had the quilt on display and we sold lots of tickets for that too. People are so impressed with the quality workmanship. All in all, I think it was a very nice day and good for SPF.

Autumn in Carolina IV Winston-Salem, NC October 8, 2005 Contributed by Don Wilson
Folks began to arrive in Winston-Salem late Friday afternoon. It had been raining for two full days causing many attendees to have major problems getting there. Saturday's meeting began with Krispy Kreme doughnuts and Orange Glazed Breakfast Buns and plenty to time for participants to

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being passed by The Shot. Cece held on for the win. Bettie "Green Thumb" Wilson, Barbara "Stormy Neely and Annette "The Blond Bombshell" (also known as "The

meet each other. Mel and Jill Ellison of Learning Resources brought Eye Response equipment along with other types of special communications gear. After their presentation, Bettie Jo Wilson and Barbara Neely had a test drive. Tracy Wilson gave a presentation on *How to Select a Massage Therapist* and described different types of massage. She was available to give reflexology massage in the afternoon. After a catered box lunch, Bettie Jo presented *Adaptive Gardening*. She made the presentation with the aid of a laptop computer and Text Aloud software. A Power Point slide show completed the presentation. Bettie Jo also had crafts and cards that she had made, and also seeds saved from heirloom plants. Racing time was approaching. Scooter and power chair drivers donned uniforms from Nextel teams of Dale Jarrett, Ricky Rudd, Elliott Sadler, and Ryan Newman along with hats from Kasey Kahne. Everyone then go a close up look at a NASCAR Modified racer, which is actually pound for pound the more powerful car racing under the NASCAR banner. The rain stopped, and the Autumn in Carolina races began. Cece "The Shot" Russell, Ronnie "Frogger" Grove and Martin "Mad Dog" Martin lined up for the two-lap scooter race. Mad Dog was the defending champion, but fell behind at the start. Frogger led most of the first lap before

Prez") Lockwood lined up for the powerchair race. Green Thumb went a little off course and The Blond Bombshell jumped out to a long lead, winning handily. Stormy Neely

blamed her loss on her crew chief (husband Arthur) claiming that he forgot to charge her chair overnight. "The Shot" Russell and "The Blond Bombshell" Lockwood raced for the Championship, with Annette easily outrunning Cece. Trophies were presented to Cece and Annette. Annette also received the Championship plaque and a special candy bouquet. Cristina Lockwood squeezed into the modified racecar for a short ride. She loved the sound and feel of the powerful engine. The group gathered for dinner at Hill's Lexington Barbeque, after which those brave enough to stay up a little longer met in the hospitality room for stories, jokes and conversation. Brenda Asbury came alone from Texas and was a great help to me. The trip home on Sunday was made in much better weather. Everyone seemed to enjoy himself or herself, and Bettie Jo and I will try to hold Autumn in Carolina V next year.

SP FOUNDATION

A Letter from our SP President Our fifth annual TeamWalk took place in Columbus, Ohio on October 2, 2005. It was a spectacular event beginning with the Mayor's office declaring October 2, 2005 HSP/PLS Day and ending at Goodale Park with cartoon characters and finger printing for the children. Over 200 people participated in the walk. Prior to the TeamWalk, the National Conference was held. We had the opportunity to hear the latest from Dr. Fink, Dr. Siddique and Dr. Pease. Attendees also had the opportunity

to see the latest Adaptive Equipment available. In addition to the National TeamWalk, seven Local TeamWalks are being held. Some have already taken place and there are few more to go. The Board of Directors has elected to invest in a software solution to manage our mailing list and database. As we are getting this in place, some things have been delayed, such as Thank You letters. Your patience and understanding is greatly appreciated. Once the software is in place, it will result in a much smoother and efficient operation. If you haven't purchased raffle tickets yet for the "Turning Leaves" quilt, you need to do so soon. <http://www.sp-foundation.org/quilt.htm> The drawing will be held in early November. The quilt was on display in Columbus and it is beautiful. My sincerest wishes to everyone for a memorable Holiday season and looking forward to a breakthrough in research that will eradicate PLS and HSP!.
Annette Lockwood The Spastic Paraplegia Foundation 2005 Research Awards Announced Compiled by Mark Weber, Chair for Grants Thank you so much to each and every one of you who made this possible. It is just incredible that this small community has joined hands and worked real hard to help scientists find our cure. And we've had fun doing it---meeting new friends---and learning how others experience and deal with these disorders. And actually hearing and meeting the top scientists and physicians who are working to find our cures. Grant I "Analysis of Spastin and Atlastin in the cell biology of neurons". Brett Peter Luring, M.D., Ph.D., of the Columbia University College of Physicians

and Surgeons in New York City. Dr. Luring was awarded a two year grant totaling \$96,701 for his project. Dr. Luring published an article in the Journal of Cell Biology this past February entitled "Linking axonal degeneration to microtubule remodeling by Spastin-mediated microtubule severing." Grant II "Revealing

the mechanisms underlying ALS2, a form of hereditary spastic paraplegia, using ALS -/- mice". Michael R. Hayden, M.D., Ph.D. and Blair R. Leavitt, Ph.D. of the Center for Molecular Medicine and Therapeutics at the University of British Columbia, in Vancouver, British Columbia, Canada.

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Drs. Hayden and Leavitt were awarded a two year grant totaling \$149,896 for their project. Dr. Hayden has been researching the ALS2/Alsin gene since the late 1990's. He is co-author of seven scientific journals about ALS2/Alsin, the most recent of which was published in March of this year in the journal "Neurobiology of Disease", and included Dr. Leavitt as another co-author. Note that some researchers argue that the disorder caused by mutations in the ALS2 gene is PLS, not HSP. Drs. Hayden and Leavitt, and a team in Italy call it HSP. Whether you call it HSP or PLS, this research will obviously help both disorders. Grant III "Molecular genetic controls over the development, connections, and survival of upper motor neurons". Jeffrey Macklis, M.D., the Director of the Massachusetts General Hospital--Harvard Medical School Center for Nervous System Repair, in Boston. Dr. Macklis was awarded a two year grant totaling \$121,660 for his project. Dr. Macklis published several papers in the past 12 months dealing with neuronal repair and replacement therapies. In particular, see "The repair of complex neuronal circuitry by transplanted and endogenous precursors" in NeuroRx. 2004 Oct;1(4):452-71. Dr. Macklis

has discovered that precursor cells already present in mouse brains can be induced to produce new motor neurons and then grow axons that project down the spinal cord. As recently as five years ago, scientific dogma insisted that this was completely impossible. The long-term goal of Dr. Macklis and his team is to repair and generate new motor neurons in human brains without transplanting any cells into the brain. He has discovered genetic "switches" that can activate stem cells that are already present in the brain and turn them into upper motor neurons, and then induce them to grow axons that project down the spinal cord and connect to lower motor neurons. Grant IV "Invertebrate model of hereditary spastic paraplegia" Peter Hedera, M.D., of the Department of Neurology at Vanderbilt University, in Nashville, Tennessee. Dr. Hedera was awarded a two year grant totaling \$90,000 for his project .

Dr. Hedera has published at least eight articles in various medical journals about HSP--most with his colleague John Fink, M.D. when Dr. Hedera was at the University of Michigan prior to 2002. Another article is currently in press and is due out soon. Grant V "Mechanistic interactions among hereditary

spastic paraplegia genes” Kendall S. Broadie, Ph.D., a Professor of Neurobiology at Vanderbilt University, in Nashville, Tennessee. Dr. Broadie was awarded a one year grant totaling \$54,673 for his project. His grant may be extended for an additional year. (We ran out of money and couldn't fund Dr. Broadie's work for a full two years as originally hoped. But if we raise enough this year, we will consider extending his grant for an additional year.) Dr. Broadie spoke about his research at the SPF's Connection earlier this year in Nashville, Tennessee. **Personal Reflections about SP History** Contributed by Mark Weber Kathi Geisler devoted over three years of her life to making the SPF happen. She traveled around the country to Patient Connections that she either organized by herself, or helped others to organize. She organized the first three annual TeamWalk/SPF Conferences that were our major source of revenues. (And she helped

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crucial. She also organized an annual SPF fundraiser in Texas each year--a Learn to Windsurf day. Frank Davis took over Marlene's responsibilities this year and is doing a superb job. He is also involved with our mailings--a huge job. Linda Gentner has also been a major force in SPF fundraising. In addition to spearheading the SPF cookbook work, she has organized a local TeamWalk in California again this year. Linda and her husband Craig have traveled to many SPF functions around the country over the years. Linda is also a valued member of our Board of Directors. Paul Brockman organized the National

with the local TeamWalks.) Kathi also created the SPF web site, which she maintains to this day. In short, Kathi has been the SPF dynamo. If you saw something happen related to the SPF, Kathi was there--if not out in front, then behind the scenes. Annette Lockwood is the SPF president. She brings her management talents as an executive with Exxon/Mobil to the SPF. In just 6 short months under Annette's leadership, the SPF has organized more fundraising events than ever before. And there are management initiatives in the works that we hope will bring the SPF to the next level. Annette also has spearheaded the SPF drive through the Exxon/Mobil Favorite Charities Campaign. Last year her work there raised \$27,500. Marlene Doolen was the SPF secretary for 3 years and kept our "back office" running. You never saw her in this role, but her work was

TeamWalk/SPF Conference in Columbus, Ohio to be held September 30th -- October 2nd. David Lewis is the Chief Financial Officer of a company in the Atlanta area. He is also the SPF Treasurer. In that role he handles all of the donations received by the SPF, maintains our financial books, and prepares our financial statements and IRS filings. His work is critical to our success. Chris Brocchini remains a large SPF fundraiser in California. There's nothing sexy about raising money--just a lot of hard work. Chris gets the job done, keeping those checks flowing in. Jean Chambers

represents the SPF in Canada. Jean is a valued member of our Board of Directors and is raising money for HSP/PLS research being carried out in Canada. Jean has already organized and held a local Teamwalk in West Vancouver, British Columbia, and, along with Linda Gentner, she is heading up our "Turning Leaves Quilt Raffle" project this year. Jim Sheorn joined our Board last year and has already organized an SPF Conference/Connection in his hometown of Nashville. He has also organized a local TeamWalk to be held this October. And there are so many others who have given of themselves to help this community through the SPF. Everyone involved is an SPF hero. They include Mark Dvorak, Betsy Baquet, and Bonnie Guzelf. Ronnie Grove held her annual Spring Fling this year--even though she was recovering from surgery. Ronnie is also spearheading the "Saving Pennies for SPF" campaign. Don Wilson continues to hold his fabulous, annual "Autumn in Carolina" event every October, along with his famous SAWCAR Race (Scooter and Wheel Chair Association of Racing) to bring people affected by these disorders together. Dolores Carron has organized a support group for PLSers and HSPers in the Connecticut area. Every year she also holds a patient Connection. The entire Milbourne Family (including Joan Heinicke, Jack Heinicke, Ruth Easterling, Rick Easterling, Randy Easterling, John Heinicke, Joe Heinicke, Sandie Heinicke, Lisa Lewald, Gail Easterling, Wendy Easterling, Lee Heinicke, Cindy Compher, Kevin Compher, LuAnn Webb, and John Webb) organized the annual Richard G.

Milbourne Memorial Golf Classic to raise money for the SPF. Harvey Mover has organized the "Loop for Life" motorcycle ride in Indianapolis, Indiana again this year. A portion of those monies goes to the SPF--\$29,000 last year. Doug Brand has donated his artistic and design talents and has designed many of the SPF promotional materials over the years. I have undoubtedly failed to mention others who have given of themselves to further our cause. I apologize in advance for my mistakes. The people who have done the work are the SPF heroes. I applaud you all. **Memorial Donations to SP Foundation** SP Foundation is happy to accept and properly acknowledge donations given in memory or in honor of friends and loved ones. You may feel moved to give a gift to SP to acknowledge a significant effort of a friend or loved one. Often a death needs to be memorialized, and the family does not have a favorite charity. You can make SP Foundation the recipient of memorials, by sending checks to the SP Treasurer, David Lewis, PO 1208, Fortson, GA 31808.

End of Year Donations of Appreciated Stock to SP Foundation Start planning now for end of the year donations to SP Foundation. Have you considered taking advantage of the opportunity to donate appreciated stock to SP Foundation? Donating appreciated stock is a bonus for both you the donor and the SP Foundation. You avoid capital gains tax, are credited with the charitable contribution at the appreciated value and SP sells

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the appreciated stock to add to our dollars for research. SP Treasurer David Lewis is making all necessary arrangements with the bank that has the SP Foundation account. We will post detailed instructions for you ASAP on the stock transfer process. You may also contact David directly at PO 1208, Fortson, GA 31808.

CAREGIVING

Ed. Note: November is National Caregiver Appreciation Month according to the MA Chapter ALSA. I hope the following articles and resources will affirm and support the caregivers of our patient community.

Four Messages for Caregivers Excerpted from National Family Caregivers Association and the National Alliance for Caregiving

Principle # 1 Choose to Take Charge of Your Life Obviously we cannot control everything that happens to us, or to our loved ones. But we can make active choices about how we are going to deal with the circumstances of our lives. We can choose to martyr ourselves in the interests of the loved one for whom we are caring, or we can set limits on what we as individuals can and cannot do without causing irrevocable damage to our own health or the other relationships in our lives. Knowing that circumstances change, including our own health and innate capabilities it is vital to learn to take charge of our lives.

Principle # 2 Love, Honor, and Value Yourself It is easy to lose your true self while being a family

caregiver. It is easy to lose your identity as an individual and that is why it is so important to cling to the core of your personality. Do the things that make you happy, that let you say: "I feel like 'me' when I am doing this or that, or I like 'me' when I am being this way." Caregiving so often keeps us off balance. It is easy to get lost in its physical and psychological maelstrom-the sadness, the frustration, the stress and strain on your body and your mind, the financial worries, the emotional pain. All the more reason to step out of the frame on a regular basis, and cultivate the other parts of yourself, to learn to give to yourself in addition to giving to others.

Principle # 3 Seek, Accept and At Times Demand Help Family caregivers regularly seek supportive relationships with other caregivers, knowing they can provide the emotional sustenance needed during difficult times. Support doesn't change the circumstances under which you are living That's the work of a different word-and that word is HELP. How do you get the help you need, and where do you find it? Many people think asking for help is a sign of weakness. Asking for help is a sign of strength because it acknowledges the difficulty of the situation at hand. It is a sign of strength because it requires putting pride aside and acting in the best interest of your loved one and yourself.

Principle # 4 Stand Up and Be Counted It is important to acknowledge your role as a family caregiver because that bonds you with the millions of other family caregivers in America who share many of the same worries and

concerns that you have. **Well Spouse Association** <http://www.wellspouse.org/> 1-800-838-0879 Illness and accident attack without warning and can happen to anyone. No two people are living in the same situation and no two illnesses exact the same toll. All well spouses face similar problems of anger, guilt, fear, isolation, grief, and financial threat whether they are full-time caregivers or whether their partners have only moderately disabling illnesses. Well Spouse is a national, not for profit membership organization which gives support to wives, husbands, and partners of the chronically ill and/or disabled. Well

Spouse support groups meet monthly. WS support groups are also an excellent source for information on a wide-range of practical issues facing spousal caregivers. Well Spouse support groups exist or are being formed in many areas of the country. Well Spouse publishes a bi-monthly newsletter, "*Mainstay*". Our organization also helps set up letter writing "round robins" to help those members who can't get out too often break the bonds of isolation. Well Spouse also works to make health care professionals and the general public aware of the great difficulties caregivers face every day. Membership fees, individual donations,

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foundation and corporate grants, bequests, endowments and commemorative gifts support our organization. Donations are tax deductible to the extent provided by law. To become a Well Spouse member complete the application, print it out, and mail it in with your dues payment. You can write to us at Well Spouse Association, 63 West Main Street -- Suite H, Freehold, NJ 07728, or call us at 1-800- 838-0879 *When one is sick...two need help.* **National Caregiving Alliance** Established in 1996, The National Alliance for Caregiving is a non-profit coalition of national organizations focusing on issues of family caregiving. Alliance members include grassroots organizations, professional associations, service organizations, disease-specific organizations, a government agency, and corporations. The Alliance was created to conduct research, do policy analysis, develop national programs and increase

public awareness of family caregiving issues. If you have questions on caregiving, would like more information on the Alliance and its programs, or would like to learn how to order paper copies of Alliance publications, please contact: National Alliance for Caregiving 4720 Montgomery Lane, 5th Floor Bethesda, MD 20814

The National Family Caregivers

Association 1-800-896-3650 The National Family Caregivers Association (NFCA) supports, empowers, educates, and speaks up for the more than 50 million Americans who care for a chronically ill, aged, or disabled loved one. NFCA reaches across the boundaries of different diagnoses, different relationships and different life stages to address the common needs and concerns of all family caregivers. We are committed to improving the overall quality of life of caregiving families and minimizing the

disparities between family caregivers and non-caregivers. We envision an America in which the contributions of family caregivers to the welfare of their loved ones and to society is recognized and supported by all societal sectors. In short, we envision an America in which the work of family caregiving is recognized and rewarded in a manner commensurate with its importance.

10400 Connecticut Avenue, Suite
500 Kensington, MD 20895-3944
Toll Free:1-800-896-3650 Phone:
301-942-6430 Fax: 301-942-2302

LIVING WITH HSP/PLS

Free Dasher Software Dasher is an information-efficient text-entry interface, driven by natural continuous pointing gestures. Dasher is a competitive text-entry system wherever a full-size keyboard cannot be used - for example, on a palmtop computer; on a wearable computer; when operating a computer one-handed, by joystick, touchscreen, trackball, or mouse; when operating a computer with zero hands (i.e., by head-mouse or by eyetracker). The eyetracking version of Dasher allows an experienced user to write text as fast as normal handwriting - 25 words per minute;

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1) "Take It All Off" from the MDA Quest Magazine, written by Armand Legault who was a tax auditor for the State of CT for more than 30 years and is a disabled person himself, so he knows the issues from both sides of the desk (he was a speaker at our last two CT Connections) 2) a follow up

using a mouse, experienced users can write at 39 words per minute . Dasher is free!

Exercise Equipment for Disabled

1-888-298-9922 Daily exercise has long been recognized as an essential component of health. The Ex N' Flex Leg Machine has been designed for those who have little or no control over lower body movement. In passive mode, the legs are rotated by the electric motor in an orbital motion similar to cycling. This prevents atrophy of joints, muscles & tendons. In active mode you can assist the motor to move the pedals with your own muscles.

Adaptive Golf Accessories Phone

1-319.268.0939 Upright Golf products are designed to reduce the amount of repetitive bending that's traditionally required when playing golf. They believe that golf is truly a great game. They encourage you to play it, and to play it UPRIGHT.

Disability Benefits Contributed by

Dolores Carron I know from personal experience that shower modification cost is tax deductible--I did that this past year. Links that I'd recommend are:

'Take It All Off 2" article in MDA Quest Magazine 3) the IRS publication document 502 entitled Medical and Dental Expenses (Including the Health Coverage Tax Credit)This document gets updated each year. These references are good starting points for the thought process of analyzing

your particular issues. As Armand emphasized in his presentations, any modifications of home or vehicle that are necessary because of medical conditions are tax deductible. Your tax advisor may help with other suggestions, as well. Not only the initial costs of such projects deductible, but also the ongoing expenses incurred by their maintenance and use--for instance, the additional electricity to run a bi-pap machine would be tax deductible. I recently added a therapeutic heated pool addition to my home so I keep track of the extra cost of gas to heat the water, chemicals to maintain the water, etc. for yearly ongoing deductions.

Stair Lift Summit Stair Lifts. 1-888-252-2205

The AC Powered Summit Stair Lift is a stair lift designed to fit into your home. The lift usually takes only a few hours to be installed in your home. **Passengers with Disabilities** The Air Carrier Access Act prohibits discrimination on the basis of disability in air travel and requires U.S. air carriers to accommodate the needs of passengers with disabilities. The Department of Transportation has a rule defining the rights of passengers and the obligations of air carriers under this law. The following is a summary of the main points of the DOT rule (Title 14 CFR, Part 382).

Prohibition of Discriminatory Practices

Carriers may not refuse transportation to people on the basis of disability. Airlines may exclude anyone from a flight if carrying the person would be inimical to the safety of the flight. If a carrier excludes a person with a disability on safety

grounds, the carrier must provide a written explanation of the decision. Airlines may not require advance notice that a person with a disability is traveling. Carriers may require up to 48 hours' advance notice for certain accommodations that require preparation time (e.g., respirator hook-up, transportation of an electric wheelchair on an aircraft with less than 60 seats). Carriers may not limit the number of disabled persons on a flight. Carriers may not require a person with a disability to travel with an attendant, except in certain limited circumstances specified in the rule. If a disabled passenger and the carrier disagree about the need for an attendant, the airline can require the attendant, but cannot charge for the transportation of the attendant. Airlines may not keep anyone out of a seat on the basis of handicap, or require anyone to sit in a particular seat on the basis of handicap, except as an FAA safety rule requires. FAA's rule on exit row seating says that carriers may place in exit rows only persons who can perform a series of functions necessary in an emergency evacuation.

Accessibility of facilities New aircraft with 30 or more seats must have movable aisle armrests on half the aisle seats in the aircraft. "New aircraft" requirements apply to planes ordered after April 5, 1990 or delivered after April 5, 1992. No retrofit is required, although compliance with on-board wheelchair requirements (see below) became mandatory on April 5, 1992 regardless of the plane's age. As older planes are refurbished, required accessibility features (e.g., movable armrests) must be added. New widebody

(twin-aisle) aircraft must have accessible lavatories. New aircraft with 100 or more seats must have priority space for storing a passenger's folding wheelchair in the cabin. Aircraft with more than 60 seats and an accessible lavatory must have an on-board wheelchair, regardless of when the aircraft

was ordered or delivered. For flights on aircraft with more than 60 seats that do not have an accessible lavatory, carriers must place an on-board wheelchair on the flight if a passenger with a disability gives the airline 48 hours' notice that

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he or she can use an inaccessible lavatory but needs an on-board wheelchair to reach the lavatory. Airport facilities owned or operated by carriers must meet the same accessibility standards that apply to Federally-assisted airport operators. **Other Services and Accommodations** Airlines are required to provide assistance with boarding, deplaning and making connections. Assistance within the cabin is also required, but not extensive personal services. Ramps or mechanical lifts must be available for most aircraft with 19 through 30 seats at larger U.S. airports by December 1998, and at all U.S. airports with over 10,000 annual enplanements by December 2000. Disabled passengers' items stored in the cabin must conform to FAA rules on the stowage of carry-on baggage. Assistive devices do not count against any limit on the number of pieces of carry-on baggage. Wheelchairs (including collapsible battery-powered wheelchairs) and other assistive devices have priority for in-cabin storage space (including in closets) over other passengers' items brought on board at the same airport, if the passenger with a disability chooses to preboard. Wheelchairs and other assistive devices have priority over other items for storage in the baggage compartment. Carriers must

accept battery-powered wheelchairs, including the batteries, packaging the batteries in hazardous materials packages when necessary. The carrier provides the packaging. Carriers may not charge for providing accommodations required by the rule, such as hazardous materials packaging for batteries. However, they may charge for optional services such as oxygen. Other provisions concerning services and accommodations address treatment of mobility aids and assistive devices, passenger information, accommodations for persons with hearing impairments, security screening, communicable diseases and medical certificates, and service animals. Additional information may be obtained by contacting airconsumer@ost.dot.gov. Office of Aviation Enforcement and Proceedings 400 Seventh Street, SW, Room 4107

Washington, DC
20590

MEDICAL RESEARCH UPDATES

Reflections on Research Benefits for HSP and PLS Contributed by Mark Weber I firmly believe that research on HSP or PLS will directly help scientists understand---and ultimately cure---both disorders. Why?

Scientists need to understand the bio-chemical processes that are defective in HSP and PLS and that cause the symptoms that we struggle with daily. Once they know what is going wrong, they can discover cures. Uncomplicated HSP Both PLS and HSP share certain symptoms-- lower body symptoms. Work on the underlying cause of uncomplicated HSP (HSP with lower body symptoms only) will help scientists to understand the cause of those same symptoms in sporadic PLS. The Spastic Paraplegia Foundation has funded several studies dealing with the causes of uncomplicated HSP. Additional work on the genes and resulting proteins that cause uncomplicated HSP is on-going in a significant number of additional labs throughout the United States and Europe. Alsin Familial PLS caused by mutations in the ALS2/Alsin gene also causes HSP. (The

symptoms are the same, but some researchers classify the symptoms differently.) Research to understand the role of the Alsin protein in a neuron, will help to explain the defective cascade of events that results in sporadic PLS. SPF has funded two genetic studies – one for PLS, one for HSP. Research on Alsin has also been reported by labs in Chicago, Quebec, London, Tokyo, and Lyon (France). Troyer Syndrome PLS can affect one's ability to speak. Troyer syndrome is a complicated form of HSP that includes loss of speech as a symptom. Research on the cause of Troyer syndrome will also shed light on the bio-chemical cause of the loss of speech experienced by those with sporadic PLS. Research on understanding how defective Spartin protein causes Troyer syndrome is being done in England in at least one lab and in at least

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one lab in the United States. Silver Syndrome The onset of PLS can sometimes occur in the hands. Further, some cases of PLS show some lower motor neuron damage--although such damage is not significant. Silver syndrome is a complicated form of HSP that attacks upper motor neurons as well as selected lower motor neurons (causing muscle wasting in the hands). Research on understanding how Silver syndrome attacks the neurons that control the hands will also help to unravel the cause of PLS. Work to identify the gene responsible for Silver Syndrome is on-going in London. Summary The bottom line is that we are all in this together. Both PLS and

HSP selectively attack the longest upper motor neurons. Neurons that control speech are also attacked in PLS and by a form of HSP called Troyer syndrome. Neurons that control hand muscles are selectively attacked in Silver Syndrome and PLS. Although the tools to identify the causes of sporadic disorders such as PLS either don't exist or have failed to determine the cause of sporadic PLS, the tools of geneticists have found genes that cause a familial form of PLS (alsin) and complicated forms of HSP that share symptoms with PLS (the uncomplicated forms of HSP, as well as complicated forms such as Troyer Syndrome and Silver Syndrome). Scientists

are now going beyond the identification of disease causing genes to determine the functions of the proteins created by those genes. Undoubtedly sporadic PLS will be found to be caused by some defect in the same cascades of events that cause the various forms of HSP mentioned above. When the cures are discovered, won't it be great to know that we all helped? By raising the monies that you do every year, we fund research that is leading to our cures.

NIH Study Seeking Volunteers Title: Movement Related Cortical Potentials in Primary Lateral Sclerosis Number:

04-N-0017 Summary: This study will examine whether the motor cortex (the part of the brain that controls movement) works properly in patients with primary lateral sclerosis (PLS), a disorder in which voluntary movements are very slow. Healthy volunteers between 40 and 75 years of age and patients with ascending PLS (a subset of PLS) may be eligible for this study. Patients with ascending PLS have a slowing of finger-tapping movements that corresponds to a particular abnormality of certain neuronal (nerve cell) activity. Participants perform a finger-tapping reaction time exercise while brain wave activity (electroencephalography, or EEG) and muscle activity (electromyogram, or EMG) are measured. The subject is seated in front of a computer screen. A signal appears on the screen and the subject taps a key as quickly as possible in response to the signal. For the EEG, brain activity is recorded by placing electrodes (small metal

discs) on the scalp with an electrode cap or glue-like substance. A conductive gel is used to fill the space between the electrodes and the scalp to make sure there is good contact between them. The brain waves are recorded while the subject taps his or her fingers very slowly. For the surface EMG, electrodes filled with a conductive gel are taped to the skin. Participants also undergo magnetic resonance imaging (MRI). This test uses a strong magnetic field and radio waves to obtain images of the brain. During the procedure, the subject lies still on a table that can slide in and out of the scanner - a narrow metal cylinder. Scanning time varies from 20 minutes to 3 hours, with most scans lasting between 45 and 90 minutes. Subjects can communicate with the MRI staff at all times during the scan and can ask to be moved out of the machine at any time. Toll Free: 1-800-411-1222 301-594-9774 (local), 1-866-411-1010 (toll free) Fax: 301-480-9793

Research Snippets

New Locus for HSP Maps to Chromosome 1p31.1-1p21.1. SOURCE: Ann Neurol. 2005 Aug 29;58(3):423- 429

We have updated the clinical description of a large Scottish pedigree, in which patients were affected by spastic paraplegia complicated by hearing impairment and persistent vomiting due to hiatal hernia inherited as an autosomal

dominant trait. Using a genome-wide mapping approach, we identified a novel locus (SPG29) for this form of hereditary spastic paraplegia on chromosome 1p31.1-21.1 and narrowed it to 22.3cM between markers D1S2889 and D1S248. Sequencing of one candidate gene in the region (sorting nexin 7, SNX7), involved in several stages of intracellular trafficking and protein transport, showed no disease-causing mutations.

Transcutaneous Electrical Nerve Stimulation Versus Baclofen in Spasticity: Clinical and Electrophysiologic Comparison. Am J Phys Med Rehabil 2005;84:584-592.

OBJECTIVES:: Clinical and electrophysiologic comparison of the efficacy of transcutaneous electrical nerve stimulation (TENS) and oral baclofen in the treatment of spasticity. DESIGN:: Patients with spinal cord injury and spasticity were included in the study. Ten patients were assigned to oral baclofen and 11 to TENS groups. CONCLUSION:: TENS may be recommended as a supplement to medical treatment in the management of spasticity.

New Study Reveals Potential for Radical New Treatment of Paralysis and Brain Disease The study suggests wide-ranging applications; Findings could impact millions with spinal cord injuries, Alzheimer's disease, ALS, Parkinson's disease and other debilitating illnesses- The study, published in the Journal of Neuroscience Research, shows how a protein called KDI tri- peptide (KDI) can block the harmful effects of a substance called glutamate that

s present in all degenerative brain diseases and spinal cord injuries, causing permanent cell death and preventing the repair of damaged nerve connections. Glutamate is produced as part of the body's natural reaction to central nervous system damage. In the new study, researchers at the Brain Laboratory at the University of Helsinki and at the Johnnie B. Byrd, Sr., Alzheimer's Center & Research Institute in Tampa, FL show KDI to be a potent and wide-ranging blocker of glutamate's damaging chemical processes. It therefore has a tremendous ability to protect the brain and spinal cord from cell death and even enable regrowth. Human clinical trials are expected to begin as soon as next year. The Byrd

Institute is dedicated to helping and coordinating research, as well as building national and international collaborations. Contact Amy Giordano Director of Clinical Research 813- 866-1611.

Research in the ALSTDF Digest Stem Cells Researchers find how newborn neurons replace dying cells
<http://www.als.net/articles/articleDetail.asp?articIeld=3750> Brain cells generated in a dish - Discovery pinpoints the true 'stem'
<http://www.als.net/articles/articleDetail.asp?articIeld=3745> Research marks giant step in potential of using stem cells to treat human disorders
<http://www.als.net/articles/articleDetail.asp?articIeld=3743> Drug Trial Thanks to efforts of ALS patients, U.Va. doctor can keep giving drug
<http://www.als.net/articles/articleDetail.asp?articIeld=3739>

Fez1 Is Required for the Birth and Specification of Corticospinal Motor Neurons

SOURCE: Neuron, Volume 47, Issue 6, 15 September 2005, Pages 817-831 (NOTE: This work was done by Dr. Jeffrey Macklis's team at Harvard Medical School. The SPF just awarded Dr. Macklis a 2 year grant to continue his work. It shows that a gene called Fez1 can turn stem cells that are present in the brain into motor neurons. This was considered impossible just 5 years ago.) Data indicate that Fez1 plays a central role in the specification of corticospinal motor neurons

and other subcerebral projection neurons, controlling early decisions regarding lineage-specific differentiation from neural progenitors.

Spastin Mutations in Sporadic Spastic Paraplegia

SOURCE: J Med Genet. Published Online First: 31 July 2005. Background: SPG4 encodes spastin, a member of the AAA protein family, and is the major gene responsible for autosomal dominant spastic

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paraplegia. It accounts for 10-40% of families with pure (or eventually complicated) hereditary spastic paraparesis (HSP). SPG4 mutations in patients without a family history has not been systematically studied. Objective: To assess the frequency of SPG4 mutation in a large series of patients with spastic paraplegia without family histories. Patients and methods: We selected 146 mostly European probands with progressive spastic paraplegia for which major neurological causes were excluded and without familial history of the disease. One hundred and three probands presented pure spastic paraplegia while 43 had additional features. DNA was screened by DHPLC for mutations in the 17 coding exons of the SPG4 gene. Sequence variants were characterized by direct sequencing. A panel of 600 control chromosomes was used to rule out polymorphisms. Results: The overall rate of mutations was 12%. We identified 19

different mutations, 13 of which were novel, in 18 different patients. In one family, where both parents were examined and found to be normal, the mutation was shown to be transmitted by the asymptomatic mother, indicating reduced penetrance. The parents of other patients were not available for analysis but were reported to be normal on history. There was no evidence for de novo mutations. The mutations found in these apparently isolated patients were mostly of the missense type and tended to be associated with a less severe phenotype than in previously described patients with inherited mutations. Conclusions: The unexpected presence of SPG4 gene mutations in patients with sporadic spastic paraplegia argues in favour of gene testing in patients with pure or complicated spastic paraplegia without family histories.

A Case of Presumptive PLS with Upper and Lower Motor Neurone Pathology.

SOURCE: J Clin Neurosci. 2005 Aug;12(6):706-9. Motor Neurone Disease (MND) is one of the commonest neurodegenerative disorders of adulthood. MND characteristically presents with a combination of both upper and lower motor neurone features. Primary Lateral Sclerosis (PLS) is thought to be a variant of MND

presenting with purely upper motor neurone signs. Debate continues over whether PLS constitutes a distinct pathological entity or whether it is part of the spectrum of motor neurone diseases that present as an upper motor neurone-predominant form of MND. We present a case of MND with purely upper motor neurone features and a prominent pain component. A pre-mortem diagnosis of PLS was made, however autopsy findings demonstrated both upper and lower motor neurone involvement. We believe these findings support the view that PLS is not a discrete pathological entity, but that it is a part of the range of motor neurone diseases that present with predominant but not exclusive upper motor neurone involvement. This case also highlights

the feature that pain may be associated with MND even though it is not appreciated to have a sensory pathology. *Ed. Note: MND=ALS*

MRI to Detect Spinal Cord Atrophy in

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mm(2) at C2, $p < 0.02$; 31.75 ± 2.76 mm(2) at T9, $p < 0.001$). These observations indicate that spinal cord atrophy is a common finding in the four genetic types of

HSP Hedera P, Eldevik OP, Maly P, Rainier S, Fink JK. Department of Neurology, The University of Michigan, Rm. 5214 CCGCB, 1500 E. Medical Center Drive, Ann Arbor, MI, 48109-0940, USA. Hereditary spastic paraplegia (HSP) is a genetically heterogeneous group of neurodegenerative disorders characterized by progressive lower extremity weakness and spasticity. HSP pathology involves axonal degeneration that is most pronounced in the terminal segments of the longest descending (pyramidal) and ascending (dorsal columns) tracts. In this study, we compared spinal cord magnetic resonance imaging (MRI) in 13 HSP patients with four different types of autosomal dominant hereditary spastic paraplegia (SPG3A, SPG4, SPG6, and SPG8) with age-matched control subjects. The cross-section area of HSP subjects at cervical level C2 was 59.42 ± 12.57 mm(2) and at thoracic level T9 was 28.58 ± 5.25 mm(2). Both of these values were less than in the healthy controls ($p < 0.001$). The degree of cord atrophy was more prominent in patients with SPG6 and SPG8 who had signs of severe cord atrophy (47.60 ± 6.58 mm(2) at C2, 21.40 ± 2.4 mm(2) at T9) than in subjects with SPG3 and SPG4 (66.0 ± 8.94

HSP. Spinal cord atrophy was more severe in SPG6 and SPG8 HSP subjects than in other types of HSP we studied. This may suggest a different disease mechanism

with more prominent axonal degeneration in these two types of HSP when compared with HSP due to spastin and atlastin mutations.

Human neural stem cells differentiate and promote locomotor recovery in spinal cord- injured mice SOURCE:

Published online before print September 19, 2005 Proc. Natl. Acad. Sci. USA, 10.1073/pnas.0507063102 We report that prospectively isolated, human CNS stem cells grown as neurospheres (hCNS- SCns) survive, migrate, and express differentiation markers for neurons and oligodendrocytes after long-term engraftment in spinal cord-injured NOD-scid mice. hCNS-SCns engraftment was associated with locomotor recovery, an observation that was abolished by selective ablation of engrafted cells by diphtheria toxin. Remyelination by hCNS-SCns was found in both the spinal cord injury NOD-scid model and myelin-deficient shiverer mice. Moreover, electron microscopic evidence consistent

with synapse formation between hCNS-SCns and mouse host neurons was observed. Glial fibrillary acidic protein-positive astrocytic differentiation was rare, and hCNS-SCns did not appear to contribute to the scar. These data suggest that hCNS-SCns may possess therapeutic potential for CNS injury and disease.

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I welcome comments and suggestions.

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North Carolina SP Gathering

Race Green flag at Start of SAWCAR Bettie Jo Wilson wife of SAWCAR
winner Mad Dog Martin down organizer Don Wilson

the home stretch **California TeamWalk**

On TeamWalk organizer Linda Gentner with

Main St. in Pleasanton husband Craig and grandson William **Arkansas TeamWalk Australia**

Gathering

Maureen, Sue Burges, Helen Garner & Sonny Garner

Adrianna & Janet gather getting ready for walk

1st ever SP gathering in Australia **Massachusetts TeamWalk**

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Bottled water for walkers – a must for a hot day

TeamWalk registrar and scooter winner Kris Brocchini

TeamWalk organizer Sue Huskey (right) with her supportive family

Synapse editor Thurza Campbell with her grandchildren

SP President Weber assisted by VP Kathi Geisler wrap up the day